This thesis has been submitted in fulfilment of the requirements for a postgraduate degree (e.g. PhD, MPhil, DClinPsychol) at the University of Edinburgh. Please note the following terms and conditions of use:

This work is protected by copyright and other intellectual property rights, which are retained by the thesis author, unless otherwise stated.
A copy can be downloaded for personal non-commercial research or study, without prior permission or charge.
This thesis cannot be reproduced or quoted extensively from without first obtaining permission in writing from the author.
The content must not be changed in any way or sold commercially in any format or medium without the formal permission of the author.
When referring to this work, full bibliographic details including the author, title, awarding institution and date of the thesis must be given.
Executive Function in Children and Adolescents with Down Syndrome: A Systematic Review and Kuwait-Based Intervention

Ghaleyah Bleah Hamad Alajmi

This Thesis is submitted in accordance with the requirements for the degree of Doctor of Philosophy

Moray House School of Education and Sport
University of Edinburgh
2020
Abstract

Individuals with Down syndrome (DS) have been found to exhibit significant atypicalities in most executive functions (EFs), such as working memory (WM), cognitive flexibility/shifting, inhibition, self-regulation and attention. However, to date there has been limited research into the relative strengths and weaknesses in their EF profiles. Moreover, EFs in DS have received very little attention in the Kuwaiti research context, despite the need for greater support and resources for those with DS in non-Western countries. There is broad agreement in the research literature, however, that WM is one of the more affected EFs in children and adolescents with DS. This thesis therefore set out to investigate EF profiles in, and WM support for, children and adolescents with DS, through two interlinked studies.

Study 1 was a systematic review of EF research in DS. Relevant databases were searched for studies that were published prior to January 2020, involving children and adolescents with DS, comparing their EFs with various comparison populations. Fifty-five studies were included in the final review. The results revealed that, in general, all EFs are atypically developed in individuals with DS. WM appeared to be the most challenged EF, particularly verbal (though not necessarily visuospatial) WM, and emotional control appeared to be the least atypical, relative to controls. Furthermore, there are significantly fewer studies focusing solely on adolescents than on children or mixed-age samples, and even fewer that compare the EF performance of children and adolescents. There are also no studies that draw comparisons between different tools measuring the same EF in the same samples, to ascertain whether different results are gained from different measures. Finally, studies use a variety of comparison approaches, including mental age and chronological age matching, or normative data. They also involve a range of different comparison groups, allowing only tentative conclusions to be drawn from current research.
In study 2 an intervention that aimed to support teachers in Kuwait to use different strategies in the classroom that could encourage the development of WM in children and adolescents with DS, was devised, conducted and evaluated. The aim of this study was to explore the effectiveness of the WM intervention on teachers’ knowledge surrounding WM, their use of WM strategies in the classroom and on cognitive, behavioural, and academic outcomes for children and adolescents with DS. Study 2 involved 31 children and adolescents with DS (aged 7 to 16 years) and 28 literacy and numeracy teachers from specialist units in 4 mainstream primaries schools. The study adopted a quasi-experimental design, involving 2 groups of teachers: an experimental group (enrolled on the WM intervention) and an active control group (enrolled on a positive behaviour support intervention). Outcomes were assessed using a range of measures to test EF, WM, and challenging and social behaviour. Teachers’ knowledge of WM was also evaluated to examine the effectiveness of the intervention, and the experimental group was also observed to explore any difference on their use of WM strategies, pre- and post-intervention. Teachers’ perspectives on the feasibility and acceptability of the intervention process were also gathered, using focus groups. Results showed that the WM in the experimental group significantly improved from pre-intervention to follow-up compared to the control group. In contrast, the control group had significantly lower incidence of challenging behaviour and an increase in prosocial behaviour compared to the experimental group at follow-up. There was no significant difference on academic achievement in literacy and numeracy outcomes between the two groups at follow-up. Moreover, teachers’ knowledge about WM was found to improve significantly in the experimental group compared to the control group, and the use of WM strategies significantly improved from pre- to post-intervention in the experimental group.

Overall, the systematic review in study 1 provides a novel contribution to understanding EFs in the DS population, revealing that WM is the EF that those
with DS struggle with the most. These findings indicate the necessity of effective interventions to address and improve this EF in children and adolescents with DS. Furthermore, the systematic review revealed significant gaps in current understanding and research knowledge of executive functioning more generally in DS: for example, a need for further studies focusing solely on WM functioning in groups of children and adolescents with DS, especially studies that compare a range of measures to test WM functioning. Moreover, the relative strengths of emotional control in those with DS should be further explored. The findings from study 2 established the effectiveness of the WM intervention aimed at teachers - a novel approach in Kuwait - in the short-term, although the long-term efficacy of the intervention is unknown. Therefore, it would be beneficial to conduct a longer-term follow-up assessment in the future. Additionally, it may be helpful in future studies to conduct an intervention to improve WM at home, so there is a consistency of approach between home and school.

The results of these studies therefore have practical implications for teachers, given the finding that when teachers employ strategies to boost WM, the WM functioning of children and adolescents with DS is improved. Helping teachers to become experts in a range of EF support strategies may also lead them to interact differently with children and adolescents with DS, which could improve students’ abilities in other areas (such as other cognitive function or social behaviour). Importantly, the study also altered teachers’ understanding of, and attitudes towards, the learning capabilities of individuals with DS, with important implications for reducing cognitive difficulties in children with DS in Kuwait.
Lay Summary

There is much variability in the skills in individuals with Down syndrome; however, on average, many struggle with particular tasks and skills, such as working memory (the short-term memory we use when we are actively thinking about a task or problem, to help us remember what we are doing), paying attention and dealing with different situations. These skills are called ‘executive functions’ and they help people to complete daily activities and maintain relationships with others. In Kuwait, there is very little research into the executive functions of those with Down syndrome; research studies however have revealed that interventions to help improve the working memory can be beneficial for children and adolescents with Down syndrome. This research project therefore involved two studies: the first aimed to investigate the executive functions of children and adolescents with Down syndrome and, the second aimed to explore whether children’s working memory could be improved.

The first study reviewed the findings of 55 published studies involving children and adolescents with Down syndrome, which specifically investigated their executive functions and how they compared to other groups of individuals, such as typically developing children and adolescents and those with other disorders. The overall finding was that, in general, all executive functions are atypical in those with Down syndrome, but in particular, working memory is significantly affected. Skills requiring verbal abilities were the most atypical or delayed skills in those with Down syndrome, although individuals diagnosed with this disorder often show relative strengths in what is known as emotional control, which means their ability to control, understand and demonstrate emotions are less affected than other executive functions. However, the results of study one remain somewhat inconclusive given the noticeable lack of studies that purely involved adolescents (instead of children or mixed-age groups, samples that appear to be more popular amongst the research literature), as well as the lack
of studies that compare the executive functions of children and adolescents with Down syndrome, which might help us to understand how these skills may develop over time. Furthermore, different studies use different tests and tasks to study the same executive function, which makes their results difficult to compare, and those that do draw comparisons use varying comparison groups, which means only tentative conclusions can be drawn from current research.

The second study was an intervention which supported teachers in Kuwait learn how to use different strategies in the classroom to encourage the development of working memory skills in children and adolescents with Down syndrome. This study sprang from a few studies that indicated that the working memory may indeed be improved through employing specific interventions focused on enhancing either the working memory specifically, or executive functions more generally. This study therefore involved both a group learning working memory skills, as well as another ‘control’ group of teachers of students with Down syndrome, who were instructed in strategies to help them reduce difficult behaviour in the classroom. This was to provide a comparison between the two groups, to establish whether the working memory intervention was effective compared to the difficult behaviour group. There were 31 children and adolescents with Down syndrome (aged 7 to 16 years) and 28 literacy and numeracy teachers from specialist units in 4 mainstream primaries schools. The students’ working memory, executive functions, academic achievement and behaviour were also assessed to see if there was an improvement. The teachers’ knowledge of working memory and use of strategies to improve working memory in the classroom were also assessed, both before and after the intervention.

This study found that students’ working memory skills improved, as did teachers’ knowledge about (and use of) working memory improvement strategies. This suggests that the intervention was effective and may help those with Down
syndrome to achieve their potential in future if it can be rolled out on a wider scale. There are also other implications from the findings of this study; more research and investment are required by governments in this area, to ensure that those with Down syndrome have appropriate, useful materials, toys and books to improve their working memory both in the classroom and at home, and that teachers (and trainee teachers) are taught the correct strategies and methods to use in the classroom when teaching pupils with Down syndrome. This can give this group the best chance of improving their working memory so as to assist them both academically as well as in their daily lives.
Table of Contents

Abstract ............................................................................................................................... i
Lay Summary ....................................................................................................................... iv
List of Tables ....................................................................................................................... xii
List of Figures ..................................................................................................................... xiii
Acknowledgments ........................................................................................................... xiv
List of Abbreviations and Acronyms .............................................................................. xv

Chapter One: Introduction ............................................................................................... 1
  1.1 Overview ................................................................................................................... 1
  1.2 The Importance of Executive Functions .................................................................. 1
  1.3 Executive Functioning in Down Syndrome ............................................................. 2
  1.4 Working Memory Functioning in Typical Development and Down Syndrome ........ 5
  1.6 Down Syndrome in the Kuwaiti Context .................................................................. 11
  1.7 Research Aims ......................................................................................................... 19
  1.8 Summary .................................................................................................................. 20

Chapter Two: Literature Review .................................................................................... 22
  2.1 Overview and Introduction ...................................................................................... 22
  2.2 Down Syndrome ..................................................................................................... 23
    2.2.1 Definitions and Prevalence .............................................................................. 23
    2.2.2 Genetic Causes ................................................................................................. 25
    2.2.3 Risk Factors ..................................................................................................... 25
  2.3 Behavioural Phenotype: Down Syndrome ............................................................... 26
    2.3.1 Medical/Clinical ............................................................................................... 27
    2.3.2 IQ and General Cognitive Functioning ............................................................ 30
    2.3.3 Verbal and Non-Verbal Ability ........................................................................ 33
    2.3.4 Visuo-Spatial Abilities in Down Syndrome ..................................................... 36
    2.3.5 Numerical Skills in Down Syndrome ............................................................... 37
    2.3.6 Memory Abilities in Down Syndrome ............................................................. 39
Chapter Three: Executive Function in Children and Adolescents with Down Syndrome: Systematic Review

3.1 Introduction ............................................................................................................. 94

3.2 Methods ....................................................................................................................... 98

3.3 Results ....................................................................................................................... 106

3.4 Results: Narrative Synthesis .................................................................................... 109
3.4.1 Global Executive Functioning ................................................................. 110
3.4.2 Multiple Executive Functions ................................................................. 120
3.4.3 Working Memory .................................................................................. 127
3.4.4 Attention Function ............................................................................... 134
3.4.5 Cognitive Flexibility............................................................................. 142
3.4.6 Self-Regulation .................................................................................... 145
3.4.7 Inhibition and Impulsivity .................................................................... 147
3.4.8 Planning/Organising ........................................................................... 151

3.5 Discussion ................................................................................................. 153
3.5.1 Possible Explanations for Executive Function Difficulties ......................... 161
3.5.2 Possible Explanations for Working Memory Difficulties ............................. 168
3.5.3 Challenges of Executive Function Research in Down Syndrome Populations 173
3.5.4 Limitations ............................................................................................ 177

3.6 Conclusions, Recommendations and Next Steps ....................................... 179

Chapter Four: Study Two .............................................................................. 218

4.1 Introduction ............................................................................................... 218
4.1.1 Overview ............................................................................................. 218
4.1.2 Executive Function Interventions ............................................................. 220
4.1.2.1 Executive Function Interventions for Typically Developing Children ........ 220
4.1.2.2 Executive Function Interventions for Children with Developmental Difficulties 226
4.1.2.3 Executive Function Interventions for Children with Down Syndrome 228
4.1.3 Working Memory Interventions ............................................................... 233
4.1.3.1 Working Memory Interventions for Typically Developing Children ............ 233
4.1.3.2 Working Memory Interventions for Children with Developmental Disabilities ... 237
4.1.3.3 Working Memory Interventions for Children with Down Syndrome .......... 240
4.1.4 Working Memory Interventions in an Arab Context ................................ 246
4.1.5 Interventions to Support the Reduction of Challenging Behaviour .............. 249
4.1.5.1 Challenging Behaviour Interventions in Typically Developing Children 250
4.1.5.2 Challenging Behaviour Interventions for Children with Cognitive Difficulties 251
4.1.5.3 Interventions to Reduce Challenging Behaviour in Children with Down Syndrome 252
4.1.6 Teachers’ Involvement in Interventions ................................................... 253
4.1.7 Summary and Present Study ................................................................. 255

4.2 Methods ................................................................................................. 259
4.2.1 Introduction to Methods ...................................................................... 259
4.2.2 Research Design .................................................................................. 259
4.2.3 Participants .......................................................................................... 262
4.2.3.1 Teachers ......................................................................................... 262
4.2.3.2 Pupils ............................................................................................ 265
4.2.4 Data Collection .................................................................................... 267
4.2.4.1 Questionnaires .............................................................................. 268
Appendix 2.1: Informed Consent Forms ................................................................. 477
Appendix 2.2: Teacher Knowledge and Demographics Questionnaire ............ 498
Appendix 2.3: Pupil Demographics Questionnaire ........................................... 505
Appendix 2.4: BRIEF-P Subscales Reliability Details ...................................... 506
Appendix 2.5: Literacy and Numeracy Assessments ........................................... 507
Appendix 2.6 Observation Form ........................................................................ 514
Appendix 2.7: Focus Group Prompt Questions .................................................. 515
Appendix 2.8: Intervention Session Resources (PowerPoint) .......................... 518
Appendix 2.9: Ethics Information and Consent Forms ....................................... 553
Appendix 3.1 Change in Teachers’ Knowledge on Working Memory and Challenging Behaviour: Comparison Between Groups .................................................. 554
List of Tables

Table 2.1: Executive Functions in Typical Development and Down Syndrome ..........81
Table 3.1 Keywords of Search Strategy ............................................................... 100
Table 3.2 BRIEF Index Subscales ............................................................... 110
Table 3.3 BRIEF-P Subscales ........................................................................ 111
Table 4.1 Distribution of Teachers Across Schools, Teaching Subject and Experimental or Control Group ............................................................... 265
Table 4.2 Pupil Demographics ...................................................................... 266
Table 4.3 Measures Used in this Study .............................................................. 268
Table 4.4 Verbal and Visuo-Spatial Working Memory Tasks in the Lanfranchi Tasks 284
Table 4.5 Operational Definitions of Child Behaviours and Teacher Strategies ....... 288
Table 4.6 Intervention Session Topics ............................................................... 303
Table 4.7 Mean (Standard Deviation) and Mean Ranks Verbal Working Memory and Visuo-Spatial Working Memory by Group ...................................................... 317
Table 4.8 Working Memory Change Scores and Mean Ranks of Change Scores by Group ........................................................................................................ 318
Table 4.9 Means and Standard Deviations of Parents’ Pre- and Follow-up BRIEF-P Scores (Global and Subscales) by Group ......................................................... 321
Table 4.10 Means and Standard Deviations of Teachers’ Pre- and Follow-up BRIEF-P Scores (Global and Subscales) by Group ......................................................... 323
Table 4.11 Means and Standard Deviations on Pre- and Follow-up Parent and Teacher SDQ Total Difficulties and Prosocial Scales by Group ........................................................... 325
Table 4.12 Literacy and Numeracy Scores Pre- and Post-intervention by Group ....... 327
Table 4.13 Teachers’ Use of Working Memory Strategies .................................... 329
Table 4.14 Means and Standard Deviations of Teachers’ Knowledge Scores on Working Memory and Challenging Behaviour Items by Group ......................... 331
List of Figures

Fig. 2.1 Baddeley’s Working Memory Model (Baddeley, 2003) .......................................................... 60
Fig. 2.2 Morton’s (2004) Developmental Causal Modelling Frame .................................................. 89
Fig. 3.1 PRISMA Flow Diagram Revealing Full-Text Articles Excluded, with Reasons ........ 104
Fig. 4.1 Frequency of Strategy Use Pre- and Post-intervention ...................................................... 330
Acknowledgments

I am grateful to all of those with whom I have had the pleasure to work alongside during my journey towards completing this PhD. My completion of this thesis could not have been accomplished without the assistance of teachers and students who generously participated in this study. A huge thank you to all of them. I am especially thankful and would like to express my gratitude to my supervisors, Dr Katie Cebula, Dr Gale Macleod and Dr Josie Booth, for their guidance and professional supervision during the completion of this thesis. Their fantastic help and feedback throughout this project is highly appreciated.

Words cannot express how grateful I am to my father who passed away during my PhD journey but his support and encouragement continue to stay with me, motivating me to continue with this important work, even when times were difficult. I cannot express how grateful I am also to my mother, who continued my father’s support and took on a significant amount of responsibility in order to help me continue this journey.

Very special thanks go to my five daughters and little son, who were very patient during this journey. A big thank you also to my loving and caring husband your encouragement and support are much appreciated. My heartfelt thanks.

Finally, I want to encourage an individual undertaking a PhD to be positive and continue with the work, even when it seems too difficult; there is something beautiful waiting for you at the end.
List of Abbreviations and Acronyms

ADHD  Attention Deficit Hyperactive Disorder
ASD  Autism Spectrum Disorder
BRIEF-P  Behaviour Rating Inventory of Executive Function (preschool version)
CA  Chronological age
CB  Challenging behaviour
CF  Cognitive Functioning
CG  Control group
CHD  Coronary Heart Disease
DBD  Disruptive Behaviour Disorder
DD  Developmental disorder
DS  Down syndrome
EC  Emotional control
EF  Executive function
EG  Experimental group
EMI  Emergent Metacognition Index
FBA  Functional behaviour assessment
FG  Focus group
FXS  Fragile X syndrome
GEC  Global Executive Composite
HFA  High Functioning Autistic
ID  Intellectual disability
LTM  Long-term memory
MA  Mental age
NSID  Non-specific intellectual disability
PBS  Positive behaviour support
PMA  Performance mental age
RT  Reaction time
SDQ  Strengths and Difficulties Questionnaire
<table>
<thead>
<tr>
<th>Acronym</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>SLI</td>
<td>Specific Language Impairment</td>
</tr>
<tr>
<td>SR</td>
<td>Systematic review</td>
</tr>
<tr>
<td>SS</td>
<td>Social skills</td>
</tr>
<tr>
<td>STM</td>
<td>Short-term memory</td>
</tr>
<tr>
<td>TD</td>
<td>Typically developing</td>
</tr>
<tr>
<td>TLD</td>
<td>Typical Language Development</td>
</tr>
<tr>
<td>VLTM</td>
<td>Verbal long-term memory</td>
</tr>
<tr>
<td>VMA</td>
<td>Verbal mental ability</td>
</tr>
<tr>
<td>VSWM</td>
<td>Visuospatial working memory</td>
</tr>
<tr>
<td>VWM:</td>
<td>Verbal working memory</td>
</tr>
<tr>
<td>WM</td>
<td>Working memory</td>
</tr>
<tr>
<td>WS</td>
<td>Williams syndrome</td>
</tr>
</tbody>
</table>
Chapter One: Introduction

1.1 Overview
Executive functions (EF) are cognitive skills with important implications for an individual’s ability to socialise, learn and undertake daily living tasks. However, in Down syndrome (DS), some EFs appear significantly different from typically developing individuals, whilst others remain relatively unaffected. This thesis therefore explores the EFs of those with DS, as well as how far one EF – working memory (WM) – may be improved in children and adolescents with DS through interventions with teachers. This chapter introduces the research topic and explains the rationale underpinning this study. It explains the gap in knowledge that the current research addressed, by highlighting the importance of executive functioning in daily life, indicating the limited literature that details the EFs that are most and least affected in individuals with DS. It also presents the impact of EF atypicalities on this population, and some of the research indicating that WM difficulties seem to present particular challenges for the DS population. This chapter subsequently presents the rationale for greater research in this area (EF in children and adolescents with DS) through a systematic review, as well as the decision to devise and conduct a novel WM intervention for children and adolescents with DS in the Kuwait region aimed at teachers of DS students. Finally, the Kuwaiti educational system is introduced, before the research questions are presented.

1.2 The Importance of Executive Functions
Cognitive abilities in general, including EFs, are critical components of people’s wellbeing and quality of life, as they are intrinsically linked to different aspects of daily living, including learning, behaviour, adaptation, relationships, social skills and vocational functioning (Najdowski et al., 2014). Goldstein and Naglieri (2014) claim that ‘EF’ has come to be an umbrella term to describe a wide range of hypothesised cognitive processes, including working memory (WM), attention,
inhibition, flexibility, planning, self-monitoring, initiation and self-regulation, all skills that are facilitated and controlled by prefrontal areas of the frontal lobes. Thus, EFs underpin intellectual, emotional, social and organisational elements of one’s life (Sereno & Bolding, 2017).

1.3 Executive Functioning in Down Syndrome

Down syndrome (DS) is the most common chromosomal cause of intellectual disability (ID) in individuals (Parker et al., 2010). Those with DS are predisposed to experiencing various biomedical issues, such as congenital heart defects, sleep disturbances and gastrointestinal difficulties. The daily functioning of individuals with DS in their home, educational and community contexts is heavily impacted by the areas of relative weakness (EF; expressive language; verbal processing) and strength (receptive language; social relatedness; visual processing) (e.g., Fernández-Alcaraz & Carvajal, 2020; Manrique-Niño et al., 2021; Thomas et al., 2020; Tungate & Conners, 2021; Van Herwegen et al., 2019).

The cognitive functioning, particularly the executive functioning, of those with DS, has gained a significant amount of attention within academic literature (e.g., d’Ardhuy et al., 2015; Daunhauer et al., 2014; Esbensen et al., 2019; Lott & Dierssen, 2010). It is also of particular interest to researchers, health care professionals and teachers, given their role in supporting and enhancing the development of children and adolescents with DS. Cognitive function includes EF, and is defined as a multitude of mental skills, such as ‘learning, thinking, reasoning, remembering, problem-solving, decision-making, and attention’ (Fisher et al., 2019, p.18).

Research has revealed that individuals with DS experience difficulties with a variety of cognitive functions (Thompson, 2003), including substantial atypicalities and difficulties in EFs, such as WM (Lanfranchi et al., 2009),
planning (Fidler et al., 2005), shifting (Kopp, Krakow & Johnson, 1983; Rowe, Lavender & Turk, 2006) and inhibition (Edgin et al., 2015; Rowe et al., 2006). Individuals with DS also demonstrate low levels of global EF (Daunhauer et al., 2014) and weaknesses in impulse control and attention (Landry, 2003).

Atypicalities and difficulties in EF in DS are associated in various ways with a range of different aspects of development. This includes motor skills (which are correlated with EF skills (Schott & Holfelder, 2015)), health problems (for instance heart disease, which is also associated with poor EF skills (Alsaied et al., 2016)), sleep difficulties such as obstructive sleep apnoea (Chawla & Heussler, 2017), which is linked with poorer WM, emotional control and shifting (Joyce et al., 2019), linguistic skills (Cuskelly et al., 2016; Polisenka & Kapalkova, 2014; Udhnani et al., 2020) and adaptive behaviour – individuals with DS are more likely to demonstrate poor adaptive behaviour as a consequence of atypicalities and difficulties in EFs (Tomaszewski, Fidler, Talapatra & Riley, 2018; Van Duijn et al., 2010). Those with DS also exhibit poorer skills in temporal orienting of attention (Mento et al., 2019) – the orientation to time cues that can impact behaviour, such as realising that time is running out, so actions must be enacted more quickly – a ‘domain-general cognitive mechanism … that may constrain the build-up of domain-specific skills in DS’ (Mento et al., 2019, p. 81).

Other aspects of development affected by poor EF in those with DS include learning, particularly as a result of a weak WM, including poor literacy and numeracy skills (Banales, Kohnen & McArthur, 2015; Henry, Messer & Nash, 2014). Daunhauer, Will, Schworer and Fidler (2020) for example recently found that compared to typically developing (TD) children, the EF challenges experienced by those with DS led to poor academic outcomes and quantitative skills. Differences in eye movement patterns have been investigated in DS, which have led to insight as to why counting skills are sometimes poorer in
those with DS (e.g., Ranzato, Tolmie & Van Herwegen, 2020), supporting Van Herwegen and Karmiloff-Smith’s (2015) argument that basic abilities may negatively impact numeracy skill development.

Challenging behaviour (CB) has also been linked to difficulties in EF in people with DS (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998; Wilding et al., 2002), as has effective decision-making (Cuskelley, Einam, & Jobling, 2001), and cognitive flexibility (Zelazo, Burack, Benedetto & Frye, 1996), which can mean those with DS struggle with mentalising and emotional intelligence, as they are less able to perceive things from the perspective of others (Theory of Mind) (Tavakoli, Demehri & Azizi, 2019). Thus, as can be seen by the multitude of EF-related developmental issues above, the genetic condition of DS affects individuals on a biological, cognitive and behavioural level (Thomas et al., 2020).

As demonstrated above, research has been able to draw some general conclusions regarding the overall EFs of individuals with DS in various domains (Daunhauer & Fidler, 2012). However, given the complexity of executive functioning (Sereno & Bolding, 2017) and the different assessment measures and methodologies adopted to research EFs in children and adolescents with DS (d’Ardhuy et al., 2015; Amadó et al., 2016; Memisevic & Sinanovic, 2014), greater research in this area is continually required. Systematic reviews are a useful way to explore the breadth and depth of research on this topic. Furthermore, previous research indicates that WM functioning is amongst the most atypical EF in individuals with DS (Lee et al., 2011; Daunhauer et al., 2014; Pritchard et al., 2015), which indicates the usefulness of conducting an intervention to explore how far the WM can be improved in individuals with DS. A systematic review was first therefore adopted, in order to better understand the EF profile in DS and confirm just how affected WM is, relative to other EFs. Gaining a deeper understanding of what is known about how EF works in children and adolescents with DS is imperative if an appropriate, effective
intervention to improve WM in this population is to be designed and implemented with any degree of success.

1.4 Working Memory Functioning in Typical Development and Down Syndrome

Research suggests that WM is an EF that is significantly atypical in children and adolescents with DS (Carretti, Lanfranchi & Mammarella, 2013; de Santana et al., 2014; d’Ardhuy et al., 2015; Godfrey & Lee, 2018; 2020; Manrique-Niño et al., 2020; Visu-Petra et al., 2007). WM is strongly related to one’s learning ability and overall academic achievement, in both typically and atypically developing populations. Alloway and Alloway (2010) for instance aimed to explore how far WM was potentially a proxy for IQ in TD children or if WM made a specific, independent contribution to learning outcomes. In their longitudinal study, they demonstrated that the WM skills of TD children at 5 years old were correlated with their numeracy and literacy skills at age 11. By comparison, IQ predicted a smaller variance in learning outcomes for these participants. They therefore concluded that WM is not interchangeable with IQ; rather, it is a specific cognitive skill that plays a unique role in academic attainment. The study findings also suggest that, for some children, there may be a need for early and ongoing WM interventions. Naturally, this may depend on pedagogical approach; WM may be even more important where rote learning is the norm. This compares to cultures that emphasise understanding in which IQ may be more important, especially in terms of a predictor of academic success. Both IQ and WM are integral to learning in most educational contexts in the 21st century, however.

Other studies have delved into the specific academic skills that may be affected in TD children by WM. For example, Christopher et al. (2012) demonstrated that WM predicted word reading ability and overall reading comprehension – both of which are integral to academic success (Peng et al., 2018). Furthermore,
research has indicated that WM is linked with language development (O’Grady, 2017). For example, Young (2000) researched children with language difficulties (those that have typically developing non-verbal intelligence yet develop language later than TD children), and found substantial WM atypicalities in these children, indicating the integral role WM plays in language development. WM has also been found to underpin numeracy skill development; Geary et al. (2000) found that students that struggled with numeracy, such as the inability to solve simple arithmetic calculations, experienced difficulties with their WM, such as retrieving correct answers from memory.

Measuring WM can prove challenging in individuals with developmental disorders, such as DS. Assessment approaches that effectively capture WM, whilst minimising confounding variables from other domains, has been the goal of various researchers, in TD groups as well as other individuals with developmental delays. In DS, the construct validity of WM assessments may be called into question given the behavioural phenotype associated with DS, which includes, for example, stronger receptive language skills compared to expressive language skills (Daunhauer & Fidler, 2011). As a result, assessments of WM that rely on verbal responses can often produce inaccurate WM data given that performance on a WM task can be hindered/confounded by the expressive mode of response (Fidler, Daunhauer, Will & Schworer, 2018). Visual tasks may also depend on significant verbal skills and processing stemming from the task instructions that can confound any WM competencies. Similarly, WM tasks requiring motor skills (an area that those with DS may struggle with (Palisano et al., 2001)) can also present challenges to valid assessments of WM. Given the integral role of WM within many cognitive processes, other EFs are perceived to impact WM, such as planning, inhibition and cognitive flexibility, which may not be truly dissociable as to be individually tested (Miyake & Friedman, 2012), generating task impurity (Fidler, Daunhauer, Will & Schworer, 2018).
Moreover, whilst models and conceptualisations of EF, including WM, appear to be similar across different cultures, there appear to be differences in the developmental trajectories of different EFs in different cultures. Schirmbeck, Rao and Maehler (2020) conducted a systematic review of 26 studies involving children and adolescents from different countries, revealing gender and cultural differences. Girls tended to perform better on both tasks and parent/teacher rating measures in some cultures (the West and in East Asia) but not others (Tanzania and Iran). Furthermore, whilst at all ages, individuals from East Asia gained consistently higher scores in different EF tests than Western peers, their parents rated them as lower. This indicates that cultural differences play a significant role in both the development and measurement of EFs more broadly, including WM in particular, in TD populations in different countries.

In addition, different measures may be used to assess WM in different settings, which can give rise to psychometric concerns as regards to whether the results from different tools are equivalent. The standardised instruments used in one country to assess WM, for example, may not be appropriate for use in a different country, although arguably there may be greater cultural similarity between some countries (e.g., Arab countries) that may render some WM assessments valid between countries. The first study to validate a WM battery in the Arab culture was conducted by Alansari and Soliman (2012). This study administered a battery of six WM tests to 192 primary-school age children in Kuwait and 192 primary-school age children in Egypt (matched by age and sex). Using multi-group confirmatory factor analysis, the authors found that there were metric, configural, strict and scalar invariances across the two settings, meaning that the same process could be used to measure WM in both Egyptian and Kuwaiti children. This indicates that much of the research surrounding WM in different TD populations – in some Arab cultures at least – may be compared. A more recent review of EF cognitive measures in various Arab countries (Fasfous et
al., 2017), however, has indicated that half of the publications reviewed (384 studies) from Arab countries ‘did not employ cognitive tests that were developed, translated, adapted, or standardized according to international guidelines of psychological measurement’ (p. 158). This included EF and specific WM assessment measures. It is clear, therefore, that a validated, standardised WM assessment in the Arab world – and cross-culturally more generally – is required, that can enable cross-cultural comparisons to be more accurately gleaned.

Given the critical role that WM in particular plays in learning and overall academic outcomes, and the plethora of research that indicates that those with DS have significant WM atypicalities and difficulties (e.g., Doerr, Carretti & Lanfranchi, 2019), this research firstly undertook a systematic review to establish the EF profile of individuals with DS. Subsequently, this research devised and conducted an intervention containing strategies to effectively improve WM in children with DS. This kind of intervention is likely to be more effective if it is delivered by teachers as this can help to guarantee sustainability (long-term outcomes). For example, research suggests that expert-led interventions (experts in this case would be teachers, who are well acquainted with the learning and behavioural needs/skills of the children in their class) are often more sustainable than one-off interventions in improving both the development of children with additional support needs (Daniel & Lemons, 2018) and the pedagogical approaches of teachers (Lin-Siegler, Dweck & Cohen, 2016). This is especially true if teachers are fully on board with, and invested in, the intervention; thus, their opinions on the subject are important (Florian & Camedda, 2020).

Designing such a WM intervention for children with DS in Kuwait addressed a gap in this area, given that (to the researcher’s knowledge), no such WM intervention had been conducted before in Kuwait. The Kuwaiti educational
system is an ideal context in which to trial and assess the effectiveness of a WM intervention with children with DS, as the Kuwaiti government facilitates and champions educational research procedures, acknowledging the proposed benefits for participants and the wider educational community in Kuwait. The significant level of support available for researchers in Kuwait also facilitated the implementation of an intervention for a control group, to ensure that their time was not wasted and to make the research more robust, by focusing on improving another common problem in children with DS: challenging behaviour (CB). This is one of the most discussed and concerning topics for teachers of students with DS, in both the literature (e.g., Broomhead, 2013; Feeley & Jones, 2006; 2008) and amongst the teacher participants for the present thesis. An intervention focusing on CB, delivered to teachers, containing strategies to reduce this problematic phenotype in children and adolescents with DS, could significantly benefit the students’ learning journey and had also never been conducted in Kuwait before. This means that both WM and CB interventions in children with DS were novel interventions in the Kuwaiti educational context, although the primary focus in the current study was the effectiveness of a WM intervention for this cohort.

1.5 Education in Kuwait

Education in Kuwait is free for native Kuwaitis from kindergarten to university, with girls and boys taught separately (except in the kindergarten), starting at age 6, and progressing onto secondary school at 11 years old, and high school at 15-years-old. There is an obligation for parents to enrol their children in school, and not doing so is punished; children leave high school at 18 years old. The government provides financial support for every family, for example a monthly salary for every person (adult or child) in the family (except those who do not enrol their children in school) according to Kuwait’s Constitution of 1962 (General Secretariat of National Assembly Information Department, 2020). Also, when in university, each student is given a monthly stipend by the government.
to cover the cost of education (General Secretariat of National Assembly Information Department, 2020). This indicates the high value that Kuwait places on the education of its citizens.

Similarly to most countries, there are public and private mainstream schools in Kuwait (special education is discussed in section 1.6). All public mainstream schools for TD children contain play areas, a cafeteria, music classes and a theatre, administration blocks, a clinic, school nurse, social worker and psychologist, a gym and teachers’ offices, in addition to classrooms. Public schools in Kuwait are large, catering for approximately 600-800 students (at primary, secondary and higher education levels) with the same format (organisation and class size) and examination/curriculum requirements. The National curriculum for elementary and secondary stages of education include Arabic, English, Religious Education, Mathematics, Science and Social Science (Kuwait Ministry of Education, 2015). Other optional subjects include art and music, but lesson frequency occurs once a week, not every day (such as the aforementioned core subjects). At the high school stage, students choose to enrol in either the scientific or humanities subjects. For the scientific subset of subjects, students learn the same subjects as in both elementary and secondary school, yet have a greater focus on scientific subjects including Mathematics, Chemistry, Physics, Biology and Geology. In the 'humanities' sub-set of subjects, students place more focus on subjects including the Social Sciences (Psychology, Geography) and humanities (History, Philosophy, French), and do not study Science or Mathematics (Kuwait Ministry of Education, 2015). In private schools, which numbered 551 in 2018 (40% of the total schools in Kuwait), with 266621 students (Central Bureau of Statistics in Kuwait (cited in Kuwait Times, 2021)), students learn the same National Curriculum as in Kuwait public schools, yet may generally follow a different educational system, such as British or American. 32% of these schools are Arab schools (not necessarily for native Kuwaitis), whereas the remaining 68% of private schools are foreign
Schools for ex-patriots (British, American, bilingual and so on) (Kuwait Times, 2021).

School is free for both sexes, although sex segregation occurs from elementary school to high school, due to conservative social and cultural norms (AlMatrouk, 2016). Boys and girls attend mixed classes in kindergarten and in some universities, but not elementary, secondary or high school (in public schools). Sex segregation in Kuwait was formalised in law in 1996, when what was common practice was discussed and legalised by the representatives of the people in the Kuwaiti National Assembly (Alqabas, 2018). Kuwait is a conservative society, yet is beginning to adapt to the changing times; the Kuwait government permits males and females to mix in some public universities. There are currently two kinds of public university: one allows mixing in some specialities whilst the other does not permit mixed classes. Mixing in some private schools and private universities is acceptable (Kuwait Ministry of Education, 2015). Thus, there is flexibility in that parents are able to choose the kind of institution they wish to send their child to. Moreover, the attitudes towards sex segregation in Kuwait are beginning to change, with many pointing out the need for males and females to interact given the need to do so in the workplace (Alqabas, 2018).

1.6 Down Syndrome in the Kuwaiti Context

The conceptualisation of disability in Arab and Islamic countries tends to be positive, although it is difficult to untangle cultural and religious views of disability. In the Qur’an, the Islamic holy book, individuals with disabilities are termed ‘disadvantaged people’ (Al-Aoufi, Al-Zyoud & Shahminan, 2012), and the book illustrates the need for society to care for and improve the wellbeing of such individuals. The social model of disability is generally championed in the Qur’an, where barriers are perceived to be produced by society that prevent ‘disadvantaged people’ from participating fully in social activities (Al-Aoufi, Al-
Zyoud & Shahminan, 2012). Various Arab countries have different conceptualisations of disability, depending on the Islamic or cultural traditions. For example, individuals with a disability are protected by Islamic law in Saudi Arabia, whereas in Jordan, the Law on people with disabilities (issued in 1993 and reviewed in 2007) urges equality for individuals with disabilities, despite the absence of Islamic values in the law (Al-Aoufi et al., 2012). In the Kingdom of Saudi Arabia, basic education for those with disabilities did not exist until 1958, given the view that individuals with disabilities were unproductive in society (Aldabas, 2015). Yet over subsequent decades, provision expanded to include those with visual impairment, low IQ and hearing difficulties (Afeafe, 2000). This highlights the significant differences in provision, understanding and conceptualisation of those with disabilities and Special Educational Needs (SEN) in different Arab nations.

As of 2018 (Public Authority for Civil Information, 2018), the population of Kuwait was 4,588,000 (of which 1,385,000 are ethnic Kuwaitis). There were approximately 52,820 individuals with a disability in Kuwait in 2015 (Kuwait National Assembly, 2015). In 2018, there were a total of 3,200 individuals with DS (Kuwaiti Down Syndrome Association, 2018), with a prevalence rate of 1-3 in 1000. Thus, perhaps due to various social and legal factors, the rate of DS in Kuwait is greater than some other countries, including the UK, where the prevalence rate is 1 in every 1,000 (Down Syndrome Association, 2020). In fact, the incidence of DS is higher in the Middle East than in many other countries (Centre of Arabic Genomic Studies (CAGS)), particularly in Kuwait (CAGS, 2013). This is likely to be because although screening exists for DS in pregnancy, according to articles 174-177, abortion is illegal in Kuwait except in exceptional circumstances, such as if there is serious danger to the mother’s health as a result of the pregnancy or due to significant foetal impairment (rendering its chances of survival outside of the womb very low) (Kuwaiti National Assembly, 2021).
Perhaps given that the rate of DS is slightly higher in Kuwait compared to other Arab countries, there is less stigma and shame associated with disability and greater government support for individuals with DS in Kuwait compared to other countries (Raman et al., 2010). There are also significant legal protections for those with disabilities, such as DS, in Kuwait. Those with disabilities are protected by law number 8 (2010) in Kuwaiti legislation (Public Authority for the Disabled in Kuwait, 2010). This comprises 72 articles that deal with a range of issues and concerns regarding disability. Some of these articles directly related to this study include Article 9 (which stipulates that the government is committed to the provision of appropriate facilities and teaching to enhance the learning and education for those with disability) and Article 10 (which indicates the government’s responsibility to ensure that those with disabilities are able to have fulfilling lives in society and the workplace) (Public Authority for the Disabled in Kuwait, 2010).

However, there is a paucity of clinical practice guidelines in the Gulf generally and in Kuwait in particular surrounding DS. Clinical practice guidelines are defined by the United States Institute of Medicine as: ‘systematically developed statements that include recommendations, strategies, or information that assists physicians and/or other health care practitioners and patients make decisions about appropriate health care for specific clinical circumstances’ (cited in Rosenfeld & Shiffman, 2009, p. 30). The diagnosis, care and management of individuals with DS follows specific rules set out by the government to support these individuals, akin to the guidelines in other countries (e.g., Ivan & Cromwell, 2014). However, these rules do not constitute clinical practice guidelines the way that the National Institute of Clinical Excellence (NICE) have set out guidelines for individuals with conditions such as DS, as they are not as extensive or as in-depth as the NICE guidelines. Rather, these ‘rules’ in Kuwait are a series of (in some cases) statements released by the Kuwaiti government
intended to advise different ministerial departments as to how they should cater for those with special needs, such as DS. These rules are generated in discussion with the head of each Ministry and the Kuwaiti Assembly and are adapted for each Ministry, similarly to a streamlined code of conduct (outlining rights and expectations) instead of specific clinical guidelines. Indeed, a systematic review of the literature regarding the development, implementation and evaluation of general clinical practice guidelines found that in Kuwait, there were only seven studies that discussed clinical practice guidelines (Koornneef et al., 2015). These tended to focus on lifestyle-associated diseases in the Gulf region, such as diabetes, and no study looked at clinical practice guidelines with respect to DS. This highlights the lack of specific, detailed clinical guidelines for the management and provision of those with DS (and other disabilities). Yet, as previously mentioned, there are specific rules to support those with special needs in every aspect of life, including education; all Kuwaiti ministries are expected to follow and adapt the rules that apply to their department regarding provision for those with special needs.

In terms of education, there has been an increasing move in the Middle East towards adopting the ideas and practices surrounding inclusive education, akin to the discourse in many countries in the West (such as the UK). This is, in part, an attempt to reflect global human rights initiatives (Almuhareb, 2007). This includes inclusivity in schools and in society more generally, including individuals with disabilities. Various Arab countries, such as Saudi Arabia and Kuwait, have passed laws guaranteeing free housing, medical care and education for those with disabilities. For instance, special education services began in Saudi Arabia in 1958 for males who were visually impaired, which was later expanded in 1960 to a special facility, the Al-Noor Institute of Riyadh the basis of public special education which was supported by the Ministry of Education (Aldabas, 2015). Other Arab countries, such as the United Arab Emirates (UAE), have also generated provision for special education; in 1979
the UAE began to provide special education programmes for those with intellectual disabilities within community-based educational centres. Since the introduction of Federal Law no. 29 in 2006 in the UAE, however, entitled *Regarding the Rights of Persons with Special Needs*, all students with disabilities are granted equal educational opportunities and access to general (mainstream) education, when this is perceived to be the most appropriate place for them, as decided by the Ministry of Education (Ministry of Education in the UAE, 2020). Various studies exploring teacher attitudes towards those with disabilities and Special Educational Needs (SEN) in Arab countries, however, have revealed that this can have a significant impact on the child’s adjustment in mainstream education. Takriti, Atkinson and Elhoweris (2019) for example recently found that teachers of children with DS in the UAE and UK developed different expectations of the child’s adjustment and outcomes, depending on their location (UK or UAE); those in the UK had greater optimism regarding the level of achievement they perceived the child to be capable of than teachers in the UAE, which impacted the child in different ways. This highlights the critical nature of gaining support from (and developing high expectations of) teachers of children with SEN in the Arab context.

In Kuwait, there is significant provision of special schools and special education programmes specifically aimed at meeting the education and social support needs of children with disabilities (Almuhareb, 2007). This inclusive policy setting out to include individuals with special needs in all areas of life, including education extends to services within the community for those with disabilities, including DS. Kuwait is arguably more progressive in this respect compared to some Arab countries, although there are plentiful areas for improvement (Almoosa, Storey & Keller, 2012; Alshemari, 2016; Elbeheri et al., 2020; Middle East Health, 2011). Inclusive practice and awareness of those with DS, for instance, extends beyond schools into society, with events being run to boost community awareness, acceptance and understanding of DS (Ahmad, 2015).
Any individual that is not able to work, for instance, is able to claim the same ‘salary’ (benefits package) from the Kuwaiti government, including those with DS; however, it is questionable whether those with DS have the same work opportunities as TD individuals and those without disabilities. Yet individuals with DS are able to access financial support from the government in Kuwait to help them achieve a considerable quality of life. The state also supports the creation and maintenance of various associations that benefit those with disabilities, such as the Kuwaiti Society for People with Disabilities (established 1971), the Kuwaiti Society for the Blind (established 1972), the Kuwaiti Autism Society (established 1994), the Kuwaiti Society for Parents of People with Disabilities (established in 1998) and Kuwaiti Society for Down syndrome (established in 1995) (Kuwait Ministry of Education, 2015).

In terms of school provision for those with SEN, according to the Kuwait Ministry of Education (2013), in Kuwait there are six governorates, and each governorate contains 4 mainstream schools that also have units attached to cater for those with special educational needs: two for those with Down Syndrome (one for girls and one for boys) and two for people with learning difficulties (one for girls and one for boys) that have an IQ of between 70-84 (Alenezi, 2016). Those with other types of SEN and other disabilities are taught in separate special schools; thus, the only individuals included in mainstream schools (albeit in special wings/suites) are those with DS and moderately low IQ. Facilities and extra provision for these children include physical and speaking therapies, for instance, as well as easier access (in terms of toilets, halls, classrooms, in and out of the building and so on). The inclusion of those with DS in mainstream schools only occurs in the elementary stage; after this, those with DS attend specific vocational training depending on their sex. For those with other learning difficulties/disabilities and SEN (low IQ), inclusion continues to secondary stage. For students with physical disabilities and blind/visually impaired students are free to attend mainstream secondary school in the same class as their TD peers
(they attend a special primary school), and are able to continue to university, similar to TD individuals, if they wish.

Parents of children with disabilities may also opt to send their child to a private, fee-paying school if they wish: they are able to ask the Committee for People with Special Needs in Kuwait (a national committee that can inform parental decisions and fund fee-paying school places for those with special needs at a range of specific private schools for those with SEN) to cover the costs of this. This may be preferable if the school is closer to them in location, although in general, government-funded public schools are considered to be a more attractive option due to their superior facilities.

The Ministerial Decree No. 350/2007, based on the Article 40 of the Constitution of the State of Kuwait (which indicates that education for all Kuwaitis is guaranteed by the state) and Decree No. 31/5/2000, generated regulations for special classes for students with DS in both kindergarten and the primary stages of education in Kuwait. These regulations stipulated that: 1) isolating students with DS by labelling their wings/suites/classrooms as ‘Down syndrome’ should be avoided (rather they should use a neutral word such as ‘the happiness wing’ etc); 2) there was to be a maximum of 10 children with DS per class; 3) three specialists (a psychologist, social worker and speech and language therapist) were to cooperate with school administrations and teachers to help deliver lessons, as well as 4) two female service implementers to assist with hygiene and self-care. To be accepted into a special class in a DS wing in a mainstream school, the child must be a Kuwaiti citizen, over four and a half years old, and have DS only (no co-morbid disabilities or health issues). Moreover, the child should have an IQ of 52 or above and be interviewed for acceptance; otherwise, they must attend a special school for students with DS.
There is a specific curriculum that those in DS units in mainstream schools follow. In elementary school, during stage one to stage five, there are four topics covered for those with DS in mainstream schools: numeracy, literacy, science, and self-care. After elementary school, students with DS undertake vocational training in a special institute run by the Kuwait Ministry of Education, with specific skills for girls (for example, painting on textiles, printing pictures in books) and boys (for example, woodwork), which are highly supervised, as well as general skills and practical training to best prepare them for adulthood and the world of work. There are four institutes in total (two for boys, two for girls), spread across two different cities in Kuwait (Mubarak Alkaabeer and Hawally).

Special schools for those with DS that either have a lower IQ or comorbid syndromes/disorders (compared to those with DS in specific wings in mainstream schools in Kuwait) focus on life and social skills, such as self-care, personal hygiene, and social behaviours, according a specific eight topics, such as food (healthy eating), or the supermarket (shopping skills), presented every 2 weeks. This is similar to the TD kindergarten curricula (Kuwait ministry of education, 2015).

However, arguably, the inclusion of those with special educational needs is limited in Kuwait, compared to some other countries; those with physical disabilities may experience greater inclusion in mainstream classes, following the same curriculum as their TD peers. Children with cognitive and EF disabilities, including those with DS, who do not have multiple difficulties (e.g., DS without co-occurring conditions) are able to attend mainstream schools (instead of special schools), yet are placed in separate classrooms or wings, following a special curriculum (Public Authority for the Disabled in Kuwait, 2010), rendering inclusion as locational only (Hodkinson, 2005).
Moreover, various studies have emerged over the last decade detailing the exclusionary and disorganised practices in special needs education in Kuwait (for example, Al-Hilawani, 2011). For example, a study investigating perspectives of 34 mothers on the inclusion of young Autistic children in Kuwait revealed that they felt the current inclusive mainstream educational model could not meet the particular needs of their child (Mutabbakani & Callinan, 2020). Whilst this study focused on children with ASD, it may be that this experience extends to parents of children with DS. Attitudes of teachers towards those with disabilities plays a significant role in the way they are educated within a society (Al-Abdulghafour, 1999). A similar finding was uncovered by Alenezi (2016), who revealed that teachers’ and mothers’ perspectives of inclusion are conceptualised by the medical model of disability (rather than the social model) and that, furthermore, the term ‘inclusion’ was merely used as the antithesis to ‘exclusion’. The participants in Alenezi’s (2016) qualitative study indicate that integration (limited access and less participation of those with special needs and disabilities in mainstream education) is more prevalent in Kuwait, where cultural understandings of disability appeared to impact policymakers in each region, as well as the (narrow) extent to which inclusion is actually practiced in this context. Thus, Alenezi (2016) concluded that there are significant obstacles to true inclusion for those with special educational needs in Kuwait, which may impact on the educational experience of those with SEN, such as individuals with DS.

1.7 Research Aims
This thesis comprises two studies. The rationale for these studies emerged from: the prevalence of DS in Kuwait; the current research indicating that WM is one of the most affected EFs in those with DS; the lack of literature investigating the broad EF profile in children and adolescents with DS; and, crucially, the lack of an existing effective, teacher-involved intervention to improve WM in those with DS in the Kuwaiti context.
These studies were as follows:

**Study One:**
The aim of study one was to explore the elements of EF that are atypical, and, critically, the most and least challenged, in children and adolescents with DS.

This was achieved by conducting a systematic review of current literature in this area.

**Study Two:**
The aim in the second study to develop an effective WM intervention for children and adolescents with DS and investigate how far this changed:

a) working memory outcomes;

b) general executive function scores;

c) behavioural outcomes;

d) literacy and numeracy scores on school assessments;

for pupils with Down syndrome in inclusive education in Kuwait.

It also aimed to assess how far the intervention changed:

1. teachers’ use of WM strategies from pre- to post-intervention;

2. teachers’ knowledge of WM and CB change from pre- to post-intervention;

It also aimed to:

3. explore teachers’ viewpoints about the WM intervention.

1.8 Summary

This chapter has explained the importance of EF in general and the impact of its weaknesses on DS in various different domains/areas. It has presented some of the key features regarding special education and DS in Kuwait, highlighting the
benefits of conducting educational research in this context. The need for a systematic review to explore the profile of EF in DS, and the decision to conduct an intervention specifically aimed at improving WM in DS, given the integral role it plays in learning, has also been presented. Furthermore, the importance of getting teachers as experts on board with an educational intervention has been expounded as a means of promoting the sustainability of any intervention.

The structure for this thesis forthwith is as follows: chapter two provides a general literature review to delve into DS in greater detail, including definitions, behavioural phenotype and co-occurring conditions, as well as exploring definitions, models, tools for measuring EF and developmental trajectories of EF in TD and Populations with DS. Chapter three then presents study one (the systematic review of EF in children and adolescents with DS), including the rationale, methodology and narrative review and discussion of the findings; chapter four then introduces study two, describing current WM interventions, the research design of the present study and presents, analyses and discusses the results from study two. Chapter five then discusses and concludes the combined, generalised findings, with implications and recommendations for future research, policy and teaching practice made.
Chapter Two: Literature Review

2.1 Overview and Introduction
This chapter sets the context for this thesis by presenting different aspects of DS, including definitions, prevalence, genetic basis, characteristics and common physical and health issues. Moreover, the behavioural phenotype commonly found in those with DS is explored, with a focus on medical/clinical features, IQ and general cognitive functioning, verbal and non-verbal abilities, visuo-spatial, numerical skills, memory abilities, language profile, social skills, challenging behaviours and daily living skills. Co-occurring conditions such as Autism Spectrum Disorder (ASD) and Attention Deficit Hyperactivity Disorder (ADHD) are also briefly discussed, to ascertain the prevalence of DS co-occurring with these disorders. Furthermore, there is review of the literature relating to the definitions and models of executive functioning (EF), developmental trajectories of EF, as well as how executive functions (EFs) are commonly assessed, before a discussion of EFs in DS and the behavioural phenotypes of DS ensues. The final section discusses the conceptual model of EF within the DS population that was adopted throughout this thesis, taken from Morton (2004), as well as assessing EF development in both TD populations and those with DS.

In reviewing the relevant literature, this chapter includes both older, classic research into cognitive functioning, EF and DS, as well as more recent work, in an attempt to harness both current understanding and the origins of this understanding. Thus, the aim of this chapter is to consider a range of sources that enable a deeper exploration of how DS affects children, which identified the need for a systematic review (SR), reported in chapter three, specifically regarding EFs in children and adolescents with DS.
2.2 Down Syndrome

2.2.1 Definitions and Prevalence

John Langdon Down first formally identified Down syndrome (DS) in 1866 (Dunn, 1991). Ekvall and Ekvall (2017) and Kazemi, Salehi and Kheirollahi (2016), describe DS as arising from atypical cell division that leads to an extra copy of chromosome 21, affecting one in every 400-1500 children, born in populations that vary in terms of maternal age and prenatal screening schedules.

There is conflicting evidence about the prevalence of DS within the general population at any given time. Examination of DS prevalence data collected between 1979 and 2003 showed that the pooled prevalence of this condition at birth in ten US regions, for example, rose from 9.0 to 11.8 in every 10,000 births, a 0.9% average increase (Shin et al., 2009). Shin et al. (2009) and Weijerman and De Winter (2010) claim that apparent increases in the prevalence of DS can partly be explained by a significant increase in the survival rate of those with this condition, as over 90% of children with this condition survive beyond 5 years, as well as the older age of mothers than historically might have been the case – a significant risk factor for DS (Coppedè, 2016). However, findings from Sherman et al. (2007), John (2012), Shin et al. (2009) and Kurtovic-Kozaric et al. (2016) show that the actual number of individuals born with DS (live births) across the globe (the incidence) has decreased, undoubtedly due to advanced screening programmes in antenatal care (Cuckle & Maymon, 2016). It is possible that the increase in the US states outlined by Shin et al. (2009) is due to various factors, such as varied anti-abortion laws in place, religious beliefs, or more positive ideas surrounding DS and disability more generally. This indicates that both incidence and prevalence rates are significantly affected by cultural attitudes, laws and medical/antenatal practices in different countries and areas.
The incidence of DS (live births) is generally lower in developed countries for example, compared to developing countries, as a result of advanced antenatal screening and greater freedom to terminate pregnancies in economically stable countries (Al-Biltagi, 2015). According to Al-Biltagi (2015), the potential risk factors for DS in different communities also influences variation in incidence and prevalence of DS in different countries. For instance, there are factors that also contribute to increased numbers of individuals with DS in developed countries compared to developing countries, such as advanced maternal age at conception and the increased survival rate of children with this syndrome, as better health care means that infants with DS tend to live longer in developed countries (Santoro, 2016). In the UK, for example, there is an increasing prevalence of individuals with DS in the 40-55-year-old age category, revealing both a rise in life expectancy and the limited availability of selective abortion at the time those individuals were born.

Various researchers note that the incidence and prevalence of DS is extremely high in the Arab world (e.g., Al-Biltagi, 2015; Alsubie & Rosen, 2018; Alkhateeb, Hadidi & Alkhateeb, 2016; Jastaniah et al., 2017). Al-Biltagi (2015) speculates that this may be caused by various factors, including social and economic conditions, social practices (such as consanguineous marriage), advanced maternal age (in Arab countries it is common for women to continue having children as long as fertility lasts) and legal/religious restrictions (abortion is illegal in the United Arab Emirates, for example). Weijerman and De Winter (2010) support this finding by indicating that the prevalence of DS is highest in countries that treat abortion as an illegal practice and lowest in countries that allow abortion, such as the UK. For example, the estimated prevalence of DS in England and Wales is 1.08 in 1000, whereas the estimated prevalence in the UAE is 3.12 in 1000 (Al-Biltagi, 2015).
In Kuwait, the last census in 2018 (Public Authority for Civil Information, 2018) reported that the population of the country is 4,588,000, although of these only 1,385,000 are ethnic Kuwaitis. There is a prevalence of between 1-3 in 1000 for DS in ethnic Kuwaitis, meaning that the prevalence of DS in Kuwait is possibly greater than in the UK; this could be due to a range of social and legal practices as outlined in the previous paragraph.

2.2.2 Genetic Causes
DS is caused, in 95% of cases, by an additional copy of chromosome 21; it is occasionally caused by other chromosomal errors, such as translocation (4%) and mosaicism (1%) (Asim et al., 2015; Sotonica et al., 2016; Sherman et al., 2007). According to Asim et al. (2015), trisomy 21 arises from the unsuccessful separation of chromosome 21 during the development of the egg or the sperm. Although it is the smallest human autosome, chromosome 21 has 48 million nucleotides that influence approximately 1.5% of the human genome (Kazemi et al., 2016). Recent studies also show that a specific section of chromosome 21 contributes to the occurrence of many aspects of the DS phenotype, such as craniofacial differences, intellectual disability and other features (Kazemi et al., 2016). Mosaicism, a rarer form of DS (2-3% of all cases) (Miller & Therman, 2001), is where some cells in the body have two copies of chromosome 21, and some have three, developing from a single zygote (Nussbam, McInnes & Willard, 2001), often with fewer/less severe symptoms of DS.

2.2.3 Risk Factors
The main risk factor for the incidence of DS could be advanced maternal age, as argued in early research such as Van der Scheer (1927) (cited in Glidden, 2008) and Thurston and Jenkins (1931), and in more recent studies including those by Kurtovic-Kozaric et al. (2016). According to Al-Biltagi (2015), the risk of mothers aged below 25 years having children with DS is 1 in 1,400, whereas in those aged 42 years and 49 years, the risk is 1 in 60 in and 1 in 12 respectively.
However, Perkins (2017) points out that 80% of children with DS are born to mothers aged below 35 years, due to greater fertility rate. According to Al-Biltagi (2015), the risk of having children with DS increases with advanced maternal age as eggs are more susceptible to ageing effects than sperm, leading to an increased formation of aneuploid oocytes, cells with either too few or too many chromosomes and meiotic errors.

In addition to maternal age, several other factors have been identified as increasing the risk of DS. According to Kling (2008), for example, short spacing of pregnancies contributes to the occurrence of birth defects. Hunter et al. (2013) indicate that disadvantaged maternal social and economic status can result in poorer nutrition and environmental toxin exposures, which augment the chances of congenital malformations. Environmental factors, such as smoking and drinking alcohol during pregnancy, maternal irradiation, fertility drugs, oral contraceptives and spermicides have also been implicated as contributing factors for having children with DS (Coppedè, 2016; Corona-Rivera et al., 2019; Sherman et al., 2007).

2.3 Behavioural Phenotype: Down Syndrome

The behavioural phenotype approach asserts that specific genetic syndromes are linked with greater prevalence of particular patterns of cognitive and behavioural development (Dykens, 1995; O'Brien, 2000). The literature highlights that there are specific clusters of behaviours and characteristics that individuals with DS have been associated with, creating a generalised ‘behavioural phenotype’ (e.g., Feeley & Jones, 2006, p. 65). Fidler et al. (2019) describe this as a pattern of challenges and relative strengths in functioning in different domains of development, including cognitive, social and linguistic development. Whilst the behavioural phenotype approach does allow for individual variability across DS, it could nonetheless be argued that a behavioural phenotype approach perpetuates a stereotype of DS. This may
perhaps affect integrations into the wider community (Wishart & Johnston, 1990) as a result of their stereotyped cognitive, social and behavioural profile (Fidler, Hodapp & Dykens, 2002; Hodapp, 1997; Hodapp & Dykens, 2004).

The following section explores some of the research surrounding the medical and clinical characteristics of those with DS, their typical cognitive functioning, as well as social development and challenging behaviour, to establish any patterns, trends and variability. Whilst this thesis is predominantly focused on cognition, it is acknowledged that individuals with DS may also be more likely to exhibit challenging behaviour and other characteristics (Feeley & Jones, 2006; 2008).

2.3.1 Medical/Clinical

Diagnosis for DS rests on a range of factors, including physical and cognitive features and tests. Children with DS generally possess distinctive physical features. There are various distinctive facial features that are characteristic of individuals with DS, including slanting eyes, a small chin, round face, Brushfield spots (white or yellow-coloured spots on the iris surface), atypical outer ears and a flattened nose (Cornejo et al., 2017). There are also dysmorphic features of the head, neck and airways (Asim et al., 2015). In terms of health issues, endocrinologic and hematologic problems (Weijerman & De Winter, 2010), various cancers (Bhella et al., 2015), gastrointestinal anomalies, congenital heart defects, and early onset dementia (Ekvall & Ekvall, 2017) can all occur, with research indicating that some of these conditions (for instance, dementia) may correlate with weak EF (Fonseca et al., 2019). People with DS also demonstrate weak neuromuscular tone, visual, hearing and sleeping problems (Joyce et al., 2019) (discussed below).

Individuals with DS are also susceptible to many health problems, which has implications for their medical and social care. For example, research has
established that individuals with DS are at increased risk (40-50%) of developing congenital heart disease (Elmagrpy et al., 2011). Of particular relevance is the finding that heart disease (including congenital heart disease) in DS is associated with poor EF. Studies have found that children with DS who had corrective heart surgery in the first year of life due to coronary heart disease (CHD) demonstrated poorer neurodevelopmental outcomes, including language and EF skills, at preschool age compared to those that did not have CHD (Alsaied et al., 2016). However, it must be noted that there was no difference in EF at school-age between those with and those without CHD in this study, indicating that the impact on EF was perhaps overcome as the child with CHD matured, especially given that they underwent corrective heart surgery. Yet the link between CHD and poorer EF is also seen in the TD population of children and adolescents (Cassidy et al., 2015), such as inhibition (Gaynor et al., 2014), planning (Bellinger et al., 2011), cognitive flexibility, working memory (Calderon et al., 2012), attention (Hövels-Gürich et al., 2007) and problem-solving (Bellinger et al., 2003).

Further health issues experienced by people with DS include delays and weaknesses in a range of gross and fine motor skills. This includes hypertonia and hyperreflexia (atypically flexible joints (Ostermaier, 2019)), deficits in motor planning, coordination delays (Weiss et al., 2010), atypical mobility (Dedlow et al., 2013) and difficulty in standing, walking and moving limbs due to a lack of balance (Davis, 2008). Sunderman (2016) found that DS affects not only the gross motor but also fine motor skills of toddlers, leading to motor dysfunction and oculomotor disturbances. Kim et al. (2017) revealed that infants with DS take twice the time taken by TD children to achieve specific motor milestones. Causes of these motor development delays have been attributed to the limited size of the cerebrum, various pathophysiological processes and brain maturation complications (Malak et al., 2015). Thus, cognitive delay along with delayed motor skills can affect the ability of children with DS to interact with the
immediate environment, manipulate objects and explore the space around them, often affecting daily activities (El-Hady et al., 2018). It is worth noting that research has found that levels of EF correlate with motor skills for people with DS: the better an individual’s EF skills, the better their motor skills tend to be (and vice versa) (Schott & Hofelder, 2015).

Over 50% of children with DS present with visual challenges (Weijerman & De Winter, 2010), which can exacerbate existing developmental issues. According to Vladareanu et al. (2017), ocular anomalies such as inner epicanthal folds (vertical skin folds that cover the eye), the palpebral slant and Brushfield spots are common in neonates with DS (Weijerman & De Winter, 2010). Visual difficulties can also make academic progress more challenging for children with DS, given associated issues such as difficulty learning to read (Chen & Dote-Kwan, 2018; Lemons et al., 2018).

Hearing difficulties are found in a third to nearly one half of people with DS (De Schrijver et al., 2019; Nightengale, 2018; Raut et al., 2011). The most common hearing difficulty is conductive (sound does not reach the inner ear). Ekvall and Ekvall (2017) also note that some children with DS experience sensorineural hearing loss (where sound does not reach the brain as a result of defects with inner ear/auditory nerve). Unfortunately, even mild hearing loss can impact educational achievement, language and emotional development (Weijerman & De Winter, 2010).

Children with DS often also experience sleep issues, with difficulties in initiating and maintaining sleep, night awaking, early waking (Stores, 1993), circadian rhythm disorders (Ekvall & Ekvall, 2017), sleep disordered breathing, excessive daytime sleepiness (Bassell et al., 2015), sleep anxiety and parasomnias (Chawla & Heussler, 2017), teeth grinding, head banging and bedwetting (Stores, 1993). Some researchers assert that sleep difficulties in individuals with DS contribute to reduced cognitive development and EF (Chawla & Heussler,
2017; Joyce et al., 2019), as well as attention issues that can be detrimental for learning (Ashworth et al., 2015). For example, Breslin et al. (2014) found that children with DS and sleep apnoea had significantly less cognitive flexibility when compared to infants with DS without sleep apnoea, matched for mental age (MA). This may be because greater effort is required to complete everyday functioning, meaning less allocation of resources to EF. Notably, sleep disturbance could also contribute to the challenging behaviours often witnessed in individuals with DS (Wood & Sacks, 2004).

2.3.2 IQ and General Cognitive Functioning

The importance of cognitive functioning has been highlighted in much of the research related to individuals with DS (e.g., Lott & Dierssen, 2010). An understanding of cognitive functioning and how this relates to individuals with DS is especially important for researchers, educators and teachers that are interested in supporting the development of children and adolescents with DS. Cognitive functioning has been conceptualised as ‘multiple mental abilities, including learning, thinking, reasoning, remembering, problem-solving, decision-making, and attention’ (Fisher et al., 2019, p.18). In terms of cognitive functioning, individuals with DS are reported in the literature to demonstrate moderate or severe intellectual disability (ID) (Wishart, 1993; Boat & Wu, 2015), as indicated by intelligence quotient (IQ) scores.

One’s IQ is established by standardised tests or subtests, developed in the West, aimed at assessing human intelligence and cognitive functioning (Braaten & Norman, 2006). It must be noted that the understanding of intelligence may vary from culture to culture; thus, the tests developed in the West do contain embedded assumptions about intelligence – including the fact that it may be measured at all, and represented by a numerical score. According to Boat et al. (2015), individuals with moderate ID would record an IQ score ranging from 36-49 and those with severe ID would record scores ranging from 20-35, which can
give an indication of their level of cognitive functioning. Yet providing a ‘score’ to determine cognitive functioning can be reductionist, and there continues to be debate surrounding whether cognitive functioning, as denoted by an IQ ‘score’, is stable or can change over time, particularly in DS (Channell et al., 2014).

The mean IQ of individuals with DS is 50 (with a range of approximately 30-70), equivalent to the mental age of 5 or 6 years, although there are many variations within the degree of ID in individuals with DS (Mégarbané et al., 2013), and some do demonstrate borderline normal levels of intelligence (Wishart, 1998). In researching the cognitive functioning of individuals with DS, many researchers refer to mental age (MA) as opposed to chronological age (CA) (e.g., Ferreira-Vasques et al., 2017), and individuals with DS generally present as much younger in terms of MA compared to their CA (Caplan, Neece & Baker, 2015). MA is a person’s mental ability expressed or described as the average age at which a typically developing (TD) person generally reaches that ability (Ayers et al., 2007). The reason for the cognitive limitations and lower MA than CA in those with DS may stem from various delays and atypicality in the neurological development in this population. In support of this claim, Bartesaghi et al. (2015) indicate that brain development in the human foetus with DS is significantly delayed at the start of the second trimester, resulting in low IQ in children born with DS.

Some research has indicated that IQ scores for individuals with DS do not change with age. For example, Channell et al. (2014) explored IQ and non-verbal cognitive abilities in a longitudinal study involving 20 adolescent males with DS. The participants completed a measure of IQ (Leiter-R Brief IQ) and a non-verbal cognitive ability test across a period of three years. Notably, the study revealed that over time, there was no significant change in IQ, similarly to TD individuals. Of course, three years is a relatively short period of time for a longitudinal IQ study, and a much greater period of time may be required in
order to gain meaningful results. Yet these researchers also advocate for the use of ‘raw’ or growth scores, which reflect absolute level of ability and can therefore be useful to chart the acquisition of skills over time for individuals with DS, where this gain may be slow to develop (Channell et al., 2014). Whilst standard IQ remained stable, the growth scores revealed a significant increase in levels of absolute non-verbal cognitive ability overtime. Subdomain test findings suggested a significant increase in participants’ abilities to complete tasks based on visual organisation and deductive reasoning, with smaller (non-significant) gains in their performance on activities requiring inductive reasoning and fluid reasoning (required for abstracting, problem-solving, planning, decision-making and other higher-order learning processes). This suggests perhaps that the latter two subdomains are particularly challenging for individuals with DS.

It is critical therefore to examine both growth scores and standard IQ scores, as well as the contribution of each of the domains and how they may strengthen or decline over time. Some research reveals that the IQ of those with DS has changed over time, depending on their age. For example, some studies document increases or declines in IQ for individuals with DS during different developmental stages (e.g., Carr, 2005; Couzens et al., 2011). Wishart conducted an early study (1987) in this area, employing the Piagetian Infant Search Task to measure the cognitive functioning of children with DS (n=12, age 3-5 years old), observing an increase in cognitive functioning over a period of 10 weeks in those with DS compared to TD children (whose performance declined). Perhaps therefore the cognitive functioning of children with DS increases (Wishart, 1987), whereas it may halt in adolescence (Channell et al., 2014), and older adults with DS experience a decline in cognitive functioning and subsequent IQ (Couzens et al., 2011). Therefore, Wishart (1987) cautions against drawing conclusions about the cognitive functioning of those with DS through single testing sessions or in the absence of normative data/a TD control
group, as there may be important qualitative differences (behaviourally and verbally, for example) identified in the way children with DS and TD children express competence in performance. However, 12 participants may be insufficient from which to draw accurate or generalisable conclusions in Wishart’s (1987) study, although further studies from Wishart with greater numbers of participants do indicate that children with DS are able to continue to learn and do continue to develop their cognitive functioning, perhaps albeit at a slower rate (on average) than TD children (e.g., Wishart, 1988; 1990; 1991; 1992; 1993; Wishart & Bower, 1984; 1985; Wishart & Duffy, 1990). Moreover, measuring cognitive performance may not be accurately achieved by measuring IQ, depending on the contents and purpose of the test, rendering it difficult to draw comparisons across studies.

Yet the stability of IQ in adolescents with DS may also be a misnomer. Even in Channell’s et al. (2014) study, which was focused on adolescence as opposed to infancy, 23-37% of participants scored at the floor level (IQ at or below 36) of standard scores at every time point within the Leiter-R Brief IQ, creating a result that appeared stable, but could actually be masking a downward trajectory of IQ. Yet as has been mentioned, this does not necessarily mean that cognitive functioning declines; Channell et al.’s (2014) study found on growth scores that adolescents with DS continued to gain and develop non-verbal cognitive skills over time, leading them to conclude that on average, ‘individuals with DS are learning and able to demonstrate new cognitive skills across adolescence’ (p. 25).

2.3.3 Verbal and Non-Verbal Ability
Separating the verbal and non-verbal elements of the DS profile/phenotype for cognitive functioning is useful as it enables educators to move beyond simplistic IQ scores, helping them to design or use the most supportive methods available to help individuals with DS in their learning journey. For example, verbal and
non-verbal performance can be distinguished to allow educators to have a good understanding of the variability in these abilities. Verbal ability involves being able to process information involving speech sounds and words (Yang, Conners & Merrill, 2014).

Verbal MA (VMA), then, is drawn from cognitive tests that predominantly assess verbal abilities, giving an approximate age for an individual’s language skills and verbal performance compared to normative data (Reese, 1962). Performance MA (PMA) (often referred to as non-verbal or visuo-spatial MA) is drawn from cognitive tests that predominantly assess non-verbal abilities, although there may be a degree of overlap in VMA and PMA, especially if verbal abilities are required to complete PMA tests (Sumowski, 2018). This could arguably indicate an issue of task impurity.

Despite the fact that the literature indicates that individuals with DS are often classified as having moderate to severe ID, it is important to not simply focus on their overall IQ score, but to consider the various facets of cognitive functioning, including adaptive and functional behaviour and explore any differences in the verbal and visuo-spatial or performance dimensions of their IQ profile (Mecca et al., 2015). This can help to ascertain syndrome specificity (Dykens, Hodapp & Finucane, 2000), establishing the features of cognitive functioning that appear to be most affected in those with DS. For example, some studies (such as Cebula et al., 2017) have used IQ scores to assess the difference between VMA and PMA, finding that the PMA is generally higher than the VMA in those with DS, although other studies (such as Campbell et al., 2013), that used other verbal assessments (not IQ tests) to measure cognitive functioning, have found no significant difference between PMA and VMA, indicating greater research is required in this area. Research does generally indicate however that those with DS initially demonstrate mild delays compared to TD children in cognitive functioning, yet these delays become more pronounced by school-age.
Furthermore, verbal delays and difficulties appear to persist more than visuo-spatial skills, which remain on a trajectory for MA (Grieco et al., 2015).

Studies of cognitive functioning in DS include that of Mecca et al. (2015), who studied 30 children with DS between 3-8 years to assess a variety of areas of non-verbal cognitive functioning. They used the Leiter International Performance Scale-Revised (Roid & Miller, 1997) and compared their participants to 30 TD children, matched by CA, sex and school. Mecca et al. (2015) found that those with DS achieved lower scores compared to the TD group in subtests assessing fluid reasoning performance and visual processing, and that girls with DS did better in inductive reasoning than boys with DS. They concluded that it was important to take into account the visual and linguistic difficulties faced by children with DS (and other ID) in completing standard IQ tests (and, presumably, gender), and that subtests within non-verbal tasks can illuminate areas of strength or weakness compared to TD children. Yet even studies such as this can be critiqued as providing merely a ‘snapshot’ of an individual’s cognitive functioning, instead of helping foster an understanding of how and why specific cognitive weaknesses arise in individuals with DS, similarly to the TD population.

Interestingly, children with DS appear to demonstrate relative strengths in non-verbal cognitive abilities, yet this may often only be because non-verbal scores are compared to the significant challenges in verbal cognitive abilities. Various studies demonstrate this; for instance, Yahia et al. (2014) found that verbal short-term memory along with expressive language difficulties impairs the communication ability of individuals with DS, resulting in the comparative strength of non-verbal skills. However, some researchers, including Grieco et al. (2015), indicate that the verbal abilities in toddlers with DS progress at a greater rate than in adolescents and adults with DS. From this, it can be surmised that advancement of verbal abilities decelerates with age in individuals with DS. As
noted, non-verbal abilities in those with DS are often perceived as a relative strength, with visuo-spatial abilities in individuals with DS in particular becoming the focus of much research.

2.3.4 Visuo-Spatial Abilities in Down Syndrome

One of the most significant areas researched is visuo-spatial ability, which involves being able to process visual information involving spatial relations (Yang et al., 2014). Various authors assert that good visuo-spatial ability compared to verbal ability remains a key feature of the cognitive phenotype for DS (for example, Chapman & Hesketh, 2000; Davis, 2008; Silverman, 2007). As Yang et al. (2014) indicate, possessing a relative strength in visuo-spatial ability generates advantages in various aspects of daily life, such as organising a cupboard or wardrobe, finding one’s way home from school, buttoning one’s jacket, understanding right from left and catching a ball. However, as Yang et al. (2014) point out, it is possible to possess superior visuo-spatial performance compared to verbal performance, yet for visuo-spatial performance to remain below CA norms.

Yang et al. (2014) conducted an extensive systematic review of 49 studies on visuo-spatial abilities, separated into five different domains: 1) visuo-spatial memory; 2) visuo-spatial construction; 3) mental rotation; 4) closure; and 5) wayfinding (these five areas were, Yang et al. (2014) note, originally conceived by Lohman et al. (1987) and Carroll (1993)). Most studies compared a TD group with a group of individuals with DS, matched on MA, on a task measuring one of the five aforementioned abilities. Yang et al. (2014) found that spatial sequential memory was commensurate with overall cognitive ability in individuals with DS, whereas spatial working memory, closure and wayfinding present particular challenges for those with DS. There were mixed findings for visuo-spatial construction and mental rotation. They concluded that there was an uneven profile of visuo-spatial abilities in those with DS, where some are commensurate
with overall cognitive ability level, and others present certain areas of weakness. Some studies (e.g., Meneghetti et al., 2017) have found that mental rotation ability is significantly poorer in children and adolescents with DS compared to TD children matched on MA, especially for smaller angles of rotation (0-45 degrees).

Thus, whilst most studies indicate that visuo-spatial abilities signify a relative strength in those with DS, this is often only relative to their verbal abilities or MA (which often includes verbal abilities, thus biasing overall strengths towards non-verbal abilities), not general cognitive ability. Moreover, despite the fact that most studies tend to group visuo-spatial abilities together, it is important to untangle the different kinds of visuo-spatial ability to truly understand the potential relative strengths and areas that present particular challenges in individuals with DS in each. This can enable specific interventions to be generated, targeting particular visuo-spatial skills (e.g., path learning in Meneghetti et al.’s (2017) study).

2.3.5 Numerical Skills in Down Syndrome

Basic numerical skills, including being able to count and solve straightforward arithmetic problems, are critical daily life skills. Studies involving individuals with DS reveal that those with this syndrome consistently demonstrate lower levels of attainment in arithmetic than their CA- and MA-matched TD peers (e.g., Abreu-Mendoza & Arias-Trejo, 2015; Agheana & Duță, 2015; Faragher, 2013; Sella, Lanfranchi & Zorzi, 2013). For example, Sella et al. (2013) compared the performance of 21 children and adolescents with DS with MA- and CA-matched TD counterparts, to investigate whether mathematical delays in individuals with DS can be attributed to their general intelligence level or to specific difficulties in basic numerical skills. The researchers were specifically assessing the Object Tracking System (Mandler & Shebo, 1982), which ‘is a domain-general system that encodes spatio-temporal characteristics of objects with a capacity limited to
three-four items’ enabling observers to ‘quickly, accurately and effortlessly perceive the numerosity of small sets, a phenomenon known as subitizing’ (Sella et al., 2013, p. 3798-3799) and the Approximate Number System (Dehaene, 1997). The Approximate Number System is activated when numbers exceed the subitizing range (greater than 4), enabling individuals to approximate numbers. They found weaker performance of those with DS in subitizing even compared to their MA-matched peers, which continued to decline with increasing numerosity, whilst both MA- and CA-groups performed at ceiling. This indicates challenges to the object tracking system in those with DS. However, the Approximate Number System was in line with their MA-matched controls, suggesting the Approximate Number System was typical for their MA albeit developmentally delayed for their CA. This is in line with other research, such as Van Herwegen, Ranzato, Karmiloff-Smith and Simms (2020), who found that for TD and DS populations, mathematical abilities were predicted by visuospatial abilities, not Approximate Number System abilities, which was in line with overall MA.

Differences in eye movement patterns have also been investigated in DS, which have led to insight as to why counting skills are sometimes poorer in those with DS. For instance, Ranzato, Tolmie and Van Herwegen (2020) compared 24 individuals with DS to TD children matched for mental age on a subitizing/counting task and found that in the DS group, inefficient scanning strategies and shorter fixation duration was employed that, whilst rendering no significant effect on subitizing performance, had a significant negative effect on counting accuracy. The authors conclude that their findings support Van Herwegen and Karmiloff-Smith’s (2015) argument that basic abilities may negatively impact numeracy skill development.

Yet despite studies indicating delays in various areas of numerical ability in DS, there are studies that assert that teaching style can assist in helping those with
DS to improve their mathematical skills. For example, a study in Australia (Faragher et al., 2008) involved 12 children with DS between 6-12-years-old in task-based interviews, using the Early Numeracy Interview (Clarke et al., 2002, cited in Faragher et al., 2008) and the Early Mathematics Understandings Instrument (Gervasoni, 2004, cited in Faragher et al., 2008), both adapted for DS. The researchers found that there were various approaches and strategies that could be adopted by both teachers and parents to assist the numerical development of children with DS. This includes taking a visual and kinaesthetic approach (instead of simply using oral language), using matching, selecting and naming of numbers, using slow counting songs (Makaton or signing as well as oral singing), using real objects, the use of ten-frames, five-frames, number lines, empty number lines and hundreds charts, and Numicon (Wing & Tacon, 2007), a multi-sensory mathematics teaching programme. Whilst children with DS still demonstrated weaknesses in their mathematical abilities compared to TD norms, these interventions were found to improve their skills and confidence in basic arithmetic.

2.3.6 Memory Abilities in Down Syndrome
There are other specific areas in which those with DS appear to experience significant challenges, alongside VMA. For example, those with DS often demonstrate difficulties in both short-term memory (STM) (which is part of working memory (WM) (Cowan, 2008) and long-term memory (LTM). In terms of STM, research shows that both non-verbal STM compared to TD children matched for MA (Lanfranchi et al., 2015) and, in particular, verbal STM (VSTM) compared to TD individuals matched for MA (Pennington et al., 2003; Jarrold et al., 2002) is affected in individuals of varying age ranges (e.g., in Pennington et al.’s (2003) study, participants were aged 11-19; in Jarrold et al.’s (2002) study, participants were children to young adults). These studies indicate that throughout development, both verbal and non-verbal STM performance remains consistently below MA expectations. Indeed early research (e.g., Hulme &
Mackenzie, 1992, cited in Pennington et al., 2003) reveals that verbal STM lags behind MA in DS, which can explain some of the language-related issues and challenges exhibited by those with DS. Yet comparisons of individuals with DS with other ID populations, such as Williams syndrome and Fragile X syndrome, reveal that non-verbal STM is often stronger (than VSTM and other populations with developmental disabilities) in those with DS (Edgin et al., 2010; Lamônica & Ferreira-Vasques, 2015), which indicates further that it is specifically VSTM that is particularly affected in individuals with DS.

VSTM in particular is considered a prevalent cognitive challenge in children with DS (Jarrold, Baddeley & Phillips, 2002) compared to TD children matched for MA (Lee, Pennington & Keenan, 2010). In terms of the specific aspects of the memory system involved, it is clear from research that those with DS do not have relative weaknesses in rehearsal compared to TD matched controls (Jarrold, Baddeley & Hewes, 2000). Rather, Sabat et al. (2020) assert that the issue lies in the storage of information in the phonological loop, not in rehearsal. However, it should be noted that this may not be the result of auditory difficulties in DS comparative to TD (Jarrold et al., 2002). This STM difficulty is a significant contributor to the language difficulties often observed in individuals with DS (Laws et al., 1996).

Regarding long-term memory (LTM), various studies have indicated that child and adolescent individuals with DS show difficulties in VLTM when compared to TD peers matched for MA (Pennington et al., 2003; Nichols et al., 2004). Godfrey and Lee (2018) found that VLTM weaknesses in individuals with DS are evidenced by their low performance in list-learning tasks and other tasks requiring VLTM tasks when compared to MA-matched TD groups. The performance of adolescents with DS on verbal list-learning long-delay tasks was similar to the performance of individuals with Williams syndrome, yet
interestingly those with DS have been found to perform better on word-list learning tasks (Godfrey & Lee, 2018).

Similarly to VLTM difficulties, compared to TD children matched on MA, adolescents with DS demonstrate reduced performance on non-verbal LTM tasks (Byrne et al., 2002; Pennington et al., 2003). Basten et al. (2018) found that people with DS demonstrate poorer performance on non-verbal LTM tasks involving recalling and recognition of new information compared to MA-matched children. However, studies indicate that the performance of adolescents with DS in visual and spatial LTM tasks is considerably higher than in verbal tasks (Jarrold, Baddeley & Phillips, 2007; Vicari, Bellucci & Carlesimo, 2005).

Working memory (WM) in individuals with DS is also significantly poorer than that of their TD counterparts (both CA- and MA-matched) (Tungate & Conners, 2021). In keeping with findings regarding the STM and LTM, researchers have found that visuospatial WM appears to present fewer challenges in individuals with DS compared to verbal WM (Pulina, Carretti, Lanfranchi & Mammarella, 2015; Godfrey & Lee, 2018). The WM in children and adolescents with DS is discussed in detail in the third chapter (the systematic review).

Given that the research reveals greater challenges in verbal cognitive functioning compared to non-verbal functioning, it is critical to delve a little deeper into the specific language-based difficulties experienced or demonstrated by those with DS.

2.3.7 Language in Down Syndrome

Language development is typically one of the main areas of difficulty for individuals with DS (Udhnani et al., 2020), lagging behind other developmental areas within cognitive development, a gap that becomes more pronounced as children with DS age (Coggins & Stoel-Gammon, 1982). Previously, researchers
suggested there was no evidence that language development deviated from the ‘normal’ developmental course in DS, claiming that it simply developed more slowly than in TD children (the ‘stretched-normal’ hypothesis) (Lenneberg, 1967). However, subsequent research suggested that certain elements of language development appear to be more delayed or challenged in those with DS (Miller, 1988), indicating that those with DS may indeed have a somewhat different linguistic profile and system (Rondal, 1988).

Research into individuals with DS typically assesses both expressive and receptive language (Kumin, 2015). Receptive language (understanding and comprehension) is generally stronger than expressive language (speaking) amongst DS populations (Channell et al., 2015), with some variability (Martin et al., 2009). This is known as the receptive-expressive gap (Gibson et al., 2014). Whilst this gap has been found to remain and even grow throughout the lifespan (Laws & Bishop, 2003), in that adults with DS tend to find tasks requiring receptive skills easier than tasks requiring expressive language, some research indicates the gap becomes more individualised (e.g., Kumin, 2015).

Expressive vocabulary has been found to develop more slowly in children with DS compared to TD peers (Mervis & Robinson, 2000; Roberts et al., 2007), with fewer words spoken during narration and conversation compared to non-verbal MA-matched TD children (Chapman et al., 1998). Polisenka and Kapalkova (2014) investigated the language profiles of 13 children with DS and 16 children with language difficulties, comparing them also to 58 TD children matched for their receptive or expressive vocabulary size. They found that those with DS had similar grammar and vocabulary development to those with language difficulties, again demonstrating difficulties in the DS population in these areas of linguistic development.
Speech intelligibility, which is the degree to which individuals can understand the acoustic signals given to them by a speaker without the need for supporting information (Duffy, 2005; Yorkston, Strand & Kennedy, 1996), thus denoting expressive language skills, has been consistently found to be reduced in children with DS (Kumin & Adams, 2000; Kumin, 1994; 2006), persisting into adolescence and adulthood (Miller & Leddy, 1999; Roberts, Chapman & Warren, 2008). Roberts et al. (2005) found that the intelligibility level was much lower than the MA for children with DS. Moreover, whereas it takes TD children approximately 48 months to master speech intelligibility (Togram, 2015), many individuals with DS never master this skill, experiencing challenges throughout their entire lives (Roberts, Price & Malkin, 2007).

