

**PSYCHOSOCIAL FACTORS AND QUALITY OF LIFE IN
ADULTS WITH CYSTIC FIBROSIS**

By

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**SUBMITTED IN PART FULFILMENT OF THE
DOCTORATE IN CLINICAL PSYCHOLOGY.
THE UNIVERSITY OF EDINBURGH 1997**



DECLARATION

I hereby declare that this thesis is entirely my own work, with no help from others except for those referred to in the acknowledgements.

Acknowledgements

I would like to thank Frank Charlton and Dr F. Buckley for all their help and encouragement in supervising me. I am grateful to the medical and nursing staff in the CF team at the Western General Hospital for their help in co-ordinating interviews and for allowing me to learn about their clinical work. Thank you also to my husband Ian for his support and particularly for keeping everything going on the domestic front. Finally I would like to thank all the subjects for taking part, particularly to those with Cystic Fibrosis who gave up their time to be interviewed.

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ABSTRACT

The life expectancy of Cystic Fibrosis (CF) patients continues to increase, and it is therefore important to understand more about the psychosocial aspects of CF in adulthood. The present study aims to compare a group of 30 adult CF patients with a healthy control group on a multidimensional quality of life measure (WHOQOL) and other measures assessing self-esteem, anxiety, depression, social support and body image. A clinical measure of severity was also included in the CF group as were objective measures of physical functioning. The experimental group were outpatients attending the CF centre, Edinburgh. Their mean age was 29 (range=18 - 49 years). Healthy controls were a community sample matched for age, sex and deprivation category. Adjustment to illness was also assessed in the CF group and additional qualitative data relating to CF specific quality of life was obtained during semi-structured interviews with 19 of the CF subjects. Statistical analysis was conducted to test the prediction that there would be no significant between groups differences on psychological measures (anxiety, depression and self-esteem). Differences were predicted in certain aspects of quality of life such as mobility and ability to work. Social support networks and body image concerns were also predicted to vary between the groups. A thematic analysis of interview data was also conducted to explore specific issues of adjustment and quality of life within the CF group.

INTRODUCTION

Cystic Fibrosis (CF) is the most common fatal genetic disease occurring in Caucasian populations. It is an autosomal recessive disease but thought to be rare in populations not of Caucasian European descent (Tsui 1990). It is characterised by abnormal secretions of the exocrine (mucous producing) glands which can result in a variety of secondary clinical effects, including chronic lung disease and pancreatic insufficiency.

CF used to be regarded as a disease which only affected children and in 1938, 70 % of children died within the first year of life (Andersen 1938). However results from a study in England and Wales now estimate that by the year 2000 approximately half of the CF population will be over the age of 16. The estimated median age of survival for a child with CF born in 1990 is now estimated at 40 years (Elborn et al 1991). The improvement in prognosis has generally been due to the institution of aggressive preventative medical treatments (e.g. medication and physiotherapy) before the onset of irreversible pulmonary changes. Changes in service delivery, such as the setting-up of specialised clinics have also allowed timely interventions to be made by health professionals.

It is therefore likely that health service provision for adults with CF will require continual expansion, and greater attention will need to be focused on the physical and psychosocial course of this disease in adulthood.

1. Medical Background

1.1 Incidence of CF

The most frequently quoted incidence figure is 1 in 2000 live births (di Sant'Agnese and Talmo 1967). The regional variations for this figure are considerable. While it may hold true for most of the UK it has been reported to vary within the U.S.A. with the highest incidence reported as 1 in 489 (Honeyman and Siker 1965) and lowest

incidence as 1 in 6667 (Hannar 1965). Different data have been given again for racial subgroups such as American blacks with an incidence of 1 in 17000 live births (Stern et al 1976).

One large scale UK study (Dodge et al 1993, 1988) following cohorts from 1968 onwards gave an incidence rate of 1 in 2,500 live births which is generally considered to be a reliable rate for this country. The survival rate for females has been found to be lower than males in all age groups (Dodge et al 1993) except in the 0-4 years age range. The reason for this is unclear as there have been no links found with genotype, pulmonary function, body mass, age at diagnosis or microbiological factors (Rosenfeld et al 1996). Environmental and psychosocial factors have not yet been considered as possible explanatory variables of this observed gender difference.