In terms of more specific areas of difficulty with language, especially with expressive language, research indicates that phonology, pragmatics and syntax appear to be particularly challenging for those with DS (Martin et al., 2009). In addition, individuals with DS also display difficulties in vocabulary, pragmatics (e.g., Roberts et al., 2007), hearing skills (Roizen, 2007), difficulties in oral-motor skills (Barnes et al., 2006) and literacy (Bird et al., 2008). For example, whilst most children with DS do gain literacy skills, there is significant variability in the level of skill obtained (Bird, Cleave & McConnell, 2000; Turner & Alborz, 2003).

Interestingly, reading skills do tend to be a relative strength in those with DS, although research from Buckley, Bird, Sacks and Archer (2006) and Laws, Byrne and Buckley (2000) reveals that those with DS attending mainstream schools outperform their peers at special schools, which indicates that this relative strength may be strongly impacted by environmental factors (in keeping with Morton’s (2004) causal developmental framework, discussed in due course later in the chapter). Reading ability has also been linked to differences in phonological skill (the ability to map sounds onto letters and reflect on sound structures within speech) (Kalayçi & Diken, 2020). Those with DS often exhibit a
different developmental path for phonological awareness compared to TD peers; they have been found to easily identify initial sounds in words but have difficulty identifying rhymes (Nash, 2007), and Næss, Ostad and Nygaard. (2021) reported in their study that the phonological awareness of children with DS was poorer than that of TD peers yet, unlike for TD children, was a significant predictor of expressive vocabulary development for children with DS. This, and other research into this topic, indicates that phonological awareness may not be commensurate with cognitive ability in children with DS (Snowling, Nash & Henderson, 2008).

Despite relative strengths in reading within receptive language comparative to expressive language skills, within the domain of receptive language development there may also be specific difficulties. Abbeduto et al. (2001) found that participants with DS had lower age-equivalent scores on the Test for Auditory Comprehension of Language (TACL-R) than a group of TD children matched for MA. This indicates that individuals with DS may be less able to comprehend spoken language, comparative to their non-verbal cognitive skills.

However, there have been conflicting findings regarding receptive vocabulary skills; some studies show that children and adolescents with DS understand spoken vocabulary, a core skill of receptive language, at the same level as MA-matched TD children (e.g., Laws & Bishop, 2003), whilst other studies indicate that children with DS actually score lower on standardised measures of receptive vocabulary compared to TD, MA-matched children (Price et al., 2007). These conflicting findings could be due to the hearing status, age or the different measures used with participants to measure receptive language skills, however. There is also a relationship between DS-related difficulties in language and some EFs, such as self-regulation. For example, Grouios and Ypsilanti (2011) found that people with DS have language difficulties that contribute to challenging behaviours and that these language delays are more pronounced in
adolescents than in children. Thus, the poor self-regulation observed in DS and subsequent challenging behaviour might stem, in part, from language difficulties as they struggle to express their thoughts and feelings and may have poorer ‘self-speech’, which is important in self-regulation (Glenn & Cunningham, 2002). Other EF difficulties may also be associated with language difficulties, as discussed by Cuskelly et al. (2016), who found an association between low levels of receptive language ability and difficulties with delayed gratification in those with DS.

Studies indicate that a number of factors may influence or cause the poor development of language in people with DS. Weaknesses in grammar and vocabulary present in early childhood are thought to be due to difficulties in verbal WM, amongst other things (Polisenka & Kapalkova, 2014). Polisenka and Kapalkova (2014) also suggest, based on the results of their study, that individuals with DS demonstrate difficulties not only with higher-order cognitive processing of information, which can affect language skills, but also difficulties in navigating interpersonal interactions, which can generate social cognitive challenges that may continue to adversely affect language development.

2.3.8 Social Development and Social Cognition

Despite demonstrating weaknesses in speech, communication and language, individuals with DS are stereotypically perceived to be highly sociable, affectionate, happy and with good ‘people’ skills, by both the general public and even some professionals working with this group (Fidler et al., 2008; Hines & Bennett, 1996; Wishart & Johnston, 1990). An observational study examining the peer-related social competence of children with DS supported this view, finding that children with DS often display social skills and a willingness to interact socially much like TD children (Guralnick, Connor & Johnson, 2011). Moreover, Freeman and Kasari (2002) found that the friendships formed in preschool by those with DS were not significantly different in stability and nature.
than friendships formed by TD children; however, they also found that the number of friendships in children with DS tended to decline with age, and were not always reciprocated. From the findings of these researchers, it seems reasonable to claim that social development is a relative strength of toddlers with DS, at least. However, as Guralnick et al. (2011) argue, much research has also established that children with DS have considerable social competence problems that go on to affect many of their relationships.

A review of the literature by Martin et al. (2009) found that children with DS form interpersonal relationships in the same way as TD children. However, Martin et al. (2009) also cites research suggesting that the social skills of those with DS often decline with age, as adults with DS often show significant behaviour disorder, which is dependent on childhood psychopathology and functioning (McCarthy, 2008). Thus, despite the sometimes mixed findings, and the stereotype of those with DS being highly sociable, it can be concluded that the majority of older children with DS have relatively poor peer relationships due to a myriad of factors, including friendships with TD peers drifting as TD individuals age, a lack of autonomy given to those with DS from family members (Gilmore et al., 2009), a lack of inclusion (McFadden et al., 2017) and the characteristics of their condition.

There are many potential reasons why children with DS may struggle in their social relationships as a result of the characteristics of their condition, often stemming from challenges in their social cognition. Social cognition is broadly defined as ‘the ability to process and respond appropriately to the behaviour, emotions and intention of others’ (Cebula & Wishart, 2008, p.44). Difficulties with social cognition may start from an early age. Research suggests that children with DS attend to people’s eyes (the ‘mutual gaze’) for longer than TD children, who begin to look around at their environment after one year of age (Berger & Cunningham, 1981). This may be because those with DS need longer to extract
the same amount of information as TD children (Sigman, 1999). They also focus more on eyes, rather than facial features (Berger, 1990), which may hamper emotion recognition; additionally, they find it more difficult to switch attention between people, objects and the environment (Krakow & Kopp, 1983). For example, Schworer et al. (2020) recently compared 58 infants with DS with 40 TD infants, assessing their performance on four dimensions that are perceived as precursors to EF: action planning, sustained attention, processing speed, and attention shifting. They found that those with DS were slower to shift their attention, focused on objects for longer and were observed to take longer to touch objects, compared to TD infants. Crucially, they found that this early attention shifting significantly predicted later EF performance in those with DS. This difficulty in shifting attention as an infant may also hamper joint attention (Kasari et al., 1995), a building block of sharing interests, intentions and desires with others, as well as language acquisition (Carpendale & Lewis, 2006). Furthermore, Kasari et al. (1995) found that difficulties in shifting attention and coordinating joint attention were more observable in activities and contexts that generated greater cognitive load.

Language weaknesses could also be associated with difficulties in social competence; for example, individuals with DS appear to experience challenges when it comes to acknowledging the information that a listener may require to understand them. Moreover, they appear less able to provide ‘scaffolding’, which can give listeners the speech references they need to interpret meaning (Abbeduto & Murphy, 2004). Language plays a fundamental role in mediating the majority of social interactions; thus, challenges in scaffolding can potentially lead to confusion or social misunderstandings, which, alongside reduced speech intelligibility, can compound existing social cognitive delays and difficulties.

Other research points to difficulties with social knowledge - the ability to analyse social situations for the ‘social rules’ and abide by these social rules.
appropriately. Barisnikov and Lejeune (2018) for example used a social resolution task in 351 TD children (4-12 years) and compared them to 20 individuals with DS (10-18 years). They found that the DS group showed significantly greater difficulties in identifying, judging and reasoning surrounding social rule transgression and demonstrated reduced social awareness.

Theory of Mind (ToM) has also been found to be delayed in those with DS (Zelazo, Burack, Benedetto & Frye, 1996) comparative to their mental age, which can mean those with DS struggle with mentalising and emotional intelligence, as they are less able to perceive things from the perspective of others (Tavakoli, Demehri & Azizi, 2019). This links with cognitive flexibility; indeed, ToM and EF ability more generally have often been associated. Wade et al. (2018) for instance established that, using brain imaging studies from a variety of individuals (TD and non-TD), despite the fact that separable mechanisms appear to underlie both EF and ToM, there are 'shared mechanisms for domain-general processing that support both abilities' (p. 2119). Abbeduto et al. (2001) for instance explored the linguistic and cognitive profile of individuals with DS, by comparing the receptive and expressive language and ToM skills of individuals with DS (n=25) to individuals with Fragile X syndrome (n=18) and TD children (3-6 years-old, n=24)) matched to nonverbal MA (average MA=4 years in the DS group). Fragile X syndrome is genetic condition also associated with ID; individuals with this syndrome often have an IQ level that is typically similar to that observed in individuals with DS (Abbeduto et al., 2001), thus providing a useful comparison. Abbeduto et al. (2001) found that participants with DS demonstrated greater challenges in each of these areas compared to TD children and individuals with Fragile X Syndrome.

Research also suggests however that children with DS often use social skills as strategies to compensate for ID, for example, by distracting other people when exposed to a challenge (Pitcairn & Wishart, 1994). Pitcairn and Wishart (1994)
asked three groups of 3-5-year-olds (those with DS, TD children matched for CA, and TD children matched for MA) to complete a shape-sorting task involving both possible and impossible shapes. Whilst the DS group was as proficient at matching ‘possible’ shapes to the appropriate holes, their responses to the impossible shapes differed. The researchers stated that those with DS behaved in ways so as to divert attention from the task, often in socially appealing ways that were inappropriate for completing the task. This behaviour may, the researchers’ opine, be why many individuals with DS are perceived as being highly sociable yet struggle with learning (Pitcairn & Wishart, 1994, p.485).

Hahn (2016) also found that individuals with DS use social skills as strategies to compensate for ID, for example by distracting other people when exposed to a challenge. Hahn (2016) noted for example that toddlers with DS increasingly use non-verbal social acts, such as pointing, smiling, play acts and gesturing, to sustain the attention of others and distract from challenging tasks. Thus, the development of social skills in the early stages of life enables individuals with DS to interact with the environment and detract from any challenges they may face (Barisnikov & Lejeune, 2018). However, the findings of Barisnikov and Lejeune (2018) show that children with DS grapple with social understanding and reasoning, externalising behaviour and sometimes demonstrating challenging behaviour as a result.

2.3.9 Challenging Behaviour
A low level of IQ is often associated with challenging behaviour. In connection with this view, Foley et al. (2016) indicated that young individuals with ID, including those with DS, demonstrate significant and lasting challenging behaviour compared to non-disabled peers. Often, challenging behaviour occurs twice as often in DS as in the general population (Dykens & Kasari, 1997). This could be due to a deceleration of cognitive development in their early childhood and cognitive delays that become more evident as they advance in age.
(McCarthy, 2008). It could also be due to a range of biological, developmental, psychological and social stressors (Allen, 2008). For example, the environmental demands on children with DS may be a significant precursor of challenging behaviour; it may be, for instance, that teachers set tasks that are too challenging for the developmental level of children with DS, or that children with DS experience bullying or goading by other peers, which may lead to challenging behaviour in a classroom environment.

However, Foley et al. (2016) found that the incidence of emotional and behavioural problems was lower in 323 school-aged children with DS when compared with 466, 3-19-year-olds in Australia with other ID, stemming from a range of other genetic and environmental causes and co-occurring conditions. The caregivers of individuals in each group completed the Developmental Behaviour Checklist over three time points, with scores lower in the DS group compared to the group without DS (with other ID), indicating fewer behavioural problems, except on the communication disturbance subscale. Interestingly, whilst for both groups the scores on the subscales of anxiety, communication disturbance, disruptive behaviour and self-absorption declined over time, the symptoms of depression remained the same for those with DS, whereas in the ID group these symptoms declined. Depression is a common occurrence in those with DS - up to 11% of those with DS exhibit depression (Walker et al., 2011).

Children as well as adolescents with DS often present with conduct disorders, such as disruptive behaviour disorder (Yahia et al., 2014), with pronounced externalising behaviours (Ersoy, Guler & Çetin, 2018). Externalising behaviours include stubbornness, disobedience, inattention and oppositional behaviour, and these increase with a child’s age. When those with DS enter adolescence, internalising behaviours (such as withdrawal, secrecy, preferring to be quiet and alone) may also increase (Dykens et al., 2002; Ersoy, Guler & Çetin, 2018).
Dykens et al. (2015) found that children with DS present fewer disruptive behaviours, in addition to distinctive social and motivational profiles, compared to adults with the same syndrome. Studies also indicate that children with DS aged between 5 and 11 years old present greater hyperactivity (Yahia et al., 2014) than TD children, often demonstrating more externalising behaviours during late childhood. Thus, DS can lead to many children with this syndrome being hyperactive and therefore generating disruption, as found by Jacola et al. (2014).

Studies indicate that there is a relationship between challenging behaviour and EF in DS. Challenging behaviour affecting children with DS can be partially attributed to delays and difficulties in EF domains (Daunhauer & Fidler, 2013; Lee et al., 2011; Memisevic & Sinanovic, 2014; Rowe, Lavender & Turk, 2006). For example, research shows that weaknesses in effective decision-making, self-regulation (Cuskelley, Einam & Jobling, 2001) and cognitive flexibility (Zelazo, Burack, Benedetto & Frye, 1996), in relation to MA, can generate challenging behaviour. Cuskelley, Einam and Jobling (2001) found that in 31 individuals with DS, only approximately one third to half of participants (36% and 48% in trials 1 and 2 respectively) were able to delay their gratification on a task. Those that waited for the experimenter, as instructed, tended to exhibit greater expressive language, which aligns with the theory that self-speech is important in self-regulation (Vygotsky, 1962; Glenn & Cunningham, 2002). This difficulty for many individuals with DS to self-regulate and delay gratification is more pronounced in childhood than adulthood and can create significant challenges in terms of behaviour when individuals with DS are required to wait, especially affective expression/outbursts (Bieberich & Morgan, 2004). This challenging behaviour may also have a significant impact on the daily living skills and quality of life for those with DS.
2.3.10 Daily Living Skills

Several studies indicate that people with DS experience difficulties in performing and completing daily activities, such as dressing (Hayton, Wall & Dimitriou, 2020). Some studies have used the Vineland Adaptive Behaviour Scale (Sparrow, Balla & Cicchetti, 1984), administered to parents, to assess areas of strength and weakness in adaptive behaviour functioning in four domains: socialising, communication, daily living and motor skills. Fidler, Hepburn and Rogers (2006) found for example that those with DS demonstrate relative strengths in socialisation (especially in play and leisure time items) compared to communication (especially expressive language) and gross motor skills, when compared to children with other developmental difficulties, matched for MA. However, other studies do indicate that whilst those with DS perform significantly worse overall on the Vineland Adaptive Behaviour Scale than TD individuals, their adaptive behaviour and daily living skills increasing with age, plateauing at approximately age 30 years (Dressler et al., 2010).

Movement can also be atypical in DS. For instance, Brantmuller, Gyuro and Karacsony (2015) highlight that the inability to walk independently poses a serious challenge to people with DS as they go about their daily lives. These researchers found that movement ability is significantly affected by their body weight, as joints can become weakened (Pavan, 2001). Other research (e.g., Dolva et al., 2004) found that children with DS demonstrate difficulties with toileting when in school, compared to age-matched TD peers. According to Ryan and Mahmood (2017), this is because of low muscle tone in bladder and bowel control (Ryan & Mahmood, 2017). People with DS also demonstrate difficulties in bathing, dressing (Hayton et al., 2020) and eating (Hudnall, 2014).

Hearing and visual difficulties in children and adolescents with DS (Dolva, Coster & Lilja, 2004) can also mean that some individuals cannot complete tasks involving daily skills to the same level of competence as their TD peers,
despite tasks involving visual and auditory processing providing a relative strength in people with DS (Chapman & Hesketh, 2001). Research furthermore indicates that there is a significant positive relationship between adaptive behaviour and EF, in particular WM, in that those with DS often display poor adaptive behaviour, which is associated with poorer functioning in some elements of EF (e.g., Tomaszewski, Fidler, Talapatra & Riley, 2018; Van Duijn et al., 2010). This can affect future life chances (Tomaszewski et al., 2018), particularly if the individual with DS displays a co-occurring condition.

2.4 Co-occurring Conditions
Those with DS can often demonstrate co-occurring conditions, such as poor mental health, ADHD and ASD, which can contribute to challenging behaviour and learning difficulties.

2.4.1 Mental Health Problems
Research reveals that young people with DS commonly experience mental health difficulties, with the prevalence of these lying between 18 and 38% (Fujino, 2017). The difficulty of diagnosing mental health conditions in the DS population however is compounded by the fact that there may be symptoms of a mental health disorder that are actually caused by something else, such as sleep disorder (a common issue in those with DS, as mentioned previously). For instance, Dykens et al. (2015) highlight the greater existence of depressive disorders in those with DS; Ersoy, Güler and Çetin (2018) for example estimate the prevalence of depression in people with DS to be up to 11%, compared to 2-10% of the general population. In children with DS, however, the frequency of depression has been found to be considerably lower than in adults with DS (Haddad, Bourke, Wong & Leonard, 2018), yet still greater than TD children. The risk factors for depression in people with DS include a small brain volume, as well as a reduced hippocampus volume, when compared with the general population, and reduced serotonin during foetal development, a deficiency that
persists into adulthood (Ersoy, Güler & Çetin, 2018). Societal factors can also play a large role in contributing to the onset and maintenance of depression in those with DS, including alienation, stigma, lack of relationships and reduced life chances (Stein et al., 2013).

In terms of anxiety, Generalised Anxiety Disorder is not thought to be common in individuals with DS compared to other groups of people with ID. Ersoy, Güler and Çetin (2018) do note that symptoms such as irritability, trembling and fear can be common in DS, yet studies have found phobia disorders are uncommon: Myers and Pueschel (1991) found that only 1% of 497 people with DS had a phobia disorder (compared to 7-10% of TD children (Lichtenstein & Annas, 2000)). Similarly, they found that 1.7% of 236 individuals with DS had obsessive-compulsive disorder (OCD), comparable to 1.65-2.5% of the general population, although another study has found this rate to be 4.5% in individuals with DS (Prasher & Day, 1995). According to Glenn (2017), however, OCD is considerably lower in people with DS, creating little consensus in the literature. However, Ersoy, Güler, and Çetin (2018) claim that the prevalence of OCD is higher in adolescents and adults compared to children, revealing an increase in its prevalence with age, which is similar to TD individuals.

Interestingly, various studies have revealed a link between EF and mental health, with those diagnosed with mental health problems and disorders demonstrating poorer EF. Akbaryan (2014) for example reviewed 106 studies in this field, involving a range of participants (not only those with DS) and found that EF promoted many resilience factors, including physical, behavioural, emotional and mental health, with mental health in particular demonstrating a significant link with EF. Those with poor EF were more vulnerable to disorders such as psychosis, post-traumatic stress disorder (PTSD) and depression; it could therefore be speculated that those with DS are more likely to have poorer
mental health due, in part, to reduced EF, although as has been highlighted, other factors (such as social factors) undoubtedly play a role.

2.4.2 Attention Deficit Hyperactivity Syndrome

Many studies suggest that attention deficit/hyperactivity disorder (ADHD) (a behavioural disorder with symptoms of impulsiveness, hyperactivity and inattentiveness) (Barkley & Murphy 2006) is more prevalent in people with DS than in the general population. For instance, Ersoy, Güler and Çetin (2018) in their review show that the prevalence of ADHD in the general population is 5% compared to between 14-43.9% in people with DS. Oxelgren et al. (2017) also found that the prevalence of ADHD is higher in children and adolescents with DS than in TD individuals and those without DS. According to Ekstein, Glick, Well, Kay and Berger (2011), the occurrence of this condition in children with DS is very high, reaching 43.9%. However, this high prevalence and the vast range (14-43.9%) in those with DS may be because ADHD is difficult to diagnose even in the TD population; in populations with DS, it is possible that some of the symptoms of ADHD have a different underlying cause, such as inattention as a result of poorer WM and EF more generally, or hyperactivity as a distraction technique from challenging tasks. Ersoy, Güler and Çetin (2018) assert that impulsivity as well as hyperactivity become more prominent at 36 months in children with DS, which coincides with the age that those with DS begin more formal education (such as preschool); this could link with struggles within the classroom environment, rather than organic causes (such as ADHD). Schreiber et al. (2014) also point out that children with DS and ADHD exhibit more delay in EF than those with DS and without ADHD; this indicates that some ADHD symptoms could, in fact, be caused by EF challenges. Thus, the issue of misdiagnosis of ADHD may remain prevalent in DS populations.
2.4.3 Autism

According to the DSM-V, Autism Spectrum Disorder (ASD) is a neurodevelopmental disorder characterised by challenges in social communication and social interaction, in addition to restricted, repetitive behaviour (APA, 2013). ASD is also common in people with DS: approximately 5 to 39% of people with ASD have DS and approximately 1-11% of persons with DS have ASD (Ersoy, Güler & Çetin, 2018; Godfrey et al., 2019). The occurrence of ASD in the general population by comparison is approximately 1.89% (Kim et al., 2011). This condition, according to Oxelgren et al. (2016), is generally more prevalent in people with DS than in many other groups of people with other ID.

Children with ASD may demonstrate difficulties with certain EF, such as self-regulation, affective sharing, cognitive flexibility and ToM (Zelazo, Burack, Benedetto & Frye, 1996), which are similar to many EF difficulties experienced by those with DS (Bieberich & Morgan, 2004). The next section discusses EF in greater detail, to gain a deeper understanding of what EF is, before delving into the EF of individuals with DS.

2.5 Executive Function: Definitions, Models and Assessments

EF begins to emerge during infancy (Miller & Marcovitch, 2015) and develops slowly until early adulthood (Anderson, 2002; Henry & Bettenay, 2010), although the rate of development depends on the trajectory of aspects of EF and individual differences. Research indicates that EF is atypical in people with DS. These difficulties can be found across a range of EFs (Rowe, Lavender & Turk, 2006), including working memory (WM) (Jarrold et al., 2000; Lanfranchi et al., 2010), planning (Fidler et al., 2005), shifting (Kopp, Krakow & Johnson, 1983; Rowe, Lavender & Turk, 2006), cognitive flexibility (Zelazo et al., 1996), self-
regulation (Bieberich & Morgan, 2004) and inhibition (Edgin, 2003; Rowe et al., 2006).

This section explores what EF is, models of EF and how it is commonly measured, as well as the pitfalls of this within research involving individuals with DS.

2.5.1 Definitions of Executive Functioning

The way EFs are defined and explained is fundamental to gaining a deeper understanding of human cognition, and for furthering knowledge and research in this field. From a historical perspective, the concept of EF has developed over time. The notion of a control mechanism was first debated in the 1840s by those trying to pinpoint the function of the frontal lobes, particularly the prefrontal cortex (Luria, 1966; Naglieri & Goldstein, 2014). Definitions from the 1970s onward predominantly developed from a neuroscience perspective, focusing on locating functions in specific brain areas, such as the prefrontal cortex. This can be seen in the first definition of EF, proposed by Pribram (1973), who argued that:

*The frontal cortex appears critically involved in implementing executive programs when these are necessary to maintain brain organization in the face of insufficient redundancy in input processing and in the outcomes of behaviour.*

(p.312)

Pribram’s (1973) definition continues to underpin current understanding of EF. For example, Best, Miller and Jones (2009, p.180) describe EF as an umbrella term for the ‘goal-oriented control functions of the prefrontal cortex’. Similarly, Hughes (2011) defines EF as a ‘complex cognitive construct encompassing the whole set of processes underlying these controlled goal-directed responses to novel or difficult situations, processes which are generally associated with the prefrontal cortex’ (p.313). For the purposes of this thesis, however, the
neurological localisation of EF function is not immediately important. Of greater interest is the notion of EF as a set of complex cognitive functions, which also relate closely to behaviour. Many definitions include reference to the variety of EFs, for example ‘a variety of different capacities’ (Stuss & Benson, 1986, p.272); ‘a collection of interrelated cognitive and ‘behavioural skills’ (Lezak, 1995, p.42); ‘a family of cognitive control processes’ (Friedman et al., 2007, p.893); and ‘a self-directed set of actions’ (Barkley, 2011, p.11).

Definitions also usually make reference to ‘levels’ of cognition, although these are not always precisely defined: for example, ‘EF coordinates two levels of cognition’ (Borkowski & Burke, 1996, p.241); a ‘directive system exerting regulatory control over the basic functions’ (Gioia & Isquith, 2004, p.139); ‘cognitive control processes that operate on lower-level processes’ (Friedman et al., 2007, p.893); and ‘the highest level of human functioning’ (Lezak, 1995, p.42). Finally, the idea of ‘goal-directedness’ is central to most definitions of EF. The purpose of EFs is seen as enabling ‘purposeful, goal-directed behaviour’ (Stuss & Benson, 1986, p.272), ‘goal-directed activity’ (Lezak, 1995, p.42); a ‘system in the service of reaching an intended goal’ (Gioia & Isquith, 2004, p.139); and ‘actions intended to alter a delayed (future) outcome (attain a goal for instance)’ (Barkley, 2011, p.11).

Thus, EF is seen as a set of higher-level, regulatory skills used to guide behaviour toward goals. Within this generally agreed notion of EFs, particular definitions highlight different EFs depending on the focus of the researchers. For example, Miyake et al. (2000) conducted seminal research that found evidence for three separate factors within EF: WM, inhibition and shifting, to which Lanfranchi, Jerman, Dal Pont, Alberti and Vianello (2010) also include ‘planning’. The three factors within Miyake et al.’s (2000) work has been adopted by many researchers investigating EFs since (e.g., Blair, 2016). Burgess and Simons (2005), discussing research in populations with neurogenetic disorders, describe
EFs as a cluster of top-down mental processes required in concentrating and paying attention. For this reason, Goldstein and Naglieri (2014) argue that ‘EF’ has come to be an umbrella term for various hypothetical processes within cognition, including WM, attention, inhibition, planning, self-regulation, self-monitoring and initiation, all of which are performed and controlled by prefrontal areas of the frontal lobes (Goldstein & Naglieri, 2014). These processes are discussed in more detail below, although it must be noted that there may be considerable overlap between them.

(i) Working Memory - WM is described as simultaneous storage and manipulation of information for tasks that are considered complex, such as learning, reasoning and mental arithmetic (Baddeley, 2003; Gathercole et al., 2004). Adams, Nguyen and Cowan (2018) define working memory as ‘a system of components that holds a limited amount of information temporarily in a heightened state of availability for use in ongoing processing’ (p. 341). There are various models of WM, such as the ‘modal model’ (Atkinson & Shiffrin, 1968, cited in Adams et al., 2018) and the embedded-processes model (Cowan, 1988; 1999; 2001, cited in Adams et al., 2018) and, perhaps most well-known of all, the working memory model from Baddeley and Hitch (1974; with further revision by Baddeley, 1986; 2000; 2003; 2010).

According to Baddeley’s (2003) model, the WM comprises the central executive, which acts as a filter to control attentional processes and direct information to one of three ‘slave’ systems: the episodic buffer, the visual spatial sketchpad and the phonological loop. Thus, WM can be divided into verbal WM (VWM) and visuospatial WM (VSWM) (Baddeley, 2003). The former temporarily stores information that can be verbalised (such as nameable objects, numbers, words and so on) within the phonological loop, maintained through subvocal repetition (Van Dun & Mariën, 2016). The latter deals with visuospatial information within the visuospatial sketchpad (Wang et al., 2018). The episodic buffer then acts as
a ‘back up’ store to connect the long-term memory and various components of WM; this feature was added later (Baddeley, 2000), given original shortcomings in the model (see Figure 2.1).

![Figure 2.1 Baddeley's Working Memory Model (Baddeley, 2003).](image)

The WM model therefore presents WM as a mental workspace in which information can be held and manipulated; in TD individuals, it has a span of 5-9 items at a time that can be increased by ‘chunking’ (combining) different pieces of information (Miller, 1956, cited in Cowan, 2015). Thus, it is a limited capacity store that is activated when using or thinking about specific information. Two key features that can be measured when assessing WM as contributing to EF are the processing of information and the storage of this coded information, often measured using complex span tasks.
There are other models of WM, such as Cowan’s embedded processes theory (Cowan, 1999, p. 62), in which he defines WM as ‘cognitive processes that are maintained in an unusually acceptable state’. This theory centres around a limited-capacity attentional focus that works across various domains of activated LTM. Cowan (2005) indicates in his research that the capacity of WM is four items (although these may include chunked items). However, this model appears to focus heavily on the link between (in Baddeley’s terminology) the central executive and episodic buffer, which limits it somewhat, and Cowan (2005) admits that the WM system, based on ‘activated LTM’, does not provide sufficient explanation for processes outside of attentional focus. Interestingly, Baddeley (2012) asserts that the differences between his WM model and that of Cowan (2005) lie predominantly within terminology and emphasis. Thus, Baddeley’s model was selected for this thesis as providing the most easily accessible, broader and more encompassing model of WM.

Other theories of WM considered for this thesis also include individual difference-based theories, given the difference in memory span between individuals. Some researchers focus on the abilities of some to capitalise on the gaps between the processing operations within a span task so as to prolong a fading memory trace (Barrouillet et al., 2004), whereas others emphasise individual differences in the ability to switch between different tasks involved in span (Towse et al., 2000) or focus on interference instead of decay (Saito & Miyake, 2004). Engle et al. (1999) emphasise the role of inhibitory processes within WM, which protect content within the memory from being disrupted. However, a theory of executive processing that is rooted almost entirely on inhibitory control was considered too narrow for the purposes of this thesis; whilst inhibitory control is deemed an important element of WM, individuals may also differ in other elements of attentional capacity. The individual difference-based theories therefore, whilst useful for highlighting the variable capacity of WM in different people, is still consistent with the idea of executive control within
the broader WM model proposed by Baddeley, albeit placing greater emphasis on inhibitory control. Most theorists do also accept that separate verbal and visual STM systems also contribute to WM (e.g., Alloway et al., 2006), which renders Baddeley’s model as more comprehensive (as it contains these elements).

Finally, computational models of WM were considered, which give a detailed account of WM alongside computer simulation; however, some of these models also include subsystems that resemble features of Baddeley’s WM model, including an auditory loop (Anderson et al., 1996) and sketchpad (Anderson et al., 2004). Yet the complexity of many of these models (e.g., Barnard, 1987; Oberauer, 2009), often developed by computer scientists, can make them difficult to apply. For this reason, computational models were rejected, as they were deemed too unnecessarily complex for the purposes of this thesis. Rather, Baddeley’s model appeared to contain sufficient features to comprehensively explain the WM, without becoming overly complicated.

(ii) Inhibition and Interference Control - This refers to the controlled, conscious suppression of prepotent responses (Miyake et al., 2000), involving the exertion of self-control over one’s behaviour to overcome extraneous stimuli in the environment, not only to maintain focus but also to retrieve information stored in the brain for update and manipulation (Will, Fidler & Daunhauer, 2014; Carlson & Moses, 2001). The term ‘self-regulation’ is often used to describe a specific kind of inhibition, which depends on self-awareness and the ability to evaluate oneself according to standards. Self-regulation is also dependent on fluctuations in willpower that can occur between and even within individuals (Baumeister, 2014). Similarly, Diamond (2013) argues that self-control/regulation is an element of inhibitory control that is associated with interference control, overcoming temptations and not making decisions prematurely. Zelazo, Blair and Willoughby (2017) share a similar view, as they define self-regulation as a
range of ways that an individual can adjust his or her behaviour using effortful EF skills. Miller, Giesbrecht, Müller, McInerney and Kerns (2012), describing their work with individuals with DS, refer to avoiding distraction within the context of inhibition and self-regulation as the conscious effort to persist in the effective completion of tasks; therefore, attention is critical for inhibition and self-regulation. Notably, avoiding distraction is categorised as an essential component of EF, since it determines the pace of the completion of tasks, especially in relation to challenging tasks in difficult conditions (Miller et al., 2012).

These definitions of WM and inhibition illustrate the potential for considerable overlap between the two functions. For example, in WM there is a need to ignore irrelevant information and maintain or manipulate the required information during operations such as calculation. Diamond (2013) supports the idea of overlap between functions by demonstrating that, as the inhibitory control function deteriorates with age, WM also becomes weaker, indicating that cognitive functioning in general can decline, which affects various EFs simultaneously. Similarly, it could be argued that there is considerable overlap between the EFs of WM and attention. WM requires selective attention to keep required information in the mind (Diamond, 2013) and, as Schworer et al. (2020) found, attention skills in early infancy can predict later EF performance, including WM. WM can therefore be seen not as a unitary EF, but as ‘complex’, as defined above either by function (Daunhauer & Fidler, 2013; Krasny-Pacini et al., 2017) or as a skill that depends on several EFs. This complexity or overlap brings into question whether WM (and, moreover, EFs more generally) can be deemed a discrete indivisible mental function, or a convenient construct.

(iii) Mental Flexibility, Set Shifting or Switching - Mental flexibility, which is also described as ‘cognitive flexibility’, switching or shifting is associated with the ability to view something in varied ways (Diamond, 2013; Zelazo, Blair &
Willoughby, 2017). This requires skill in altering one’s cognitive set on demand, which involves responsively changing/alternating between strategies, or abandoning a strategy if it is not effective (Goldstein & Naglieri, 2014). According to O’Hare and Sowell (2008), ‘set shifting’ connotes the ability of an individual to effectively coordinate various tasks and states of mind simultaneously. Moreover, it could be said that task switching involves adapting to a different set of rules when their current set is not working in the present environmental conditions (Manoach, 2009).

The three core EFs above also affect a range of other cognitive skills, including planning/organising (being able to organise cognition in time and space, which is critical in circumstances that are goal-oriented (Owen, 1997)) and problem-solving (working in different phases to comprehend a problem and be able to plan for and find a solution through the appropriate selection and ordering of effective strategies (Zelazo et al., 1997)).

Given the complexity of EF, there are still some limitations due to the vagueness in EF definitions, primarily due to the overlap between functions and the multitude of skills that stem from these functions. For instance, two functions may be assessed with the same tools (discussed in due course). As a result, some researchers such as Barkley (2012) argue that different definitions of EF mean it is insufficient to consider EF as an umbrella term, given the lack of consensus regarding an operational definition of EF. Moreover, Barkley (2012) argues that there is no clear understanding about how the separate, ‘core’ functions are related to each other, claiming that the absence of clear explanation means the absence of clear EF theory. However, as has been noted, there are some specific indications about the relationship/overlap between some EFs to one another (Diamond, 2013), including between WM, inhibition and attention.
EF is therefore not a unitary cognitive process; rather, this construct may well consist of various interlinked higher order cognitive skills. This was found by Miyake et al. (2000) in 137 undergraduates that performed a variety of tasks designed to measure different EFs (shifting, inhibition and ‘updating’ (also known as WM)). This included tests such as the Tower of Hanoi (TOH), Wisconsin Card Sorting Test (WCST), operation span, random number generation (RNG), and dual tasking. Using factor analysis, the researchers concluded that whilst there was a moderate correlation between the three EFs in question, they were clearly separable. This was also found by Friedman et al. (2008) who, using both 316 monozygotic and 216 dizygotic twin pairs and their families, found that EFs are correlated due to the presence of a largely heritable (99%) common factor beyond general IQ, yet also separable as a result of other genetic influences that are specific to different EFs. However, within the literature, definitions of EFs are often generalised and broad, which attempt to describe the construct’s multiplicity whilst also acknowledging the umbrella construct. Yet it is clear that common themes arise from the literature, presenting some degree of agreement regarding the skillset that EF encapsulates (Anderson, Jacobs & Anderson, 2008). Moreover, there are a range of models of EF that seek to describe this construct.

2.5.2 Models of Executive Functioning

There has been increasing interest in the concept of EF since it was first proposed by Pribram (1973), and various theoretical models of EF have since been developed, influencing research and clinical practice (Anderson, Jacobs & Anderson, 2008). The development of these EF models has been informed to a considerable extent by the study of individuals with frontal lobe damage (e.g., Vinken & Bruyn, 1969), which of course carries its own limitations, given that this damage may have affected more than just EF, but cognitive functioning more generally. Notably, Luria (1973) described the disorganisation and inability to develop strategies to control behaviour in people with pre-frontal cortex (PFC)
injuries. This came to be known initially as ‘dysexecutive syndrome’. Yet in tests of fundamental cognitive processes, such as memory or reasoning, little atypicality was found, leading researchers to postulate a separate control system that coordinated other cognitive abilities (Goldstein & Naglieri, 2014). This has been borne out by more recent functional magnetic resonance imaging (fMRI) studies (Nowrangi et al., 2014), which have found that two parts of the prefrontal cortex, the anterior cingulate cortex and the dorsolateral prefrontal cortex, are critical to tasks thought to be sensitive to EF (Goldstein & Naglieri, 2014). This does indeed indicate that there may be a separate control system that coordinates EF. In this section, different models are presented chronologically in a review largely informed by Goldstein and Naglieri (2014) and Barkley (2012). It must be noted, however, that the very fact that so many models have been proposed is testament to the major issues underpinning the definition of EF itself (Kluwe-Schiavon et al., 2017).

The foundation of many modern theories of EF relies on a two-part theory of human cognition, which asserts that cognitive processes are either automatic or controlled processes, depending on the amount of ‘processing power’, effort and attention that is required by a process (Schmidt & Lee, 2011). Such theories originated with Broadbent’s (1958) automatic or controlled processes model, also called the filter model. In this model, a filter determined the information that is going to be consciously thought about (Broadbent, 1958). If other stimuli are competing for this awareness, the filter prioritises them in terms of the conscious attention that they should be allocated, with part of the prioritisation ‘criteria’ being their relevance to current tasks or goals (Barkley, 2011). Without the filter, information would overload the brain’s conscious information processing system (Broadbent, 1958; Driver, 2001). Broadbent’s model postulates that system components, such as the ‘sensory store’ and the ‘sensory filter’, are involved in the processing of stimuli at the pre-attentive level. This processing can be shown diagrammatically, with parallel lines representing the processing of
information, which narrow to a ‘bottleneck’ at the point at which the filter selects information for conscious attention (Shiffrin & Schneider, 1977). This gives Broadbent’s model the name by which it is often known: the ‘bottleneck theory’ (Driver, 2001). Yet whilst the bottleneck theory and other theories like it (e.g., the three-component model from Atkinson and Shiffrin (1971)) could distinguish between automatic and controlled cognitive processes, they were not able to fully explain how information could be consciously, deliberately inhibited or selected during attentional tasks.

This criticism led to Posner and Snyder (1975) developing the ‘cognitive control’ model, to explain individuals’ abilities in managing their thoughts and emotions and to adapt behaviour according to context and needs. Focusing on the bottleneck theory, they examined the role of attention during specific higher-level tasks, claiming that emotions and thoughts play a significant role in deliberatively managing cognitive control (Rueda, Posner & Rothbart, 2004). Posner and Snyder (1975) asserted that cognitive control is used to override automatic responses, both inhibiting automatic responses and selecting stimuli for conscious attention, enabling behaviour to be adapted to the situation and to the goals of the individual (Checa, Rodriguez-Bailon & Rueda, 2008).

A criticism of the cognitive control model came from Shiffrin and Schneider (1977), who postulated that the allocating of conscious attention to different stimuli was not innate - the individual could learn to assign stimuli to either unconscious processing or conscious attention. This is important given the fact that attention is limited, meaning that certain stimuli must be favoured over others. This ‘Controlled Processes’ theory proposed that in automatic processing, there is activation of learned sequences of patterns, in contrast to the temporary activation of a sequence of elements that require conscious attention (Shiffrin & Schneider, 1977). Automatic processes are effortless, rapid and unavailable to consciousness. Controlled processes are slow and effortful
yet completely conscious. Thus, the Controlled Processes theory asserts that through practice or training, individuals can develop the permanent neural connections required to transfer sequences of behaviour from needing conscious attention to being automatic (Shiffrin & Schneider, 1977).

Yet it is not always possible to learn how to respond to situations when the circumstances presented are novel and unique. A model of EF, based on the dual-processing role of EF, was developed by Norman and Shallice (1983) and Shallice (1988; 2002) to address this issue. These authors formulated a model of the executive system entitled the ‘contention scheduling/supervisory attentional system’ (SAS). Contention scheduling is the process of controlling the mediator of inhibition in which one selects an action to be performed rather than competing possible actions. The SAS is a mediator used in nonroutine or novel situations in which inhibition is required for decisions to be made in such novel situations (Shallice, 1988; 2002). If there are challenges or delays within this supervisory attentional system, disorders within EF may arise (e.g., disinhibition) (Shallice, 2002). The dual-processing models described above have been integral in including the prefrontal cortex as the primary brain structure involved in cognitive control, responsible for managing/regulating automatic behaviours (Norman & Shallice, 1983).

The delineation between controlled and automatic processes underpinning many EF models, has arguably promoted a hierarchical, categorical approach to understanding and explaining EF, wherein cognitive functions are seen as independent yet related components, playing specific roles in the journey from stimulus to behavioural response (Kluwe-Schiavon et al., 2017). As an example, the central executive model of WM (Baddeley, 1996; 2012), which proposes a coherent, unified ‘central executive’ system, has three distinct ‘slave’ systems: the phonological loop, the visuospatial sketchpad, and the episodic buffer (this is also the model of WM discussed earlier). Below the level of the central
executive, the model proposed specific functions, such as time-sharing, selective attention, temporary activation of LTM and switching of retrieval plans (Baddeley, 1986). This demonstrates the hierarchical, categorical nature of EFs, especially the WM, which this model explains in greater detail. Yet even though Baddeley (2012) describes the central executive as a ‘homunculus’, suggesting that it was fraught with issues for which there was no explanation, it is still seen as the pinnacle or starting point in a hierarchy of functions that make up EF. Miller and Cohen’s (2001) integrative model of EF for example focuses on cognitive control, placing particular emphasis on tasks that represent goal maintenance, describing EF as an umbrella of cognitive processes under goal-directed behaviour. Here again, EF is a top-down system coordinating various motor and sensory processing domains, often situated in the prefrontal cortex (Miller & Cohen, 2001).

Some researchers (e.g., Banich, 2009) point to the potentially flawed nature of many other multicomponent models of EF that perceive the functions as separate categories. For instance, Lezak’s (1995) four-component model perhaps oversimplifies EF by dividing EFs into the different components that allow a person to enact self-serving, purposeful, autonomous behaviour, including planning, volition, action and effective performance. Another example is Diamond’s (2013) description of the three core functions of EF, presented in the previous section (inhibition and selective attention, WM and cognitive flexibility). These three functions are, of course, established based on the assumption of a general agreement these are three of the key components of EF. Some research, such as that of Miyake et al. (2000) for example, does assert that EFs may be correlated with each other, yet are clearly separable. Miyake et al.’s (2000) study involved 137 college students who completed tasks measuring shifting, updating and inhibition, finding that the three functions contributed independently to performance on executive tasks. The researchers
concluded that future research should acknowledge both the unity and diversity of EF.

A further development in EF models has been the proposition of various ‘hot’ and ‘cold’ cognitive information processing systems (Sahlin, Wallin & Persson, 2010), perhaps because they enable a greater understanding of otherwise complex processing systems in the human body. For example, Zelazo and Müller (2011) described cold EFs as those relatively free of affect or emotional ‘charge’, essential to cognitive tasks involving WM and planning (Zelazo & Müller, 2011; Zelazo & Carlson, 2012). By contrast, hot EFs are those cognitive abilities considered necessary for decision-making and goal-setting, which are more motivationally or emotionally laden and associated with reward, reinforcement and motivation (Zelazo & Müller, 2011). Other researchers also utilise this idea of two systems; system 1 requires stronger activation of subcortical structures and is specifically unconscious, rapid, automatic and emotional; system 2 on the other hand requires greater activation of cortical structures, is more rational, deliberative, conscious, slow and primarily in charge of serial information processing (Noël et al., 2013). The emotional versus rational (hot versus cold) divide has received much support in various studies, with some relying on this model to explain behaviours such as risk-taking in adolescence (Gladwin et al., 2011). ‘Hot’ or emotional/affective, bottom-up EFs or systems are perceived as maturing more quickly than cold, top-down control systems, a theory used to explain impulsive behaviours and lack of self-regulation (Benningfield et al., 2014), lack of risk aversion (Pripfl et al., 2013) and lack of self-reflection (Kluwe-Schiavon et al., 2016). Yet these models have received criticism from some recent authors for being too simplistic (e.g., Moors & De Houwer, 2006), although dual-processing models of EF (including automatic vs. controlled and ‘hot’ vs. ‘cool’) have arguably been relied on to aid scientific enquiry and enable researchers to grasp such a complex system (and the resulting complex behaviours).
Whilst it is beyond the scope of this thesis to explore emerging neuroscientific research in any depth, it appears to point to an integrated EF circuit, involving the prefrontal cortex, neuroendocrine and autonomic systems, suggesting that earlier EF models may be simplistic in their dual-processing, hierarchical approach. Moreover, most models of EF apply to TD individuals within the general population. In other areas of research, models of EF have been developed for or adapted to specific populations, such as those with ADHD (Barkley, 1997) or ASD (Demetriou et al., 2019). There have been very few models of EF developed specifically for populations with DS, however, although it is noteworthy that Zelazo and Müller’s (2011) model was used in DS research by Lee et al. (2015), in a study that found that people with DS demonstrate more difficulties in ‘cool’ EFs than ‘hot’ EFs. The ‘cool’ and ‘hot’ model therefore appears to be useful for the present research, therefore. Most recently, Sabat et al. (2020) generated a model of EF taking the three core components (WM, inhibition and flexibility) and relating them to aspects of adaptive behaviour. They found moderate teacher- and parent-reported correlations between these EFs and adaptive behaviour skills. This indicates that adaptive behaviour could be considered within EF models in a way that has not yet currently been addressed. Sabat et al.’s (2020) article is therefore useful as illuminates how models of EF can be related to behaviour, rendering them practically useful, which has relevance for this thesis.

Yet even these researchers state that their model linking core EFs to adaptive behaviour domains was generated based on previous research involving participants from TD and other disability groups, not individuals with DS. Some researchers however have indicated that EF may be linked to IQ, which suggests that similar brain areas are involved in each of these aspects of development (EF and cognitive functioning as measured by IQ). For example, a study by Campbell et al. (2013) examined the difference in mean scores
between verbal mental age (VMA) and performance mental age (PMA), and the association of these with cognitive flexibility for 22 children, adolescents and young adults with DS (mean CA was 15.2 years) who completed the Flexible Item Selection Task. Campbell et al. (2013) found that VMA was more significantly associated with cognitive flexibility than PMA, best accounting for individuals’ performance in this skill, thus highlighting the pivotal role of verbal abilities for cognitive flexibility in individuals with DS (although 21 out of the 22 participants were female and there may be gender differences that are unaccounted for in this study). Yet simply linking EF to IQ is rudimentary and inaccurate, given the potentially different developmental trajectories for different EFs across the lifespan (Best & Miller, 2010).

2.5.3 Developmental Trajectories of Executive Functioning

Developmental trajectories for both TD populations and those with varying disorders and disabilities are gaining ground in the research literature (Thomas et al., 2009). This section explains and argues for a developmental, trajectorial approach, before explaining the (limited) literature surrounding the trajectory of EF development in TD groups and populations with DS.

2.5.3.1 A Developmental, Trajectorial Approach

There appear to be two broad approaches in the literature regarding the perception of those with developmental disabilities. These individuals are perceived by difference theorists to be qualitatively ‘different’ to others in certain areas. This is in contrast to developmental theorists, who use growth models to ascertain how far individuals with developmental disorders may be delayed in acquiring certain skills (Thomas et al., 2009). This thesis adopts a developmental approach, which emphasises that there may be delays in different areas in the cognitive functioning in those with DS compared to their typically developing peers of the same chronological age (CA) (Thomas et al., 2009).
The distinction between a developmental and difference approach has also led to discussions regarding developmental trajectories; specifically, the methods used to reflect this change, which has moved from participant matching to trajectory studies. Within the research literature, participant matching has been one of the main indicators of developmental difficulties. Matching can involve individuals being matched to both CA and mental age (MA) control groups; if individuals with a developmental disability show difficulties on a task and/or perform less well compared to the CA-matched group, but their performance is consistent with the MA-matched group, these individuals are perceived to be developmental delayed on this task or ability. This could be due to delayed onset of ability, slow rate of ability acquisition, or both; yet individuals with a disability are expected to show a similar general qualitative pattern of development (including developmental milestones) as TD populations. However, if the ‘disorder group’ exhibits challenges completing a task compared to both CA- and MA-matched groups, within a difference approach, this is understood as atypicality or developmental deviance (Caplan, Neece & Baker, 2015). However, within a developmental approach, the difficulties experienced by the disorder group are perceived as being qualitatively different to that of a TD group, as they may demonstrate non-linearity in their development trajectory or their rate of ability acquisition may begin to prematurely slow and level off.

The matching methodology has been criticised by some; Thomas et al. (2009) point out its shortcomings, indicating that it is liable to floor effects (the measure no longer accurately captures ability), is theory-driven, often involves group comparisons (which can miss many nuances, including instances of TD individuals scoring significantly below average expectations for their age, and profiles of difficulties experienced by those with developmental disabilities). As a result, the concept of developmental trajectories has gained increasing attention in the research literature over previous decades, in a bid to place development
at the centre of descriptions and explanations of developmental disabilities (Karmiloff-Smith, 1998; Karmiloff-Smith et al., 2004; Rice, 2004; Rice, Warren & Betz, 2005; Singer Harris et al., 1997; Thomas et al., 2001, 2006). This is because phenotypes linked to different developmental disabilities are often not detectable at birth but develop gradually and can change with age (as found by Paterson et al., 1999). Therefore, the trajectorial developmental approach is beneficial as it shifts the focus towards change overtime and ‘discourages static interpretations of developmental deficits as if they represented focal damage to preformed systems’ (Thomas et al., 2009, p. 338). This approach aims to link performance with age on specific experimental tasks before investigating if and how far a function differs between TD groups and those with developmental disabilities (Thomas et al., 2009). It also seeks to ascertain the relationship between various experimental tasks, establishing the extent to which task performance in one domain/area relates to and predicts performance on another task across development, given that behaviour and cognition change with age (Thomas et al., 2009). In addition, it is not assumed that a delay in one area necessitates a delay in other (or all) cognitive domains (Karmiloff-Smith et al., 2003).

Age is a critical feature when it comes to understanding the characteristics of a developmental disorder/disability, with performance and outcomes on a range of factors remaining qualitatively different to an adult-acquired deficit (Levy, 2018). This is often because any cognitive or behavioural deficit present from birth is likely to be ‘the outcome of an adaptive, developmental process likely to be characterised by features such as interactivity, compensation, and redundancy’ (Thomas et al., 2009, p. 318), as evidenced in the literature (Bishop, 1997; Karmiloff-Smith, 1997, 1998; Thomas, 2007, cited in Thomas et al., 2009). Thus, Thomas et al. (2009) highlights that the delay versus difference approach is somewhat reductionist; including the notion of change with age when considering developmental disability, richer ways of understanding
characterising developmental change can be found. A matching approach may promote a simplistic contrast between the terms ‘deviance’ and ‘delay’; however, a developmental trajectorial approach asserts there are seven ways in which a population with developmental disabilities can differ to a control/TD group: (a) delayed onset, (b) slowed rate, (c) delayed onset + slowed rate, (d) nonlinear, (e) premature asymptote, (f ) zero trajectory, and (g) no systematic relationship with age (Thomas et al., 2009, p. 345). Thus, the terminology and approach used within this thesis reflects the theory underpinning the developmental, trajectorial approach more generally.

2.5.3.2 Executive Functioning Development in Typical Development and Populations with Down Syndrome

In TD populations there appears to be rapid development of most EFs in the early years, particularly during the ages of 3-6-years-old (Shonkoff et al., 2011). Global EF development may begin to increase less sharply around the age of five, and level off in early adulthood. A review of the research surrounding the development trajectories of inhibition, WM and cognitive flexibility (shifting) (Best & Miller, 2010) revealed that, despite some inconsistencies, in general, inhibition shows a marked improvement over the preschool years and changes less later on. Shifting and WM, however, appear to improve more gradually in a linear fashion throughout development to adulthood, which supports Miyake’s et al. (2000) assertion that these three components of EF are interrelated but dissociable (cited in Best & Miller, 2010).

Poon (2018) looked specifically into the development trajectory of ‘hot’ and ‘cool’ EFs during adolescence (from 12-17) in TD individuals and revealed that abilities in cool EFs tended to continue to increase with age, whilst hot EFs show a bell-shaped development. Poon (2018) asked 136 TD adolescents to undertake four cool EF tasks to measure WM, attentional control, cognitive
flexibility, goal-setting and planning, and inhibition (including the Backward Digit Span, Contingency Naming Test, Stockings of Cambridge, and Stroop Color and Word Test), and one hot task to measure both reward-related decision-making and delay discounting (Cambridge Gambling Task). The results for the WM test (Backward Digit Span) indicated that processing speed and WM functioning increases in a linear fashion, with a spurt between the ages of 14 and 15. By comparison, hot EFs were shown to peak between the ages of 14-15, before declining (Poon, 2018). Performance in delay aversion tests for example peaked at age 15, before declining rapidly over the next year.

There is a paucity of literature that specifically compares the EF development of those with DS by CA, perhaps given the vast individual differences in abilities in individuals with DS (Fidler et al., 2018). Most studies comparing those with DS to either CA- or MA-matched controls utilises a matched groups approach, which has its shortcomings. However, from various studies utilising this approach, it may be possible to deduce some basic developmental trajectories in EF development in those with DS. In fact, the development of EF in individuals with DS appears to follow a similar trajectory to that of TD individuals in some studies. A cross-sectional study by Lee et al. (2015), comparing EF performance on the BRIEF in 30 participants with DS (age range=5-18) and 30 TD participants matched on CA, and later, in a second study, 85 individuals with DS and 43 TD individuals aged between 4-24, provided support for similar age-EF relations in both the TD and DS groups. In almost all areas of EF, children and adolescents with DS were found to have a similar developmental trajectory (similar rate of development but with delayed onset) to TD children and adolescents. This supports the developmental stability hypothesis.

However, Lee et al. (2015) reported that the results for the DS group on the inhibit scale revealed greater skill acquisition, concluding that throughout mid/late adulthood, difficulties in inhibition would continue to reduce and
eventually be similar to that of the TD group. This supports the notion of developmental ‘delay’, instead of atypicality or deviance. In addition, skills in organisation of materials improved sharply for participants with DS, comparative to TD populations, perhaps suggesting that this is one skill that those with DS are able to improve on significantly over adolescence – more so than TD populations. Naturally, this is simply the findings of one study, and given its use of the BRIEF (a self-report measure used by teachers and parents, instead of using alternate—or complementary—tasks to measure EF) and the small sample sizes, it is unclear how far these findings are generalisable to other DS and TD child and adolescent populations. Moreover, this was a cross-sectional study involving different individuals of specific ages, and further longitudinal research is required to examine the EF developmental trajectory in individuals with DS to ascertain how this compares to TD individuals over time. However, in so far as Lee et al.’s (2015) study assessed EFs over a wide age range in childhood and adolescence and directly compared TD individuals to those with DS, it provides some evidence of a similar developmental trajectory that is not contradicted (to the best of this researcher’s knowledge) within the current literature base.

However, Lee et al. (2015) presented linear trends in development across participant ages. Another study using the BRIEF and BRIEF-P to explore EF profiles in DS cross-sectionally by age (2-35 years) was undertaken by Loveall et al. (2017). This study compared the scores from 112 individuals with DS with normative data and presented both linear and curvilinear trends in EF development. Their study revealed that, for 2-5-year-olds, Emergent Metacognition Index (EMI) only scored significantly higher (worse performance), and, within that, WM (not planning/organising (PO)) was significantly delayed, with PO and inhibition being marginally significant and shift and emotional control not being significantly different to normative scores. For those aged 6-18, results revealed that global EF scores (GEC) were significantly delayed. The
areas of greatest weakness were WM, monitor, shift, and POS; inhibit and initiate, whereas inhibit and initiate were marginally significant (moderate difficulty in those with DS). However, emotional control and organisation of materials were not significantly different to normative data.

When assessing EFs by CA (using raw scores on the BRIEF/-P), Loveall et al. (2017) found that in linear terms, most EFs followed a similar pattern to Lee et al. (2015), remaining consistent across the age range. A curvilinear analysis noted significant trends for WM, PO, shift and inhibit. In all these domains, the scores increased (greater difficulty) between preschool (age 4-5) and middle childhood, before decreasing until the early/mid-30’s, when they were observed to increase once again (quadratic trend). This fluctuating of skill particularly applied to shift; Loveall et al. (2017) found that those with DS encountered significantly greater difficulties in shifting in late childhood (compared to normative data), indicating skills in this area slow in middle childhood, but ‘recovered’ (resumed pace) during adolescence, before again declining in early/mid-30s. The authors note that this finding indicates that shifting in particular may have different growth rates at different points during childhood/adolescence, although this has not yet been corroborated through replication.

A review of the research surrounding WM development in particular in individuals with DS compared to their TD counterparts (Fidler et al., 2018) indicated that one of the main issues of research in this area is the wide age range of participants included. Often, mean EF performances have been reported for groups, which means that in some studies, school-age children have been grouped with adolescents and young adults (e.g., Carney et al., 2013; Costanzo et al., 2013; Lanfranchi et al., 2012). Whilst a wide age range may be justified given the wide differences in developmental status of individuals with DS (e.g., in Lanfranchi et al.’s (2009a) study, the sample of 8-19-year-olds
with DS all had a MA of between 4-7-years-old, indicating a far narrower window of development), it is possible that such wide groupings can mean age-related features, areas of vulnerability and important developmental trends can be missed (Fidler et al., 2018).

General trends have emerged in terms of EF performance in studies with a broad CA range; for example, when reviewing EF literature regarding WM, Pennington et al. (2003) found that performance of individuals with DS between 11-19 years was similar to TD children matched for MA of 3-6 years, suggesting performances that paralleled anticipated performances based on developmental status. This finding, however, has not been replicated. In fact, Lanfranchi et al. (2009a; 2012) found that when comparing groups of individuals with DS from 8-23-years-old with 4-6-year-old TD children matched for MA, WM skills appeared to be significantly poorer in the DS group.

This could indicate that, whilst the performance in WM for some children and adolescents with DS is similar to that of children between 3-6-years-old, there may be a more pronounced weakness in WM than may be expected based on MA or general developmental status in DS. It could also be due to the matching of those with DS with TD children based on MA, which generates an ‘experience’ effect when compared to very young children; older individuals with DS may have more overall experience, which could mean they perform similarly to MA-matched younger children (again indicating the issues with the matching approach, as highlighted by Thomas et al. (2009)). Other studies that have incorporated narrower CA groupings to explore WM (e.g., primary school aged children with DS (5-11-year-olds) (Daunhauer et al., 2014; Will et al., 2016); adolescents (11-18-years-old) (Lanfranchi et al., 2010; Pennington et al., 2003); and adults over 30-years-old (Ball et al., 2008)) all indicate similar patterns of WM performance in those with DS as reported in studies involving broader age
ranges previously discussed, indicating substantial difficulties in WM as well as cool EFs more generally.

It was of interest in the current research to explore how far the EF development trajectories of both TD groups and populations with DS are reflected in the literature. An extensive search resulted in a significant dearth of information regarding this, especially given that most studies utilise the matching method to compare these groups, involving wide age ranges and mean CAs and MAs, as well as using different tools to compare the same EF (or perhaps they look at different elements of the same EF, such as sustained or selective attention). All of these factors render discerning the development trajectories extremely difficult. What scant information there is presented in Table 2.1.
Table 2.1: Executive Functions in Typical Development and Down Syndrome

<table>
<thead>
<tr>
<th>Executive Function</th>
<th>Pattern of Development in TD (all ages are approximate)</th>
<th>Pattern of Development in DS (all ages are approximate)</th>
<th>Comparison with CA-Matched TD Populations</th>
</tr>
</thead>
</table>
| Emotional Control  | 2+: Development appears to increase in a linear fashion
14-15: Declining rate of development until late adolescence
18+ Development appears to increase in a linear fashion | 2-adulthood: Development appears to increase in a linear fashion similar to TD.
In line with MA. | Least delayed (relative strength) |
| Attention          | 3+: Rapid development.
Development trajectory pattern unknown. | 3+: Significantly slower development than TD.
Mixed findings; some studies suggests attention is in line with MA. | Delayed |
| Self-Regulation    | 2+: Development appears to increase in a linear fashion.
14-15: Declining rate of abilities until late adolescence.
18+ Development appears to increase in a linear fashion. | Mixed findings; some studies suggests self-regulation is below MA.
Development trajectory pattern unknown. | Delayed |
| Cognitive Flexibility | 2-4: Rapid increase in skill  
4-15: Steady linear increase  
15+: Performance reaches adult levels | 2-9/10: Development increases as sharply as in TD  
9/10: Development rate slows substantially  
13: Development begins to recover and increase  
Early 30s: Development begins to slow. | Delayed |
|---------------------|-------------------------------------------------|-------------------------------------------------|--------|
| Inhibition          | 3-5: Rapid development.  
5-8: Skills improve but more slowly (linear)  
8+: Skills improve but more slowly still (modest linear improvement)  
30-40+: Skills begin to decline. | 3-5: Rapid development (only slightly delayed compared to TD groups)  
5+: Development slows (significantly greater difficulties upon reaching late childhood).  
Development trajectory pattern unknown. | Delayed |
| PO                  | 10-30: Linear increase in skill  
30-70: Skills begin to decline  
70+: Small increase in skill | Infancy+: Development falls significantly below MA  
Development trajectory pattern unknown. | Substantially delayed |
| WM                  | 3-6: Rapid development  
6-14 Linear development  
14/15: spurt in development  
15-early adulthood: Linear development  
30-60+: Abilities begin to decline | 2+: Development significantly delayed from infancy and remains so into adulthood.  
Development trajectory pattern unknown. | Most substantially delayed |

Note: Numbers refer to CA. Sources: Breckenridge et al., 2013; Brown et al., 2003; Cuskelly et al. 2003, 2016; Lanfranchi et al., 2010; Lee et al., 2011; Loveall et al., 2017; Lee et al., 2015; Molina & Perez, 1993; Shalev et al., 2019.
Table 2.1 gives an overview of the general consensus within the literature regarding the least to the most delayed EFs in those with DS, although a more thorough investigation in the form of a systematic review of all the literature comparing EFs in populations with DS and TD populations is required to confirm this. It is also important to indicate where there may be a delay versus a difficulty/weakness in EF ability; a ‘delay’ may imply that the individual may ‘catch up’ with their TD peers, although this may not be the case. Thus, terminology must be selected carefully when reviewing studies; a systematic review of studies his nature is presented in chapter three of this study.

Whilst a general consensus appears to emerge from the literature surrounding EF development in those with DS, the issues and challenges pertaining to understanding the specific age-related developmental trajectories of EFs in populations with DS is complicated by the multiple issues surrounding task purity and validity. Some studies, e.g., Loveall et al. (2017) and Lee et al. (2015) have used the BRIEF/BRIEF-P to ascertain EF skill development. However, this parent-reported form does not necessarily indicate EF ability – it may simply indicate parents’ views of how their child with DS responds to certain tasks, which in turn could reflect wider contextual factors, such as the challenges encountered when starting school. This could be why, as Loveall et al. (2017) discuss, skills in certain EF domains appear to develop less slowly or even decrease in middle/late childhood. This also reflects the wider issues surrounding EF measurement.

2.5.4 Measuring Executive Functioning

The issue of EF assessment has increasingly been debated by researchers in all fields, including those the field of cognitive disability, especially given the varied definitions and conceptualisations of EF. There are a variety of EF assessment tools and tasks that can be used in studies of populations with DS. Indeed, studies often make use of multiple EF tasks to assess children and
adolescents with DS (Lanfranchi et al., 2010). These tasks include the Go/No Go task (Purser et al., 2015), the A-not-B task (Roberts & Richmond, 2015), the Tower of London task (Lanfranchi et al., 2010), and the Stroop test (Lobaugh et al., 2001), as well as parent/teacher questionnaire measures, such as the Behaviour Rating Inventory of Executive Function (BRIEF) and its preschool version (BRIEF-P) (Daunhauer et al., 2014; Lee et al., 2011; Pritchard et al., 2015; Memisevic & Sinanovic, 2014). It is generally accepted that the tasks have a high degree of reliability and validity, as reported in these studies, and their diversity allows researchers to assess EFs from various perspectives. In addition, most of these assessments have been designed to minimise effort and disruption for both researchers and children by assessing multiple functions at one time (d’Ardhuy et al., 2015). There are, however, still some concerns about these tests and their applicability to the study of populations with DS.

One of these concerns is the complexity of EF assessments, as there are several different tools to assess specific aspects of EF (Henry & Bettenay, 2010). A researcher can either use multiple tests to assess multiple EFs, or they can use a combination of subtests focused on a particular EF (Goldstein & Naglieri, 2014). This could be seen as a positive given that one of the problems of EF tests that assess more than one function is that they are often ill-adapted to isolating individual EFs. In the Tower of London task, for example, performing well involves skills in planning, but planning also requires WM; yet the test does not allow for these abilities to be assessed independently of each other (Larochette et al., 2009). Moreover, to negate ceiling effects, for instance, researchers may utilise complex EF tasks that may tap into numerous EFs and necessitate the coordination of various processes (e.g., Asato et al., 2006), despite the researchers often classifying tasks into a single cognitive construct. As mentioned, the Tower of London/Hanoi tests have either been described as assessing WM, planning or inhibition, depending on the study’s purposes (e.g., D’Antuono et al., 2017; Huizinga et al., 2006; Nitschke et al., 2017). The fact that
the same task can measure multiple EFs, or different tasks measuring the same 
EF can achieve different results from children of the same MA and CA indicates 
the ongoing need for reliable, valid EF measures.

Another concern highlighted by researchers, with assessments of EF in general 
(Barkley, 2012) and their use in DS in particular (Bevins & Hurse, 2016), is that 
both the identity of the person conducting the assessment and the context in 
which the assessment is conducted could plausibly be expected to influence the 
test results. For example, differences in test/assessment results could reflect the 
different contexts in which the tests were conducted. For example, there may be 
a greater focus on intellectual considerations in a school environment, where 
WM may for instance be seen as fundamental to learning (Alloway & Alloway, 
2010), while emotional considerations might have more influence when the test 
is conducted in a home environment. Similarly, the BRIEF or BRIEF-P, which 
can be used by either parents or teachers, might yield different scores when the 
tests are conducted by parents or teachers, because the person conducting the 
test will focus more on what they are most familiar with. In addition, parents and 
teachers might view the child with a different kind/level of emotional 
involvement, and this could affect their evaluation of the child.

Another issue is that the variation in results from assessments could be due to 
differing perceptions of the Likert scale form used, for example, in the BRIEF. 
With this scale, one person might perceive 2 out of 3 as medium, while others 
could see it as high, creating variation in test results as there is no control 
regarding how people should interpret each number (Bishop & Herron, 2015). 
This limitation is discussed in more detail in chapter four (the methodology 
chapter for Study 2).

Another concern is that the child's physical, linguistic and memory difficulties 
might impact test performance, meaning that it may be difficult to tell whether
test results reflect EF difficulties or something else. For example, sometimes children may gain a low score in EF tests not because they have weaknesses in EF, but because they have hearing impairment, as is common amongst children with DS (Weijerman & De Winter, 2010). Such hearing difficulties could prevent them from hearing and understanding assessment instructions or prevent them from correctly repeating the required verbal information in WM tasks. Similar considerations would apply to EF tests involving linguistic abilities in people with DS, as language development is typically one of the main areas of difficulty for individuals with DS (Togram, 2015). Thus, if an assessment for VWM required the child to repeat some sentences, the child might repeat them incorrectly as a result of pronunciation difficulties, rather than because of WM difficulties.

Moreover, visual differences in those with DS could also affect the results of EF assessments. People with DS have often been found to struggle with certain tasks that contain a visual element (Weijerman & De Winter, 2010), despite the relative strength of the visuo-spatial versus verbal domain in the DS profile. For example, some assessments, such as the Tower of London test, contain disk transfer tasks that require children to distinguish between different coloured rings in order to put them in their correct places (Le Bouc et al., 2020); this may pose difficulties if those with DS have greater difficulty than TD populations in differentiating colours (as found in an early study by Pérez-Carpinell, De Fez & Climent (1995). Other EF tests may require specific/local processing skills, which research indicates is a particular area of difficulty for those with DS. For example, studies have revealed that individuals with DS are more skilled in focusing on the global configuration and more holistic processing of objects (Bellugi et al., 1994; Porter & Coltheart, 2006), rather than focusing on local features – that is, they are better at recognising the whole, rather than specific elements of an object or image. However, these findings have been challenged, with some authors pointing to the wide range of abilities and individual differences within DS (D'Souza et al., 2016; Karmiloff-Smith et al., 2016), and
others asserting that in order to better understand the challenges that those with DS experience with visual processing, it may be important to analyse the challenges exhibited in other underlying processes, such as selective attention (Cornish et al., 2007) and visual memory (Lanfranchi et al., 2012). However, whilst there may be individual differences in the visual profile of DS, performing poorly in an EF task could reflect the visual difficulties experienced or exhibited by those with DS, rather than poor EF skills.

The same consideration would apply to physical abilities, as some EF tasks require fine motor skills (for example EF assessments that require the use of a mouse or keyboard, such as the TOL or CANTAB) yet these skills are typically somewhat delayed in children with DS (Smith, 2014). As a result, low test scores could reflect these disabilities, rather than EF weaknesses.

A further concern is that there is a lack of normative data for children with DS. As a result, it is likely that assessments of DS children may be conducted using EF tasks, such as the BRIEF-P, that are designed for younger, sometimes preschool, TD children (for example, Chen et al., 2014; Lee et al., 2011). Tests of EF have therefore often been constructed using normative data from TD children and then adapted for use for children with DS. However, it may be that normative data generated from studies of TD children does not reflect factors that affect children with DS, such as their lack of fine motor skills and their visual, hearing and linguistic delays, or take into account the greater CA of those with DS, and their subsequent greater (and more varied) life experiences.

To overcome this, EF assessments must be designed for the population they are intending to assess, and take into account the impact of any differences in location and sample group on assessment outcomes (Sabat et al., 2020). There is one EF assessment designed for those with DS: the informant-rated Cognitive Scale for Down Syndrome (CS-DS) (Startin et al., 2019). This is designed to
measure cognitive abilities within EF, memory and language domains in adults and has been found to be a valid measure. Yet the scale has so far only been used with adults, and therefore further validated measures of EF, specifically designed for use with children and adolescents with DS, are required.

Regardless, any assessments with children with DS must follow correct protocols for how the assessments should be conducted, not only to ensure accurate measurements are recorded, but to make sure that the administration of the test accommodates the patterns of disability typical of those children (Daunhauer et al., 2017; Roth et al., 2014).

2.6 Conceptual Model of Executive Function Down Syndrome

In studies of EF in DS, there are a number of ways in which theory may be drawn upon, such as including consideration of specific theories of EF (as outlined in the previous section) or broader theories of disability (including the medical model or social model of disability (Anastasiou & Kauffman, 2013; Bricout et al., 2004). These theories have been considered and drawn upon throughout the thesis where relevant. However, a conceptual model is also a useful component within a research study, as it enables key variables that may be critical to a phenomenon of interest to be drawn together (Rocco & Plakhotnik, 2009). A model is arguably more useful than a theory as a theory is conceptual framework of an idea, presented as a set of statements and an end product of scientific theorising. Conversely, a model is a set of assumptions often presented as a visual representation of a phenomena (Psillos, 2005). One such conceptual model that was specifically developed to represent development in children with developmental disabilities is the developmental causal model approach (Morton, 2004) (Figure 2.2).

Figure 2.2 reveals that, the developmental causal model approach acknowledges that environmental factors (including social, economic and
cultural factors), can affect development on three levels: biological, cognitive and behavioural. Biological factors include genetics and brain conditions, which partially determines cognitive factors, which in turn leads to behavioural phenotypes and patterns of behaviour. Morton’s model describes a one-way process; in this thesis, the behavioural element in particular is acknowledged to be two-way (e.g., behaviour can impact the environment, which, in turn, can then impact cognitive outcomes and so on), as the researcher is a strong advocate of this idea.

Fig. 2.2 Morton’s (2004) Developmental Causal Modelling Frame

This conceptual model presents a structure to understanding DS, as it allows individuals to ascertain explanations for any developmental disorder at three levels, each of which, in turn, is influenced by environmental context. As Oliver and Woodcock (2008) assert, this modelling approach enables a holistic account of a disorder or condition. The model has been used in relation to a number of different developmental disabilities. For example, Morton (2004) used it to explain the central coherence, executive functioning and affective deficit within
autistic individuals. Separating causal elements into three primary levels enables researchers and practitioners in mental and physical health/development fields to describe a condition beginning with biological/genetic causes first, before hypothesising about cognitive structures and documenting observed behaviours.

In the case of DS, this causal modelling framework is useful because explanations at all three levels help in understanding DS as a condition. DS is a genetic disorder (as highlighted previously) (Sotonica et al., 2016), which subsequently affects brain development. (Bartesaghi et al., 2015; Ersoy, Güler & Çetin, 2018). Various environmental factors can influence DS causation at a genetic level (e.g., antenatal screening, antenatal care, maternal age of conception, maternal behaviours during pregnancy and attitudes towards DS and abortion) (Corona-Rivera et al., 2019).

This altered brain development then affects cognitive skills, including IQ (Boat & Wu, 2015; Mégarbané et al., 2013), WM, STM and LTM (Lanfranchi et al., 2015) and EFs more generally. The environment can also influence cognitive abilities; for example, health and educational policy, resources and practitioners, and family members/caregivers and peers, can all play an essential role in the development of cognitive abilities. For example, teachers’ knowledge and perspectives surrounding EFs and general cognitive functioning in children and adolescents with DS strongly influences their teaching strategies, some of which may help to improve EF (Gilmore & Cragg, 2014; Rapoport, Rubinstein & Katzir, 2016). This is discussed in greater detail in chapter four.

Finally, both the environment and cognitive functioning can affect behavioural outcomes. For example, IQ level is often associated with level of challenging behaviour (e.g., Foley et al., 2016) Difficulties at the cognitive level with EF can also lead to challenging behaviour, such as a lack of self-regulation and challenging EF tasks that lead individuals with DS to try to distract or escape
from the task (Feeley & Jones, 2006; 2008). This can also make relationships more difficult to establish and maintain with those with DS (Bieberich & Morgan, 2004). Sabat et al. (2020) also illuminates how models of EF can be related to behaviour, rendering them practically useful, which has relevance for this thesis. Again, the environment is influential at the behavioural level (for example, a noisy classroom where a child with DS cannot adequately hear teacher instructions could influence levels of challenging behaviour).

Morton’s (2004) modelling framework is therefore deemed to be a suitable conceptual model through which this thesis can launch an investigation into the EFs of those with DS, given that it takes various integrated levels of explanation for DS into account. It is a useful modelling approach that has inspired subsequent visual modelling approaches (such as Moore and George’s (2011) causal model of ASD) and social cognition in DS (Cebula et al., 2010).

The visual model (Figs. 2.2) can help in the development of explanations of how DS affects cognitive function and its impact on behaviour, and the role of the environment within this. However, this thesis was not focused on testing models, and cognitive functioning - and EFs specifically - is the central component of this research: there is little focus on DS at the biological level, and there is more focus on the cognitive than the behavioural level. Study one in this thesis was focused in the main on developing a clear picture of EF at the cognitive level in DS (as opposed to how EF is influenced by brain development in DS or the environmental influencers, for example). Morton’s model played a greater role in informing the design of the second study in this thesis, including the approach to the intervention, which aimed to help teachers to understand WM briefly in terms of the role of the brain, how WM can affect behaviour in the classroom, and the impact of the environment on WM development. Whilst this study did not focus greatly on genetics or brain development, it did investigate whether changes at the cognitive level (in terms of WM) would impact children’s behaviour (see
chapter four for more details). Thus, the model is useful within the thesis because it arguably provided a holistic framework, facilitating a greater understanding of the biological, cognitive and behavioural phenotypes exhibited by this population group, rooting the particular focus of cognitive elements (EF) within a wider context.

2.7 Conclusion
This chapter presented different aspects of DS. It investigated the prevalence and common characteristics of DS and associated issues, including the genetic causes, risk factors, and physical features and health issues associated with DS. It also reviewed the different behavioural phenotypes and the relationship between these phenotypes and EFs in DS and investigated definitions, models, developmental trajectories of EF and assessment of EFs.

Since the 1840s, important advancements have occurred in our understanding of how the brain manages, regulates and organises information and coordinates how humans react to their environment. Furthermore, the effective functioning of the brain demands a system of EFs. This system of EFs is predominantly controlled by prefrontal areas of the frontal lobes, parts of the brain that, whilst from an evolutionary perspective, have recently developed, enable a complex, sophisticated EF system. However, this literature review has revealed that there are limitations in exploring EF, given the lack of a clear operational definition, which can impact on the design of effective, a paucity of literature that specifically compares the EF development of those with DS by CA, accurate EF assessments. Future research must define, understand, and develop clinical strategies and interventions to facilitate that understanding of how the EF system operates.

From this review, it is clear that the role of EFs in various aspects of the lives of those with DS needs greater consideration within the research literature.
Moreover, whilst much is now known about the specific cognitive and behavioural phenotypes of those with DS, aspects remain unclear; there is therefore a need for greater research in this area, specifically in the domain of EF, that is worth exploring through a systematic review. This can enable a much deeper insight into the executive functioning of those with DS, especially in terms of those EFs that are relatively strong or unaffected in those with DS, as well as those EFs that present particular areas of difficulty. It should also assist in situating EFs within the causal model framework of DS discussed in the previous section. The next chapter presents a systematic review of current research into the executive functioning of children and adolescents with DS. The research questions for the systematic review are as follows:

1. With reference to the existing literature, which elements of EF are delayed or differ amongst children and adolescents with DS compared to matched populations (TD matched for MA or CA, or those with developmental and/or chromosomal disorders)?
2. What are the main, relative EF strengths and weaknesses in DS?
Chapter Three: Executive Function in Children and Adolescents with Down Syndrome: Systematic Review

3.1 Introduction

This chapter reports on study one of the thesis, a systematic review of the studies that specifically explore executive function (EF) abilities in children and adolescents with Down syndrome (DS) compared to either typically developing (TD) individuals, normative data or other groups of individuals with intellectual disability (ID). Firstly, a justification of this study is given, before the methods that were used to source and review the studies are detailed. The results are presented in a narrative synthesis, before being discussed, with implications and recommendations given.

Patterns of relative cognitive strengths and difficulties are not yet fully specified in DS because, as Landry (2003) notes, the cognitive profiles of persons with DS are highly complex. This complexity includes areas of strength and weakness. For example, there are relative strengths in the visuospatial processing of those with DS, skills that are often more pronounced than verbal processing skills (Jarrold, Baddeley & Hewes, 1999; Klein & Mervis, 1999; Wang & Bellugi, 1994).

As revealed in this emerging profile, people with DS have difficulties across a range of cognitive functions (Thompson, 2003). Importantly, this also involves significant deficits in EFs. These include working memory (WM) (Lanfranchi et al., 2009), planning (Fidler et al., 2005), shifting (Kopp, Krakow & Johnson, 1983; Rowe, Lavender & Turk, 2006) and inhibition (Edgin, 2003; Rowe et al., 2006). There are also findings of low levels of global EF (Daunhauer et al., 2014) and weaknesses in attention, self-regulation and impulse control (Landry, 2003). In a study by Costanzo et al. (2013), participants with DS demonstrated various executive difficulties depending on the EF domain and modality of the
task (whether it required visuospatial or verbal skills), compared to mental age (MA) matched TD children and to children and adolescents with Williams syndrome (WS). Those with DS were particularly poor on tests of shifting, inhibition and verbal aspects of WM. Similarly, Carney et al. (2013) compared EF measures (WM, fluency, inhibition and set-shifting) in 25 children and adolescents with DS to 26 TD children matched for MA and 24 children with WS matched for MA and CA; they found that the DS group demonstrated relative difficulties within set-shifting (verbal, not visuospatial). Using a questionnaire given to parents and teachers, Daunhauer et al. (2014) also found that DS is associated with difficulties in WM, planning and inhibition compared to MA-matched TD children.

The cognitive profile of persons with DS has been brought further into focus by recent studies that have conceptualised ‘hot’ EFs as distinct from ‘cool’ EFs in an attempt to clarify the specific difficulties encountered by those with DS (Lee et al., 2015). ‘Cool’ EFs are those considered to be relatively free of affect or emotional ‘charge’ and which are essential to cognitive tasks involving WM and planning (Zelazo & Müller, 2011). By contrast, ‘hot’ EFs are those cognitive abilities considered necessary for decision-making, goal-setting and delaying gratification, which are more motivationally or emotionally laden (Zelazo & Müller, 2011). ‘Hot’ EFs are associated with reward, reinforcement and motivation. On the basis of standardised test scores in Lee et al.’s (2015) study, one can conclude that individuals with DS exhibit greater difficulty with ‘cool’ EFs, especially in WM and monitoring. However, the scores for organisation of materials (OM) and emotional control (EC) were lower on the BRIEF-P measurement compared with other EFs, indicating relative strengths in these areas (Lee et al., 2015). In fact, when compared with the normative mean and a CA-matched TD group, the scores of children and adolescents with DS in OM and EC, although slightly worse, were not significantly so. Overall, data suggests that young people with DS have greater difficulties in the domains of
‘cool’ EFs than in the domains of ‘hot’ EFs, although some individuals with DS demonstrate atypical development scores compared to TD children matched for CA across both ‘hot’ and ‘cool’ test scores. This was confirmed in Lee et al.’s (2015) study, which found that deficits in both ‘hot’ and ‘cool’ EFs are present in persons with DS, beyond the general intellectual functioning deficit, although the difference in ‘hot’ EF ability between individuals with DS and TD individuals is not always significant (depending on the individual).

In addition to this complexity in the EF profile of people with DS, the picture is further complicated by variations in research approaches to investigate this profile, for example, in methods, samples, controls and the specific EF investigated (d’Ardhuy et al., 2015; Amadó et al., 2016; Memisevic & Sinanovic, 2014). This can render it difficult to form a clear overall picture, given the discrepancy in results. For example, some studies have found significant difficulties in specific EFs in those with DS, whilst other studies (using a different method or age group) have not. For example, Lanfranchi et al. (2010) found adolescents were significantly poorer in set shifting (cognitive flexibility) using the Rule Shifting Card Test (Wilson et al., 1996), compared to CA-matched TD groups. By comparison, Roberts and Richmond (2015), investigating CA-matched TD and DS groups of preschool age, found no significant differences on the A-not-B task in set shifting. It is unclear whether this difference is due to the age of the participants, the measurement used or some other variable, such as sleep quality, which can affect EF (Esbensen & Hoffman, 2018). Moreover, the performance of people with DS on the tests and tasks mentioned above may vary depending on the individual, or may change from one day to the next depending on the mood of the individual being tested, as is typical across cognitive testing in DS (not just in tests of EF) (Wishart & Duffy, 1990).

Moreover, the samples of individuals with DS are often compared to different groups matched on a range of factors, such as CA, MA, normative data or with
participants with other difficulties, such as Autism Spectrum Disorder (ASD) or Disruptive Behaviour Disorder (DBD). This, again, renders it a challenge to draw firm conclusions regarding specific EF deficits in those with DS, comparative to other groups.

Another issue is, as alluded to above, we do not yet understand clearly how these difficulties vary with age; EF competence has been found to change over time in TD individuals (Salthouse et al., 2003), and Lee et al. (2015) showed that there were fairly similar age-EF relations between a sample of 85 individuals with DS and 43 TD individuals, aged 4-24, revealing fewer difficulties with increasing age in both groups. This lends support for the developmental stability hypothesis in DS. However, it is not yet clear whether EFs that are shown to be areas of difficulty in adolescence in those with DS also present difficulties in childhood for individuals with DS. It is possible that each age level may have a characteristic profile; understanding these differences across age groups could help inform the design of interventions aimed at people with DS in specific age groups (Lee et al., 2015). Therefore, despite the emerging profile discussed briefly above, there are still several aspects of EF in the DS cognitive profile that remain to be further explored.

It becomes pertinent, therefore, to explore the existing literature further surrounding the EF strengths and weaknesses of children and adolescents with DS. This might enable, despite the complexity and variations in research approach and sample, some tentative general conclusions to be drawn from the research, allowing the emergence of a broad profile that maps the overall EF of individuals with DS in various domains. Therefore, this systematic review aimed to clarify characteristic patterns of EF delay and/or difficulty in children and adolescents with DS and identify the tools most common for testing levels of EF in these children and adolescents. In doing so, it also aimed to assess how the results varied, depending on the control groups chosen (TD children, children
with other ID or a within-groups comparison of individuals with DS that possessed specific characteristics) and on the matching variables (CA or MA) used. Another factor that was considered was whether the tasks used were verbal or non-verbal in nature. Finally, through the systematic review, a comparison of results was made, in an effort to identity how consistent the results of various studies are. This then enabled a foundation for study two, as it allowed some light to be shed on the EF that was most delayed or challenged in those with DS, thus indicating which EF should be targeted for a practical intervention to assist the skills of children with DS in this area.

The research questions for this systematic review were therefore as follows:

- With reference to the existing literature, which elements of EF do children and adolescents with DS experience/exhibit most difficulty in, compared to matched populations (TD matched for MA or CA, or those with developmental and/or chromosomal disorders)?
- What are the main, relative EF strengths and weaknesses in DS?

3.2 Methods

In this section I explain the research strategies, databases used, type of sources included, inclusion and exclusion criteria and procedures to undertake this systematic review. Registries were checked to confirm that no such systematic review had already been undertaken. These included the YORK prospective systematic review register and the PROSPERO International prospective register of systematic reviews. This study was registered on PROSPERO (study number: CRD42016042595). The protocol is provided in Appendix 1.1.

Systematic reviews are a useful tool to explore and synthesise the findings of multiple studies within a particular field. This renders conclusions drawn more reliable and accurate than that of a single study (Gopalakrishnan &
Ganeshkumar, 2013). It can therefore facilitate more useful implications and ensure best practice in a field. However, limitations of systematic reviews include limited search criteria, databases and studies of poor/reduced quality, which needs to be taken into account when synthesising results (Bartolucci & Hillegass, 2010). It can also be difficult to compare findings of very disparate studies using different methods; furthermore, given potential publication bias as a result of preferences of peer reviewers and journal editors, some studies that may provide a different viewpoint may not be included in the review (Bartolucci & Hillegass, 2010). It is a challenge to overcome this latter obstacle; however, the previously mentioned limitations were all addressed in this review.

3.2.1 Databases
A comprehensive search of the academic literature was conducted using electronic bibliographic databases. Initial pilot searching indicated that ASSIA, Cochrane Library, Medline, PsycINFO, Web of Science and ERIC were the most suitable databases, as they allowed me to identify articles or studies that met the review inclusion criteria. The EMBAS database was initially also included, but was subsequently excluded at the pilot searching phase, because the search results obtained failed to meet my inclusion criteria; most of the search results produced were irrelevant to the research topic, focusing instead on animal and plant research. Reference lists of included study articles were searched for further potential sources.

3.2.2 Types of Sources
I included book chapters and journal articles published prior to 31 January 2020 (the search date) in my initial search. Other sources, such as thesis, reviews and conference proceedings were considered ‘grey literature’ and were excluded. However, after screening the full texts, I found that all sources identified as meeting the inclusion criteria were journal articles; therefore, no book chapters are included in the review.
3.2.3 Search Strategies

The search strategy consisted of Boolean combined keywords and subject headings related to the aims of the systematic review. The same search string was used for each database (see Appendix 1.2). Some initial keywords were excluded or amended, for example ‘impulse’ was changed to ‘impulse control’, ‘mental planning’ was changed to ‘planning’ and ‘mental recall’ was changed to ‘recall’. This was done because their meaning was ambiguous or returned results not relevant to the field of EF. The final list of keywords is given in Table 3.1 below; between the words within each list ‘OR’ was used, and between each column, the word ‘AND’ was used.

Table 3.1 Keywords of Search Strategy

<table>
<thead>
<tr>
<th>(Executive function)</th>
<th>(Syndrome, Down)</th>
<th>(Child*)</th>
</tr>
</thead>
<tbody>
<tr>
<td>(Problem Solving)</td>
<td>(Down’s Syndrome)</td>
<td>(Adult*)</td>
</tr>
<tr>
<td>(Decision*)</td>
<td>(Downs Syndrome)</td>
<td></td>
</tr>
<tr>
<td>(Inhibit*)</td>
<td>(Syndrome, Down’s)</td>
<td></td>
</tr>
<tr>
<td>(Distract*)</td>
<td>(Syndrome, Downs)</td>
<td></td>
</tr>
<tr>
<td>(Attention)</td>
<td>(Trisomy 21)</td>
<td></td>
</tr>
<tr>
<td>(Impulse control)</td>
<td>(Chromosome 21)</td>
<td></td>
</tr>
<tr>
<td>(Mental Flexibility)</td>
<td>(Mosaicism)</td>
<td></td>
</tr>
<tr>
<td>(Set Shifting)</td>
<td>(Translocation)</td>
<td></td>
</tr>
<tr>
<td>(Working memory)</td>
<td>(Intelectual<em>disabilit</em>)</td>
<td></td>
</tr>
<tr>
<td>(Recall*)</td>
<td>(Intelectual* Impair*)</td>
<td></td>
</tr>
<tr>
<td>(Planning)</td>
<td>(Retard*)</td>
<td></td>
</tr>
<tr>
<td>(Switch)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Self Regulat*)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
3.2.4 Inclusion and Exclusion Criteria

Inclusion criteria for sources were as follows:

- Studies should involve participants aged from 2 years up to adolescent (adolescent age range was determined by using the definitions of adolescent and youths from the United Nations General Assembly (1989) and United Nations Human Rights Committee on the Rights of the Child (2003). These stipulate that adolescents are 10-19-years-old and ‘youth’ are 15-24 years old, as well as what the authors of each study determined their sample age to be);
- The studies should focus on EF in DS populations;
- Studies should have research designs that were either randomised/quasi-randomised, non-randomised controlled, cohort, case-control, observation, cross-sectional, or longitudinal;
- Studies had to either include a distinct DS group OR include samples with mixed developmental diagnoses where the sub-sample of DS participants could be clearly identified in the data results;
- Studies had to include at least one of the following measures of EF: standardised/objective tests, assessment by trained staff or caregiver report/questionnaire, standardised observation methods;
- Finally, all sources used had to be available in the English language.

The exclusion criteria were as follows:

- Qualitative studies, case studies, and expert opinion papers;
- Studies in which the sample populations consisted entirely of children with DS who were less than 2 years old;
- Samples with mixed developmental diagnoses if the sub-sample of DS participants could not be clearly and separately identified in the data results;
• Studies that reported EF exclusively in other, non-DS populations;
• Studies that did not report on EF;
• Intervention studies that did not include comparison with non-DS populations or with normative data;
• Conference abstracts.

After conducting the primary trial searches, it was found that a larger than expected number of studies were returned, so further exclusion criteria was applied: adult studies were excluded, to allow me to focus on children and adolescents.

3.2.5 Procedures
3.2.5.1 Screening Phases
The results from the searches of all databases used were downloaded into Endnote and were merged into one master library. After the initial database searches (record \( n=36610 \)), duplicates were excluded, and the remaining articles totalled 29557 (this number also includes studies \( n=31 \) added after the initial full text screening by checking the reference lists of included studies for any other relevant studies). Results were then transferred to Excel and were screened in three phases: titles, abstracts and full texts. After screening 29557 returned studies by title (excluded \( n=27113 \)) and subsequently 2444 for abstract using the criteria above (excluded \( n=2214 \)), 230 studies remained. The final screening phase was a complete screening of the full texts, using the inclusion and exclusion criteria described above. The final included studies numbered 55. In all screening phases I used the criteria above.

This process is summarised in the PRISMA diagram on the following page (see Figure 3.1). Studies excluded at the full-text screening stage are noted, with reasons for exclusion. The data from the final set of studies were then extracted into study data sheets (see 3.2.5.3).
3.2.5.2 Reliability of Each Phase

In each screening phase, 10% of documents were randomly chosen and screened by both myself and my supervisors, which the *Cochrane Training Manual* (Higgins et al., 2021) asserts is an appropriate, acceptable method to follow. The reliability percentages were as follows:

- Screening titles reliability was 97.2% and Cohen’s kappa value=0.95, which indicates substantial reliability;
- Screening abstracts reliability was 78.24% and Cohen’s kappa=0.66, which indicates substantial reliability;
- Screening full texts reliability was 100% and Cohen’s kappa=1, which indicates perfect reliability;
- Screening quality assessment reliability (see 3.2.5.4) was 80% and Cohen’s kappa=0.65 indicating substantial reliability. Any disagreements were discussed to reach a consensus.
Records identified through database searching:
- MEDLINE, Psych INFO, WEB OF SCIENCE, ERIC, COCHRANE LIBRARY, ASSIA ($n=36580$)
- Additional records identified through other sources ($n=31$)

Records after duplicates removed ($n=29557$)

Titles screened ($n=29557$)

Abstracts screened ($n=2444$)

Full-text articles assessed for eligibility ($n=230$)

Studies included in synthesis ($n=55$)

Full-text articles excluded, with reasons:
- Qualitative studies ($n=5$); 
- Studies including populations with mixed developmental diagnoses while the sub-population of DS participants could not be clearly and separately identified in the data results ($n=55$); 
- Not DS studies ($n=7$) 
- Contained adult population only ($n=44$); 
- Intervention did not meet inclusion criteria ($n=12$); 
- Did not explore EF in DS but explored correlations of EF with other variables ($n=34$); 
- Did not explore WM and attention as EFs ($n=15$). 
- Children with DS who were less than 2 years old ($n=3$)
3.2.5.3 Data Extraction and Synthesis

All data from included sources were extracted into study data sheets, which detailed the following: citation; sample (child, adolescent and mixed ages studies that included both children and adolescents) and DS group size; sample characteristics including any co-occurring diagnoses; mental age (MA), chronological age (CA), sex and ethnicity; sample controls/comparators; country and setting; study research design and methods; EF measured; measures of EF; summary of main findings; EF specific findings; and quality assessment. Regarding the categorisation of studies into ‘child’, ‘adolescent’ or ‘mixed’, I both followed the researchers’ description of the sample as well as the United Nations (1989; 2003) recommendations (detailed earlier); this is to ensure I did not miss some studies that may have contained adolescents if the title and abstract of a study described them as ‘young adults’, for example. Also, it is recognised that different cultures and researchers may have slightly different ideas about the conventional age range of children and adolescents (Blakemore & Choudhury, 2006); thus, the UN guidelines provided a useful tool to standardise the age range.

3.2.5.4 Quality Assessment Methods

Assessment forms (‘Randomised control trial’, ‘Case control study’ and ‘Cohort study’) from the CASP (2018) toolkit were employed to appraise the quality of evidence generated from the selected studies. Some included studies did not match exactly the type of study given in the titles of these three assessment forms. Inspired by Katrak, Bialocerkowski, Massy-Westropp, Kumar and Grimmer (2004) and Murakami, Fox and Dijkers (2017), who struggled to find a CASP form to fit all the studies in their systematic reviews and thus modified it to create their own assessment, a quality assessment form was created containing five questions drawn from the CASP forms mentioned above. These questions fitted all study designs in this review. The first two questions in the created checklist were scored out of 2 (yes=2 or no=1) and the remaining questions
were marked out of three (yes=3, somewhat=2, no=1) for a maximum score of 13. The quality assessment score is included in Table 3.4. Articles that scored 8 or less on the quality assessment scores were considered low quality, while those scoring 9 and above (out of a possible score of 13) were considered high quality, which was similar to the scoring system used by Murakami et al. (2017).

These questions were:

- Did the study or trial address a clearly focused issue?
- Did the authors use an appropriate method to answer the research question?
- Were cases recruited in acceptable ways? (ethics)
- Have the authors taken account of potential confounders in the design or analysis?
- Was the analysis and presentation of results precise and rigorous?

3.3 Results
Table 3.4 (at the end of this chapter) summarises the studies, and is organised by participant age group and then alphabetically by surname of first author. For each study, details are given that summarise and simplify the greater level of detail contained within the initial study data extraction sheets. Study details given in Table 3.4 include study sample, location, design, EF measured (specific EF or general profile), measures, results and methodological quality.

This information was then summarised into a narrative synthesis (Boland, Cherry & Dickson, 2014), arranged according to ‘global’ (a range of) EF, or specific EFs. Studies are then organised within this according to the age of participants (child, adolescents, or mixed-age group).
3.3.1 Sample Size

Regarding studies with child participants \((n=23)\), the number of participants ranged from 12 to 183. It can be seen from Table 3.4 that five studies involved between 10 and 20 participants, eight involved between 21 and 30 participants, two involved between 31 and 40 participants and one involved 41 to 50 participants. Four studies had between 70-80 participants, one had 112 participants and another had 142. The largest number of participants in a study was 183.

There were eight studies with adolescent participants, ranging in number from 12 to 49. Six studies included between 10 and 20 participants, one study contained 49 participants and one included 112 participants. These studies sometimes included young adults (not solely adolescents), which were not the focus of this investigation; therefore, the results of the adolescent-aged samples in these studies were focused on, where they were reported separately from the young adults. Otherwise, the results were included from the whole sample (adolescents and young adults), where they could not be separated.

Of the 24 mixed child/adolescent sample studies, the number of participants ranged from 11 to 369. Nine studies included from 10 to 20 participants, seven studies included between 21 to 30 participants, one study included 31 to 40 participants, one study included 41 to 50 participants and three studies involved between 61 and 70 participants. One contained 97 participants and one contained 128 participants. Finally, the largest mixed-age population study involved 369 participants. From the above, it can be seen that the studies with mixed-age populations involved the largest sample sizes.

3.3.2 Locations

Regarding research locations, the largest number of studies \((n=14)\) were conducted in the USA. The other studies were conducted in the UK \((n=9)\),
Canada \( (n=2) \), Italy \( (n=9) \), Australia \( (n=7) \), Brazil \( (n=2) \), Spain \( (n=2) \), Scotland \( (n=1) \), one from Greece and one from Germany. A few studies were conducted in East-Central Europe, in Bosnia \( (n=1) \) and Romania \( (n=1) \). There was only one study from the Middle East (Israel) and one study from India. Three studies were joint research projects involving researchers from different countries (see Table 3.4).

### 3.3.3 Research Design

Most of the studies were cross-sectional studies \( (n=52) \), including an observational study. Two were longitudinal research studies \( (n=2) \) (see Table 3.4).

### 3.3.4 Quality Assessment Results

The fifty five studies were assessed for quality in this review; all scored between 11 and 13 out of a possible total score of 13. Specifically, 32 studies of 55 scored 13, while 13 studies scored 12 and finally 10 studies scored 11, as some (predominantly older studies) lost marks given they had ‘somewhat’ as the response, predominantly in questions 4 (Have the authors taken account of potential confounders in the design or analysis?) and 5 (Was the analysis and presentation of results precise and rigorous?), lacking clarity in either their design or results. Some studies did not rigorously address the confounding variables in their designs or results, whilst a few others could have analysed and presented their results more clearly to allow a clearer picture of group differences (for instance, some did not report on the significance of their results). The more recent studies tended to score more highly in the quality assessment.

Use of the CASP tool in this study was necessary, as this critical appraisal provided a systematic platform for assessing the quality of the research, taking into account validity and reliability (Katrak et al., 2004). The abbreviated list of CASP questions, however, were perhaps simplistic, enabling many studies to
obtain a high score as they predominantly focused on the studies’ clarity of focus, appropriateness of method and acceptability of recruitment. This simplicity was necessary, however, as the CASP tool needed to be applicable for a range of research designs. As a result, it must be noted that it is possible that each study was not assessed for quality as rigorously as it might have been, as the generalised CASP questions did not allow for specific questions to be asked of particular studies.

3.4 Results: Narrative Synthesis

This section presents a narrative synthesis of EFs in children and adolescents with DS. A narrative synthesis encapsulates data that is summarised narratively, that is, with text and words to explain the findings (Boland, Cherry & Dickson, 2014). A narrative syntheses was useful given the range of very different studies in this review, focusing on varied aspects of EF explored using a range of measurement tools (Popay et al., 2006). Firstly, a preliminary synthesis was undertaken by way of categorising the findings according to the EF(s) studied in the articles (for example, WM, attention); then, the included studies were summarised in a narrative synthesis within this thematic framework (Popay et al., 2006). Likewise, EF profiles by age are also presented in this synthesis. The groups for comparison are typically developing (TD) participants, and participants with other disabilities. In addition, the tools used in each study are presented.

The narrative synthesis is presented in the following framework of EFs; global EF; multiple EF; working memory; attention function; mental/cognitive flexibility, shifting and switching; self-regulation; inhibition and impulsivity; planning/organisation. A narrative synthesis summary is then provided. As can be seen from this framework, studies that used standardised questionnaires, such as the BRIEF-P or BRIEF to measure global EF, and studies that employed tasks in which several EFs were measured (multiple EFs), are
presented. Equally, studies that focused on individual EFs are discussed narratively.

3.4.1 Global Executive Functioning

Eleven studies investigate global EF in DS. Seven studies used the standardised BRIEF-P (Lee et al., 2011; Daunhauer et al., 2014; Pritchard et al., 2015; d’Ardhuy et al., 2015; Loveall et al., 2017; Joyce et al., 2020; Wilde & Oliver, 2017) while five used the BRIEF (or also used the BRIEF alongside the BRIEF-P) (Camp et al., 2016; Memisevic & Sinanovic, 2014; Mason, Spano & Edgin, 2015; Lee et al., 2015; Joyce et al., 2020). Four studies (Lee et al., 2011; Joyce et al., 2020; Daunhauer et al., 2014 and Pritchard et al., 2015) looked solely at children with DS, although two (Camp et al., 2016; d’Ardhuy et al. (2015) focused purely on adolescents, and five studies (Lee et al., 2015; Loveall et al., 2017; Mason et al., 2015; Memisevic & Sinanovic, 2014; Wilde & Oliver, 2017) involved mixed-aged participants. Table 3.2 and 3.3 show the indices and subscales of BRIEF and BRIEF-P, which contain similar measures yet are grouped differently. In each, the total score from each scale combined generates a global executive composite (GEC).

<table>
<thead>
<tr>
<th>Table 3.2 BRIEF Index Subscales</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Index/Subscale</strong></td>
</tr>
<tr>
<td>Behavioural Regulation Index (BRI)</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Metacognition Index (MI)</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>
Table 3.3 BRIEF-P Subscales

<table>
<thead>
<tr>
<th>Index/Subscale</th>
<th>Component/Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inhibitory Self-Control Index (ISCI)</td>
<td>Inhibit (IN)</td>
</tr>
<tr>
<td></td>
<td>Emotional Control (EC)</td>
</tr>
<tr>
<td>Flexibility Index (FI)</td>
<td>Shift (cognitive flexibility) (S)</td>
</tr>
<tr>
<td></td>
<td>Emotional Control (EC)</td>
</tr>
<tr>
<td>Emergent Metacognition Index (EMI)</td>
<td>Working Memory (WM)</td>
</tr>
<tr>
<td></td>
<td>Plan/Organise (PO)</td>
</tr>
</tbody>
</table>

In terms of the child studies, Lee et al. (2011) used the BRIEF-P to assess EF skills in young children (mean chronological age (CA) of 6.3 years). The comparison of the BRIEF-P scale with normative mean scores showed that children with DS presented significant challenges in EF when compared with children with a similar mental age (MA) of approximately 3 years old. The T-score for children with DS was significantly higher (an indication of poorer functioning) on the Global Executive Composite (GEC) and Emergent Metacognition Index (EMI) (which tests WM and Planning-Organising (PO)) on the BRIEF-P scale, compared to the normative mean. However, the T-score for the Inhibitory Self-Control Index (ISCI) and Flexibility Index (FI) on the BRIEF-P scale were considerably lower (an indication of better functioning) compared to the other EFs in the sample of children with DS, although they were still higher than the normative mean (indicating some weakness in these EFs), although this difference was not statistically significant at this age. Emotional control (EC) had a significantly lower T-score compared with other EFs, denoting that this domain presented the least difficulty in the sample, especially as it obtained a similar score to the normative mean for TD children.

These findings were similar to that of a UK study conducted by Joyce et al. (2020), who, in a sample of 80 children with DS (mean CA of 4.7 years), used the parent-reported BRIEF-P to compare EFs to normative data. This study
investigated whether obstructive sleep apnoea (OSA) contributed to worse EF in children with DS. However, using MA to score the BRIEF-P, their comparisons of BRIEF-P scores with MA-based normative data across the entire sample revealed, like the results from Lee et al. (2011), that the greatest weaknesses could be found in EMI (WM and PO). This was followed by the ISCI and FI. Similarly to Lee et al. (2011), EC was found to be a relative strength compared to inhibit, WM and PO indexes. These findings do suggest that, even at this young age, children with DS do exhibit challenges in most (although not all) areas of EF compared to mean averages for their age.

Likewise, Daunhauer et al. (2014) used teacher and parent reports on the BRIEF-P to investigate a wide range of EFs in children with DS. Unlike Lee et al. (2011) and Joyce et al. (2020), who both compared the EFs scores of children with DS with the normative mean, Daunhauer et al. (2014) compared the EF outcome in children with DS with TD children matched for MA. Similarly to Lee et al.’s (2011) study, both teachers and parents reported the highest T-scores for WM compared to other domains in children with DS (indicating that this was the area of greatest difficulty), followed by PO. Therefore, as in Lee et al.’s (2011) and Joyce et al.’s (2020) studies, children with DS demonstrated difficulties in the EMI (WM+PO). Their GEC scores were also significantly higher than the TD group (similar to Lee et al.’s (2011) study), indicating global EF difficulties in children with DS. The T-score for ISCI, shifting (S) (also known as cognitive flexibility) and EC were not at a clinical level of difficulty in the DS group compared to the TD group according to the teacher reports, although results from the parent group showed significant difficulties in ISCI in the DS group compared to their MA-matched, TD counterparts (which conflicts with both the teacher reports and the findings of Lee et al. (2011)). Daunhauer et al. (2014) indicate that children with DS, according to parents’ reports, had an EC T-score similar to that reported in Lee et al.’s (2011) study, revealing least difficulty in this area in children with DS.
The observation that GEC is significantly affected in individuals with DS is also supported by other studies, including that of Memisevic and Sinanovic (2014). These researchers also used the BRIEF teacher version in Bosnia, comparing T-scores for a mixed sample of children and adolescents with DS with both normative values (assumedly for CA, as the MA of the DS group was not calculated) and two groups of children with intellectual disabilities (ID) (ID with unknown aetiology and ID from organic/other genetic cause aetiology group). The study revealed clinically significant challenges and weaknesses in S, EC, PO, organisation of material (OM), Initiate (I), WM and monitoring (M) domains, but not in ISCI - although this was near statistical significance. In addition to S, WM gained the highest score amongst the seven scales, as in Lee et al. (2011), Joyce et al. (2020) and Daunhauer et al.’s (2014) studies, indicating the area of greatest difficulty. According to the results obtained in Memisevic and Sinanovic’s (2014) study, shifting in children and adolescents with DS is significantly worse than children and adolescents with ID with unknown aetiology.

Another study involving a mixed-age sample of children, adolescents and young adults with DS was conducted in the USA by Loveall et al. (2017), who aimed to assess how global EF compared to normative data for same-aged individuals in different age brackets. Their cross-sectional study involved 112 participants from three age groups: 2-5-years old; 6-18-years-old; and 19-35-years old. Results from the adult group are not reported here given that adults were not the focus in this review. They found, using the BRIEF-P and the BRIEF where appropriate, that in all age groups, the GEC was significantly worse in the DS sample compared to normative data. However, only EMI in the 2-5-year-old age group was significantly above the TD norm (indicating significant difficulty in this index, which includes WM and PO). WM was significantly higher, whereas PO and inhibition was marginally significantly higher, and other areas—S and EC—were
not significantly different to the TD norm. For the 6-18-year-old age group, areas of significant weakness included WM, M, S, and PO, and areas of moderate weakness included IN and I. An area of relative strength was OM and, as the studies before, EC. Interestingly, whilst most areas remained consistent between ages 2-18 in terms of the difference they represented in children and adolescents with DS, comparative to normative data, S abilities saw a marked (significant) decline with increasing age, suggesting this declines over time. This indicates that, throughout childhood and adolescence, skills in S or CF present an increasing challenge for individuals with DS.

Overall, the findings of Lee et al. (2011), Joyce et al. (2020), Memisevic and Sinanovic (2014), Daunhauer et al. (2014) and Loveall et al. (2017) indicate that WM was the weakest EF and EC the strongest EF in individuals with DS compared to TD children or normative data. However, as previously discussed, Memisevic and Sinanovic (2014) do not note in their study whether they compared their results to normative MA or CA values; since they did not calculate the MA of their participants, it is assumed that they compared the performance of their participants to the normative values for CA. It therefore may be expected that the EF performance of their participants with DS and ID would be significantly poorer compared to the normative data, although findings from Lee et al. (2011) and Joyce et al. (2020), who compared participants with DS to MA-based normative data, also found significant challenges in various areas.

The previously discussed studies however only investigated EF in populations of children with DS that do not have co-occurring conditions. Pritchard et al. (2015), by contrast, investigated EF (using the BRIEF-P) in children with DS with no simultaneous conditions, comparing them to the EF abilities of similar-aged (CA) children with DS and Autism Spectrum Disorder (ASD) or DS and Disruptive Behaviour Disorder (DBD). They found that children with DS that do not have co-occurring conditions experienced significantly lower scores (fewer
EF difficulties) on the three factors (EMI, ISCI and FI) of BRIEF-P than in the groups of children with DS plus ASD or DBD. All three groups performed best on FI (flexibility factor including Shift and EC subscales), in line with previous findings, given this contained EC, and DS (without co-occurring conditions) and DS+ASD groups performed worst on EMI (which includes WM + PO), similar to the findings from Lee et al. (2011), Memisevic and Sinanovic (2014) and Daunhauer et al. (2014). Interestingly, those with DS+DBD performed worst in ISCI (inhibition), not EMI (Pritchard et al., 2015). From this study, it can be concluded that children with DS and co-occurring conditions such as ASD and DBD are associated with significantly poorer EF performance than children with DS (without co-occurring conditions) (Pritchard et al., 2015).

However, it must be noted that Pritchard et al. (2015) conceptualised the BRIEF-P differently to the original measure, in that their ISCI factor was comprised of the inhibition subscales, instead of including the EC subscales (which were moved to the Flexibility Index (FI)). Moreover, their EMI factor (which in the original BRIEF-P is comprised of WM and PO subscales), also included some Inhibit items (12%) and Shift items (4%) from the respective subscales. This was undertaken as a result of exploratory factor analysis to explore the BRIEF-P’s structure in a sample of individuals with mixed ages, finding that the Bayesian Information Criterion (BIC) was smallest (indicating that there was a better fit of factor solutions for the data they collected) for the three-factor solution. Fewer items were dropped as a result of cross-loading for the three-factor solution compared with 2, 4 and 5-factor solutions. Therefore, the three-factor solution, comprised of EMC (which includes memory, learning and cognitive problem-solving, which demonstrated a variance of 44%), Flexibility (that contains elements of behavioural flexibility and emotion regulation, with a variance of 30%) and Inhibition (aspects of response inhibition and motor activity, with a variance of 25%) was utilised in Pritchard et al.’s (2015) study.
This may mean difficulties could be encountered when attempting to compare Pritchard et al.’s (2015) findings with that of other studies using the BRIEF-P; however, the researchers note that their version of BRIEF-P could ensure a factor of inhibition (IN) that is more focused, a Flexibility factor that is relatively similar, and an EMC factor that is also similar yet ‘not as clean’ as the original BRIEF-P index (Pritchard et al., 2015, p. 1138). Thus, their confidence of the similarities of their conceptualisation indicate findings may be compared, although should be done so with a degree of caution.

Regarding studies that focused purely on adolescents, d’Ardhuy et al. (2015) also assessed EF using BRIEF-P and found that adolescents with DS showed statistically greater atypicalities in global EF composite (GEC) compared to adults with DS, despite the slightly higher mean IQ of the adolescent group. Both groups had low scores on the Spatial Span (SSP) construct within the Cambridge Neuropsychological Test Automated Battery (CANTAB) that measures WM, indicating difficulties in this area, which remained stable over time. This perhaps indicates that some EF skills (but not WM) improve as individuals with DS progress into adulthood, and that EF may not be linked to IQ.

Interestingly, some studies opted to use the BRIEF instead of the BRIEF-P when investigating groups of adolescents with DS. Camp, Karmiloff-Smith, Thomas and Farran (2016) aimed to compare the ratings of real-world EF with problem-solving in DS, WS and TD groups, in a bid to establish the possible mechanisms underpinning success or failure on problem-solving tasks in everyday scenarios. In Camp et al. (2016) study, 31 parents/carers of adolescents with DS completed the BRIEF and a Problem-Solving Questionnaire (PSQ) developed by these researchers. The results were compared with 47 parents/carers of adolescents with WS and 34 parents/carers of TD children, whose CA was 9 years younger, on average, than the DS and
WS group, although their MA was similar. Results from the BRIEF revealed performance for the WS group was significantly poorer than the DS and TD groups, whilst performance of the DS group was significantly poorer than the TD group only in certain scales, notably the Shift, Working Memory, Plan/Organize, Monitor and Initiate scales (at p<0.5). The DS group scored highest on the scale Inhibit followed by Organisation of Materials, which indicate areas of greatest difficulty in those with DS. This perhaps contrasts with many findings that indicate the WM is the weakest EF in those with DS. However, findings can generally reveal individual differences in results, as indicated by Mason, Spano and Edgin’s (2015) study.

Mason, Spano and Edgin (2015) looked at a mixed age group and found in their study of children and adolescents with DS that there was an interaction of EF and 7-repeat (7R). 7R is an allele of the dopamine receptor D4 (DRD4) on chromosome 11 that codes for dopamine activity and helps modulate excitatory (glutamatergic) and inhibitory (GA-BAergic) receptory currents in brain areas important for EF (Wang, Zhong & Yan, 2002). Those with 7R often demonstrate specific behaviours, disorders (e.g., ADHD; Faraone & Mick, 2010) and individual differences in EF, including impulsivity (Congdon & Canli, 2008) and effortful control (Smith et al., 2012). White individuals with DS and at least one 7R in Mason et al.’s (2015) study demonstrated greater EF difficulties than individuals with DS without 7R of a similar CA, according to caregiver reports and experimenter observations (although not laboratory-based EF tasks) (see section 3.5.3). Individuals with DS+7R had extremely high scores on BRIEF BRI (the behavioural regulation index, otherwise called ISCI), EMI (the metacognition index), and GEC (global EF).

Interestingly, this finding did not extend to Hispanic children with DS. Thus, there is a clear interaction with ethnicity, although the researchers note that the White group alone had a sample size large enough to merit inclusion in the main
analysis. There was no TD control group for comparison in Mason et al.’s (2015) study, which means it is unclear how far those with DS and 7R are more at risk of EF difficulties than TD individuals with 7R, given that 7R expression was found by Mason et al. (2015) to have a similar prevalence in both DS and TD populations. Yet Mason et al. (2015) clearly demonstrate that those with DS often demonstrate difficulties in EF, specifically in ISCI, EMI and GEC.

A similar finding was gained by Lee et al. (2015), which also employed the BRIEF. These researchers found that, according to parent-reported BRIEF scores, children and adolescents with DS had significant difficulties in all EFs compared with TD children matched for CA. They also found there were no significant differences between children and adolescents with DS and those with sex chromosome trisomy (XXX or XXY) matched for CA, except in measures of EC, where children and adolescents with DS scored significantly lower than those with Klinefelter and Trisomy X syndromes (indicating fewer difficulties). Lee et al. (2015) found that the weakest functions in DS were monitoring and WM, while the least difficulty was in EC and organising of materials (OM). Similar to the previously discussed studies, Lee et al.’s (2015) findings support the idea that youth with DS have fewer hot than cool EF difficulties, with significant difficulty in WM. It must be reiterated, however, that relative strengths in EC are not absolute strengths, as participants with DS were still significantly more challenged than their TD counterparts and the normative mean in all areas, even those providing a relative strength.

A study that compared children and adolescents with DS to another group of children and adolescents with ID using the BRIEF-P was conducted by Wilde and Oliver (2017), who compared DS individuals with those with Smith-Magenis syndrome. This UK study revealed that individuals with Smith-Magenis syndrome demonstrated greater weaknesses compared to those with DS on some scales in the BRIEF-P (Inhibit, Shift and EC scales) but not others (WM;
PO). Similar to previous findings, in the DS group, the greatest challenges in the BRIEF-P were observed in WM and there were fewer difficulties in EC. Compared to normative data, children with Smith-Magenis syndrome were found to have significantly greater difficulties on all clinical scales of the BRIEF-P, whereas children with DS only had significantly greater difficulty in Inhibit, WM and PO, but not Shift or EC. This is useful in revealing the extent to which those with DS may demonstrate delay and/or challenges in different EFs compared to another developmental disorder, such as Smith-Magenis syndrome.

Summary
The studies included that explore global EFs in general reveal strong evidence that there are significant difficulties and challenges in GEC and EMI, especially WM, in children and adolescents with DS, comparative to TD individuals of a similar CA and/or MA. The evidence suggests that challenges in ISCI (inhibition), FI (flexibility, including shifting) (Daunhauer et al., 2014; Lee et al., 2011; Memisevic & Sinanovic, 2014) and, in particular, EC (Daunhauer et al., 2014; Lee et al., 2011; 2015) are not as significant as the difficulties in GEC and EMI in those with DS, compared to TD individuals (matched for CA or MA, or normative data). Compared to individuals with other ID or those with DS and co-occurring conditions, those with DS without co-occurring conditions exhibit consistently fewer difficulties in EC and hot EFs more generally (Prichard et al., 2015; Lee et al., 2015).

However, there are discrepancies. Whilst Memisevic and Sinanovic (2014) for example found that WM and shifting were significantly worse in those with DS compared to either those with other ID matched for CA or normative data, children with DS in Daunhauer et al.’s (2014) study were not deemed by teachers or parents to demonstrate particular problems in shifting when compared to TD children of a similar MA.
Moreover, the studies generally reveal that the WM difficulties and weaknesses in children and adolescents with DS appear to remain relatively stable over time at different age points (d’Ardhuy et al., 2015; Lee et al., 2015), although the GEC was found to improve with CA. However, the breakdown of which EFs improved the most was not given in d’Ardhuy et al.’s (2015) study. Loveall et al. (2017) found that the gap between EF abilities in those with DS, compared with normative data, only widened for shifting abilities as individuals with DS aged, with all other abilities remaining consistent. There are, therefore, significant limitations in the data that renders drawing conclusions somewhat difficult, especially given the challenges presented by the use of self-report surveys (the BRIEF or BRIEF-P) to collect data, something that will be discussed more extensively in due course. However, the research does indicate that global EFs, in most studies, are significantly more challenging for those with DS compared to TD children and adolescents, with particular difficulties in WM, followed by PO, and relative strengths in EC.

3.4.2 Multiple Executive Functions

Eleven studies investigated multiple (but not global) EF tasks in children and adolescents with DS. Five studies compared children with DS to TD children (Amadó, Serrat & Valles-Majoral, 2016; Daunhauer, Gerlach-McDonald, Will & Fidler, 2017; Klotzbier et al., 2020; Roberts & Richmond, 2015; Schott & Holfelder, 2015) and two studies involved adolescents with DS and compared them with TD children (Chen et al., 2014; Lanfranchi et al., 2010). A further four compared a mixed-age group with DS to TD children (Borella et al., 2013), those with WS (Landry, Russo, Davkins, Zelazo & Burack, 2012) and WS and TD groups (Carney et al., 2013; Costanzo et al., 2013).

In terms of the child studies, Amadó, Serrat and Valles-Majoral (2016) employed EF tasks to compare the EF abilities in 30 children with DS with two groups of TD children (one group matched for CA and one group matched for language
development (LD)). They assessed visuospatial WM (using the Frog task), inhibition (Day-Night Task), and cognitive flexibility (Wisconsin Card Sorting Task). They concluded that the performance on all three tasks was significantly lower in children with DS when compared to TD children matched for either CA or LD, indicating significant difficulties in these EFs.

Similarly, Schott and Holfelder (2015) used tasks assessing inhibitory control and set switching\(^1\) (including attention and distraction) on the Trails-P. The results showed that the DS group demonstrated significantly poorer performance in all functions than the TD group, matched for CA and sex. In the most complicated assignment, distraction, the children in the DS group showed the lowest efficacy scores on the Trails-P compared to TD children. Thus, both Schott and Holfelder (2015) and Amadó et al. (2016) found that children with DS demonstrated difficulties in inhibition, although it must be noted that both used CA-matched TD control groups for comparison; most studies (especially those using the BRIEF and BRIEF-P) match children with DS to TD children on MA, as this provides a better understanding of the difficulties that children with DS have in EF compared to children of a similar mental age. This is therefore more illuminating than matching children with DS to TD children on CA, as it would be expected that children with DS demonstrate difficulties in EF and other domains compared to other children their age, given their specific genotype.

Roberts and Richmond (2015) also measured several aspects of EF (WM, inhibition and set shifting), this time using the A-not-B task. The results showed that there were no differences in any of the EFs measured between children with DS of preschool age and the TD group matched for receptive language scores as a proxy for MA. They concluded that that these EF difficulties are not

\[^1\] In general, it could be said that task switching refers to moving flexibly from one set of rules to another in response to changing environmental conditions (Manoach, 2009). It is used interchangeably with ‘shifting’, which Schott and Holfelder (2015, p. 861) state is ‘the ability to shift between different mental sets or tasks’. See chapter two for a discussion of definitions of EF
demonstrated in preschool-age children with DS, and therefore are likely to become more evident as individuals age, given the prevalence of difficulties in these areas in older children, adolescents and adults.

Klotzbier et al. (2020) used the dual-task paradigm as to assess cognitive and executive function in relation to motor demands, that is, what dual-task costs were present when participants were asked to perform a complex change of direction walking task. These authors assert that EF challenges have been found to be pivotal in locomotor delay and interference; thus, the dual-task paradigm was deemed to be a useful means of assessing EF. Using a Trail-Walking-Test, Klotzbier et al. (2020) assessed performance walking along a specific pathway delineated by certain target markers that either increased in number and/or letter. They found that compared to a CA-matched TD group, children with DS (n=12) experienced significantly greater motor costs (errors) and significant decreases in speed as the cognitive demands increased (rendering the pathway more challenging to accurately follow). This difference was not observed between children with DS and a TD group matched for MA, however. The researchers did find that there were differences between DS and TD-MA groups in the execution times on the task, which they assert is a strong argument that attention in particular is an EF that is substantially reduced in children with DS. It is also inferring that cognitive flexibility skills are challenging for those with DS, given their reduced performance in switching between cognitive and motor tasks compared to a CA-matched TD group.

Daunhauer, Gerlach-McDonald, Will and Fidler (2017) used a battery of EF laboratory tasks to investigate working memory/inhibition, inhibition, planning and cognitive flexibility (shifting) in 42 children with DS, compared to non-verbal MA-matched TD children. They found that there was a significant mean difference on overall performance in the EF laboratory tasks, with the DS group performing significantly more poorly than the TD group. However, there was no
significant difference between inhibition and shifting (when looking at individual variables) between DS and TD groups; although the DS group performed marginally worse than the TD group in shifting, they actually scored marginally better than the TD group in inhibition (moderate effect size for each). This latter finding is perhaps surprising, although potential reasons for the similar performance of TD and DS groups in both shifting and inhibition are given by the researchers, including methodological aids to assist the DS group, the small sample size and the young chronological age of both groups of participants. Yet it still reveals that there may be areas of EF in which children with DS are just as competent as their TD counterparts, especially at a young age, although challenges in overall (global) EF competency are present on EF laboratory tasks in children.

In terms of the studies looking specifically at adolescents, several tools were used by Lanfranchi et al. (2010) (see Table 3.4) to investigate different EFs in DS compared to TD children, matched for MA. The results showed that adolescents with DS performed significantly worse than TD children in all EF tasks including verbal, visuospatial and dual WM tasks, inhibition tasks, sustained attention, set shifting and conceptual shifting tasks, and planning/problem-solving (PO) tasks. The areas of greatest difficulty were found to be WM, planning and conceptual shifting, in line with all previous studies investigating these EFs. However, no difference between the groups was observed in Lanfranchi et al.’s (2010) study in the performance on tasks to measure verbal fluency, where participants are asked to produce as many words as possible belonging to the same letter or semantic category.

Similarly, specific EFs (PO and verbal/visual WM) were also investigated by Chen et al. (2014). They used the Corsi-Block tapping test to assess visual WM, an auditory memory span test to assess verbal WM, and the Tower of London test to assess PO ability. The results showed that the performance of
adolescents with DS was significantly poor relative to performance in the typical population on the Tower of London task (their mean score was equivalent to the performance of TD children with the CA of 8 years). These findings corresponded with earlier studies that used the BRIEF to assess PO in children with DS (Memisevic & Sinanovic, 2014; Daunhauer et al., 2014; Lee et al., 2011). However, Chen et al. (2014), whilst providing mean WM results, do not compare these results with TD individuals of a similar CA or MA; therefore, whilst the performance of adolescents with DS was deemed by these researchers to be ‘poor’ in these areas, it is difficult to draw conclusions as to whether their performance was significantly worse than that of the TD population.

For mixed-age groups, Landry, Russo, Davkins, Zelazo and Burack (2012) researched WM and cognitive flexibility EFs in individuals with DS and WS. They used the Dimensional Change Card Sort task to assess cognitive flexibility and the Self-Ordering Pointing task to assess WM. The sample was comprised of children, adolescents and young adults (with an age range of 8-21 years) with either DS or William syndrome (WS). The results, which were not separated into age groups (therefore it is difficult to assess how children, adolescents and young adults may have differed from each other) showed that participants with DS performed slightly worse on the Dimensional Change Card Sort (cognitive flexibility) than those with WS, yet were deemed to have a slightly higher WM span (3-4 items compared to 2-3 items in the WS group) as recorded by the Self-Ordering Pointing task. However, the authors do note that comparisons could not be accurately made between the two groups given that they were not matched for MA.

Similar to Landry et al. (2012), Borella et al. (2013) investigated WM, this time with inhibition instead of cognitive flexibility. They employed the Verbal Dual Task (Lanfranchi et al., 2004) to assess verbal WM, and results showed that
children and adolescents with DS performed significantly worse than TD children matched for MA. These authors link this to inhibition, postulating that verbal WM is weak in individuals with DS as they struggle to suppress information that is no longer relevant. The animal Stroop task, used to assess inhibition, revealed the DS group performed significantly less well than the MA-matched TD children, making more mistakes, indicating general inhibitory difficulties.

Costanzo et al. (2013), like Landry et al. (2012), also assessed multiple EFs (both visual and verbal domains) in children and adolescents with DS compared to children and adolescents with WS and a TD group, all matched for MA, using various tools (see Table 3.4). Results showed that children and adolescents with DS demonstrated significantly lower ability in inhibition, attention, shifting and WM compared to the TD group, while there was no difference in PO accuracy (although the DS group took significantly longer in execution time compared to both the TD and WS groups). Both DS and WS groups demonstrated difficulties in EFs, although participants with DS were poorer in shifting (similar to Landry et al., (2012) and Memisevic and Sinanovic (2014)) and verbal aspects of memory (unlike Landry et al., 2012) and inhibition than those with WS. This indicates some commonalities between the two groups with ID compared with the TD group, although there were some distinct EF differences characterising those with DS and WS, despite being matched for MA.

In the same manner, Carney et al. (2013) compared EF measures (WM, fluency, inhibition and set shifting) (for tools see Table 3.4) in children and adolescents with DS or WS with TD children, matched for MA. They found that individuals with DS displayed relative (significant) difficulty in the domain of verbal set shifting and performed significantly worse than the TD group on both verbal and visuospatial Executive-Loaded WM. However, they did not find significant differences between DS and TD groups in inhibition and fluency tasks, contrary to previous studies that have analysed these EFs on groups matched for MA.
(Costanzo et al., 2013; Lanfranchi et al., 2010). Carney et al. (2013) however did not directly compare WS and DS groups, remaining focused on comparing DS and WS groups with TD children.

Summary
In sum, the studies exploring multiple EFs in those with DS indicate that these individuals experience significant weaknesses in various EF domains. Similarly to the studies researching global EF in the previous section, the generalised findings from studies investigating multiple EFs indicate that, compared to TD individuals matched for CA, individuals with DS demonstrate greatest difficulties on WM, followed by PO tasks (Amadó et al., 2016; Chen et al., 2014) and inhibition (Amadó et al., 2016; Schott & Holfelder, 2015). This might be expected, given that matching on CA would mean a greater discrepancy between the MA of TD and DS groups and therefore delays and difficulties in various EF are not wholly surprising. Yet the fact that WM is one of the weakest EFs supports findings in the previous section exploring global EF.

However, there are some discrepancies when comparing individuals with DS and TD individuals matched for MA. Carney et al. (2013) only found differences between MA-matched DS and TD groups in verbal set shifting and WM, not inhibition and fluency tasks, and Roberts and Richmond (2015) found no difference between preschool age groups in WM, inhibition and set shifting. This compares to findings from Lanfranchi et al. (2010), who showed significant difficulties in adolescents with DS compared to MA-matched TD children in WM, PO, conceptual shifting, inhibition, sustained attention and set shifting (with worst performance in the first three EFs). Similarly, Costanzo et al. (2013) found significantly lower abilities in WM, inhibition, attention and shifting in MA-matched DS and TD children and adolescents.
3.4.3 Working Memory

Studies that investigated global EF in children with DS, with measures that contained a WM measure, such as Lee et al. (2011), Daunhauer et al. (2014), Pritchard et al. (2015) and Lanfranchi et al. (2010) (tools and controls mentioned previously) have shown that participants with DS have difficulties in WM compared to controls and normative data. In this review, 14 studies were found that investigated WM in DS, with three focusing on children (Carretti & Lanfranchi, 2010; de Santana et al., 2014; Estigarribia, Martin & Roberts, 2012), two on adolescents (Edgin et al., 2010; Trezise et al., 2014), and the rest (n=9) in a mixed age group of participants (Borella et al., 2013; Carretti et al., 2013; d’Ardhuy et al., 2015; Fortunato-Tavares et al., 2015; Kogan et al., 2009; Lanfranchi et al., 2004; 2009a; Pezzuti et al., 2018; Visu-Petra, Benga, Tincas & Miclea, 2007).

Studies investigating WM in those with DS have used a range of tools and participant ages and have matched them either with TD controls on the basis of MA and CA or compared them to normative data, which does provide a challenge when drawing comparisons. Pezzuti et al. (2018) for instance found that 128 children and adolescents with DS had lower scores on the WM index subscales on the WISC-IV (digit span and letter-number sequencing) compared to any of the other subscales. This corroborates previous studies indicating that the WM is a specific area of weakness, yet this study did not explicitly compare the findings to TD controls. Rather, the WISC-IV has normative levels of performance in TD populations, thus providing a means of comparison. de Santana et al. (2014) also established from a subscale of the WISC-III that assessed WM generally, that children with DS had lower scores than TD children matched for CA (mean age of 8.8 years), although it was unclear from this study if this difference was significant.
Some studies appear to indicate that some areas of WM provide more difficulty to those with DS compared to other areas. Overall, studies that focused on WM evidenced that children with DS demonstrated difficulties compared to CA-matched TD children in verbal and visuospatial tasks (Lanfranchi et al. 2004; Visu-Petra et al., 2007). Amadó, Serrat and Valles-Majoral (2016), as previously discussed, employed the Frog task for visuospatial WM, and found that performance on the task was significantly lower in children with DS compared with a CA-matched TD control.

Other studies have matched participants with DS and TD individuals on MA to ascertain specific and/or generalised WM differences. Visuospatial WM was investigated by Visu-Petra et al. (2007), Carretti, Lanfranchi and Mammarella (2013), Lanfranchi et al. (2009), Carretti and Lanfranchi (2010) and d’Ardhuy et al. (2015). Carretti and Lanfranchi (2010) explored spatial simultaneous WM and found that children with DS performed significantly worse in all WM tasks (random and structured conditions). They also exhibited a significantly greater decline in performance as a result of increased WM load compared to TD children, whose performance remained substantially stable across four levels of memory load.

Other studies matching individuals with DS on MA to TD samples involved mixed-age samples. Visu-Petra et al. (2007), for example, using the CANTAB, found that 25 children, adolescents and young adults (however, adults are not the main focus in this review) with DS (age range: 8-21 years) recorded significantly lower performance when compared to 25 TD children matched for MA in most tasks/subtests measuring visuospatial WM (VSWM). The strategic self-ordered task however, measuring VSWM, revealed that whilst the number of errors made by participants with DS were significantly greater than the TD participants, their search strategy was the same.
Carretti, Lanfranchi and Mammarella (2013) employed the VSWM task in assessing WM in 20 children and adolescents with DS and MA-matched TD children. The outcome of their study showed that children and adolescents with DS presented with greater difficulties in the spatial-simultaneous element of VSWM, although their performance did not differ on the pattern configuration of the spatial-sequential VSWM task. The same conclusion was drawn by Lanfranchi et al. (2009a), who used a battery of four VSWM tasks (see Table 3.4) to compare VSWM in 34 children and adolescents with DS compared to 34 TD children, matched for MA. The results showed that those with DS performed significantly worse than the TD group in the spatial-simultaneous tasks, but not in the spatial-sequential tasks (in which, in the coding subtest, the DS group actually performed better than the TD group). This is also therefore, similar to Carretti, Lanfranchi and Mammarella’s (2013) study, indicates that VSWM weaknesses in individuals with DS are selective rather than pervasive.

Studies included in this review investigating visual WM also focused solely on those with DS that were of adolescent age upwards (instead of a mixed sample of children and adolescents). The outcome of the CANTAB assessment by d’Ardhuy et al. (2015) for example showed that 24% of the adolescents (and 12% of adults) with DS scored 0 in the reverse task in the spatial span subtest (SSP), which indicates a significant weakness in this particular domain (the mean score for TD individuals is 30 on this task). The poor overall performance of the DS group on the CANTAB SSP (especially the reverse SSP) significantly correlated (moderate effect) with the WM scores the participants with DS gained on the BRIEF-P WM scores, which further highlights challenges in this area for those with DS (especially adolescents).

Similarly, Kogan, Cornish, Graham, Berry-Kravis, Drouin and Milgram (2009) investigated VSWM and visual-perception WM abilities in adolescents with DS, yet they compared them to those with Fragile X Syndrome (FXS), as well as TD
children matched for MA and TD adolescents matched for CA. They utilised a modified version of a tool referred to as the Wisconsin General Test Apparatus to investigate both VSTM and visual-perception WM abilities, revealing that both ID groups demonstrated relative strengths on a visual-perceptual WM task and weaknesses on a VSTM task, compared to both MA- and CA-matched TD control groups. Furthermore, whereas individuals with DS performed relatively poorly on visual-perceptual and visuospatial reversal learning tasks, they showed relative strengths in egocentric spatial learning and object discrimination tasks, performing significantly better than the FXS group and as well as the MA- and CA-matched TD groups. The researchers suggested that this may have been due to the relatively low WM load of the object discrimination learning task. The findings of their study concurred with those of Carretti et al. (2013) and d’Ardhuy et al. (2015) (who found individuals with DS have a significant weaknesses in reverse spatial tasks), indicating that individuals with DS have difficulties in certain elements of VSTM only.

Verbal WM only was investigated by both Borella et al. (2013) (discussed in the multiple EFs section) and Estigarribia, Martin and Roberts (2012). Estigarribia, Martin and Roberts (2012) used the WM test known as the comprehensive test of phonological processing to compare children with DS to participants with either FXS, FXS with ASD, or TD children, all matched for non-verbal MA. The results indicated that all three ID groups performed significantly worse than TD children on the test, although the FXS with ASD group performed slightly worse than children with DS.

Thus, there are a variety of studies that assess either verbal or visuospatial WM in individuals with DS. However, there does appear to be a distinction between the relative strengths and difficulties in visual and verbal WM in those with DS. Many studies looked at both aspects of WM to ascertain which was weaker in participants with DS. For example, as discussed in the previous section
regarding multiple EFs, Lanfranchi et al. (2010) used a battery of measures from previous studies (for tools used see Table 3.4) to assess WM in adolescents with DS, comparing them with MA-matched TD children. They found that the adolescents with DS performed significantly poorer on both the verbal WM as well as VSWM and dual WM tasks. From these researchers’ findings, it can be established that adolescents with DS have weaknesses in their verbal and visuospatial WM.

This assertion was supported by earlier research from Lanfranchi, Cornoldi and Vianello (2004), which conducted two studies, the first assessing verbal WM (4 tasks) and the second investigating VSWM (5 tasks) (see Table 3.4). Each study involved a separate group of children and adolescents with DS (to avoid over-taxing individuals with DS by studying them twice) and compared them to TD children, matched on MA. The tasks in each study increased in difficulty (‘control’). Thus, the researchers distinguished between ‘low control' WM tasks (which included simple operations, such as reversing the word order), and ‘high control' tasks (which required remembering pathways (visuospatial) and selective word recall (verbal)). Results showed that, in tasks involving low control, children with DS performed significant more poorly specifically on verbal WM tasks (not VSWM) when compared to TD children matched for MA. However, the DS group in study 2 demonstrated increasing difficulties in VSWM tasks that required higher controls relative to TD children. Thus, individuals with DS were found to show significant weaknesses in both verbal and VSWM tasks as the requirement for control increased, although it must be noted, the DS groups showed less overall difficulty in the VSWM tasks than the verbal tasks, indicating greater difficulty in verbal WM.

Various studies have explored WM in general (and specific elements of WM) in DS compared with a number of other developmental disabilities. For example, as noted in the previous section, Landry et al. (2012) used Self-Ordering
Pointing to assess VSWM. The results showed that children and adolescents with DS recorded better performance than children with WS. Other studies comparing groups of individuals with DS to those with other ID include Fortunato-Tavares et al. (2015). This study compared verbal WM in children and adolescents with DS, those with typical language development (TLD), those with specific language impairment (SLI), and high-functioning autistic (HFA) children and adolescents, all of a relatively similar CA. The researchers found that participants with DS performed worse than all 3 other groups on the short WM condition, performed equally poorly with SLI group on long WM condition and did not demonstrate a WM load effect (given that their performance on simple tasks was already low, and remained so). By comparison, the other groups performed better when WM load/demand was reduced, demonstrating a WM load effect as tasks increased in complexity. This finding is in line with that of Lanfranchi, Cornoldi and Vianello (2004), who indicate that verbal WM presents challenges for those with DS even in relatively simple, low load tasks. However, Fortunato-Tavares et al.’s (2015) study can be criticised given that it did not compare participants based on MA, but on CA, which was similar between the groups, although the participants were not ‘matched’. This may mean that other variables, such as differences in MA, may be to blame for the relatively poor performance of the DS group on the verbal tasks.

Trezise, Gray, Taffe and Sheppard (2014) researched the performance of adolescents with DS, adolescents with non-specific ID (NSID) and autistic adolescents, all with a similar MA and CA, on verbal and visuospatial WM tasks. The results of their research showed that adolescents with DS recorded significantly poor performance on both verbal and visuospatial tasks compared to the NSID, and worse (although not significant) performance compared to the ASD group. Trezise et al. (2014) did acknowledge the trend for individuals with DS to show stronger performance in visual versus auditory tasks, in line with research from Lanfranchi, Cornoldi and Vianello (2004). However, this trend was
not significant, which the researchers assert may be due to the small sample size (15 individuals with DS).

In a similar vein to Trezise et al.’s (2014) study, Edgin et al. (2010) utilised a Corsi span task along with the TAPS-R backward auditory number memory test to evaluate both verbal and visuospatial WM in adolescents with DS as well as adolescents with WS matched for CA and IQ, to enable group comparisons. The researchers established that there was no significant difference in the WM performance (verbal or visuospatial) of either group.

**Summary**

More studies in this review examining WM in DS populations appear to match these individuals on MA when comparing WM functioning with that of TD populations, in order to provide a more accurate assessment of their WM functioning (than comparisons based on CA). Most studies appear to contain mixed samples, with either children and adolescents, or a wide age range, from children to young adults. The studies, in general, reveal that individuals with DS have significant difficulties in both visuospatial and verbal WM (e.g., Visu-Petra et al., 2007); however, significantly poorer performance was found in verbal WM (Borella et al., 2013; Fortunato-Tavares et al., 2015) than in VSWM. Indeed, some studies reveal those with DS possess relative strengths in VSWM, especially when either the WM load is low (Lanfranchi et al., 2004) or in specific areas, such as egocentric spatial learning and object discrimination (Kogan et al., 2009) and spatial-sequential tasks (e.g., Carretti et al., 2013; Lanfranchi et al., 2009).

Assessment of the studies examining WM in children with DS reveals that these individuals perform significantly worse in WM tasks when contrasted to TD control groups, matched for either MA (Estigarribia et al., 2012) or CA (de Santana et al., 2014). However, research into the WM of samples of children
with DS is lacking, given that only three studies in this review solely focused on this age group. Whilst Estigarribia et al. (2012) found children with DS have poorer verbal WM than TD children, de Santana et al. (2014) did not differentiate between verbal WM or VSWM and did not state whether the poorer performance of children with DS was significantly worse compared to their TD, CA-matched peers.

There is also a lack of studies specifically reviewing the performance of adolescents with DS on WM tasks; those studies that do exist compare adolescents with DS to CA- and MA-matched (Trezise et al., 2014) or CA and IQ-matched (Edgin et al., 2010) peers with other ID, such as NSID and ASD (Trezise et al., 2014) or WS (Edgin et al., 2010). This may be because it is less illuminating to compare adolescents with DS with their CA-matched TD peers, given the expectancy of significant weaknesses in all areas of WM between the two groups in adolescents with DS.

3.4.4 Attention Function

Twelve studies in total focused on exploring the attentional capabilities of individuals with DS. Six studies investigated attention in children with DS (Ashworth et al., 2013; Brown et al., 2003; Kirk et al., 2017; Lewis & Brooks-Gunn, 1984; Schott & Holfelder, 2015; Shalev et al., 2019), whilst one specifically focused on adolescents (Lanfranchi et al., 2010). Five studies involved a mixed-age group of children and adolescents (Breckenridge et al., 2013; Faught et al., 2016; Goldman et al., 2005; Purser et al., 2015; Trezise et al., 2008) to study attention as an EF in those with DS. Attention has often been studied as either selective or sustained attention, although some do not explicitly make this distinction, and can encompass tasks measuring visual or auditory attention, or both (e.g., Trezise et al., 2008).
In terms of the child studies, Kirk et al. (2017) explored sustained attention in 22 children aged between 5-11 with DS, and compared their findings to control groups involving children with ASD or non-specific intellectual disability (NSID). This Australian, cross-sectional study employed the Wilding Attention Battery to explore sustained attention using vigilance tasks. This involved asking participants to monitor a range of non-target distractors over time, and pointing out when target objects appeared. Kirk et al. (2017) reported that children with DS had significantly poorer vigilance and, crucially, sustained attention abilities than children with ASD and NSID. However, no significant group differences were present on parent-rated inattention; parent-report measures therefore may tell a different story in terms of a child with DS’s attentional skills, which is why most studies utilise attention tests, such as Ashworth et al. (2015).

Ashworth et al. (2015) examined both the selective and sustained attention of children with DS compared to both children with WS and TD children with a similar CA (mean age of 9 years old). They found for the TD and WS group, but not the DS group, there was a significant positive correlation between CA and reaction times for correct responses on the attention test. They also found that there was a significant positive correlation between the CA and MA and correct responses in the TD group, but not the DS or WS groups. Overall, the findings revealed that there was significant difference in attention between children with DS and their CA-matched TD peers, with children with DS demonstrating significantly greater difficulty than their TD counterparts, although they do not distinguish between selective and sustained performance in their results.

Some researchers conducted studies specifically to assess sustained attention in children with DS. For instance, Brown et al. (2003) assessed sustained attention in children with DS in comparison with TD children matched on MA, TD children matched on CA and children with WS, of a similar CA to the DS group. They established that children with DS had shorter as well as fewer periods of
sustained attention than all groups, including MA-matched TD children. A similar observation was made by Shalev et al. (2019) who, in their UK-based study, compared sustained attention in groups of children with DS and WS to different groups of TD children that were either aged approximately 3-, 5- or 7-years-old. Results from a Continuous Performance Task revealed that children with DS and WS performed significantly poorly compared to all TD groups; however, the DS group and all TD groups performed better in the second half of the task, whereas WS group performed worse in the second half. Shalev et al. (2019) therefore concluded that children with WS experience a selective difficulty in their ability to sustain attention over time, whereas children with DS have a more general performance difficulty. However, more children in the DS group did not complete the task (demonstrating a lack of engagement), which could have skewed the results somewhat. Nevertheless, results from the Continuous Performance Task did indicate poorer sustained attention in children with DS.

Other studies have also assessed sustained attention in those with DS and other developmental disorders, without finding significant differences, however. Lewis and Brooks-Gunn (1984) used a visual attention task as their assessment tool to understand how the fixation (sustained attention) of various groups of children with different disabilities changed over time, using different age groups for comparison. Infants between 3 months and 3 years old with either cerebral palsy, developmental delay or multiple disabilities were the comparison groups in this study, grouped into similar CA categories. According to their findings, children with DS looked for significantly longer at stimuli during trials as their CA increased, although, like the other participant groups, demonstrated a significant decrement in attention as the trials went on. However, there were no differences between the four groups in terms of attention paid to the stimuli in the trials.

There was only one study of sustained attention with adolescents. Lanfranchi et al. (2010) found that adolescents with DS demonstrate significant weaknesses
in sustained attention compared to TD children matched for MA. Using the Self-Ordering Pointing task, they established that participants with DS made a greater number of errors and rarely employed strategies necessary for sustaining attention, such as focusing on a particular spatial position to help solve a task (which was observed in the TD children).

In terms of the mixed-age samples in studies of sustained attention, Trezise et al. (2008) used both visual and auditory Sustained Attention to Response Tests (SART) to assess sustained attention in children and adolescents with DS in comparison to their peers with non-specific intellectual disability (NSID), matched for MA. Their findings indicated that participants with DS made significantly fewer errors in the visual modality of the SART than in the auditory version, although the reverse was true for the NSID group. Those with DS still outperformed the NSID group on auditory sustained attention, yet this difference was not statistically significant. MA was found to be associated with error rates in the visual condition, but no effect of group or MA was observed on the auditory task. This could perhaps indicate that visual sustained attention is less affected in DS than auditory sustained attention, and that increased MA reduces visual sustained attention errors.

To explore this finding further, Faught et al. (2016) used the same auditory and visual SART used by Trezise et al. (2008), yet this time comparing youth with DS (10-21 years) with TD children (7-10 years), matched for non-verbal MA as well as receptive vocabulary. The results showed that both groups performed similarly on auditory and visual sustained attention tasks, with the auditory and visual sustained attention deemed to be consistent with the developmental age of the participant. Therefore, Faught et al. (2016) did not replicate the findings of Trezise et al. (2008), that those with DS had stronger visual sustained attention relative to their auditory sustained attention. This indicates that the
auditory/visual sustained attention discrepancy within some studies in individuals with DS is an area that requires further research.

Other studies have specifically assessed selective attention in mixed samples. Goldman et al. (2005) used mixed ages (children and adolescents) to evaluate the selective attention skills of people with DS compared to TD children matched for visuospatial MA. Contrary to the findings of the majority of other studies that have assessed sustained attention, Goldman et al. (2005) found that there was no significant difference in their ‘orienting behaviour’ to information cues within the task, suggesting comparable skills in selective attention. They also learned that the reaction time (RT) for participants with DS in responding to the stimuli was significantly faster compared to MA-matched TD children. The researchers indicate that this is likely due to the link between faster RTs and CA in TD children that is often observed, and therefore may be due to the greater physical maturity of the DS group in this study, given their greater CA. This supposition however is contradicted by the findings of Ashworth et al. (2015), who found a correlation between RT and CA in TD participants, but not those with DS.

Other studies looked at attentional control, indicating the significant overlap between some EFs, such as inhibition and attention. Using a child sample, for example, Schott and Holfelder (2015) employed the trail-making test for preschool children (Trails-P) to assess attention and inhibition (as it was in Purser et al.’s (2015) study, discussed below). This study showed that children (age range 7-11 years) with DS had low attentional control and greater distraction when compared to a TD group matched for CA. Reasons for this are unclear; it could be speculated that the DS group were more likely to be distracted, affecting their attention, or perhaps the task was too difficult for this group. Either way, the findings indicate that attention, avoiding distraction and inhibitory control are related EFs that can be difficult to extricate in measures of EF, and that all are weak relative to similar aged TD children.
Purser et al. (2015) also researched a sample of DS, WS and TD children and adolescents (the WS and TD children were matched for verbal MA, although the DS subset was not matched to either, although were of a similar CA to the WS group). Using the Go/No Go task, they assessed attention and executive control during route learning, revealing that children with DS were poorer performers in Go/No Go task than participants in both control groups. However, they also found that participants with DS that had a relatively high level of non-verbal ability performed at a similar level to TD children on route learning. The researchers do note however that there was no significant correlation between reaction time (which they assert reflects the general attentional resources required for concentrating on a task) and route learning in the DS group, although there was a significant correlation between the number of errors made and route learning, which is indicative of reduced inhibitory control. It could be argued that the TD children may have experienced a lapse in sustained attention when they made errors, whereas poorer inhibition in the disorder groups (DS and WS) is the more likely cause of greater errors (Purser et al., 2015). However, since the DS group was not matched to either the TD or WS group on CA or MA, definitive conclusions regarding the attention and inhibitory differences between the three groups may only be cautiously drawn.

A study that drew on a mixed-age sample to explore all areas of attention (sustained, selective, auditory, visual and attentional control) was conducted by Breckenridge et al. (2013). These researchers developed the Early Childhood Attention Battery (ECAB), which contains 8 sub-tests, to test a wide range of attentional skills in both TD populations and groups with developmental delays and disabilities. 32 children and adolescents with DS and 32 children and adolescents with WS completed the ECAB in this UK-based study, with the findings compared to normative values. They found that in the DS group, scores on 3 of the 8 subtests on the ECAB were significantly lower than the expected
norm value of 10 for their MA. On two tests, the DS group performed significantly better than the norm for MA (auditory sustained and dual task) and on two tasks performed significantly better than their WS counterparts (auditory sustained and counterpointing). Thus, their attentional battery revealed that sustained attention, in general, was an area of relative strength. They also revealed that challenges were most apparent in visual selective attention tasks, especially those requiring selection from a complex, crowded array, which conflicts with findings from Trezise et al. (2008). This may be because Trezise et al. (2008) used a sample aged 10-21, so predominantly adolescents, compared to Breckenridge et al. (2013) (age range 5-14-years-old). It may be that difficulty with visual attention begins to increase with CA. In addition, Trezise et al. (2008) used the SART, not the ECAB; different tests may lead to different conclusions regarding auditory/visual attention skills. Furthermore, Trezise et al. (2008) compared adolescents with DS with those with NSID, not normative data; again, the comparison group may result in differences in findings.

**Summary**

In terms of attention in children and adolescents with DS, the studies that exist present some discrepancies and contradictions. Some recent studies reveal that sustained attention in particular is affected in children with DS (Ashworth et al., 2015), as children with DS have demonstrated fewer and shorter episodes of sustained attention (Brown et al. 2003), especially compared to CA-matched TD individuals (Ashworth et al., 2015; Lewis & Brooks-Gunn, 1984). Selective attention skills were found to be comparable between MA-matched children amongst DS and TD populations (Goldman et al., 2005). Yet most studies involved mixed samples of children and adolescents with DS, in order to match them on MA with other groups (i.e., TD children) without reporting on the data of the two age groups (children or adolescents) separately; thus, it is not possible to explore whether children with DS face specific difficulties compared to adolescents or adults with DS.
Sustained attention appears to have received more interest in the research literature, with some studies finding significant weaknesses in individuals with DS compared to MA-matched groups (Brown et al., 2003; Lanfranchi et al., 2010), whilst others involving a mixed sample of children and adolescents found that individuals with DS performed significantly better than expected for their developmental level/MA in certain areas, such as auditory sustained and dual tasks. Some researchers, such as Shalev et al. (2019), suggest that findings indicating a weakness in sustained attention may in fact be explained by more general performance difficulties in tests, instead of specific weaknesses in sustained attention, given the number of individuals in their DS sample that failed to complete the task.

Selective attention has received less attention within the research literature, although some studies indicate it may be less affected than sustained attention, with Goldman et al. (2005) finding no difference between participants with DS and MA-matched TD children for selective attention. Other studies present findings that contradict this, however, such as Breckenridge et al. (2013), which found that the greatest weaknesses in the DS group of children and adolescents they studied were found specifically in visual selective attention. Therefore, strengths and weaknesses in the DS attentional profile may depend on the modality (auditory or visual) of selective or sustained attention being tested.

Perhaps attention cannot be extricated from measures of control, as demonstrated by Schott and Holfelder (2015) and Purser et al. (2015). These studies highlight the lack of attentional control when comparing DS and TD CA-matched (Schott & Holfelder, 2015) and MA-matched (Purser et al., 2015) groups. Yet interestingly, reaction times have been postulated to increase with greater CA (Goldman et al., 2005; Purser et al., 2015), although this finding was contradicted by Ashworth et al. (2015).


### 3.4.5 Cognitive Flexibility

Cognitive flexibility (CF) is often a term used synonymously with set shifting or switching to describe the ability of viewing something in varied ways (Diamond, 2013). Therefore, this section combines studies exploring these skills in those with DS, despite the different terms adopted by various studies. It also assesses behavioural flexibility, as a related concept. This systematic review found that three studies assessed CF in children (Amadó et al., 2016; Roberts & Richmond, 2015; Schott & Holfelder, 2015), one study assessed CF in adolescents only (Lanfranchi et al., 2010) and three studies adopted mixed-age samples (Carney et al., 2013; Green et al., 2006; Memisevic & Sinanovic, 2014).

In terms of the studies assessing CF in children, Amadó, Serrat and Valles-Majoral (2016) (discussed in previous sections) employed the Wisconsin Card Sorting Task to assess CF and found that performance on this task was significantly lower in children with DS when compared with controls (a TD group matched for CA and a group of children with language development difficulties). The same observation was made by Schott and Holfelder (2015) in their study, as they also found that children with DS demonstrated significantly lower performance in switching than the TD group, matched for CA (like in Amadó, Serrat & Valles-Majoral’s (2016) study) and sex. By comparison, Roberts and Richmond (2015) found in their study that there was no significant difference in shifting performance between children with DS of preschool age and the TD group matched for either receptive language or non-verbal scores as a proxy for MA.

There was only one study that looked at CF in adolescents only: Lanfranchi et al. (2010) assessed a range of EFs and found that adolescents with DS recorded poor scores in all EFs compared to TD children matched for MA, including CF (measured as both set shifting (for example, switching attention
between objects) and conceptual shifting (for example, switching attention between intangible things, such as rules, categories or tasks)). In fact, whilst the performance of the DS group is reported as ‘worse’ on the set shifting task (whether this was significant is not made clear by the authors), Lanfranchi et al. (2010) do note that on a Modified Card Sorting Test (Nelson, 1976), adolescents with DS showed significantly greater challenges in conceptual shifting, when compared to TD children matched for MA.

In terms of the mixed samples, Memisevic and Sinanovic (2014) used the BRIEF and found that the shifting functioning of children and adolescents with DS was significantly worse when compared with normative data for TD children, although it is unclear whether they compared their participants with DS to this data based on MA or CA. They did, however, compare the results of individuals with DS to individuals with ID either with unknown aetiology or other genetic cause (matched on CA), finding that there were no significant group differences on the BRIEF except in shifting, where the participants with DS scored significantly higher (indicating greater difficulty) than the ID group of unknown aetiology. This indicates that CF may be an EF weakness that distinguishes DS from some other IDs. Although Carney et al. (2013) involved children and adolescents with DS or WS in their study of EFs, they compared both groups with TD children, with a similar MA. Yet they also found, similar to Memisevic and Sinanovic (2014), that individuals with DS displayed relative (statistically significant) difficulty in set shifting, although their findings were domain-specific, as they pertained to verbal set shifting only.

Another study involving a wide age range of individuals with DS in their sample was conducted by Landry et al. (2012). This study used the Dimensional Change Card Sort to evaluate the CF in mixed-aged participants (children, adolescents and young adults, 8-21 years) with DS and WS with different Mas. The results of this study showed that participants with DS had greater difficulty
with CF than participants with WS, although it is unclear if this difference was significant, given that the authors state they do not draw comparisons between the two groups as they were not matched for MA. They did find, however, that verbal development was a unique predictor of CF in both groups.

Whilst flexibility is generally explored in terms of CF, behavioural flexibility has also been explored by one study in those with DS. Green et al. (2006) explored behavioural flexibility using the parent-rated Behavioural Flexibility Rating Scale (BFRS) to compare a sample of individuals with DS of mixed ages with groups of participants with ASD and Asperger syndrome (High Functioning Autism (HFA)) (not matched on CA or MA, but also varying in CA). The results showed that, when compared in groups according to CA, participants with DS had a mean BFRS score that was significantly lower than the averages for both the ASD and HFA groups, indicating fewer difficulties. There were no sex or age differences, moreover, in the DS group, indicating the behavioural flexibility scores (according to parents) did not differ dependant on these variables.

**Summary**

Studies assessing CF in children reveal mixed results – those comparing children with DS to a TD group matched on CA reveal significant weaknesses in the DS group (Amadó et al., 2016; Schott & Holfelder, 2015), whilst the study that assessed preschool children compared to a TD group matched on MA did not find a significant difference (Roberts & Richmond, 2015). The one study that looked at adolescents with DS and their MA-matched TD counterparts found that the DS group were ‘poorer’ on set shifting, and significantly worse in conceptual shifting. The mixed sample studies also show significant weaknesses in individuals with DS on CF, compared to normative data (Memisevic & Sinanovic, 2014) and some groups of individuals with ID of unknown aetiology (Memisevic & Sinanovic, 2014).
Yet Carney et al. (2013) in their mixed-age sample study found that individuals with DS were only significantly poorer in verbal set shifting (not visuospatial set shifting) compared to a MA-matched TD group. This indicates that difficulties in CF may be modality specific, with conceptual and verbal set shifting providing greater difficulties than visuospatial CF difficulties, although this should be explored further to provide any definitive conclusions on that front. Finally, behavioural flexibility was found to be less of a challenge in children and adolescents with DS compared to individuals with other disorders, such as HFA and ASD.

3.4.6 Self-Regulation

There were only four studies that specifically assessed self-regulation in populations of individuals with DS. The dearth of studies in this area could be due to the close ties between self-regulation and inhibitory control, with the greater number of studies being focused on inhibitory control as a related function. Two of the reviewed studies focused on self-regulation in children with DS (Bieberich & Morgan, 2004; Gilmore et al., 2003), whereas two focused on a mixed sample of children and adolescents (Cuskelley et al., 2003; 2016). Interestingly, all studies matched individuals with DS to either TD or other groups based on MA.

In their study, Gilmore et al. (2003) used the Goodman Lock Box task (Goodman, 1981) to assess self-regulation in children with DS, compared to a TD group matched for MA as the control. They established in their cross-sectional, observational study that both groups demonstrated similar performance in the task in terms of competence, playfulness and distractibility. They do however note that the DS group showed more task-avoidant behaviour, indicating either a sustained attention weakness, greater distractibility, helplessness after repeated failures in the task or the inability to structure their activities independently.
To investigate differences in self-regulation between verbal MA-matched children with DS or ASD, Bieberich and Morgan (2004) used The Minnesota Preschool Affect Rating Scale (MN-PARS) (Shapiro et al., 1994), together with a behaviourally-anchored rating scale, analysing four elements of self-regulation (attention, object orientation, persistence and adaptability) over 2 years. They found that children with ASD received significantly worse scores than children with DS on 3 of the 4 self-regulation factors (attention, object orientation and persistence) over time, with the fourth scale (adaptability) approaching significance. This indicates that children with DS have better self-regulation skills than children with ASD. Moreover, the self-regulation behaviours of children with DS were more stable over the different time points compared to those with ASD.

Two studies investigated mixed-age groups. Cuskelly et al. (2003) researched self-regulation in children and adolescents with DS, using several tools to test delay of gratification, ascertaining whether children and adolescents with DS would wait for a similar length of time compared with a MA-matched group of TD children. The results however, contrary to Gilmore et al.’s (2003) study, indicated that the DS group were significantly less able to self-regulate (through an inability to delay their gratification) than TD children. In addition, according to parent ratings, TD children were found to have significantly greater self-control than the children and adolescents with DS. This suggests that participants with DS have reduced self-regulation skills than MA-matched TD children. These findings were further replicated by Cuskelly et al. (2016), who also found that children and adolescents with DS had significantly greater weaknesses in delay of gratification and therefore self-regulation compared not only with MA-matched TD children, but also compared to individuals with moderate ID.
Summary
From the review of studies that assessed self-regulation in children with DS, one indicated that children with DS have better self-regulation compared to children with ASD, and one indicated that children with DS do not demonstrate self-regulation difficulties compared to an MA-matched TD group. However, both the two studies containing mixed groups of children and adolescents with DS found that the DS group demonstrated significant weaknesses in delaying gratification and in self-control, compared to TD children matched for MA.

Thus, it could be speculated that the addition of adolescents to the sample generated results that denote significant difficulties for the DS group. However, as the two studies involving mixed samples (Cuskelly et al., 2003; 2016) did not separate their results to discern any differences between children and adolescents, it is difficult to support this speculation with data. Clearly, greater research is required regarding self-regulation in children and adolescents with DS compared to TD individuals to untangle the contradictions within these findings.

3.4.7 Inhibition and Impulsivity
This section reviews studies that investigated inhibition and impulsivity in children, adolescents and mixed-age groups with DS. Three studies investigated inhibition in children with DS (Amadó et al., 2016; Schott & Holfelder, 2015; Roberts & Richmond, 2015), one study only investigated adolescents (Lanfranchi et al., 2010), whilst five studies investigated this function utilising a mixed-age group (Bhattacharyya et al., 2009; Borella et al., 2013; Johns et al., 2012; Purser et al., 2015; Traverso et al., 2018).

Compared to TD children of a similar CA, children with DS were found by Schott and Holfelder (2015) to have significantly poorer performance in all tasks involving inhibitory control. This was also found by Amadó, Serrat and Valles-
Majoral (2016) who, using the Day and Night task, revealed significant difficulties in children with DS compared to CA-matched TD children and children with language development difficulties.

However, unlike previous studies comparing children with DS to TD children matched on the basis of CA, Roberts and Richmond (2015) found in their study that there was no significant difference in inhibition performance in the A-not-B task between children with DS of preschool age and the TD group matched for either receptive language or non-verbal scores as a proxy for MA. This indicates that inhibition differences are magnified between DS and TD groups when compared by CA, although matching on MA, especially at preschool age, may not reveal significant differences in inhibition between groups.

In terms of studies looking purely at adolescents with DS, Lanfranchi et al. (2010) however, contrary to Roberts and Richmond’s (2015) findings, indicated that adolescents with DS performed significantly worse on the Day and Night task (like Amadó et al., 2016) when compared to MA-matched TD controls. Thus, perhaps MA-related difficulties in inhibition increase with CA (as MA-matched preschoolers in Roberts and Richmond’s (2015) study did not show significant differences whereas MA-matched adolescents with DS in Lanfranchi et al.’s (2010) study did).

In terms of the studies employing mixed-age groups, Borella et al. (2013) explored three inhibition-related functions: 1) prepotent response inhibition (this enables prepotent and dominant motor or cognitive responses that are activated automatically by a stimulus to be blocked); 2) response to distracter inhibition (the ability to focus on relevant information by blocking simultaneously presented irrelevant items); and 3) resistance to proactive interference (being able to prevent the activation of items that are no longer relevant and thus reduce memory intrusions (intrusion errors)).
They compared the performance of children and adolescents with DS to TD children matched for MA, on a range of tasks assessing the three inhibition-related functions and found that the DS group performed significantly less well than the TD children on all three measures, indicating a general inhibitory weakness. This weakness in a mixed-age sample was also found by Purser et al. (2015), who researched samples of children, adolescents and young adults with DS and WS, matching them to TD children on non-verbal MA, using the Go/No Go task to assess executive control (inhibition). The results showed that individuals with DS performed significantly more poorly in Go/No Go tasks than children in both control groups, making more errors as a result of poorer inhibition.

The findings of Borella et al. (2013) were somewhat replicated by Traverso et al. (2018), who compared the performance of children, adolescents with DS on a battery of tasks assessing inhibition to a group of TD children aged 5 (TD5) and a group of TD children aged 6 (TD6). They found that children from the DS and TD5 groups performed similarly, whereas children from the TD6 group significantly outperformed both the other groups. This indicates that inhibition development in those with DS may match developmental age. However, there was variability in the scores from different tasks used to measure inhibition; those with DS outperformed both TD5 and TD6 groups in the Flanker task in terms of accuracy and response time, yet exhibited significantly greater scores (performed worse with a longer reaction time) than the TD5 group on the Preschool Matching Familiar Figure Task. Thus, there was an inconsistent pattern across different measures, perhaps as a result of the non-executive abilities required to complete different tasks or verbal versus visual stimuli. However, in general, those with DS performed significantly more poorly across most (if not all) inhibition tasks.
Whilst the above studies matched mixed-age samples on MA, Bhattacharyya et al. (2009) used a range of ages (child-adult) in their study, comparing those with DS to TD individuals and a group of individuals with other ID with a similar CA (not matching). They employed a cross-sectional survey method using the Diagnostic Assessment for the Severely Handicapped (DASH II). This revealed that just over half of the child and adolescent participants with DS were rated as having high impulsivity, which was significantly greater than the TD group, although not as high as the group of individuals with other ID. If impulsivity denotes a lack of inhibitory control, this indicates greater difficulties in inhibitory control in those with DS compared to TD individuals.

Other studies that investigated inhibition included Wishart (1987) and Johns et al. (2012). Wishart (1987) used Piagetian infant search tasks to assess inhibition over six sessions. The study compared infants with DS and TD children, matched for gender, number of siblings and birth order (not MA or CA). The results showed that children in the DS groups were significantly less competent on the tasks than those in the TD groups in the first session (although they significantly improved, unlike those in the TD group, who demonstrated more consistent performance). Furthermore, Johns, Homewood, Stevenson, and Taylor (2012) used reverse categorisation tasks known as the ‘hand’ and the ‘animal’, which can assess inhibition (as well as WM, as the participants must remember the rules of the game) (Carlson et al., 2004). They compared the performance of children and adolescents with DS to TD children matching for MA, as well as participants’ siblings and undergraduate students as the control groups. They found that the participants with DS demonstrated significantly lower inhibition ability when compared with all groups, including the MA-matched TD children.
Summary

An analysis of studies assessing inhibition reveals that both children and adolescents with DS perform more poorly in tasks measuring inhibition and impulsivity than TD children matched for either CA or MA. All studies in this review, except one (Roberts & Richmond (2015), assessing inhibition in preschoolers (DS and TD groups) matched for MA), revealed that performance was worse on a range of tasks in both the child, adolescent and mixed-age sample groups. Perhaps Borella et al.’s (2013) study is of particular interest here, as they uniquely break inhibition down into three components (prepotent response inhibition; resistance to proactive interference; and response to distractors inhibition), revealing that their mixed-age sample of individuals with DS performed significantly worse on all three areas of inhibition compared to their MA-matched TD peers.

3.4.8 Planning/Organising

Eight studies that investigated planning/organising (PO) (often just named ‘planning’ in the studies) are reviewed in this section: five investigated PO in children with DS (Daunhauer et al., 2014; Joyce et al., 2019; Lee et al., 2011; Molina & Perez, 1993; Pritchard et al., 2015), two explored this in adolescents with DS (Chen et al., 2014; Lanfranchi et al., 2010), whereas one (Costanzo et al., 2013) employed a mixed-age sample. Some focused primarily on PO, whereas others included this EF whilst exploring multiple or global EFs.

Of the studies looking primarily at child samples, Molina and Perez (1993) compared information processing of children with DS with children with ID (with a similar MA and CA) as well TD children with a similar MA. Using 3 PO tests from the Dynamic Assessment of Learning Potential and Cognitive Strategies Battery, they found that, without mediation (cues and prompts from pre-test to post-test to help participants with the answers), children with DS demonstrated significantly lower performance in PO processes when compared to both groups
(Molina & Perez, 1993). Other studies involving child samples also found significant weaknesses in PO, such as Lee et al. (2011) and Daunhauer et al. (2014), who both used the BRIEF-P, compared to MA-matched controls. Interestingly, however, the PO scores of children with DS without co-occurring conditions on the BRIEF-P were not found by Pritchard et al. (2015) to be significantly different to groups of children with DS+ASD or DS+DBD, although those with DS without co-occurring conditions did perform better in all EFs more generally (just not PO).

Joyce et al. (2019) assessed PO using the Tower of Hanoi in children with DS, WS and a TD sample, all with similar mean CA (between 9-10-years old). Their aim was to investigate the impact of sleep on test performance; they found that, in general, DS and WS groups made significantly more rule violations on the TOH task compared to the TD group. Another study using a similar task, the Tower of London (TOL) task, also revealed similar findings. Lanfranchi et al. (2010) looked at the EF of PO as part of an investigation into a range of EFs in adolescents with DS, comparing them to TD children matched for MA, finding significantly poorer performance in the DS group in this EF on the TOL task. In fact, PO and conceptual shifting, after WM, were found to be the EFs presenting the most difficulty in adolescents with DS. Similarly, Chen et al. (2014) also used the TOL task to assess PO in adolescents with DS, revealing that, relative to performance in the typical population, the performance of adolescents with DS was significantly poorer than what might be expected for TD individuals of the same CA (the performance was equivalent to TD children at the age of eight). However, these results differ from that of Costanzo et al. (2013), who compared performance on the TOL between a mixed-age sample of individuals with DS, a TD group matched for MA and a group with WS, with a similar MA and CA. They found that there were no significant differences between participants with DS and the TD group on accuracy on the Tower of London; there was, however, a significant difference in execution time, with the DS group taking significantly
longer on the task than both the WS and TD groups. This may indicate that it takes longer for those with DS to execute the same level of skill in PO as a TD group.

**Summary**

An assessment of studies investigating the EF of PO reveals that both children and adolescents with DS appear to score poorly compared to TD children matched for CA or MA, either using the BRIEF-P, Tower of London/Hanoi tasks or EHPAP to assess PO skills. However, children with DS without co-occurring conditions seem to fare somewhat (although not always significantly) better than those with DS and co-occurring conditions (specifically ASD and DBD). Moreover, there was some discrepancy in the findings using the Tower of London with a mixed-age sample, with the accuracy of individuals with DS being comparable to a TD (MA-matched) group, although time taken to complete tasks was significantly longer. This could shed some light on why performance on the Tower of London test is often noted as being significantly poorer in adolescents with DS.

**3.5 Discussion**

This review indicates that, overall, children and adolescents with DS have weaknesses in all EFs (working memory (WM), planning and organisation (PO), inhibition, attention, cognitive flexibility, self-regulation, monitoring and emotional control (EC) (although this latter term is only included in the BRIEF assessment)), although the level of weakness in these domains differed depending on study, comparison group, age group and the domain itself, as some present more difficulty than others. It must be noted that no study focused specifically on emotional control; this was only explored as part of global EFs, using the BRIEF or BRIEF-P. A synthesis of studies that focused on global as well as multiple EFs revealed that children and adolescents with DS demonstrate considerable weaknesses in EF domains when compared to their
TD peers matched for MA and CA. Some ‘cool’ EFs, notably WM, appeared to be the most severely affected EF in children and adolescents with DS, given that most studies investigating global or multiple EFs in individuals with DS found more significant weakness in these areas when compared to TD individuals matched for MA or CA (e.g., Daunhauer et al., 2014; Joyce et al., 2020; Lee et al., 2011; 2015; Memisevic & Sinanovic, 2014).

There is less compelling evidence for significant weaknesses in ISCI (inhibition), FI (flexibility, including shifting) (Daunhauer et al., 2014; Lee et al., 2011; Memisevic & Sinanovic, 2014) and EC (Daunhauer et al., 2014; Lee et al., 2011; 2015) in those with DS, compared to either TD individuals (matched for CA or MA, or normative data), results of which tends to be mixed or conflicting (e.g., Daunhauer et al., 2017; Traverso et al., 2018). Furthermore, compared to individuals with other ID or those with DS and co-occurring conditions, those with DS without co-occurring conditions exhibit consistently fewer difficulties in EC and ‘hot’ EFs more generally (Lee et al., 2015; Prichard et al., 2015).

These findings correspond with the study by Loveall et al. (2017), included in the systematic review, who describe the general EF difficulties faced by individuals with DS and also the specific patterns of strengths and weakness across EFs. For children with DS aged between 2 and 5 years old, EC and shifting were relative strengths compared to other EFs, falling more in line with normative data, while for the 6–18 year-old group, only EC provided a relative strength, indicating that shifting (CF) becomes more challenging as CA increases. This could perhaps compare with the development of shifting for TD infants, which was found by Best and Miller (2010) to develop more gradually in a linear fashion. Perhaps in those with DS, shifting develops more rapidly than other EFs in early infancy (thus being in line with TD development), before slowing down significantly, at which point it appears to present significant challenges for older children and adolescents, compared to TD infants matched for CA. In Loveall et
al.’s (2017) study, for both age groups (2-5-years-old and 6-18-years-old), WM continued to provide the EF area of greatest difficulty for those with DS. Whilst PO, which is grouped in the BRIEF with WM under the EMI scale, would potentially appear to be affected to the same extent as WM on the BRIEF, other studies employing tasks to measure PO specifically reveal contradictions in their findings regarding the extent to which PO is affected in individuals with DS. For instance, Costanzo et al. (2013) reported no significant differences between participants with DS and the TD group on accuracy on the Tower of London, although the DS group took significantly longer on the task than both the WS and TD groups, suggesting it takes longer for those with DS to execute the same level of skill in PO as a TD group. Therefore, whilst PO certainly is an area of marked weakness for those with DS, it may be surmised that PO only appears to be as affected as WM on the BRIEF given its grouping with WM on this scale. In reality, tests do not reveal that PO is perhaps quite as challenged as WM is in individuals with DS.