1.2 Defining and diagnosing CF.

Another factor influencing epidemiological studies is the way in which CF is defined, either by its genotype or phenotype. The term “phenotype” refers to the clinical signs and symptoms of CF and historically diagnosis has relied on the interpretation of these. This process has been refined with some clinicians attempting to categorise patients into groups with a diagnosis of “proven”, “probable” and “possible” CF (Ten, 1977).

A diagnostic test which is commonly used is the sweat test. Patients with CF have a raised level of sodium chloride in their sweat, however this test is generally performed on patients with a “phenotype” already suggestive of CF. Routine infant screening is not generally carried out unless there is a specific programme established or there is a family history of CF. This has therefore meant that diagnosis has sometimes been delayed. In approximately 10% of cases the diagnosis is made by sweat test a few weeks after birth and a further 15 % of babies present immediately after birth with intestinal obstruction (meconium ileus). The majority of cases present later in the first

year of life with failure to gain weight, recurrent respiratory tract infections and diarrhoea. Excluding babies with meconium ileus, the diagnosis has been made before one year in only about half of the cases (Jackson 1989). This is probably due to the fact that symptoms vary in severity and some patients have a mild form of CF. In one study of 65 adult patients, 12 had been diagnosed after the age of 16 (Swachmann et al 1965). In a large study of 316 CF patients seen at the Brompton Hospital (Penketh et al. 1987) only 6% had been diagnosed in adult life with three diagnosed after the age of 30.

1.3 Genetics and CF

Since the gene for CF was identified on the long arm of chromosome seven (Knowlton et al 1985; Wainwright et al 1985; White et al 1985) current estimates suggest there are at least 200 different gene defects which give rise to the CF phenotype. Since no exhaustive list of genotypes is yet available, it is therefore difficult to diagnose by genetic screening alone. It is possible that some genotypes can give rise to asymptomatic CF phenotypes and presentations of differing severity, so the relationship between genotype and phenotype requires further elucidation.

One plausible assumption that is made on the basis of the incidence rate of Cystic Fibrosis occurring in 1 in 2,500 live births (in a Caucasian population) is that one in 25 people are carriers of the CF genotype. There is therefore a one in four chance that the offspring of two carriers will be affected by CF and a one in two chance that offspring will be carriers. There is a common CF gene defect, Delta F508, which has an occurrence of 45 per 100 cases, six defects which each have an occurrence of 3% giving a frequency of 24 in 100 cases. The remainder, a variety of rare genotypes therefore occur on average in 31 out of 100 cases, (The Cystic Fibrosis Genetic Analysis Consortium, 1990).

Understanding the genetics of CF has been helpful in that parents of a child with CF could be informed of the 25% recurrence risk for their next child. Pre-natal diagnosis

by amniocentesis offers further control over recurrence of CF but not its occurrence in a first child (Ten 1996). Screening programmes have been set-up to detect carriers outside of known CF families. However there are complex ethical issues as well as genetic problems surrounding attempts to control CF.

Very rare genotypes only occur in 1 in 100,000 live births which can make detection difficult and could present a serious impediment to any screening programme as does the fact that it is not always the case that both parents of a CF patient are carriers. The range of clinical variation in CF is wider than was previously assumed and genotype-phenotype correlations have proved to be poor (Rosenstein 1994). It is therefore difficult to counsel carriers or predict the clinical course, quality or length of life of a child born with a particular mutation, and this must be taken into account when counselling is carried-out.

Recent studies of knowledge of and attitudes towards carrier-testing have found that willingness to consider genetic testing or abortion of a foetus with CF decreased significantly in a group of college students after being lectured about recent advances in CF treatment (Neiger et al 1992). Denayer et al (1996) also found that only 25% of their sample of 200 people who had received genetic screening for CF were aware of the high prevalence of CF carriers; genetic counselling and information-giving by trained professionals is therefore a vital part of any antenatal screening programme for CF.

1.4 The Impact of diagnosis

Families may experience a considerable delay before receiving an explanation for their child's symptoms. In a study of 30 children who had been conventionally diagnosed with CF the average duration of symptoms before diagnosis was 15 months (Helton et al 1991). The stage of diagnosis can be a highly stressful one for families (see 2.5), however there is a dearth of research about the impact of a late diagnosis on the child or adult with Cystic Fibrosis.