According to Zelazo and Müller (2011), ‘cool’ EFs include nonemotionally laden functions such as WM and PO, which are central to cognitive tasks, while ‘hot’ EFs are psychological processes more influenced by emotions, such as EC and self-regulation. Some researchers (e.g., Lee et al., 2011; 2015; Memisevic & Sinanovic, 2014; Daunhauer et al., 2014) established that the WM is the EF domain presenting the greatest difficulty in children and adolescents with DS, therefore indicating greater challenges in the ‘cool’ EF domains. This is further supported by research that, using the BRIEF or BRIEF-P, found greater teacher/caregiver reports of difficulties in the Emergent Metacognition Index (EMI) (‘cool’) domains (containing WM and PO), compared to the ‘hot’ inhibitory self-control (ISCI) and EC domains (also known as the Behavioural Regulation Index (BRI)). This is supported by Watson, Gable and Morin (2016), who revealed that students with DS demonstrate difficulties in tasks such as reading comprehension and note-taking as a result of the related planning difficulties
they experience. Furthermore, the ‘cool’ EF of both sustained and selective attention was found to be significantly worse than TD groups, both in children matched for MA (Brown et al., 2003; Goldman et al., 2005) or CA (Ashworth et al., 2015; Schott & Holfelder, 2015), and adolescents matched for MA (Lanfranchi et al., 2010).

Shifting is an EF considered to be relatively free of affect or emotional ‘charge’. In this review, shifting task problems were found to be more common than EC problems in adolescents with DS (Lanfranchi et al., 2010). These findings are also consistent with Loveall et al. (2017). Children with DS also appeared to perform more poorly in shifting tasks than children with other ID (Memisevic & Sinanovic, 2014) and it has been suggested that the lack of verbal skills typical of children with DS may contribute to these weaknesses in the domain of cognitive flexibility (Carney et al., 2013). Some studies do match on this to overcome this verbal difference, however; Campbell et al. (2013) for example, using the Flexible Item Selection Task showed that the CF scores of children and adolescents with DS with a mean MA of 5 years on the Flexible Item Selection Task were positively correlated with verbal MA. This indicates that CF abilities in individuals with DS may be predicted by verbal MA (VMA). Whilst these researchers do not compare the CF scores to TD children, normative data or any other group (hence it was not included in this review), it does support the idea that some EFs, such as CF or shifting, are linked to VMA, and that this is a factor that must be controlled if more accurate comparisons are to be made to other control groups (e.g., TD or those with other ID).

However, from the reviewed studies, it can be concluded that children with DS do not experience shifting task problems to the same degree as in other functions, such as WM (Roberts & Richmond, 2015). This finding is also consistent with Loveall et al. (2017), who showed that shifting was a relative strength in children with DS compared to other EFs, although arguably still
presents a challenge for this population compared to the TD population. This may depend on the age of the individual, however; Daunhauer et al. (2017) revealed no significant difference between DS and TD groups of children matched on non-verbal MA (mean=4.2 years). Yet they explain this finding by indicating the trend towards less competence on the task employed to measure shifting (Dimensional Change Card Sort) with a moderate effect size, and parallel this with the performance trend in the literature in adolescents and adults with DS (Edgin, 2003; Rowe et al., 2006; Zelazo, Burack, Benedetto, & Frye, 1996). They also indicate that the lack of significant group differences in shifting may be due to a floor effect in the performance of the group with DS, the small sample size or the young CA of participants. This tallies with literature indicating that the development of some EFs in young children with DS may align with developmental norms in very early infancy, and that EF weaknesses become more pronounced over time (Lee et al. 2015).

Although this review did not include studies that made a direct comparison between children and adolescents with DS in terms of shifting, it appears that adolescents with DS demonstrate poorer levels of shifting ability compared to children with DS (Lanfranchi et al., 2010). This would tally with the previously discussed findings from Daunhauer et al. (2017), that young infants with DS demonstrate no significant difference in shifting ability compared with MA-matched TD peers. As Loveall et al. (2017) suggest, this could demonstrate that children of preschool age children that have DS may switch between different activities without seeming distressed (according to parent reports), although children that are older have been found to demonstrate greater difficulties with changes in plans or routines. It could be argued, however, that this apparent difference is a product of the particular environment that school creates, which can draw attention to these difficulties or, as indicated earlier, shifting skill becomes comparatively worse over time, with this difference between skill in DS
and TD groups becoming increasingly pronounced, as it does not develop as quickly as the gradual, linear skill development of TD children and adolescents.

Turning to EC, this review found that EC was the least challenged function in children and adolescents with DS (e.g., Loveall et al., 2017). This supports previous research revealing that people with DS demonstrate less difficulties in ‘hot’ EFs, compared to ‘cool’ EFs (Lee et al., 2011; Daunhauer et al., 2014). This relative strength could be due to potentially higher levels of empathy and social skills demonstrated by some people with DS, when compared against people with other ID (Buckley, 2012), although evidence supporting this is not consistent. When compared against CA-matched TD peers, however, difficulties with EC are greater (Dykens et al., 2007).

Thus, whilst the research clearly indicates a greater ‘cool’ EF/EMI weakness in those with DS, it is imperative to note that ‘hot’ EF aspects are also affected in children and adolescents with DS. Yet the research is admittedly more mixed in this field, with some researchers finding significant weaknesses (compared to MA- or CA-matched TD groups) in supposedly ‘hot’ EFs (although less significant/smaller effect size than WM and PO), such as self-regulation (e.g., Cuskelley et al., 2003; 2016, both MA-matched), compared to other ‘hot’ EFs, especially EC.

This complexity is furthered by some studies that indicate that some EFs, such as inhibition and cognitive flexibility (shifting) (Poon, 2018) do not present as much difficulty relative to other EFs, with some studies even finding similar performance or no significant differences in individuals with DS to TD children matched for either CA or MA in inhibition (e.g., Carney et al., 2013, MA-matched; Daunhauer et al., 2017, MA-matched; Roberts & Richmond, 2015, CA-matched; Memisevic & Sinanovic, 2014, normative data) and cognitive flexibility (CF) (Daunhauer et al., 2014, teacher (not parent) reports on the BRIEF-P;
Daunhauer et al., 2017, MA-matched comparisons). Some studies give explanation for this admittedly surprising finding. For example, Daunhauer et al. (2017) presented the lack of a significant difference between MA-matched DS and TD infants on the snack delay (inhibition) task, and highlighted that actually, the DS group performed better than the TD group with a moderate effect size. These researchers indicate the mixed findings, and differentiate between different types of inhibition, claiming behavioural inhibition presents difficulty for young children with DS (Kopp, Krakow & Johnson, 1983), whilst verbal response inhibition has been found to be significantly weaker in adolescents compared to their TD counterparts (Constanzo et al., 2013). However, Daunhauer et al. (2017) point out the lack of evidence for significant weaknesses in response inhibition and teens in young adults with DS (Pennington et al., 2003; Rowe et al., 2006). Daunhauer et al. (2017) utilised a behavioural inhibition task, yet used both visual and auditory cues to support the task, by placing a bell on a table, which was rung when participants were allowed to retrieve the snack. Daunhauer et al. (2017) admit the possibility that this bell played the role of a cue or distractor for infants with DS and allude to previous research indicating that visual perceptual skills are an area of relative strength for people with DS (Daunhauer & Fidler, 2011). Given this, the performance of those with DS on the snack delay task in Daunhauer et al.’s (2017) study could have been supported, although note that greater research is required on inhibitory control development in those with DS.

However, the picture is complicated by the different studies that have found significant differences in inhibition in individuals with DS compared to MA-matched TD peers (Borella et al., 2013; Costanzo et al., 2013; Lanfranchi et al., 2010; Purser et al., 2015) and CA-matched TD peers (Amado et al., 2016; Bhattacharyya et al., 2009; Schott & Holfelder, 2015). In addition, more studies have found significant weaknesses in CF compared to MA-matched TD children (Lanfranchi et al., 2010); CA-matched TD peers (Amadó et al., 2016; Schott &
Holfelder, 2015) or normative data (Memisevic & Sinanovic, 2014), with verbal set shifting (Carney et al., 2013) and conceptual shifting (Lanfranchi et al., 2010) domains faring significantly worse. This indicates that there is no binary finding that all ‘cool’ EFs are relatively weak and all ‘hot’ EFs are a relative strength in those with DS. Rather, the results generally indicate that all EFs are challenged to some degree, with more agreement in the research that WM is the worst affected EF, and EC in particular is one of the least affected EFs, but less agreement regarding the extent of the weaknesses and challenges in inhibition, self-regulation and CF.

Moreover, features of some EFs, such as problem-solving, were not exclusively explored in the research studies involved in this systematic review; as noted, problem-solving appears to be less of an EF in its own right and more imbued in the functioning of other EFs, such as WM. For instance, Drigas and Karyotaki (2019, p. 76) assert that, ‘problem solving is tied to metacognitive awareness processes, necessary for applying executive function skills in goal setting and decision-making situations’. However, it is not an EF per se; it forms a bidirectional relationship with EF, in that EFs are critical for problem-solving ability, and problem-solving is an inherent part of various EF tasks (Rapado-Castro et al., 2019). Camp et al. (2016), in a bid to establish the possible mechanisms underpinning success or failure on problem-solving tasks in everyday scenarios using the Problem-Solving Questionnaire (PSQ). On the PSQ, TD children (mean CA=8.3 years old) scored significantly higher than DS and WS adolescents, although the DS group scored significantly higher than the WS group. The findings indicated therefore that adolescents with DS, whilst demonstrating challenges with problem-solving comparative to TD children, were not as challenged in their problem-solving EF abilities as individuals with WS. Camp et al. (2016) also found more significant relationships between parent-rated EFs on the BRIEF and reaching the solution to a problem-solving task for individuals with DS and WS compared to TD children (on 12 and 10 out
of 16 possible correlations respectively, compared to only one (inhibit) for the TD sample). These findings suggest that adolescents with DS and WS are able to solve everyday problems depending on the level of weakness in their other EFs. Therefore, problem-solving, is inherently linked to EF and generally embedded within different tasks measuring EF (Diamond, 2013).

Yet research highlights that the strategies that those with DS may use to solve problems differ to those of the TD population (e.g., Lanfranchi et al., 2010), although this may be due to less engagement with and focus on a task. For example, the fact that sustained attention is affected in those with DS can mean that those with DS fail to stay on-task (e.g., the Goodman Lock Box task in Gilmore et al.’s (2003) study), rendering it difficult to ascertain precisely what these problem-solving strategies might entail.

Thus, whilst there is some discrepancy in the findings, a generalised picture does begin to emerge; as noted, both children and adolescents with DS appear to have the most significant weaknesses in WM and the least problems with EC.

3.5.1 Possible Explanations for Executive Function Difficulties
This section discusses various potential explanations for the weaknesses in EFs experienced by those with DS, as well as why these weaknesses may be found in EF measures. Morton’s (2004) developmental causal model, outlined in chapter two as the conceptual model for this study, suggests that it can be helpful to assess development disorders and difficulties from the perspective of biological causes, cognitive difficulties, behavioural influences and environmental context and responses. It may be, for example, that the responses of others (for example, peers) to those with DS (such as being very helpful towards them) may mean that individuals with DS have to make fewer demands on their EF skills. The idea that those (especially children) with DS are treated differently and preferentially as a result of their facial features, for example, was
found by Fidler and Hodapp (1999); these researchers revealed that babyfaced children with DS were more subject to overgeneralisation of babyish/immature personalities (and therefore more likely to be assisted more). This could mean that EF development is delayed because caregivers, peers and adults provide fewer opportunities for individuals with DS to challenge and improve their EFs through scaffolding (Yussof & Zaman, 2011).

It is possible that reduced opportunities for scaffolding may stem from language delays in children with DS (Abbeduto, Warren & Connors, 2007), which may prevent them from either understanding a more knowledgeable other/teacher, or prevent them from communicating their understanding so that the teacher is happy to continue with the lesson (Abbeduto & Murphy, 2004). Given the importance of language and social interaction for learning, it is possible that a diagnosis of DS can lead to delays and difficulties in language, which then limits children’s opportunities for scaffolded learning, consequently negatively affecting their EF ability. This explanation may be supported by the idea that the development of some EFs in children with DS may not be significantly different to the development of EFs in TD children (e.g., Daunhauer et al., 2017, who found no significant differences in CF and inhibition in young children with DS compared to TD peers). By the time a child with DS begins formal education, it could be that their learning is not sufficiently scaffolded, especially given preconceptions regarding the learning ability of those with a SEN (Demetriou, 2020; Enea-Drapeau, Carlier & Huguet, 2017; Takriti, Atkinson & Elhoweris, 2019). Interestingly, research does indicate that, when those with severe and multiple learning difficulties are taught from an early age within their zone of proximal development, their learning outcomes improve (Chalaye & Male, 2011). This further supports Morton’s (2004) developmental causal model as it indicates that the environment can play a substantial role in the learning outcomes (such as EF development) of those with DS.
Linguistic weaknesses, typically one of the main areas of difficulty for individuals with DS, especially expressive language (Channell et al., 2015), may also play a role in the results found within different studies. Some tasks that require expressive rather than receptive language skills may present greater difficulty for those with DS, although there is some variability with this (Kumin, 2015). Studies have found that, on the Stanford-Binet subtest, individuals with DS demonstrate greater difficulty in understanding spoken language compared to their non-verbal cognitive skills (Abbeduto et al., 2001). This may be why studies found greater difficulties in verbal WM compared to VSWM for instance (Borella et al., 2013; Fortunato-Tavares et al., 2015). The difference in auditory or visual skill areas is therefore difficult to untangle, given the contradictory evidence provided by different tests. For instance, the test being used to measure the EF of attention (and whether selective or sustained attention is being explored) appears to generate conflicting findings regarding visual and auditory skill level. For instance, Breckenridge et al.’s (2013) findings conflict with that of Trezise et al. (2008), that those with DS had stronger visual sustained attention relative to their auditory sustained attention, as that finding was not replicated using a different study (the ECAB). The role that language may play in the development of different EFs – and different domains within a particular EF (e.g., visual or auditory skill requirements) – would benefit from greater research.

An explanation for some of the findings may be the factor on which the samples are matched on, which could also account for some discrepancies within the findings. This is another reason why, as Thomas et al. (2009) assert, using the developmental trajectory approach can provide a much richer method (compared to the matching approach) for characterizing/describing developmental patterns, including that of EF, in developmental disabilities. However, as can be seen from the results of this systematic review, much of the literature on EF in DS appears to take a cross-sectional, matching approach, which can make drawing firm conclusions about the developmental trajectories
very difficult. Whilst Memisevic and Sinanovic (2014) for example found that WM and shifting were significantly worse in those with DS compared to either those with other ID matched for CA or normative data, children with DS in Daunhauer et al.’s (2014) study were not deemed by teachers or parents to demonstrate particular problems in shifting when compared to TD children of a similar MA. Perhaps this discrepancy is due to the different comparison measures, including whether the participants were matched for MA or CA; studies that only match participants on CA cannot necessarily provide meaningful comparisons to one another, since one would expect difficulties in individuals with DS matched to TD individuals or normative data of the same CA. It may also be due to the comparison groups used, including whether they are comprised of those with other ID, or normative data (instead of TD children), and of course the sample size, as smaller samples lack power and could lead to greater inconsistencies in the findings as a result of individual differences (Fritz & MacKinnon, 2007; Scherbaum & Ferreter, 2009).

Biomedical factors could affect EFs in DS. Bartesaghi et al. (2015) believe that EF limitations are a result of differences in the neurological development in people with DS. In support of this claim, Bartesaghi et al. (2015) demonstrate that the brain in the human foetus with DS shows significant delays at the start of the second trimester, resulting in low intellectual abilities in children born with DS. Pennington et al. (2003) found that adolescents with DS had a smaller hippocampus compared to MA-matched TD controls; in addition, they identified a specific weakness in hippocampal functions in those with DS alongside general cognitive dysfunction in PO, inhibition, VSWM and verbal WM. Lubec and Engidawork (2002) demonstrated that the area of the brain that is most affected in individuals with DS is the cortex, and this is associated with weaknesses in cognitive abilities, particularly memory; it is posited that an atypical neural density within the cortex could be responsible for the cognitive deficits linked with DS (Pennington et al., 2003; Grouios & Ypsilanti, 2011).
Weaknesses in EFs in DS could be also due to the fact that the IQ in people with DS is lower than what might be expected for their CA. According to Grouios and Ypsilanti (2011), the intellectual potential of persons with DS ranges from mild to severe ID. Ersoy, Güler and Çetin (2018) assert that children with DS are likely to demonstrate ID at mild (IQ of between 50 and 70), moderate (IQ between 35 and 50), or severe (IQ between 20 and 35) levels. Thus, one possibility is that the lower performance of children and adolescents on EF tasks may stem from differences in ID (Grouios & Ypsilanti, 2011); it could be that the cognitive demands of the tasks asked of individuals with DS may overwhelm their cognitive abilities. However, if children with DS and TD children have a similar MA, IQ is ruled out as an explanation. Some TD children of a similar MA outperform children with DS on EF tasks, which indicates that EF difficulties found in DS are not likely to be due to broad lower IQ; rather, they are more likely to be due to a specific manner in which DS affects cognition (generating an uneven profile in cognitive abilities).

Moreover, the aspect of MA that children/participants are matched on in studies may affect whether EFs are as weak as studies show. This is supported by the discrepancies when comparing individuals with DS and TD individuals matched for MA. Carney et al. (2013) only found differences between MA-matched DS and TD groups in verbal set shifting and WM, not inhibition and fluency tasks, and Roberts and Richmond (2015) found no difference between preschool age groups in WM, inhibition and set shifting. This compares to findings from Lanfranchi et al. (2010), who showed significant challenges in adolescents with DS compared to MA-matched TD children in WM, PO, conceptual shifting, inhibition, sustained attention and set shifting (with worst performance in the first three EFs). Similarly, Costanzo et al. (2013) found significantly lower abilities in WM, inhibition, attention and shifting in MA-matched DS and TD children and adolescents.
In addition, the demands of some EF tasks may be too great for individuals with DS if they have a visual or hearing difficulties (Weijerman & De Winter, 2010), as vision and hearing challenges and weaknesses are known to be associated with cognitive disabilities (O'Malley, 2013). This could significantly hinder performance on EF tasks. However, some studies only included children with corrected/controlled vision and hearing; some studies have specifically ruled hearing difficulties out of explanations for cognitive difficulties in individuals with DS (e.g., Jarrold & Baddeley, 2001). Yet if there were variability in these physical senses, the inconsistency within the findings from different studies could be more easily explained.

Task difficulty is a particular issue within EF research; some tasks measuring the same (or different) EF may not have the same difficulty level, meaning there may be a difference in EF results across different kinds of tasks. For example, the finding that WM is weaker in those with DS compared to controls than EC ability in those with DS (compared to controls), this might because the task measuring EC may not be as difficult as the task measuring WM. Thus, it is difficult to compare different EFs unless the tasks are of a similar level of difficulty, although the use of control groups as a baseline, using the same tests, can help to illuminate any potential issues with task difficulty. The idea of task impurity creating challenges in drawing conclusions about EF is supported by research from Lanfranchi et al. (2010). In their assessment of various EFs, these researchers found no difference in verbal fluency skills between adolescents with DS and TD children matched for MA. This could suggest that this skill ‗is relatively preserved in individuals with DS‘ (Lanfranchi et al., 2010) (which may be surprising, given that individuals with DS generally demonstrate weak verbal abilities) or that the demands of the task did not exceed the cognitive capabilities of those with DS in this instance. It could also indicate that the verbal fluency tasks were not as difficult as the visuospatial tasks.
The age of participants may also play a role in EF difficulties presented, comparative to normative means or TD individuals. CA may therefore interact with MA, for in Roberts and Richmond’s (2015) study, where no differences were found between MA-matched groups, participants were all preschool age (although these researchers did use receptive language scores as a proxy for MA; using other measures of MA may reap different MA scores, and therefore different findings, as noted earlier). In studies that found significant difficulties in the DS group (Costanzo et al., 2013; Lanfranchi et al., 2010), participants were all above preschool age. Thus, perhaps differences in EFs become more pronounced between TD and DS groups as both CA and MA increases, either as a result of developmental trends, environmental factors, or it may simply be more difficult to measure EF accurately in preschool children. Perhaps this is why studies including adolescents with DS often reveal greater group difficulties in most EFs, compared to studies that only involve young children with DS. However, the small number of studies that involve adolescent individuals with DS limits the confidence with which conclusions regarding EF developments over time can be made, especially given findings from d’Ardhuy et al. (2015), who found that GEC improves over time in individuals with DS. Therefore, further research in this area would be beneficial.

Another factor that may exacerbate EF weaknesses in people with DS could be co-occurring conditions. There is a clear indication that people with both DS and co-occurring conditions present with greater difficulty in all EFs than those without co-occurring conditions. Children and adolescents with DS and other disorders such as DBD and ASD (Pritchard et al., 2015) or obstructive sleep apnoea syndrome, have more weaknesses in EFs when compared to those with DS who do not have additional difficulties. According to Joyce et al. (2020) for example, obstructive sleep apnoea contributes to the advancement of EF deficits as it negatively impacts on cognitive ability. They also note that
obstructive sleep apnoea is associated with poorer EC, shifting and WM. Similarly, ASD makes it difficult for children and adolescents with DS to exercise EFs. Thus, other co-occurring conditions appear to worsen the EF performance of children and adolescents with DS, perhaps due to reduced EF abilities or because they make it more difficult for the children to engage in the assessment of EFs.

Yet despite the evidence indicating that EF abilities may be worsened by various other factors, such as co-occurring conditions, low IQ and hearing/visual difficulties, the different genetic and neurological structure of those with DS indicates that EF weaknesses may be a specific product of DS; therefore, ascertaining just which EF domains are most affected can help provide a better explanation of the neurological and cognitive differences in this population group. EF measurements, tests and tools used commonly by researchers to test different EFs can therefore provide an insight into the specific challenges faced by individuals with DS.

In sum, it may be difficult to separate EF abilities from other cognitive abilities (such as language, problem-solving and avoiding distraction) given the requirements of these abilities to complete EF tasks (Faria, Alves & Charchat-Fichman, 2015). However, given the range of EF tasks and measures (including teacher and parent reports, observation and EF-specific tasks), it is still possible to measure EF performance in different domains to create a generalised picture of specific deficits, although the precise reasons for these weaknesses requires greater research.

3.5.2 Possible Explanations for Working Memory Difficulties

Given the finding that WM is the EF that appears to be the weakest domain in individuals with DS, it is important to explore the trajectory of WM development
in DS, the relative strengths in the visual domain, and potential reasons for why the significant deficits and challenges are experienced by this population.

Some assert that WM weaknesses may occur as a result of generalised cognitive diabilities and difficulties associated with DS (Atkinson & Braddick, 2012; Grouios & Ypsilanti, 2011). For instance, as Colom, Rebollo, Palacios, Juan-Espinosa and Kyllonen (2004) assert, general intelligence (g) or IQ are inextricably related to WM; thus, given that children and adolescents with DS typically have a low IQ, it is potentially to be expected that individuals with DS will exhibit challenges in WM. Moreover, the language difficulties faced by those with DS may also hinder performance on EF tasks, or generate weaknesses in verbal WM (although it could be that weaknesses in verbal WM negatively affect aspects of language acquisition, such as vocabulary development) (Jarrold, Nadel & Vicari, 2007).

Yet individuals with DS appear to exhibit greater challenges and weaknesses in verbal WM than VSWM. Lanfranchi et al. (2004) for example found using a battery of verbal and VSWM tasks in a group of children and adolescents with DS and a control group of children, that individuals with DS demonstrated greater weaknesses in verbal WM relative to their VSWM skills. This difference was more evident when control levels were low; as the requirement for control increased, the DS group showed more difficulty in both domains. Yet Borella et al. (2013) in particular asserts that verbal WM may be poor in those with DS due a link with another EF, inhibition, claiming that they may struggle to suppress irrelevant information, leading to confusion and errors on tasks assessing verbal WM as the load increases.

Various studies indicate relative strengths in VSWM, as well as visuospatial STM and LTM (Visu-Petra et al., 2007). Whilst the latter do not form part of this review, these related functions indicate that visuospatial memory faculties more
generally are less affected than verbal memory faculties in DS. Visu-Petra et al. (2007) concluded from their study that basic VSWM skills are relatively spared in children with DS, yet this strength decays/declines with increased demand (e.g., with greater memory load or when greater executive requirements are experienced by the children). Moreover, Visu-Petra et al. (2007) do note that, whilst their participants were matched for MA, the similarity in basic VSWM skills between DS and TD groups at the mean MA of 5.5 years could ‘conceal a still developing ability in TD children’ (p. 948), meaning that weaknesses may become more pronounced in individuals with DS as their MA increases, compared with MA-matched TD groups.

However, even within an aspect of WM (e.g., VSWM), there may still be a profile of strengths and difficulties in individuals with DS, as shown by Carretti, Lanfranchi and Mammarella (2013). These researchers revealed that children and adolescents with DS presented with greater difficulties in the spatial-simultaneous element of VSWM compared to MA-matched TD children, although their performance did not differ on the pattern configuration of the spatial-sequential VSWM task. This indicates that the presentation format can generate difficulties for those with DS, with spatial-simultaneous WM tasks generating difficulties for those with DS in processing one item at a time, supporting previous research that indicates distinctions in VSWM functioning (Pazzaglia & Cornoldi, 1999). Carretti, Lanfranchi and Mammarella (2013) propose that one explanation for this difference is that those with DS may have visual perception difficulties, including visual crowding, which poses challenges when they must distinguish between different items of information.

One potential additional explanation for why some individuals with DS display significant weaknesses in WM may be due to the presence of the co-occurring condition of attention deficit/hyperactivity disorder (ADHD). Many studies suggest that ADHD is more prevalent in people with DS. For instance, Ersoy,
Güler and Çetin (2018) show that the prevalence of ADHD in the general population is 5% compared to between 14% and 43.9% in people with DS. WM performance has been explored in individuals that have been diagnosed with ADHD, and some of these studies suggest that ADHD negatively affects WM performance in children (Kasper, Alderson & Hudec, 2012; Martinussen, Hayden, Hogg-Johnsen & Tannock, 2005). Some indicate that ADHD and DS are genetic conditions that affect the same brain regions (Gathercole & Alloway, 2006); the idea that genetics may contribute to poorer WM is supported by studies such as Mason et al. (2015), included in this review, which found that people with one 7R are more likely to have ADHD or DS, and that possession of this allele affects WM in white people (although curiously, not in their smaller Hispanic sample). Of course, not all individuals with DS also have ADHD, and yet these individuals also show WM weaknesses. Therefore, ADHD may not be a cause of WM deficit, but perhaps a contributing factor exacerbating this. Either way, it supports Morton’s (2004) developmental causal framework, as genetics clearly play a role in the rate and proficiency of EF development from early infancy onwards (Friedman et al., 2008).

The collated findings from the reviewed studies showed that there was a significant difference between WM performance in adolescents with DS compared to TD children, matched for both CA and MA, whereas there was less evidence that this significant difference was present in children with DS. All studies that used adolescents as participants indicated that adolescents with DS had WM difficulties when compared to their peers matched for MA (Carretti et al., 2013; Chen et al., 2014; Kogan et al., 2009). One study that investigated WM in children found that when children with DS were matched to TD children for either receptive language or non-verbal scores as a proxy for MA, their performance in these tests was similar (Roberts & Richmond, 2015). The consistency in adolescent studies suggests that WM challenges as a result of DS become more evident as age advances (Grouios & Ypsilanti, 2011). This
corresponds with Bopp and Verhaeghen (2007), who, in their meta-analysis of age and WM, found moderate to large negative correlations between WM and age.

A systematic review by Godfrey and Lee (2018) found that adolescents with DS had a demonstrably weaker WM ability compared to TD peers (both MA- and CA-matched) than children with DS compared to similar controls. More particularly, they found that both children and adolescents with DS performed significantly more poorly on both verbal and visual WM tasks, and this performance gap compared to TD peers becomes more significant with age. This falls in line with d’Ardhuy et al. (2015), who found that whilst general EFs tend to improve with CA in people with DS (although at a slower rate than normative data), WM difficulties in children and adolescents with DS appear to remain relatively stable over time. This indicates very little improvement in this area. Godfrey and Lee (2018) speculate that this could be due, in some young adults with DS, to early onset dementia, which is more prevalent amongst DS populations. This decline could be associated with a gradual weakening of multiple brain areas, such as the hippocampus or the cerebellum (Grouios & Ypsilanti, 2011). This does not explain the significant WM performance gap in adolescents however, who do not exhibit early onset dementia.

Thus, although the results of the current systematic review showed that children and adolescents with DS demonstrated substantial difficulties in WM, there is still no clear evidence for causes of WM weaknesses in individuals with DS (Lanfranchi et al., 2004). Similarly, research on how WM skill in DS may change across the lifespan is scarce, so there is a paucity of evidence to indicate why WM abilities appear to progress at a much slower rate than other EFs from childhood into adolescence (Godfrey & Lee, 2018).
One limitation of research specifically into WM in children and adolescents with DS is that, although there is a consensus that these children and adolescents have poor WM as indicated on a range of WM tests, the conclusions lack clarity as these results could represent different performance patterns on very different types of WM tests. It should be noted that WM can be divided into different components, such as visual WM or verbal WM, as indicated by Baddeley’s model of WM discussed in the literature review; these different components can be investigated by different WM tasks. Failures and challenges on tasks by adolescents with DS could stem from some, although not all, of these components. More generally, there is considerable debate about which factors influence WM skills and about how they influence WM (Blasiman & Was, 2018).

3.5.3 Challenges of Executive Function Research in Down Syndrome Populations

It was evident from the systematic review that there are various challenges and limitations of research into EFs in individuals with DS. A majority of the studies focused predominantly on separate EFs, such as WM, inhibition and attention, which left out other EF domains, despite their contributions to the performance of individuals with DS. According to Watson, Gable and Morin (2016), almost all EFs affect the behaviours and activities of participants with DS; however, studies did not seem to give the same level of weight and interest in each EF.

For example, there were no studies that focused specifically on problem-solving, perhaps because problem-solving is not generally identified as an EF; rather, it is present within and supports several EFs. Drigas and Karytotaki (2019) explain the link between problem-solving and EF as ‘bidirectional’, given that both are based on self-control. Both hot and cool EFs contribute towards an individual’s ability to exert cognitive control, whereas problem-solving is interlinked with metacognitive awareness processes. Thus, problem-solving is essential for individuals to apply EF skills in situations requiring goal-setting and decision-
making (Drigas & Karytotaki, 2019). These authors assert that those with cognitive control weaknesses or impairments, either through developmental delay or traumatic brain injury, can ‘counterbalance their cognitive control deficits through enhancing their problem-solving skills’ (Drigas & Karytotaki, 2019, p. 76). The variable of problem-solving could be a useful area to explore in individuals with DS, given its link with EF ability, to assess how far it impacts on EF skill. This is similar to the idea of avoiding distraction, which is a necessary compositive of various EFs such as inhibition and self-regulation (Miller, Giesbrecht, Müller, McInerney & Kerns, 2012) and attraction (Schott & Holfelder, 2015), as discussed in the previous chapter. However, this issue is rarely focused on in its own right, as a distinct contributor to EF capability and development. The lack of focus on associated skills, such as problem-solving and avoiding distraction, may generate challenges for truly understanding EFs in a range of populations, not least the DS population, warranting further investigation.

Although different EF assessment tools and multiple EF tasks can be used in studies to assess children and adolescents with DS (Lanfranchi et al., 2009; 2010), BRIEF (and BRIEF-P), a global measurement tool, is the most widely used (Lee et al., 2015) for assessing EF skills in DS, in addition to other developmental disorders. Various researchers assert that BRIEF is an appropriate tool for categorising EF difficulties specifically in individuals with DS, as they present significant variation in cognitive abilities, making it a useful tool for identifying relative strengths and weaknesses in EF (Lee et al., 2015; Loveall et al., 2017; Pritchard et al., 2015). Yet it does come with various challenges, especially as it was not designed specifically for DS populations; for example, this review revealed that individuals with DS faced significant challenges in PO, similarly to WM, in studies that used either the BRIEF or BRIEF-P. However, this finding may be due to the fact that PO is found in the EMI; this index contains 17 WM items and only 10 PO items. Thus, the challenges seemingly found on PO
may have actually been due to the individual’s observed poor performance on the WM. Thus, the EMI will gain, in general, significantly worse scores compared to controls or normative data, yet this is potentially only due to the WM deficits, not extreme deficits in PO. In other studies that measured PO using different tools (such as Costanzo et al. (2013), which employed the Tower of London task), PO was not found to be an area of severe difficulty in individuals with DS (at least not to the same extent as WM).

Moreover, whilst BRIEF-P reports from teachers and parents showed that children with DS have significant difficulties in WM and EMI (Daunhauer et al., 2014), some studies have suggested that the reports on EF performance of children with DS vary according to who undertook the report (caregivers or teachers). This presents a significant challenge for any researcher attempting to draw conclusions regarding EFs in those with DS. It may be that these differences reflect the different contexts in which the report was conducted, with a greater focus on intellectual considerations in a school environment and emotional considerations in a home environment (e.g., Daunhauer et al., 2014). Yet significant differences between child self-report, parent reports and teacher reports are fairly common, even in TD populations. Cebula et al. (2019) for instance explored the psychosocial adjustment of neurotypical (TD) siblings of children with Williams syndrome. They revealed that their behavioural and emotional adjustment was similar to the norm for the TD population; however, the siblings themselves reported somewhat greater difficulties than caregivers perceived them to have. This illustrates that parent and teacher reports can often be inaccurate regarding their children’s abilities.

Furthermore, parent- or teacher-reported tasks may come with additional considerations, such as bias. Perhaps to overcome this, other studies used different tests to assess the same EF, such as A-not-B task (Roberts & Richmond, 2015), the Day and Night task (Lanfranchi et al., 2010) or the Go/No
Go tasks (Purser et al., 2015) to measure inhibition. Whilst each measure might be valid in and of itself, it is unclear whether the same participant with DS would perform to a similar level on each of the tasks. In addition, as some EFs, such as inhibition, may consist of several functions or skills (as found by Borella et al., 2013), it is unclear which of these specific inhibitory control skills each standalone measure assesses. Given this lack of clarity, it is difficult to draw concrete conclusions. As most studies were cross-sectional, it is possible that their scores may change over time on the same test; as Wishart (1987) found, there was a significant increase in the scores of children with DS on the Piaget Infant Search tasks (measuring inhibition) over the course of the testing sessions. These findings indicate that the cognitive ability of children with DS may be poorly measured by single-session testing and that there may be variability across test sessions, in that skills may not be consolidated quite as quickly in individuals with DS, compared to TD individuals.

Moreover, the studies in this review contained a range of control groups, matched on different features, such as MA or CA. Edgin et al. (2010) asserted that comparisons between participants with IDs and younger TD children ‘are often inaccurate due to differences in the development trajectories of various measures’ (p. 409); this may be especially true for tasks involving memory (Mervis & Robinson, 2005). Even those that matched on MA often used different measures or determinants of MA, such as verbal MA, non-verbal MA and receptive language as a proxy for MA. Again, only tentative conclusions can be drawn, given the disparity of the definitions and measures of MA, even if both the DS and TD groups are measured on the same thing, as the results may not be comparable to other studies using different measures of MA. Furthermore, some did not match participants at all (e.g., Mason et al., 2015, and some only included participants with ‘similar’ (not matched) MA or CA (e.g., Carney et al., 2013). Yet the similarity is not specified and may skew the results or the effect
size if the MA are not matched precisely. These factors all increase the challenge of drawing firm conclusions from study comparisons.

Another limitation is that many of the analysed studies involved a sample of less than 30 participants, although a considerable number (n=40) used a small number of participants (25 studies contained less than 21 participants whilst 15 studies contained fewer than 31 participants). Sample sizes should be relative in size to the population they are attempting to represent – given the percentage of those with DS worldwide, and the number in each country (see chapter two), a sample with only 20 or 30 individuals with DS does not present a sizeable representation of that population (Hackshaw, 2008). The fact that many studies involved a small sample size provides a limitation to the generalisability of their findings (Faber & Fonseca, 2014). A small sample may contribute to the occurrence of type 2 errors (Banerjee et al., 2009), which lowers the power of a study, and may give an incorrect effect size in the findings. This must be taken into account when drawing conclusions regarding the findings and in terms of executive functioning in individuals with DS more generally. However, some studies (e.g., Carretti & Lanfranchi, 2010) do indicate that the effect size values were corrected to avoid small-sample bias. This addresses this limitation in some studies, although this is not noted in all studies.

3.5.4 Limitations
The current review included a wide range of primary studies on EFs in children and adolescents with DS. These were all journal articles. There were no ‘grey’ materials such as PhD theses, working papers or government documents. The year of publication of these studies ranged from the late 1980s to 2020. Use of such a wide scope of sources provided a significant strength of this review, enabling more relevant data to be captured and synthesised. Likewise, the current review reported a substantial number of different tools that assessed a wide range of EFs, giving a fuller image of the investigated EFs.
Despite these overall strengths, there were some limitations that weakened this review. First, there were an extremely large number of papers (29557) which, whilst it was exciting to have so many potentially useful resources and studies, meant that the researcher may have overlooked an important study during the screening process. The search terms used may not have caught all the potentially important studies in the area of EF in DS; thus, which could have affected the results. It is also possible that the cut off date (January 2020) meant that some studies could not be included in the review; whilst this was necessary to prevent an inexhaustible amount of data being included, more recent studies could have shed more light on different EFs investigated in DS.

Secondly, it is noteworthy that many of the studies reviewed (23/55) involved only children as participants, whereas this study was interested in determining the EF functioning in both children and adolescents with DS. Only 8 studies solely involved adolescents as participants, and those studies that included a mixed-age sample did not report the results for adolescents and children separately, rendering it difficult to compare the findings for each age group. Some also included young adults (18+), who may not necessarily be classified as adolescents, especially if they are no longer in school/education. This limitation affected the understanding of the effects of DS on EFs such as WM as individuals advanced in age, and also means that conclusions regarding WM performance differences between children and adolescents can only cautiously be drawn from these findings. The studies with mixed-age populations involved the largest sample sizes. This may be expected however, as this gives the researcher(s) a greater sample from which to draw conclusions. Moreover, it may assist with overcoming any obstacles encountered with recruiting participants with DS from a narrower age range. However, studies also used different age ranges and classifications for children, adolescents and adults, which meant some studies may have compared individuals with different ages
against one another under the same category (e.g., children could be 5 years old or 12 years old).

A further limitation is the fact that no meta-analysis of the studies was carried out. Although such an analysis had been planned in the early stages of this review, it was found that the studies were too diverse in controls, tools and assessed functions for a meta-analysis to be possible. This issue was compounded by the fact that some studies (e.g., Wishart, 1987) did not make it clear which EF they were measuring, which could limit the validity of the study as pertaining to particular EFs. The wide range and variation of SR studies rendered synthesising the studies very difficult, as this review included studies that investigated different populations using a variety of methodologies and assessments. Thus, in some instances it was challenging to compare studies, meaning that any conclusions drawn from the literature regarding EF in DS had to be done so cautiously.

Finally, the critical appraisal tool (CASP) to evaluate the quality, reliability and validity of the included studies, may have been too simplistic; arguably, this tool allowed for too high a number of studies to be rated as high quality. However, the simplicity of the quality control assessment tool was arguably necessary to cater for the wide range of studies exploring EF and DS that were included in the final review.

### 3.6 Conclusions, Recommendations and Next Steps

The systematic review showed clearly that children and adolescents with DS have significant deficits in EFs. People with DS demonstrate considerable disparity in performance in different tasks, including those involving working memory, inhibition, attention, cognitive flexibility and planning. Similarly, findings from the synthesis also show that children and adolescents with DS demonstrate significantly poorer results in all functions than TD groups, matched
for either CA or MA. However, it was established from the studies that specific EF domains are highly affected. Studies that focused on global EF skills, often using BRIEF and BRIEF-P, presented WM and PO as the most affected. These findings were also supported by the studies that focused exclusively on WM (although to a lesser extent the planning/organising EF). The comparison of EF abilities in participants of different ages demonstrated that deficits in these abilities in individuals with DS become more prominent (comparative to TD individuals) as this population advanced in age, indicating that the development trajectory of EFs in those with DS falls behind that of their CA-matched TD counterparts.

Although a considerable amount of information was retrieved from the reviewed studies, it is imperative to note that there are few studies on adolescents with DS as participants, as a majority of the reviewed studies focused on children or mixed-age group as participants. Based on this, there is a need for more research to be conducted using adolescents with DS as the participant base. Likewise, it can be established from the reviewed studies that there are not enough studies using participants of mixed ages. For EF in children and adolescents to be compared effectively, future research should be conducted that includes both children and adolescents as participants and very clearly distinguishes between these age groups in the results.

It is evident that DS affects all EFs as well as associated skills, such as problem-solving and avoiding distraction. Based on this, it is imperative for future research to assess the implications surrounding a diagnosis of DS on all EFs, as well as associated skills, as they affect the performance of children and adolescents with DS. A focus on the problem-solving abilities of those with DS, for instance, can better help understand (and perhaps even improve) the EF abilities of those with DS. The idea that learning rooted in problem-solving, and problem-solving interventions more broadly, can help to improve overall EF
ability and academic achievement has been documented in the literature (Monari Martinez & Pelligrini, 2010; Zelazo et al., 2018). Learning within problem-centred, collaborative environments could enable students (both TD and those with DS) gain higher-order executive skills, including EFs (Zelazo et al., 2018), although there is conflicting research surrounding this (e.g., Beck et al., 2016). Therefore, it would be beneficial to focus research attention on the role that associated skills such as problem-solving may play in the development of EFs.

Moreover, the current study focused predominantly on children and adolescents with DS despite the fact that the EF deficits demonstrated by those with DS become more apparent, relative to their CA, as individuals age. Thus, it is necessary for future research to assess EFs in children, adolescents and adults to understand how they are affected by advancement in age and whether the developmental trajectory differs to that of the TD population in different EFs. It would also be useful, to avoid the challenges inherent within the BRIEF and BRIEF-P, to adopt the principle of evaluating EFs in one measure, yet giving each EF the same weighting in that measure, to avoid the risk that some EFs appear to be significantly challenged because they are reported as part of a collapsed/merged set of data.

Overall, many studies have concluded that EC was the least challenged EF in those with DS. It would be useful to investigate which factors may contribute to this relative strength, to ascertain if these factors may be employed to help strengthen other EFs in those with DS. Furthermore, this review concluded that WM is the most challenged EF in people with DS. For this reason, it is arguably necessary to conduct interventions with the aim of improving WM in children and adolescents with DS. Therefore, future studies should focus on WM and the efficacy of specific interventions designed to improve this specific EF in children with DS.
This being the case, the next stage of this thesis focuses on WM in children with DS. It aims to design and test the efficacy of an intervention specifically aimed to improve WM functioning in children, before the observed gap in WM performance between individuals with DS and TD individuals becomes significantly large, as it is in adolescence. The next chapter therefore details the methodology that was used in the creation of such an intervention.
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amado et al., 2016</td>
<td>Children DS sample size: 30 F=18; M=12; mean CA=8.54(\text{SD}=2.36); mean MA=4.49 (SD=1.69)</td>
<td>Spain School, home or clinic</td>
<td>Cross-sectional Tests WM Inhibition Cognitive flexibility</td>
<td>Visual-spatial WM: Frog task; Inhibition: Day-night task; Cognitive flexibility (CF): Wisconsin card sorting task</td>
<td>• DS group scored significantly worse than both control groups on all tasks of EF, denoting significant WM, inhibition and CF difficulty.</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group TD x 2: CA-matched (n=30) LD(^3)-matched (n=30)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ashworth et al., 2015</td>
<td>Children DS sample size: 22 F=11; M=11; mean CA=9.42 (SD=1.98; range=6.09-12.23); Mean MA&lt;5 (range=&lt;5-8; RCPM=12.6 (SD=3.53)</td>
<td>UK Home and school</td>
<td>Cross-sectional Tests Attention</td>
<td>Visual Continuous Performance Task (CPT)</td>
<td>• Children with DS showed significantly poorer attention than TD children and children with WS (fewer 'correct hits' on the CPT than both groups and more errors than the TD group). • Reaction time of children with DS was significantly slower when compared to WS and TD children.</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group x 2 = CA-matched: Control 1: WS group (n=22); CA mean=9.24 (SD=2.13; range=6.08-12.58); mean MA=6</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

\(^2\) Ages given in years

\(^3\) Language development
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bieberich &amp; Morgan, 2004</td>
<td>Children DS sample size: 15 (F=3; M=12); mean CA=8.3 at start, 10.3 at follow-up</td>
<td>USA University research room</td>
<td>Longitudinal Tests</td>
<td>Self-regulation</td>
<td>The Minnesota Preschool Affect Rating Scale (MN-PARS) (Shapiro et al., 1994)</td>
<td>Children with ASD received significantly lower ratings than children with DS on 3 of the 4 self-regulation factors (Attention, Object Orientation; Persistence) across time, with the fourth scale approaching significance (Adaptability); Over time, DS group showed significantly greater stability in self-regulation compared to ASD group</td>
<td>12</td>
</tr>
<tr>
<td>Brown et al., 2003</td>
<td>Children DS sample size: 19 Mean CA=2.4 (range=2.3-1). Control group x 3:</td>
<td>UK</td>
<td>Cross-sectional Tests</td>
<td>Attention</td>
<td>2 x sets of experimental trials: 1) double-step saccade task 2) sustained attention task</td>
<td>WS group poorer performance on saccade tasks compared to DS and TD control groups; DS group demonstrated shorter and fewer periods of sustained attention compared to WS and TD control groups.</td>
<td>13</td>
</tr>
</tbody>
</table>

4 Verbal mental ability
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>mean CA=2.5 (range=1.9-3.1) Control 3: MA-matched TD control group (n=15), mean CA=1.25 (range=1.1-1.75).</td>
<td>Italy</td>
<td>Cross-sectional Tests</td>
<td>WM</td>
<td>Spatial-simultaneous WM task</td>
<td>● DS group performance was significantly worse in all WM tasks (random and structured condition); ● Individuals with DS are able to take advantage of structured material for raising their performance, but to a less extent TD children; ● Index of Benefits results suggested that DS group benefitted from the presence of pattern less than the TD group; ● Individuals with DS had a significantly greater decline in performance as a result of WM load compared to TD children, whose performance remained substantially stable across four levels of memory load.</td>
<td>13</td>
</tr>
<tr>
<td>Carretti &amp; Lanfranchi, 2010</td>
<td>Children DS sample size: 20 (F=14; M=6); mean CA=7.45 (SD=1.1); mean MA=4.8 (SD=1.1) Control group: TD control group matched for VMA (n=20); mean CA=5.4 (SD=0.3); mean MA=5.4 (SD=0.7).</td>
<td>Italy Location not reported</td>
<td>Cross-sectional</td>
<td>WM</td>
<td>Spatial-simultaneous WM task</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Daunhauer et al., 2014</td>
<td>Children DS sample size: 25</td>
<td>USA</td>
<td>Cross-sectional</td>
<td>Global EF BRIEF-P⁵</td>
<td></td>
<td></td>
<td>13</td>
</tr>
</tbody>
</table>

⁵ Behavioural Rating Inventory of Executive Function-Preschool Version
⁶ Inhibitory Self-Control Index
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Daunhauer et al., 2017</td>
<td>Children. DS sample size: 42 (M=24; F=18). Mean CA=7.6 (SD=1.4; range=5.1-11.1); Mean MA=4.2 (SD=0.78); USA; laboratory</td>
<td>Cross-sectional Test</td>
<td>WM Inhibition</td>
<td>WM/inhibition task: Pony and gator Inhibitory</td>
<td>EMI) submitted by both teachers and parents showed significant difference in EF between DS and TD children. • WM = most significant area of weakness in children with DS compared to TD group. • No significant interaction effect for group by the type of reporter. • Teacher-reported scores showed DS students had significantly greater deficits in GEC and EMI • No significant between-group differences in the flexibility/shifting and EC scales; • Significant difference in ISCI between TD and children with DS in parent group only (not teachers • Both teachers and parent agreed that children with DS had difficulties in global executive function, WM and P/O</td>
<td>13</td>
<td></td>
</tr>
</tbody>
</table>

7 Flexibility Index
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>de Santana et al., 2014</td>
<td>Children DS sample size: 30 (F= 13; M=17); mean CA=8.83 (SD=1.84); MA not provided. Control group: TD Group (n=30) matched for CA (F=16; M=14); mean CA=8.17 (SD=1.72)</td>
<td>Brazil Participant's house, school, and a private office.</td>
<td>Cross-sectional Tests</td>
<td>WM</td>
<td>Wechsler Intelligence Scale for Children, 3rd Edition (WISC-III) – including Working Memory Index (WMI)</td>
<td>DS performing significantly more poorly. No significant difference between inhibition and shifting (when looking at individual variables) although DS group performed worse than TD group in shifting but better in inhibition (non-significant).</td>
<td>11</td>
</tr>
<tr>
<td>Estigarribia et al., 2012</td>
<td>Children (Boys, all matched for MA) DS sample size: 31 Mean CA=10.19(SD=2.8) Control group 1: FXS group (N=33); Mean CA=10.56(SD=2.41)</td>
<td>USA Child’s home, school or the Frank Porter Graham Child Development</td>
<td>Cross-sectional Tests</td>
<td>Phonological WM</td>
<td>Nonword Repetition Task of the Comprehensive Test of Phonological Processing (CTOPP; Wagner, Rorgesen &amp;</td>
<td>Participants with DS performed poorer on WMI of WISC-III than TD group (unclear if significant).</td>
<td>13</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>-------------------------------------------------------------------------------------</td>
<td>---------------------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>-----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Control group 2: FXS + ASD group (N=28); Mean CA=9.98(SD=3.04)</td>
<td>ent Institute at the University of North Carolina.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Both TD children and children with DS showed a similar performance in self-regulation tasks matched for MA;</td>
<td></td>
</tr>
<tr>
<td>TD group (n=46); Mean CA=5.03(SD=1.18)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• DS group better at competence with the lockbox task;</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• DS group showed more ‘leaving’ – task-avoidant behaviour.</td>
<td>12</td>
</tr>
<tr>
<td>Gilmore et al., 2003</td>
<td>Children DS sample size: 25 (F=15; M=10); mean CA=5.31 (SD=0.73); mean MA=2.5 (SD=0.44)</td>
<td>Australia research room</td>
<td>Cross-sectional Observation</td>
<td>Self-regulation</td>
<td>The Goodman Lock Box task (Goodman, 1981).</td>
<td>• Both TD children and children with DS showed a similar performance in self-regulation tasks matched for MA;</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group: TD group (n=43) matched for MA. (F=20; M=23); mean CA=2.57 (SD=0.31); mean MA=2.49 (SD=0.34).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• DS group better at competence with the lockbox task;</td>
<td></td>
</tr>
<tr>
<td>Joyce et al., 2019</td>
<td>Children. DS sample size: 17 (M=8; F=9). Mean CA=10.11 (SD=1.68; range=7.19-12.23). Mean MA=under 5.</td>
<td>UK. Child's school or home</td>
<td>Cross-sectional Test Probl. solving</td>
<td>Tower of Hanoi</td>
<td></td>
<td>• DS and WS groups made significantly more rule violations on TOH task than TD group.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 1: WS. N=21 (M=10; F=11). Mean CA=9.39 (SD=2.05; range=6.16-12.58). Mean MA=6.25.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• The TD Wake-sleep condition showed no significant change in number of rule violations after either sleep or wake.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 2: TD.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Children with DS showed no significant change in scores after wake or sleep.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Children with WS in both circadian conditions had significantly reduced number</td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/ setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>---------------------</td>
<td>------------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>------------------------------------------</td>
<td>---------------</td>
</tr>
</tbody>
</table>
| Joyce et al., 2020| Children. DS sample size: 80 (M=50; F=30). Mean CA=4.7 (SD=0.9; range=3-5.9). Reliable data obtained for 75 children. Compared to normative data. | UK Children's centre | Cross-sectional Survey | Global EF | BRIEF-P | DS sample had significantly poorer EFs on every measure relative to the mean of 50 in TD reference group.  
  - DS group performed most poorly on WM, then plan/organise.  
  - Emotional control was an area of relative strength compared to inhibit, WM and Plan/Organize.  
  - EMI was significantly poorer than both Inhibitory Self-Control Index and Flexibility Index. | 11 |
| Kirk et al., 2017 | Children. DS sample size: 22 (M=12; F=8). Mean CA=8.92 (SD=1.51; range=5-11).  
  Control group 1= ASD group (n=23) (M=19; F=3). Mean CA=7.2 (SD=1.74; range=4-10).  
  Control group 2=NSID group (n=32) (M=18; F=14). | Australia Laboratory/school | Cross-sectional Test | Sustained attention | Wilding Attention battery (Wilding, Cornish & Munir, 2002) |  
  - Children with DS had significantly poorer vigilance and sustained abilities than children with ASD and NSID.  
  - No significant group differences were present on parent-rated inattention. | 11 |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
</table>
| Klotzbier et al., 2020 | Children. DS sample size: 12 (M=6; F=6). Mean CA=10.5 (SD=1.08). Control group 1: CA-matched TD (n=12) (M=6; F=6). Mean CA=10.5 (SD=1.07). Control group 2: MA-matched TD (n=12) (M=6; F=6). Mean CA=5.98 (SD=1.21). | Germany Laboratory | Cross-sectional Test   | Attention Cognitive flexibility | Trail-Walking-Test | - Children with DS and TD-MA exhibited overall greater performance deficits in tasks with increased cognitive load compared to tasks with low cognitive load.  
- Differences between DS and TD-MA in the execution times of the motor ST and in the motor DTC are strong arguments for reduced attention resources and cognitive flexibility skills in children with DS. | 13            |
| Lee et al., 2011    | Children DS sample size: 26 F=11 M=15; mean CA=6.3 (range=4-10; SD=1.9); mean MA=3 (range=2-4.75; SD=8.98). No control group; results compared to normative means for TD children with similar MA. | USA University lab | Cross-sectional Caregiver survey and clinical measurement | Global EF | BRIEF-P         | - Those with DS indicate a significant and specific pattern of EF (global EF, WM, planning/organisation) weaknesses compared to the MA-matched TD normative data.  
- In particular, significant relative weaknesses found on GEC$^8$ and EMI$^9$ indexes of BRIEF-P.  
- Scores on WM scale were significantly higher on WM scale compares to other scales (indicating greater | 13            |

---

$^8$ Global Executive Composite  
$^9$ Emergent Metacognition Index
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lewis &amp; Brooks-Gunn, 1984</td>
<td>Children DS group (n=50) Divided into four groups: [3-7 months, n=15 mean, CA=5.5 ]; [8-16 m, n=9, mean CA=12.1]; [17-27 m, n=14, mean CA=22.2]; [28-36 m, n=12, mean CA=32.5]. MA RANGE (3-27 months) Control group 1: cerebral palsy group (n=17) CA=range 0-4 months, MA range=3-4 months. Control group 2: Developmentally delayed group (n=14) CA= range 0-5 months, MA= range 4-36 months. Control group 3: Multiple disabilities group (n=21) CA range=2-10, MA</td>
<td>USA clinic Cross-sectional Test</td>
<td>Visual attention</td>
<td>Visual attention task.</td>
<td>• No group differences; • Children with DS looked for significantly longer during trials as their CA increased and demonstrated a significant decrement in attention as the trials went on</td>
<td>12</td>
</tr>
</tbody>
</table>

10 Emotional Control
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Loveall et al., 2017</td>
<td>Children. DS sample size: 112 (M=62; F=50). Overall mean CA=12.8 Ages 2-5, n=22 (M=9; F=13). Mean CA=3.6 (SD=1.2; range 2-5). Ages 6-18, n=64 (M=28; F=36). Mean CA=11.5 (SD=3.3; range=6-18). Ages 19-35, n=26 (M=13; F=13). Mean CA=24 (SD=5; range=19-35).</td>
<td>USA. Child's home</td>
<td>Cross-sectional Survey</td>
<td>Global EF</td>
<td>BRIEF, BRIEF-P</td>
<td>• 2-5 years: GEC sig. worse than normative data. Only EMI significantly above TD norm. WM=significantly different. P/O and Inhibition=marginal significant; shift and EC=non-significant. • 6-18: GEC sig. worse than TD norm. BRI, EMI also sig. worse than TD group. Areas of sig. weakness=WM, monitor. shift, P/O. Areas of moderate weakness=inhibit and initiate. Area of relative strength=organisation of materials &amp; EC. • From 2-18 years, sig. decline in shift abilities. All others=consistent.</td>
<td>13</td>
</tr>
<tr>
<td>Molina &amp; Perez, 1993</td>
<td>Children DS sample size: 30 2 DS groups (Experimental and Control groups, randomly matched for CA and MA). Experimental group (n=15); CA range=9-12; mean MA=6.57 (SD=1.46). DS control group (n=15);</td>
<td>Spain Research lab</td>
<td>Cross-sectional Test and Re-test</td>
<td>Planning</td>
<td>Dynamic Assessment of Learning Potential (3 tests) Spatial Structuring Designs (SSD); Mazes (M); Puzzles (P).</td>
<td>Children with DS had poorer planning processes (although whether the difference was significant was not noted) than TD group of similar MA; • Some children with DS performed significantly poorer in all tasks compared to some children (similar MA and CA) with ID (in control group only – the experimental groups did not differ); • Indicates that mediation (prompts and cues for solving</td>
<td>11</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>--------------------------------------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>-----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td><strong>Pritchard et al., 2015</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Results from an exploratory factor analysis of item-level BRIEF-P data supported the theoretically derived three-factor structure originally proposed for the BRIEF-P (Emergent Metacognition (EMI), Flexibility (FI) and Inhibitory Self-Control (ISCI)); • The result shows the item composition of each factor varied somewhat in comparison to the original structure of the measure. • All DS groups showed greatest deficit in EMI, then ISCI, then FI. • Youth with typical DS evidence fewer executive function difficulties across all domains.</td>
<td>13</td>
</tr>
<tr>
<td>Children</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DS sample size</td>
<td>183</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>mean CA</td>
<td>6.99</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(SD=2.47)</td>
<td>range=3-13</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>MA not provided.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DS co-occurring</td>
<td>with ASD (n=61) and DS co-occurring with DBD (n=98). 86% had documented trisomy 21, while 3% had complete Robertsonian translocation, and 2% had mosaicism.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control group:</td>
<td>children with Typical DS (n=24) controlled for CA and MA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>USA</td>
<td></td>
<td>Cross-sectional Caregiver reports</td>
<td>Global EF</td>
<td>BRIEF-P</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Clinic</td>
<td></td>
<td>Records</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Cross- sectional Caregiver reports Records</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>----------</td>
<td>---------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>---------------</td>
<td>-------------------------------------------</td>
<td>---------------</td>
</tr>
</tbody>
</table>
| **Roberts & Richmond, 2015** | Preschool children DS sample size: 17 (F=8; M=9); mean CA=4.3 (SD=0.9); mean MA (Receptive Language Equivalent)=1.9 (SD=0.7) years. Control group: TD children (n=17), matched on MA (Receptive Language Equivalent). Mean CA=1.9 (SD=0.5); mean MA=1.9 (SD=0.7). | Australia Early learning lab, University of New South Wales. | Cross-sectional Test | WM Inhibition Set shifting (SS) | A-not-B task | ● Youth with DS+ASD show the greatest weaknesses in EMI.  
● Youth with DS+DBD show significant difficulties in both EMI and ISCI. | 13 |
| **Schott & Holfelder, 2015** | Children DS sample size: 18 (F=7; M=11); mean CA=9.06 (SD=0.96; range=7-11); MA not provided. Control group: TD group matched for CA and gender; mean CA=8.99 (SD=0.93); MA not provided; mean BMI=18.2 (SD=3.55). | Greece School | Cross-sectional Test | Inhibitory control Set switching (attention and distraction) | Trails-Preschool test – Revised (Trails-P) (Espy & Cwik, 2004). | ● DS group demonstrated significantly poorer performance in all functions involved in inhibitory control and set switching.  
● EF correlated positively with motor skills for the group with DS.  
● Children with DS had worse scores in tasks assessing distraction and inhibition, compared to TD children. | 11 |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shalev et al., 2019</td>
<td>Children. DS sample size: 18. Mean CA=7.2 (SD=1.1). Mean VMA=3.5 (SD=0.8). Mean NVMA=3.3 (SD=0.8). Control group 1=WS group (n=25). Mean CA=6.62 (SD=0.92). Mean VMA=5.3 (SD=1.6). Mean NVMA=3.21 (SD=1.6). Control group 2: TD younger (n=31). Mean CA=3.8(SD=0.5). Mean VMA=4.3(SD=1). Mean NVMA=4.1(SD=0.8). Control group 3=TD-middle (n=34). Mean CA=5.5(SD=0.5). Mean VMA=5.9(SD=1.2). Mean NVMA=5.2 (SD=0.9). Control group 4=TD-older (n=34). Mean CA=7.1(SD=0.4). Mean VMA=4.9(SD=1.4). Mean NVMA=7.3 (SD=1.6).</td>
<td>UK Laboratory</td>
<td>Cross-sectional Test</td>
<td>Attention (specifically sustained attention)</td>
<td>Continuous Performance Task</td>
<td>- Children with DS and WS performed significantly poorly compared to all TD groups. - DS and all TD groups performed better in the second half of the task whereas WS group performed worse in the second half. - Researchers concluded that children with WS suffer from a selective deficit in the ability to sustained attention over time, whereas children with DS have a more general performance difficulty. - More children in the DS group did not complete the task (lack of engagement).</td>
<td>12</td>
</tr>
<tr>
<td>Wishart, 1987</td>
<td>Children DS sample size: 12 (F=6, M=6); mean CA=</td>
<td>Scotland Infant</td>
<td>Longitudinal study</td>
<td>Inhibition</td>
<td>Piagetian Infant Search Tasks</td>
<td>- DS group performed significantly more poorly than TD group, denoting worse</td>
<td>11</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
<td></td>
</tr>
<tr>
<td>----------</td>
<td>---------------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>---------------</td>
<td>-------------------------------------------</td>
<td>--------------</td>
<td></td>
</tr>
<tr>
<td><strong>3.73 (SD=0.81): 10 participants with Trisomy 21, 1 Mosaic and 1 due to translocation.</strong>&lt;br&gt;Control group: TD group (n=12; (F=6; M=6); mean CA=3.75 (SD=0.56). matched on Sex, Number of Siblings and Birth order</td>
<td>Laborator y Test</td>
<td>Inhibition;&lt;br&gt;Over the 6 sessions, the performance of children in the DS group significantly improved, whereas the performance of those in the TD group did not.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Adolescent sample studies</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Camp et al., 2016</strong>&lt;br&gt;Adolescents. DS sample size: 31 (M=14; F=17). Mean CA=18 (range=10.4-23.9).&lt;br&gt;Control group 1: WS group (n=47) (M=24; F=23). Mean CA=18 (range=10.7-26.6).&lt;br&gt;Control group 2: TD group (n=34). (M=18; F=16). Mean CA=8.3 (range=4.10-11.5).</td>
<td>UK Child’s school Cross-sectional Survey Problem-solving BRIEF Problem-solving questionnaire (PSQ)</td>
<td>BRIEF: performance for WS group was significantly poorer than DS and TD, whilst performance of DS group was significantly poorer than TD only in Shift, Working Memory, Plan/Organize, Monitor and Initiate (at p&lt;0.5). The DS group scored highest on Inhibit then Organisation of Materials.&lt;br&gt;PSQ: TD scored significantly higher than DS and WS, but DS scored significantly higher than WS.&lt;br&gt;Significant relationships between parent-rated EFs and reaching the solution to a problem-solving task are present more often for</td>
<td></td>
<td></td>
<td>13</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>--------------------------------------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>---------------</td>
<td>-------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Chen, et al., 2014</td>
<td>Adolescents and adults DS sample size: 90 (49 adolescents and 41 adults) Adolescents: F=20; M=29; mean CA=14.5(SD=1.6; range=12-17); Mean IQ=41.8(SD=7.1). Adults: F=21; M=20; mean CA=22.7(SD=3.4; range=18-30); mean IQ=39(SD=6). Control group: group comparison matched for CA and MA</td>
<td>USA, UK, Spain, France, Italy, Canada Argentina Laboratory</td>
<td>Cross-sectional Tests</td>
<td>WM Planning</td>
<td>WM: Corsi-Block taping for Visual WM; Auditory Memory span test for verbal WM Planning: Tower of London (TOL) test</td>
<td>Compared to normative data, the performance of people with DS was poor on the TOL score (the performance was equivalent to TD children with a CA of 8 years) measuring planning.</td>
<td>12</td>
</tr>
<tr>
<td>d’Ardhuy et al., 2015</td>
<td>Adolescents and adults DS sample size: 12 mean CA=18.58 (SD=3.79); mean MA=5.95 (SD=2.32).</td>
<td>USA Laboratory</td>
<td>Cross-sectional Tests</td>
<td>Global EF WM</td>
<td>Global EF: BRIEF-P WM: CANTAB (Spatial Span [SSP])</td>
<td>BRIEF-P: Adolescents with DS demonstrated statistically greater difficulties than adults with DS in GEC. WM, PO and I subscales did not correlate with IQ. WM domain of BRIEF-P correlated with reverse SSP results. CANTAB SSP: Low scores for both groups in all tasks (showing significant WM deficits).</td>
<td>13</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>---------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>-----------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Edgin et al., 2010</td>
<td>Adolescents and adults DS sample size: 18 (M=7; F=11); mean CA=17.81 (SD=2.90; range=13-23)</td>
<td>USA University lab</td>
<td>Cross-sectional Tests</td>
<td>WM</td>
<td>Backward Corsi span task; TAPS-R backward auditory number memory test.</td>
<td>• No significant differences between adolescent with DS and WS in WM.</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>Control group: Williams Syndrome (WS) group (n=18). CA- and IQ-matched. (M=13; F=5); mean CA = 18.63 (SD=3.98; range=12-26)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Faught et al., 2016</td>
<td>Adolescents and adults DS sample size: 20 (F=10; M=10); mean CA=15.98 (SD=3.53; range=10-21)</td>
<td>USA Home, school or University lab</td>
<td>Cross-sectional Tests</td>
<td>Attention (Auditory and visual sustained attention)</td>
<td>Auditory SART; Visual SART using two tasks Omission errors and Commission errors.</td>
<td>• DS and TD groups performed similarly on both auditory and visual sustained attention. • Auditory and visual SA aligns with developmental level.</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>Control group: TD group matched for non-verbal MA (n=20); mean CA=4.84 (SD=1; range=3-7).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kogan et al., 2009</td>
<td>Adolescent and adults DS sample size: 15 Mean CA=17.16 (SD=5.80; range 11.09-36.01); Mean MA=6.35 (SD=1.50; range 2.58-8.75)</td>
<td>Canada Research lab</td>
<td>Cross-sectional Tests</td>
<td>WM</td>
<td>The adapted Wisconsin General Test Apparatus (WGTA)</td>
<td>• Individuals with DS had significantly poorer performance on visual-perceptual and visual-spatial reversal learning tasks compared to egocentric spatial learning and object discrimination tasks. • Both ID groups performed better on a visual-perceptual</td>
<td>13</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------</td>
<td>---------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>--------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td>Lanfranchi et al., 2010</td>
<td>Adolescents with DS sample size: 15 (F=7, M=8); mean CA=15.2 (SD=2.2; range=11-18.42); mean MA=5.75 (SD=0.7; range=4.5-6.83).</td>
<td>USA School room</td>
<td>Cross-sectional Tests</td>
<td>WM Sustained attention</td>
<td>WM: Verbal and visuo-spatial dual tasks (Lanfranchi et al., 2004). Inhibition: Stroop Type Task – Day/ Night Version (Gerstadt et al. 1994).</td>
<td>Results showed that adolescents with DS performed significantly worse than TD children for all EF tasks, including sustained attention, verbal and visuospatial dual tasks, inhibition, set and conceptual shifting tasks and planning/problem-solving.</td>
<td>13</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>----------------</td>
<td>-----------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>--------------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Trezise et al., 2014</td>
<td>Adolescent (males only)</td>
<td>Australia</td>
<td>Cross-sectional Tests</td>
<td>WM</td>
<td>The verbal and visuospatial WM task.</td>
<td>• Adolescents with DS demonstrated significantly poorer WM than the NSID group;</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>DS sample size: 15</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Adolescents with DS performed less well than autistic adolescents, although</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mean CA=14.84 (range=10.83-18.82).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 1: non-specific intellectual</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-----------------------</td>
<td>--------------------------------------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td></td>
<td>disability (NSID) (n=12); mean CA=13.72 (range=11.7-18.42) matched for MA and CA.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>the difference was not significant; • DS group showed a non-significant trend for stronger performance in visual versus auditory task.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 2: Autism and ID group (n=11); mean CA=14.48 (range=10.83-18.42)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• DS group showed a non-significant trend for stronger performance in visual versus auditory task.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>matched for MA and CA.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mixed sample studies</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bhattacharya et al.,</td>
<td>Children, adolescents and adults</td>
<td>India</td>
<td>Cross-sectional survey</td>
<td>Impulsivity</td>
<td>DASH scale</td>
<td>• The DS group showed significantly higher scores in impulsivity compared to TD groups (although not as high as non-DS group with other ID matched on CA).</td>
<td>13</td>
</tr>
<tr>
<td>2009</td>
<td>DS sample size: 70 mean CA=16.30 (SD=10.288; range=3-37); mean IQ =45 (range=25-70); MA not provided.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 1: TD group (n=70) with similar CA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Control group 2: non-DS group with other ID (n=70) with similar CA</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Borella et al., 2013</td>
<td>Children and adolescents.</td>
<td>Italy School</td>
<td>Cross-sectional Tests</td>
<td>WM Inhibition</td>
<td>WM: Verbal Dual Task (Lanfranchi et al., 2004)</td>
<td>• Verbal Dual Task: DS children performed significantly less well than TD children; • Animal Stroop Task: DS performed significantly less</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>DS sample size: 19 (F=12; M=7); mean CA=14.5 (SD=2.5);</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measures</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>-------------------------</td>
<td>---------------------------------------------------------</td>
<td>-----------------</td>
<td>---------------------------</td>
<td>-------------</td>
<td>--------------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td></td>
<td>Controls group: TD group (n=19) matched for MA. (F=11; M=8; Mean CA=5.22 (SD=0.8; range=3-6).</td>
<td></td>
<td></td>
<td></td>
<td>Inhibition: Animal Stroop test (Wright et al, 2003); Proactive Interference (PI) task (Borella et al., 2010); directed forgetting-blocked method (Harnishfeger &amp; Pope, 1996).</td>
<td>well than TD children, making more mistakes, indicating general inhibitory deficit; <strong>PI Task</strong>: Main effect of group (DS group more susceptible to interference); <strong>Directed forgetting-blocked method</strong>: main effect of Group = marginally significant for intrusion errors, with the DS group recalling more words that they should have forgotten than the TD group; TD group performed significantly better than DS group in the second half of the word list.</td>
<td>11</td>
</tr>
<tr>
<td>Breckenridge et al., 2013</td>
<td>Children and adolescents. DS sample size: 32. Mean CA=9.76 (range=5.01–14.07). Mean MA=4.51 (range=3.01–5.11). Control group=WS group (n=32). Mean CA=8.45 (range=5.0–15.11). Mean MA=4.89 (range=3.10–5.11).</td>
<td>UK Laboratory</td>
<td>Cross-sectional Test</td>
<td>Attention</td>
<td>Early Childhood Attention Battery (ECAB)</td>
<td>• For DS: 3 of the 8 subtests on ECAB gained significantly lower scores than the expected norm value of 10 for MA (for WS it was 4 of the 8 subtests); • On two tests, the DS group performed significantly better than the norm for MA (auditory sustained and dual task); • On two tasks the DS group performed significantly better than WS (auditory sustained and counterpointing);</td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td></td>
</tr>
<tr>
<td>------------------</td>
<td>--------------------------------------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>---------------</td>
<td>----------------------------------------------------------------------------------------------------------</td>
<td></td>
</tr>
</tbody>
</table>
| Carney et al., 2013 | Children and adolescents DS sample size: 25(M=10; F=15); mean CA=13.64 (SD=2.64; range=10.33–18.92); mean MA=6.03 (SD=0.93; range=3.92–7.67). Control group 1: WS group (n=24); (M=11; F=13); mean CA=13.59 (SD=3.03; range=8.08–18.92); mean MA=6.74 (SD=1.1; range=5.5–10). Control group 2: TD group (n=26); (M=16; F=10); mean CA=6.14 (SD=0.97; range=5–8); mean MA=6.53 (SD=1.18; range=4.92–8.92) | UK Participant home/school | Cross-sectional Tests | WM Inhibition Shifting | Executive-loaded working memory ELWM. Verbal ELWM using adapted version of the Listening Span task (Leather & Henry, 1994). Visuospatial ELWM test was a modified version of the Odd-One-Out test (Henry, 2001). Verbal Inhibition, Motor Inhibition (VIMI) task (Henry, Messer & Nash, 2012). Switching subtest from Delis–Kaplan Executive Function System | • Sustained attention in general was a relative strength for both groups.  
• Weaknesses were most apparent in visual selective attention tasks, especially those when selection from a complex, crowded array is required.  
• DS group showed significant difficulties in most EFs in comparison to a TD group, especially in verbal set shifting;  
• DS population showed weaknesses and difficulties across modality in ELWM compared to TD group;  
• No significant differences between DS and TD groups in inhibition and fluency tasks;  
• WS and DS groups not directly compared; both compared to TD group only. |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/ setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
</table>
| Carretti et al., 2013 | Children and adolescents DS sample size: 20 mean CA=14.16 (SD=2.42; range=9.41-17.92); mean MA=5.17 (SD=5.17) Control group: TD group matched for MA | Italy research room | Cross-sectional Test     | WM          | Visuospatial memory task: Spatial-simultaneous and spatial-sequential VSWM task. | • DS individuals performed significantly worse in the spatial-simultaneous task compared to the TD group;  
  • Individuals with DS took less advantage of the pattern configuration in the spatial-simultaneous task than TD children;  
  • No significant difference in performance in spatial-sequential tasks between TD and DS groups.                                                                 | 13           |
| Costanzo et al., 2013 | Children, adolescents and young adults DS sample size: 15 (F=8; M=7). Mean CA=14.5 (SD=3.7; range=8.6–21.2); mean MA=6.2 (SD=0.9; range=4.8–8.7); mean IQ=53 (SD=13.5; range=36–83) Control 1: WS group (n=15) (F=7; M=8). Mean CA=17.6 (SD=7.4; range=10.7–34.9); mean MA=6.7 (SD=0.9); | Italy            | Cross-sectional Tests    | WM Attention | Selective attention (SA) (auditory): BVN test (Bisiacchi et al., 2005); Selective attention (SA) (visual): Sky Search/TEA-Ch (Manly et al., 2001)  
  Sustained attention (Sus/A) (auditory): Score task (SCO; TEA-Ch, Manly et al., 2001) | • Attention:  
  • Auditory SA: No group differences on BVN;  
  • DS and WS groups performed sig. worse (more errors) than TD group on SCO and SKY with large effect sizes;  
  • DS group scored sig. worse than WS group on BELLS; no sig. difference between ID groups and TD.  
  • WM:  
  • DS sig. worse than TD and WS on verbal WM (B-DST; NWR);  
<p>| 13           |</p>
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>2001</td>
<td>Range=5.10–7.8); Mean IQ=53 (SD=10.2; range=36–71)</td>
<td>Control 2: TD group (n=16) Matched for MA (F=78 M=8). Mean CA=7.4 (SD=0.8; range=6.1–8.4); mean MA=6.9 (SD=0.7; range=5.7–10); mean IQ=94 (SD=8.7; range=85–119).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Note: this review focuses on child and adolescent results only.</td>
<td></td>
<td>2001</td>
<td>Sus/A (visual): Bells Test (BELLS; Gauthier, Dehaut, &amp; Joanette, 1989)</td>
<td></td>
<td>• Both DS and WS groups sig. worse than TD on visuo-spatial WM (B-CBT).</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Verbal WM: backward version of the Digit Span (B-DST); Nonword Repetition task (NWR) (Vicari, 2007).</td>
<td></td>
<td>• Planning:</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Visual-spatial WM: backward version (of Corsi Block Test (B-CBT) (Orsini et al., 1987).</td>
<td></td>
<td>• No differences between participants with DS and TD on accuracy; sig. diff in execution time (DS spent more time than WS and TD on task)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Planning: Tower of London test (TOL; Shallice, 1982).</td>
<td></td>
<td>• Categorisation:</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Categorisation (verbal): Category Fluency Test (CAT) Mantyla et al., 2007).</td>
<td></td>
<td>• Verbal (CAT): DS scored lower than WS, but both comparable to TD scores;</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Categorisation (visual): Weigl</td>
<td></td>
<td>• Visual (WEIGL): ID groups scored sig. lower than TD.</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Shifting: Verbal (CAT-A): DS groups sig. worse than WS and TD;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Visual-spatial (TMT): DS sig. longer than TD in execution;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Visual-spatial (F/F): DS sig. longer than TD and WS in execution.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Inhibition: Verbal (STROOP): DS group sig. more interference than TD and WS;</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Visual (G/NG): no difference between groups in accuracy or execution time.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>---------------------</td>
<td>---------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>-------------------------------------------</td>
<td>---------------</td>
</tr>
<tr>
<td>Cuskelly et al., 2003</td>
<td>Children and adolescents</td>
<td>Australia</td>
<td>Cross-sectional Self-regulation</td>
<td>Tea Task; Gift Task (Block)</td>
<td>Color Form Sort Test (WEIGL; Spinnler &amp; Torgoni, 1987). Shifting (verbal): Alternate Category Fluency Task (CAT-A; Mantyla et al., 2007). Shifting (visual-spatial): Trail Making Test (TMT) (Reitan, 1958); Forma/Forma task (F/F) (Scarpa, 2006). Inhibition (verbal): the Stroop task. Inhibition (visual): go-no go task (G/NG; Van der Meere, Marzocchi, &amp; De Meo, 2005). Conclusion: • Both ID groups were challenged in auditory SusA (but not in auditory SA); visual SA (but not visual SusA); visual categorisation (but not verbal) and WM. • DS group were especially poor in shifting and verbal aspects of memory and inhibition compared to WS; • WS group were specifically poor in planning.</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td></td>
</tr>
<tr>
<td>------------------------</td>
<td>---------------------</td>
<td>----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>----------------------------------------------------------------------------------------------------------------</td>
<td></td>
</tr>
<tr>
<td>DS sample size: 25</td>
<td></td>
<td></td>
<td>laboratory of the</td>
<td>Tests</td>
<td></td>
<td>children on two of the three tasks (Gift Task and Self-imposed delay of gratification task):</td>
<td></td>
</tr>
<tr>
<td>(F=14; M=11); mean</td>
<td></td>
<td></td>
<td>FESSER C.</td>
<td></td>
<td></td>
<td>• There was no significant difference between groups on the Tea Task;</td>
<td></td>
</tr>
<tr>
<td>CA=10.3 (SD=2.05;</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• TD children were rated by their mothers as having significantly more self-control than were the children with</td>
<td></td>
</tr>
<tr>
<td>range=6.25-14.25);</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>DS;</td>
<td></td>
</tr>
<tr>
<td>mean MA=4.7 (SD=0.7;</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• Individuals with DS were less able to wait when working for a self-directed goal;</td>
<td></td>
</tr>
<tr>
<td>range=2.5-5).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>• The ability to delay gratification in DS group increases with MA.</td>
<td></td>
</tr>
<tr>
<td>Control group: TD</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>control group (n=32)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>F=14; M=18); mean</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CA=3.9 (SD=0.2), mean</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>MA=4.6 (SD=0.9).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cuskelly et al., 2016</td>
<td>Children and</td>
<td>Australia &amp;</td>
<td></td>
<td>Self-</td>
<td>Delay of</td>
<td>• DS group demonstrated significantly less waiting time compared to TD and MID groups;</td>
<td></td>
</tr>
<tr>
<td>adolescents</td>
<td>adolescents</td>
<td>UK University</td>
<td></td>
<td>regulation</td>
<td>gratification</td>
<td>• DS group had significantly less self-regulation than the other controls.</td>
<td></td>
</tr>
<tr>
<td>DS sample size: 22</td>
<td>22 F=10; M=11);</td>
<td>laboratory or</td>
<td></td>
<td>Cross-</td>
<td>task (Mischel</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(F=10; M=11); Mean</td>
<td>Mean CA=12.26 (SD=</td>
<td>conveniences</td>
<td></td>
<td>sectional</td>
<td>&amp; Baker, 1975).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CA=1.67; range=7-14.42;</td>
<td>1.67; range=7-14.42;</td>
<td>venue for family</td>
<td></td>
<td>Tests</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>mean MA=3.89 (SD=0.62;</td>
<td>mean MA=3.84 (SD=</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>range 3–5.25).</td>
<td>0.62; range 3–5.25).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control group 1: TD</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>group matched to DS</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>group for MA (n=43);</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(F=21; M=22); mean</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CA=3.84 (SD=0.45);</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>mean MA=3.84 (SD=0.45).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>--------------------------</td>
<td>-------------------------------------------------------------------------------------</td>
<td>-----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>----------------</td>
<td>-----------------------------------------------------------------------------------------------------------------</td>
<td>---------------</td>
</tr>
</tbody>
</table>
| Fortunato-Tavares et al., 2015 | Children and adolescents DS sample size: 15 (F=7; M=8) mean CA=10.25 (SD=2.58; range=7-14.17); mean IQ = 54.5 (6.9) | Brazil Research Lab | Cross-sectional Test | WM (verbal) | ABFW Child Language Test; TONI-III reflexive assignment sentences | - Children with DS had significant WM weaknesses compared to children with Typical Language Development (TLD)  
- Children with DS performed worse than all 3 other control groups on short WM condition;  
- Children with DS performed similarly to SLI group on long WM condition (both groups performed significantly worse than the other control groups);  
- No significant difference between children with DS and children with SLI and HFA when WM demands were higher (no WM load effect). | 13            |
| Green et al., 2006       | Children and adolescents DS sample size: 369 F = 169; M=200; CA range <5->19.       | USA Website     | Cross-sectional Survey  | Behaviorally flexibly function | Behavioral Flexibility Rating Scale | - Results show that children with DS scored in significantly lower in the BFRS than those with autism and Asperger Syndrome.  
- Children with DS had most problems in the situations represented by activity | 11            |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
</table>
| Goldman et al., 2005   | Children and adolescents DS sample size: 12 (F=9; M=3); mean CA=15 (SD=2.93); mean MA=5.6 (SD=0.64) Control group: TD children: (F=6; M=7); mean CA=5.6 (SD=0.45); mean MA=5.6 (SD=0.45) | Israel School | Cross-sectional Test | Attention | Reaction Time Tests for cued target location | • Results showed reaction time for participants with DS was significantly shorter/faster when compared to TD children, MA-matched (perhaps due to greater CA of DS group);  
• No significant difference between groups in ‘orienting behaviour’ to information cues within the task, suggesting comparable skills in selective attention. | 12            |
| Johns et al., 2012     | Children and adolescents DS sample size: 15 (F=8; M=7); mean CA=14.8 (SD=3.7; range=8-20). 11 siblings Control group: TD children (n=17) matched for MA | Australia Home, child care centre, school and university research room | Cross-sectional Tests | Inhibition | Hand reverse categorisation test  
Animal reverse categorisation test | • Participants with DS demonstrated significantly lower scores in the hand and animal reverse categorisation tasks, demonstrating poor inhibition ability when compared with TD children matched for MA. | 12            |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Landry et al., 2012</td>
<td>Children, adolescents and young adults. DS sample size: 11 Mean CA=14.6 (SD=4.15; range=8.75-21.25); mean verbal MA=3.5 (SD=1.6); mean non-verbal MA=4.42 (SD=0.9). Control group: Williams Syndrome (n=14). Mean CA=13 (SD=12.9); mean verbal MA=6.2 (SD=3.4); mean non-verbal MA=6.2 (SD=1.8).</td>
<td>Canada Research lab</td>
<td>Cross-sectional Test</td>
<td>WM Cognitive flexibility WM: Self-Ordering Pointing (SOP) Cognitive flexibility (CF): Dimensional Change Card Sort (DCCS) (Frye et al., 1995)</td>
<td>• DS group performed somewhat worse than WS group in CF but better in WM; • Comparisons were not drawn between the groups as they were not MA-matched; • EF performance did not increase with CA; • Verbal development was uniquely correlated with (and a good predictor of) CF and WM in both groups.</td>
<td></td>
</tr>
<tr>
<td>Lanfranchi et al., 2004</td>
<td>Children and adolescents Study 1: DS sample size: 18 Mean CA=11.75 (SD=3; range=7-16); mean MA=5.42 (SD=0.8; range=4.42-6). Control group: TD group (n=18), matched for MA. Mean CA=5.17 (SD=0.58; range=4.42-5.83); mean MA=5.17 (SD=0.83). Study 2: DS sample size: 22</td>
<td>Italy School and research room</td>
<td>Cross-sectional Tests</td>
<td>Verbal and visuospatial WM 4 x verbal WM tests 4 x visuospatial WM tasks</td>
<td>• The higher the WM control required, the greater the differences in the performance of DS children and TD controls; • The differences between the performance of DS children and controls was related to the nature of the task, namely low level or high level WM control; • For tasks requiring low control, DS children demonstrated difficulties in verbal but not visuospatial WM tasks; • For tasks requiring higher controls, DS children showed</td>
<td></td>
</tr>
</tbody>
</table>
## Table: Study Design and Methods

<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Lanfranchi et al., 2009a</strong></td>
<td>Children and adolescents DS sample size: 34 (F=15; M=19); Mean CA=12.5 (SD=2.42; range=7-17.92)</td>
<td>Italy Research lab</td>
<td>Cross-sectional Test</td>
<td>Visuospatial WM (VSWM)</td>
<td>A battery of VSWM tasks</td>
<td>Individuals with DS scored significantly poor than TD groups in a series of VSWM tasks; WM deficit in DS is selective rather than pervasive; Individuals with DS performed significantly worse in the spatial-simultaneous tasks, but not in the spatial-sequential tasks. In fact, the DS group performed significantly better in the coding subtest of the spatial sequential tasks.</td>
<td>13</td>
</tr>
<tr>
<td><strong>Lee et al., 2015</strong></td>
<td>Children and adolescents DS sample size: 30 (F=15; M=15; Mean CA=11.34 (SD=3.02; range=7-17) Nonverbal IQ mean=52.41 (SD=13.2;</td>
<td>USA</td>
<td>Cross sectional Caregive r report</td>
<td>EFs</td>
<td>BRIEF (parent)</td>
<td>DS group gained highest T-scores in monitoring (MO), WM and PO on the EMI (metacognition index) respectively, and highest scores on the shifting and inhibition scales of the behaviour regulation index</td>
<td>13</td>
</tr>
</tbody>
</table>
Control 1: CA-matched TD (n=30), mean CA=11.28 (SD=2.69; range=6-17). Nonverbal IQ Mean=110.37 (SD=11.97; range=86-139)

Control 2: CA-matched Sex Chromosome Trisomy (XXX & XXY [+1X]) (n=30) CA mean=11.61 (SD=3.29; range=5-18). Nonverbal IQ Mean 100.75 (SD=15.70; range=74-135)

Mason et al., 2015

Children, adolescents and young adults
DS sample size: 62 Mean CA= 11.89 (SD=3.35; range=7-21), with (n=27) and without (n=35) 7R

USA Research lab
Cross-sectional Tests Caregiver reports

Global EF: BRIEF Attention: Caregiver reports Cognitive Flexibility: CANTAB Intra/Extra Dimensional Set Shift (IED)

DS and +1X groups gained significantly higher scores (greater difficulty) on both BRI and MCI/EMI index compared to TD group.

The weakest functions in DS were WM and MO.

The strongest EFs in DS were EC and organisation of materials (OM).

Nontrisomy genetic factors may contribute to individual differences in ADHD symptoms in persons with DS.

White individuals with DS and at least one 7R (genotype) had elevated scores on the BRIEF, especially in ISCI, EMI and GEC, which indicates more difficulties;

Hispanic children with DS and 13

7-repeat allele, which mediates dopamine activity and relates to individual differences in EF, including effortful control (Smith et al., 2012) and impulsivity (Congdon & Canli, 2008) in those without DS.
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
</table>
| Memisevic & Sinanovic, 2014 | Children and Adolescents DS sample size: 30 F=15; M=15; mean CA=11.8(range=7-15; SD=2.8); Mild Intellectual Disability =12%; Moderate Intellectual Disability =18%  
Control group 1: Unknown aetiology group (n=30); F=11; M=19; mean CA=11.5(2.6); Mild Intellectual Disability =20%; Moderate Intellectual Disability =10%.  
Control group 2: Organic cause/ other genetic cause group (n=30); F=20; M=10; mean CA=10.6(2.7); Mild Intellectual Disability =10%; Moderate Intellectual Disability =20%.  
All 3 x groups compared to normative data for TD | Bosnia Special education school x 2  
Cross-sectional  
Caregiver report | Global EF  
BRIEF (parents) | • BRIEF: Children with DS, similar to other ID children, had a significant deficit in EF.  
• All groups had a clinical significant or near-significant weaknesses in EF behaviours on all the BRIEF scales.  
• DS group had clinical significant difficulties in all scales except Inhibit scale (which was approaching significance).  
• No statistically significant differences in EF between sex.  
• Children with DS scored significantly worse than children with unknown aetiology of ID group on the Shift scale (but not compared to children with organic/other genetic cause aetiology ID group).  
• Apart from Shifting, no other statistically significant differences were found between the groups. | 13 |
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pezzuti et al., 2018</td>
<td>Children and adolescents. DS sample size: 128 (M=72; F=56). Mean CA=12.4 (SD=2.59; range=7-16).</td>
<td>Italy Laboratory</td>
<td>Cross-sectional WISC questionnaire</td>
<td>WM</td>
<td>WISC-IV including the working memory index subscales (digit span and letter-number sequencing).</td>
<td>• Working memory index and processing speed index gained worst scores from all the subscales.</td>
<td>12</td>
</tr>
<tr>
<td>Purser et al., 2015</td>
<td>Children and adolescents</td>
<td>UK and France University lab</td>
<td>Cross-sectional Tests</td>
<td>Attentio n  Inhibitio n</td>
<td>Go/No Go task</td>
<td>• Participants with DS who had relatively high levels of non-verbal ability performed at a similar level to TD participants; • Participants with DS were significantly poorer performers in Go/No Go tasks than participants in both control groups; • DS participants not matched to either WS or TD group, so difficult to draw definitive conclusions regarding performance differences on the tasks.</td>
<td>13</td>
</tr>
<tr>
<td>Traverso et al., 2018</td>
<td>Children and adolescents. DS sample size: 32 (M=10; F=22). Mean CA=14.4(SD=5.4; range=6-25). Control group 1=TD group 5 years old (n=35) (M=17;</td>
<td>Italy Treatment centre</td>
<td>Cross-sectional Test</td>
<td>Inhibitio n</td>
<td>Inhibition battery (Go/No-Go task; preschool matching familiar figure task; fish flanker task; dots task)</td>
<td>• Children from the DS and 5TD group performed similarly; • Children from the 6TD group significantly outperformed both the DS and 5TD groups.</td>
<td>13</td>
</tr>
<tr>
<td>Citation</td>
<td>Sample and controls</td>
<td>Country/setting</td>
<td>Study Design and Methods</td>
<td>EF Measured</td>
<td>Measures of EF</td>
<td>Summary of Main Findings, only EF findings</td>
<td>Quality score</td>
</tr>
<tr>
<td>----------</td>
<td>---------------------</td>
<td>----------------</td>
<td>--------------------------</td>
<td>-------------</td>
<td>---------------</td>
<td>-------------------------------------------</td>
<td>--------------</td>
</tr>
</tbody>
</table>
| Trezise et al., 2008 | Children and adolescents; DS sample size: 11 (F=6; M=5); mean CA=14.1 (SD=2.91; range=8.75-18.4); mean MA=6.12 (SD=1.32; range=4.83). Control group: non-specific intellectual disability (NSID) group (n=16), matched for MA. (F=5; M=11); mean CA=11.6 (SD=2.94; range=7.58-18.7); mean MA=7.12 (SD=1.56; range=5.08-10.58). | Australia | Cross-sectional Test | Attention (sustained) | SART (visual and auditory) | • Significant group differences were found in the visual modality as the DS group performed better than the control, and mental age was also found to effect error rates in the visual condition.  
• No effect of group or mental age was observed in the auditory SART.  
• For individuals with DS, the presentation of education material in a visual medium may facilitate sustained attention and thus learning. | 11 |
| Visu-Petra, et al., 2007 | Children, adolescents and young adults; DS sample size: 25 (F=13; M=12; mean CA=14.4 (SD=3.5; range=8-21); mean MA=5.75(SD=1.08; range=4-7.3). Participants institution (generally school) | Romania | Cross-sectional Tests | Visual-spatial memory (VSWM) | Cambridge Neuropsychological Test Automated Battery (CANTAB) – 5 VSWM tasks. | • As memory load increased, in recognition tasks (spatial or visual), or when visual and spatial demands were combined, the performance of individuals with DS performance was significantly weak compared with MA controls; | 13 |

F=18. Mean CA=5.5 (SD=0.2; range=5.1-5.9). Control group 2=TD group 6 years old (n=30) (M=17; F=13). Mean CA=6.1(SD=0.4; range=6-7).
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wilde &amp; Oliver, 2017</td>
<td>Control group: TD group (n=25) matched for MA. (M=11; F=14). CA range=4.25-8); mean MA=5.5 (SD=1.16)</td>
<td>UK Home</td>
<td>Cross-sectional Survey</td>
<td>Global EF</td>
<td>BRIEF-P</td>
<td>DS group demonstrate significantly poorer performance than TD group in one task from WM assessment (returns to previously investigated locations); Non-significant spatial STM difference in mean span (TD group = higher); VSSTM = relatively spared in DS sample, which may conceal a still developing ability in TD children.</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td>Control group 1=Smith-Magenis syndrome (n=13) (M=7; F=8). Mean CA=8.9(SD=3.8; range=3-11.25).</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

- Children with SMS showed more difficulties than children with DS on the Inhibit, Shift and Emotional Control scales, but not Working Memory, Plan/Organize;
- In SMS there were difficulties only in Working Memory relative to both Shift and Emotional Control and in Inhibit relative to Shift;
- In DS there were weaknesses in Working Memory relative to Inhibit, Shift, Emotional Control and Plan/Organize, but fewer weaknesses in Emotional Control compared to Inhibit, Plan/Organize and Working Memory.
- When compared to the...
<table>
<thead>
<tr>
<th>Citation</th>
<th>Sample and controls</th>
<th>Country/setting</th>
<th>Study Design and Methods</th>
<th>EF Measured</th>
<th>Measures of EF</th>
<th>Summary of Main Findings, only EF findings</th>
<th>Quality score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>normative mean T score of 50, children with SMS had higher mean T scores (denoting greater difficulties) on all clinical scales of the BRIEF-P, whereas children with DS had significantly higher mean T scores for Inhibit, Working Memory and Plan/Organise but not Shift or Emotional Control.</td>
<td></td>
</tr>
</tbody>
</table>
Chapter Four: Study Two

4.1 Introduction

4.1.1 Overview

The findings of the systematic review revealed that working memory (WM) is the executive function (EF) that is most significantly challenged in children and adolescents with Down syndrome (DS). The adverse impacts of EF disorders necessitate research to identify intervention approaches with high levels of efficacy in promoting EF development and learning. Primarily, study two aimed to develop, implement, and evaluate an intervention that helped teachers to improve WM for children with DS; as mentioned in chapter one, this is the first time that such a WM intervention for children with DS has been carried out in Kuwait. Study two also rests on the idea that, as described within the conceptual model underpinning this thesis (Morton’s (2004) causal model framework), the environment can potentially influence the cognitive outcomes of those with DS, including EF, if it is appropriately harnessed.

This chapter therefore reviews existing EF interventions in different contexts, focusing in particular on WM interventions for individuals with DS, with the aim of highlighting the extent to which these interventions have been effective, particularly with children with DS. Moreover, this chapter aims to assess the benefits and limitations of different elements/characteristics of EF interventions, a process that informed the design of the intervention that was used in the present study. In each case, typically developing (TD) and intellectually disabled (ID) populations are considered first, before focusing on DS populations. A discussion of WM interventions in Arab contexts is also given. This section also discusses the importance of teachers’ involvement and the participation of both teachers and parents in WM intervention work with children. This is because research indicates that, by training teachers, there is greater potential for any positive effects of the intervention to be sustained, as teachers may continue to
use these strategies with their pupils after the intervention has officially ended (Friend et al., 2014; Lai et al., 2009).

While this review of EF interventions is structured as outlined above, it should be noted that there are several complicating factors in such a review. For example, EF interventions do not all have the same aims, they use a wide variety of techniques and, as Lanfranchi et al. (2010) point out, the term ‘executive function’ is very general, and covers a wide range of behaviours and cognitive functions. These complicating factors make the field of EF interventions very broad.

Moreover, whilst there are numerous different kinds of EF interventions, this chapter primarily focuses on school-based interventions that are/have been delivered by teachers, which adopt interventions that focused on teaching approaches within a classroom/school context. This was the tentative design of my own intervention at this stage; hence, these interventions are of particular interest.

When developing my own intervention, a crucial factor to consider was the suitability of the intervention for the target population (children with DS), and whether the strategies involved would be suitable for the abilities of those with DS. Furthermore, it was essential to consider the sustainability of any intervention effects, that is, whether it could create long-term WM benefits, long after the intervention officially ends (Yeung et al., 2016; Roesken-Winter, Hoyles & Blömeke, 2015). Thus, the sustainability of interventions is also discussed where possible. Finally, after reviewing the literature on EF interventions and methods adopted in this study, the results and discussion surrounding the efficacy of my own intervention are presented.
4.1.2 Executive Function Interventions

EF interventions can be beneficial in supporting the development of EF and other cognitive skills/functions (Doebel, Rowell & Koenig, 2016; Ahmed et al., 2019). According to Neitzel (2018), EF interventions are especially important when delivered by teachers, as it increases teachers’ abilities to improve the learning, academic attainment and behaviour of students. First, there is a need to determine the effectiveness of EF interventions for TD children, as well as those with DS. Additionally, it is essential to evaluate the impact of teacher involvement in the development and implementation of interventions that improve EF, and especially the WM, for both TD children and those with DS.

4.1.2.1 Executive Function Interventions for Typically Developing Children

EF interventions, as mentioned above, may use range of techniques, as seen in the empirical studies of Traverso, Viterbori and Usai (2015), Diamond and Lee (2011), Serpell and Esposito (2016), Dawson (2014) and Spencer-Smith and Klingberg (2015), which share a common general aim of reducing developmental disabilities through the EF interventions that they investigate. This applies both to TD children and to children with ID, for all of whom EF is fundamental in their development.

Traverso et al. (2015) for example conducted a group-based EF intervention with 75 five-year-old children, targeting cognitive flexibility, inhibitory control and WM. The intervention aimed to improve EF skills via various game activities conducted in small groups, which required increasing levels of inhibitory control, cognitive flexibility and WM. These activities were conducted through a series of 12 intervention sessions over four weeks. The findings show that the implementation of the intervention significantly enhanced all domains of EF in these preschool children. Specifically, Traverso et al. (2015) found that compared to controls, the intervention group exhibited increased abilities to control on-going responses and to delay gratification (inhibitory control), to
manage high cognitive conflict (cognitive flexibility) and to update and process information (WM). This indicates that group-based interventions with young children can be effective for EF promotion. This indicates that this intervention focused on increasing children's abilities in line with age-appropriate norms of what might be expected in TD children. All the aforementioned EF skills increase rapidly over ages 5-12 years, thus age-appropriate interventions focusing on these specific areas should take into account what might normally be expected for a TD individuals’ CA.

Unfortunately, the training of teachers to deliver the intervention in Traverso et al.'s (2015) study, whilst undertaken by a trained psychologist, was not specifically evaluated for its effectiveness (as opposed to the effectiveness of the intervention more generally). There are also concerns with generalisability, as teachers in other contexts may not receive the training or deliver the EF strategies in the same way as the teachers in Traverso et al.'s (2015) study, which was conducted in Italy. According to Mumford and Schisterman (2019), generalisability in research is critical to ensuring that an intervention measure can be applied to wider populations. Moreover, this study did not conduct any follow-up evaluations in EF; thus, it is unclear whether there was any long-term benefit of the intervention conducted. Finally, there was no active control group; the authors themselves note that in future, an active control group matched in terms of intervention time and effort to the training group should be incorporated (Brehmer et al., 2012). This is for both ethical reasons (ensuring a benefit for the control group), as well as to uncover whether simply being part of any intervention group benefits EF or leads to demand characteristics that might bias the results.

Another form of EF intervention investigates the effect of physical activities in enhancing the development of EFs. The review of research into EF interventions by Diamond and Ling (2016) asserted that teachers need to provide students
with a diverse range of activities, as they all contribute towards the development of children’s EF. The review mainly focused on the incorporation of aerobics as one activity in school (focusing on studies such as Erwin et al., 2012; Mahar et al., 2006); it found that children and adolescents with weaker EFs significantly benefitted from physical activity, as global EF improved when movement was incorporated into learning. For this reason, it can be argued that the implementation of aerobic exercise as an EF intervention could be critical in the education system as it can help in managing underachievement. Findings from this study are consistent with those of Traverso et al. (2015) and may seem to support the claim that EF interventions can lead to the positive EF development of children. It must be noted, however, that in adults at least, ‘mindless’ physical activity has been found to be less effective in improving EF than physical activity combined with EF challenges/cognitive training (Moreau et al., 2015; Oswald et al., 2006). Oswald et al. (2006) also found that the combination resulted in significantly higher levels of EF than the cognitive training alone in older adults, although it is unclear whether this applies to children and adolescents.

Programmes such as TAKE10! and Move for Thought (Vazou & Smiley-Oyen, 2014) have been implemented in schools as a result of findings that physical activity, combined with academic instruction, helps to further understanding of academic concepts (Erwin et al., 2012; Kibbe et al., 2011; Kubesch & Walk, 2009).

Yet the extent to which programmes such as these specifically relate to improvements in EF has not been fully investigated, nor has it clearly discriminated between children and adolescents; for instance, there appear to be rapid increases in EF during the early years, especially inhibition (Best & Miller, 2010). The question remains, therefore, whether interventions ought to be focused on periods of rapid skill acquisition, such as between ages 3-6, or whether they should be used during more stable, gradual EF development,
during adolescence – or indeed, throughout the entirety of an individual’s development into adulthood.

Some studies, however, have cast doubt on the value of physical activity to improve EF. In response to Diamond and Ling (2016), Hillman et al. (2019) conducted a review of the research in this area and concluded that interventions based on physical activity have been fraught with errors, with intervention methods often not sufficiently detailed and, on occasion, the findings misrepresented in some studies. Therefore, interventions using physical exercise to improve EF need greater investigation. Moreover, physical exercise has been found to improve sleep (Kline et al., 2014) and reduce stress and symptoms of depression (Haslacher et al., 2015); lack of sleep has also been found to impair EF (Labelle et al., 2015), as does stress (Liston et al., 2009). However, there is no study exploring whether EF benefits from exercise may be due to the indirect impact exercise has on reducing stress and improving sleep, with better EF as a bi-product of this.

Moreover, EF interventions generally focus on improvements to global EF; it is unclear whether interventions may be more effective when they focus predominantly on one specific EF, such as WM. In many ways, it may be difficult to target only one EF, given that other EFs may be involved in any task and are therefore elicited also. Yet it is possible that broad EF interventions may be less effective in boosting academic achievement than interventions that target specific EFs; Serpell and Esposito (2016) for example found limited evidence of the transfer of any EF gains from global EF interventions to academic outcomes throughout childhood and adolescence. Interventions often focus on a range of EF skills however, although some EF skills (particularly working memory) have a stronger association with academic outcomes than others (Pascual, Muñoz & Robres, 2019). Therefore, broad interventions may not necessarily impact academic skills. Targeting specific EFs may be more impactful on academic
achievement (Daunhauer et al., 2020); for example, WM has been shown to correlate very strongly with academic achievement (Blair & Razza, 2007; St. Clair-Thompson & Gathercole, 2006). Therefore, there may well be value in schools structuring their learning environment into one that supports the development of WM in children. WM interventions are discussed in more detail in due course.

It is also possible that interventions focused on a single EF will benefit other EFs. For example, inhibition training skill is another kind of single EF-focused intervention that teachers can employ, as inhibition indirectly impacts WM. Zhao, Volckaert and Noel (2015) investigated the impact of inhibition on WM using a training programme for 20 minutes on a daily basis. The study involved 47 TD pre-schoolers with a mean age of 5 years – an age when rapid increases in inhibition abilities in particular have been found to occur (Best & Miller, 2010). Inhibition training strategies in the intervention groups consisted of exercises and games within a go/no go task, that were aimed at increasing different inhibition components, such as impulse management, inhibition of external distractors, and interruption of on-going responses, while the control group only had handicraft sessions. The study found that both inhibition and the WM of the children improved (the latter as a transfer effect). It could be argued that this study also indicates that interventions focused on one function may be as effective or even more effective than interventions aimed at multiple EFs, as there may well be transfer effects to other EFs. Some other studies have confirmed this finding; Aydmune, Introzzi and Lipina (2019) for example found that inhibition training in a sample of 6-8-year-old TD children did indeed result in improvements to visual spatial WM. However, this transfer effect only represented a short-term effect. Moreover, as far as this researcher is aware, there is currently no study that has directly compared EF outcomes of broad EF versus specific EF interventions.
EF interventions for TD children have therefore been shown in some studies at least to significantly improve inhibition, working memory and flexibility, although not necessarily emotional control (Traverso et al., 2015) and also have positive effects on a trainee’s organisation, time management, prioritisation, task initiation, metacognition, goal-directed persistence and response inhibition (Dawson, 2014). It must be noted however that there could be a publication bias at play, with some interventions with non-significant outcomes less likely to be published. Moreover, there is a lack of consensus about which kinds of interventions are most successful in terms of improving EF, and there is limited evidence that any such interventions directly improve academic outcomes. Some researchers have compared explicit training to implicit training (EF activities embedded in children’s everyday activities) and found that implicit training has greater ecological validity and is more enjoyable than explicit training (Takacs & Kassai, 2019). Finally, it is critical that the age of participant is taken into consideration when designing appropriate interventions; not simply due to the intervention content, which must be age-appropriate, but in terms of the age at which an intervention may have the most impact. For instance, with research indicating a spurt in WM development between the ages of 14-15 years (Poon, 2018), it may either be more prudent to conduct interventions during this age range to maximise this rapid increase, or to conduct these interventions during a period of less rapid development, such as during middle childhood. In addition, given the finding that ‘hot’ EFs tend to decline from the age of 15 years (Poon, 2018), it may be effective to conduct some interventions around the age of 15, so as to reduce or mitigate any significant declines around this CA. This can be accompanied by interventions in the early years to ensure that individuals are receiving as much attention and input as possible on EF skill development, to lay a strong foundation in these skills as they progress through childhood and adolescence.
4.1.2.2 Executive Function Interventions for Children with Developmental Difficulties

The previous section reviewed several interventions with TD populations. This section focuses on different interventions for children with cognitive and/or development difficulties except those with DS, which are covered subsequently.

Some school-based interventions can be considered as a form of EF training. ‘Unstuck and On Target’ (UOT) (Kenworthy et al., 2014) is a home- and school-based EF intervention for children aged 7-11 years with autism spectrum disorder (ASD), delivered by school staff and parents, targeting flexibility, planning, goal-setting and reducing insistence on sameness, using a cognitive-behavioural programme. The UOT intervention involved classroom activities with supportive materials for children, plus sessions for teachers and parents. In the study, children with ASD received one of two interventions delivered by school staff to compare UOT and a social skills intervention. The results showed significantly greater EF improvements after UOT than the social skills intervention, although there was no indication about the sustainability of the intervention effect as there were no follow-up assessments. It must also be noted that the age range of 4 years may mean that the intervention was too challenging for some individuals and not sufficiently challenging for others; perhaps using targeted, age-appropriate interventions, or better still, tailored interventions depending on the individuals’ current EF abilities, may be more effective. However, recruiting across a narrow age range may be challenging, especially given the wide variety in individual differences in skillset, IQ and so on. It may also be potentially unethical to exclude some individuals based on CA that may benefit from an intervention.

Whilst the age range may still be up for debate, the findings from Kenworthy et al. (2014) do support, however, the effectiveness of contextually-based EF interventions for children with developmental disabilities, in particular via a
school-based intervention, which forms the context of the present study. The study by Kenworthy et al. (2014) also shows that such interventions can be effective for children with ASD, who often experience some difficulties with EF similar to the difficulties experienced by children with DS, in addition to which, children with DS often have co-occurring conditions such as ASD (Pritchard et al., 2015). These observations strengthen the rationale for a school-based DS intervention.

There is also research investigating whether the improvement of one aspect of EF could support improvement in other abilities, such as challenging behaviour (CB) (which was the main focus of the control group in the present study). For example, de Vries et al. (2015) investigated a computer game entitled ‘Braingame Brian’, designed for children aged 8-12 years with ASD who experienced EF difficulties. Participants were randomly assigned to either adaptive WM training, adaptive cognitive flexibility training (both on the computer game) or to non-adaptive control training (mock training). The outcome measures included attention, inhibition and parent ratings of daily life, EF, social behaviour, ADHD-behaviour and quality of life. It was found that children in all groups who completed the training improved in all measures except inhibition. In the WM condition, there was a general pattern of improvement on near transfer WM and far transfer (ADHD behaviour). By comparison, those in the cognitive flexibility condition demonstrated a non-significant trend toward improvement on near transfer flexibility at a 6-week follow-up. Yet de Vries et al. (2015) themselves assert that this improvement may be due to practice effects, and that, furthermore, the parental attention received as a result of jointly visiting the test sessions and supporting their child to participate in the training may underpin any improvements in behavioural problems (see also Forehand, Jones & Parent, 2013).
The types of EF interventions reviewed above have targeted other disabilities, such as ASD, with varied effectiveness. Some drawbacks were noted in these studies, such as the absence of follow-up tests to evaluate the sustainability of the intervention effect. However, from this review, it appears that interventions based on strategies taught by teachers can – in some cases – achieve improvement. To illuminate whether such strategies might be applied to children with DS or if specific approaches should be adopted, the following section reviews studies of EF interventions for children with DS.

4.1.2.3 Executive Function Interventions for Children with Down Syndrome

Having considered EF interventions in TD children and children with developmental disabilities, this section focuses specifically on EF interventions for children with DS. Children with DS perform poorly in measures of various EFs, as explored previously in chapters two and three, and the variety of EF difficulties corresponds to a variety in intervention aims and types. In this section therefore, the different types of EF interventions for children with DS are reviewed, in order to ascertain the effectiveness of EF interventions for this population group. Moreover, it aims to explore any gaps in the research in this area, and to ascertain which intervention elements are deemed to be beneficial when conducting an EF intervention for children with DS in particular.

Physical activity is one strategy to enhance EF amongst children with DS. For example, Holzapfel et al. (2016) conducted a study to investigate whether sports interventions can be used by teachers to enhance the EF of adolescents (aged 18 years) with DS. The study evaluated how various EF abilities were affected by 8 weeks of assisted cycling therapy (where participants’ pedalling rates were augmented with the use of a motor to maintain a particular speed), voluntary cycling (self-selected pedalling rate), and no cycling and found inhibition significantly improved in the assisted cycling therapy group; set-shifting (cognitive flexibility) and cognitive planning improved in the voluntary cycling
group. There were no significant improvements in the no cycling group. The study was replicated by Ringenbach et al. (2016) to further study the impact of cycling on WM; they revealed that significant improvements in WM in adolescents with DS were only found in the group undertaking assisted cycling therapy. Yet these studies only focused on adolescents in their late teens; it may be that cycling differentially affects the EFs of children with DS.

Tungate (2016) also examined the relationship between exercise and EF in individuals with DS, through correlating steps per day and caregiver reports of physical activity with EF measures (specifically, the Behaviour Rating Inventory of Executive Function (BRIEF)). This study found that exercise in terms of steps per day and physical activity ratings did not positively correlate with EF in individuals with DS aged between 6 and 18 years. Tungate’s (2016) study thus contradicts the findings from Holzapfel et al. (2016) and Ringenbach et al. (2016) on the importance of exercise in promoting EF. However, this might be because the exercise type differed in the two groups (steps per day may not provide the same level of physical exertion as cycling for instance), and there was no specific intervention in Tungate’s (2016) study; it could be that the intervention in and of itself contributed to greater EF, not necessarily the activity, for instance. It may also be that EF interventions involving physical exercise have greater impact for different age groups, such as adolescents, perhaps given that physical development (motor control and coordination, balance and muscle development) may develop more slowly in DS populations compared to TD peers (Pitetti, Baynard & Agiovlasitis, 2013). Adolescents may therefore be more able to benefit from physical exercise-based interventions once their physical skills have become sufficiently developed. As a wide age range was included in Tungate’s (2016) study, it is possible that any improvements in adolescents in this study were masked by a potential lack of improvement in the younger participants. Therefore, the age at which individuals with DS are
involved in any EF intervention may play a role in contributing to the findings, although this requires further research to clarify.

Although some sport interventions have achieved promising results, there are several considerations that might also limit their applicability with the DS population. For example, the use of exercise as an intervention would involve adults with significant training in supporting children with DS in sports, to reduce the risk of injury demands significant attention, as children with DS are known to have poor muscle balance (Smith, 2001). Specialised staff may also be required, such as sports coaches or physiotherapists, to assess the children to firstly ascertain what kind of intervention might be suitable, which may be costly or logistically difficult. Also, it may be difficult to conduct such an intervention in an inclusive school, as it would require access to sporting facilities at specific times of the week, which might not always be possible, due to timetable clashes with other classes. However, in disability-specific schools in Kuwait not under the inclusion system, this would not present a problem, as their facilities are not shared.

Pharmacological interventions are another approach for people with ID and for people with DS in particular. Some studies include EF measures. Lobaugh et al. (2001) for example used Piracetam in a double-blind placebo-controlled design together with several tools for evaluating the EF (cognitive flexibility, inhibition and shifting) of 25 children (aged 6.5-13 years) with DS, and observed no differences between the control and experimental groups after the intervention. The researchers concluded that the medication did not enhance cognition or behaviour; thus, such an intervention might be inappropriate in the current study. Indeed, a review of nine studies investigating the use of pharmacological interventions to treat cognitive decline in adults with DS by Livingstone et al. (2015) found that adults with DS receiving medication did not improve their cognitive abilities compared to those that did not receive any medication.
Livingstone et al. (2015) also identified various negative side-effects of medication, including dizziness, headache and nausea. Whilst this review focused on adults with DS, instead of children and adolescents as in the present study, it does suggest that at this time, pharmacological interventions do not seem to be an effective strategy for the improvement of EF in children with DS.

Similarly, Johnson et al. (2003), aimed to investigate whether acetylcholinesterase inhibitor donepezil could improve cognitive functioning (language, attention, motor skills, memory, arithmetic, conceptualisation and visuospatial ability) in 19 adults with DS and without dementia. However, although language scores improved somewhat, there was no demonstrable improvement from baseline in any of the cognitive subtests, behavioural scores or caregiver ratings at 4 and at 12 weeks. No significant improvement was found in cognitive function, especially in attention, which is considered an EF, and in visuospatial ability, which is relevant to WM. Part of the evaluation of participants was caregiver assessment, which might be beneficial in giving a different perspective from others (such as teachers or scientists). Moreover, this study included follow-up assessments to ascertain the sustainability of the intervention effect. Thus, despite the fact that pharmacological intervention does not seem to be an appropriate route for enhancing EFs, nor an approach that would be feasible in the present study (given the need for extensive medical knowledge/experience), some of the elements of this research, such as caregiver assessments and follow-up assessments, may be beneficial to incorporate in the evaluation of an intervention to establish a different view on any immediate and longer-term EF changes/improvements.

Summary
There are therefore various findings surrounding interventions for improving EFs for TD population. Research indicates a range of cognitive interventions for TD individuals that may assist in their development of EFs, including increased
abilities to control on-going responses and to delay gratification (inhibitory control), to manage high cognitive conflict (cognitive flexibility) and to update and process information (WM) (Traverso et al., 2015). EF interventions also have positive effects on an individual’s/participant’s organisation, time management, prioritisation, task initiation, metacognition, goal-directed persistence and response inhibition (Dawson, 2014). There is therefore some (limited) evidence that improving one EF (such as inhibition) can have a transfer effect on other EFs (such as WM, as found by Zhao, Volckaert and Noel (2015)), although this may not always be the case. Physical activity/movement combined with cognitive training/interventions has been found to improve EFs more than either approach alone for TD children (Erwin et al., 2012; Kibbe et al., 2011; Kubesch & Walk, 2009; Mahar et al., 2006; Moreau et al., 2015; Oswald et al., 2016), although the issues with some studies, as reported by Hillman et al. (2019), indicate that findings must always be interpreted with a degree of caution.

There is more limited research regarding EF interventions for children and adolescents with DS. Research has hitherto focused on interventions involving exercise (e.g., Holzapfel et al., 2016; Ringenbach et al., 2016; Tungate, 2016), revealing contradictory evidence (research suggests that aerobic exercise with a mindful/cognitive component or challenge (such as yoga or taekwondo) is more effective than those that do not include such a component (Diamond & Ling, 2016)) or pharmacological interventions (Lobaugh et al., 2001), which may not appear to be the most suitable or practical choice for improving EF in this group of individuals (Hart et al., 2017). Most pharmacological interventions focus on adults, and most exercise-based interventions focus on adolescents, examining global EF instead of focusing on a specific EF. There does appear to be a lack of research focusing on EF interventions for children with DS in particular.
Although the studies reviewed in this section enabled identification of some useful elements of intervention evaluation (e.g., the use of caregiver reports to provide another perspective on any EF changes in the children with DS), they do seem to rule out the use of physically-based interventions and pharmacological interventions in the present study. Therefore, it appears that a cognitive-behavioural intervention may, by default, be the best option, particularly given the evidence of their effectiveness with children with other developmental disabilities, such as ASD. However, broad cognitive-behavioural EF interventions for children with DS have not been reviewed here, given the lack of these interventions for improving general EFs in this particular population group. The next section therefore investigates WM interventions only, as WM was shown in the previous chapter to be the most delayed function in children and adolescents with DS, and therefore formed the focus of the intervention in the present study.

4.1.3 Working Memory Interventions

This section reviews various interventions that aim to improve WM for a variety of populations (both DS and non-DS) in order to understand the efficacy and benefits of different types and features of interventions.

4.1.3.1 Working Memory Interventions for Typically Developing Children

The structure of WM was described in section 2.5.1. WM interventions can potentially help boost the cognitive development and academic achievement of children (Alloway & Alloway, 2010), in particular, in literacy and numeracy (Christopher et al., 2012; Geary, Hamson & Hoard, 2000). WM interventions are usually conducted with the TD population. For instance, Roberts et al. (2016) found that WM interventions temporarily improved cognitive function, such as visuospatial STM, in 452 TD children aged 6-7 years who had low WM. This effect was retained at 12-month but not 24-month follow-up. However, Rode et al. (2014) and Sala and Gobet (2017) reported no measurable benefits of WM
interventions in terms of literacy or numeracy performance in TD children. Although there are concerns about the transfer effectiveness of WM interventions to other domains such as academic achievement, some school-based WM interventions have achieved promising results, as discussed below. In these interventions, an essential role is played by the teachers.

Because of the association between WM and academic achievement, as mentioned above, some interventions focus on improving academic skills and explore the effect of this on WM. Banales, Kohnen and McArthur (2015) conducted a study to determine the relationship between weak word reading accuracy and poor verbal WM, as well as to establish their causal relationship. The study involved four children with a mean age of 9.92 years. Teachers provided an eight-week programme of verbal WM training activities and another eight weeks of reading training activities for each child. The findings showed that verbal WM activities were effective in improving verbal WM in two children, but did not improve their reading accuracy. Interestingly, the reading training intervention was critical in improving word accuracy reading in all four children, although it did not have any impact on their verbal WM. The study thus suggests that an intervention offered by teachers to improve WM will only lead to positive results on the task being focused on in the intervention, limiting the near- and far-transfer effects of the intervention. Unfortunately, the results from Banales et al. (2015) were obtained from a very small sample, which diminishes the reliability of the study, and the children were all between 9-10-years-old – a narrow, specific age range that may provide a limitation to understanding the efficacy of the intervention. A wider age range could perhaps have established whether different age groups may have benefitted more from an intervention of this kind, given that WM is believed to mature steadily in TD individuals, with individual differences and confounding factors playing a role once children are at school (Cowan, 2016).
Similar to Banales et al. (2015), Henry, Messer and Nash (2014) examined the impact of a face-to-face WM training intervention offered by teachers on performance in WM, word reading and mathematics. The intervention was provided in 18 sessions of 10 minutes, three times a week over a six-week period. The study was conducted with TD children between 5 and 8 years, and results showed that the training intervention significantly improved WM tasks and comprehension reading for the intervention group, but there were no differences in single word reading and arithmetic abilities between students in the control and intervention group. Skills in mathematics and reading were assessed alongside six WM skills, both pre- and post-test and at a six-month follow-up. Further measures to assess mathematics, word reading, reading comprehension and spelling were also administered at a 12-month follow-up. At post-test, the group that had received the intervention demonstrated significantly greater performance on the two trained executive loaded WM tasks (Listening Recall and Odd One Out Span) as well as on two untrained WM tasks (Word Recall and Counting Recall) than the control group. These ‘near transfer’ effects remained at a six-month follow-up, although at the 12-month follow-up, higher reading comprehension scores were the only significant ‘far transfer’ effect for the trained group (there were no WM differences at 12-month follow-up). The authors do suggest that this benefit may be derived from greater WM performance, with a greater skill in processing and storage, contributing to reading comprehension (and potentially academic achievements) in the long-term. It is possible that this study achieved different (more positive) findings than Banales et al.’s (2015) study due to the different (younger) mean age of the children, meaning that any WM benefits (and their transfer to real-world and academic outcomes) were gained at a younger age. This could mean that they laid the foundation early on to practice and build on more advanced skills, such as reading.
WM benefits as a result of direct WM interventions is supported further by Passolunghi and Costa (2016). These researchers found that a direct adaptive WM training intervention (that is, adaptive where task difficulty is adjusted to individual performance; von Bastian & Eschen, 2016) improved the WM and early numeracy abilities of 48 five-year-old TD children, whilst an early numeracy training programme improved their numeracy abilities only. The WM training involved a variety of paper-and-pencil exercises, all designed to engage and improve the three components of Baddeley’s WM model (Baddeley, 1986). There was no indication in the study that follow-up assessments were conducted, however; it is possible that, as Henry, Messer and Nash (2014) found, WM intervention measures might be more effective as long as the training continues, as WM-specific benefits seemed to diminish (except reading comprehension) when the intervention stopped. This study, involving pre-school children, extended research from other studies that has also found WM training to benefit the early numeracy skills of older children and low-performing children (e.g., Kroesbergen et al., 2014; Kuhn & Holling, 2014; Witt, 2011), despite mixed findings on this front (Melby-Lervåg & Hulme, 2013). It is entirely possible that WM training outcomes could vary based on the age of the children involved, and most studies investigating the impact of WM training focuses on school-aged children. Passolunghi and Costa (2016) assert that WM training may be more effective in younger, preschool children when, as Wass et al. (2012) found, the neural system is more malleable to experience. However, this may be more difficult to implement, if parents of participants are less likely to be recruited. Conducting WM training in a school setting can form part of lessons, meaning that parents do not necessarily need to be involved (if they do not wish).

Play could be another effective form of intervention for children to improve WM. Thibodeau et al. (2016) conducted a study of TD children aged between 3 and 5 years, where the teacher used pretend play as a method of instruction in the intervention group, while the control group used non-imaginative play. The study
found that pretend play significantly improved the WM of the TD children that participated in the intervention. Therefore, it is possible that, with further research to support the findings of Thibodeau et al. (2016), teachers in an institution could incorporate pretend play as an intervention measure to improve the WM of TD children, especially pre-schoolers, given the idea that WM training may be more effective for this age group (Wass et al., 2012). However, the long-term benefits of this intervention is called into question, given that no follow-up assessment was conducted in this study.

Thus, research into WM interventions with TD children within an educational context appears to suggest that specific, adaptive interventions are generally effective, especially in the short-term; moreover, the ‘near’ and ‘far transfer’ effects for specific academic skills, such as numeracy and reading comprehension are demonstrated in some studies. Yet with a lack of follow-up assessments, it is difficult to conclusively suggest that WM interventions have a long-term effect on both WM and subsequent academic achievement. These issues of longer-term effect may be even more of a concern when working with children with developmental disabilities. The next section therefore explores literature surrounding WM interventions for children with developmental disabilities other than DS.

4.1.3.2 Working Memory Interventions for Children with Developmental Disabilities

Interventions that have focused on improving WM and, in turn, academic skills, have also been tested on children with developmental disabilities and developmental disabilities (e.g., de Vries et al., 2015; Dunning, Holmes & Gathercole, 2013; Loomes et al., 2008; Mezzacappa & Buckner, 2010). A number of WM interventions for children with developmental disabilities take teaching approaches similar to those described in 4.1.3.1. (e.g., Atia, 2010; Bennett et al., 2013; Moalli, 2006; Smith & Jarrold, 2014; Söderqvist et al.,
For example, Orsolini et al. (2018) investigated the impact of a direct training intervention in five participants between the ages of 9-12 years with MID, which included card games and exercises targeting verbal WM. The experimental training, which took place in two weekly sessions over eight weeks, significantly improved problem-solving and verbal WM post-test (which occurred eight weeks after the final session), although there was no indication of subsequent follow-ups to determine longer-term effects. Yet this does demonstrate that direct training by teachers can be an important intervention to enhance WM performance of children with MID.

One area of particular interest in the literature, however, is the use of computer-based WM training programmes, that have been developed commercially by several companies. The effectiveness of these programmes in improving memory, academic ability, fluid intelligence, response inhibition and recall in adolescents (13-16 years) with mild intellectual disability (MID) that were enrolled in special education was evaluated by Van der Molen et al. (2010). Two computer-based training programmes were found to lead to significantly higher scores in WM performance, demonstrating that WM can be enhanced in adolescents of this age range at least with MID (this may somewhat coincide with the developmental spur in WM at age 14 in TD adolescents (Best & Miller, 2010; Sippl, 2021). The best-known programme is CogMed, which is commonly used both in clinical settings as well as schools. Klingberg et al. (2002) found that CogMed improved performance of trained WM tasks in participants with ADHD; the effect of the training was furthermore extended to nontrained tasks that required WM. Moreover, performance on tasks that involved prefrontal regions was also significantly improved by training, as was the motor activity in children with ADHD. These findings indicate that training specific to WM functioning could potentially be used in clinical settings to reduce ADHD symptoms. It could therefore be argued that computerised training might transfer positive effects to other non-trained areas.
Dunning, Holmes and Gathercole (2013) instigated the first randomised controlled trial (RCT) of CogMed with 94 children (aged 7-9 years) with low WM in schools in the North-East of England. Children were randomly allocated to an adaptive training group, a non-adaptive training group or a no intervention group. The intervention groups both received six weeks of training using CogMed. Post-training assessments were then completed. Some students were then tested 12 months after the intervention, to establish any long-term gains. The study found that adaptive training was significantly correlated with particular improvements in numerous untrained tests of WM, with a substantial effect size, compared to both the non-adaptive intervention group and the non-intervention group. However, there were no far transfer effects, given that the participants in the intervention groups did not show improvements in related classroom tasks requiring WM or in other cognitive assessments. Yet the follow-up did show that improvements in verbal WM were still present one-year post-training for some students. Given that there were no significant differences in WM between groups at baseline, this RCT usefully reveals that students can benefit, even in the long-term, from computerised WM interventions, although these benefits may not be transferable to other tasks requiring WM. Perhaps an earlier intervention (at a younger age) may have worked to benefit WM in other aspects of life requiring WM, rendering the gains more transferable in later life.

However, other research contradicts these findings. A review of computerised WM training indicated that these programmes rarely facilitate sustainable or transferable benefits to WM, classroom functioning or academic performance (Colmar & Double, 2017). Some studies (e.g., Holmes, Gathercole & Dunning, 2009) that report significant gains in WM and mathematics tests in children with low WM at a 6-month follow-up, did not also re-test the control group in these abilities. Thus, it becomes difficult to separate WM gains from an intervention from any improvements that would be gained during natural development.
Moreover, Roberts et al. (2016) found in their large-scale Australian study, assessing the impact of CogMed after two years, that there was no near transfer effect or generalisability to academic outcomes from the programme, and given the expense and time-consuming nature of CogMed, its use was not deemed to be justified. Most studies focus on ADHD populations (Colmar & Double, 2017; Rapport et al., 2013); thus, there also needs to be further investigation of how computerised interventions fit with children that have more than one disability, such as fine motor skills disabilities, which restrict their use of keyboards, which may be the case in some with DS. It is also imperative to understand the availability and view of such computerised programmes in contexts such as Kuwait, where programmes such as CogMed may not be available in Arabic, for example.

4.1.3.3 Working Memory Interventions for Children with Down Syndrome

Fidler et al. (2018) indicate that an understanding of the early origins of ID and EF difficulties in DS is pivotal for the design and implementation of effective interventions that capitalise on early neuroplasticity. These researchers state, ‘Waiting until the school years, or even the preschool years, to understand and address cognitive challenges in DS may sacrifice an important window of opportunity’ (Fidler et al., 2018, p. 207). This is because, even at age 12 months, children with DS have been detected as demonstrating poorer performance on cognitive assessments comparative to TD norms. They even demonstrate less cognitive gains between 12-30 months compared to children with the same MA with other developmental delays (Visootsak et al., 2013). Given research indicating that difficulties with memory are central to the cognitive phenotype of DS throughout infancy, childhood and into adulthood (Oxelgren et al., 2017), this indicates that interventions for improving the WM of individuals with DS may be best placed in infancy – or indeed, as early as is practicable. However, whilst infancy may provide the optimal focus for WM interventions, WM training throughout the lifespan may be beneficial. WM
interventions for TD children in a school context have been found to enable a rich opportunity for transfer effects to everyday life and academic skills (e.g., Kroesbergen et al., 2014; Kuhn & Holling, 2014; Witt, 2011), especially as it may be delivered in a more structured manner by teachers than by parents at home (Redick at al., 2015). It is likely that the same applies for children with DS as TD children.

As discussed in chapter three, WM appears to be the most challenged EF in children with DS, with those with DS demonstrating greater difficulty in this EF compared to other EFs. For this reason, WM interventions are critical to enabling children with DS to improve their WM abilities to complete daily tasks. Such interventions include computerised games, specific WM strategies taught by teachers and physical exercise activities. Several studies have also investigated the role of WM in academic achievement for children with DS; for example, Laws, Brown and Main (2016) highlight the importance of WM in reading comprehension in children with DS, given its association with phonological awareness, listening comprehension and word reading. WM interventions (such as rehearsal) to improve word reading, phonological awareness and listening, may ultimately help in improving not only WM but also reading comprehension amongst children with DS.

Physical activity can also be used to improve the WM of children with DS. As mentioned previously, Ringenbach et al. (2016) conducted a study to investigate the impact of assisted cycling therapy compared with voluntary cycling on the WM of adolescents with DS. 17 participants completed the assisted cycling therapy and 16 participants completed the voluntary cycling therapy. Findings from the study showed that assisted-cadence cycling resulted in improvement to WM, but the voluntary cycling or no cycling conditions did not. Moreover, Ringenbach et al. (2015) conducted an intervention study similar to that by Holzapfel et al. (2016) and found that both assisted cycling therapy and
voluntary cycling improved WM in adolescents with DS, with a moderate effect size. Individuals in the assisted cycling therapy group improved the most, while those that did not do any exercise did not improve their cognitive abilities.

Similarly, Alesi et al. (2014) conducted a study to investigate the impact of exercise training on three children (one girl aged 14 years and two boys aged 10 and 14 years respectively) with DS. The study found that in these individuals, exercise improved performance on tasks using WM. Despite the small, non-generalisable sample in Alesi et al.‘s (2014) study, the findings of the combined studies exploring exercise and WM show that physical activities have the potential to enhance the WM performance for adolescents with DS. Yet these studies were focused predominantly on older children and adolescents, rendering it unclear what the effect of assisted cycling therapy may be on younger children. However, this was perhaps necessary given to the nature of the intervention, which required gross motor skills and sufficient physical health to participate. Infants with DS do not always possess refined skills in these areas, rendering some exercise interventions impractical for younger age groups, despite the fact that many researchers are calling for EF interventions to be conducted earlier than adolescence (e.g., Fidler et al., 2018).

Several studies have examined the efficacy of computer-based WM interventions for children with DS. For example, similar to the studies of Van der Molen et al. (2010) and Klingberg et al. (2002), Pulina et al. (2015) investigated the effects of computer-based WM training specifically for children with DS. The sample of 39 children and adolescents (age range 7-19 years) was divided into two groups; the training was delivered by an expert in WM in one group, and by parents that had been appropriately instructed in the other. In both groups, spatial-simultaneous components of WM performance improved and the results were sustained one month later. The findings suggest that WM performance can be improved in children and adolescents with DS; interestingly, the involvement
of parents could suggest that the intervention strategies can also be used within the home context, by people that individuals with DS know and trust, for further gains.

Although computer-based interventions may be effective, it presents difficulties in a population with cognitive disabilities. For example, computerised training needs skill in keyboard use, and DS is characterised by weaknesses in muscle balance (Smith, 2001), presenting difficulties in selecting options onscreen. It requires a hugely expensive bespoke computer lab to accommodate children with disabilities, potentially with trained, specialist experts to deliver this intervention or coaching teachers or parents in how to deliver it. There are also costs involved in accessing programmes such as CogMed (Gathercole, Dunning & Holmes, 2012), which may not be possible for schools. This could represent yet another task additional to teachers’ current workloads. Despite the fact that in Kuwait every school has a computer lab, children’s muscle weakness, the lack of availability of CogMed in Arabic, together with the lack of trained staff, could present problems. In addition, it is unclear whether younger children (especially preschoolers) are able to navigate and undertake CogMed or computerised training more generally, when potentially the greatest gains in WM may occur, as they arguably do in other populations (Wass et al., 2012).

Another form of intervention is classroom games-based training (paper-and-pencil tasks created to enhance WM skills) by teachers to improve the WM of children and adolescents with DS. Costa, Purser and Passolunghi (2015) conducted a study to investigate the possibility of training STM and WM abilities in adolescents by implementing a school-based games training that targeted the visuo-spatial WM of students with DS for six weeks. The results from the group of 17-year-old and the group of 15-year-old participants with DS showed that training the WM with given tasks improved visuo-spatial performance post-test; however, the study did not conduct a follow-up assessment to ascertain the
sustainability of the intervention’s effectiveness. Thus, the reliability of the findings from the studies is questionable as the long-term stability of the training effects was not tested and the sample sizes used were relatively small and involved adolescents in their mid-late teens only.

Teaching children with DS a specific WM strategy may also be effective in improving WM performance. Laws et al. (1996) describe using a rehearsal strategy for 27 children and adolescents with DS aged 5–19, all attending schools for individuals with severe learning difficulties. The findings suggested that there was a significant (albeit weak) improvement in memory span when the children engaged in these strategies as delivered in the classroom by teachers or teaching assistants. Memory and verbal recall of longer words particularly improved and there was also a significant correlation between reading and memory scores. This suggests that there was an underlying difference in the levels of rehearsal used pre- and post-intervention. There was no control group in this study with whom to compare the findings, however. Moreover, the one-way analysis of variance of the mean gains in word span confirmed there was a significant effect for school attended, which could suggest either inherent variations in the abilities in the children at each school, or that the teachers may have delivered the intervention differently in each school. Standardisation of the WM intervention would need to be monitored to ascertain the effect of this on outcomes.

In another example of a rehearsal strategy intervention, a study by Comblain (1994) exposed one group of four teenagers and four young adults with DS to intensive rehearsal training for eight weeks, while another group received no training. The trained participants improved their memory span significantly and the others not at all. At that time, the trained individuals showed clear signs of systematic rehearsal, yet at six weeks and six months post-intervention, they no longer appeared to rehearse systematically and their memory performance had
fallen significantly, although still remained significantly higher than at the start of the study. Again, this suggests the importance of creating interventions in such a way that any benefits are likely to be sustained, and involving samples that may be more developmentally susceptible to benefitting from WM interventions (e.g., a younger age range).

Of relevance to any rehearsal strategy to improve WM is the child with DS’s language ability, as the condition is generally accompanied by deficits in memory span, particularly auditory verbal memory span (Conners et al., 2008). The ability to learn language therefore may require adequate skills in memory span, which suggests that if those with DS were able to develop greater skill in memory span performance, any linguistic interventions they undergo may be made more effective. A study that set out to investigate whether auditory memory span may be enhanced in those with DS was conducted by Conners et al. (2008). Participants were 16 children with DS, aged 6 to 14, divided into two groups that alternated in receiving training in overt cumulative rehearsal and auditory strategies, which targeted verbal WM (not visual). The training was implemented by parents in the home environment. Whilst the participants did not demonstrate any significant improvement on verbal WM or sentence memory, all children improved a little in digit span, which was the main proximal outcome measure, which indicates greater use of phonological codes in memory. This indicates that some individuals with DS may be able to enhance their auditory verbal memory span as a result of rehearsal training received within the home, to some extent. The study conducted pre- and post-testing, but there was no follow-up test, so it is unclear whether any gains were sustained over time. It also did not highlight whether greater gains were made by participants that were younger or older in terms of CA; such an insight may go some way to highlighting which age range to focus on in future WM training research.
Children with DS that possess relatively good verbal WM and language skills may benefit most from rehearsal-type interventions, despite the fact that their improvements may only be minimal. However, even small improvements when function is severely challenged are to be celebrated, justifying further investigation (Conners et al., 2008). On the other hand, the use of this intervention might be limited, as this strategy requires good language skills, while children with DS often demonstrated language difficulties as mentioned in chapter two. However, given that these earlier studies show some promising findings, rehearsal may be a useful strategy to incorporate in a WM intervention.

4.1.4 Working Memory Interventions in an Arab Context

Research into WM provision and intervention for children with DS specifically in Arab countries is almost non-existent. Whilst there is some WM research conducted in TD populations in other Arab countries, the findings may not be applicable to the Kuwaiti context. For example, Hussein and Reid (2009) examined whether redesigning the presentation of the chemistry curriculum to TD senior school pupils in the United Arab Emirates so as to lower the WM demand would improve outcomes. This study revealed that altering teaching materials improved performance by 13%, and required no teacher training, extra time or alteration of curriculum content. Yet this process may not be applicable to DS populations of a younger age range in Kuwait, especially given that it was focused on teaching chemistry, a subject that is not explicitly taught in Kuwait to students with DS.

The idea that culturally-specific, tailored interventions for a Kuwaiti context is supported by the finding that Kuwaitis may differ from non-Kuwaitis in terms of performance on some WM tests. Al-Dyiar and Salem (2013) investigated phonological processes and WM performance in 500 Kuwaiti and non-Kuwaiti Arabic dyslexics. This study found that the performance of non-Kuwaiti participants was significantly better on the Non-Word Repetition Test, Sentence
Repetition Test and Forward Digit Recall Test, measuring WM. Despite this study being focused on dyslexics (not those with DS), this suggests that results from Arab countries regarding WM performance and interventions (the few that there may be) may not necessarily be transferable to specific countries within the Gulf (e.g., Kuwait). However, this was the finding of only one study, and the results do not appear to have yet been replicated. Furthermore, the study was not DS-specific; yet it does suggest that it is important to remain cautious when interpreting the findings of other studies, given that they may not be transferable to the Kuwaiti context.

A qualitative study by Alfaraj and Kuyini (2014) into the use of technology to support the learning of children with DS in Saudi Arabia, for instance, found that the 20 teachers involved highlighted key challenges, such as the lack of software designed in Arabic, limited resources (such as computer software) and insufficient teacher training to assist them in supporting the children using the software. This indicates that computerised interventions discussed in the literature are not necessarily available or appropriately designed for an Arab context such as Kuwait. Those that do exist in a limited capacity (generally as trials for the DS population in Arab countries) have shown promising results, however, such as the use of a translated ‘See and Learn’ reading intervention programme for children aged 3-6 with DS in Lebanon in Abu Khadra’s (2014) study.

Overall, there are mixed findings on the best intervention measures to improve WM in this population. The WM intervention measures show conflicting results, indicating a need for further rigorous research. There is evidence that computer training programmes can be useful in improving spatial simultaneous WM (Pulina et al., 2015). Similarly, some physical activity interventions have been shown to have a positive impact on WM abilities (Holzapfel et al., 2015; 2016; Ringenbach et al., 2016). Thus, important elements that might be useful in a
WM intervention appear to be the use of computer training programmes, physical activity, a rehearsal strategy, class-based activities and games, and perhaps the use of play (e.g., Von Bastian & Eschen, 2016). Involving parent and teacher participation in the research design can also provide benefits. Conners (2008) made a positive contribution by involving parents in the intervention, an approach that can be expected to widen parent knowledge about their child’s WM disability, for example.

However, there are a number of issues with some of the WM interventions for children with DS to date, often surrounding the lack of long-term follow-up assessments to ascertain the sustainability of the intervention, the lack of causality (it could be that there are other factors at play in the correlation between physical activity and improved WM, such as better sleep as a result of exercise, for instance, which was not considered by the author), a lack of control groups for comparison and the lack of generalisability, often as a result of small sample sizes, and variable age ranges. Moreover, specialist equipment and personnel are often required in devising and administering either exercise- or computer-based interventions, which is not always practicable or affordable for some researchers (including myself). Without these resources securely in place, there may be increased safety risks for the children involved in the intervention.

Taking into consideration the factors discussed above, this study opted for an intervention delivered to teachers (as opposed to children) model, training teachers in techniques and strategies designed to promote WM in children with DS. Teachers were chosen as the main focus of the intervention partly because previous interventions that involved teachers showed high levels of effectiveness in improving WM (Banales et al., 2015; Conners et al., 2008; Costa, Purser & Passolunghi, 2015; Henry, Messer & Nash, 2014; Laws et al., 1996; Orsolini et al., 2015; Thibodeau et al., 2016). Another consideration is that, by training teachers, there is greater potential for any positive effects of the
intervention to be sustained, as teachers will be encouraged to continue using their new skills with the children during the school year after the intervention has officially ended, and then potentially with different children each year.

The present study also included an active control group to strengthen my research design by providing a comparison group, enabling the effects of the WM intervention to be more easily discerned, whilst still providing a benefit to the control group (Oberste et al., 2017). In this control group, the intervention focused on strategies to improve challenging behaviour in children with DS, given that this was reportedly an ongoing issue for the teachers in my study. Thus, as the present study included an active control group (a Positive Behaviour Support intervention to reduce challenging behaviour (CB)) for comparison, the literature surrounding interventions to reduce CB is now briefly reviewed.

4.1.5 Interventions to Support the Reduction of Challenging Behaviour

CB amongst TD children and those with disabilities may include calling out in class, tantrums, refusing to follow instructions, screaming and swearing (Skiba et al., 2016; Myers et al., 2017; Veletsianos, Reich & Pasquini, 2016). There are various factors that may contribute to CB, including anxiety, depression, apathy, aggression (Dekker et al., 2018), stubbornness, mood disturbance, impulsivity and over-reactivity (Sheth et al., 2015). The CB exhibited by many children, especially those with cognitive difficulties (such as those with DS), could also occur as a result of developmental difficulties, poor non-verbal and verbal skills, delayed comprehension, attention deficit, speech and hearing difficulties, or sleep disturbance (Wiggs & Stores, 1996). Other causes could include the classroom environment, difficult/challenging tasks or teaching that does not match the child’s needs (Grieco et al., 2015).
It is also possible that EF difficulties and atypicalities may be an underlying cause for (or could exacerbate) CB (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998; Wilding et al., 2002) and resulting in challenges with effective decision-making (Cuskelly, Einam & Jobling, 2001) and cognitive flexibility (Zelazo et al., 1996). Teachers experience various challenging scenarios when they must manage the behaviours of children with disabilities, including children with DS, whilst ensuring suitable learning so that students can attain the highest level of performance. Such behavioural challenges require teacher intervention to help create an environment that supports learning. Teachers need to be prepared to use evidenced-based intervention techniques when encouraging and enhancing prosocial behaviours. Therefore, it could be argued that teachers play an essential role in interventions to reduce CB in children. The following section thus aims to briefly identify forms of intervention that can best support teachers to mitigate CB in their students.

4.1.5.1. Challenging Behaviour Interventions in Typically Developing Children

Strategies for tackling CB vary; some draw on principles requiring behavioural modification, such as positive and negative reinforcement, to facilitate appropriate behaviour in students (e.g., see Petscher, Rey and Bailey, 2009, for a review). Other interventions are more relationship-focused, looking at teacher relationships (e.g., Spilt et al., 2012) and parental attachment (e.g., Moretti & Obsuth, 2009). Positive behavioural support (PBS) is a person-centred approach that has become common in reducing CB, as it provides long-term, individualised, tailored strategies to tackle the causes of CB and improve the individuals’ quality of life (MacDonald & McGill, 2013). Despite its critics, many of whom claim PBS is open to corruption (e.g., Baker & Allen, 2012), various studies have shown it is effective in reducing CB.

For example, in a review conducted by Stormont et al. (2015), which focused on teachers of both TD children and children with special needs, it was found that
PBS interventions are fundamental to increasing learning amongst children, including preschool age children. Caldarella et al. (2015) investigated the effect of PBS strategies in TD children from kindergarten to second grade, and it emerged that increased praise and reduction of reprimand amongst teachers helped in reducing classroom disruption and increasing appropriate behaviour in a class setting. This reveals that positive reinforcement is perhaps the most effective strategy, although a limitation of this is that it does not uncover the root cause of the CB in the first place.

4.1.5.2 Challenging Behaviour Interventions for Children with Cognitive Difficulties

Functional behaviour assessment (FBA) or analysis is one popular method to help reduce CB, especially in children with cognitive difficulties. FBA is based on the idea that all behaviour serves a function or purpose (Barnhill, 2005); thus, it aims to assess the antecedents and consequences of behaviour (Gresham, 2003). FBA generally involves both indirect assessment methods, such as rating scales and interviews, as well as direct observations of behaviour (O’Neill et al., 1997). This can allow a picture of types and triggers of CB in particular individuals to emerge, which may then even be tested for confirmation of causal relationships (Doggett et al., 2001). Flynn and Lo’s (2016) study attested to the effectiveness of FBA with children with cognitive difficulties, although some students exhibited unacceptably high rates of replacement behaviour, with the intention of gaining reinforcement from teachers; as a result, teacher instruction was disrupted. Yet it is possible that the teachers’ approach, rather than children’s lack of engagement with the intervention, may give rise to CB; eliminating the teachers’ use of rule statements was effective in decreasing the students’ problem behaviours, for example (Albin et al., 1995).
Ultimately, FBA, the incorporation of social and emotional learning and use of reinforcement intervention by teachers are all PBS strategies that appear to have a positive impact on the management of CB amongst students.

4.1.5.3 Interventions to Reduce Challenging Behaviour in Children with Down Syndrome
A number of interventions have been developed for teachers to utilise and help manage the CB of students with DS. Feeley and Jones (2008) researched the factors most associated with CB in DS and identified four strategies that teachers can use to reduce levels of CB in children with DS: 1) providing caregivers with information regarding identification of, and interventions in response to, potential CB; 2) creating strategies to reduce escape behaviours, for example by reducing task difficulties; 3) teaching children how to use requesting behaviours, one of the PBS techniques; and 4) reducing levels of self-stimulatory behaviours in children, such as rocking, tongue clicking, teeth grinding or placing hands in mouth, as such behaviours may disrupt classroom learning and participation, although such behaviours must be assessed in light of the individual child’s needs.

Functional assessment and intervention are important for maintaining pro-social behaviours amongst children with DS that facilitate learning. Wadsworth, Hansen and Wills (2015) in their study identified escape, and access to tangible items, as primary and secondary reasons for CB in children with DS. Intervention measures implemented were self-monitoring and teacher monitoring. The results from the study found that monitoring increased the following of teachers’ instruction amongst students with DS. Neil and Jones (2016) found that differential reinforcement helped in reducing repetitive behaviour in children with DS, although the intervention worked differently for each student, where some will respond positively, some negatively, and no
changes may be experienced in other students. This suggests teachers may need to tailor any intervention to the child.

In sum, teachers’ participation could be a factor in successful interventions, although in some cases teachers’ participation could be one of factors leading to CB. Positive Behaviour Support, FBA and differential reinforcement have been shown to be effective in reducing CB, perhaps because the former two predominantly focus on causes of CB. However, whilst different behavioural interventions can be implemented by teachers to reduce CB among children with DS, most of these interventions provide for the management of one specific behavioural challenge, while children with DS usually demonstrate several behavioural challenges. It becomes clear therefore that teachers must be able to adapt any intervention to the particular pupil with whom they are working.

4.1.6 Teachers’ Involvement in Interventions

Interventions that support development and learning can be delivered by the research team directly to the child, or can be delivered to those in close contact with the child, such as teachers, parents or pupil support assistants, who then implement the intervention with the child. One key aspect of the intervention in the current study was that it was delivered to teachers, who then implemented it with their particular class of children with DS. This section reviews relevant previous research and considers the advantages and disadvantages of such an approach.

Teachers’ involvement in interventions (or ‘teacher-focused interventions’) are programmes and strategies that teachers can employ in a learning environment to promote student achievement. This kind of intervention is critical to the support of children with a wide range of disabilities and behavioural difficulties. According to Lee et al. (2016), teacher-focused interventions can be used with TD children to enhance their academic achievement and reduce the
performance gap, as well as to support students with disabilities, including those with DS, to improve their academic achievement and abilities. According to Lin-Siegler, Dweck and Cohen (2016), teacher-focused interventions may lead to demonstrable gains on assessment measures, promote engagement within the classroom and encourage greater school satisfaction amongst students. Consequently, it is possible that there may be positive outcomes for students as a result of teachers being involved in interventions.

There are also other benefits of teacher involvement in interventions. It has been found to be more sustainable than one-off, expert-led interventions and improves the development of special needs children (Daniel & Lemons, 2018), perhaps because students gain greater exposure to the intervention and it is delivered by someone known to them (as opposed to a stranger). It may also be more cost-efficient and widespread, as teachers may use the same intervention in subsequent school years with other pupils. Darling-Hammond, Hyler and Gardner (2017) also found that teacher-focused interventions are important as they promote student-teacher interactions, helping teachers to understand their students better and thus ensure responses that are tailored to students’ particular needs. This is especially important in children with a variety of CB and EF profiles, such as children with DS (Neil & Jones, 2016).

For example, a teacher-focused intervention to improve WM and academic performance in 256 primary school children with WM difficulties was tested by Elliott et al. (2010). This study included a teacher-focused intervention programme, where teachers received training regarding how to ensure the classroom environment was tailored to meeting the needs of those with WM difficulties. A second group received a behavioural teaching intervention approach, and were provided with brief, regular and specific instruction in basic WM skills; and a third group formed the control group. However, at the close of the academic year, the children in both intervention groups showed no
significant improvement in terms of academic performance or WM skills, compared to the controls. This might suggest that these kinds of interventions do not initiate near- or far-transfer effects. However, the study did find via observational data that teachers’ effective, regular use of WM strategies in their teaching, in any of the three research groups, predicted the children’s attainment. This indicates that teacher involvement in intervention groups could boost their knowledge and use of WM strategies, which could, in turn, increase the academic attainment of students, although the intervention type itself may not have a significant effect.

However, there are drawbacks and challenges of teacher-focused interventions, most of which arise from implementation. Daniel and Lemons (2018) for example report that some teacher-focused interventions used in schools are not evidenced-based, although this could be due to insufficient planning in the implementation of the intervention. According to Swain-BRADway, Pinkney and Flannery (2015), the implementation of a teacher-focused intervention is most feasible in smaller groups and challenging to implement with larger groups. Moreover, implementation is resource-intensive and expensive for school administration and can increase teachers’ workload. Swain-BRADway, Pinkney and Flannery (2015) also found that resistance to new interventions amongst teachers also lead to implementation problems. Ultimately, the benefits of teacher-focused intervention are high, but implementation strategies should be designed carefully, to try to reduce the potential challenges and limitations that can reduce the effectiveness of teacher-focused interventions.

4.1.7 Summary and Present Study
This review has shown that various EF, WM and CB interventions have be researched in groups of TD children as well as those with developmental disabilities and DS. It has also investigated teachers’ involvement during interventions, and the advantages and drawbacks of this on achieving positive
intervention outcomes on the development of children. There appear to be a number of gaps in the literature reviewed. Firstly, there is insufficient research on WM interventions where teachers are able to take ownership of the intervention and decide how to implement it with their own pupils. Teachers must feel fully engaged and autonomous in any intervention process for it to be effective, as well as being supported to develop or possessing the skills to adapt any intervention to meet the needs of a particular child. Yet there is a paucity of research in this area, especially pertaining to teacher-focused interventions for children with DS; training teachers in strategies is useful as they can then use any beneficial strategies with other students with DS that they teach, either now or in future.

Moreover, there is a gap in the literature concerning interventions that involve both parents and teachers in the evaluation to ascertain whether any WM gains are seen across both home and school contexts. Furthermore, there is a discernible lack of literature concerning interventions for children with DS in Arabic countries. Studies in an Arab context are sorely lacking, and to date there appears to be no study of WM interventions for children with DS in Kuwait. Interventions used in other contexts may not be applicable for the Kuwaiti context given the language barrier (very few have been translated into Arabic) and cultural differences that may render some English, Western interventions less appropriate (they may contain material that has little relevance to children growing up in Kuwait, for instance).

Moreover, there appears to be very little emphasis on WM interventions for those with DS in the educational context in Kuwait; some countries have a collaborative body that gathers evidence-based research to inform government and organisational policy and practice in education and public health, such as the National Collaborating Centre for Methods and Tools (NCCMT) in Canada or the British Educational Research Association (BERA) in the UK. However, there
is no such body in Kuwait, which often means that decision-makers do not prioritise the implementation of educational interventions as a result of a lack of evidence-based information. There is no such association between the administrators of interventions in educational settings and organisations that specialise in the development of training programmes or tools in Kuwait. Therefore, this is a gap that needs addressing if the WM of children with DS, and the best ways to support it, in this particular context, is to be more fully understood and improved.

Previous research has highlighted that the important elements of any WM intervention design include the following features: supporting rehearsal strategies (Comblain, 1994; Laws, Brown & Main, 2016), physical activity (Alesi et al., 2014; Holzapfel et al., 2016; Ringenbach et al., 2015; 2016), class-based activities and games (e.g., Costa et al., 2015; Orsolini et al., 2018), and (potentially) computer-based training programmes (e.g., Pulina et al., 2015), where practical and financially viable (physical activity and computer-based training was not feasible in the current setting of the present study, however). Other important elements of an intervention to improve the outcomes of children in an educational context more generally include teacher-focused interventions (Lee et al., 2016), the involvement of parents/carers (Conners et al., 2008) and delivery over a number of weeks (Comblain, 1994; Costa et al., 2015).

In terms of the important elements in the evaluation of any WM intervention, research indicates that the inclusion of an active control group is useful (Oberste et al., 2017), as is a follow-up assessment to measure the long-term effect/sustainability of the intervention (Holmes et al., 2009). A large enough sample is also required to ascertain any significant effects. Moreover, gaining both parent and teacher reports on improvements in WM is also beneficial to ascertain the effectiveness of the intervention across different contexts (Pulina et al., 2015).
It becomes clear, given the gaps in the current literature surrounding WM improvement of children with DS in a Kuwait educational setting, that a new, teacher-focused intervention is required. The present study therefore aimed to devised, implement and evaluate a Kuwait-based teacher-focused intervention strategy to improve WM in children with DS as its main focus, to ascertain its effectiveness in improving WM performance and academic abilities compared to an active control group.

The research questions of the current study were:

RQ1: Does the WM intervention programme aimed at teachers lead to changes in:
  a) working memory outcomes;
  b) executive function scores;
  c) behavioural outcomes
  d) literacy and numeracy scores on school assessments;
    for pupils with Down syndrome in inclusive education in Kuwait?

RQ2: Does teachers’ use of WM strategies change from pre- to post-intervention in inclusive education in Kuwait?

RQ3: Does teachers’ knowledge of WM and CB change from pre- to post-intervention in inclusive education in Kuwait?

RQ4: What are teachers’ viewpoints about the WM intervention in inclusive education in Kuwait?

The next section explains the methodology and methods adopted to answer these research questions in detail.
4.2 Methods

4.2.1 Introduction to Methods
This section explains and justifies the research design, participant criteria, tools (measures, observation and focus group), procedures, ethical considerations and methods of data analysis used for designing and assessing my intervention.

4.2.2 Research Design
The study was a quasi-experimental intervention study; as with many quasi-experimental studies, the study included an intervention and a control group, although the participants were not randomly allocated to groups. This kind of design is useful as it enables students to remain in their natural environment during educational research, providing comfort and familiarity (Whitely & Kite, 2013, cited in Cebula, 2018) and helping to provide a deeper insight into ‘what works’ within the school setting (Cebula, 2018). However, it must be noted that there are critics of quasi-experiments within education (e.g., Howe, 2004, cited in Cebula, 2018); this includes the argument that some findings cannot be replicated outside of a tightly-controlled laboratory setting, calling the reliability of experiments within the classroom into question (Howe, 2004, cited in Cebula, 2018). However, as Cook (2003) asserts, experiments can still generate usable knowledge within certain conditions, such as schools.

There are also concerns regarding internal validity in quasi-experiments, given the impossibility of controlling extraneous variables (especially inherent differences in participants as they have not been randomly allocated to conditions) that may have a confounding impact on the dependent variable (Campbell & Stanley, 1963; Shadish et al., 2002, both cited in Cebula, 2018). Moreover, as Pawson and Tilley (1997, cited in Cebula, 2018) indicate, not only
the mechanisms but the *context* of change must be taken into account; whilst the mechanisms of how the dependent variable may have been changed by the independent variable are often unreported in studies (Howe, 2004, cited in Cebula, 2018), quasi-experiments conducted in schools can at least enable the context of any causal mechanisms to be known (Goldacre, 2013, cited in Cebula, 2018). This can lead to a deeper understanding of the effect of a particular intervention within specific classroom settings (Cebula, 2018).

Quasi-experimental research designs are common in educational research when randomisation is not possible, and has been used in a variety of school- or home-based studies, often involving children with ID (e.g., Bruning et al., 2008; Bianco, Lecce & Banerjee, 2016; Remington et al., 2007 all cited in Cebula, 2018). The quasi-experimental research design in the present study was chosen over an experimental design, given that randomisation was not possible in this research situation (Campbell & Stanley, 1963, cited in Cebula, 2018). Although randomisation provides stronger evidence for causal relationships (between the independent variable and the dependent variable), given that it minimises the risk of bias or confounding factors influencing the results (Cebula, 2018), in the present study, randomisation would have entailed mixing children and teachers from different schools. This was not logistically possible given where the schools were located. It would also not have been culturally appropriate, as girls and boys are taught separately in Kuwait, and randomisation would have required mixing girls and boys. In addition, randomisation might have led to ‘intervention contamination’ (Sanson-Fisher et al., 2007) between the two groups, as single schools would then have contained both EG and CG teachers. By adopting a quasi-experimental design, therefore, external validity was potentially increased, given the intervention was conducted in the students’ natural environment with their current classmates (Walser, 2014).
Using non-identical, non-randomised groups can threaten internal validity (Cohen, Manion & Morrison, 2018), given the possibility of other variables (other than the intervention) affecting the results. However, I sought to limit the possibility of this within my research, by ensuring that variables such as the number of participants, educational resources and other school facilities, and the diagnosis of Down syndrome (DS), were similar in both the control group (CG) and experimental group (EG), so the groups were well matched, if not randomly allocated.

Using a quasi-experimental research design within a school setting enhances ecological validity, compared to controlled laboratory settings (Thyer, 2012). My quasi-experimental study was conducted with children in their regular schools and with their regular teachers, so it can reasonably be assumed that there was minimal threat to ecological validity. Findings may therefore be more readily applied to other children and settings than would have been the case had the study taken place in a lab setting, allowing for some generalisations to be made about populations.

However, there is also the possibility that the specific person delivering the intervention, or the specific teachers, or the specific school setting, may be instrumental in the success of the intervention (Witt et al., 1984). This would mean it was not possible to generalise any findings to other school settings. This possibility can be reduced however (as it was in this study) but creating a fairly manualised intervention, to ensure that it was at least implemented in a similar way by teachers in different settings (Bradshaw et al., 2015). Also, the design of the intervention (which allowed for a high degree of individualisation in implementation by different teachers) had some flexibility built into the design to allow teachers to tailor the intervention to their own class and teaching preferences (and child learning preferences), yet each with the same aim or addressing the same feature of WM. Again, this kept the intervention as
standardised as possible, to increase the internal validity and generalisability of the intervention (Yu & Frempong, 2012).

In terms of research methods, rating scales, cognitive assessments, WM assessments, questionnaires, school records of academic achievement and observations were all used to explore research questions 1, 2 and 3, which sought to uncover cause and effect relationships. These methods were quantitative in the sense that they were highly structured, yielding quantitative data (as explained below). Research question 4, which focused on teachers’ perspectives and understandings, was answered through focus groups, which was the only purely qualitative research method adopted in this study.

4.2.3 Participants
4.2.3.1 Teachers
The research was conducted in two different governorates in Kuwait, with no key differences between them in terms of student intake or school provision (Central Statistical Bureau, 2019). The research was conducted in DS units (two girls’, two boys’) within four mainstream schools (one unit in each school). The schools were all in urban cities in Kuwait, with similar levels of economic affluence, with approximately 600-800 pupils in the entire school (an average school size in Kuwait), with no more than 5 children with DS in each class (there may be several classes with students with DS, however, in a school). The children with DS have their own suite in the school, with separate teachers and classrooms (detailed in section 1.6); however, they mix with the mainstream children during breaks and at morning assembly. The schools all follow the same curriculum for children with DS, which includes teaching in literacy and numeracy. For practical purposes, participants (both teachers and pupils) were limited to these four schools.
There are some similarities between provision in Kuwait for those with DS and provision for students with DS in mainstream primary schools in the UK. In the UK, like in Kuwait, many children with DS are taught in mainstream schools, particularly in the primary years (Faragher, Robertson & Bird, 2020). There may be an additional member of staff (e.g., a pupil support assistant in Scotland) to support the pupil in the UK. In the UK, similarly to Kuwait, some children with DS are taught in special schools, but these tend to cater for children with a range of additional support needs, rather than being DS-specific (like in Kuwait).

However, there are also specific differences in the provision and inclusion of students with DS in mainstream schools in Kuwait, compared to the UK. In Kuwait, those with DS are taught in special suites/wings of mainstream schools. However, within mainstream schools in the UK, children with DS tend to be included in a mainstream class for most/all of the time, though they may spend some time outside the class, such as working 1:1 with a pupil support assistant or with other pupils with additional support needs (e.g., in an additional support needs base in Secondary Schools). Furthermore, whilst there are different teachers in Kuwait primary schools for different subjects (such as numeracy, literacy and so on), in mainstream and special primary schools in the UK, there would usually be one teacher who would teach almost every subject, although there may be additional teachers for some specialist subjects such as physical education or music. In mainstream secondary schools in the UK, however, there are usually different teachers for each subject. In the UK, children with DS in mainstream and special schools will usually be in a class with same/similar age peers, following the national curriculum, although this might include an adapted version of it [note that some terminology and policy varies across the different countries in the UK]. In Kuwait, those with DS have their own curriculum, as discussed previously (see section 1.6 in chapter one), focusing on key skills such as life skills.
The teachers’ sample consisted of 28 female teachers who taught either numeracy or literacy in these units (14 teachers for each subject). 27 of the 28 teachers had a graduate qualification (96.43%); only one teacher in the control group had a post-graduate qualification (3.57%). The mean length of teaching experience was 12.54 years \((SD=8.15, \text{ range}=1-37)\), whilst the mean length of teaching experience of children with DS was 1.39 years \((SD=.49, \text{ range}=1-22)\). The distribution of teachers across the different schools can be seen in Table 4.1.
Table 4.1 Distribution of Teachers Across Schools, Teaching Subject and Experimental or Control Group

<table>
<thead>
<tr>
<th>School</th>
<th>Experimental Group or Control Group</th>
<th>Girls’ or boys’ school</th>
<th>No. of teacher participants</th>
<th>No. of numeracy/literacy teachers</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>EG</td>
<td>Boys</td>
<td>6</td>
<td>4/2</td>
</tr>
<tr>
<td>2</td>
<td>EG</td>
<td>Girls</td>
<td>8</td>
<td>3/5</td>
</tr>
<tr>
<td>3</td>
<td>CG</td>
<td>Boys</td>
<td>8</td>
<td>4/4</td>
</tr>
<tr>
<td>4</td>
<td>CG</td>
<td>Girls</td>
<td>6</td>
<td>3/3</td>
</tr>
</tbody>
</table>

Teachers taught either literacy or numeracy, and each taught only one class. All classes consisted of between one and four pupils. Teachers were required to complete a consent form prior to the study commencing.

4.2.3.2 Pupils

The pupils’ sample comprised 31 pupils (15 male and 16 female, aged from 6.58 years to 15.42 years), who attended one of these four special units in Kuwait for pupils with DS. All pupils were Kuwaiti and of Arabic ethnicity.

The inclusion or exclusion criteria for child participants included that the children be aged between 6 and 16 years and be diagnosed as having Down syndrome, but not diagnosed as having any other developmental disabilities, such as ASD. In fact, no students were excluded, as this criteria coincided with the schools’ own criteria for pupil admissions, which excludes children with DS and another significant disability, such as ASD or epilepsy (designed to ensure conformity to the national inclusion system). These individuals (with co-occurring conditions) attend specific schools for students with DS, given that they are more equipped to support these individuals.

Table 4.2 provides further information about the demographic and assessment scores of the children in the two groups. The assessment measures are
described in further detail in section 4.2.4.2. A t-test revealed the groups did not differ significantly in age as sig level was .482, which is not significant as it is greater than .05. Also, a chi-square revealed no significant between-group differences of gender as significance level was .72, which is not significant as it is greater than .05. Table 4.2 shows that the children were spread across the educational stages (years) of primary schools, with a similar spread for each of the groups. There was no student in stage 4. Each class has a specific teacher for literacy and another specific teacher for numeracy, in addition to teachers for other subjects that were not the research focus, such as science and life skills. Each teacher taught all pupils in the class, and all pupils (and both the numeracy and literacy teachers) in the class participated in the study.

Table 4.2 Pupil Demographics

<table>
<thead>
<tr>
<th>Variable</th>
<th>Intervention (n=18)</th>
<th>Control (n=13)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pupil mean age in years</td>
<td>10.08 (2.26)</td>
<td>9.50 (1.48)</td>
</tr>
<tr>
<td>(SD)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of males (%)</td>
<td>8 (44.4%)</td>
<td>7 (53.8%)</td>
</tr>
<tr>
<td>Number of females (%)</td>
<td>10 (55.6%)</td>
<td>6 (46.2%)</td>
</tr>
<tr>
<td>n (%) at each school stage:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>6 (33.3%)</td>
<td>4 (30.8%)</td>
</tr>
<tr>
<td>2</td>
<td>3 (16.7%)</td>
<td>2 (15.4%)</td>
</tr>
<tr>
<td>3</td>
<td>5 (27.8 %)</td>
<td>5 (38.5%)</td>
</tr>
<tr>
<td>4</td>
<td>0 (00.0 %)</td>
<td>0 (00.0 %)</td>
</tr>
<tr>
<td>5</td>
<td>4 (22.2%)</td>
<td>2 (15.3 %)</td>
</tr>
</tbody>
</table>

*Nb: after assessment, pupils may be able to move up to the next stage/level of education, although if they are not ready, they will remain in their current stage.*

Parents were asked to complete a consent form prior to the study commencing, and report any health problems that their children had. Parents of only 4 of the 31 children reported that their children had health difficulties (3 in the EG, 1 in the CG). The health problems were as follows: 1 visual difficulties (male in stage
one); 1 cardiac problem (female in stage five); 1 dental anomalies (male in stage three); and 1 sleep difficulties (female in stage five, in the CG).

All children included agreed to participate and gave their consent to do so. The process of acquiring informed consent from children with a developmental delay is described in more detail in section 4.2.6 and in Appendix 2.1. Briefly, however, child consent forms were created consisting of pictures and very simple language, informing them what I would do with them if they participated (permission from caregivers was also gained (British Education Research Association, 2011; Mietola, Miettinen & Vehmas, 2017; O’Kane et al., 2019; Yan & Kerim, 2004)). Simple, colourful ‘forms’ were devised through which children were invited to give or withhold their consent to participate in the research by ticking either a happy face or a sad face respectively; they were also repeatedly told they could stop (withdraw) from the assessments at any time. For children with ID, the researcher and teachers needed to be especially aware that pupils may give non-verbal indications that they wish to withdraw (Skånfors, 2009), given the power differentials that exist between adult researchers and child participants (perhaps especially those with ID) (Khoja, 2016).

4.2.4 Data Collection

A summary of the measures used in this study are shown in Table 4.3. Some measures were not conducted at post-intervention or at follow-up (see 4.2.5.1 Procedures for a detailed rationale for why each measure was implemented at certain points) to avoid making too many demands on teachers’ time or because time was needed the intervention to have any significant impact on these measures. Finally, the numeracy and literacy tests were conducted at follow-up, as the school conducts this assessment at the end of each term, which coincided with the follow-up timeframe.
### Table 4.3 Measures Used in this Study

<table>
<thead>
<tr>
<th>Measure</th>
<th>Pre-intervention (weeks prior to intervention)</th>
<th>Post-intervention (weeks post-intervention)</th>
<th>Follow-up (weeks post end of intervention)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Teacher’s Demographics and Knowledge questionnaire</td>
<td>2 weeks ✓</td>
<td>1 week ✓</td>
<td></td>
</tr>
<tr>
<td>Pupils Demographics questionnaire</td>
<td>2 weeks ✓</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Strengths and Difficulties questionnaire</td>
<td>1 week ✓</td>
<td>4 weeks ✓</td>
<td></td>
</tr>
<tr>
<td>BRIEF-P</td>
<td>1 week ✓</td>
<td>4 weeks ✓</td>
<td></td>
</tr>
<tr>
<td>RCPM</td>
<td>3 weeks ✓</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WPPSI-III</td>
<td>3 weeks ✓</td>
<td></td>
<td></td>
</tr>
<tr>
<td>WM assessment</td>
<td>1 week ✓</td>
<td>1 week ✓</td>
<td>4 weeks ✓</td>
</tr>
<tr>
<td>Academic achievement record in literacy and numeracy</td>
<td>3 months ✓</td>
<td></td>
<td>4 weeks ✓</td>
</tr>
</tbody>
</table>

**4.2.4.1 Questionnaires**

Questionnaires are practical, cost-efficient and often supply standardised answers (Basit, 2010). These standardised answers allow the results to be easily quantified, particularly when the questionnaires include only questions eliciting pre-coded answers (Cohen, Manion & Morrison, 2018). The quantitative nature of such data means that results can be analysed in such a way that comparisons can easily be made with other studies of a similar nature and design (Patten, 2016).

However, there are also limitations to using questionnaires. The use of pre-coded answers in questionnaires, while facilitating quantification, limits the scope of the respondents’ answers (Krosnick, 2018). Respondents might find this frustrating, alienating them from the research. More significantly, perhaps, the researcher’s choice of questions limits the frames of reference within which
respondents can provide answers, biasing the findings towards the researcher’s rather than the respondent’s worldview (Denscombe, 2014; Krosnick, 2018). There is also a danger that poorly phrased, ambiguous questions allow participants to interpret the questions in different ways, diminishing the quantitative value of the data or invalidating the final results (Johnson & Christensen, 2019).

In this study, I was aware that the teachers might not be able to provide extensive, detailed explanations and reflections of their experiences before, during and after the intervention solely through the questionnaires that were used. That is why I also took a more qualitative approach in the focus group interviews, allowing interviewees to explore the full significance of the intervention and the strategies they adopted (Johnson & Christensen, 2019). The combination of these two methodologies provided more scope to fully understand the data obtained and the experience and effectiveness of the intervention. This is discussed further in due course.

Teacher Knowledge and Demographics Questionnaire
A questionnaire (Appendix 2.2), designed specifically for this study, was given to teachers at the two weeks’ pre-intervention and one-week post-intervention stages. The pre-intervention questionnaire asked teachers to provide details of their teaching experience, specialised DS teaching subject and qualifications (as presented in 4.2.3.1). The pre- and post-intervention questionnaires also included 22 multiple-choice questions: 14 questions that assessed teachers’ knowledge of WM in general, as well as specifically in DS, along with knowledge of WM improvement strategies and the relationship between WM difficulties and literacy and numeracy ability; and 8 questions that assessed teachers’ knowledge of CB specifically in DS, and strategies of supporting positive behaviour. These questions were built around what was being taught in the intervention sessions with teachers, so knowledge could be assessed both
before and after the intervention, as well as drawing on relevant literature (e.g., Emmons & Esken, 2014; Gathercole & Alloway, 2008; Pawan, 2016). There began with a long list of items that were then narrowed down through discussion with teachers. Whilst there are steps in questionnaire development (such as factor analysis) (Rattray & Jones, 2007), these were not used in this study because this questionnaire built on specific topics to assess teacher's knowledge of the cognitive abilities in DS, including EFs, as well as CB in DS. These questions were created using existing literature on the topic, with references such as those appearing under Table 4.6. Whilst factor analysis can be used to assess validity, this questionnaire had high validity given that the final questions were generated through discussion with teachers of students with DS. The questionnaire was also shown to have a high internal consistency in both pre- and post-tests.

The answers to these knowledge questions were in the form of a 5-point Likert scale (where 1=‘not at all confident’ and 5=‘very confident’ in their knowledge). Likert scales do not require a simple yes/no answer from the respondent, but allow for degrees of opinion, and the quantitative data generated is easily analysed. Although some authors (e.g., Leung, 2011) suggest an 11-point scale, in the present study a five-point scale was chosen as it was deemed more practical, in the sense that it allowed for fast responses, yet it still facilitated the expression of opinion in the sense that respondents need not struggle to find words to express themselves.