One recent study of adults diagnosed later with Cystic Fibrosis by Widerman (1996) found that subjects self-reported not being severely affected. They also described unfamiliarity with CF prior to diagnosis and feeling different from those diagnosed in childhood. Most of the subjects had been symptomatic prior to diagnosis and had been referred by their physician to a specialist Cystic Fibrosis centre, thus diagnosis was made over a period of time. Circumstances surrounding diagnosis had a strong impact on the patient's response. In children with CF, factors predicting good adaptation to diagnosis were good communication with the Cystic Fibrosis team and extended family supports. Poor adaptation was associated with geographical isolation from the CF team, financial deprivation and previous experience of CF death (Burrows et al 1996).

A search for information was noted in newly diagnosed adults (Widerman 1996) but patients often came across outdated material describing CF as a fatal childhood disease. Finally late-diagnosed patients presented with a great need for support and information at a time when they were facing other life-stage events.

1.5 Clinical features of CF and treatments.

There are a number of different bodily systems affected during the course of CF and current treatment is aimed "towards prevention and control of secondary and tertiary manifestations of the disease" (Dodge 1989). Both the symptoms themselves and the treatments will inevitably have implications for quality of life and are likely to be specific to the individual .

Respiratory disease

Pulmonary disease is almost universal in adult CF populations, with an incidence of 99.7% quoted in the Penketh et al study (1987) . It was also responsible for 97% of all deaths among CF patients and three quarters of hospital admissions in this sample.

Shortly after birth, mucous glands in the lungs can be seen to be distended, and secretions can block the airways resulting in infections. Later in childhood and adult life, some CF patients (about 40% in the Penketh et al study 1987) demonstrate an increased sensitivity to a range of allergens. This sensitivity is sometimes termed asthma or “reversibility” since it responds to inhaled bronchodilators. The development of a cough producing mucus is also characteristic of most CF patients by an early age. This is as a result of mucous over-production by exocrine glands in the lungs which subsequently blocks the airways; this obstruction can cause lesions in the lung tissue over time.

Chest physiotherapy has been the main treatment for respiratory disease, aimed at removing mucous from the lungs. This can take about 20-30 minutes a day to perform, and responsibility for this can be taken by family and carers in childhood, transferring to the patient in adolescence. Independence in treatment-related responsibility generally increases with the age of the child (Drotar and Ievers 1994) and self-efficacy (confidence in being able to perform a behaviour) was the most important educational factor predicting self-management behaviour for monitoring and treatment of respiratory problems in CF patients under the age of 18 (Parcel et al 1994). Adherence to physiotherapy in adulthood may be poor in about 25% of patients (Pownceby 1996) the main reasons given being lack of time and low motivation. This treatment could be perceived by a sub-group of patients as having a high personal cost and a low immediate benefit on the quality of their life. This will be investigated in the present study.

Other preventive treatments on which trials are currently being conducted include DNase, or the drug (P-Imosyne). This drug is administered by nebuliser once a day and works by breaking down mucous in the lungs to make it less sticky and thick and therefore less receptive to infection (Shak 1990). Treatment with this has been associated with improvements in lung function, fewer chest infections and an improvement on quality of life measures (Wohl 1993). Gene therapy is another new

preventative treatment which is in the process of being developed. It may be useful in children or patients who are reasonably fit but it can not reverse the scarring of lung tissue which has already occurred in some older patients (Hodson 1995).

Inevitably mucous retained in the lungs will become infected with a range of bacteria, certain fungi and viruses such as influenza, which may lead to secondary infection. Some of these bacteria can be damaging and hard to treat which is an argument in favour of preventative treatments. Combinations of antibiotics can be given to treat infections either intravenously during hospital visits or via a port-o-cath (“port”) which is inserted into the chest wall and through which the patient can administer their own antibiotics at home. The presence of a port could conceivably have implications for improved quality of life but could also exert a negative effect on body image. This has as yet received little attention in the literature.