Yet what can be seen as a strength in some ways can also be seen as a weakness; when respondents are restricted to pre-formulated statements, they are unable to express any opinion or make observations that are not contained in/described by these statements, and the value of the data gathered may therefore be reduced. Moreover, regardless of the number of points on a Likert scale questionnaire, the value of this method for measuring attitudes can be
questioned, especially regarding social desirability. Questionnaire respondents might seek to cast themselves in a more positive light, compromising the validity of the data. Paulhus (1984) found that more socially desirable personality characteristics were reported when respondents had to include identifying information on a questionnaire, so offering anonymity on such questionnaires might help minimise social desirability bias. Applied to my study, although there were no questions asking for explicitly identifying data, such as names or addresses, there were demographic questions about, for example, years of teaching experience, which could be seen as partially identifying data. Thus, it is reasonable to consider whether the problem of perceived social desirability might have influenced the data generated. An experienced teacher might, for example, claim to possess greater knowledge than she really had, as a result of feeling that a teacher of several years’ experience should be more knowledgeable. To overcome this, I verbally reiterated the fact that all data gathered would remain entirely confidential, to encourage honest, open answers, without judgement from me regarding their answers.

In the present study, the WM items on the teachers’ knowledge questionnaire were analysed for internal consistency and reliability. Values of Cronbach’s alpha scale showed high internal consistency in pre- and post-test (.91 and .97 respectively). Furthermore, the CB items of the teacher’s knowledge questionnaire for pre- and post-test also showed acceptable to high internal consistency (.83 and .91 respectively).

*Pupil Demographics Questionnaire*

The pupil demographics questionnaire (Appendix 2.3) for parents was designed for this study, and included questions on the age, gender, nationality and ethnicity of the children involved in the study, as well as questions about any additional diagnoses, apart from DS, that the child might have.
**Strengths and Difficulties Questionnaire (SDQ)**

The Arabic version of the Strength and Difficulties Questionnaire (SDQ: Goodman, 1997) was completed by parents and half of the teachers in this study (the other half completed the BRIEF-P for each child) one week before the intervention (a week after the teacher knowledge questionnaire, to ensure they were not overloaded with questionnaires at any one time) as well as four weeks after the intervention to measure behavioural and emotional difficulties in the pupils. The SDQ is a 25-item measure, with five subscales (five statements for each subscale): four subscales are summed to measure total difficulties (emotional symptoms, conduct problems, hyperactivity, peer problems) and one subscale measures prosocial behaviour. The statements within each subscale may be responded to with either, ‘not true’, ‘somewhat true’ or ‘definitely true’ (rated 0, 1 and 2 respectively). There are also five statements that are phrased in the opposite direction; these must be reversed prior to scoring. In the total difficulties scale, a score of 20 and above (parent version) or 19 and above (teacher version) is classified as ‘very high’ (Ehcap, 2014; SDQ Info, 2016).

The SDQ was chosen as it is recognised as a reliable and valid measure of behavioural difficulties (Dahlberg et al., 2017); there is evidence for convergent validity with other measures, as well as good internal consistency of the SDQ Total Difficulties scale (Kersten et al., 2016). It has also been widely used in previous studies of children with DS (Næss et al., 2017; Sarimski, 2018; Glenn et al., 2013; Martínez et al., 2011). The SDQ was also chosen because it is very convenient and practical for teachers and parents to complete as it is relatively short.

A further reason why the SDQ was identified as the most appropriate measure of behaviour for this study was that it applied well to both the CG and the EG group. It is appropriate for the CG as it is a measure of CB. It is also appropriate
for the EG for it is possible that, as Otten and Tuttle (2011) have argued, improvements to WM will lead to reduction in the level of CB.

Although there are other measures of child behaviour, such as the Child Behaviour Checklist (Achenbach, 1991; Achenbach, Edelbrock & Howell, 1987) and the Conners’ Teacher Rating Scale (Conners et al., 1998), they are not available in Arabic. I could find only one study in Arabic that used the Conners’ Teacher Rating Scale (Mutwally & Zeyadah, 2016), and in this study the researchers produced their own translation of the scale.

Furthermore, I chose to use the SDQ because it assesses both strengths and difficulties, while other measurements of behaviour simply identify CBs. This inclusion of a strengths criteria in SDQ gave me a more complete measure of how the intervention affected the children. Examples of positive behaviour criteria measured in the SDQ are whether the child is ‘Considerate of other people's feelings’ and ‘Kind to younger children’.

The Arabic version of the SDQ (SDQ info.com, 2018) was used in the present study. The SDQ was originally developed and validated within the UK, so in the present study it was possible that cultural differences would affect the SDQ results. The reliability and validity of the SDQ test have, however, been replicated in several countries, and there is evidence to support the validity of the Arabic SDQ in particular. In a survey of the use of the SDQ across a variety of cultures and languages, Woerner et al. (2004) found that it demonstrated good psychometric properties and clinical utility. Studies reported in this survey include research in another Arabic country (Yemen), which supported the validity of the Arabic SDQ (Woerner et al., 2004). Another study conducted in Oman found similar results, verifying the validity of the Arabic SDQ through different measures (Kazem, 2016).
While I have not been able to find baseline normative data for Kuwait, the evidence for the reliability and validity of the SDQ across different cultures justifies comparison with UK norms, albeit cautiously. The scores on the 20 items for each individual were added to make the four SDQ Total Difficulties scales. Regarding the baseline data for this tool, in my study there are some cases that scored higher than 20 or 19 according to parents’ and teachers’ views, respectively, which is classed as a very high score, indicating behavioural difficulties. Regarding the parent view, there were 9 cases classified as demonstrating behavioural difficulties, while teachers’ scores demonstrated they believed there to be 6 cases among the participant group. 9 of these identified cases were in the EG, while 6 were in the CG, which means the EG had more children with a high rate of total difficulties than the CG. Also, it worth noting that there were two cases reported by both teachers and parents as children who have a very high rate of behavioural difficulties. Except these two individuals, there was very little overlap between the ratings of parents and teachers in their identifications of those with difficulties, perhaps indicating different behaviours in different contexts, or perhaps different ideas from parents and teachers about what constitutes difficult behaviour.

In terms of reliability, in the present study, all 20 items for the Total Difficulties scale were analysed for internal consistency reliability; values of the Cronbach’s alpha scale showed acceptable internal consistency for the parent-report on pre-test and follow-up (.71 and .72 respectively). Similarly, the teacher-report showed acceptable internal consistency on pre-test and follow-up (.77 and .72 respectively). The Prosocial Behaviour scale showed high levels of internal consistency for both the parent-report pre-test and follow-up and the teacher-report pre-test and follow-up (.88, .85, .77 and .79, respectively).
The Arabic version of the Behaviour Rating Inventory of Executive Function – Preschool version (BRIEF-P) Questionnaire was completed by half of the teachers (the other half completed the SDQ) and parents one week before the intervention and four weeks after the intervention. Having half of the teachers (e.g., all the literacy teachers) complete the BRIEF-P and half the teachers (e.g., the numeracy teachers) complete the SDQ meant it was less taxing on their time, yet all children had both scales completed about them. BRIEF-P is a 63-item rating form designed to assess children’s EF in the home and school environments and is appropriate for use by both teachers and parents (Isquith, Gioia & Espy, 2004). It assesses five functions via five subscales: inhibition, shift, emotional control, WM and planning/organising (PO), and then it provides a total score for these five functions, called a global EF. Each statement in these items can be answered ‘never, ‘sometimes’ or ‘often’ (rated 1, 2 and 3, respectively). Having teachers and parents complete the same form enabled an assessment and comparison of the relevant aspects.

Because performance-based measures of EF are usually administered in a structured, quiet, new testing environment on a one-on-one basis, they may not facilitate the demonstration of executive deficits or strengths to emerge. This is why standardised questionnaires that measure EF within a more ecologically-valid setting, such as the home or classroom environment (such as the BRIEF-P rating form), can therefore provide useful insight into the assessment of executive deficits and relative strengths. This includes the extent to which these deficits and strengths in EF are observed by others and how they may impact on everyday life (Sherman & Brooks, 2010).

The BRIEF-P has been used to assess the EF of a range of pre-school age children, with different developmental and acquired neurological conditions (Daunhauer et al., 2014). Memisevic and Sinanovic (2014) showed that there is
evidence for the validity of its use in various clinical populations and argued that
the BRIEF (and therefore BRIEF-P) scale can be used in identifying
weaknesses and supporting the creation of programmes for enhancing EF in
children. My systematic review showed that it has previously been widely used
to assess EF in individuals with DS (Memisevic & Sinanovi, 2014; Pritchard et
al., 2015).

Although a small number of studies of children with DS have used the BRIEF
(e.g., Memisevic & Sinanovi, 2014), the majority have used the BRIEF-P (e.g.
Lee et al., 2011; Daunhauer et al., 2014; Pritchard et al., 2015 and d’Ardhuy et
al., 2015). This is because it is a better fit with the developmental age range of
children with DS; it is clear and easy to use, with most scale items expressed in
simple sentences. It also covers different domains for assessing children, not
only in terms of their cognitive abilities, but also some aspects of personality and
life skills. As the research focuses on WM and academic achievement in class, it
was appropriate to conduct the BRIEF-P scale research with teachers, as they
could be expected to produce a more accurate assessment than parents in
relation to children’s EF in school. However, for other EF aspects that might
affect life skills, parents’ evaluations could be considered more valuable.
Involving both parents and teachers in these assessments also helped make the
data gathered less prone to any respondent bias. A possible drawback of the
BRIEF-P form however is that, with 63 items, it is time-consuming to complete,
and I tried to minimise difficulties here by allowing one week for teachers and
parents to complete the form.

The BRIEF-P scale was also appropriate for my study given that it is available in
Arabic, provided by the company Psychological Assessment Resources. This
company denies researchers the permission to make any alterations to the
translation, so it was used as provided. There are very few examples of
research in Arab countries using the BRIEF or the BRIEF-P. I only found one
study (Ismael, 2007) that investigated attention and EF in children with ADHD, using the BRIEF. This study, however, did not include children with DS and did not focus on the preschool age group, so my study is making a novel contribution to this area of EF research, in using the Arabic version of the BRIEF-P in relation to children with DS for the first time, which worked well as an evaluation tool; the respondents understood the items and the consistency scores were high.

The Cronbach’s alpha values for the BRIEF-P items for teachers and parents reveal that the internal consistency of the BRIEF-P is acceptable to high (values over 0.7 are deemed acceptable), with a range of .78-.96 and .78-.96 for items on the teacher and parent reports respectively, details for each subscale’s consistency are provided in Appendix 2.4.

4.2.4.2 Cognitive Assessments
I conducted cognitive assessments at the 3 weeks pre-intervention stage to identify child participants’ levels of ability. I used the Raven’s Coloured Progressive Matrices (RCPM: Raven, Court & Raven, 1990; Arabic version: Qurashi, 1987) to measure non-verbal cognitive ability and tests from the verbal subscale of the Wechsler Preschool and Primary Scale of Intelligence (Wechsler, 2002; Arabic version: Hadi & Murad, 2011) as a measure of verbal IQ. Cognitive assessments were conducted at the pre-intervention stage only to facilitate group-matching in terms of Raven’s score and as indicators of the pupils’ verbal IQ.

Cognitive assessments can give a unique insight into a child’s functioning in different cognitive domains to assist in identifying their specific areas of strength or weakness. There are, however, limitations to cognitive assessments. In particular, given that these kinds of tests provide a ‘snapshot’ of the child’s
abilities at a specific time point, it is possible that the child may demonstrate poorer results than they are actually capable of achieving, as a result of, for example, shyness, tiredness, anxiety or discomfort with the process of test-taking, perhaps linked to cultural background especially their experience/expectations of interacting with adults or strangers (Decker et al., 2013). Some of the tests take time to complete, which can generate challenges as many individuals with DS are easily distracted or tired. However, in the present study, this issue was somewhat mitigated by conducting the tests in the morning, after the children had had breakfast, when they might have been less tired and more alert. They were undertaken in a quiet, comfortable area, a special assessment room next to the DS suite in each school, that the children should have been familiar (and therefore comfortable) with.

Moreover, breaks were permitted, with toys for the participants to play with, when participants demonstrated they wanted a break from the tests. Research indicates that breaks are essential within tests that measure cognitive abilities (e.g., Janssen et al., 2014; Sievertsen et al., 2016), given that performance appears to peak directly after a break and can decline as time goes on. This demonstrates the very real impact that fatigue can have on the validity of the results. In students that have ID, such as DS, it seems even more important that they are given regular breaks to avoid test fatigue, which may affect the validity of the test. Other studies investigating the cognitive functioning of children with DS use this approach, allowing more breaks as the child loses focus or becomes tired, for this very reason (e.g., Edgin et al., 2010).

In my study, liaising with teachers who were very familiar with the children was important, and every effort was taken to ensure that none of the children were tired, unduly distracted or otherwise influenced in a way that might distort their test performance. This consideration applied to all assessments conducted during my research.
Raven’s Coloured Progressive Matrices

According to Qurashi (1987), the RCPM is a test designed to assess a child’s non-verbal mental age. It is suitable for children aged 5½ to 11 years of age. It contains three sections, each of which contains twelve questions. The range of scores achieved in current research is between 1-16 for the entire test.

The Arabic version of this test was chosen because, like the English language version, it is clear and colourful, so appropriate for use with children with ID, and is widely used in DS research to assess mental age (MA) (Amadó et al., 2016; Hippolyte et al., 2010; Lanfranchi et al., 2009a; 2009b; Laws, 2002; Numminen et al., 2001; Purser, 2015). Facon and Nuchadee (2010) investigated whether RCPM tests can facilitate similar performance in different groups, including study participants with DS. They concluded that the RCPM may be used with considerable confidence in studies comparing TD children, children with DS and ID participants of undifferentiated aetiology. However, this is refuted by Gunn and Jarrold (2004), who found that DS populations appear to make unusual errors on the RCPM compared to TD groups and those with moderate learning disabilities. They found that there was no significant age-related change in terms of the types of errors made by individuals with DS, compared to TD individuals; Gunn and Jarrold (2004) conclude that this unusual pattern of errors in individuals with DS may be due to challenges experienced by these individuals in integrating perceptual information, in their visual acuity and reduced task completion. This may mean that DS and TD groups, for example, are matched on flawed results that do not accurately depict MA. Regardless, the RCPM remains a validated measure of MA and is one of the most widely used measures of MA in the DS literature, enabling this study to draw comparisons with previous studies adopting this measure.
However, in the current research, the raw scores were used to assess similarities in scores between children in both the experimental and control groups, because in the Arabic version there was no information provided on standard scores. Using standard scores to compare groups is a technique widely used in educational research, and this is deemed to be preferable to using raw scores in general (Pind et al., 2003). This is because standard scores allow the probability of a score occurring within normal distribution to be calculated and allows for a comparison of two scores that are from different normal distributions (Stafford, 2006). However, in the Kuwai context, the only available version of RCPM is from 1987 (Qurashi, 1987). This does not provide information on standard scores. Instead, it provides information on percentage categories (e.g., 5%, then 10, 25, 50, 75, 90 and then 95%) to provide estimated levels of non-verbal MA. Thus, it does not provide norm-based comparisons; rather, scores are divided into seven general percentage categories of achievement on the RCPM and the differences between each percentage category is large. This can underestimate students' abilities if they achieve a score between two levels, classifying them incorrectly. For example, if they achieve a score that would be reflected as 24%, they would not quite achieve a place on the 25th percentile – instead, they would be classified as possessing an estimated NVMA of around the 10th percentile. I avoided this and used the raw scores instead, as I deemed this to be a more accurate reflection of the actual ability level of the child. Furthermore, the use of raw scores was justified as in this thesis, I did not aim to compare children with DS to TD children; rather, the RCPM was used to assess any differences between children with DS in the experimental group and the control group. In addition, the general protocol is that assessment tools should be consistent with the research context (Mohajan, 2017). Given the use of raw scores in Kuwait, I felt confident using raw scores in this thesis to reflect the NVMA of Kuwaiti participants in lieu of the unavailability of standardised scores. Furthermore, the use of raw scores on the RCPM to compare and/or match different groups of participants with ID is not uncommon.
in the literature (see, for example: Van Herwegen, Farran & Annaz, 2011; Riby & Hancock, 2008; Smits et al., 2011).

A drawback to using RCPM is that, because it can take up to 30 minutes to complete, children with DS might not be able to maintain focus on the task, as children with DS demonstrate difficulties in attention span (Carrasco et al., 2005). To help reduce distractibility or fatigue, consideration was given to test time, location and the allocation of breaks, as previously discussed in 4.2.4.2. The test was not counterbalanced in any form since it was only used once, and it is a standardised assessment task; therefore, it did not require counterbalancing.

Several studies conducted in Arabic countries have made use of the Arabic version of RCPM, used in the present study. This Arabic version is a translation from the English version with no modifications, and with a manual in Arabic (Qurashi, 1987). The Qurashi (1987) version has previously been extensively used in Kuwait where, if teachers suspect that a pupil may have cognitive or learning disabilities, there is an obligation for school psychologists working for the Ministry of Education to conduct RCPM tests, along with other cognitive and IQ assessments, such as the WIPPSI and the Binet (Roid & Pomplun, 2012). In Oman, a different translation (Kazem, 2008) has been used in schools for children with ID.

*Wechsler Preschool and Primary Scale of Intelligence—Third Edition (WPPSI-III)*

The WPPSI-III (Wechsler, 2002) is a standardised intelligence test, designed for use with children aged from $2^{1/2}$ to 7.25 years. It is used around the world, and widely used in DS research (e.g., Conners et al., 2001; de Santana et al., 2014; de Sola et al., 2015; de la Torre et al., 2016). The Wechsler school-age test was not used because the selected WPPSI-III subtests were designed for the
developmental age of preschool and school-age children; the Wechsler school-age test is not designed for the age of the oldest participants in my study and was therefore inappropriate.

I used three of the verbal subtests because I had already used the RCPM to assess non-verbal cognitive abilities, rendering the other non-verbal subtests unnecessary. I used the Receptive Vocabulary, Information and Picture-Naming subtests because they are designed for use with children across a wide age range as mentioned above. In addition, these subtests have the strongest correlation with full-scale IQ scores, compared to the other verbal subtests (Hadi & Murad, 2011). The correlations are: Receptive Vocabulary=0.91, Information=0.93 and Picture-Naming=0.90. The test, like the RCPM, was not counterbalanced in any form since it was only used once. The raw scores from these subtests were combined to give one single verbal ability score for each child. It is worth noting that for the Arabic version, information is not provided to allow the scores to be converted into MA.

In my experience, the only drawback of this IQ test is that it is time-consuming to use, as the researcher should continue the subtest until all questions have been asked, unless the child fails five question items in a row, at which point the test should be abandoned. The WPPSI-III was selected over other potential measures of verbal ability, such as the British Picture Vocabulary Scale, given that there is no Arabic version of British Picture Vocabulary Scale or other measures, that have proven validity in Arabic. There was one being developed in the United Arab Emirates, yet the researcher was still working on this when I began conducting my research. The Arabic version of the WPPSI-III was used, which was adapted from the English version by Hadi and Murad (2011) who established the reliability and validity of the Arabic version in tests with a large number (1,574) of children in Kuwait under the Ministry of Education's supervision. The reliability for the WPPSI-III 3 subtests combined in the current
The study was .959 on Cronbach's alpha, which indicates a high level of internal consistency.

It must be noted however that the Arabic version is not identical to the English version. For example, some of the individual items on the subtests are different, possibly for cultural reasons. For instance, there is a picture of a mosque and the Kuwait towers in the Receptive Vocabulary test, which is famous in Arabic culture but not available in the English version. This means that normative data from the English version cannot be used for comparison (Husni & Newman, 2015; Khalaila, 2013). However, the name and aim of the subtests are the same in the two versions.

**4.2.4.3 Working Memory Assessment**

The Lanfranchi tasks were selected to assess WM as they measure both verbal WM (VWM) and visuo-spatial WM (VSWM) (Lanfranchi et al., 2004), and have been extensively used in DS research (Borella et al., 2013; Carretti & Lanfranchi, 2010; Carretti et al., 2013). These were administered pre- and post-test and at follow-up.

The Lanfranchi tasks used were three VWM tasks and three VSWM tasks. The verbal tasks were the Word Span task, the Selective Word Recall task and the Dual task. The VSWM tasks were the Pathways task, the Selective Pathways task and the Dual task. All tasks have 4 levels of difficulty. Table 4.3 shows the name and a description of the different tasks used, with the range of possible scores that may be received. The scores were combined from the three verbal tasks to give a total VWM score (the same occurred for the visuo-spatial tasks).

This assessment is quick and easy to use, making it appropriate for the children in my study, especially as they were asked to participate in multiple tests.
Table 4.4 Verbal and Visuo-Spatial Working Memory Tasks in the Lanfranchi Tasks

<table>
<thead>
<tr>
<th>Task name</th>
<th>Task procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verbal WM Tasks</td>
<td></td>
</tr>
<tr>
<td>WORD SPAN</td>
<td>The child is given lists of 2-5 words and is required to repeat the list immediately and in the same order.</td>
</tr>
<tr>
<td>SELECTIVE</td>
<td></td>
</tr>
<tr>
<td>WORD RECALL</td>
<td>The child is given one or two lists of words and is required to repeat the first word of each list after seeing the entire set of materials.</td>
</tr>
<tr>
<td>DUAL TASK</td>
<td>The child is given a list of 2-5 words and is asked to remember the first word of the list and to tap his hand on the table when the word “BALL” is presented. Two lists of words for every level of difficulty are presented.</td>
</tr>
<tr>
<td>Visuo-Spatial WM Tasks</td>
<td></td>
</tr>
<tr>
<td>PATHWAYS</td>
<td>Children observe the journey of a small frog on a 3x3 or 4x4 chessboard. They are asked to recall the pathway immediately and move the frog from cell to cell.</td>
</tr>
<tr>
<td>SELECTIVE</td>
<td></td>
</tr>
<tr>
<td>PATHWAY</td>
<td>The child is shown one or two paths taken by a frog on a 4x4 chessboard and asked to remember the frog’s starting positions.</td>
</tr>
<tr>
<td>DUAL TASK</td>
<td>The child must remember a frog’s starting position of a pathway on a 4x4 chessboard; the child must also tap on the table when the frog jumps onto the red square.</td>
</tr>
</tbody>
</table>

For each task, the minimum possible score was 0 and the maximum was 8 across the 4 levels of difficulty for each task.

For use in my study, with Lanfranchi’s permission (personal correspondence), I had the Lanfranchi task instructions and questions (for the tasks mentioned above) translated from English into Arabic by the Oxford Translation Office, which is under the supervision of the Kuwaiti government. There was counterbalancing of tasks involving VWM and VSWM to combat fatigue, boredom and practice effects (50% had VWM first followed by VSWM tasks, the remaining 50% had VSWM first then VWM tasks later). Within the VWM or VSWM test, in the post-intervention test, the individual items were always presented in the same order, which increased in difficulty, so as to standardise this. The same words were used as stimuli in the verbal tasks as in the pre-test, but in a different order, to minimise practice effects. Furthermore, whichever task
(VWM or VSWM) was presented first to an individual in the pre-intervention test and were presented last (counterbalanced) in the post-intervention test.

The Lanfranchi assessment was used over other WM assessments given that the systematic review revealed it has been widely used, and is an accepted measure, of WM in children and adolescents with DS. This seemed to be the most appropriate measure, therefore. Moreover, the reliability for this WM assessment was satisfactory, as alpha coefficients (WM total pre-intervention=.791, WM total post-intervention=.852, WM total follow-up intervention=.836), all indicated acceptable levels of internal consistency as they were all above .70. Other WM assessments were considered, such as COGME (Klingberg et al., 2002); however, this automated, computer-based assessment lacks an Arabic version, and may prove unsuitable for some students (or could become intensely time-consuming) given the need for a certain level of computer literacy to operate and access, skills that some children with DS may lack as a result of varying cognitive or muscular challenges. Therefore, given these limitations, the Lanfranchi assessment was perceived to be the most appropriate assessment model.

4.2.4.4 Academic Achievement Assessment (Literacy and Numeracy Scores)
Academic scores from existing school tests of literacy and numeracy were chosen as an indicator of whether the WM intervention also affected academic achievement. These school tests were already available to me as they are routinely conducted by teachers at the end of each semester and therefore required no additional time-consuming tasks for the children. These tests are designed for pupils with special educational needs by a department within the Ministry of Education in Kuwait and are all built on the same stage-specific curriculum for children with DS. Because the tests are given to various schools on different dates, the tests are not identical across different schools, in order to avoid any leaking of questions. They are, however, of a consistent level of
difficulty. A maximum score of 20 is gained by each child on these assessments (reading and writing for literacy and calculation and arithmetic for numeracy). These were accessed in each child’s record. These tests were delivered in Arabic by their teacher. Translated English versions of these assessments can be found in Appendix 2.5.

It must be noted that while UK/US studies have the possibility of using standardised measures of literacy and numeracy (for example, the Test of Preschool Early Literacy Skills (TOPEL) (Lonigan et al., 2007) and the Preschool Early Numeracy Skills (PENS) test (Purpura, 2009)) no such measures exist in Arabic (at least, not in Kuwait), and it would not have been possible to have produced meaningful translations of the English measures, because of the curriculum and cultural differences across countries (Hassan, 2014).

**4.2.4.5 Structured Non-Participant Observation**

In this section, the use of an overt, structured, non-participant, event coding observation technique (Borich, 2016) is described as the most appropriate form of observation and coding to use to collect classroom data. Structured non-participant observation is commonly used for educational classroom research (Wragg, 1999/2012; Kane & Staiger, 2012) and this method was chosen to enrich and complement the data gathered using the other research tools. The observation was ‘structured’ in the sense that it took an event coding approach in which the behaviours that formed the focus of the observation were specified precisely in advance (e.g., Duff & Van Lier, 1997; Heath et al., 2010). This coding allows these specific behaviours to be counted, making structured observation a form of quantitative research (Hill et al., 2012).

One aim of the observation was to uncover how frequently the teachers used teaching strategies that have been shown to improve WM in the class (the
process by which these strategies were identified is discussed in the intervention section 4.2.5.1). To fit with this aim, it was decided that event coding was the most appropriate technique. Other coding strategies were considered, but not deemed a good match for the aims of this study. Interval coding, which measures behaviour occurring during specific time intervals (Cohen et al., 2018; Simpson & Tuson, 1995), would have offered no additional benefit and would have involved the risk of missing coded behaviours that occurred between intervals. State coding (Cohen et al., 2018; Simpson & Tuson, 1995) might have been an appropriate method for investigating the effect of WM interventions on children’s overall behaviour in terms of the duration of particular behaviours, but because the focus of the observation was how teachers responded to particular behaviours, the duration of these behaviours was not particularly relevant.

In the schedule of observation (Table 4.5), the extent to which teachers used classroom teaching strategies to improve pupils’ WM was the focus, and specific behaviours or responses identified as being indicative of this were defined operationally and coded as shown. Pupil behaviours to be observed were simply events that could be taken as indications of WM difficulties. Teacher behaviours to be observed were the use of strategies taught during the intervention (although, some strategies are not easily observed as they are ‘internal’ mental events, rather than ‘external’ observable events). Operational definitions were prepared using WM strategies taken from Gathercole and Alloway (2008) and modified after a pilot observation (see section 4.2.4.7).
<table>
<thead>
<tr>
<th>Behaviour/Strategy Name</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child Behaviours</strong></td>
<td></td>
</tr>
<tr>
<td>1. Failure to follow instruction</td>
<td>Child does not do what the teacher has asked within the time of the task. Also include partial failure (e.g. teacher gives 3 steps and child only completes 1).</td>
</tr>
<tr>
<td>2. Incomplete recall</td>
<td>In response to a question from the teacher, the child either states that they do know and then then says that they have forgotten OR starts to give an answer and says they have forgotten or then trails off.</td>
</tr>
<tr>
<td>3. Place keeping error</td>
<td>In a verbal task the child incorrectly repeats or skips an element.</td>
</tr>
<tr>
<td>4. Task abandonment</td>
<td>Child stops doing task before it is complete without instruction from the teacher to stop.</td>
</tr>
<tr>
<td><strong>Teacher Behaviours</strong></td>
<td></td>
</tr>
<tr>
<td>1. Teacher responds to child showing possible WM difficulty.</td>
<td>This is coded when the child shows one of the following behaviours: failure to follow the instruction; incomplete recall; place keeping error; task abandonment (see definitions under child behaviour) AND within 60 seconds of this occurring, the teacher does one of the following:</td>
</tr>
<tr>
<td></td>
<td>Talks to the child about the task: e.g. asks them what they are doing, whether they require help, asks whether they need an instruction repeated.</td>
</tr>
<tr>
<td></td>
<td>Looks at the child’s written work</td>
</tr>
<tr>
<td>2. Teacher reduces distraction</td>
<td>Teacher does one of the following:</td>
</tr>
<tr>
<td></td>
<td>Removes material unrelated to the task from the child’s desk, the blackboard, or the noticeboard.</td>
</tr>
<tr>
<td></td>
<td>Reminds pupils not to talk</td>
</tr>
<tr>
<td></td>
<td>Posts a “Do Not Disturb” sign on classroom door.</td>
</tr>
<tr>
<td>3. Teacher uses repetition</td>
<td>Teacher repeats a statement, questions or instruction within the same task. This can be a full repetition or partial (e.g. same meaning but a reduced number of words).</td>
</tr>
<tr>
<td>4. Teacher uses rhythm and rhyme</td>
<td>Teacher uses song (recording or their own voice) or makes a rhythm (e.g. clapping or tapping) or uses rhyming words.</td>
</tr>
<tr>
<td>5. Teacher uses visual memory aids</td>
<td>Teacher shows the pupil a picture, diagram, video, symbol, writing, or gesture which is either a clear stand-alone statement, question, or instruction OR reinforces a verbal statement, question or instruction.</td>
</tr>
<tr>
<td>6. Teacher uses chunking</td>
<td>Teacher: gives two or more statements, questions, or instructions as separate sequential units (rather than run together using connectives). gives a list of items (numbers, words) either semantically grouped, or with pauses to indicate grouping.</td>
</tr>
<tr>
<td>7. Teacher encourages student to use WM strategies</td>
<td>The teacher: tells/asks the child to rehearse or repeat information (could be out loud or silently), or tells/asks the child to use/create visual memory aids, or tells/asks the child to request if they need help remembering</td>
</tr>
<tr>
<td>8. Teacher evaluates WM load of task</td>
<td>The teacher asks the child if the task is too difficult to remember.</td>
</tr>
<tr>
<td>9. Teacher reduces task WM load</td>
<td>Teacher simplifies a previously used statement/question/instruction in a manner not already covered above (e.g. chunking, repetition, visual memory aid).</td>
</tr>
</tbody>
</table>

It was hoped that the observation (form in Appendix 2.6) would allow me some insight into how much the teachers understood and were correctly applying the WM strategies taught during the intervention. Observation was considered preferable to only using teacher self-reporting, which might be prone to biases, filters and preconceptions, given the teacher’s emotional investment in the object of study: her pupils and her own teaching practice (Cohen et al., 2018). Of course, the observer cannot observe from a completely neutral position (Cohen & Goldhaber, 2016) and will bring her own preconceptions and biases to the observation. However, the observer will be less emotionally involved and has the advantage of being able to observe and record without also being involved in the behaviours under investigation (Grosvenor & Rose, 2013). Using
observation alongside self-reporting (teacher knowledge questionnaires) and the focus groups allowed some degree of triangulation of the data gathered, which avoids some of the problems of relying on one method alone (Zohrabi, 2013).

In deciding the design of this research, I was aware of, and tried to compensate for, some of the limitations of observation as a research technique. Perhaps the main limitation of observation as a research approach in general is that it places focus on observable behaviour and therefore emphasises what happens, yet does not explore why this behaviour happens (O’Leary, 2013). Thus, it cannot uncover the intentions underpinning or motivating the observed behaviour (Simpson & Tuson, 1995). Observation can give the researcher largely unmediated access to behaviours and interactions, avoiding the distortions inherent in self-reporting through interviews or questionnaires, but it cannot explain these behaviours (Bell et al., 2012). For example, observation can only record whether a teacher displays strategy A or strategy B, or neither, but cannot easily offer any explanation for this. Such limitations can be minimised by supplementing observational research with other tools (Bell et al., 2012). In this study, the observations were supplemented by the use of teacher focus groups, in which questions regarding causes and motivations could be investigated.

A related limitation of structured observation is that behaviour should be given an operational definition that allows the researcher to clearly identify it, but some behaviours are difficult to define operationally. For example, one common WM strategy to support children’s memory is ‘evaluating memory load’ (Gathercole & Alloway, 2008). Although there are different signs that might allow a teacher to evaluate WM load and they can all be defined operationally, it is still hard to say whether a teacher has evaluated WM load or not, because it is a mental process that cannot be observed. I minimised such limitations by not only observing signs of WM difficulty in the children, but also observing teachers’ responses to
these signs. In addition to this, I asked teachers in the focus group interviews whether and how they evaluated a pupil's WM load.

**Main Study Observations**

The focus of the observation was the teachers, and I observed each teacher for 20 minutes one week pre- and two weeks post-intervention (see 4.2.5.1). Observations were conducted only with EG teachers as this was the only group that was taught WM strategies, which is the main focus of the current study. Each teacher in each school, whether a numeracy or literacy teacher, had only one class, so there was no issue regarding which class or children to observe the teacher with. Children were only observed in the sense that I was looking to see whether teachers would identify signs of WM difficulties in the child and respond using the strategies discussed during the intervention workshops, so no individual children were selected as a focus of observation.

**4.2.4.6 Focus Groups**

A focus group is a small group, usually between six and nine people, who, through discussion, explore their experiences, beliefs, feelings and opinions about a specific topic (Denscombe, 2014). This method of gathering data was chosen because it was hoped that it would enable the researcher, through interacting with and listening to the participants, to explore in a relatively quick and efficient manner (Dilshad & Ijaz, 2013; Denscombe, 2014), teachers' views on the effectiveness of the WM intervention more fully. Analysis of these focus group discussions also allowed a clearer picture to emerge concerning how teachers felt about the intervention, generating insights not available simply from the test result data. As Morgan (1988) and Palomba and Banta (1999) argue, focus group data lends itself to triangulation with data from other sources, such as questionnaire and observation data, and thus has the ability to complement and illuminate data generated from the questionnaires, assessments and observations in my own study.
In general, focus groups can elicit the opinions and feelings of participants more fully than might be the case with individual interviews, as the process of listening to and commenting on contributions from other interviewees in a focus group can often stimulate individuals into considering their opinions more fully and volunteering them more freely (Denscombe, 2014; Russell, 2000). Of course, this might not be the case when the topic under discussion is a very sensitive or personal one (Morgan, 1998), but it was felt that this consideration did not apply to my study. Other limitations of focus groups include the risk of a kind of ‘groupthink’, or the influence of power dynamics within the group (MacDougall & Baum, 1997), such as social pressure to conform to the opinion of more senior teachers. In my study, however, all teachers were at the same level in the school hierarchy as there were no senior teachers involved, reducing the risk of power differentials between teachers influencing contributions to the discussion.

In this study, the focus group contained teachers from the experimental group only, because the aim was to investigate teachers’ viewpoints about the WM intervention. The focus groups included both literacy and numeracy teachers, with one focus group of six teachers, and the other of eight teachers (as there was one focus group in each of the two experimental group schools). It was conducted one-week post-intervention.

During the focus group discussions, I first welcomed participants and thanked them for their participation. I presented a brief summary and feedback about the intervention, reviewing only the titles of the previous sessions, and avoiding any ‘leading’ comments that might have influenced the teachers to give an unduly positive assessment. I emphasised that the teachers should feel free to be completely honest, as the aim was simply to further develop the intervention. So, if they had found it difficult, or if they felt that it had made no difference to their children, or indeed had made teaching more difficult, they should feel free to say
so. Teachers were reminded of the intervention strategies with a handout distributed during the focus group discussion. The questions I posed to the teachers during the discussions were divided into main questions and supporting questions or ‘prompts’, to stimulate further discussion if the main questions appeared not to fully engage the teachers. A full list of main and prompt questions can be found in Appendix 2.7.

The focus group interviews were conducted in quiet surroundings, and were audio recorded using high-quality recording equipment. To facilitate the identification of participants, each mentioned their name every time they spoke. In the transcription process, all comments were anonymised, with names being replaced with numbers (Wilson, 1997). Participant consent for the recording and transcription had already been received. The actual transcription process was largely verbatim, but with fillers, background noises, repetitions and so on taken out. I avoided excessively cleaning up the focus group transcripts. If, for example, a participant used colourful language, made grammatical errors, misused or mispronounced words, I did not correct these while typing. Timestamps were inserted into the transcript every two minutes. This facilitated finding and cross-checking any quotes (Barbour, 2018). Also, the teachers were respectful and considerate, allowing one another to finish speaking, rarely interrupting and avoiding talking over one other, which facilitated the transcription process.

It was critical that I remain reflexive throughout the process of conducting and analysing the focus group. It is a given in much research literature that the researcher’s experiences, beliefs and knowledge affects the data collection and analysis, with interpretations potentially being influenced by the researcher’s preconceptions and worldview (Darawsheh & Stanley, 2014). My own experience and background working in education, for example, could have influenced the participants’ responses to both the intervention experience and
effectiveness as well as to me within the focus group (Greenbank, 2003). It is possible the participants felt more about to open up to me and be honest about their experiences given my background; equally, it is possible that, as I delivered the intervention, the participants may have wanted to be polite. My own desire to develop an effective WM intervention may also have affected the ways in which I analysed the data; I was conscious that I could therefore be biased towards reporting the positive experiences of the teachers and underreporting the negative experiences. Therefore, to help me overcome this, I kept a reflexive journal to assist me in the data collection and analysis process, as this encouraged me to reflect on and challenge my own thoughts, feelings and interpretations.

4.2.4.7 Pilot Work
A pilot process of all measures and methods used in this study was undertaken so as to hone each measure prior to the commencement of the actual study. These pilot processes are discussed briefly in this section. Note that there was no pilot study for the assessment of academic achievement, as this is a test set and conducted by the schools themselves. Therefore, the scores were taken from students’ records.

Pilot Questionnaires
Pilot studies were conducted of the questionnaires (BRIEF-P; Teacher Knowledge and Demographics Questionnaire; SDQ) used in this study, involving teachers, in order to gain their feedback on the clarity and useability of each questionnaire, especially those that had been translated into Arabic (e.g., the BRIEF-P). These measures were piloted on a different sample to that included in my final sample so as to avoid any practice effects on the final sample of participants.

In terms of the BRIEF-P pilot, four teachers of students with DS in the mainstream schools I selected for my sample were recruited. These teachers
were not numeracy or literacy teachers and therefore would not form part of my final sample. The pilot of the BRIEF-P revealed that no real changes to the measure were required, as the teachers were clear on the content of each item. However, it became apparent that the translated title of the form was confusing and perhaps had been mistranslated, with the title apparently referring to business or jobs – perhaps a mistranslation of ‘executive’. Apart from the title, no other errors were identified in the translation, so it was felt that this would not unduly affect the survey results.

In terms of the Teacher Knowledge and Demographics Questionnaire, four teachers of students with DS in different mainstream schools from those that would form my final sample were recruited as an opportunity sample. These were not numeracy or literacy teachers (this was not intentional). They were asked verbally during one of my many visits to the school to complete the paper questionnaire and return it to me. They were also asked to provide me with feedback about the clarity of questions and how long it took to complete. This taught me that some teachers did not understand some of the words used, such as ‘executive function’ and ‘working memory’. I then added another open question: ‘is there anything else you would like to add about teaching children with DS’, in case the teacher participants in the final sample did not know the terminology presented in the questionnaire. The final questionnaire was amended to provide greater detail and description regarding some of the more confusing terms, e.g., executive function.

In terms of the SDQ, the Arabic version of the SDQ for teachers (sdqinfo.com, 2018) was piloted on a sample of three teachers of children with DS from different mainstream schools that were not included in the actual sample for this thesis. These schools have the same set-up and environment as the schools in the present study. No amendments were required following this pilot as the
teachers found the Arabic version of the SDQ easy and quick to understand and complete.

**Pilot Cognitive Assessments**

The cognitive assessments (RCPM and Wechsler scale) were piloted with four children with DS (two for each assessment) from different mainstream schools that were not included in the final sample for this thesis, although these schools have the same set-up and environment and timetable as the schools in the present study. The cognitive assessments were piloted on a different sample so as to avoid order and practice effects on the final sample of participants. The reason for piloting the cognitive tests was to test their use for children with DS, such as the potential behavioural responses to the tests and the time it took to complete the assessment, and the best time of day to conduct the tests (given that all the schools, the pilot and final sample schools, have the same timetable and the same facilities). Piloting these tests meant that a dialogue with the pilot participants’ teachers could inform me of the best ways to help maintain the child’s attention, how far apart to sit the students, what time of day to conduct the tests, and so on. This was hugely informative for conducting these tests more effectively with participants in the final sample.

**Pilot Working Memory Assessment**

The WM assessment was piloted with 6 children with DS: 1 child from stage 1, 2 children from stage 2, 2 children from stage 3 and 1 child from stage 5. All were between the ages of 8-14. These children were not from the final research sample (for obvious reasons); rather, they were from different mainstream schools that were not included in the study.

Piloting the WM assessment revealed that I needed to make some minor changes to the tests, so that they would be appropriate for the Kuwaiti context. For example, the original verbal tests mention ‘pears’, which in Arabic would
require using a word unfamiliar to the children, so I substituted this with ‘bananas’ instead. In direct correspondence with Lanfranchi, I was assured that such changes were appropriate (Lanfranchi, 2018, personal communication, 26 July 2018). Lanfranchi also advised me that, in the Word Span task, the number of syllables in a word should be the same in Arabic as in the original English, and this was taken into consideration in the final version of the translation.

Pilot Observations

In the pilot phase, I tested the structured observation to make sure that the operational definitions could be observed in a real classroom environment. Prior to the live pilot observation, I looked at videos of teachers in the classroom, to train myself as to how they conducted the strategies and how I could code them. The live pilot observation was then conducted in one school only, over two sessions, with teachers that were not participants in my main study and with pupils who had typical levels of development and were not diagnosed with DS or any other learning difficulty. This was because I did not want to use study participants and I was unable to access any other children with DS or other ID in these governates.

To further enhance the reliability of my observations, during the pilot study my own observations were compared with observations made by another observer (a teacher not otherwise involved in the study) using the same schedule at the same time and place, and were found to be largely identical, which indicated that the operational definitions were appropriate for my study. One change that was made as a result of the pilot studies was that all of the operational definitions (child behaviour and teacher strategy) were merged into one single record table. This helped to make the recording of behaviours more efficient, ensuring that behaviours were less likely to be missed.
The pilot observations were also conducted to identify the optimal length of time needed for these observations. Through these pilot observations it was decided that 20 minutes of observation would allow me to gather sufficient data whilst minimising disruption to class activities.

**Focus Group Pilot**

So as to mitigate any potential limitations surrounding the risk of groupthink and power dynamics coming into play, I conducted pilot focus groups with 6 other female teachers not involved in the study sample. These participants were selected from mainstream schools in my sample, but not literacy or numeracy teachers, and the pilot was conducted in a classroom at the school. This pilot assisted me in practicing the skills needed to successfully carry out focus group research and so minimise the risks outlined above. These skills included taking note of and asking for the input of individuals that had not spoken in a while or seemed more reluctant to offer an opinion, as well as active listening and tactful intervention skills, especially when the conversation went off-topic or was dominated by particular individuals.

In this pilot phase, I experimented with different ways to manage the focus group discussions, including timing, how to arrange seating and different ways of recording, settling on discussions of one hour, with participants seated around a circular table, with the recorder passed from teacher to teacher. I made it clear that the recorder was to be passed to whoever wanted to speak; in the focus groups, however, all participants wanted to speak and appeared to have a response for every question asked of them, so the recorder was passed around the circle, from teacher to teacher, the majority of the time. I invited different teachers to begin responding to different questions, to avoid one teacher consistently giving their opinion first (which could have influenced the responses of other participants).
Pilot Intervention Sessions
It was important that I designed the WM and CB intervention sessions to fit into teachers’ existing schedules so as to cause only minimal disruption. This was established through the pilot study. Teachers were recruited from one of the mainstream schools in the sample (not the same one as the focus group pilot), but did not involve literacy or numeracy teachers. The outcome of this pilot was that I amended the intervention session to allow more time for explaining and exploring the participants’ driving questions, especially in the first session, which was already due to be the largest intervention session. This was because this session involved an in-depth introduction to the aim of the research, EFs, WM, and my intervention, as well as covering important points such as driving questions, that were integral to the rest of the intervention. The first session took an hour and 10 minutes, but as a result of the pilot sessions, I allowed for this. Furthermore, the pilot intervention sessions revealed that the participants were happy for subsequent sessions to last between 45 to 60 minutes, as this time frame did not affect their timetable.

Also, I was aware that I should not waste the time of the control group participants, so I designed and provided an intervention that I considered would be useful to all teachers of children with ID and this opinion was confirmed through pilot study feedback.

4.2.5 The Intervention
The experimental group participated in the WM intervention while the control group participated in the CB intervention. Both the experimental and the control interventions aimed to work with teachers to support them in the development of teaching strategies that would help children in the class. These strategies had originally been developed by expert practitioners and researchers who have researched WM, including well-known researchers such as Jarrold (2017), Hall and Jarrold (2015) and Gathercole and Alloway (2006; 2007; 2008), and CB
(Positive Behaviour Support, PBS) in people with intellectual disabilities, such as Otten and Tuttle (2011), ProfHastings (2018) and Allen (2018). For each intervention group, there was a session every week for six weeks, during which I presented information about important aspects of WM or CB in class. The sessions were varied and included lectures, film clips, group discussions and role play, with information taken and curated from a range of sources to best suit my purposes. In all these sessions, teachers were encouraged to contribute their own opinions and expertise to these exercises and discussions. For clarification of my exact materials, I will now outline each intervention.

Regarding WM, the material used in the intervention was drawn from Gathercole and Alloway’s (2006; 2007; 2008) and Jarrold’s (2017), Hall and Jarrold (2015) studies, with some additional materials to give greater clarification on WM to the teachers, many of whom were unfamiliar with this concept. Gathercole and Alloway’s (2006; 2007; 2008) studies were used to explain WM, including the ways to evaluate WM load in the classroom, recognise WM difficulties, teach strategies to support WM and ways to develop the child’s own strategies to support WM. Similar to Gathercole and Alloway's contents, Hall and Jarrold's (2015) recent research was drawn upon to inform teaching about children who experience difficulties with WM, including how WM works, ways to assess WM, WM in DS and strategies to improve WM (these are predominantly the same strategies that are mentioned in Gathercole and Alloway’s (2006; 2007; 2008) studies), with the main focus on dealing with distraction (see Table 4.6 for a list of session contents).

I also translated small portions of previous studies that focused on WM in DS, such as Wang and Bellugi (1994), Lanfranchi et al. (2004; 2009a; 2009b) and Hughes (2006), with the aim of giving teachers a greater understanding about WM in DS. I also drew on the results of various studies that focused on WM interventions, such as Conners et al. (2008) and WM and learning, such as
Alloway and Alloway (2010), Christopher et al. (2012), Young (2000) and Geary et al. (2000), to show teachers the possibility of improving WM and its impact on learning.

Further reading for the teachers was provided by handing out translated segments of Gathercole and Alloway’s (2007) *Understanding WM: A Classroom Guide* and from Hall and Jarrold (2015), who also discuss WM as a cognitive function in addition to its strategies on fact sheets (University of Bristol, 2020a). In each further reading, I always indicated the relevant pages to be read for each specific session and strategy that was being focused on.

In the CB intervention (PBS) sessions, we discussed definitions of CB (Emerson et al., 2001; Royal College of Psychiatry, 2007), CB in DS, how to recognise different types of CB (external and internal) (Smith, 2014; Huxley et al., 2005), causes of CB (National Down Syndrome Society, 2017), the correlation between EF and CB in general (Hughes & Ensor, 2008), the correlation between EF and CB in DS (Costanzo et al., 2013; Oosterlaan, Scheres & Sergeant, 2005; Oosterlaan & Sergeant, 1996), common functions of CB (Otten & Tuttle, 2011), and types of CB in people with DS (Evans & Gray, 2000; Coe et al., 1999; National Down Syndrome Society, 2017).

I also drew on the following materials to discuss the effect of CB on teachers, pupils and the class in general, as well as teachers’ perception and emotional responses towards students that demonstrate CB (Silberman, 1969; Jones & Hastings, 2003), common and effective practices in the education of children with social, emotional and mental health difficulties in students with special needs (Carroll & Hurry, 2018). I also drew on the ProfHastings’ blog (2018) when discussing CB in children with DS. I also explained how to evaluate CB by creating an observation form and explained: the questionnaire adapted from
Mustapha (2006)\textsuperscript{12}; Positive Behaviour Support (PBS) interventions for CB in DS and functional behaviour assessment (Otten & Tuttle, 2011); and strategies to address CB in children with DS (Feeley & Jones, 2008).

The intervention sources, including any films used, were all translated from English into Arabic by the Oxford Translation Office, which is under the supervision of the Kuwaiti government. A list of the contents of the intervention sessions are available in Appendix 2.8.

\textsuperscript{12} This was designed specifically for children with ID and was developed and published in the Um Al Khura University Magazine for Human, Social and Educational Science.
Table 4.6 Intervention Session Topics

<table>
<thead>
<tr>
<th>Session No:</th>
<th>Experimental group WM INTERVENTION&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Control group CB INTERVENTION (PBS)&lt;sup&gt;b&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Introduction.</td>
<td>Introduction.</td>
</tr>
<tr>
<td></td>
<td>WM definitions</td>
<td>Challenging behaviour definition.</td>
</tr>
<tr>
<td></td>
<td>Model of WM</td>
<td>Challenging behaviour in Down Syndrome</td>
</tr>
<tr>
<td></td>
<td>WM in Down syndrome</td>
<td>Causes of challenging behaviour: internal and</td>
</tr>
<tr>
<td></td>
<td>WM and learning</td>
<td>external factors.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Teachers' attitude and response toward children</td>
</tr>
<tr>
<td></td>
<td></td>
<td>with CB.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Common functions of challenging behaviour.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>The effect of challenging behaviour on teachers,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>school and other children.</td>
</tr>
<tr>
<td>2</td>
<td>Recognise WM difficulties in the classroom.</td>
<td>Recognising types of CB (external and internal</td>
</tr>
<tr>
<td></td>
<td>Incomplete recall</td>
<td>CB).</td>
</tr>
<tr>
<td></td>
<td>Failure to follow instructions</td>
<td>Stubbornness</td>
</tr>
<tr>
<td></td>
<td>Place keeping errors</td>
<td>Not following instructions</td>
</tr>
<tr>
<td></td>
<td>Task abandonment</td>
<td>Social withdrawal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Aggression</td>
</tr>
<tr>
<td>3</td>
<td>Evaluate WM load of tasks taught.</td>
<td>Evaluating CB.</td>
</tr>
<tr>
<td></td>
<td>1. Ask the child</td>
<td>Observation</td>
</tr>
<tr>
<td></td>
<td>2. Observe how the child responds to the task:</td>
<td>Behaviour checklist.</td>
</tr>
<tr>
<td></td>
<td>* Incomplete recall</td>
<td>Discussing with parents</td>
</tr>
<tr>
<td></td>
<td>* Failure to follow instruction</td>
<td></td>
</tr>
<tr>
<td></td>
<td>* Place keeping errors</td>
<td></td>
</tr>
<tr>
<td></td>
<td>* Task abandonment</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3. Evaluate the task (with the child in mind):</td>
<td></td>
</tr>
<tr>
<td></td>
<td>There are many features of a classroom task</td>
<td></td>
</tr>
<tr>
<td></td>
<td>which could lead to overload, such as:</td>
<td></td>
</tr>
<tr>
<td></td>
<td>* Long sequences</td>
<td></td>
</tr>
<tr>
<td></td>
<td>* Unfamiliar and meaningless content</td>
<td></td>
</tr>
<tr>
<td></td>
<td>* Demanding mental process activities</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Teaching strategies to support WM in Down</td>
<td>Positive Behaviour Support for CB in Down</td>
</tr>
<tr>
<td></td>
<td>syndrome.</td>
<td>syndrome.</td>
</tr>
<tr>
<td></td>
<td>Reducing task WM load</td>
<td>How is Positive behaviour supported in the class?</td>
</tr>
<tr>
<td></td>
<td>Reducing distraction</td>
<td>The relationship between Positive Behaviour</td>
</tr>
<tr>
<td></td>
<td>Repetition</td>
<td>Support and Functional Behaviour Assessment</td>
</tr>
<tr>
<td></td>
<td>Rhythm and rhyme</td>
<td>How to use Functional Behaviour Assessment</td>
</tr>
<tr>
<td></td>
<td>Memory aids</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Chunking</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Develop the child’s own strategies to support</td>
<td>Model of Positive Behaviour Support intervention.</td>
</tr>
<tr>
<td></td>
<td>WM.</td>
<td>Prevention</td>
</tr>
<tr>
<td></td>
<td>Asking for help/repetition</td>
<td>Instruction (teaching replacement behaviour)</td>
</tr>
<tr>
<td></td>
<td>Rehearsal information</td>
<td>Reinforcement</td>
</tr>
<tr>
<td></td>
<td>Note taking</td>
<td>Negative consequences</td>
</tr>
<tr>
<td>6</td>
<td>Review and reflection.</td>
<td>Review and reflection.</td>
</tr>
</tbody>
</table>

<sup>a</sup> For most of sessions, there were also videos clips presented that were derived either from YouTube, uploaded by researchers, or from psychology-based websites (e.g., in session 1: Gathercole, 2011; Archibald, 2012; BBC Earth Lab, 2014; Komodo Maths, 2017; Digital Promise, 2017; University of Bristol, 2020a; University of Bristol, 2020b; Emmons & Esken, 2014; Pawan, 2016).

<sup>b</sup> Videos clips from YouTube were used throughout to emphasise points, including Teachings in Education (2017) to explain Functional Behavioural Assessment and the Challenging Behaviour Foundation (CBF, 2018) to explain PBS. Further reading was also provided to the teachers, including Hutchinson et al. (2014), Fidler et al. (2006), Johnson (1998), and the National Down Syndrome Society (2020), which were translated into Arabic.
One important component of both interventions was the concept of ‘driving questions’. This was adapted from materials originally created by Perth and Kinross Educational Psychology Service (EPS) in the UK. Approval to use this technique was obtained from Perth and Kinross EPS (2021); their materials were then translated into Arabic with no adaptations necessary for Kuwaiti participants.

At the end of each session, teachers had time to develop their own personal driving question(s), which would be discussed at the start of the next session. The intention of these driving questions is to help teachers reflect on, and understand, the aims and goals of each session and the implications of the session content for their own classroom practice (Perth and Kinross EPS). They did this by designing a task for themselves to complete before the next intervention session. These driving question(s) could reflect either a specific challenge with a particular pupil that they wanted to understand and explore, or a change that the teacher wanted to make in their own teaching strategies. Driving questions are more open than direct questions as they allow teachers to think about their personal response to session input. In my study, they were intended to prompt a constructive discussion between teaching colleagues at the start of each intervention session, and a chance to prepare the driving questions in the end of each session.

4.2.5.1 Procedure for Teachers
After receiving approval from the School of Education and Sport, University of Edinburgh ethics committee (Appendix 2.9), I contacted the Educational Research Department in the Kuwait Ministry of Education and obtained approval to conduct my research in four different schools in Kuwait. I explained my research to the schools’ management personnel, teachers and parents, and I obtained their written permission to conduct my research, providing them with information about the research and a consent request form for teachers and
parents (Appendix 2.1). These teachers were initially provided with information about the study at a meeting arranged with the cooperation of the school management. I gave teachers an information leaflet about the proposed study, explaining why I was asking them to participate, and I discussed my research and answered their questions. All literacy and numeracy teachers of pupils with DS in the schools agreed to take part.

Pre-intervention
Teachers completed the ‘teachers’ knowledge’ questionnaire two weeks pre-intervention, and either the BRIEF-P or SDQ questionnaires one-week pre-intervention. For every pupil in each class, the BRIEF-P or SDQ was completed by either their literacy teacher or their numeracy teacher. In the first city, the EG group numeracy teachers assessed children using BRIEF-P, while the literacy teachers assessed children using the SDQ, and the opposite in the CG. In the second city, EG group literacy teachers assessed children using BRIEF-P while numeracy teachers assessed children in the SDQ and the opposite in the CG. This was done for three reasons; firstly, to avoid the same teacher assessing the same child in two different assessments; secondly, to give teachers of both literacy and numeracy a chance to try each type of assessment in each (EG and CG); and thirdly, by spreading the questionnaires across all the teachers, it reduced the time demands on them. Note that the same teachers assessed the same child pre-test and at follow-up, using the same measure and each teacher assessed all pupils in her class. This means that each child had a pre-assessment and follow-up SDQ and BRIEF-P.

Teachers in the EG only were observed one week before the intervention for 20 minutes, because the EG was the only group that was trained in the WM strategies. The aim of the initial observation was to identify the WM strategies that the teacher already used before the intervention and to assess the frequency of their use. A one-week period between the initial observation and
intervention was considered short enough to avoid any other significant influences on teaching practice occurring, such as other training inputs or the headteacher’s instructions.

**Intervention**
Teachers were enrolled into either the WM in DS intervention group or the CB in DS intervention group. All teachers attended all training sessions for the intervention.

**Post-Intervention**
One week after the intervention, teachers again completed the ‘teachers’ knowledge’ questionnaire and I conducted the focus groups. The one-week time gap is justified by the fact that the teachers were likely to still be using the strategies within their classrooms; I also wanted the feedback to remain fresh in the teachers’ minds; and finally because the teachers would soon begin preparing for their students’ final exams, therefore it was unwise to wait any longer given the extra time pressure they may feel. I then observed the teachers two weeks after the intervention for 20 minutes to assess the effect of the intervention. A two-week interval was chosen as it was felt that this was long enough to allow teachers to implement the strategies that they had learnt during the intervention, but short enough to avoid any decline in teachers’ use of the intervention strategies (Moyles, 2002). I was careful to make the conditions of the pre- and post-observations similar in term of the time and place, and the class observed.

**Follow-Up**
The BRIEF-P and SDQ questionnaires for teachers were completed four weeks after the intervention. To achieve a valid test of the intervention’s effectiveness, it was necessary to ensure adequate time for the development of any changes as a result of the intervention. A common criticism of similar studies is that tests
of an intervention’s effectiveness follow too soon after the intervention and do not allow adequate time for effects to develop (Kersten et al., 2016). In my study, I avoid this drawback, as the follow-up was 10 weeks after the baseline assessment and 4 weeks after the end of the intervention (similar to Sherman and Brooks’ (2010) study, which also used the BRIEF-P to measure EF in preschool children), which was enough time for any such changes to become apparent.

The WM intervention materials used with the EG and the CB intervention materials used with the CG were made available to all teachers after the follow-up data had been collected. This was done so as to be as inclusive as possible of the teachers who had helped in my research, and to provide them with materials and information that they would likely find interesting and useful for their own teaching practice. Also, I in the final session, I supported teachers to prepare a plan to be used by them with pupils in the class to continue using interventions strategies in the longer term and informed them that I would be available to help them if there were any questions during the academic year. This was to ensure that the benefits of the intervention would be continuous.

4.2.5.2 Procedure for Children and Parents

Pre-Intervention

Children were assessed over a three-week period prior to the intervention. Pre-assessment tests were the Ravens Matrices (RCPM: Raven, Court & Raven, 1990; Arabic version: Qurashi, 1987) and the Wechsler Preschool (verbal subtest) assessment (Wechsler, 2002; Hadi & Murad, 2011). These assessments were completed three weeks pre-intervention while the Lanfranchi (2004) WM tasks were completed one-week pre-intervention. Parents were given the pupil demographic questionnaire, SDQ (Goodman, 1997) and BRIEF-P (Isquith, Gioia & Espy, 2004) one-week pre-intervention. The literacy and
numeracy assessment records were accessed, which were last completed in each school 3 months pre-intervention.

In order to avoid putting the child under excessive pressure, which might upset the child and distort the results of the assessments, I decided not to use more than one assessment for each child in one day. This was also important given the attentional and motivational factors that may affect performance; Purser (2014) notes that this may be a common occurrence with the use of the RCPM in DS, therefore emphasising the importance of spacing out the assessments to ensure maximum focus and comfort for each child.

Post-Intervention
The Lanfranchi (2004) WM assessments were conducted one week after the intervention.

Follow-Up Assessment
Follow-up WM assessments for children, parents’ SDQ, and BRIEF-P were conducted four weeks after the intervention. As noted above, this follow-up was 10 weeks after the pre-intervention assessment, which was considered sufficient time to allow any changes to become apparent.

4.2.6 Ethical Considerations
Research ethics specific to disability research are located within a wider research ethics framework. In the UK, research ethics generally are not governed by specific statute laws – no law compels researchers to observe ethics guidelines on consent, although the EU General Data Protection Regulation (GDPR) and UK Data Protection Act 2018 govern the processing (acquiring, holding, using etc.) of personal data, both in the UK and the EU. This means that social science research is obliged to maintain confidentiality for the participants involved (UK Research and Innovation (UKRI), 2020). However, in
the University of Edinburgh, the College of Arts, Humanities and Social Sciences has a research ethics framework, based on the Code of Practice for Research produced by the UK Research Integrity Office (UK Research Integrity Office, 2018). This, along with the British Psychological Society (BPS, 2020), provided a code of practice, where researchers should adhere to principles of dignity, care for others, respect, integrity, objectivity, accountability, honesty, openness and leadership. I have tried throughout to ensure that my research conforms to these principles. In particular, when conducting research in the field of disability, issues of dignity, respect and care for others are paramount, as there is a risk that already marginalised and disempowered individuals might be further marginalised and disempowered by a research approach in which the research is done on the individuals with disabilities, rather than with them (Iacono, 2006).

Children with disabilities, as with all children, should be able to exercise self-determination and their right to say yes or no (Dalton & McVilly, 2004). Children’s rights must be considered therefore when gaining consent, both from children and from parents of children with an ID, given that from a human rights perspective, children with an ID are arguably especially vulnerable. This can therefore create a special ethical challenge, given the delicate balance between viewing them as children with agency and protecting their vulnerability (Iacono, 2006; Mietola, Miettinen & Vehmas, 2017; Yan & Kerim, 2004). Cameron and Murphy (2007) argue that since children with ID may find it a challenge to understand and communicate with others, they should be provided with ample time to consider whether they wish to participate in research. The researcher must be mindful that gaining consent from a child does not automatically offer protection from harm, and so an ongoing awareness of ethical considerations is necessary throughout the research, even after consent has been obtained (Mietola, Miettinen & Vehmas, 2017).
Regarding consent, for all participants in my research, I tried to explain each component of my research clearly, through information included on consent forms and through verbal communication. Although the primary focus of my research was on the teachers rather than the pupils, every effort was made to ensure that the research was explained to the pupils with DS in an appropriate way, and their informed and free consent was obtained. The child consent form I created consisted of pictures and very simple language and I informed them what I would do with them if they participated. Moreover, it was necessary to gain permission from their caregivers and clearly detail the research purpose and methods (British Education Research Association, 2011; Mietola, Miettinen & Vehmas, 2017; O’Kane et al., 2019; Yan & Kerim, 2004), doing my best to ensure that potential power differentials between school, parents and researcher did not create any suggestion, even unconsciously, of obligation. For parents and teachers, informed consent forms were signed and these included the statement that participants had the right to withdraw at any time. For the children, I created a simple, colourful ‘form’ through which they were invited to give or withhold their consent to participation in the research by ticking either a happy face or a sad face respectively. Both verbally and in writing it was made clear to all participants that, even having given consent, they retained the right to withdraw from all assessments and observations at any time. For children with ID, the researcher and teachers need to be especially aware that pupils may give non-verbal indications that they wish to withdraw (Skånfors, 2009), given the power differentials that exist between adult researchers and child participants (perhaps especially those with ID) (Khoja, 2016).

The principle of care for others, of avoiding harm was an important consideration in my research design. For the teachers, it was important that I designed the intervention sessions to fit into their existing schedules so as to cause only minimal disruption. This was established through the pilot study. Also, I was aware that I should not waste the time of the control group participants, so I
designed and provided an intervention that I considered would be useful to all teachers of children with ID and this opinion was confirmed through pilot study feedback. Many teachers of children with DS struggle to manage classes due to CB (McFadden et al., 2017). For this reason, I provided an intervention that might help the teachers understand better how to evaluate and prevent CB and to support positive behaviour.

Regarding care of the pupils, I was aware of the importance of the relationship of trust between pupils and teachers, so I took steps to make sure that my presence in the classroom during observations disturbed the pupils as little as possible (Khoja, 2016). During the study I was introduced to the pupils when distributing consent forms, so they were already familiar with my presence in the school when I was actually conducting the study (Skånfors, 2009). The consent forms for children explained my presence in class, and the testing activities that were part of my study, as a kind of ‘play’.

All assessments and observations were scheduled so as not to create undue stress for both teachers and pupils. I carried out only one assessment per day for children, making sure that this did not disrupt their ordinary routines (play times in particular) and allowed enough time (two weeks) for teachers to complete their questionnaires.

Regarding confidentiality and data protection, there are both legal and ethical duties to consider. In order to maintain confidentiality, I assigned an identification number to each teacher and pupil, and used this on all written data collected. I asked the teachers’ permission to record the focus group and guaranteed that participants would not be identifiable in the written report of the research. In all report documents, pupils’ names are withheld in order to maintain confidentiality. I regularly updated a log that contained a record of any decisions made throughout the research process.
It was also important to be clear and transparent about data storage, retention and secure destruction. I explained to teachers and parents that all data would be securely stored and would be deleted 3 years after publication of my research, in accordance with the University of Edinburgh Data Protection Policy research ethical approval (for information and consent sheets see Appendix 2.1).

4.2.7 Data Analysis
The quantitative data were inputted into SPSS and checked for completion of data entry (Kinnear & Gray, 1999). The EG and CG pre-intervention data were then compared, as discussed in section 4.2.3.2 (participant sampling). Missing data are discussed in the results section.

The normality of the distribution of the scales used and the type of data involved were assessed to decide whether to use parametric or non-parametric tests. If data showed normal distrubtion, then parametric tests were used, which was advantageous, given that parametric tests have greater statistical power than non-parametric tests (Pallant, 2005). The specific tests used were determined by assumptions of inferential statistics, such as reliability, linearity, homogeneity of regression slopes and equality of variances (discussed in due course). Justification for the statistical approach taken for each scale/questionnaire is presented in the results section. Effect sizes were used given that whilst results may show a significant statistical difference, the effect sizes show the magnitude of effect between data sets, which can give greater confidence in my findings (Kraft, 2020).

To answer study two research question, RQ1a: Does the WM intervention programme aimed at teachers lead to changes in WM outcomes for pupils with
Down Syndrome?, non-parametric analyses were used, which required the use of the Friedman test and the Mann Whitney test.

To answer RQ1b: Does the WM intervention programme aimed at teachers lead to changes in executive function for pupils with Down Syndrome (parents’ and teachers’ BRIEF-P)? and RQ1c: Does the WM intervention programme aimed at teachers lead to changes in behavioural outcomes for pupils with Down Syndrome? (parents’ and teachers’ SDQ), ANCOVA tests were used.

To answer RQ1d: Does the WM intervention programme aimed at teachers lead to changes in literacy and numeracy outcomes for pupils with Down Syndrome? (students’ school records), Mann Whitney tests were used.

To answer RQ2: Does teachers’ use of WM strategies improve from pre- to post-intervention? (observations), the use of strategies by each teacher was counted within the observational data, to evaluate the most and the least commonly used strategies. This was analysed using paired sample t-tests, to explore the changes in use of each strategy from pre- to post-intervention.

To answer RQ3: Does teachers’ knowledge of WM and CB improve from pre- to post-intervention? (teachers’ questionnaire), both in the EG and CG, teacher knowledge of WM and CB in DS pre- and post-intervention, were analysed using paired sample t-tests and independent sample t-tests.

Regarding the qualitative data, RQ4: What are teachers’ viewpoints about the WM intervention in inclusive education in Kuwait?, the focus groups were recorded and the discussions transcribed in Arabic. The analysis was completed without the support of software, as the relatively small data set did not necessitate this. Some approaches to the analysis of focus group data are fully qualitative, such as the approach to thematic analysis propounded by Clarke
and Braun (2018); however, in my research, the focus group discussions were structured around a limited number of specific questions designed to elicit opinions and feelings regarding specific teaching strategies. This was predominantly to support and explain the quantitative data gathered, in a triangulated approach (Palomba & Banta, 1999). In addition, there was not a very large amount of data from each participant, thus the data did not lend itself well to a detailed thematic analysis (Braun & Clarke, 2006).

Rather, I divided the responses into five different areas that the focus group discussion was centred/structured around. These key areas were: 1) Impact on Teachers’ Knowledge and Confidence around WM; 2) Teachers’ Beliefs About Children’s Abilities; 3) Teachers’ WM Strategy Use; 4) Teachers’ Understanding of the Associations between Pupils’ WM and Behaviour; and 5) Teachers’ Views on the Feasibility of the Intervention and Suggestions for Future Development. The identification of specific categories to discuss in the focus groups meant that the key points from each category could be extracted from the data set, by noting down the most common or popular responses (including how many participants said something similar), as well as any other outlier responses (individuals that offered an opinion that may have differed to that of the majority) (Creswell & Poth, 2016). Patterns across the data set were also assessed, including whether there were differences in the responses of numeracy and literacy teachers, for instance. The transcripts were not translated from Arabic to English to stay as close to the original meanings in the data as possible, to avoid over interpretation (Al-Amer et al., 2016), although some quotes were translated into English to be presented in the thesis, in order to illustrate the findings to the reader.

4.3 Results
This section presents the statistical results and qualitative findings for the following aims, which correspond to the study two research questions as follows:
• To identify whether the working memory intervention programme aimed at teachers led to changes in working memory outcomes for pupils with Down syndrome (verbal and visual WM assessment) (RQ1a);
• To identify whether the working memory intervention programme aimed at teachers led to changes in executive function for pupils with Down syndrome (parents’ and teachers’ BRIEF-P) (RQ1b);
• To identify whether the working memory intervention programme led to changes in behavioural outcomes for pupils with Down syndrome (parents’ and teachers’ SDQ) (RQ1c);
• To identify whether the working memory intervention programme led to changes in literacy and numeracy outcomes for pupils with Down syndrome (students’ assessments in school records) (RQ1d);
• To identify whether teachers’ use of working memory strategies improved from pre- to post-intervention (observations) (RQ2);
• To identify whether teachers’ knowledge of WM improved from pre- to post-intervention (teachers’ questionnaire) (RQ3).
• To explore teachers’ perspectives of the WM intervention (focus groups) (RQ4).

4.3.1 Data Screening and Preliminary Handling
First the data were checked to identify missing data; it was found that there were seven responses missing on the BRIEF-P (3 from the EG and 4 from the CG, 3 for parents and 4 for teachers) items; this was addressed using the scale manual (BRIEF-P, manual). This advised that none of the subscales on which there were missing items should be excluded, as no respondent had missed more than two items (any more than two per subscale and that subscale must be excluded). Then the scale manuals were followed in order to group the items according to the different BRIEF-P subscales and, for the SDQ, to reverse the
scoring on negatively worded items and compute the assessment subscales within SPSS.

4.3.2 Results by Aim

Aim 1: To identify whether the working memory intervention programme aimed at teachers led to changes in working memory outcomes for pupils with Down syndrome (RQ1a).

This analysis sought to establish whether children in the EG improved from pre- to post-intervention in their WM scores, compared to the children in the CG. The WM assessment was divided into two subscales: verbal working memory (VWM) and visuo-spatial working memory (VSWM).

The means and standard deviations of the VWM and VSWM scores for the whole sample at pre-test, post-test and follow-up were computed (Table 4.7), and the normality of distribution of each subscale was checked using the Shapiro-Wilk test and by visual inspection of the frequency histograms. The Shapiro-Wilk tests were p<.05 for each subscale, indicating that they were significantly different from a normal distribution. This was confirmed by the histograms, which showed positive skewness, with the scores clustering at the left at the low values. This presents a problem with using a parametric ANOVA, which requires normally distributed data. A non-parametric equivalent, the Friedman test, was therefore appropriate (see Pallant, 2005, p.296). Unfortunately, this test is less effective than the parametric test that would have been used, namely the mixed between–within subjects ANOVA (Pallant, 2005, p.239). The latter would have allowed comparison of the two groups’ WM scores at the three time points, whereas the Friedman test only allows comparison at the three time points for the two groups separately.
<table>
<thead>
<tr>
<th>Variable</th>
<th>Experimental group (n=18) Mean (SD)</th>
<th>Mean rank</th>
<th>Control group (n=13) Mean (SD)</th>
<th>Mean rank</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Verbal working memory total:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>2.94 (1.66), 1.50</td>
<td></td>
<td>3.00 (2.24), 1.92</td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>3.44 (1.62), 1.75</td>
<td></td>
<td>2.92 (1.98), 1.77</td>
<td></td>
</tr>
<tr>
<td>follow-up</td>
<td>4.61 (1.79), 2.75</td>
<td></td>
<td>3.23 (2.17), 2.31</td>
<td></td>
</tr>
<tr>
<td><strong>Visuo-spatial working memory total:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>2.11 (1.64), 1.39</td>
<td></td>
<td>2.38 (1.80), 1.92</td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>2.61 (1.61), 1.75</td>
<td></td>
<td>2.46 (1.85), 2.04</td>
<td></td>
</tr>
<tr>
<td>follow-up</td>
<td>4.39 (2.03), 2.86</td>
<td></td>
<td>2.54 (1.94), 2.04</td>
<td></td>
</tr>
</tbody>
</table>

The improvement in WM scores in the EG and CG groups were analysed separately using the Friedman test. The mean ranks of each group on each test are shown in Table 4.7, as is the output from the Friedman test (shown as the 'mean rank').

There was a statistically significant improvement in both VWM and VSWM for the EG between pre-test and follow-up [p<.001 for each], but not for the CG [VWM p=.187 and for VSWM p=.905]. However, the test does not indicate between which pairs of time points the EG change was significant. For that, the improvements were calculated by subtracting the initial from the final scores and performing a Mann-Whitney test. The change scores and the mean ranks of these change scores, together with the significance levels of the differences between groups, are shown in Table 4.8.
Table 4.8 Working Memory Change Scores and Mean Ranks of Change Scores by Group

<table>
<thead>
<tr>
<th>Change Variable</th>
<th>Exptal group</th>
<th>Exptal group</th>
<th>Control group</th>
<th>Control group</th>
<th>Z</th>
<th>Sig</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Change score</td>
<td>Change score</td>
<td>Change score</td>
<td>Change score</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>(N=18)</td>
<td>Mean rank</td>
<td>(N=13)</td>
<td>Mean rank</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Verbal WM:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>post-intervention minus pre-intervention</td>
<td>0.05</td>
<td>17.81</td>
<td>-0.08</td>
<td>13.50</td>
<td>-1.411</td>
<td>.196</td>
</tr>
<tr>
<td>follow-up minus pre-intervention</td>
<td>1.67</td>
<td>19.92</td>
<td>0.23</td>
<td>10.58</td>
<td>-2.909</td>
<td>.004</td>
</tr>
<tr>
<td>Visuo-spatial WM:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>post-intervention minus pre-intervention</td>
<td>0.50</td>
<td>17.61</td>
<td>0.08</td>
<td>13.77</td>
<td>-1.305</td>
<td>.192</td>
</tr>
<tr>
<td>follow-up minus pre-intervention</td>
<td>2.28</td>
<td>21.00</td>
<td>0.16</td>
<td>9.08</td>
<td>-3.674</td>
<td>.000</td>
</tr>
</tbody>
</table>

Table 4.8 shows that there is significant difference between the groups in their VWM change scores between pre-test and follow-up (p=.004), and also in their VSWM change scores between pre-test and follow-up (p<.001). On the other hand, there is no significant difference between the groups in either VWM or VSWM scores between pre-test and post-test. In sum, the analysis shows that the EG improved significantly more than the CG in both dimensions of WM and that this improvement occurred between pre-test and follow-up, rather than between pre-test and post-test. The values of eta squared were .79 for VWM and .73 for VSWM, which indicated large effect sizes.
Aim 2: To identify whether the working memory intervention programme aimed at teachers led to changes in executive function for pupils with Down syndrome, according to parents’ and teachers’ reports (RQ1b).

*Parents’ BRIEF-P Results*

The parents’ pre- and follow-up means and standard deviations on BRIEF-P (global) and its subscales (inhibition, shift, emotional control, working memory, plan/organise) were compared in order to ascertain whether there had been change over the course of the intervention.

To test for the significance of any differences in outcomes, ANCOVA, was appropriate, because this is a two-group pre-test, post-test design (Pallant, 2005, p.263). In ANCOVA, the scores in the pre-test are treated as a covariate to control for pre-existing differences between the groups. ANCOVA tests for the significance of the difference in the means in the follow-up test of each group while controlling for the pre-test scores. It removes the influence of the pre-test scores and this increases the power of the test. In order to carry out ANCOVA, the data must meet certain assumptions: reliability, normality, linearity, homogeneity of regression slopes and equality of variances. Therefore, the analysis started by testing these assumptions.

Reliability tests were performed on the five BRIEF-P subscales and global executive function at pre-intervention and follow-up, as described in the methodology chapter. All but one of the Cronbach's alpha values were >.8 (the other was >.7), indicating a sufficiently high level of reliability (the reliability details of BRIEF-P subscales can be found in Appendix 2.4).

Testing for normality of distribution, the Shapiro-Wilk test, gave a non-significant result (p>.05) for each of the scales, indicating normal distributions. Linearity and homogeneity of regression slope requirements were also met. For all these
scales, the Levene’s test was $p > .05$, indicating equality of variances in the two groups, EG and CG. Therefore the assumptions of ANCOVA were met.

The parents’ pre- and follow-up means and standard deviations on BRIEF-P (global) and its subscales (inhibition, shift, emotional control, working memory, plan/organise) were compared (Table 4.9). A higher EF score indicates greater difficulties.
Table 4.9 Means and Standard Deviations of Parents’ Pre- and Follow-up BRIEF-P Scores (Global and Subscales) by Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>Experimental group (N=18) Mean (SD)</th>
<th>Control group (N=13) Mean (SD)</th>
<th>ANCOVA F-value</th>
<th>p-value</th>
<th>Effect size (Partial Eta squared)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Global EF:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>132.60 (24.05)</td>
<td>127.90 (21.70)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>122.50 (24.99)</td>
<td>127.80 (23.80)</td>
<td>(1,28)=8.94</td>
<td>.006</td>
<td>.242</td>
</tr>
<tr>
<td><strong>Inhibition:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>31.27 (5.88)</td>
<td>30.50 (6.64)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>29.72 (6.46)</td>
<td>30.46 (7.74)</td>
<td>(1,28)=1.74</td>
<td>.198</td>
<td>.059</td>
</tr>
<tr>
<td><strong>Shift:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>18.27 (5.34)</td>
<td>18.61 (5.41)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>18.22 (5.20)</td>
<td>17.46 (3.71)</td>
<td>(1,28)=1.130</td>
<td>.297</td>
<td>.039</td>
</tr>
<tr>
<td><strong>Emotional control:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>18.83 (4.60)</td>
<td>19.54 (3.95)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>17.72 (4.56)</td>
<td>18.92 (4.71)</td>
<td>(1,28)=.997</td>
<td>.327</td>
<td>.034</td>
</tr>
<tr>
<td><strong>WM:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>40.72 (8.40)</td>
<td>37.80 (7.40)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>35.67 (7.30)</td>
<td>39.40 (7.74)</td>
<td>(1,28)=16.855</td>
<td>.000</td>
<td>.376</td>
</tr>
<tr>
<td><strong>Plan/organise:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>23.00 (4.90)</td>
<td>21.50 (4.62)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>21.22 (5.30)</td>
<td>21.54 (4.66)</td>
<td>(1,28)=3.070</td>
<td>.091</td>
<td>.099</td>
</tr>
</tbody>
</table>

From Table 4.9, it can be seen that there were significant differences between pre-intervention and follow-up scores on one of the EF subscales (working memory) and on the global EF scale. None of the other subscales showed a
significant difference between pre- and follow-up. On WM, the EG showed an improvement, unlike the CG. The effect size, as given by partial eta squared, was .376, which is considered large (see Pallant, 2005, p. 201). Similarly, on the global EF score, the EG showed an improvement but the CG did not, and here the effect size was also large (partial eta squared = .242).

**Teachers’ BRIEF-P Results**

The same procedures were followed for the teachers’ reports on the BRIEF-P variables as for the parents’ reports. The scales were tested for the assumptions of ANCOVA, followed by performing the ANCOVA tests to assess whether the follow-up measures differed significantly by group once the pre-intervention measures were controlled. Reliability tests showed values of Cronbach’s alpha of greater than .7 on all the scales. All scales had a normal distribution (p>.05 in all Shapiro-Wilk tests). To test for linearity, the relationship between the dependent variable (global EF follow-up score) and the covariate (global EF pre-test score) was inspected by a scatterplot and line of best fit, which confirmed linearity. When graphs for the two groups were separated, the slopes were seen to be similar, confirming the homogeneity of regression slopes. Levene’s test showed equality of variances on the global scale and on each subscale.

Having confirmed the assumptions of the teachers’ BRIEF-P scales, the teachers’ pre- and follow-up means and standard deviations on BRIEF-P (global) and its subscales (inhibition, shift, emotional control, working memory, plan/organise) were compared (Table 4.10).

From Table 4.10, it can be seen that the only significant difference between the groups occurred between pre- and follow-up scores on WM; in the teachers’ view, the EG children improved their WM more than did the CG children. The effect size, as given by partial eta squared, was large.
Table 4.10 Means and Standard Deviations of Teachers’ Pre- and Follow-up BRIEF-P Scores (Global and Subscales) by Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>Experimental group (N=18) Mean (SD)</th>
<th>Control group (N=13) Mean (SD)</th>
<th>ANCOVA F-value</th>
<th>p-value</th>
<th>Effect size (Partial Eta squared)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Global EF:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>122.83 (27.21)</td>
<td>132.53 (21.50)</td>
<td>(1,28)=2.680</td>
<td>.113</td>
<td>.087</td>
</tr>
<tr>
<td>Follow-up</td>
<td>122.72 (23.30)</td>
<td>125.40 (16.80)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Inhibition:</td>
<td></td>
<td></td>
<td>(1,28)=.872</td>
<td>.358</td>
<td>.030</td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>29.67 (7.62)</td>
<td>32.50 (6.60)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>27.94 (6.42)</td>
<td>29.07 (4.90)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Shift:</td>
<td></td>
<td></td>
<td>(1,28)=.315</td>
<td>.579</td>
<td>.011</td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>18.00 (5.92)</td>
<td>18.10 (4.30)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>16.94 (5.20)</td>
<td>17.40 (4.21)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Emotional control:</td>
<td></td>
<td></td>
<td>(1,28)=.001</td>
<td>.973</td>
<td>.000</td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>19.05 (5.66)</td>
<td>19.92 (3.97)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>18.17 (5.04)</td>
<td>18.92 (4.00)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>WM:</td>
<td></td>
<td></td>
<td>(1,28)=14.863</td>
<td><strong>.001</strong></td>
<td>.347</td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>36.05 (8.20)</td>
<td>39.53 (8.23)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow up</td>
<td>30.61 (6.70)</td>
<td>38.70 (6.80)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Plan/organize:</td>
<td></td>
<td></td>
<td>(1,28)=.328</td>
<td>.571</td>
<td>.012</td>
</tr>
<tr>
<td>Pre-intervention</td>
<td>20.05 (4.70)</td>
<td>22.53 (4.23)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>19.05 (4.05)</td>
<td>21.30 (3.61)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Aim 3: To identify whether the working memory intervention programme aimed at teachers led to changes in behavioural outcomes for pupils with Down Syndrome (parents’ and teachers’ SDQ) (RQ1c).

ANCOVA assumptions were checked. As described in the methodology section, the 20 SDQ total difficulties items and 5 prosocial items on the parents’ and teachers’ questionnaire were tested for internal consistency and values of Cronbach’s alpha of greater than .7 were obtained. The normality of the scales was ascertained by the Shapiro-Wilk test, which give non-significant results for the parent and teacher total difficulties and the teachers prosocial behaviour scales at both pre-intervention and follow-up. However, for the parents’ pre-intervention and follow-up prosocial scales the Shapiro-Wilk test gave a significant result while the Kolmogorov-Smirnov test gave a non-significant result. Thus, most of the scales were normally distributed, although there was some ambiguity with the parent prosocial scales. Nevertheless, it was felt that, overall, the assumptions of ANCOVA had been adequately met. ANCOVA was therefore used to test for the significance of the difference between the groups in the follow-up test means while controlling for the pre-intervention scores.

The means and standard deviations on the parents’ and teachers’ pre-test and follow-up SDQ total difficulties and prosocial scales were then computed (Table 4.11).
Table 4.11 Means and Standard Deviations on Pre- and Follow-up Parent and Teacher SDQ Total Difficulties and Prosocial Scales by Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>Experimental group (N=18) Mean (SD)</th>
<th>Control group (N=12&lt;sup&gt;a&lt;/sup&gt;) Mean (SD)</th>
<th>ANCOVA F-value</th>
<th>p-value</th>
<th>Effect size (Partial Eta squared)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Parent SDQ Total Difficulties:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>17.06 (4.44)</td>
<td>16.58 (3.50)</td>
<td>(1,27)=7.825</td>
<td>.009</td>
<td>.225</td>
</tr>
<tr>
<td>follow-up</td>
<td>16.56 (5.53)</td>
<td>13.50 (4.12)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parent Prosocial:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>5.11 (2.60)</td>
<td>7.00 (3.02)</td>
<td>(1,27)=3.988</td>
<td>.056</td>
<td>.125</td>
</tr>
<tr>
<td>follow-up</td>
<td>5.17 (2.57)</td>
<td>7.70 (2.60)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Teacher SDQ Total Difficulties:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>12.28 (4.79)</td>
<td>14.31 (5.14)</td>
<td>(1,27)=4.478</td>
<td>.043</td>
<td>.138</td>
</tr>
<tr>
<td>follow-up</td>
<td>13.67 (4.76)</td>
<td>13.15 (5.40)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Teacher Prosocial:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>4.33 (2.11)</td>
<td>5.23 (2.69)</td>
<td>(1,27)=5.23</td>
<td>.030</td>
<td>.158</td>
</tr>
<tr>
<td>follow-up</td>
<td>4.77 (2.01)</td>
<td>6.53 (2.56)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Note: <sup>a</sup> There were only 12 because case 24 in CG was an outlier and was therefore removed.

The significance level of .044 for Levene’s test for parents’ follow-up SDQ total difficulties indicated that the assumption of equality of variances has been violated. According to Pallant (2005, p.234), because the variance in SDQ total difficulties parent follow-up is different between the two groups, a more stringent significance level should be set for the ANCOVA. Therefore, a significance level of .01 (instead of .05) was adopted in the following analysis of parent data (this was not the case for the teacher data).
Nevertheless, the difference between groups was statistically significant (p=.009) and the effect size was large. Parents of CG children saw fewer total difficulties, compared to the EG children, following the intervention than before it began (with the proviso that case 24 had been eliminated from the analysis). There was no significant difference between groups on the parents’ prosocial scale.

Regarding the teachers’ view, there were significant differences between the groups at the p=.05 level on SDQ total difficulties and prosocial scales in their pre-intervention and follow-up scores. The effect sizes are classified as moderate for the SDQ total difficulties and large for the prosocial scale. These differences were in favour of the CG children, supporting the greater effectiveness of the positive behaviour support intervention.

Aim 4: To identify whether the working memory intervention programme aimed at teachers led to changes in literacy and numeracy outcomes for pupils with Down syndrome (students’ school records) (RQ1d).

Literacy and numeracy scores were collected from students’ records in school and these scores contained the marks of examinations plus class activities, such as verbal participation in answering teachers’ questions or writing on the board. To achieve aim 4, the mean scores on literacy and numeracy for each group, before and after the intervention were computed and compared (Table 4.12).
Table 4.12 Literacy and Numeracy Scores Pre- and Post-intervention by Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>Experimental group (N=18) Mean (SD)</th>
<th>Control group (N=13) Mean (SD)</th>
<th>Mann-Whitney Z-value</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Literacy:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>13.06 (5.64)</td>
<td>14.15 (5.86)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>14.94 (5.38)</td>
<td>16.31 (2.90)</td>
<td>-.871</td>
<td>.384</td>
</tr>
<tr>
<td><strong>Numeracy:</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>14.11 (5.46)</td>
<td>14.31 (5.98)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>17.44 (4.78)</td>
<td>17.38 (2.40)</td>
<td>-1.119</td>
<td>.263</td>
</tr>
</tbody>
</table>

From Table 4.12, it appears that both the EG and CG improved in literacy and numeracy. Improvements were then calculated by subtracting the initial from the final scores to obtain ‘change’ scores. Since the data did not meet the requirements of ANCOVA, a Mann-Witney test was used to ascertain whether differences in improvement between groups was statistically significant. However, the Mann-Whitney test showed that these differences between groups in the literacy and numeracy change scores were not statistically significant.

**Aim 5:** To identify whether teachers' use of WM strategies improved from pre- to post-intervention (RQ2).

To obtain data on teachers' use of WM strategies, the EG teachers were observed before and after the intervention and the observed frequency of use of the nine strategies during the 20-minute observation was recorded. The CG teachers did not participate in this part of the research.

The mean number of times the 14 EG teachers were observed to use each of the nine WM strategies included in the training, both before and then after the
intervention, is recorded in Table 4.13. These means were then added to give totals for the frequency of use of the strategies.

Table 4.13 shows that use of all strategies increased from pre-intervention to post-intervention. The mean total frequency of use increased from 10.71 pre-intervention to 19.93 post-intervention. Strategy 8 (evaluating working memory load) was not used at all before the intervention. The greatest improvements were in use of strategy 1 (responding to child’s WM difficulties), strategy 7 (encouraging student to use working memory strategies) and strategy 9 (reducing WM load). These figures are shown in graphical form in Figure 4.1.
Table 4.13 Teachers’ Use of Working Memory Strategies

<table>
<thead>
<tr>
<th>Strategy</th>
<th>Mean (SD)</th>
<th>t</th>
<th>p</th>
<th>Eta squared</th>
</tr>
</thead>
<tbody>
<tr>
<td>1- Responding to child's WM difficulties:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>0.21 (0.43)</td>
<td>-7.80</td>
<td>.000</td>
<td>0.82</td>
</tr>
<tr>
<td>post-intervention</td>
<td>2.36 (0.93)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2- Reducing distraction:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>1.43 (0.51)</td>
<td>-2.88</td>
<td>.013</td>
<td>0.39</td>
</tr>
<tr>
<td>post-intervention</td>
<td>2.36 (1.08)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3- Repetition:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>3.07 (1.07)</td>
<td>-2.19</td>
<td>.047</td>
<td>0.22</td>
</tr>
<tr>
<td>post-intervention</td>
<td>3.57 (1.28)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4- Rhythm and rhyme:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>1.29 (0.82)</td>
<td>-3.68</td>
<td>.003</td>
<td>0.51</td>
</tr>
<tr>
<td>post-intervention</td>
<td>2.00 (1.11)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5- Memory aids:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>3.93 (1.07)</td>
<td>-2.88</td>
<td>.013</td>
<td>0.39</td>
</tr>
<tr>
<td>post-intervention</td>
<td>4.86 (0.86)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6- Chunking:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>0.36 (0.50)</td>
<td>-2.19</td>
<td>.047</td>
<td>0.22</td>
</tr>
<tr>
<td>post-intervention</td>
<td>0.86 (0.66)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7- Encouraging student to use working memory strategies:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>0.29 (0.61)</td>
<td>-4.18</td>
<td>.001</td>
<td>0.57</td>
</tr>
<tr>
<td>post-intervention</td>
<td>1.64 (1.22)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8- Evaluating working memory load:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>0.00 (0.00)</td>
<td>-8.00</td>
<td>.000</td>
<td>0.83</td>
</tr>
<tr>
<td>post-intervention</td>
<td>1.14 (0.53)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9- Reducing WM load:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>0.14 (0.36)</td>
<td>-4.77</td>
<td>.000</td>
<td>0.64</td>
</tr>
<tr>
<td>post-intervention</td>
<td>1.14 (0.66)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10- Total strategies:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>1 0.71 (2.33)</td>
<td>-8.78</td>
<td>.000</td>
<td>0.86</td>
</tr>
<tr>
<td>post-intervention</td>
<td>19.93 (3.41)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
To test for the normality of distribution of the pre- and the post-intervention strategy counts, the Shapiro-Wilk test was used and gave non-significant results, indicating normality. Paired samples t-tests were therefore used to test for the significance of the differences between the pre- and post-intervention frequencies. The effect sizes (eta squared) were also calculated. From Table 4.13 it can be seen that all pre/post differences in use of strategies were statistically significant and all of the values of eta squared indicated large effect sizes. Therefore, the intervention was successful in increasing the teachers’ use of WM strategies.

Aim 6: To identify whether teachers’ WM or CB knowledge improved from pre- to post-intervention (RQ3).
The teachers’ questionnaires were divided into two sections, working memory (WM) knowledge and challenging behaviour (CB) knowledge, as explained in chapter four. The total scores were found to be normally distributed on the Shapiro-Wilk test. The means and standard deviations of the teachers’ scores on the WM and CB knowledge items are given in Table 4.14.

Table 4.14 Means and Standard Deviations of Teachers’ Knowledge Scores on Working Memory and Challenging Behaviour Items by Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>Exptal group (N=14) Mean (SD)</th>
<th>t (df=13)</th>
<th>p</th>
<th>Eta sq</th>
<th>Contrl group (N=14) Mean (SD)</th>
<th>t (df=13)</th>
<th>p</th>
<th>Eta sq</th>
</tr>
</thead>
<tbody>
<tr>
<td>WM</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>knowledge:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>32.29 (8.43)</td>
<td>-10.66</td>
<td>.000</td>
<td>.141</td>
<td>35.57 (10.62)</td>
<td>.000</td>
<td>.814</td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>63.50 (5.56)</td>
<td>1.25</td>
<td>.234</td>
<td>.057</td>
<td>34.71 (10.64)</td>
<td>1.25</td>
<td>.234</td>
<td></td>
</tr>
<tr>
<td>CB</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>knowledge:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pre-intervention</td>
<td>26.64 (7.50)</td>
<td>2.60</td>
<td>.012</td>
<td>.207</td>
<td>26.71 (4.91)</td>
<td>2.60</td>
<td>.012</td>
<td></td>
</tr>
<tr>
<td>post-intervention</td>
<td>29.00 (6.26)</td>
<td>10.51</td>
<td>.000</td>
<td>.810</td>
<td>38.36 (2.06)</td>
<td>10.51</td>
<td>.000</td>
<td></td>
</tr>
</tbody>
</table>

From Table 4.14, it is clear that WM knowledge scores of the EG group increased by a large amount, whereas this was not the case in the CG group. To test whether the improvement in the WM scores was statistically significantly greater for the EG group than the CG group, an ANCOVA test was considered. However, scatter plots indicated a lack of linearity and homogeneity of regression slopes in the EG group. Therefore, since the assumptions for ANCOVA were not met, a paired sample t-test was used in each group separately instead, with the results shown in Table 4.14. The results of the paired samples t-test showed that for the EG there was a statistically significant increase in WM knowledge from pre- to post-intervention with a very large effect.
size, while for the CG there was no statistically significant change in WM knowledge from pre- to post-intervention.

For the CB items, there was a statistically significant increase in CB scores for the EG between pre- and post-intervention with a large effect size. For the CG, there was a larger improvement in CB knowledge scores between pre- and post-intervention, which was also statistically significant, with a very large effect size. Therefore, while both groups of teachers improved their knowledge of CB over the course of the intervention, the effect appeared to be greater for the CG.

The previous analysis looked at the groups of teachers separately before and after the intervention and found different amounts of change. A further analysis did not compare ‘before and after’ but compared the two groups on the same tests at the same points in time. To find whether the differences between EG and CG in WM and CB items were statistically significant, independent samples t-tests were conducted.

The analysis showed that there was no significant difference on pre-intervention WM knowledge between the EG and CG \[t(26)=.907, p=.373, \text{Eta squared}=.030\], indicating that the two groups were at the same level on their WM knowledge before the intervention. However, there was a significant difference between the groups on WM items post-intervention \[t(26)=8.975, p<.001, \text{Eta squared}=.756\]. Thus, after the intervention, the EG scored significantly higher than the CG on WM; this is a very large effect size.

There was no significant difference between groups on CB knowledge items pre-intervention \[t (26)=-0.030, p=0.976, \text{Eta squared}=.00003\]; so the two groups started at the same level in CB knowledge before the intervention. However, there was a significant difference between the groups on CB items
post-intervention \[t(26)=-5.310, \ p<.001 \ \text{Eta squared}=0.520\], indicating that the CG scored significantly higher than the EG on CB, with a large effect size.

The two analyses together show that there was significant improvement in teachers’ knowledge of WM for the EG and of CB for the CG between pre- and post-intervention. For further explanation, paired samples t-test were used on each item individually to ascertain on which items the scores had improved statistically and on which they had not, in both the WM subscale and CB subscale, for each group (see Appendix 3.1).

Aim 7: To explore teachers’ perspectives of the WM intervention (focus groups) (RQ4).

The aim of this focus group was to gain the teachers’ perspectives on the WM intervention, to support the interpretation of their responses to both the questionnaire (assessing their pre- and post-intervention knowledge of WM) and the systematic observation (assessing their pre- and post-intervention use of WM strategies). Because this aim was achieved using qualitative data, the results were analysed and presented in a slightly different way; as mentioned earlier, the aim was to gain a deeper understanding of the teachers’ viewpoints regarding learning about and implementing the WM intervention.

The teachers in the focus group discussed the organisation of the intervention and its overall impact on their understanding of DS and on their methods for dealing with children with DS in the classroom. The teachers were asked to focus on WM strategies and their perceptions of the effect they had on pupils’ academic achievement. They also discussed what effects the intervention had on themselves, specifically on their understanding of WM and their attitudes towards children with DS. The teachers also provided their own suggestions for future interventions, given their status as experts on teaching children with DS.
This section provides a summary of the key points made in the focus group, grouped together under five salient categories. Analysis of the focus group discussion revealed five areas in which the teachers’ perspectives were particularly helpful in understanding reasons for the effectiveness of the intervention: 1) Impact on Teachers’ Knowledge and Confidence around WM; 2) Teachers’ Beliefs About Children’s Abilities; 3) Teachers’ WM Strategy Use; 4) Teachers’ Understanding of the Associations between Pupils’ WM and Behaviour; 5) Teachers’ Views on the Feasibility of the Intervention and Suggestions for Future Development.

1. Impact on Teachers’ Knowledge and Confidence

The teachers agreed that a significant advantage of this intervention was that they had expanded their knowledge of cognitive abilities, especially with regard to WM in children with DS.

The teachers added that it also increased their knowledge of the intangible cognitive abilities of children with DS, as they became more familiar with the signs pointing to WM difficulties, which could then lead to low academic achievement. The teachers also felt that that this intervention helped them become more confident when discussing children with DS with specialists or experts, as they learnt scientific terminologies, such as WM load, signs of weak WM and reducing distractions. One literacy teacher for example commented:

*Previously I knew nothing about the complex terminologies, but now I have some understanding and I am more confident about being able to discuss this issue with experts.*
A numeracy teacher also commented:

*This is the first time I’ve ever heard of memory load; previously I only knew about this in terms of smart devices, but it is interesting that the brain and technological devices work through similar concepts, when I reduce my device load it works more effectively – the same with the brain.*

These quotes illustrate how all teachers felt they had increased their knowledge and confidence of using WM strategies.

2. Teachers’ Beliefs about Children’s Abilities

There were mixed opinions prior to the intervention regarding how far the teachers believed that WM could be improved in children with DS. Some teachers asserted that they did not believe that children with DS would be able to implement the proposed strategies within the intervention. They also implied that they did not believe that the children’s WM could be improved, as they believed that weaknesses in WM formed a key characteristic of DS. Thus, some teachers frequently referred to the idea that WM deficits just needed to be ‘coped with’ as far as possible, and that their role was to teach their pupils whatever they could, with the strong belief that their current curriculum was suitable for children with DS. These teachers assumed that the curriculum may include tasks that were too difficult for some children with DS, who might fail in certain areas, and that this was, unfortunately, the way it was, because the curriculum had to cater for individual differences. However, 4 teachers – notably those with a long history of teaching children with ID (ranging from 17-37 years) – explained that they believed they could improve a child’s ability in any domain, as long as they used suitable strategies repeatedly until the child understood. Once the child understood, they would move on and teach the child something new.
After the intervention, all teachers reported that they also changed their views of how children with DS learn, especially when they witnessed children with DS could conduct some strategies by themselves in the class that the teachers previously assumed would be too hard for these children. The teachers therefore changed their attitudes and teaching strategies by focusing on strategies that aimed to develop the child’s own strategies to support their WM. The teachers suggested they became more positive about what children with DS can achieve in future as a result. One numeracy teacher for instance commented:

*When you said that children with DS can use their own strategies I laughed to myself, and I thought researchers always judge children by studies they read but not by actual time spent with them, as teachers do. However, after you explained the strategy for note taking by stickers, I tried this strategy just to show you that sometimes academic recommendations differ from realistic practice, and I was shocked that my student could use stickers much easier than I thought. That really alerted me as to how much I underestimate their abilities.*

Therefore, most teachers said that before the intervention, they believed that it was impossible for DS children to use some of the WM strategies to improve their WM; however, after the intervention, they had changed their minds. The strategies that teachers were most sceptical of initially included those that were to be conducted by the children themselves, including requesting help, asking for repetition, note-taking and rehearsing information. The teachers (and children) did not implement these strategies before the intervention, as they were unaware of them being potentially useful, and some teachers thought they would be ineffective or too difficult for the students to enact. The majority of teachers said that during the first intervention training session they found it difficult to believe that DS children could use certain strategies to support their WM unassisted. However, after the teachers taught these strategies within the classroom, they found them to be effective, although they said in the focus group that the strategies had different levels of difficulty. For example, the
teachers all commented that teaching children to ask for help or for repetition was the most difficult strategy to implement, given that many of the children had verbal difficulties.

Moreover, all the teachers commented that they thought the note-taking strategy would be impossible before the intervention, given that they spent a great deal of time and effort trying to teach each child a specific word and helping them to write it. Therefore, the idea of the children taking notes independently was seemingly impossible, given their lack of training and skill in this area. The teachers also said they had never heard of children with DS taking notes during lessons. Interestingly, one numeracy teacher stated she was ‘not convinced that this strategy could work’, so she decided to test it to show me that it was impossible. After using this strategy, however, this teacher (along with all seven others in this particular group of eight in the girls’ school) asserted that they were shocked by their students’ ability to use it. They said that using stickers in a book was the most effective note-taking strategy if drawing or writing was difficult. By comparison, the teachers found the rehearsing strategy was the easiest method for the children when they were creating their own strategies to support their WM. All the teachers said that this strategy was effective and useful to all children, even those with verbal difficulties.

Therefore, as a result of the intervention, the teachers had changed their attitudes towards and beliefs about the children’s learning methods and therefore the teaching strategies they used with the children, by reducing WM load, after they recognised signs of WM overload. They also stated that they found this, in turn, improved learning, as did their altered strategy use in general, discussed presently.
3. Teachers’ Strategy Use

All the teachers stated that they already used some of the intervention strategies in their teaching, such as repetition, rhythm, rhyme and other WM supporting material, without realising that these strategies could improve WM.

For example, a literacy teacher commented:

> Since studying in the college of education, I learnt that repetition is the fundamental teaching strategy for all subjects, not just for literacy, as this helps students save the information, but I did not know that this could also be a working memory strategy that could improve the memory more generally.

Another (numeracy) teacher expressed that:

> It was always important to present creative support materials every time when explained any lesson, but I thought this was just to present the lesson as more interesting than in the pupils' books or to attract student attention during the teaching, but … I never thought this was one of the strategies that could improve mental skills as a memory aids.

The teachers said that before the intervention they used different teaching strategies randomly; however, after the intervention, they created a plan, using the most effective WM strategies for each child and calculating the most suitable time in the day to use them. There was wide agreement that the morning was the most effective time to conduct/teach WM strategies, as their experience was that after 11am, the children began to lose their concentration.

One of the strategies that teachers indicated that they felt they had a greater understanding of was reducing distraction (which was used to reduce WM load). To illustrate, before the intervention, the teachers thought that the more supportive materials they presented, the more effective the teaching/learning methods were for the child. However, after the intervention, they recognised that
too many supportive materials, such as pictures or stickers, may serve to distract the student from the key learning. A numeracy teacher commented for example

*Before the intervention, my understanding was that a lot of supporting materials meant a clearer presentation for the lesson, so I always chose shiny coloured cards with lovable cartoon characters. But I found the children were more attracted to these characters rather than the information. When I presented supportive materials but with simpler cards, such as a white coloured card without as much detail, I found the children were more attracted to the information on the cards, not to the other things.*

Therefore, the teachers often changed their teaching strategies to involve fewer or less distracting supporting materials. It became clear in the FG that the teachers’ understanding of WM and WM strategies had effectively changed over the course of the intervention, with most teachers (except, perhaps, the most experienced teachers) initially perceiving the WM strategies to be ineffective, until they witnessed first-hand how effective they could be. Moreover, the strong belief that the curriculum for children with DS was appropriate remained; the teachers that initially felt that some children with DS would simply ‘fail’ in some tasks/learning goals however changed. They all felt that as a result of the greater learning potential in their students, that the current curriculum could be accessed more readily by more students.

4. *Teachers’ Understanding of the Associations between Pupils’ WM and Behaviour*

Most teachers commented that as a result of the intervention, they increased their knowledge of DS behaviour, as they became more familiar with the signs of WM difficulties, which could then lead to challenging behaviour (CB). The teachers reported that they changed their ways of dealing with the children, for example, by reducing WM load, which, in turn, reduced CB. After the
intervention, all the teachers recognised that difficult academic tasks could lead to CB; these difficulties could be due to the fact that EFs or WM are weak/limited and the children demonstrated inappropriate behaviour in an attempt to avoid class activities. Therefore, many teachers reported that this new understanding led to a change in the way they dealt with the children. Previously, the teachers stated they often thought that CB was due to stubbornness, defiance or due to the child’s personality or environment, without realising CB could stem from WM weaknesses or overload. One numeracy teacher stated:

*I couldn’t imagine that challenging behaviour might be due to the fact an academic task might be too difficult until I decided to examine this myself, so I asked one child who always demonstrated challenging behaviour to do a Mathematics task with three steps. He started fighting with his classmate and I thought that might be because he wanted to distract my attention from this task. So I decided to wait for a while and ask him again to do the mathematic task with one step only. He did this easily and he was polite for the rest of the class.*

5. Teachers’ Views on the Feasibility of the Intervention and Suggestions for Future Development

All 14 teachers from the EG in both schools (one girls’ school and one boys’ school) agreed that this intervention was well organised, flexible, suited their timetables and fitted in with their teaching obligations, giving them sufficient time between sessions to implement the suggestions they received. They commented that the driving questions, workshop activities and further reading were very useful and gave them a better understanding of the subject.

Some teachers wished the researcher could observe them in their classroom after each session to ensure they were implementing the strategies correctly, and to enable them to discuss any limitations/difficulties they had immediately (so they did not forget). One literacy teacher commented:

*When I conducted any strategy for the first time, I tried to make sure that I was conducting it in the right way so I had to go to my notebook and check*
if I am doing it in the right way or not; actually, this made me less confident in the beginning of each strategy until the next session a week later, which allowed me to discuss what I did in the class with you. I was afraid sometimes that I might forget what I should discuss with you, but the driving questions in the beginning of each session were so helpful.

All teachers without any exception agreed with her. This indicates that the way the intervention was set up – including driving questions at the beginning – cemented their strategy use and enabled them to gain greater confidence in using these strategies in the classroom, through discussing their concerns and anxieties with me at the beginning of each session and gaining reassurance. Yet, it might be helpful to provide support between each session in the form of observation and personal one-on-one meetings and mentorship.

Moreover, one teacher introduced the idea of conducting the intervention over the whole school year, as she found teaching children one new strategy a week too difficult given that she believed each strategy needed time to be learnt properly, by both students and teachers. Some teachers also suggested that the WM intervention could be conducted alongside behaviour management strategies to reduce CB in the classroom, which limited their ability to teach the children new strategies.

**Conclusion for the Focus Group Discussion**

In conclusion, the focus group revealed that all teachers believed this intervention improved their knowledge of cognitive abilities in general and of the WM of children with DS in particular. The intervention therefore positively affected the teachers’ knowledge and confidence surrounding WM and WM strategy use. Moreover, the teachers’ beliefs in and attitudes towards the ability of their pupils to improve their WM and use the strategies in the classroom were changed; as a result of the intervention, all teachers agreed that most of the WM strategies were not only effective for the children, but they increased the teachers’ expectations of their pupils, given their initial surprise at the unexpectedly positive adoption of the strategies by their students. Additionally,
most teachers stated that they changed their opinion on why most children with DS behaved inappropriately or demonstrated CB, realising that it could be due to the difficulty of certain tasks that needed a high level of WM. The WM strategies led the teachers to feel confident that more students could master more of the learning goals set by the curriculum (which, necessarily, cannot take all individual differences into account).

Thus, the WM intervention was positively received by all teachers, perceiving it to give them the practical knowledge and strategies to help children with DS improve their WM. They also highlighted that it gave them a more positive attitude towards the children and their learning abilities.

4.4 Discussion

4.4.1 Introduction

This study set out to design, implement and evaluate the effectiveness of a teacher-focused WM intervention with children with DS across four schools in Kuwait. The key findings were that verbal and visual WM in the EG improved significantly on the Lanfranchi measures from pre-intervention to follow-up; moreover, the WM scores given by both parents and teachers on the WM subscale in the BRIEF-P also improved significantly from pre-intervention to follow-up. Furthermore, teachers’ knowledge about and use of WM strategies improved significantly in the EG from pre- to post-intervention. This also related to the changes reported in the FG in the teachers’ understanding about WM and attitude towards children with DS and learning methods, as well as their understanding of the effect that a weak WM has on children’s behaviour.

There was also a significant change according to parents on EF more broadly, according to the BRIEF-P, which indicates that these changes were generalised to all/more EFs. However, there were no differences between the EG and CG in literacy and numeracy scores following the intervention. There was a significant
reduction in total behaviour difficulties in the CG not the EG, in the view of both teachers and parents; teachers also reported a significant increase in prosocial behaviour, indicating the effectiveness of the control intervention in the CG.

These results will now be discussed in relation to the literature in this field, as well as considering potential explanations for these findings, limitations, and implications for practice.

4.4.2 Discussion of Results
The results reveal that the EG improved significantly more than the CG in both dimensions (verbal and visual) of WM; this improvement occurred between pre-test and follow-up rather than between pre-test and post-test. Moreover, the improvement in WM was evident across both parent and teacher viewpoints on the BRIEF-P. The parents of EG children gave lower scores (and therefore reported fewer difficulties) in the follow-up BRIEF-P WM subscale than the parents of CG children. The teachers of EG children reported fewer difficulties on the WM subscale at follow-up than teachers of CG children, once the pre-intervention scores were controlled for.

This shows that WM in children with DS can improve as a result of a teacher-directed intervention. This parallels findings from Pulina et al. (2015), who also taught interventions to teachers and parents to use WM interventions with DS children. It also supports findings from Costa et al. (2015), who found WM in children with DS improved following a WM intervention produced by teachers involving classroom games, which had a similar duration to the present research (6 weeks), as well as a similar WM assessment (a verbal and visuo-spatial task devised by Lanfranchi et al., 2004). The results of the present study also support findings from Orsolini et al. (2018), who demonstrated the effectiveness of WM interventions via the direct training of teachers, which enhanced the WM
performance of children with mild to borderline ID. The present study also aligns with Laws et al. (1996), whose study is the most similar to this one; it showed that training teachers in specific WM strategies (such as rehearsal) was effective in improving the WM of children with DS (especially memory span).

However, whilst there are similarities with previous findings, the present study is novel given that it is the first to conduct such an intervention with children with DS specifically in the Kuwait context, a country where there is a great deal of support (in terms of finances and resources) for pedagogical research and practice. Moreover, it is unique in that, whilst measuring quantitative WM (and CB) differences, it also captures the qualitative perspectives of teachers regarding their experiences throughout the intervention. This has usefully illuminated how far these teachers at least underestimated the abilities of children with DS to adopt strategies to improve their WM within the classroom context.

Improving WM in individuals with DS using specific strategies is supported in the literature more generally. For instance, when Comblain (1994) exposed 8 individuals with DS to intensive rehearsal training for eight weeks (EG) and compared them to a CG, the EG showed significant, immediate improvements in their memory span and clear signs of systematic rehearsal; however, at six-week and six-month follow-ups, the EG no longer demonstrated rehearsal and their memory performance had fallen significantly (although it was still significantly higher than at the start of the study), indicating these benefits were not sustained. Since the present study did not conduct a longitudinal follow-up, it is also unclear if the participants in the EG will sustain any long-term improvement from the WM interventions. Much of this may depend on the teachers’ use of the WM strategies with their students from the end of the intervention onwards.
In the present study, the WM improvement was only significant between pre-intervention and follow-up (1 weeks before and 4 weeks after the intervention); it was not significant between pre- and post-intervention. One possible reason for this is the fact that the intervention was taught to teachers, not directly to the children with DS by the researcher. Perhaps the teachers needed more time to understand and practice the WM strategies they were taught before they could effectively teach/use them with the children. Another possible reason is that the children may have improved through practice on the Lanfranchi assessments, thereby demonstrating a practice effect. However, whilst this is a possibility, these practice effects were not seen in the CG, which supports the interpretation that there was a genuine improvement in WM in the EG.

Another reason for this lack of immediate improvement from pre- to post-intervention could be that the teachers were not sufficiently convinced/persuaded of the effectiveness of the strategies at the beginning of the intervention to want to change their teaching style. There was no quality control that guaranteed the teachers would use the WM intervention exactly as taught to them in the intervention sessions; moreover, some teachers said that they initially demonstrated some hesitation in using some strategies, suggesting they may not have perceived them to be initially useful/beneficial. However, a number of teachers were found to change their minds regarding the benefits of the intervention, as revealed in the focus group. This change in attitude might also relate to teacher confidence; it appears that teacher confidence increased during the course of the intervention, supported to some extent by the teacher questionnaire findings that reveal teachers’ confidence in their knowledge increased, as well as confidence in the pupils’ abilities. The focus groups revealed that teachers believed the WM intervention could be even more effective if it was conducted under researcher supervision to correct any mistakes they made. Research reveals that teacher involvement, engagement, confidence and expectations can play a pivotal role in educational outcomes.
(e.g., Florian & Camedda, 2020), indicating that any reluctance to engage with, or lack of confidence in using, new interventions/teaching strategies can heavily influence the outcome of research results, such as in this study. Conversely, increased confidence over time could also influence the outcome of the research results.

Another reason for the lack of immediate improvement from pre- and post-intervention could stem from a lack of engagement or concentration from the children with DS. The teachers reported many times during the intervention sessions that the children were bored by, and disengaged from, the teachers’ instructions. Engagement is recognised as a critical skill for young children with special needs (McCormick, Noonan & Heck, 1998). Moreover, children with DS are highly sociable and often distracted from class activities through their desire to play (Wishart & Johnston, 1990; Fidler et al., 2008). These factors could reduce the effectiveness of any intervention unless it is repeatedly practiced, as it was in the 4 weeks from post-test to follow-up, which could explain the observable improvement in WM from pre-test to follow-up, but not between pre- and post-test. This is supported by the BRIEF-P findings, which showed an improvement in WM at follow-up.

The success of the WM intervention is potentially due to the significant change in teacher knowledge of WM and subsequent use of WM improvement strategies in the classroom. For the EG, there was a statistically significant increase in WM knowledge from pre-intervention to post-intervention, unlike in the CG. These results - especially regarding the improvement of WM knowledge - support previous research; White et al. (2011) for instance revealed that teachers who enrolled in training sessions around other developmental disabilities improved their knowledge significantly. The improvement in teachers’ knowledge might be due to several other factors; one factor could be the teachers’ desire to widen their knowledge, especially given that the school's
administration offered them a suitable time to enrol in the sessions with no additional impact on school activities, which may not be the situation in a different context/study. Also, teacher interest and perspectives in teaching children with DS might be associated with their desire to expand their knowledge in WM; Eren (2012) found in his study that there was a significant relationship between teachers’ interest in teaching and their professional plans and satisfaction with their career choice. Moreover, teacher experience played a role in the current study, with more experienced teachers demonstrating greater belief that WM improvement could occur in children with DS, which indicates that they may have witnessed unexpected improvements in their pupils in the past (and may therefore even stereotype the abilities of their students less than newer, less experienced teachers).

The increase in teacher knowledge of WM more generally, and WM improvement strategies in particular, was supported with the increased use of strategies to enhance WM in their teaching. The observation results revealed that the use of all strategies increased from pre-intervention to post-intervention. To illustrate, strategy 8 (evaluating WM load) was not used at all before the intervention, yet was used at follow-up. The greatest improvements were observed in the use of strategy 1 (responding to the child’s WM difficulties), strategy 7 (encouraging the student to use WM strategies) and strategy 9 (reducing WM load). Yet a significant difference was observed in teacher use of all WM strategies with large effect sizes. Therefore, the intervention was successful in increasing the teachers’ use of WM strategies in the EG. This likely explains the increase in WM seen in the children with DS in the EG.

The significant change in the teachers’ use of WM strategies could be due to the fact that they previously had little knowledge of the majority of WM strategies used in the intervention; after they were trained in these strategies, they became more familiar with and therefore used them more. However, some of these
strategies were used prior to the intervention as teaching methods, such as repetition, rhyme, rhythm, and other supporting methods/materials. However, the teachers did not know that these strategies were WM strategies. This might suggest that increases in teachers’ knowledge and understanding led to changes in teaching style, which resulted in improved teaching methods (as found by Voss, Kunter & Baumert, 2011 and König et al., 2011) and greater confidence in various teaching strategies. Even those strategies that the teachers had previously used were used significantly more, perhaps given the greater confidence and reassurance that these strategies were ‘known’ to be effective in improving WM in children.

Of course, it is also possible that the Hawthorne Effect was present, where the teachers knew they were being watched by the researcher during the observation (Rosenthal et al., 2009), who was expecting to see the WM intervention strategies being used. This would explain why they used significantly more strategies after the intervention, perhaps more than they would usually use in the classroom if the researcher had not been present. Given that the observation only lasted 20 minutes, it is possible that each teacher was keen to demonstrate their competence in using the strategies, either through social desirability bias, perhaps hoping their teaching would be viewed positively, or simply to please the researcher. This is difficult to ascertain, yet it is a common issue in social, psychological and pedagogical research (Arborelius & Timka, 1990). To address this, it might be useful for the teachers to keep a diary of strategy use to gain a more accurate reflection of their actual strategy use.

Yet the findings of the observation and knowledge questionnaire are supported by the teachers’ discussions in the focus group. The teachers in the focus group demonstrated that they changed their understanding of, and improved their knowledge about, the WM of children with DS, which led to a change in their
teaching methods and attitudes toward their students. This is supported by studies in pedagogy research, where previous research indicates teachers often make assumptions about their students’ learning abilities and that their beliefs affect their teaching, as well as potentially limiting student outcomes (Rebmann et al., 2015; White et al., 2011). The focus group also supports the questionnaire results, which showed that teachers’ knowledge about WM increased significantly in the EG but not in the CG. It is also in line with the results of the observation. Significantly, the teachers in the focus group revealed that as their knowledge of WM increased, they became more aware of why some children may behave inappropriately or demonstrate CB, due to WM overload or difficulties. This aligns with the teacher questionnaire results that showed a significant improvement in total CB items. Thus, it is also possible that a greater understanding of the link between WM difficulties and CB in children with DS changed the attitudes and teaching styles of teachers, who saw the benefit of using WM strategies in their classroom. This may have, in turn, improved the WM performance of their students.

The improvements in WM were also transferred, according to the parent view, to global EFs, given there was a significant difference in the BRIEF-P global EF scores (including inhibition, shift, emotional control, WM, and planning/organising) between pre-intervention to follow-up. However, the differences between the EG and CG were not statistically significant in the following specific subscales at follow-up: inhibition, shift, emotional control and planning/organising, once the pre-intervention scores were controlled. Moreover, for teachers, the difference at follow-up between the EG and CG was statistically significant in the WM subscale, but not statistically significant for global EF or the subscales of inhibition, shift, emotional control and planning/organising. This is unsurprising, however, given that the intervention in the EG was solely focused on WM (e.g., Pulina et al., 2015; Costa et al., 2015; Orsolini et al., 2018; Laws et al., 1996; Comblain, 1994).
There are various potential reasons for this discrepancy in findings. Firstly, the fact that parents of children in the EG may have reported a significant improvement in the total score of global EFs (from pre-intervention to follow-up) concurs with previous research that found interventions could improve EFs in the general population, especially in research that uses a similar methodology to the current research (a school-based intervention delivered by school staff and includes parent participation (e.g., Kenworthy et al., 2014)). EFs in the population with DS can also be improved via interventions (Holzapfel et al., 2016).

However, the fact that teachers and parents can often give different reports is commonly found in research. For example, Daunhauer et al. (2014) found teachers and parents of children with DS gave different scores on the BRIEF-P for EFs. It may be that these differences reflect the different contexts in which the tests were conducted, with a greater focus on intellectual considerations in a school environment by teachers and emotional considerations in a home environment by parents, as well as the different points of reference for each group. Research indicates teachers and parents perceive and appraise DS children differently; according to Schworer et al. (2018), parenting behaviours affect the development and perceptions of EF abilities in their children.

In the present research, parents of children in the EG continually asked me whether this intervention would improve their children’s academic achievement, which could mean they may have overrated their children’s EF capabilities, giving slightly higher scores for each subscale (not statistically significant in themselves, but combined can create a significant change in their child’s global EF). This could either be due to bias (perceiving their child as cleverer), social desirability bias (wanting their child to appear to perform better than perhaps they actually do) (Bornstein et al. 2015), or the expectation of a benefit as a result of the intervention (Shen et al., 2017). Culture can play a role here:
parents of children with DS in Kuwait generally want to see improvements in their children as they need to have a certain level of cognitive ability and academic achievement for them to continue being taught in mainstream schools as part of locational inclusion (Public Authority for the Disabled in Kuwait, 2010). Without this achievement level, their child may have to attend a special school far from home. By comparison, teachers may understand the intervention better, especially given the training they have received in it, and can appraise each child more accurately, which is why compared to parents, teachers reported no global EFs improvement in the EG.

Regardless of whether or not there were improvements in global EFs, the improvement in WM is not only important in and of itself, but for potential improvements in academic outcomes in literacy and numeracy. However, the Mann-Whitney test showed that the difference between the EG and CG in literacy and numeracy scores was not statistically significant, indicating that this WM intervention did not effectively improve literacy and numeracy in DS children. Indeed, this is common in educational research, either with TD children or children with ID, where WM training often produces some benefit in terms of specific gains on WM tasks, yet does not appear to benefit academic outcomes, such as achievement-based reading and arithmetic (Melby-Lervåg & Hulme, 2013; Rapport et al., 2013; Redick et al., 2015).

However, as noted, WM was seen to improve between pre-test and follow-up in the EG, and studies demonstrate WM is associated strongly with literacy and numeracy (Alloway & Alloway, 2010). The finding of this study therefore may be explained by the relatively weak cognitive abilities (in particular WM) in children with DS, which could explain the lack of observable change in their literacy or numeracy scores. It may have been that further and sustained improvement in WM was required before there would be a change in their literacy and numeracy scores. As the follow-up was only conducted 4 weeks post-intervention, it is
possible that there was insufficient time for any WM improvement to have any significant impact on literacy and numeracy; a further follow-up (to enable the children to practice beneficial WM strategies and improve further in this area) or longer intervention may help clarify this. This supports findings from Ross and Begeny (2015), who demonstrated that the longer the time period between an oral reading fluency intervention and the follow-up assessment, the more effective the oral reading fluency skills were within TD populations. It may also be the case that given the multitude of other skills required for literacy and numeracy (not simply WM), such as inhibition and cognitive flexibility (Purpura et al., 2017), in which children with DS also show weaknesses (Amadó, Serrat & Vallès-Majoral, 2016; Purser et al., 2015), the change in WM in the EG was not sufficient for a change in literacy or numeracy.

An unexpected finding in the present study was the success of the control intervention. Analysis of the SDQ data revealed that both parents’ and teachers’ reported that children in the CG had significantly fewer CB difficulties at follow-up than the EG group, once pre-intervention scores were controlled for, suggesting that the positive behaviour support (PBS) intervention received by the CG was more effective than the WM intervention in reducing CB. Additionally, while parents did not report any significant difference in prosocial behaviour at follow-up between the EG and CG, there was a greater improvement in prosocial behaviour for the CG children as reported by teachers, which supports the effectiveness of the PBS intervention within the classroom. This supports previous research surrounding the effectiveness of interventions to reduce CB (Feeley & Jones, 2008) and improve prosocial behaviour in DS children (Neil & Jones, 2016; Wadsworth et al., 2015).

Overall knowledge of CB was found to increase in both the EG and the CG, despite the fact that the teachers in the EG did not receive specific training in CB. The fact that the CG reduced total behavioural difficulties might be due to
the fact that the teachers were very interested in and engaged with this type of intervention; all teachers of children with DS in the current study complained about CB, supporting the findings of Kokkinos (2007) that this is an area of significant concern for teachers. The teachers in this study also described the CB they experienced from children with DS as disrupting their teaching and preventing the class from focusing on the lesson (in line with Barmby (2006) and Jennings and Greenberg (2009)). The PBS intervention was therefore well-received by the teachers throughout (conversely, the WM intervention was more reluctantly received by teachers at the beginning of the intervention, although they later appreciated the intervention), and this significant teacher investment could have meant greater refinement of prosocial behaviour and reduced CB in a way that was not available in the EG. As stated previously, research does demonstrate that teacher expectations and support significantly affect the outcomes of research (De Boer et al., 2018).

Therefore, both interventions appeared to have a positive impact on the intended aspect of the children’s development – the children in the EG showed improved WM and the children in the CG demonstrated reduced CB. This – alongside the fact that the CG received an intervention also – strengthens the argument that the WM intervention was effective.

4.4.3 Implications
Having discussed the findings in light of current literature, this section outlines the implications that the findings indicate. If the success of this WM intervention in improving WM outcomes in children with DS is replicated in future studies, this would suggest that it is imperative to incorporate WM strategies in the everyday teaching practice of children and adolescents with DS. Specifically, verbal and visuo-spatial WM could be improved by training teachers how to incorporate effective strategies within their teaching methods to improve WM. This supports and extends the current literature base that has found redesigning
teaching materials, strategies and subject presentation can reduce WM load for pupils and lead to better outcomes for TD students (Hussein & Reid, 2009). Moreover, as part of this, teachers can be shown evidence of how students with DS can harness these strategies to assist students’ WM development in future. This can help teachers to invest more in the WM training, as they may feel more confident in its success.

This study has shown that improving teachers’ knowledge of WM is critical if children’s WM abilities are to be enhanced. Having a greater understanding of WM encouraged teachers to change their teaching methods, including implementing the newly learned WM strategies. This seemed to apply across the board, for all subject matter, as teachers were able to draw on knowledge across a range of aspects of WM, such as evaluating WM load to improve both the learning and concentration/focus of their students and the use of note-taking to improve memory. This therefore suggests that teacher training courses and placements for teachers of students with DS should include some teaching surrounding WM, to improve their general knowledge about this EF, as well as to help them understand how it can be applied to their teaching practice on a daily basis. This kind of training is something that is already given to many teachers of TD populations, indicating that it can extend to teachers of DS populations also (e.g., Walk et al., 2018).

This new knowledge and subsequent experience of strategies to enhance WM in children with DS cultivated a new attitude in teachers towards children and adolescents with DS. Specifically, their perceptions of the learning capacity and potential of their students was altered, which was an important, unexpected finding of the focus group. It may be that, alongside improving knowledge of WM and confidence in using WM strategies in future interventions, changing teachers’ beliefs about and attitudes towards the ability and potential of their pupils could (and should) form an integral component of interventions for
teachers’ teaching practices more generally (Fetters et al., 2002). This is something that requires greater exploration in future research.

Improving teachers’ perceptions of the abilities of children with DS to improve EFs such as WM may also have a positive impact on teachers’ and headteachers’ views of inclusion: research indicates many working within mainstream and special primary schools in Kuwait generally hold a negative view of inclusion (Almotairi, 2013). This is not helped by the translation of DS in Arabic as ‘mental retardation’ (in Arabic, majnon). Almotairi (2013) revealed many Kuwaiti teachers regularly misname the disability as this, and states that teachers and headteachers in this context need training to help them adapt and prepare for students with DS in order to better meet their needs in mainstream schools. The findings of the present study suggest that including a focus on the ability of students with DS to improve their WM performance in such teacher training could assist in cultivating a new, more optimistic attitude towards the expectation of success in students with DS. Whilst the present study only focused on working memory, it is possible that, if teacher attitudes change towards the inclusion of those with DS in mainstream settings in Kuwait, true inclusion can be proposed and even implemented. This may include students with DS not being taught in special suites (locational inclusion) but rather being included in the same classroom settings and following the same curriculum as mainstream students themselves, learning side by side with them. This is an attitude that Nouf, Bader and Abbas (2019) found was becoming increasingly more prominent in pre-service teachers (those in training) in Kuwait; greater awareness and training regarding the WM capabilities of those with DS may form one element of further improving attitudes regarding true inclusion.

The teachers also became more familiar with the factors that can lead to the low academic achievement and appropriate behaviour of children with DS in the classroom. For example, they learned that some CB stems from difficult
academic tasks, which leads children to desire to escape from the class to avoid these tasks, or exhibit ‘switching out’ strategies and avoidance behaviour, in line with current literature (e.g., Feeley & Jones, 2008; Pitcairn & Wishart, 1994; Wadsworth, Hansen & Wills, 2015). Thus, the teachers were able to apply their WM knowledge to other areas they struggled with in class, such as CB, to not only assist with the students’ learning, but lead to a calmer, more productive working environment for all. The implication of this is that WM training for teachers should include modules/sessions on how WM strategies can affect other areas that are salient for teachers, such as reducing CB, with practical strategies and tips on how to manage this. In fact, managing PBS and FBA were found to be effective models to conduct with children and adolescents with DS, demonstrated by their effectiveness in reducing CB in the CG; therefore, given the teachers’ enthusiasm for improving CB and the transferability effect of managing both CB and WM, any WM intervention should give sufficient teaching in both these areas, given the impact one has on the other (Holmes, Gathercole & Dunning, 2010).

It may be the case that the implications of the study findings for teaching practice in Kuwait are, at least to some extent, context/culturally specific. For example, this study was conducted in Kuwait, in a specific cultural and educational context that receives a significant level of support from the Ministry of Education. This may mean that teachers are more readily available to engage in the type of professional learning that took place in this study, than would be the case in other countries. It may therefore be more difficult in other countries to impact the WM development of children with DS without such support. The success of the current intervention, alongside the support received from the Kuwaiti government, means that this context may provide fertile ground for investigating whether other EF interventions may also provide successful for students with DS. Moreover, given the relatively reduced stigma surrounding DS in Kuwait (Alenaizi, 2017) (compared to other countries, especially some Arabic
countries (Hadidi & Al Khateeb, 2015), it is possible that, the parents of children with DS may be happier for their children to be enrolled in an intervention in Kuwait (compared to parents in other countries). Teachers’ beliefs about WM capacity and their willingness to change their attitudes towards the abilities of students with DS may also differ from those in other cultural contexts. Therefore, given the specific context in which the study was conducted, one cannot know the extent to which there may be implications for practice in other countries. It may be beneficial in different contexts to first investigate teacher perspectives prior to conducting any intervention, to help focus on the areas that teachers believe are the most in need of improvement in their students.

There may be wider implications of the findings outside of the Kuwait context. For example, if WM may be improved in a classroom context in those with DS, it is possible that materials (including toys, books and other useful resources) can be developed by manufacturers across the globe, to enable WM interventions to be more readily undertaken within a home context. Moreover, the findings also imply that governments should invest more heavily in WM research and interventions within education for those with DS. This can include training courses or modules for teachers and trainee teachers in enhancing WM inside and outside of the classroom for pupils with DS. Such state investment could also extend to other EFs, given the potential transfer effects that may exist when focusing on one EF (Zhao, Volckaert & Noel, 2015).

Other implications, on an international plane, include the need to inform researchers and educational experts from other countries of the potential for improving WM in the DS population, particularly in childhood. It may be beneficial to highlight the need to: a) change attitudes regarding this amongst teachers, to enhance their understanding of WM and to motivate them towards incorporating WM interventions within their everyday teaching of students with DS with optimism; b) generate greater research interest in investigating the strategies that may be more beneficial and advantageous in terms of producing
demonstrable WM gains in students with DS, the context(s) that can provide the most ideal settings for such interventions (whether they be school, home, laboratory or community-based interventions), as well as the most appropriate age at which to introduce these strategies. Greater research is also required to ascertain how far the results of this study can be extrapolated to DS populations in other settings in Kuwait (such as special schools) and in other countries both in the Gulf and beyond.

4.4.4 Limitations and Future Research
This study was not without limitations that may have affected the results, some of which may be overcome in future studies. One of the main limitations was that as the researcher, I designed, implemented and evaluated the intervention myself. In order to ensure that I remained as objective as possible, I remained neutral to both groups (the EG and CG), gave the same standardised instructions and followed the same procedure (for example, keeping the driving questions the same) (Galdas, 2017). Moreover, I kept a journal to analyse and challenge my own expectations, prejudices and perspectives throughout the entire process, to minimise any bias creeping into the results when evaluating and interpreting them (Chenail, 2011).

Of particular concern was the importance of ensuring that participants felt able to discuss any negative or ineffective aspects of the intervention during the FG. In order to help them remain as honest as possible, I impressed upon them that there was no judgement attached to their perspectives and experiences of the intervention (Greener, 2018), that the FG was being conducted in a private space and that their honest account of their experience could provide invaluable insight into whether this kind of intervention could help children with DS in future. I was clear that my feelings were not important in the overall purpose of the study, and I accepted all comments in a calm, neutral and open manner (Greener, 2018). I also anonymised the data and continually reassured the
participants of confidentiality throughout the process (Wiles et al., 2008). Perhaps in future it may be better for an independent researcher, one blind to the EG/CG status of the teacher and children, to analyse the children’s assessment results following the interventions, to encourage even greater objectivity (Milner IV, 2007).

Another consideration was that in the EG group, the teachers were encouraged to focus on the WM intervention, rather than CB; however, despite their awareness of the importance of the WM intervention, the teachers continually complained about CB in their students with DS. Thus, it is possible that there was a lack of teacher engagement with the WM strategies, which could have affected how far they were implemented by the teachers in the EG and therefore how effective they were at boosting the WM of their students. Future research should therefore assess how far the teachers’ investment in the intervention topic impacts their use of the teaching strategies in the classroom.

Moreover, although there was much broad theory and research to draw upon, such as the theoretical model of WM adopted (Baddeley, 1986), and the previous WM interventions for TD children, there was no clear framework or theoretical basis for developing a WM intervention for pupils with DS. This meant that the researcher was required to start from scratch and design the WM intervention by drawing on a range of existing strategies to improve WM that were not intended specifically for the DS population. This could arguably be a strength as well as a limitation; it meant that the researcher covered a broad range of relevant points for teachers, yet also meant that some areas may have been overlooked, which may not have happened had there been a theoretical ‘blueprint’ or roadmap to follow to guide the researcher in improving WM. Therefore, it was unclear whether the WM intervention would be effective, although this in itself would have been illuminating. The leap into the unknown is therefore arguably a strength of the research, given the originality of the design,
which drew together multiple strategies proposed to improve WM from the literature.

This study also required a great deal of assessment, which provided a significant challenge for both the researcher and all participants, especially the students with DS, who were easily tired and distracted. Whilst this was in part resolved by dividing the assessment period up to ensure participants were assessed comfortably, future research may attempt to reduce the assessment load or combine elements of different assessments into one test, to minimise the time required for assessment. This can help maintain the focus and enthusiasm of all participants.

On rare occasions, some teachers (2) could not attend the sessions; I attempted to overcome this problem by sending them the session materials and focused on them at the beginning of the next session (see driving questions discussion, section 4.2.5) to ensure that they did not miss any element of the intervention. However, it is possible that the two teachers that missed a session did not engage with that session’s content and therefore did not use those WM strategies, potentially affecting the results. Future studies should ensure that catch-up sessions are scheduled with teachers to ensure they all receive the same amount of time spent on each WM strategy in the intervention.

Additionally, the follow-up assessment time point scheduled was linked to the school timetable, as the schools were on the brink of conducting the final exam of the term before the school holidays. Thus, I was obliged to conduct the follow-up assessment 4 weeks following intervention. Allowing more time between the intervention and follow-up assessment could give a clearer image about the sustainability of the results and a more accurate reflection of how far the intervention actually improved the WM of the EG.
Finally, the materials and some pivotal assessments used in this intervention were not available in Arabic (e.g., BRIEF-P and the WM assessment by Lanfranchi et al., 2004), so the researcher had to employ a specialist translation office to translate all session materials (including journal articles, fact sheets and video clips) into Arabic. Moreover, whilst it is possible that mistranslation or cultural difference impacted the findings, this was addressed as far as possible through pilot work and the use of a specialist translation office. Future research could of course benefit from these materials as they are; or, funding could be made available for these materials to be translated into the target language by a reputable translation company to ensure that other countries can access any benefits of a WM intervention.

4.4.5 Challenges
The researcher faced various challenges in conducting this study. Due to the vulnerability of the groups of children with DS involved, the researcher had to be extremely careful to preserve the safety and comfort of the children involved. This was especially important when the researcher was interacting with the pupils themselves, such as during assessment or observation, when faced with CB or a bad mood, and being mindful of the indicators that a pupil may wish to withdraw themselves from the study. This was overcome by the researcher removing themselves from the environment and coming back later, in which instance, the child/ren appeared happy to participate or showed reduced CB, enabling the assessment or observation to continue.

This intervention was conducted in 4 schools located in 2 governorates in Kuwait; whilst these Governorates were very far from each other, which meant practically, the researcher spent a great deal of time travelling for all pre-, post- and follow-up tests and for the 6 intervention sessions. Future research could standardise the intervention materials and teaching and employ several researchers to deliver the intervention to teachers in one school each, providing
a specific point of contact for the teachers/administrators/parents of children in each school, and to reduce the load for one researcher. This could, however, introduce the issue of lack of standardisation in the intervention, which would need to be addressed as far as possible to ensure the researchers were all delivering the intervention and contact to teachers/administrators/parents in the same way.

4.5 Conclusion
This study reveals that a WM intervention specifically targeting teachers of children with DS can be effective in improving the WM of their students. In the EG, WM knowledge in teachers and WM performance in children with DS improved, as shown by the WM assessment and the parent/teacher responses in the BRIEF-P, and supported by observation of teaching strategies and teacher perspectives gained in the focus groups. This study did however report elements that did not improve as a result of the WM intervention, such as literacy and numeracy scores. Thus, whilst the ‘far effects’ and sustainability of the intervention could not be established, the near effects, in terms of improving the performance of children with DS on specific WM-related tasks, was established. This, in and of itself, renders the WM intervention worthy of pursuit, given the potential benefits that WM improvements can have on the quality of life and everyday activities of individuals with DS.

There were also other areas that were not found to significantly improve as a result of the WM intervention, including other EFs or behaviour (in terms of prosocial behaviour or reducing CB). Teachers in the CG however reported that their students improved their behaviour by a reduction in total difficulties and an improvement in social behaviour. Whilst the main focus of this study was not CB, the importance of this issue was highlighted by teachers in both groups, as reducing this issue could generate greater positive outcomes in terms of the effectiveness of the WM intervention. Thus, one of the various implications
arising from this study is the necessity of conducting both WM and CB interventions simultaneously to assist teachers of children with DS more holistically.

The possible reasons and potential limitations of the findings have been explored; yet overall, this study reveals that it is possible to improve WM in children with DS through teacher-based intervention, and that sustained results could be achieved if the limitations of the intervention duration were avoided. The implications for future interventions and teaching practice are far-reaching, as the findings indicate that, despite the specific Kuwait context, WM can be improved in children with DS and that, as a result, other teaching and educational establishments and home environments could potentially adopt similar WM improvement strategies to enhance this critical EF in individuals with DS.
Chapter Five: General Discussion and Conclusion

5.1 Overview
This chapter provides a general discussion regarding the thesis as a whole, highlighting the key findings, implications, recommendations, strengths and shortcomings of the study, to indicate where future studies can build on, and improve, research into working memory (WM) functioning and interventions for children with Down syndrome (DS).

5.2 General Discussion
This body of work explored the EFs most and least atypical in DS, and investigated whether, as previous research indicated, WM functioning is one of the most challenged EF in children and adolescents with DS (study one). Additionally, this thesis reported on the implementation and effectiveness of a WM intervention, delivered to teachers, for children with DS in Kuwait (study two). The findings from study one support the current literature base that WM functioning appears to be the most atypical EF in children and adolescents with DS (with emotional control being the least atypical EF) (e.g., Carretti, Lanfranchi & Mammarella, 2013; d’Ardhuy et al., 2015; de Santana et al., 2014; Visu-Petra et al., 2007). Study two built on the findings within the literature base, by developing the first teacher-focused intervention on WM in Kuwait. The findings from study two indicate that a WM intervention can be effective in improving WM functioning in children and adolescents according to current WM measures. These findings are discussed, and the implications in terms of teaching practice, policy and future research are highlighted. Recommendations in these three areas are also then made.

This thesis has revealed that EF is generally atypical in children and adolescents with DS, and that there are EF variations between children and adolescents with DS (Lee et al., 2015), depending on whether mental age
and/or chronological age is used to draw comparisons with other groups and populations. The systematic review (SR) is a novel contribution to research in the DS field, as it has helped to consolidated existing knowledge regarding EF functioning in children (and adolescents) with DS. This knowledge includes the most reliable measures of EF, areas where further research is required, the overall quality of the evidence base, whether previous findings are methodological artefacts or reliable knowledge, and an unbiased overview of the key issues in the field beyond what one study can provide. The findings in the SR confirmed the decision to design, conduct and evaluate a WM intervention to attempt to improve WM functioning in this population. This was especially important given that whilst there are numerous EF interventions for various groups of individuals, especially typically developing (TD) individuals (e.g., Erwin et al., 2012; Mahar et al., 2006; Moreau et al., 2015; Oswald et al., 2006; Passolunghi & Costa, 2016); Traverso et al., 2015; Vazou & Smiley-Oyen, 2014; Zhao et al., 2015), there is a lack of EF interventions aimed at those with DS, and an even greater paucity of empirically tested WM interventions for those with DS. Those that do exist focus on aerobic exercise and physical activity (e.g., Alesi et al., 2014; Holzapfel et al., 2016; Ringenbach et al., 2015; 2016; Tungate, 2016)), pharmacological interventions (e.g., Johnson et al., 2003; Lobaugh et al., 2001) or computer-based WM interventions (e.g., Klingberg et al., 2002; Pulina et al., 2015; Van der Molen et al., 2010), all of which may present issues when being implemented with the target age group of the DS population (children and adolescents).

The WM intervention in study two of this thesis was designed to be delivered to the teachers so they could implement WM-improvement strategies with children with DS. The findings indicated that the teachers' knowledge of WM and use of teaching strategies to boost WM in the classroom had increased. A particularly important finding was that the teachers’ attitudes towards the learning capabilities of children with DS in the classroom had been positively affected by
the intervention. The idea that teachers of children with DS significantly underestimate their potential and abilities has been found in some literature; for example, a study from Wishart and Manning (1996) of 231 trainee teachers on their attitudes towards students with DS found that many of the teachers underestimated the potential levels of achievement of their pupils. Other studies have revealed that children with intellectual disabilities (including those with DS) are not often included in collaborative learning exercises, ‘possibly on the assumption that the metacognitive and communicative skills on which joint problem-solving activities capitalize are less likely to be available to children with significant cognitive impairment’ (Wishart et al., 2007). The attitudes of teachers therefore play a substantial role in the kinds of activities that children with DS are engaged in within the classroom, which can have a significant impact on limiting their learning and development (Rebmann et al., 2015; Ridgway, 2017; White et al., 2011). Moreover, the finding that some teachers underestimated the learning abilities of their students in WM may also extend to their beliefs about their abilities in other areas, such as learning to read. This can also have a profound effect on their overall attainment (Ridgway, 2017). Study two revealed that the teachers possessed some negative attitudes towards students with DS in this particular setting, which may not always have been consciously acknowledged by the teachers themselves, until they reflected on them in the study. This finding falls in line with research from Almotairi (2013), who revealed that teachers working with children with DS within mainstream and special primary schools in Kuwait generally held a negative view of inclusion. One of the cited reasons for this was the difficulties that teachers expected to encounter given their view that the learning of students with DS is extremely challenged. The change in teachers’ attitudes towards their students’ potential for learning in the present study therefore may positively affect their beliefs about their learning in other areas and alter their classroom activities, strategy use and interaction with their pupils, which could in theory lead to greater student learning in the future.
The other major finding of study two in this thesis related to the students, as the intervention was shown to successfully improve the children’s WM. Whilst it is not currently known whether this intervention has long-term sustainability, or if it may be effective with individuals with DS of other ages (for example, adults), the study demonstrated the ability of teaching strategies, used within the classroom with children with DS, to improve the WM functioning of pupils with DS. This aligns with the conceptual model adopted by this thesis, implying that, as per Morton’s (2004) causal model framework, the environment can indeed have a significant impact on the cognitive and behavioural outcomes of children with DS. This study focused on the classroom environment in particular, revealing that a WM intervention can improve the cognitive functioning (WM outcomes) of those with DS in the class. The WM improvement strategies used were embedded within the curriculum content, instead of delivered as standalone strategies. As Takacs and Kassai (2019) found, this kind of embedded training has greater ecological validity and is more enjoyable than standalone training (which they describe as ‘explicit’ training), which could indicate a potential reason as to why it was effective in boosting WM, without making undue demands on teachers.

5.3 Implications
This section discusses the implications for practice, policy and future research in light of the findings of the thesis.

5.3.1 Implications for Practice
The intervention was aimed at teachers as a result of their heavy involvement with the schooling and teaching of children with DS, as well as obtaining data from parents, although this was limited to their assistance in assessing their child for any improvements in EF or behaviour. There are multiple implications for practice that may be drawn from this body of work. Clearly, there is the
implication that, given the effectiveness of the WM intervention, teachers of students with DS across both mainstream and specialist schools could adopt the WM improvement strategies to help enhance the WM of students with DS in a range of educational settings. By utilising implicit WM training—embedded within the curriculum teaching strategies adopted by teachers of students with DS in Kuwait—WM improvement strategies could be applied more holistically across different subjects, not simply numeracy and literacy. Furthermore, whilst the use of play to improve WM has not (to the best of this researcher’s knowledge) been used with students with DS, it has been used to great effect to boost WM in TD preschoolers (Thibodeau et al., 2016). Therefore, it may be beneficial to also incorporate different factors that have been shown to boost WM (and EF more generally) as well as general health and wellbeing to drive a multifaceted approach towards WM improvement. For example, teachers could use imaginative play to teach different elements of the curriculum to students with DS, as well as other WM improvement strategies and gentle physical activity (depending on the ability level of the student and, naturally, in line with caregiver consent and the requirements of their individual care plans). This could harness the potential benefits of a range of different interventions that have been shown to improve EFs—especially WM—in children and adolescents with DS.

Moreover, it may be beneficial in future to include parents more substantially in the intervention, asking them to use the same WM strategies in the home with their child to ascertain if this would generate greater intervention efficacy and lead to more successful and/or more sustainable WM outcomes. This has been undertaken successfully by some studies, such as Kenworthy et al. (2014), who found that an EF intervention for those with ASD that used the same scripts, vocabulary, visual cues and training for parents and teachers) were effective due to the transferability of the intervention between home and school contexts. Involving parents more fully may help to generate more positive attitudes towards the abilities of their children to learn; whilst some studies have indicated
that many parents of children with DS create a home environment conducive to learning (e.g., Ricci, 2011), Phillips et al. (2017) found that parents with children with DS often use different parenting styles compared to parents of TD children, such as less reasoning and induction and ignoring CB. Other studies have shown that parents of children with mild ID have lower expectations of their academic achievement (Taylor et al., 2010), which was the biggest predictor of actual attainment. Thus, less positive attitudes towards learning, parenting style and negative expectations of achievement of children with DS in parents could potentially negatively impact the cognitive (and EF) development of these children. Parental involvement in an EF/WM intervention may therefore work to change parental attitudes, expectations and subsequent parenting styles and behaviour towards their children with DS, encouraging greater EF/WM and overall cognitive development and perhaps even lower parental stress (Phillips et al., 2017).

Moreover, a similar intervention could be tailored and used with adults with DS where is it applicable; whilst these individuals may not be in a school environment, they may benefit from the intervention either in the home or in other services within the community. Therefore, the intervention could be rolled out to different professional and home-based services that assist those with DS, to provide a more holistic approach to improving WM in those with DS. However, some significant adaptations to the intervention would be required, such as making the activities used within the WM-promoting strategies more relevant to adults, such as a greater focus on daily living skills. This might be useful to implement during early adulthood, given the finding that many adolescents with DS aged 16-19 experience serious difficulty in mastering basic daily living and personal hygiene tasks and activities. This can present substantial challenges to independent living as an adult (Van Gameren-Oosterom et al., 2013). Implementing such an intervention would need significant consideration and planning however, as well as the involvement of key carers in the individuals’
lives, such as parents, key workers, health care professionals and any others that could assist, given that, without being in formal education, adults with DS would not have the benefit of seeing the same teacher for a significant portion of the day, from whom the strategies to improve WM can be learnt.

That said, some researchers assert that WM training may be more beneficial if it is delivered earlier to children with DS. Wass et al. (2012) and Fidler et al. (2018) highlight that interventions must be designed and implemented so as to capitalise on early neuroplasticity, therefore the intervention from study two could be adapted for the pre-school years (3-5 years-old), or younger. This would require not only future research, but greater collaboration between parents, healthcare providers and relevant early years teachers to ensure that children with DS are targeted and able to undergo such suitable interventions before they attend school.

The findings from this study may also have relevance for stakeholders outside of Kuwait; WM strategies could be implemented in other countries that have a similar educational and government support system to Kuwait, such as Saudi Arabia or the UAE. There is a Gulf convention regarding education in the Gulf Cooperation Council (GCC, 2020), as these countries similarly strive for excellence in and share research/ideas/practices regarding this field, including for children with SEN. Effective WM strategies for students with DS could be shared within the GCC. Furthermore, the findings of this study may have implications in Arab countries outside of the field of education, both in and beyond Kuwait. WM interventions are not limited to the classroom; they may be implemented in the workplace, for example, to assist those with DS that have jobs or voluntary/community work to improve their WM in the workplace. In fact, simple WM strategies, such as encouraging rehearsal in those with DS, could be made part of the way wider society approaches people with DS in the community more generally; raising awareness of ways in which individuals with
DS can benefit from simple yet effective strategies when communicating and interacting with individuals in the wider community. Not only can these strategies assist in improving WM in those with DS, allowing them to participate more fully in society, but they can also serve to educate the general public about DS with the aim of reducing stigma, shame and negative attitudes towards individuals with DS more generally.

In addition to difficulties in EF, challenging behaviour (CB) in students with DS was the most pressing issue that teachers stated they struggled with within the classroom. This was supported previously in the literature review (Otten & Tuttle, 2011); moreover, the literature revealed that there is a correlation between EF and CB (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998; Wilding et al., 2002), as well as a relationship between EF difficulties and academic achievement, which in turn can lead to CB (Otten & Tuttle, 2011). This study had the beneficial effect of changing teachers’ attitudes towards the CB exhibited by children, given their increased understanding of why they may be behaving inappropriately, instead of simply intending to disobey the teachers’ instructions. Given that CB is a concern for teachers, it seems as though a WM/EF intervention should also support teachers to develop their knowledge of the link between EF and CB. In future, therefore, it would be pertinent to involve some elements of the CB intervention within the WM interventions to best improve the classroom environment and facilitate individual and group learning in such a context.

A more research methods-focused implication drawn from this thesis is the need for a non-verbal IQ assessment measure for use in Kuwait that does compare scores with normative data, such as the use of Raven in many countries (the English-language version). In Kuwait, the old assessment tools (the Arabic Raven version by Qurashi from 1987) are still being used, involving the use of percentages that accord different levels to determine where a child with DS fits.
into the (large) percentage categories for estimated MA levels, but without using the actual score or MA proxies. I was concerned that this, as noted in section 4.2.4.2, may potentially underestimate an individual’s abilities, if their score lies between two percentage categories. Clearly, to avoid this, a more updated version of Raven – and indeed, greater use of a range of measures to measure both IQ and non-verbal MA – is required in practice for future research in DS, both in Kuwait and beyond. Measurement development would also be important for future cross-cultural research, given that it would currently be difficult to compare outcomes across countries because of differences in the measures use.

5.3.2 Implications for Policy
The demonstrable effectiveness of the intervention and the lack of facilities and resources currently available to implement it may also carry implications for educational policy. For example, it could encourage educational decision-makers to produce more support for teacher-involved interventions, as these kinds of interventions could have a sustainable effect on outcomes for both children and teachers.

The results of this research also revealed the potential to replicate the study on a larger scale, which could (if the results are in line with the present smaller-scale study) highlight to key educational decision-makers, such as managers of educational establishments and funding bodies, how such an intervention may be important to be conducted on a wider-scale across Kuwait. Attracting funding for such an intervention is critical to its success, given the need for resources (including manpower) to deliver the intervention to multiple institutions and continually assess its efficacy.

There are very few published teaching strategies that focus on improving WM in DS as teaching methods, bar some exceptions (for example, Buckley and Bird
(2001)), so it may be important to introduce WM improvement strategies to teachers within teacher training courses or as part of the schools' protocols/guideline for teachers through the teaching methods administration. It could even be designed to form a part of the curriculum. Making information on how to support WM development in children with DS in the classroom more readily available to teachers of pupils with DS, particularly in Arab contexts, where there may be fewer existing resources, is both a policy and practical consideration in school contexts.

It might also be worthwhile for governments, such as the Kuwaiti government, to employ education researchers to continue conducting WM assessments, or any cognitive assessments, with pupils with DS. This can add to the growing body of knowledge regarding the cognitive abilities (and any improvements therein) in children with DS, which can give a clear image about how much input and support particular children need during the school year, so that teachers can tailor their strategies and identify areas where pupils may need some more support. It can also help to indicate whether the WM strategies adopted in the present study are effective for all children or whether some children might need support in a different way. In Kuwait, the rules surrounding educational research prevent any person except highly trained researchers from assessing the cognitive skills of individuals with DS (Psychological Services of the Ministry of Education, 2004); however, the findings of the present study indicate that a WM intervention, such as the one used in study two, can give sufficient background about how WM works to teachers, as well as training them how to improve and assess the WM in children. If the intervention were rolled out to all teachers of pupils with DS, teachers may then be able to witness on a daily basis how WM may improve (or decline), which could help the teachers to support the child more effectively over the course of the year, perhaps in turn making any intervention benefits more sustainable.
There appears to be a need for clearer governmental guidelines for the care and development of children and adolescents with DS in Kuwait. As outlined in chapter one, in Kuwait, each ministry follows the government rules to facilitate the care of individuals with special needs, according to the specialisation of each ministry. However, there are no clear clinical guidelines for this population comparable to the NICE guidelines in the UK, including regarding research with DS populations. Therefore, there is a need to generate set clinical guidelines that are in line with the governmental rules regarding special needs care and education, that each ministry can follow for a number of purposes, including assisting conducting research with individuals with DS, and improving their educational setting and experience. Although it was not a focus of the research in this thesis, it was clear from study two that the education context of children with DS in Kuwait is not entirely inclusive.

Inclusion is locational only; compared to the UK, for instance, those with DS do not take part in mainstream classrooms, but are taught in a separate wing with their own classroom suites. Given that research indicates that full inclusion can have benefits for both TD students and those with special educational needs and learning difficulties (Qvortrup & Qvortrup, 2018; Tomlinson & Parrish, 2018), this is recommended.

5.3.3 Implications for Future Research
This study has highlighted the need for greater research into the area of improving WM in children and adolescents with DS, and in the field of EF and cognitive development in this population more generally. The present study is novel in many ways, which opens to door for greater research into the findings. This is the first time that an intervention to improve WM in children with DS conducted in Kuwait has been reported in academic literature; there is no evidence that a similar intervention has been conducted in any Arab country (similar to Kuwait in terms of cultural context) to date. There is a clear lack of
Arabic research in the area of EF or WM in DS, nor any known intervention conducted to boost WM in DS population; thus, the study not only filled a very clear gap in the research, and met the urgent need for interventions in this area, but it also highlights the importance of extending the intervention to other EFs in those with DS in Arabic-speaking countries, given the lack of such an intervention for individuals with DS in these contexts.

Of course, the context in Kuwait is very different to other countries; the laws surrounding educational research mandated by the department and ministry of education supports research in this area, through grants, administrative support and facilities, which makes schools and teachers more amenable to being the focus of research or facilitating research in their schools (Kuwait Ministry of Education, 2013). Thus, Kuwait is an ideal context to conduct research into WM intervention strategies in children. However, it must be noted that this may not be the situation for other contexts. Yet other Arabic-speaking countries may have a similar cultural attitude towards education (Moosq et al., 2001) and similar special education curriculums, especially in the Arabian gulf, where there has been educational cooperation as part of the Gulf Cooperation Council since 1975 (GCC, 2020). Therefore, future research could first assess the potential of such a WM intervention, similar to the one in the present study, in countries that are culturally similar to Kuwait, such as in the Arab Gulf, before its potential in culturally-dissimilar countries (such as the West) is explored.

This study focused on the possibility of improving WM in children with DS through the design of an appropriate intervention. Future research could adopt the same intervention design with the purpose of improving a different EF, such as attention or inhibition, to ascertain its effectiveness in other EF domains. This is especially important given that the general literature review in this study found that there are no studies that compare the effectiveness of interventions that aim to improve one EF or several EFs; this indicates a gap in the research literature
that could be filled by investigating the effectiveness of the intervention between one function or several functions. However, research does indicate that focusing on interventions for one EF can improve the abilities in another EF; Zhao, Volckaert and Noel (2015) found that inhibition training for 20 minutes each day improved WM in TD preschoolers. However, this may not necessarily lead to a long-term transfer effect, as Aydmune, Introzzi and Lipina (2019) found that inhibition training only led to short-term visuo spatial WM improvements. In addition, it may be more difficult to assess EFs other than WM in those with DS; for example, teachers may struggle to evaluate students’ attention without the use of specialist equipment or technology (e.g., as used in studies by Raca and Dillenbourg (2013), Climent et al. (2019) and Goldberg et al. (2019)); it may be easier to ascertain overt demonstrations of lapses in attention, rather than quieter students that may also not be focused on the task, compared to WM.

The intervention was delivered by teachers; this is a very important issue that should be taken into consideration to achieve intervention sustainability. It was found in this study that teachers’ knowledge about cognitive functioning in DS—especially WM—significantly improved as a result of teaching them the intervention strategies to improve WM. The teachers indicated they became more familiar with the role that WM plays in learning, knowledge that could arguably improve the quality of their teaching more generally through using more WM strategies than before and evaluating children’s WM load. Thus, not only can this intervention benefit the recipients of the strategies (children with DS), but it can also benefit the teachers that are being asked to deliver such an intervention, enhancing their professional skills both now and in the future. Moreover, as evidenced by the driving questions that played a fundamental role in this intervention and discussion every session, the ability of teachers to share their knowledge and experiences of using the WM strategies could be harnessed to ‘train’ other teachers. In future research, it would be useful to ascertain if key elements of the intervention may be lost if some teachers were
selected to deliver the intervention to other teachers, or whether this might be a useful strategy to roll the intervention out more widely. In general therefore, future research could focus on how best to deliver a teacher-involved or driven intervention, including the most effective ways of teaching intervention strategies to teachers, and how to engage and empower them to use the strategies effectively throughout their lessons.

The systematic review found that most studies on EF in those with DS were focused on children; there was a paucity of research on EF in adolescents with DS when searching for literature on this topic, despite the inclusion criteria of individuals within the age bracket of adolescence. This indicates that more extensive empirical research on EF in adolescents with DS is required to understand how EF may change (if at all) over time, with age.

Another implication for research drawn from this thesis is the need for the development of a standardised test to evaluate EF in a population with DS. As seen in study one, there are multiple tests for the various EFs, some performance-based and other self-report by either parents or teachers. As Manrique-Niño et al. (2020) indicate, there is currently no specific, standardised EF measure, although they note some new tests have been proposed and are currently in development, including the ‘NIH’ Toolbox cognitive battery for ID (see Shields et al., 2020). Further research could focus on developing a feasible, sensitive and accurate measure to test EF in DS populations. This goes hand in hand with EF interventions, given that, once a valid, standardised EF battery has begun to be used more widely, including cross-culturally, clinical trials for EF interventions for children with DS are proposed, to assess the efficacy of the intervention(s). This could lead to greater insight into variations that may be found in results, including the intervention type and duration, the EF being tested, the age of participants, who was delivering the intervention, and near/far and short-/long-term transfer effects observed.
Finally, this thesis was grounded in Morton’s (2004) causal modelling framework. This meant that this conceptual model was considered within both study one and study two, particularly within study two, for the intervention design, implementation and evaluation. The benefits of this for the thesis included a reference by which ideas could be cross-checked and from which could be developed, such as the driving questions and focus groups with teachers. These elements of the research meant that the different levels through which individuals with DS can be considered, and the pivotal role they can play in the cognitive and behavioural development of the child with DS, could be impressed. Moreover, their experiences and perspectives could be gained. Thus, future research would benefit from being grounded in theory, as various studies have shown the effectiveness of using a theory to provide greater depth and insight to their studies within the field in education (Nicholson & Cleland, 2015).

5.4 Strengths of the Research
There are many strengths of the present research study; study one (the SR) covered a large range of dates of published literature, which enabled the researcher to incorporate older relevant literature that may not have been using EF terminology but that nonetheless assessed EF functions. It also looked at children and adolescents, to ascertain whether there was research into how EFs developed over this age range. It investigated all EFs, to facilitate a better understanding of the EF profile in DS and did not overlook studies that included (for example) WM within a broader battery of assessments. It also was not limited to only one way of assessing EF (e.g., only teacher/parent report), and looked at EFs across a range of different comparison groups, to help foster a better understanding of how EF capabilities in individuals with DS may differ from different populations (not just TD norms), including other groups with other
ID. This allowed a better insight as to the specific EF profile (strengths and weaknesses) within DS. This addressed a gap in this research area.

In study two (the WM intervention), a wide range of assessment tools were used, aimed at children and teachers, with the participation of parents. This strengthened the research as it meant the intervention could be assessed in a number of different ways and from different angles, such as whether the teachers were able to implement the strategies, how the intervention was perceived and experienced by the teachers in terms of feasibility, whether it had an impact on pupils and if that impact extended to outside the school. For example, although this study adopted quasi-experimental design, a qualitative method (focus group) was incorporated to investigate the teachers’ perspectives for EG in greater depth. This was intended to enable participants to feel that they had a ‘voice’ and were able to provide insights as to how they felt about the intervention and its overall effectiveness (Silverman, 2016); this can help to improve the intervention in future. Moreover, adopting a qualitative method enabled the results to be triangulated (Wilson, 2014), to ascertain whether there was a correlation between any demonstrable improvements in the WM scores of the children with DS, and the teacher’s perceptions of any WM improvements in the classroom.

Thus, one of the strengths of this study is the highly holistic evaluation of the intervention in terms of a range of different respondents. The holistic approach to WM assessment and the intervention more generally aligns with Morton’s (2004) causal modelling framework, used as the conceptual model for this study, as it highlights the inextricable interlinking between each child, their cognitive abilities and development and their environment (see chapter two for more details on this). Grounding the research in Morton’s (2004) model meant that the different aspects of the model were taken into consideration when designing, implementing and evaluating the intervention. Teachers in the intervention were
made aware of the different levels at which DS can be considered, and the profound impact that the environment can have on their development.

The intervention in study two was aimed at teachers to help enhance the sustainability of the research effect; moreover, the researcher kept in touch with the teachers to help them in case they needed any clarification after the intervention. This was intended to continue to improve their confidence and skills in using the intervention strategies, and therefore help improve the effectiveness of the intervention in the longer-term, providing a strength of the research study (Aldon et al., 2017; Avgitidou, 2020; Moore et al., 2017).

Study two was also strengthened through the use of an active control group with the aim of overcoming the ethical issue of including children and adults (teachers) in the study that may not ultimately benefit from the research and to strengthen the research design (Kinser & Robins, 2013). Using an active control group meant that participants' time was rewarded by gaining something beneficial from the study; given that CB was reported to be a common occurrence, a CB intervention was deemed to be the most useful intervention for teachers in all the schools involved. The teachers reported benefitting from this intervention immensely, providing a strength of this study, as this meant their participation was sustained (avoiding participant attrition).

A final strength of this study was that the researcher gained a significant degree of support from the Kuwaiti Ministry of Education and from all of the schools enrolled in the intervention. This meant that the research could go ahead as planned, despite taking several years between inception and completion, and might have had the additional bonus that the teachers thought it a worthwhile intervention to invest time in.
5.5 Limitations

Despite the many strengths of this study, there are also various limitations to the research. Limitations of the individual studies are discussed in the relevant chapters and will not be repeated here. Rather, there is some consideration of limitations of the thesis as a whole. Firstly, although the SR highlighted a number of areas of potential EF difficulty, it was only possible to take one forwards to an intervention, given the time and practical constraints experienced by the researcher. Furthermore, there were perhaps other ways that study two could have been developed from study one, such as placing greater focus on better understanding WM/EF in adolescents specifically; however, time demands meant that this was not practicable, despite the greater understanding this would have lent to the body of literature on WM/EF in this population. Moreover, the researcher wished for the thesis to have a practical impact in Kuwait, which is why an intervention was conducted, instead of simply focusing on EF/WM capabilities in different population groups.

A considerable limitation is the researcher’s lack of training specifically in EF; whilst the researcher has worked with children with DS for numerous years, the thesis might have been improved with the insight of EF specialists. This could perhaps have lent a deeper understanding to the neurological underpinnings of EF in both TD and DS populations to ascertain how they might differ.

Another limitation is the lack of theory testing undertaken in this study; whilst Morton’s (2004) causal modelling framework was used as a conceptual model for this thesis, this predominantly informed the design, implementation and evaluation of the intervention. It might have been useful to test the theory as part of the thesis, to ascertain its usefulness for future studies of this kind; however, time constraints and the concern that this might broaden the research focus too much meant that the researcher decided not to test any theory as part of the thesis.
In the literature surrounding EF, there are numerous definitions, conceptualisations, theories and tools to describe and measure EF. This has led to a lack of agreement amongst researchers regarding EF, which arguably led to a level of vagueness surrounding the concept of EF in this study. However, this issue did not affect the research question in this study, given that a very specific understanding of EF was adopted in the thesis that of Goldstein and Naglieri (2014), who argue that ‘EF’ is now used as an umbrella term to describe a range of hypothesised cognitive processes (for more details see 2.5.1). Yet the fact that there are so many theories and definitions of EF indicates that the narrowing down of EF (to operationalise the concept) may provide limits to the scope of the study in its understanding of EF.

5.6 Conclusion
As has been discussed in this chapter, there are key implications and recommendations stemming from the findings of this study, in order to improve the WM functioning (and therefore learning skills and quality of life/wellbeing) of the DS population both in Kuwait and potentially in different cultures across the globe. Moreover, this chapter has highlighted the strengths of this thesis, indicating the benefits it posed to both the participants and wider research in the field of EF in DS more generally, whilst also noting the limitations of the thesis and areas for future study.

The production of this thesis proved to be a hugely enriching, illuminating experience, providing a better understanding of EFs in children and adolescents with DS and illuminated the research gaps in this area. It shows this area still requires an extensive amount of research, especially in terms of comparisons between different age ranges and different measures that assess the same function, to deepen current understanding about existing measures of EFs in DS. It also gives a positive impression about how an intervention similar in
design to the WM intervention could potentially improve other cognitive functions, especially if teachers are able to participate in the intervention as ‘experts’ of the children that they teach, taking their perceptions into account when discussing different elements (and the overall success) of the intervention.

The thesis clearly highlights the potential for interventions such as the WM intervention to change the perspectives and attitudes of teachers regarding the learning potential of children/students with DS; given that in Kuwait, there is arguably a more positive view of individuals with DS compared to other cultures, the potential for the attitudes and beliefs of teachers in other, less positive cultures to be changed and the ensuing impact this could have on the learning and attainment of children with DS cannot be underestimated. This carries exciting possibilities; moreover, encouraging the involvement of parents in the improvement of the WM (or wider EF) capacity of their child/ren with DS within Kuwait may also help to provide a more holistic intervention to improve WM from multiple angles, perhaps helping to change attitudes towards those with DS within the community as well as the classroom.

Focusing on the different effects of DS on various aspects of their lives can help experts and family to understand the child’s behaviour, indicating how cognitive function and behaviour can be significantly affected by both biological factors and the environment. This may not only explain differences and difficulties in skills and abilities, but can hopefully pave the way to a greater understanding of how these can be improved by generating a more tailored environment that can be conducive to learning, potentially improving the quality of life for not only the child with DS, but those around them, on a daily basis.
References


Almoosa, A. S., Storey, V., & Keller, C. (2012). Meeting the needs of all: Why schools in Kuwait are failing to meet their moral obligation and what can be learned from the US education system. *Journal of Alternative Perspectives in the Social Sciences, 3*(4), 997-1009.


Broomhead, K. E. (2013). Preferential treatment or unwanted in mainstream schools? The perceptions of parents and teachers with regards to pupils with special educational needs and challenging behaviour. *Support for Learning, 28*(1), 4-10.


Cameron, L, & Murphy, J. (2007). Obtaining consent to participate in research: The issues involved in including people with a range of learning and communication disabilities. British Journal of Learning Disabilities, 35, 2, 113-120.


405


Digital Promise (2017). *Research @ Work: Working Memory & Reading* [video]. Digital Promise [online]. Available at: https://www.youtube.com/watch?v=nwk38zLk84&t=34s [Accessed 20 April 2020].


Ehcap (2014). *Scoring the Strengths and Difficulties Questionnaire* [online]. Available at: https://www.ehcap.co.uk/content/sites/ehcap/uploads/NewsDocuments/236/SDDQEnglishUK4-17scoring-1.PDF [Accessed 8 June 2020].


Kuwait National Assembly (2015). Initial report of the state of Kuwait regarding the implementation of convention on the right of persons with disabilities submitted to committee in 2015 under paragraph (1) of article 35. Committee of People with Disabilities.


O'Malley, P. (2013). Evolving insights about the impact of sensory deficits in the elderly: Comment on "The prevalence of concurrent hearing and vision impairment in the United states" and "Hearing loss and cognitive decline in older adults". JAMA Internal Medicine, 173(4), 299-305.


Psychological Services of the Ministry of Education (2004). Psychological Services in Education. Al Qabas, November 18 2004 [online]. Available at: https://alqabas.com/article/140140-%D8%A7%D9%84%D9%80%D8%AE%D8%AF%D9%85%D8%A7%D8%AA-%D8%A7%D9%84%D9%86%D9%81%D8%B3%D9%8A%D8%A9-%D9%81%D9%8A-%D8%A7%D9%84%D8%AA%D8%B1%D8%A8%D9%8A%D8%A9%D8%A7%D8%B3%D8%AA%D9%82%D8%A8%D9%84 [Accessed 21 August 2020].


Appendices
Appendix 1.1: PROSPERO Study Protocol

Systematic review

   Give the working title of the review, for example the one used for obtaining funding. Ideally the title should state succinctly the interventions or exposures being reviewed and the associated health or social problems. Where appropriate, the title should use the PREDICOS structure to contain information on the Participants, Intervention (or Exposure) and Comparison groups; the Outcomes to be measured and Study designs to be included.
   Proposed study: a systematic review of executive functioning in children and adolescents with Down’s Syndrome

2. Original language title.
   For reviews in languages other than English, this field should be used to enter the title in the language of the review. This will be displayed together with the English language title.
   A systematic review of executive functioning in children and adolescents with Down’s Syndrome

3. * Anticipated or actual start date.
   Give the date when the systematic review commenced, or is expected to commenced.
   11/07/2016

4. * Anticipated completion date.
   Give the date by which the review is expected to be completed.
   31/12/2019

5. * Stage of review at time of this submission.
   Indicate the stage of progress of the review by ticking the relevant Started and Completed boxes. Additional information may be added in the free text box provided.
   Please note: Reviews that have progressed beyond the point of completing data extraction at the time of initial registration are not eligible for inclusion in PROSPERO. Should evidence of incorrect status and/or completion date being supplied at the time of submission come to light, the content of the PROSPERO record will be removed leaving only the title and named contact details and a statement that inaccuracies in the stage of the review date had been identified.
   The field should be updated when any amendments are made to a published record and on completion and publication of the review. If this field was pre-populated from the initial screening questions then you are not able to edit it until the record is published.
   The review has not yet started. No
Grant number(s)

13. * Conflicts of interest.
List any conditions that could lead to actual or perceived undue influence on judgements concerning the main topic investigated in the review.
None

Give the name and affiliation of any individuals or organizations who are working on the review but who are not listed as review team members. NOTE: email and country are now mandatory fields for each person.

State the question(s) to be addressed by the review, clearly and precisely. Review questions may be specific or broad. It may be appropriate to break very broad questions down into a series of related more specific questions. Questions may be framed or refined using PRECOS where relevant.

To identify which elements of executive functioning are impaired amongst children and adolescents with Down’s Syndrome (DS), as compared to matched populations (typically developing or those with developmental and/or chromosome disorder).

To identify executive functioning strengths and weaknesses from within-group comparison design studies drawn from Down syndrome populations.

State the sources that will be searched. Give the search dates, and any restrictions (e.g. language or publication period). Do NOT enter the full search strategy (it may be provided as a file or attachment.)

Books chapters and journal articles up until July 2016. (In English)

A comprehensive international search of the academic literature will be conducted using electronic bibliography databases:
Assia, Cochrane Library, MEDLINE, PsyCHINFO, Web of Science, ERIC.

Lead researchers in the field will also be contacted to identify in-press studies, and registries such as...
20. *Intervention(s), exposure(s).*

Give full and clear descriptions or definitions of the nature of the interventions or the exposures to be reviewed.

Studies of executive function interventions will be included only where baseline data on a Down syndrome group are available or if the comparison group is non-DS population.

21. *Comparator(s)/control.*

Where relevant, give details of the alternatives against which the main subject/topic of the review will be compared (e.g. another intervention or a non-exposed control group). The preferred format includes details of both inclusion and exclusion criteria.

Studies will be included that employ a chronological age (CA)-matched control group and/or a mental age (MA)-matched control group, or those matched on other measures (e.g. language assessments). Studies will be included which have a control group of typically developing children or those with developmental and/or chromosome disorder. Additionally, studies will be included of a within-group comparison design based solely on a Down syndrome population.

22. *Types of study to be included.*

Give details of the types of study (study design) eligible for inclusion in the review. If there are no restrictions on the types of study design eligible for inclusion, or certain study types are excluded, this should be stated. The preferred format includes details of both inclusion and exclusion criteria.

Included: Randomised/ quasi-randomised; non-randomised controlled; cohort; case-control; observation; cross-sectional; longitudinal; before/after.

Describe the method of assessing risk of bias or quality assessment. State which characteristics of the studies will be assessed and any formal risk of bias tool that will be used.
The content will be compared for inconsistencies and omissions once data extraction has been independently completed. Relevant toolkits such as CASP (www.casp-uk.net) will be employed to appraise the quality of evidence generated from the selected studies.

Provide details of the planned synthesis including a rationale for the methods selected. This must not be generic text but should be specific to your review and describe how the proposed analysis will be applied to your data.
Narrative synthesis will accompany the data tables and statistical outputs. Data tables and statistical outputs will report on e.g. observed mean differences between research participants with DS, and comparison groups on each test measure extracted from each study. The purpose of the narrative synthesis will be to produce a profile of executive functioning domains for child and adolescents populations with DS, compared to other populations. It will explore individual differences arising from within-group comparisons, as well as differences between Down syndrome and other populations. It will comprise analyses of individual study design, sample size, sample characteristics (including co-morbid diagnoses), objective/outcome measures, intervention (where applicable as part of setting description) to show study context, duration of study, methods, and main findings. It will identify differences between studies in methodological quality and highlight directions for future research.

29. * Analysis of subgroups or subsets.
State any planned investigation of ‘subgroups’. Be clear and specific about which type of study or participant will be included in each group or covariate investigated. State the planned analytic approach. If sufficient sources are located to warrant statistical synthesis, meta-analysis models will be generated to show overall effect size. This will require consideration of data heterogeneity and the quality of the studies that are finally entered into the systematic review. Effect sizes will be calculated based on the mean difference between cognitive test scores for Down syndrome populations, compared to control comparison groups. This will provide a random effects statistical model of strengths/weaknesses in executive function amongst Down syndrome populations, compared to other populations. Where data allow, subgroup analysis will also be conducted to explore the effects of variables including: age; differences between types of executive function assessment, country of study and participant gender.
Findings will be extracted and discussed in the narrative synthesis, and these may include comparison of ability in different aspects of executive functioning (e.g. working memory, impulsivity, planning) different groups (e.g. Down syndrome and typically developing control groups) and comparison of different subgroups (e.g. based on chronological or mental age, or language ability) within the Down syndrome groups.

30. * Type and method of review.
Select the type of review and the review method from the lists below. Select the health area(s) of interest for your review:

**Type of review**
- Cost effectiveness
- Diagnostic
- Epidemiologic
- Individual patient data (IPD) meta-analysis
- Intervention
- Meta-analysis
- Methodology
- Narrative synthesis
- Network meta-analysis
- Pre-clinical
- Prevention
- Prognostic
- Prospective meta-analysis (PMA)
- Review of reviews
- Service delivery
- Synthesis of qualitative studies
- Systematic review
- Yes

Other
31. **Language.**
Select each language individually to add it to the list below, use the bin icon to remove any added in error.

*English*

There is an English language summary.

32. **Country.**
Select the country in which the review is being carried out from the drop down list. For multi-national collaborations select all the countries involved.

*Scotland*

33. **Other registration details.**
Give the name of any organisation where the systematic review title or protocol is registered (such as with
The Campbell Collaboration, or The Joanna Briggs Institute) together with any unique identification number assigned. (N.B. Registration details for Cochrane protocols will be automatically entered). If extracted data will be stored and made available through a repository such as the Systematic Review Data Repository (SRDR), details and a link should be included here. If none, leave blank.

34. Reference and/or URL for published protocol.
Give the citation and link for the published protocol, if there is one
Give the link to the published protocol.
Alternatively, upload your published protocol to CRD in pdf format. Please note that by doing so you are consenting to the file being made publicly accessible.
Yes I give permission for this file to be made publicly available
Please note that the information required in the PROSPERO registration form must be completed in full even if access to a protocol is given.

35. Dissemination plans.
Give brief details of plans for communicating essential messages from the review to the appropriate audiences.
Publication in a peer review journal, inclusion in PhD thesis, presentation at academic conferences.

Do you intend to publish the review on completion?
Yes

36. Keywords.
Give words or phrases that best describe the review. Separate keywords with a semicolon or new line.
Keywords will help users find the review in the Register (the words do not appear in the public record but are included in searches). Be as specific and precise as possible. Avoid acronyms and abbreviations unless these are in wide use.
Down syndrome
Executive function
Intellectual disabilities

37. Details of any existing review of the same topic by the same authors.
Give details of earlier versions of the systematic review if an update of an existing review is being registered, including full bibliographic reference if possible.

38. * Current review status.
Review status should be updated when the review is completed and when it is published. For new registrations the review must be Ongoing.
Please provide anticipated publication date
Review_Completed_not_published

39. Any additional information.
Provide any other information the review team feel is relevant to the registration of the review.

40. Details of final report/publication(s) or preprints if available.
Appendix 1.2: Search String

Search strings in each data base.

<table>
<thead>
<tr>
<th>Database</th>
<th>Search String</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eric</td>
<td>((executive function) OR (problem solving) OR (decision making) OR (inhibit**) OR (distract*) OR (Attention) OR (impulse control) OR (Mental* Flexib*) OR (set shifting) OR (working memory) OR (recall*) OR (planning) OR (Switch) OR (Self Regulat*)) AND ((Down Syndrome) OR (Syndrome. Down) OR (Downs Syndrome) OR (Down’s Syndrome) OR (Syndrome. Down's) OR (Syndrome. Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (Translocation) OR (Intellectual* Disab*) OR (Intellectual* Impair*) OR retard*)) AND ((Child*) OR (Adult*))</td>
</tr>
<tr>
<td>ASSIA</td>
<td>((Executive function) OR (problem Solving) OR (Decision making) OR (Inhibit*) OR (Distract*) OR (attention) OR (Impulse control) OR (Mental* Flexib*) OR (set shifting) OR (Working memory) OR (Recall*) OR (Planning) OR (Switch) OR (Self Regulat*)) AND ((Down Syndrome) OR (Syndrome, Down) OR (Down's Syndrome) OR (Syndrome, Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (translocation) OR (intellectual* disab*) OR (Intellectual* Impair*) OR (Retard*)) AND ((Child*) OR (Adult*))</td>
</tr>
<tr>
<td>COCHRANE</td>
<td>((Executive function) OR (problem Solving) OR (Decision making) OR (Inhibition ) OR (Distraction) OR (attention) OR (Impulse control) OR (Mental Flexibility) OR (set shifting) OR (Working memory) OR (Recall) OR (Planning) OR (Switch) OR (Self Regulation)) AND ((Down Syndrome) OR (Syndrome, Down) OR (Down's Syndrome) OR (Syndrome, Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (translocation) OR (intellectual* disab*) OR (Intellectual* Impair*) OR (Retard*)) AND ((Child*) OR (Adult*))</td>
</tr>
<tr>
<td>Database</td>
<td>Query</td>
</tr>
<tr>
<td>-------------------</td>
<td>----------------------------------------------------------------------</td>
</tr>
<tr>
<td>MEDLINE</td>
<td>((Executive Function) OR (planning) OR (Problem Solving) OR (Decision Making) OR (inhibition) OR (Distract*) OR (Attention) OR (Impulse control) OR (mental* adj3 Flexib*) OR (set shifting) OR (working memory) OR (Recall**) OR (Switch) OR (Self adj3 Regulat*)) AND ((Down Syndrome) OR (Syndrome, Down) OR (Down's Syndrome) OR (Downs Syndrome) OR (Syndrome, Down's) OR (Syndrome, Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (Translocation, Genetic) OR (Intellectual Disability) OR (Intellectual* Impair*) OR (retard*)) AND ((Child*) OR (Adult*))</td>
</tr>
<tr>
<td>Web OF Science</td>
<td>(((Executive function) OR (problem solving) OR (decision making) OR (Inhibit*) OR (Distact*) OR (attention) OR (Impulse control) OR (Mental* Flexib*) OR (set shifting) OR (Working memory) OR (Recall) OR (Planning) OR (Switch) OR (Self Regulat*)) AND ((Down Syndrome) OR (Syndrome, Down) OR (Down's Syndrome) OR (Downs Syndrome) OR (Syndrome, Down's) OR (Syndrome, Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (translocation) OR (Intellectual* Disab*) OR (Intellectual* Impair*) OR (Retard*)) AND ((Child*) OR (Adult*))</td>
</tr>
<tr>
<td>PsycInfo</td>
<td>(Executive function) OR (problem solving) OR (decision making) OR (Inhibit*) OR (Switch*) OR (Impulse control) OR (mental* adj3 Flexib*) OR (distraction) OR (set shifting) OR (working memory) OR (Recall*) OR (planning) OR (Self-Regulation) OR (Attention)) AND ((Down Syndrome) OR (Syndrome, Down) OR (Down's Syndrome) OR (Downs Syndrome) OR (Syndrome, Down's) OR (Syndrome, Downs) OR (Trisomy 21) OR (chromosome 21) OR (Mosaicism) OR (Translocation, Genetic) OR (Intellectual Disability) OR (Intellectual* Impair*) OR (retard*) AND ((Child*) OR (Adult*))</td>
</tr>
</tbody>
</table>
(chromosome 21) OR (Mosaicism) OR (translocation) OR (Intellectual* Disab*) OR (Intellectual* Impair*) OR (retard*)) AND ((Child*) OR (Adult*))

From the table above it can be seen that the keywords for search strategies are similar, but the way of searching was different between databases. For example, most databases could be searched using the key words of EF inhibition or mental flexibility by adding (*) at the end of the word to allow me to gather the most possible results, such as (inhibit**) OR (Mental* Flexib*). However, this is not the case in, for example, the COCHRANE Library, where I had to insert the full keyword as shown in the table above. Moreover, databases like PsycInfo required a specific way of searching; for example, the keyword for mental flexibility function had to be searched for differently from the other databases - it required me to input (mental* adj3 Flexib*) (for more details see the table).
Appendix 2.1: Informed Consent Forms

*Ethical Approval (children, teachers and parents)*

Child Consent Form

Hello! My name is Ghaleyah.

I would like to come and see you at school.

You would:

- Talk to me
- Read and write
- Play games

Is it okay?

Yes

No
Working memory intervention with teachers of children with Down syndrome

About the researcher:
This research is being carried out by researcher Ghaleyah Alajmi under the supervision of Drs Katie Cebula, Josie Booth and Gale Macleod (University of Edinburgh, UK). The study is being completed as part of Ghaleyah’s PhD degree at the University of Edinburgh. Ghaleyah has been a social worker for more than 15 years. She works with both typically developing children and children with special needs, such as children with intellectual disabilities and children with Down syndrome. She has considerable experience in working with children with Down syndrome, their families and their teachers, as these are among the most important requirements in her job.

About the research:
Children with Down syndrome may experience difficulties with cognitive abilities that affect learning. One such area is memory, a skill which is essential for mastering school tasks and life skills. This research focuses on children’s working memory. This type of memory is responsible for short-term storage and manipulation of information. There are several teaching methods for children with intellectual disabilities which focus on supporting working memory. Support for working memory should help pupils’ learning. In this study, I will not provide intervention directly to the children but will instead present an intervention to teachers of children with Down syndrome. This will consist of providing weekly workshops over 6 to 8
weeks, to share with teachers methods to support working memory and academic achievement in children with Down syndrome.

Very little research has looked at the best ways to help teachers support working memory in their pupils with Down syndrome, and no study has yet been conducted to explore the best ways to teach children with Down syndrome in Kuwait. Most research investigating working memory in children with Down syndrome has focused directly on the children themselves, not on the techniques used by their teachers. That is why this research aims to work with teachers, to explore some ways of supporting working memory in children with Down syndrome.

Who can take part in this research?
I am looking for teachers who teach literacy and numeracy to children with Down syndrome, and for children with Down syndrome who are studying a curriculum for students with Down syndrome (aged from 8 to 15 years).

What will teachers be asked to do?
All teachers will be asked to

- participate in the intervention (from 6 to 8 weeks) by attending one workshop a week, lasting 45 to 60 minutes. They will learn certain principles and strategies to support their students' working memory with the aim of improving academic achievement.
- complete a questionnaire about working memory before and after the intervention.
- complete questionnaires about their students that evaluates behaviour and cognitive ability (such as executive function). Executive functions are a set of cognitive abilities, including working memory, that are important for reaching goals. These questionnaires should take no more than 30 minutes to complete per student.
• consider implementing some of the principles and strategies that they learn in the workshops.
• participate in audio-recorded group interviews to discuss their views about the intervention.
• agree to be observed before and after the intervention, with the aim of focusing on the efficiency of the intervention.

What about ethics and confidentiality?
The research involves collection of some personal information about you (e.g. your professional qualifications, age, name and level of class that you teach), provided through questionnaire.

This information will be stored securely, according to the University of Edinburgh Data Protection Policy, and only the researcher and her supervisors will have access to it. We will only share personal information with others if we have a serious concern that you or someone else is at risk of harm. You will not be identified by name in any PhD thesis, research report, conference presentation, journal article nor other academic publication, as all data will be anonymised. The research has been approved by the University of Edinburgh ethics committees.

What should I do if I have concerns during the research?
In the first instance, please contact Ghaleyah Alajmi (94945949). If your concerns are not resolved, please contact the Educational Research Administration in Kuwait Ministry of Education (25417942). If you wish to contact Ghaleyah’s PhD supervisors and are able to do so in English, please contact her supervisor, by email: (Katie.Cebula@ed.ac.uk) or tel: 44 131 651 6463

Is there any obligation to take part?
We would very much appreciate your involvement, but you are not obliged to take part. You can also change your mind at any time and leave the study.

What are the benefits of participating? We will send you a ‘thank you’ gift token for participating, and you will also be sent a summary of the final report.

We also hope that teachers will find it interesting to share their experiences. The research aim is to benefit teachers and students with Down syndrome in the future, by identifying the best ways to support them.

How can we take part in this research? If you agree to take part in this research, please complete the attached consent form and questionnaire.
Teacher consent form

Working memory intervention with teachers of children with Down syndrome

Please Tick (only if you agree)

I have read and understood the information provided. □

I would be happy to take part in the study to learn new approaches to support working memory in children with Down syndrome. □

I am happy to be observed and interviewed (audio-recorded) for the research. □

I understand that participants will not be individually identified in any way in this research and that all publications resulting from this research, such as PhD thesis, conference presentation, journal article will only contain completely anonymised data. □

I understand that anonymised data will be stored indefinitely, but all personal data and audio recordings of interviews will be securely destroyed five years after the research is fully completed and no individual participants will be named in any research reports. □

________________________________________
Name (Printed)

________________________________________
Signature

________________________________________
Date

=======================================================================================================

Note: this information was translated into Arabic
Positive behaviour support intervention with teachers of children with Down syndrome?

About the researcher
This research is being carried out by researcher Ghaleyah Alajmi under the supervision of Drs Katie Cebula, Josie Booth and Gale Macleod (University of Edinburgh, UK). The study is being completed as part of Ghaleyah’s PhD degree at the University of Edinburgh. Ghaleyah has been a social worker for more than 15 years. She works with both typically developing children and children with special needs, such as children with intellectual disabilities and children with Down syndrome. She has considerable experience in working with children with Down syndrome, their families and their teachers, as these are among the most important requirements in her job.

About the research:
Some children with Down syndrome sometimes show ‘challenging behaviour’, such as temper tantrums or shouting out in class, which can make learning difficult. However, not all children show this behaviour. Teachers of children with Down syndrome sometimes face difficulties in managing students’ challenging behaviour.

No study has yet been conducted to explore the best ways to work with teachers to reduce the likelihood of challenging behaviour in children with Down syndrome in Kuwait. This research aims to explore the use of an approach known as Positive Behaviour Support (PBS), a multicomponent framework used to understand which contexts and factors maintain an individual's challenging
behaviour. The researcher will provide a multicomponent framework used to understand which contexts and factors maintain an individual's challenging behaviour. Weekly workshops over 6 to 8 weeks, to explore PBS with teachers, with the intention that teachers will adapt what they learn for use in their own classroom teaching.

Who can take part in this research?
I am looking for teachers who teach literacy and numeracy to children with Down syndrome, and for children with Down syndrome who are studying a curriculum for students with Down syndrome (aged from 8 to 15 years).

What will teachers be asked to do?
All teachers will be asked to

- participate in the intervention (from 6 to 8 weeks) by attending one workshop a week, lasting 45 to 60 minutes. They will learn positive behaviour support approaches for children with challenging behaviour.
- complete a questionnaire about challenging behaviour before and after the intervention.
- complete questionnaires about their students that evaluates behaviour and cognitive ability (such as executive function). Executive functions are a set of cognitive abilities, including working memory, that are important for reaching goals. These questionnaires should take no more than 30 minutes to complete per student. This will allow some exploration of associations between these abilities and students’ behaviour.
- consider teaching students according to the new principles and strategies that they learn in the workshops.
- agree to be observed before and after the intervention, with the aim of focusing on the efficiency of the intervention.
What about ethics and confidentiality?
The research involves collection of some personal information about you (e.g. your professional qualifications, age, name and level of class that you teach), provided through questionnaire.

This information will be stored securely, according to the University of Edinburgh Data Protection Policy, and only the researcher and her supervisors will have access to it. We will only share personal information with others if we have a serious concern that you or someone else is at risk of harm. You will not be identified by name in any PhD thesis, research report, conference presentation, journal article nor other academic publication, as all data will be anonymised. The research has been approved by the University of Edinburgh ethics committees.

What should I do if I have concerns during the research?
In the first instance, please contact Ghaleyah Alajmi (94945949). If your concerns are not resolved, please contact the Educational Research Administration in Kuwait Ministry of Education (25417942). If you wish to contact Ghaleyah’s PhD supervisors and are able to do so in English, please contact her supervisor, by email: (Katie.Cebula@ed.ac.uk) or tel: 44 131 651 6463

Is there any obligation to take part?
We would very much appreciate your involvement, but you are not obliged to take part. You can also change your mind at any time and leave the study.

What are the benefits of participating?
We will send you a ‘thank you’ gift token for participating, and you will also be sent a summary of the final report.
We also hope that teachers will find it interesting to share their experiences. The research aim is to benefit teachers and students with Down syndrome in the future, by identifying the best ways to support them.

How can we take part in this research?
If you agree to take part in this research, please complete the attached consent form and questionnaire.
Teacher Consent Form for PBS Intervention

Positive behaviour support intervention with teachers of children with Down syndrome

Please Tick (only if you agree)

I have read and understood the information provided.   

I would be happy to take part in the study to learn new approaches to support positive behaviour in children with Down syndrome.   

I am happy to be observed for the research.   

I understand that participants will not be individually identified in any way in this research and that all publications resulting from this research, such as PhD thesis, conference presentation, journal article will only contain completely anonymised data.  

I understand that anonymised data will be stored indefinitely, but all personal data and audio recordings of interviews will be securely destroyed five years after the research is fully completed and no individual participants will be named in any research reports.   

__________________________________________
Name (Printed)

__________________________________________    _________________________________
Signature                                      Date

Note: this information was translated into Arabic
Parent research information sheet and consent forms for two interventions.

Working memory intervention with teachers of children with Down syndrome

About me:
This research is being carried out by researcher Ghaleyah Alajmi under the supervision of Drs Katie Cebula, Gale Macleod and Josie Booth (University of Edinburgh, UK). The study is being completed as part of Ghaleyah’s PhD degree at the University of Edinburgh. Ghaleyah has been a social worker for more than 15 years. She works with both typically developing children and children with special needs, such as children with intellectual disabilities and children with Down syndrome. She has considerable experience in working with children and their families, as this is one of the most important requirements in her job.

About the research:
Children with Down syndrome may experience difficulties with cognitive abilities that affect learning. One such area is memory, a skill which is essential for mastering school tasks and life skills. This research focuses on children's working memory. This type of memory is responsible for short-term storage and manipulation of information. There are several teaching methods for children with intellectual disabilities which focus on supporting working memory. Support for working memory should help pupils' learning. In this study, I will not provide intervention directly to your
child, but will instead present an intervention to teachers of children with Down syndrome. This will consist of providing weekly workshops over 6 to 8 weeks, to share with teachers methods to support working memory and academic achievement in children with Down syndrome.

Very little research has looked at the best ways to teach children with Down syndrome or explored ways of helping teachers support working memory in pupils with Down syndrome, and no study has yet been conducted to explore the best ways to teach children with Down syndrome in Kuwait. That is why this research aims to work with teachers, to explore some ways of supporting working memory in children with Down syndrome.

Who can take part in this study?
I am looking for children with Down syndrome (aged from 8 to 15 years) who are following a curriculum for students with Down syndrome. Please give your child the ‘Young Person’s Information & Consent’ sheet and / or discuss the study with them in a way they would understand – whatever is best for you and your child.

What will my child and I be asked to do?
The study involves carrying out assessments of the cognitive abilities and academic achievement of children with Down syndrome, before and after the intervention. Each assessment takes 10 to 30 minutes and will be done on separate days. The children will be asked to spend around 60 minutes on assessments in total. There will also be several short observations of the teachers interacting with children in class. This will all be done at a time that is convenient for your child and their teacher.

Parents will be asked to complete a questionnaire pack. This contains questions about their child, and their child’s schooling, memory and thinking skills. It should take no more than 30 minutes to complete.

What about ethics and confidentiality?
The research involves collecting some personal information about your child (e.g. your child’s diagnosis). This information will be stored securely, according to the University of Edinburgh Data Protection Policy, and only the researcher and her supervisors will have access to it. We will only share personal information with others if we have a serious concern that your child or someone else is at risk of harm. You and your child will not be identified by name in any research report. The research has been approved by the University of Edinburgh ethics committees.

What should I do if I have concerns during the research?
In the first instance, please contact Ghaleyah Alajmi (94945949). If your concerns are not resolved, please contact the Educational Research Administration in Kuwait Ministry of Education (25417942). If you wish to contact Ghaleyah’s PhD supervisors and are able to do so in English, please contact her supervisor, by email: (Katie.Cebula@ed.ac.uk ) or tel : 44 131 651 6463

Is there any obligation to take part?
We would very much appreciate your taking part, but neither you nor your child are obliged to do so. If you do decide to participate but then change your mind, you can leave the study at any time.

What are the benefits of participating?
We will send your child a small ‘thank you’ gift token for participating, and you and your child will be sent a summary of the final report.
The overall aim of the research is to benefit children with Down syndrome in the future, by identifying the best ways to support them in the classroom.

How can we take part in this research?
If you agree to take part in this research, please complete the attached consent form and questionnaire, making sure to sign the consent form, and return them to your child’s school / teacher.
If you have any questions, please e-mail gphnm@yahoo.com, or phone number (94945949)
Parent consent form: Working memory intervention for teachers of children with Down syndrome

Please Tick (if you agree)

I have read and understood the information provided. [ ]

I am happy for my child to take part in the study. [ ]

I am happy for my child’s assessment results to be saved by the researcher. [ ]

I am happy to complete questionnaires about my child. [ ]

_________________________________________  ___________________________________________
Parent’s name (Printed)  Child’s name (Printed)

_________________________________________  ___________________________________________
Parent’s signature  Date

_________________________________________  ___________________________________________
Name of person obtaining consent (Printed)  Signature of person obtaining consent

_________________________________________
Telephone number

_________________________________________

Note: this information was translated to Arabic.
Positive Behaviour Support intervention with teachers of children with Down syndrome

About me:
This research is being carried out by researcher Ghaleyah Alajmi under the supervision of Drs Katie Cebula, Gale Macleod and Josie Booth (University of Edinburgh, UK). The study is being completed as part of Ghaleyah’s PhD degree at the University of Edinburgh. Ghaleyah has been a social worker for more than 15 years. She works with both typically developing children and children with special needs, such as children with intellectual disabilities and children with Down syndrome. She has considerable experience in working with children and their families, as this is one of the most important requirements in her job.

About the research:
Some children with Down syndrome sometimes show ‘challenging behaviour’, such as temper tantrums or shouting out in class, which can make learning difficult. However, not all children show this behaviour. The intervention described here is not intended to address any issues associated with any individual child, but is simply an attempt to investigate behaviour management techniques for children with Down syndrome in general.
In this study the researcher will not provide intervention directly to your child but will instead present an intervention to ALL teachers of children with Down syndrome in your child’s school. This will consist of providing weekly workshops over 6 to 8 weeks, to explore with teachers the use of techniques to manage students’ behaviour known as Positive Behaviour Support (PBS), a behaviour
management system used to understand what maintains an individual's challenging behaviour.

No study has yet been conducted to explore the best ways to manage behaviour in children with Down syndrome in Kuwait. That is why this research aims to work with teachers, to explore some ways to modify and manage behaviour of children with Down syndrome.

Who can take part in this study?
I am looking for children with Down syndrome (aged from 8 to 15 years) who are following a curriculum for students with Down syndrome. If you are happy for your child to participate in the study, please give your child the ‘Young Person’s Information & Consent’ sheet and / or discuss the study with them in a way they would understand – whatever is best for you and your child

What will my children and I be asked to do?
The study involves carrying out assessments of the cognitive abilities and academic achievement of children with Down syndrome, before and after the intervention. Each assessment takes 10 to 30 minutes and will be done on separate days. The children will be asked to spend around 60 minutes on assessments in total. There will also be several short observations of the teachers interacting with children in class. This will all be done at a time that is convenient for your child and their teacher.
Parents will be asked to complete a questionnaire pack. This contains questions about their child, and their child’s schooling, memory and thinking skills. It should take no more than 30 minutes to complete.

What about ethics and confidentiality?
The research involves collecting some personal information about your child (e.g. your child’s diagnosis). This information will be stored securely, according to the University of Edinburgh Data Protection Policy, and only the researcher
and her supervisors will have access to it. We will only share personal information with others if we have a serious concern that your child or someone else is at risk of harm. You and your child will not be identified by name in any research report. The research has been approved by the University of Edinburgh ethics committees.

What should I do if I have concerns during the research?
In the first instance, please contact Ghaleyah Alajmi (94945949). If your concerns are not resolved, please contact the Educational Research Administration in Kuwait Ministry of Education (25417942). If you wish to contact Ghaleyah’s PhD supervisors and are able to do so in English, please contact her supervisor, by email: (Katie.Cebula@ed.ac.uk) or tel : 44 131 651 6463

Is there any obligation to take part?
We would very much appreciate your taking part, but neither you nor your child are obliged to do so. If you do decide to participate but then change your mind, you can leave the study at any time.

What are the benefits of participating?
We will send your child a small ‘thank you’ gift token for participating, and you and your child will be sent a summary of the final report. The overall aim of the research is to benefit children with Down syndrome in the future, by identifying the best ways to support them in the classroom.

How can we take part in this research?
If you agree to take part in this research, please complete the attached consent form and questionnaire, making sure to sign the consent form, and return them to your child’s school / teacher.
If you have any questions, please e-mail gphnm@yahoo.com, or phone number (94945949)
Parent consent form:

Positive Behaviour Support intervention with teachers of children with Down syndrome

Please Tick (if you agree)

I have read and understood the information provided. □

I am happy for my child to take part in the study. □

I am happy for my child’s assessment results to be saved by the researcher. □

I am happy to complete questionnaires about my child. □

________________________________________________________
Parent’s name (Printed) Child’s name (Printed)

________________________________________________________
Parent’s signature Date

________________________________________________________
Name of person obtaining consent (Printed) Signature of person obtaining consent

________________________________________________________
Telephone number

Note: this information was translated to Arabic.
Appendix 2.2: Teacher Knowledge and Demographics Questionnaire

This was translated into Arabic for the teachers.

The questions in this section are for background information purposes only.

What do you teach?

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Literacy</td>
</tr>
<tr>
<td>2</td>
<td>Numeracy</td>
</tr>
</tbody>
</table>

How many lesson do you teach per week?

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td></td>
</tr>
<tr>
<td>More, and if so how many?</td>
<td></td>
</tr>
</tbody>
</table>

At which stage do you teach? ‘Please tick all that apply’

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>First</td>
</tr>
<tr>
<td>2</td>
<td>Second</td>
</tr>
<tr>
<td>3</td>
<td>Third</td>
</tr>
<tr>
<td>4</td>
<td>Fourth</td>
</tr>
<tr>
<td>5</td>
<td>Fifth</td>
</tr>
</tbody>
</table>

At what level is your teaching qualification?

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Graduate</td>
</tr>
<tr>
<td>2</td>
<td>Post-graduate</td>
</tr>
</tbody>
</table>
Do you have any qualification, certificate or training related especially to Down syndrome?

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>No</td>
</tr>
</tbody>
</table>

If so, can you mention the content and duration of the courses?

For how many years have you been teaching since you graduated?

How long have you been teaching pupils with special needs?

If you have taught pupils with special needs, what sort of special needs did they have? (You can choose more than one.)

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Physical impairment</td>
</tr>
<tr>
<td>2</td>
<td>Intellectual disability not Down syndrome</td>
</tr>
<tr>
<td>3</td>
<td>Down syndrome</td>
</tr>
<tr>
<td>4</td>
<td>Autism</td>
</tr>
<tr>
<td>5</td>
<td>Sensory impairment</td>
</tr>
<tr>
<td>Other</td>
<td></td>
</tr>
</tbody>
</table>

In the following section, please tick the response that best describes your current level of knowledge of each topic (you should not be concerned if some terms are unfamiliar – the questions are simply to gauge current familiarity in order to help you to plan the intervention and assess its impact.).
<table>
<thead>
<tr>
<th>Question</th>
<th>not at all confident</th>
<th>little confident</th>
<th>neither</th>
<th>somewhat confident</th>
<th>very confident</th>
</tr>
</thead>
<tbody>
<tr>
<td>How confident do you feel in your knowledge about</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the cognitive abilities of children with intellectual disabilities?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the profile of cognitive abilities of children with Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the term ‘executive function’?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>‘executive function’ in Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>different types of memory? (long term, short term, and working memory)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>memory in Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>working memory?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>working memory in Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the role of working memory in reading in Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
the role of working memory in numeracy in Down syndrome?

strategies that teachers can use to support working memory of pupils?

working memory strategies that children can use to support their own working memory?

working memory interventions for children?

<table>
<thead>
<tr>
<th>Question</th>
<th>High Priority</th>
<th>Fairly Priority</th>
<th>Neutral Priority</th>
<th>Not very priority</th>
<th>Not at all priority</th>
</tr>
</thead>
<tbody>
<tr>
<td>To what extent is learning about memory in children with Down syndrome a priority for you at the moment?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Question</td>
<td>not at all confident</td>
<td>little confident</td>
<td>neither</td>
<td>somewhat confident</td>
<td>very confident</td>
</tr>
<tr>
<td>------------------------------------------------------------------------</td>
<td>----------------------</td>
<td>------------------</td>
<td>---------</td>
<td>--------------------</td>
<td>----------------</td>
</tr>
<tr>
<td>How confident do you feel in your knowledge about the challenging behaviour of children with intellectual disabilities?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the challenging behaviour of children with Down syndrome?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>different forms of challenging behaviour? (Stubbornness, Social withdrawal, Aggressive, Noncompliance, etc.)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>the effect of challenging behaviour on learning in Down</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
syndrome?

strategies that teachers can use to support reductions in challenging behaviour?

‘Positive Behaviour Support’ in Down syndrome?

‘Positive Behaviour Support’ interventions for children with Down syndrome?

<table>
<thead>
<tr>
<th>Question</th>
<th>High Priority</th>
<th>Fairly Priority</th>
<th>Neutral Priority</th>
<th>Not very priority</th>
<th>Not at all priority</th>
</tr>
</thead>
<tbody>
<tr>
<td>To what extent is learning about Positive Behaviour Support in children with Down syndrome a priority for you at the moment?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Is there anything else you would like to add about teaching children with Down syndrome, or areas of teaching you would like to develop?

............................................................................................................................................................................................
............................................................................................................................................................................................
............................................................................................................................................................................................
......
Appendix 2.3: Pupil Demographics Questionnaire

Date of birth: _____________ 2. Gender: Male/Female

3. Child’s ethnicity?:
   - Arab
   - Non-Arab

4. Child’s nationality __________________________

5- Does the child have any of the following? Please tick

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>No</th>
<th>Yes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epilepsy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Visual impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hearing impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mobility difficulties</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac (heart) problems</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypertension (high blood pressure)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kidney/bladder problems</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dental anomalies</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gastrointestinal issues/coeliac disease</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endocrine (thyroid) abnormalities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Slow growth rate/feeding problems</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypersensitivity (e.g. to noise)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sleep difficulties</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other diagnosis, please specify:</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Other diagnosis, please specify:
Appendix 2.4: BRIEF-P Subscales Reliability Details

Table 1: Cronbach’s alpha for BRIEF-P items for parents

<table>
<thead>
<tr>
<th>Variable</th>
<th>Cronbach’s Alpha</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inhibition pre intervention</td>
<td>.834</td>
</tr>
<tr>
<td>Inhibition follow-up intervention</td>
<td>.903</td>
</tr>
<tr>
<td>Shift pre intervention</td>
<td>.865</td>
</tr>
<tr>
<td>Shift follow-up intervention</td>
<td>.851</td>
</tr>
<tr>
<td>Emotional control pre intervention</td>
<td>.785</td>
</tr>
<tr>
<td>Emotional control follow-up intervention</td>
<td>.845</td>
</tr>
<tr>
<td>Working memory pre intervention</td>
<td>.937</td>
</tr>
<tr>
<td>Working memory follow-up intervention</td>
<td>.931</td>
</tr>
<tr>
<td>Plan organize pre intervention</td>
<td>.873</td>
</tr>
<tr>
<td>Plan organize follow-up intervention</td>
<td>.879</td>
</tr>
<tr>
<td>Global executive function pre intervention</td>
<td>.956</td>
</tr>
<tr>
<td>Global executive function follow-up intervention</td>
<td>.965</td>
</tr>
</tbody>
</table>

Table 2: Cronbach’s alpha for BRIEF-P items for teachers

<table>
<thead>
<tr>
<th>Variable</th>
<th>Cronbach’s Alpha</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inhibition pre intervention</td>
<td>.894</td>
</tr>
<tr>
<td>Inhibition follow-up intervention</td>
<td>.850</td>
</tr>
<tr>
<td>Shift pre intervention</td>
<td>.880</td>
</tr>
<tr>
<td>Shift follow-up intervention</td>
<td>.866</td>
</tr>
<tr>
<td>Emotional control pre intervention</td>
<td>.842</td>
</tr>
<tr>
<td>Emotional control follow-up intervention</td>
<td>.826</td>
</tr>
<tr>
<td>Working memory pre intervention</td>
<td>.937</td>
</tr>
<tr>
<td>Working memory follow-up intervention</td>
<td>.922</td>
</tr>
<tr>
<td>Plan organize pre intervention</td>
<td>.847</td>
</tr>
<tr>
<td>Plan organize follow-up intervention</td>
<td>.778</td>
</tr>
<tr>
<td>Global executive function pre intervention</td>
<td>.964</td>
</tr>
<tr>
<td>Global executive function follow-up intervention</td>
<td>.952</td>
</tr>
</tbody>
</table>
Appendix 2.5: Literacy and Numeracy Assessments

Whilst each class stage had their own assessment, and whilst assessments varied somewhat from school to school, the examples below provide a flavour of the typical contents of these assessments. These examples are from classes 1 and 5, to illustrate the differences at these different stages.

Literacy Class 1 Assessment

(The first left) The first question presents two pictures and the child had to identify ‘Father’ and ‘Mother’.

(The Second left) The question presents one letter with different shapes that create different sounds, and asked the child to match the letter with the sound.
The child was asked to draw the word ‘I’.

(Above) The child was asked to match the picture with the correct letter that the animal in the picture starts with. The child was then asked to write the letter three times.
(Above) The child is presented with two similar words and they are asked to match them. One word means ‘peace’, one means ‘be on you’.

(Above) The child was asked to draw inside the word (which means ‘mercy’) to include the missed letter.

There were then three words and three pictures, and the child was asked to circle the letter ‘h’ in each word (in Arabic): Cat; Arrow; Face

(Left) The question asked the child whether the following act was right or wrong:

1. Blowing on cup – wrong in Kuwait.
2. How to eat – need to say grace before eating.
3. This is my personal ID (this encourages learning of life skills).
This question asked the child to circle the number of apples they were instructed to in the figure. The figure in the first line is ‘3’, the figure in the second line is 5.

The next question asked the child to count and write the number of strawberries in each box (from right to left this should be 1, 4 and 2).

Under the empty square, there is a number (3, then 4). The child was asked to draw anything the number of times corresponding with the figure.

The next question asked the child to write a mathematical equation for the number of cats (from left to right, $2+1=3$, $2+2=4$).

The child was then asked to continue the number (1, 2 ...)
The child was asked to repeat the number they saw in the box above, to test their writing ability for numbers.
Numeracy Class 5 Assessment

(Left) - The question asked the child to circle the correct number of pictures (from the 10s and 1s) to add up to the correct figure (shown on the right). In this example the figure is 43.
- This question was the same as the first question, with the figure of 49.
- The third question asked the child to count and write the number of pebbles (10s and 1s). In this question, the correct answer is 25.

(Left) - The question asked the child to match the right number (in the box) with the right picture. The first number is 18, the second number is 72.
The second question asked the child to add the money together (also a daily life skill). The correct answer is 20 Kuwaiti dinars.
(Left) The question asked the child to write the correct number of ‘ones’ on the right and the ‘tens’ on the left. The second question on this page gave the number 22 and asked the child to draw the tens and draw the ones. The third question gave the number 25 and the child was asked to complete the picture by drawing the remaining sticks (5).
Appendix 2.6 Observation Form

<table>
<thead>
<tr>
<th>Child Behaviour</th>
<th>Teacher No:</th>
<th>Date:</th>
<th>Time:</th>
<th>School:</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. <strong>Failure to follow instruction</strong>&lt;br&gt;Child does not do what the teacher has asked within the time of the task. Also include partial failure (e.g. teacher gives 3 steps and child only completes 1).</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. <strong>Incomplete recall</strong>&lt;br&gt;The child either states that they do know and then then says that they have forgotten OR starts to give an answer and says they have forgotten or then trails off.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. <strong>Place keeping error</strong>&lt;br&gt;In a verbal task the child incorrectly repeats or skips an element.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. <strong>Task abandonment</strong>&lt;br&gt;Child stops doing task before it is complete without instruction from the teacher to stop.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Teacher Behaviours

<table>
<thead>
<tr>
<th>Teacher Behaviour</th>
<th>Teacher No:</th>
<th>Date:</th>
<th>Time:</th>
<th>School:</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Teacher responds to child showing possible working memory difficulty&lt;br&gt;(A) Talks to the child about what they are doing, whether they require help, asks whether they need an instruction repeated&lt;br&gt;(B) Looks at the child’s written work</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Teacher reduces distraction&lt;br&gt;Teacher does one of the following:&lt;br&gt;(A) Removes material unrelated to the task from the child’s desk, the blackboard, or the noticeboard.&lt;br&gt;(B) Reminds pupils not to talk&lt;br&gt;(C) Posts a “Do Not Disturb” sign on her classroom door</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Teacher uses repetition&lt;br&gt;Teacher repeats a statement, questions or instruction within the same task. This can be a full repetition or partial (e.g. same meaning but a reduced number of words).</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Teacher uses A. rhythm and B. rhyme&lt;br&gt;Teacher uses song (recording or their own voice) or makes a rhythm (e.g. clapping or tapping) or uses rhyming words.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Teacher uses visual memory aids&lt;br&gt;Teacher shows the pupil a A. picture, B. diagram, C. video, D. symbols, E. writing, or F. gesture which is either a clear stand-alone statement, questions, or instruction OR it reinforces a verbal statement, question or instruction.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Teacher uses chunking&lt;br&gt;Teacher: (A) gives two or more statements, questions, or instructions as separate sequential units (rather than run together using connectives).&lt;br&gt;(B) gives a list of items (numbers, words) either semantically grouped, or with pauses to indicate grouping.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Teacher encourages student to use working memory strategies&lt;br&gt;The teacher:&lt;br&gt;(A) Tells/asks the child to rehearse or repeat information (could be out loud or silently)&lt;br&gt;(B) Tells/asks the child to use/create visual memory aids&lt;br&gt;(C) Tells/asks the child to request if they need help remembering</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Teacher evaluates working memory load of task by asking child&lt;br&gt;The teacher asks the child if the task is too difficult to remember.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Teacher reduces task working memory load&lt;br&gt;Teacher simplifies a previously used statement/question/instruction in a manner not already covered above (e.g. A. chunking, B. repetition, C. visual memory aid).</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Child (CH)</th>
<th>CH 1</th>
<th>CH 2</th>
<th>CH 3</th>
<th>CH 4</th>
<th>CHS</th>
<th>CH6</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>A</th>
<th>B</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>A</th>
<th>B</th>
<th>A</th>
<th>B</th>
<th>C</th>
</tr>
</thead>
</table>

**NOTE:**
Appendix 2.7: Focus Group Prompt Questions

Focus Group Questions

How did you find the intervention?

Prompts:
What did you think of the format of the intervention, with the weekly sessions?
How did you find the time commitment?
Do you think that the intervention (strategies) had any effect on the working memory outcomes for pupils with DS?
Did you find any advantages in this intervention? If so can you mention them?
Did you find any drawbacks in this intervention? If so can you mention them?
How do you think the drawbacks you mentioned could be avoided?
Is there anything else (like the materials, methods of delivery, schedule or duration) that you would like to see included the intervention? If so, can you mention it?

Do you think the intervention changed your knowledge of working memory or did it not have much impact?

Prompts:
Do you think that the intervention made any difference to your knowledge about cognitive abilities in Down syndrome in general and about working memory in particular? If so, can you explain what you have learnt?
Do you think that the intervention did not make any difference to your knowledge about cognitive abilities in Down syndrome in general and about working memory in particular? If so, is this because you were already familiar with research into WM, or already aware of these strategies? Or is this because you felt sceptical about the effectiveness of the strategies covered in this intervention? Or was there any other reason why you felt that the intervention did not make any difference to your knowledge?
Do you think the intervention changed how you work with the children or not really?

From the table of strategies on your handout, which (if any) have you had a chance to use so far? What was your experience of that?

<table>
<thead>
<tr>
<th>Working memory strategies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Recognise working memory failures and responds to child showing possible working memory difficulty</td>
</tr>
<tr>
<td>2. Evaluate working memory loads.</td>
</tr>
<tr>
<td>3. Teaching strategies to support working memory:</td>
</tr>
<tr>
<td>a. Reducing task working memory load</td>
</tr>
<tr>
<td>b. Reducing distraction</td>
</tr>
<tr>
<td>c. Repetition</td>
</tr>
<tr>
<td>d. Rhythm and rhyme</td>
</tr>
<tr>
<td>e. Memory aids</td>
</tr>
<tr>
<td>f. Chunking</td>
</tr>
<tr>
<td>g. Develop the child’s own strategies to support working memory:</td>
</tr>
<tr>
<td>i. Request help or repetition</td>
</tr>
<tr>
<td>ii. Note-taking.</td>
</tr>
<tr>
<td>iii. Rehearsing information</td>
</tr>
</tbody>
</table>

Were any particularly effective or ineffective?

Do you think that the intervention strategies had any effect on the outcomes for pupils with DS?

*Prompts:*
If you think that the intervention (strategies) had any effect on the working memory outcomes for pupils with DS? If so, can you explain further what changes you observed?
Did you feel the intervention (and the strategies) had any impact on children literacy and numeracy, or not really?
Did you observe changes (either positive or negative) in any other areas (apart from working memory, literacy and numeracy)?
Are there any strategies which you already use (other than those covered in this intervention) which might lead to improvements in working memory, literacy or numeracy? If so, can you mention them? How do they compare to the strategies used in this intervention?

Summary and conclusion of the focus group session.
Appendix 2.8: Intervention Session Resources (PowerPoint)

The resources in this Appendix include the PowerPoint presentations for each of the 6 sessions in the WM intervention and the 6 sessions in the CB intervention.

Working Memory Intervention Session 1:

Session 1: Introduction

- Introduction

Session Outline

- Researcher's introduction
- Brief introduction to the intervention
- Driving questions
- Brief introduction to the research
- Teachers introduce themselves
- Memory
- Working memory
- Working memory in Down syndrome
- Working memory and learning
- Relationship between working memory and reading
- Relationship between working memory and numeracy
- Summary
- Driving questions discussion

Researcher's Introduction

Ghaleyah Alajmi

Brief Introduction to the Intervention

Session 1: Introduction
Session 2: Recognising working memory difficulties in the classroom.
Session 3: Evaluating the working memory load of taught tasks.
Session 4: Teaching strategies to support working memory in Down Syndrome.
Session 5: Developing the child's own strategies to support working memory.
Session 6: Review.

Introduction to 'Driving Questions'

- What are driving questions?
- How do we develop them?
- What purposes do they serve?
- How might they be answered?
- How will we use them in this intervention?

This question should be your response to the material that has just been presented, and it should be based on your reflections about the implications for what you have just heard for your own classroom practice. The driving question(s) could reflect either a specific challenge you are facing with a particular pupil, or a change you want to make in your own teaching strategies in order to assess its impact.

Brief Introduction to the Research

- PhD degree
- Special education
- University of Edinburgh
- Working memory and challenging behaviour in DS
  - Evaluation of an intervention in working memory and challenging behaviour, involving child assessment, teacher and parent questionnaires, teacher observation and focus groups.
  - Teachers’ input and perceptions are vital!
Teachers Introduce Themselves

- Now it is time to introduce yourselves!

What is Cognition?

Cognition is the mental action or process of acquiring knowledge and understanding through thought, experience, and the senses.
- Oxford English Dictionary

Memory

Thinking about cognitive abilities in Down syndrome...

1. What different cognitive abilities help children to learn?
2. How would you like to develop the cognitive abilities of the children with Down syndrome in your classes?

Memory is the means by which we draw on our past experiences in order to use this information in the present – Sternberg (1999)

Working Memory

Working memory is the small amount of information that can be held in mind and used in the execution of cognitive tasks, in contrast with long-term memory, the vast amount of information saved in one’s life (Cowan, 2014)

- Working memory refers to the ‘active’, current information that must be kept in mind to carry out everyday tasks, and underpins many key skills such as reading, vocabulary development and mathematics.

Working Memory

- Can think of when you used your working memory today?
- What kind of tasks did you use it for?
- When do children use their working memories in the classroom?

Activity – watch the following videos:
https://www.youtube.com/watch?v=S6D2oazr8M (0–1:37 min)
https://www.youtube.com/watch?v=F5Ehp3KVGmY (0–7:50 min)
(what WM is and how it is related to learning)
Working Memory

- What is verbal working memory?

Verbal working memory (VWM) is responsible for temporarily storing verbalizable information, such as letters, words, numbers, or nameable objects. — van Dun & Marlin (2016)

- What is visuospatial working memory?

Visuospatial working memory (VWM) is responsible for temporarily storing visual and spatial information. — Allen et al. (2019)

- What is your experience of verbal and visuospatial WM in DS?

- When do children use verbal and visuo-spatial working memory in the classroom?

Working Memory in Down syndrome

- Activity

1) Can you think of types of task where children with DS use these skills?
2) Can you talk about experiences of children with DS having difficulties with verbal and visuospatial WM?

Working Memory and Learning

Alloway & Alloway (2010): WM predicts academic achievement

- Aim: to investigate whether working memory is simply a proxy for IQ or whether there is a unique contribution to learning outcomes.

- Method: 98 children 9 years ago, tested on IC, memory and learning in the UK, and again 6 years later (11 years apart).

- Findings:
  1. Children's working memory skills at 5 years of age predicted literacy and numeracy 6 years later.
  2. Working memory is not a proxy for IQ but rather represents a dissociable cognitive skill with unique links to academic attainment.
  3. Working memory at the start of formal education is a more powerful predictor of subsequent academic success than IQ. This result has important implications for education, particularly with respect to intervention.

Relationship Between Reading and Working Memory

- WM predicted word reading and reading comprehension in typically developing children and adolescents (Christopher et al., 2012).

- WM problems are associated with language delay (Young, 2000).

Working Memory and Numeracy

- Difficulties in solving simple arithmetic calculations is because of difficulties in retrieving information from memory (Geary et al., 2000).

- Success with reading and mathematics seems to depend more on working memory than short-term memory (National Association of Special Education Teachers, 2007).
### Working Memory & Learning Summary

- Working memory capacity is an essential requisite for attaining reading and numerical skills.
- Academic performance is related to working memory capacity.
- Children with intellectual disabilities struggle with working memory.
- Practicing to improve working memory skill may therefore lead to improvements in reading and numeracy skills.

### Introduction about Working Memory difficulties in classroom

- How these difficulties affect learning: [https://www.youtube.com/watch?v=F5Ehe3KVGmY&list=FL23](https://www.youtube.com/watch?v=F5Ehe3KVGmY&list=FL23)

- [https://www.youtube.com/watch?v=waRS5oEdYluY](https://www.youtube.com/watch?v=waRS5oEdYluY)

- [https://www.youtube.com/watch?v=mwk3bIiKk8&list=FL34](https://www.youtube.com/watch?v=mwk3bIiKk8&list=FL34)

### Summary

- Working memory is an essential component of cognition that enable learning. It is related to academic achievement in all subjects but particularly literacy and numeracy.
- Working memory is often impaired in those with intellectual disabilities, including those with Down syndrome.
- Individuals with Down syndrome often have a stronger visuospatial working memory compared to verbal working memory (which might explain observable language difficulties).

### Any questions?

### Driving Questions Discussion

- Please complete the driving question, but complete the rest of the form after the session if need be.
- Please bring this sheet with you next week.
Working Memory Intervention Session 2:

WM in Down Syndrome in the Classroom

Session Outline

- Driving questions discussion
- Some signs about working memory difficulties
  - Incomplete recall
  - Failure to follow instruction
  - Place keeping errors
  - Task abandonment
- Summary
- Driving questions discussion

Driving Questions Discussion

Activity

- What does it look like when a child has working memory difficulties?
- What might they do?

Some Signs of Working Memory Difficulties

- Incomplete recall

The child forgets some or all of the information that is required to complete successful a particular task or activity.

Some Signs of Working Memory Difficulties

- Failure to follow instruction

Child does not follow multi-step instructions. The child may remember only part of the instruction (typically the first step or steps).
Some Signs of Working Memory Difficulties

- Place keeping errors

Children with poor working memory often lose track of what they have done and what has yet to be completed.

Some Signs of Working Memory Difficulties

- Task abandonment

Child gives up a task completely, often after one of the kinds of error described previously. In most cases, the task is abandoned because the child can no longer remember the information needed to guide an activity and, as a result, attention moves away from the task in hand.

Activity

Acting activity

- Which of previous signs have you seen in the class?
- Can you share some examples?

Summary

Four signs of working memory difficulties in the classroom:
1) Incomplete recall;
2) Failure to follow instruction;
3) Place keeping errors;
4) Task abandonment.

Any questions?

Driving Questions Discussion
Working Memory Intervention Session 3:

Working Memory in Children with Down Syndrome in the Classroom

Session Outline

- Driving questions discussion
- How to evaluate working memory load
- Activity
- Summary
- Driving questions discussion

Driving questions discussion

Evaluating working memory load

- Ask the child
- Observe how the child responds to the task
- Evaluate the task (with the child in mind)

How to evaluate working memory load?

1 - Ask the child
   - Evaluate the overload by asking the child in simple language whether she or he needs the teacher to repeat the question or divide the task into smaller parts.
   - This can be facilitated by use of supporting materials which allow child to communicate more easily with the teacher (e.g., a symbol card for ‘I have forgotten’).
2 - Observation
Review of signs of working memory difficulties:
- Failure to follow instruction
- Place keeping errors
- Task abandonment
- Incomplete recall

Observe
- Connect the previous signs with nature of required tasks in the class.
- Bear in mind that working memory difficulties could be related to the load placed on memory, but it should not be assumed that this is the main or only reason.

3 – Evaluate the task (with the child in mind)

Long Sequences
- Lengthy sequences that exceed a child's WM capacity will not be remembered (Gathercole & Alloway, 2008, p72).

Activity
- How many steps can you hold in working memory? Can you devise a long and shorter sequence for each other to remember?
- In your experience, how many steps can your children hold in working memory? (For example, 'Get your book, turn to page 8, circle the first sentence')
- Can you give examples of long sequences of instruction that would not work with the children in your classes? What about examples of shorter sequences which would be appropriate?

Unfamiliar and meaningless content
Low meaningfulness and high unpredictability place heavy demands on working memory, because children are not able to use their existing knowledge (in other words, long term memory) to support their performance (Gathercole & Alloway, 2008, p73).

E.g., Higher in meaningfulness: 1, 2, 3, 4, 5
Lower in meaningfulness: 8, 4, 9, 5, 7
Demanding mental process activities

The WM capacity available to support storage in an ongoing activity is directly affected by whether or not the child is also engaged in another mental activity that demands attention (Gathercole & Alloway, 2008).

E.g., less demanding activities = remembering a list
More demanding activities = remembering a list whilst counting backwards

Activity

• From your experience, what sorts of tasks do children usually fail in and do you think this is related to how meaningful or meaningless the tasks are?

Summary

Evaluating working memory load:
1) Ask the child
   - Involve them in the process
2) Observe how the child responds to the task
   - Review signs of WM difficulties
3) Evaluate the task (with the child in mind)
   - Long sequences, unfamiliar and meaningless content, demanding mental processes

Any questions?
Working Memory Intervention Session 4:

**Working Memory in Down Syndrome in the Classroom**

Session 4: Strategies to Support Working Memory in Down Syndrome

**Session Outline**
- Driving questions discussion.
- Evidence that working memory in a class can be improved.
- Working memory strategies:
  - Reducing task working memory load.
  - Reducing distraction.
  - Repetition.
  - Rhythm and rhyme.
  - Memory aids.
  - Chunking.
- Summary & driving questions discussion.

**Evidence that working memory in class could be improved**

Improving memory span in children with Down syndrome (Conners et al., 2008):
- A home-based parent-implemented intervention.
- Designed to improve auditory memory span in children with DS.
- Children improved in training sessions and a small amount on digit span.
- Digit span improvement was linked to the memory training, as indicated by control comparisons and correlations.

**Driving Questions Discussion**

**Working Memory Strategies**
- Reducing task working memory load (Task instructions)
- Reducing distraction
- Repetition
- Rhythm and rhyme
- Memory aids
- Chunking

**Reducing Working Memory Loads**

Ways to reduce working memory load
- Reduce the amount of materials to be remembered
- Increase the meaningfulness and familiarity of the materials
- Simplify mental processing and restructured complex tasks
Reducing Distraction
Ways to reduce distraction:

- Remove all materials unrelated to the task, such as sporting equipment, games and art materials, from near the child.
- Remove all writing on the blackboard unrelated to the task.
- Teacher states classroom rules consistently, for example reminding students that they shouldn’t chat or get up from their seat for any reason, if it is a time of concentrated individual working.
- Teacher posts a ‘Do Not Disturb’ sign on her classroom door during important activities such as tests or exercises that require a great deal of concentration.
- Video about how to reduce distraction.

Repetition
Children with working memory problems benefit greatly from judicious repetition of information to guide their ongoing activities. This information might relate to:
- General classroom management instructions
- Task-specific instructions
- The detailed content intrinsic to an activity.
Because not all of the children in a class or group have the same need for repetition, it is necessary to employ strategies that tailor repetition opportunities to needs of individual child. (Catherall & Milroy, 2008).

Activity:
In your experience, how often should you repeat and how soon? Can you explain why?

Rhythm and Rhyme
Rhythm definition:
A strong, regular repeated pattern of movement or sound.
Rhyme definition:
Correspondence of sound between words or the endings of words.
Rhythm and rhyme importance:
- May help some children to remember words and other information
- May make learning more fun (so again, more memorable)

Memory Aids
The importance of memory aids:
- Can help to trigger information in the child’s memory.
Memory aids can be anything teachers show learners in a classroom to assist their understanding, and may be visual or auditory.
- Kinds of memory aids:
  - Pictures, diagrams, videos, symbols, visual timetables, writing on the board in large, clear and colourful letters, or verbal gestures, songs, clapping.
In summary, memory aids could be anything teachers show learners in a classroom to assist their understanding.

Activity
What visual memory aids do you prefer to use in class? Why?

Chunking
‘Chunking is the technique of organizing or combining individual pieces of information into “chunks”’
– Professional Learning Board.

- How to Chunk?
Three main parts of chunking:
1. Identifying the chunks.
2. Grouping and memorising the chunks.
3. Retrieval of chunks.
Summary

A video about working memory strategies in general.
https://www.youtube.com/watch?v=WbQXEDaCDmU
By Chris Jarrold.

Any questions?

Driving Question
Working Memory Intervention Session 5:

**Session Outline**
- Driving questions discussion
- Developing the child's own strategies to support working memory
  - Asking for help/repetition
  - Rehearsing information
  - Note taking
- Summary
- Any questions?
- Driving question

**Driving question**

**Activity**
- Do you believe the children in your classes have their own strategies that they have developed to help them remember things?
- If so, what do you think these strategies are?

**Developing the child’s own strategies support working memory**
- Asking for help/repetition
- Rehearsing information
- Note taking

**Asking for help/repetition**
- Requests for help can be either verbal or gestural
- Teachers can discuss with children which person he or she should ask for help
- A child with poor WM can be partnered with another child who has good memory abilities (Gathercole & Alloway, 2008)
- Use of cards (traffic light system, or Makaton) to indicate the child’s desire for the teacher to ‘repeat’ instructions or ‘help’ them.
- What cards can you use in your classroom for the above?
Rehearsing Information

• Teachers can ask children to rehearse information for a brief period, such as one minute.

Activity: Testing the effect of rehearsal
Video: https://www.youtube.com/watch?v=A_0yNiW-tM

Make rehearsal a game! E.g., remembering the picture cards below (Hughes, 2006).

Note-taking

• Teachers can encourage note taking appropriate to the child’s literacy and numeracy skills, including pictures

Activity: What do you think are the most appropriate note-taking techniques to use in class with your pupils with Down syndrome?

Summary

In this session we discussed:
• Developing the child’s own strategies to support working memory, such as:
  • asking for help/repetition;
  • rehearsing information;
  • Note-taking.
• We practiced some activities to show the effectiveness of rehearsing information.

Any Questions?

Driving Questions Preparation
Working Memory Intervention Session 6:

**Working Memory in Children with Down Syndrome in the Classroom**

Session 6: Review and Reflection

**Driving Question**

---

**Session 1: Introduction to Working Memory and this Intervention**

- Memory
- Working memory
- Working memory in Down Syndrome
- Working memory in learning
- Relationship between working memory and literacy and numeracy

**Working Memory: Model from Baddeley & Hitch (1974)**

---

**Session 2: Recognising Working Memory Difficulties in the Classroom**

- Incomplete recall
- Failure to follow instruction
- Place keeping errors
- Task abandonment

---

**Reflection**

- Had you previously thought about literacy and numeracy tasks in terms of working memory demands?
- Has learning about working memory changed how you think about teaching literacy/numeracy?
Reflection

• Which, if any, of these indicators have you seen in pupils with Down syndrome in your classroom?
• In your experience, how easy or difficult is it to identify working memory difficulties in pupils with Down syndrome?

Session 3: Evaluating the Working Memory

Loads of Tasks Taught

Reflection

• Have you tried to evaluate working memory load of your teaching tasks?
• If so, which of these methods have you used?
• How easy or difficult did you find it to evaluate working memory load?

Session 4: Strategies to Support Working Memory in Down Syndrome

• Working memory strategies:
  - Reducing task working memory load
  - Reducing distraction
  - Repetition
  - Chunking
  - Memory aids

Reflection

• Did you try any of these strategies with your pupils?
• Which strategies did you find most helpful?
• Did this vary across different pupils?

Session 5: Developing the child’s own strategies to support working memory

- Asking for help/repetition
- Rehearsing information
- Note taking

7  8  9  10  11  12
Reflection

• Did you try any of these strategies with your pupils?
• Which strategies did you find most helpful? Did this vary across different pupils?
• How easy or difficult is it to teach a child how to improve his/her working memory?

Summary

• Session 1: Introduction: working memory and this intervention
• Session 2: Recognising working memory difficulties in the classroom
• Session 3: Evaluating the working memory loads of tasks taught
• Session 4: Strategies to support working memory in Down syndrome
• Session 5: Developing the child’s own strategies to support working memory

Video: https://www.youtube.com/watch?v=GLVc3woBkFQ

Any questions?

• To conclude this review and reflection, is there anything that you feel has been missing from our discussion today?
• Is there anything about which you wish to learn more, or skills that you would like to improve?
• Do you have any other questions?
• Can you make an action plan of things you would like to implement over the coming months?
• What resources might you require?
PBS Intervention Session 1:

Challenging Behaviour in Down Syndrome in the Classroom

Session 1: Introduction

Session One Outline

- Brief introduction to the intervention.
- Driving question.
- Challenging behaviour in Down syndrome
- Causes of challenging behaviour: internal and external factors.
- Common functions of challenging behaviour
- The effect of challenging behaviour on teachers, school and other pupils,
- Teachers' attitude and responses.
- Summary
- Driving questions discussion

Introduction to the Intervention

<table>
<thead>
<tr>
<th>Session 1: Introduction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Session 2: Recognising types of challenging behaviour (external and internal challenging behaviour)</td>
</tr>
<tr>
<td>Session 3: Evaluating challenging behaviour</td>
</tr>
<tr>
<td>Session 4: Strategies to address challenging behaviour in young children with Down syndrome</td>
</tr>
<tr>
<td>Session 5: Proposed model to support positive behaviour in Down syndrome</td>
</tr>
<tr>
<td>Session 6: Review</td>
</tr>
</tbody>
</table>

Introduction to 'Driving Questions'

- What are driving questions?
- How do we develop them?
- What purposes do they serve?
- How might they be answered?
- How will we use them in this intervention?

This question should be your response to the material that has just been presented, and it should be based on your reflections about the implications for what you have just heard for your own classroom practice. The driving question(s) could reflect either a specific challenge you are facing with a particular pupil, or a change you want to make in your own teaching strategies in order to assess its impact.

Introduction to the Research

- PhD degree.
- Special education.
- University of Edinburgh.
- Working memory and challenging behaviour in DS.
- Evaluation of an intervention in working memory and challenging behaviour, involving child assessment, teacher and parent questionnaires, teacher observation and focus groups.
- Teachers' input and perceptions are vital.
Teachers Introduce Themselves

• Now it is time to introduce yourselves!

Challenging Behaviour Definition

“Challenging behaviour is usually taken to include impulsive and aggressive behaviour, self-injurious behaviour, socially inappropriate behaviour, withdrawal”, and refusing to follow instructions
  - Emerson et al. (2001)

Behaviour “of such an intensity, frequency or duration as to threaten the quality of life and/or the physical safety of the individual or others’ and is likely to lead to responses that are restrictive, aversive or result in exclusion”
  - Royal College of Psychiatrists et al. (2007)

Activity

In small group discuss:
• Your experiences of challenging behaviour in class
• The types of challenging behaviour you have experienced
• How closely does your experience fit with the definition here?
• Has your opinion of what constitutes ‘challenging behaviour’ always been the same, or has it changed? (If so, why?)
• Might behaviours be experienced as ‘challenging’ by one person and not by another? Why?

Challenging Behaviour in Down Syndrome

Around one quarter of children and/or adolescents with DS have clinically significant behaviour problems (Smith, 2014).


Internal factors:
✓ Medical conditions (e.g., hearing)
✓ Digestive problems
✓ Emotional problems
✓ Discomfort
✓ Executive function
✓ Language and communication

External factors:
✓ Family
✓ Peers
✓ School
✓ Teachers’ attitude, strategies, communication and goal-setting
✓ Society and government rules and attitudes towards disability

Often there may be a combination of factors, and/or the causes may not be clear.

Common Functions of Challenging Behaviour

□ Communicating feelings, wants and needs.
□ Getting something tangible.
□ Meeting a sensory need.
□ As a result of lack of understanding.
□ Escape or avoid something.
□ Getting attention or reaction from peers and adults.
□ Obtain sense of power or control.
**Activity**

- Do you think there is another function not mentioned here?
- Have you observed some of these functions in your class children?

**The Effect of Challenging Behaviour**

- **On Teachers.**
  - Their ability to teach effectively. Generates tension if threatening.
- **School.**
  - Costs them valuable time and resources, investing in appropriately equipped professionals to intervene, instead of the educational environment.
- **Other pupils.**
  - Affects their concentration and attention in the classroom, disrupts learning.

**Teachers’ Attitudes and Responses**

- Teachers develop negative attitudes towards students that are difficult (Silberman 1969, 1971).
- Successfully changing the behaviour of disruptive students does not necessarily lead to a change of teacher attitude towards them (Toftel, 1983).
- A close, positive and supportive relationship between teachers and students are essential for developing learning potential and for responding appropriately to challenging behaviour. Langley’s study (2009).
- Approaches that avoid seeing the problem as entirely located in the child and which encourage pupils to feel secure and foster good relations with teachers, resulted in pupils who were more motivated to learn (Carroll & Hurry, 2018).

**Summary**

Although there are external and internal factors which influence challenging behaviour, it is clear that it is not possible for educators to consider them separately when managing the issue in their own classes. It is important to analyse both of them in order to reduce children’s challenging behaviour.

**Any Questions?**

**Driving Questions Discussion**

-
PBS Intervention Session 2:

Challenging Behaviour in Down Syndrome in the Classroom

Session Outline
- Session Outline
- Driving question discussion
- External and internal behaviour
- Forms of challenging behaviour in the class:
  - Stubbornness
  - Noncompliance
  - Social withdrawal
  - Aggression
- Driving questions discussion

Driving Questions Discussion

Forms of Challenging Behaviour

- Externalising & Internalising Behaviour Problems
  - Types of internal behaviour:
    - Behavioural difficulties characterised by disordered mood or emotional distress.
    - E.g., depression, anxiety, social withdrawal and somatic complaints (may not be challenging in themselves but could lead to challenging behaviour)
    - May not be overtly visible to others

Forms of Challenging Behaviour

- Types of external behaviour:
  - Includes aggressive, delinquent, hyperactive, inattentive, disruptive and oppositional behaviours
  - Hyperactive behaviour describes excessive motor activity, e.g., fidgeting, talking or yelling inappropriately, have difficulties remaining seated when expected.
  - Overlap between internal and external behaviour.

Forms of Challenging Behaviour

- What do you consider to be challenging behaviour?
- A brief introduction about types of challenging behaviour in DS.
Stubbornness

Discuss: How do you define stubbornness?

Definition:
Refusing to change one’s mind or course of action despite pressure to do so.

Why might some children sometimes show this behaviour?

Non-compliance

- Students who are defiant or non-compliant can frequently interrupt instruction, often do poorly academically, and may show little motivation to learn.
- Does your student:
  - Ignore your requests?
  - Directly defy you?
  - Say no a lot?
  - What might lead to non-compliance?
  - Is non-compliance behaviour always negative?
  - Choice & incentives

Social Withdrawal

- The retreat from society and interpersonal relationships often accompanied by indifference and aloof behaviour;
- Associated with depression, ASD and schizophrenia;
- Is social withdrawal an issue for your pupils?
- If so, what kind of things do you currently do to try to help kids be less isolated?
- What have you found that works? Are there things you’ve tried that really haven’t worked?

Aggression

- Definition: A forceful action or procedure (such as an unprovoked attack), especially when intended to dominate or master.
- How common is this kind of challenging behaviour in your classroom? How do you respond to it?

Activity

Discuss:
- Are there any challenging behaviours not covered?
- What are the factors that you think cause inappropriate behaviour in students?
- What are the best things to do when a student demonstrates challenging behaviour?
- In this blog: [http://prohastings.blogspot.com/p/blog-page.html](http://prohastings.blogspot.com/p/blog-page.html)

Summary

- Challenging behaviour can be external and/or internal
- Forms of challenging behaviour in the class:
  - Stubbornness
  - Noncompliance
  - Social withdrawal
  - Aggression
- Tips for dealing with the above challenging behaviours
- Reasons underpinning challenging behaviour
- Challenging behaviour is not always negative
Any Questions?

Prepare Driving Question

13

14

Further Reading

• https://www.ndss.org/resources/mental-health-issues-syndrome/
• http://profhastings.blogspot.com/p/blog-page.html

15
PBS Intervention Session 3:

Challenging Behaviour in Children with Down Syndrome in the Classroom
Session 3:
Evaluating Challenging Behaviour

Session Outline
• Driving questions discussion
• How to evaluate challenging behaviour?
• Summary
• Any questions?
• Driving questions for next session

Driving Questions Discussion
Discuss:
What is the purpose of evaluating behaviour?

How to evaluate challenging behaviour

Activity

Observation

<table>
<thead>
<tr>
<th>Behaviour Observation Planning</th>
<th>Name</th>
<th>Behaviour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Operational Definition:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Throwing items. Child throws items such as book or toys, either on the floor or at other children.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Date</td>
<td>Time</td>
<td>Location</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


Activity

• Please watch this video of challenging behaviour in the classroom and use the table to make notes on what you observe:
  - https://www.youtube.com/watch?v=0AlHFsQ7k
  - Do you think it is possible to observe children in the class at the same time that you deliver the lesson? How easy or difficult would it be? How could you manage this?

Behaviour checklist

Strengths and Difficulties Questionnaire (SDQ)
The SDQ is a measure of children’s characteristics and behaviour. The researcher has given you (teachers) a version of the SDQ measure. It contains many items describing child behaviours and allows for an estimate of the degree to which this behaviour occurs.

Behaviour Checklist

The behaviour checklist (Mustapha, 2006) was designed specifically for children with intellectual disabilities and was developed and published in the Um Al Khura University Magazine for Human, Social and Educational Science.

Activity

• Think of a particular pupil and (without identifying who that pupil is) and have a go at filling in a checklist.
• Do you think checklists/observations are useful?
• Can they tell us anything more than what is observed in the course of teaching?
• In which situations might they be useful?

Discussing with Parents

• What might teachers discuss with parents about their child’s challenging behaviours?

• What points should be considered when meeting parents to discuss their child’s challenging behaviour?

Activity

• In your experience, what is the most effective method (or combination of methods) for evaluating challenging behaviour?

• Can you evaluate the challenging behaviour in other ways, apart from observation, behaviour checklist and discussing with parents?
Summary

There are three ways of evaluating challenging behaviour:

1) Observation
2) Behaviour checklist
3) Discussing behaviour with parents.

You have discussed the benefits and pitfalls of each one and how to approach it, as well as having a go at the behaviour checklist yourselves.

13

Any Questions?

14

Driving Questions for the Next Session.

15
PBS Intervention Session 4:
Challenging Behaviour in Down Syndrome in the Classroom

Session 4:
Positive Behaviour Support for children with Down Syndrome

Session Outline
- Driving questions discussion
- Positive behaviour support in the classroom
- Functional behaviour assessment (FBA)
- Summary
- Driving questions discussion

Driving Questions Discussion

Positive Behaviour Support in the Classroom

What is Positive Behaviour Support (PBS)?
- "...a person-centred approach to supporting people who display or are at risk of displaying behaviours which challenge (Positive Response Training and Consultancy, 2020).
- Holistic approach
- Research validated practices
- Video: https://www.youtube.com/watch?v=VMsfe5CI6pQ

Positive Behaviour Support Triangle

For children with intellectual disabilities...

But PBS is suitable for everyone!

10% of children with Down Syndrome
5 to 15% - at risk
80 to 90% - no serious behaviour problems

PBS Involves...

- Understanding the reasons for behaviours which challenge
- Assessing the broad social and physical context in which the behaviour occurs - including the person's life history, physical and mental health, and the impact of any traumatic life events
- Planning and implementing ways of supporting the person which enhance quality of life for both the person themselves and their carers.
How to Implement PBS?

<table>
<thead>
<tr>
<th>Creating high quality care and support environment</th>
<th>Functional, contextual and skills-based assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Ensuring the classroom is values-led</td>
<td>1) Working in partnership with stakeholders</td>
</tr>
<tr>
<td>2) Knowing the child</td>
<td>2) Assessing match between the child and their environment</td>
</tr>
<tr>
<td>3) Matching support with the child’s capabilities and tailored goals</td>
<td>3) Knowing the health of the child</td>
</tr>
<tr>
<td>4) Effective teamwork amongst teachers and carers</td>
<td>4) Understanding the principles and functions served by that behaviour</td>
</tr>
<tr>
<td>5) Choice and communication</td>
<td>5) Assessing the functions of behaviour</td>
</tr>
<tr>
<td>6) Supporting both physical and mental health</td>
<td>6) Assessing a child’s skills and abilities, preferences and motivations</td>
</tr>
<tr>
<td>7) Predictability of environment</td>
<td></td>
</tr>
<tr>
<td>8) Understanding legislation</td>
<td></td>
</tr>
<tr>
<td>9) Teacher commitment to behaviour skills training</td>
<td></td>
</tr>
</tbody>
</table>

Adapted from PBS Coalition UK

Functional Behaviour Assessment (FBA)

- What is Functional Behaviour Assessment (FBA)?
- Three key concepts of FBA:
  - Setting events
  - Triggering antecedent
  - Maintaining consequences

Watch video (FBA)
https://www.youtube.com/watch?v=GboNGPSHA64

Activity

- PBA Form
- FBA Form

Emotional responses to challenging Behaviour
- How do you respond emotionally to challenging behaviour?
- Might this sometimes reinforce the behaviour in some way?

Summary

- Positive Behaviour Support: holistic support, evidence-based practice
- Understanding the reasons underpinning challenging behaviour.
- PBS Triangle
- Functional Behaviour Assessment – how to evaluate challenging behaviour.
- Activities

Any Questions?

Driving Question for the Next Sessions
PBS Intervention Session 5:

Challenging Behaviour in Down Syndrome in the Classroom

Session 5:
Model of Positive Behaviour Support Intervention.

Session Outline
- Driving questions discussion
- Model of positive behaviour support intervention
- Summary
- Driving questions discussion

Driving Questions Discussion

Model of Positive Behaviour Support Intervention
- Prevention
- Instruction
- Reinforcement
- Negative consequences

Prevention
- Identify and minimise setting events and possible triggering antecedents.
- Positive relations with children, helping them to feel valued and to develop a sense of belonging, are also essential in reducing challenging behaviour.

Activity
- Thinking back to the FBA sheets you completed in the previous session, what do you do to minimise the effect of setting events and/or triggering antecedents?
- How can you avoid having to intervene more directly, for example by increasing your presence with the child and providing direct support in the classroom?
- Can you think of times when you have tried to build a more positive relationship with a child? How did you go about this?
Instruction (replacement behaviour training)

- Effective intervention to reduce challenging behaviour can be carried out only when the function(s) of that behaviour have been identified.
- Teachers then need to design an intervention that encourages children to meet that function in a more socially acceptable way.
- There is evidence that teaching children skills to engage in positive, pro-social behaviours can lead to better academic performance and reduced levels of challenging behaviour.

A Replacement Behaviour

'A replacement behaviour is a behaviour that the teacher is encouraging the child to use as an alternative to the unwanted target behaviour. Focusing on the more positive behaviour may be preferable to focusing on the problem behaviour, as focusing on the problem behaviour may just reinforce it, especially if the consequence (reinforcer) is attention. Behaviour which a teacher might seek to teach replacements for would typically include aggression, destructive behaviour, self-injury, or tantrums' - (Webster, 2018).

A Replacement Behaviour

Rather than teaching social skills generally, there is a more specific sense of instruction in response to challenging behaviours, which is the very specific targeted teaching of a specific socially acceptable behaviour, which satisfies the same function as the challenging behaviour.

Teaching social instruction steps

(A replacement behaviour)

- Break the desired skill into specific steps.
- Describe each step.
- Provide a rationale based on child’s personal goals.
- Provide modelling through video or role plays.
- Provide guided practice through role plays and activities.
- Give feedback.
- Put the students in situations where the skill can be applied and generalised.
- Highly reinforce the student for exhibiting the skill.

Activity

- Can you mention some replacement behaviours that you teach children?
- How do you identify that the replacement behaviours serve the same function as the challenging behaviours?
- What methods do you use to teach these more socially acceptable replacement behaviours?
- How easy or difficult is it to teach these skills?

Reinforcement

- Behaviour is more likely to be repeated if it is reinforced.
- One kind of reinforcement might work for some children but not for others.
- Extrinsic and intrinsic reinforcement.
Activity

• Could you give me some examples of reinforcements that you use with children?
• Are there some reinforcements that are received well by some pupils but not with others?

Negative Consequences

• The correlation between ‘action and consequence’, between a child’s actions and their consequences.
• Types of negative consequences.
• Points to be considered in conducting negative consequences.

Summary

• In this session, we discussed the model of Positive Behaviour Support intervention, through the different phases of:
  • Prevention, Instruction, Positive Reinforcement and Negative Consequences.
  • We highlighted some points that need to be considered in the stages of reinforcement and negative consequences.
  • These phases of Positive Behaviour Support intervention are summarised in the following diagram...

Model of Positive Behaviour Support

• Prevention:
  - Identifying setting events and triggering event matrix
  - Identify, function of the challenging behaviour
  - Replace the challenging behaviour with socially acceptable behaviour

• Instruction:
  - Identifying intervention activities
  - Identifying intervention activities

• Positive Reinforcement
  - Positive reinforcement
  - Positive reinforcement

• Negative Consequence
  - Negative consequence

Driving Questions Preparation

• Any Questions?

13
14
15
16
17
18
PBS Intervention Session 6:

Challenging Behaviour in Children with Down Syndrome in the Classroom

Session 6: Review and Reflection

Session 1: Introduction to Challenging Behaviour and this Intervention

Reflection

• Which challenging behaviours are common in your class?
• What functions of challenging behaviour have you identified in your class?
• Which challenging behaviour did you find has the most impact on the class?
• How have you responded to challenging behaviour in the past?

Session 2: Recognising Types of Challenging Behaviour

Reflection

• Did you find these behavioural definitions helpful in recognising challenging behaviour?
• Do you see different challenging behaviours in different contexts, or in different pupils?
Session 3: Evaluating Challenging Behaviour

- Observation
- Behaviour checklist
- Discussing with parents

Reflection

- Did you try any of these methods for evaluating challenging behaviour?
- What was your experience of this?
- Did you use a combination of methods? If so, which methods did you combine? How effective did you find this combination?

Session 4: Strategies to Address Challenging Behaviour in Children with Down Syndrome

- Positive Behaviour Triangle

Session 4: Strategies to Address Challenging Behaviour in Children with Down Syndrome

- Relationship between Functional Behaviour Assessment and Positive Behaviour Support:
  1) Setting events
  2) Triggering antecedents
  3) Maintaining consequences

- Assessment Tools:
  1) Functional Behaviour Assessment form
  2) Teacher response questionnaire

Reflection

- How did the Functional Behaviour Assessment form and the teacher response questionnaire work for you?
- Did you find any benefit in your class?

Session 5: Proposed Model to Support Positive Behaviour in Children with Down Syndrome

- Prevention
- Early identification and triggering of problems
- Before intervention: prevent them (preventing or removing the cause)

- Induction (idem)
- Identify the function of the challenging behaviour and prevent the challenging behaviour with socially acceptable behaviour that produces the same outcome.

- Goal setting: challenging behaviour and development of the skills needed to reduce challenging behaviour

- Positive reinforcement

- Positive consequences (to top level)
Reflection

- Did you conduct this model in your class? If so, which stages did you find easy or difficult? Did you use all phases or just some? Why?
- Did you combine the phases of Positive Behaviour Support? Why?

Summary

- Session 1: Introduction about challenging behaviour and this intervention
- Session 2: Recognising types of challenging behaviour (external and internal challenging behaviour)
- Session 3: Evaluating challenging behaviour
- Session 4: Strategies to address challenging behaviour in children with Down syndrome
- Session 5: Proposed model to support positive behaviour in children with Down syndrome

Any questions?

- To conclude this review and reflection, is there anything that you feel has been missing from our discussion today?
- Is there anything about which you wish to learn more, or skills that you would like to improve?
- Do you have any other questions?

Action Plan

- Please design an action plan for use in your own classes, showing how you can put these ideas into action.
- What are your goals over the next 2 weeks, one month, and 6 months?
- I will be available over the next two weeks to answer any questions or to help with implementation.
Appendix 2.9: Ethics Information and Consent Forms

Our Ref: 1287

Date: 29th June 2018

Dear Ghaleyah,

Title: Working memory intervention with teachers of children with Down syndrome

The School of Education and Sport Ethics Sub-Committee has now considered your request for ethical approval for the studies detailed in your application.

This is to confirm that the Sub-Committee is happy to approve the application and that the research meets the School Ethics Level 3 criterion. This is defined as “applies to novel procedures, research without consent, sensitive personal data, or the use of atypical participant groups. Also projects in which ethical issues might require more detailed consideration but are unlikely to prove problematic”.

You are reminded that if the research changes in any way from that described on your application form, you may need to re-apply for approval.

Should you receive any formal complaints relating to the study you should notify the MHSE Ethics Committee immediately by email to MHSE@ed.ac.uk

Yours sincerely

On behalf of:
Dr Ailsa Niven
Convener, School Ethics Sub-Committee

The University of Edinburgh is a charitable body, registered in Scotland, with registration number SC005336
Appendix 3.1 Change in Teachers’ Knowledge on Working Memory and Challenging Behaviour: Comparison Between Groups

Comparing EG and CG on WM and CB knowledge pre- and post-intervention

The previous analyses on (p.261) showed that there was significant improvement in teachers’ knowledge in working memory for the experimental group and in challenging behavior for the control group between pre- and post-intervention, and how large the differences between groups was. This was achieved by analysing the total score of working memory items and total score of challenging behaviour items. For further explanation, paired samples t-test were used on each item individually to find on which items the scores had improved statistically and which they had not, in both the working memory subscale and challenging behaviour subscale, for each group.

Experimental group paired samples t-test paired for Teachers’ Knowledge on Working Memory and challenging behaviour

<table>
<thead>
<tr>
<th>Pair</th>
<th>Q1 Working Memory knowledge Pre intervention Post- intervention</th>
<th>df</th>
<th>Sig. (2-tailed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pair 1</td>
<td>Q1 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.002</td>
</tr>
<tr>
<td>Pair 2</td>
<td>Q2 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair 3</td>
<td>Q3 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair 4</td>
<td>Q4 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair 5</td>
<td>Q5 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.002</td>
</tr>
<tr>
<td>Pair 6</td>
<td>Q6 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair 7</td>
<td>Q7 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair 8</td>
<td>Q8 Working Memory knowledge Pre intervention Post- intervention</td>
<td>13</td>
<td>.000</td>
</tr>
<tr>
<td>Pair</td>
<td>Item</td>
<td>Change</td>
<td>P-value</td>
</tr>
<tr>
<td>------</td>
<td>------</td>
<td>--------</td>
<td>---------</td>
</tr>
<tr>
<td>Pair 9</td>
<td>Q9 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention knowledge Pre intervention – Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 10</td>
<td>Q10 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 11</td>
<td>Q11 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 12</td>
<td>Q12 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 13</td>
<td>Q13 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 14</td>
<td>Q14 Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 15</td>
<td>Total Working Memory knowledge Pre intervention</td>
<td>–13</td>
<td>.000</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 16</td>
<td>Q15 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.082</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 17</td>
<td>Q16 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.054</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 18</td>
<td>Q17 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.165</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 19</td>
<td>Q18 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.336</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 20</td>
<td>Q20 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.174</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 21</td>
<td>Q21 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.136</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 22</td>
<td>Q22 Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.189</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 23</td>
<td>Q total Challenging Behaviour knowledge Pre intervention</td>
<td>–13</td>
<td>.022</td>
</tr>
<tr>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

From the table above, it is clear that for the EG, there was a significant improvement in all items of WM from item 1 to item 14; all these items have p<.01. Regarding the CB items, it is clear that for the experimental group there was no significant improvement in individual CB items, although there was a significant improvement in the total CB score, as p=.022, which is less than .05 (p<.05). However, it is worth mentioning that CB item 17 nearly achieved a significant improvement (p=.054). This item’s question was: “How confident do
you feel in your knowledge about the challenging behaviour of children with Down syndrome?“

Control group paired samples t-test for Teachers’ Knowledge on Working Memory and challenging behaviour

<table>
<thead>
<tr>
<th>Pair</th>
<th>Question</th>
<th>Knowledge Pre intervention</th>
<th>Post-intervention</th>
<th>df</th>
<th>Sig. (2-tailed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pair 3</td>
<td>Q3</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 4</td>
<td>Q4</td>
<td>Working Memory</td>
<td>13</td>
<td>.218</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 5</td>
<td>Q5</td>
<td>Working Memory</td>
<td>13</td>
<td>.189</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 6</td>
<td>Q6</td>
<td>Working Memory</td>
<td>13</td>
<td>.104</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 8</td>
<td>Q8</td>
<td>Working Memory</td>
<td>13</td>
<td>.189</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 9</td>
<td>Q9</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 10</td>
<td>Q10</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 11</td>
<td>Q11</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 13</td>
<td>Q13</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 14</td>
<td>Q14</td>
<td>Working Memory</td>
<td>13</td>
<td>.336</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>– Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 15</td>
<td>Q15</td>
<td>Total Working Memory</td>
<td>13</td>
<td>.234</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pre intervention – Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 16</td>
<td>Q15</td>
<td>Challenging Behaviour</td>
<td>13</td>
<td>.189</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pre intervention – Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 17</td>
<td>Q16</td>
<td>Challenging Behaviour</td>
<td>13</td>
<td>.003</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pre intervention – Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pair 18</td>
<td>Q17</td>
<td>Challenging Behaviour</td>
<td>13</td>
<td>.000</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>knowledge</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pre intervention – Post-intervention</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
From the table above, it can be seen that in the CG, there were no significant improvements on any of the 14 WM items (questionnaire questions). Regarding the CB items, there were significant improvements in all except two items, as all p-values were less than .01 (p<.01). For item 15, p=.189: “How confident do you feel in your knowledge about the challenging behaviour of children with intellectual disabilities?” For item 22, p=.218: “To what extent is learning about Positive Behaviour Support in children with Down syndrome a priority for you at the moment?”