Some new strains of treatment antibiotic resistant bacteria (For example, *Pseudomonas Cepacia*) have been identified in some CF centres (Pedersen et al 1986). This has led to infected individuals being segregated to reduce spreading of the bacteria in hospital or clinics or even during social contact (Smith et al 1992). This could have an influence on the social networks of CF patients and could lead to a sense of stigmatisation of infected individuals within CF centres.

Intestinal Obstruction

In new-borns, one of the commonest presentations is meconium ileus or intestinal obstruction which can lead to perforation of the gut. When present in older patients, it is referred to as meconium ileus equivalent or distal intestinal obstruction syndrome. The obstruction may be partial or complete and can recur. In a large cohort of 316 CF patients aged 12 to 51 years (Penketh et al 1987), acute meconium ileus equivalent was seen in 16% of the sample and a partial obstruction with episodic symptoms was seen in a further 19%. This condition may require surgery in acute presentations or more often can be treated by enema or intestinal lavage.

Pancreatic insufficiency and nutrition

Deficiencies in pancreatic secretions can give rise to abdominal distension and fatty stools. One diagnostic test for pancreatic insufficiency can therefore be the measurement of faecal fat. Babies tend to fail to gain weight adequately, and adults may experience appetite loss which contributes to under-nourishment and malabsorption. In the Penketh et al (1987) study, 11% of patients had no malabsorption problems so could be described as “pancreatic sufficient”. It has been observed that this sub-group of patients have a biologically less severe variant of CF, and that this is determined by the inheritance of at least one copy of a mild mutation which is phenotypically dominant over severe mutations (Kerem 1989).

Pancreatic enzyme supplements are generally taken in tablet or granule form with meals in order to replace the patient’s own lack of enzymes. In most cases vitamins A, D and E are also routinely supplemented since poor absorption of these is common. If adequate replacement of pancreatic enzymes are given, then normal growth and weight can be expected. Later loss of weight and faltering of growth is usually associated with deteriorating respiratory function (Dodge 1989).

Energy levels in CF can also be low due to increased resting energy expenditure and increased work of breathing during infective exacerbations. Therefore a high energy diet is important and an intake of 120-150% of the recommended daily allowance is appropriate (O’Neill et al 1983). Not surprisingly high energy expenditure has been linked with malnutrition, and the fact that appetite loss is common during pulmonary infections may result in pressure from the family for the child to eat and the child attempting to resist this.

Behavioural feeding disorders have been found to co-exist with other nutritional problems in children with CF, (Bowen and Stark 1991) and behavioural interventions

have been shown to be effective in increasing the long-term oral intake in this group (Stark et al 1990). In adulthood compliance with pancreatic enzymes and vitamins has been reported at around 44 % in a recent “coming of age survey” (Pownceby 1996) with reasons for non-compliance being forgetting to take them and embarrassment at having to take them in public.

Other complications

As CF patients are living well into adulthood means that the other long-term sequelae of CF are increasing. There is evidence that mild liver disease which is present in about 29% of the adult CF population (Penketh et al 1987) increases in prevalence with age (Roy et al 1983). One study (Thorsteinsson et al, 1995) has shown that 32% of 25 year old CF patients had insulin dependent diabetes. This means that adult CF patients may have to contend with further diagnoses and treatment regimens as they get older.

Transplant

CF patients are generally considered to be good candidates for lung and heart-lung transplants because they are young, often well motivated and used to taking medications (Madden 1995). The main problems are the lack of donor organs and some complications post-transplant. Presently 50% of patients die on the waiting list (Wrightson 1996) but if transplantation takes place, the rate of survival can be 69% at one year and 52% at two years (Yoacoub et al 1990; Madden et al 1992).

Selection criteria for heart-lung transplantation are: Deteriorating chronic respiratory failure, severely impaired quality of life in spite of the best available medical treatment, and the patient's positive attitude towards a transplant. Factors contraindicating transplant include poor compliance, psychological difficulties, and end-organ failure (except for the liver which may also be able to be transplanted) Madden (1995).

Once the patient has been referred by the CF centre they are assessed by the transplant team. If they are considered suitable they are placed on a waiting list and can be called by bleep at any time to go in for transplant. This can be a very stressful process with possible false alarms resulting from the fact that it is important to have a good match between donor and recipient (Madden 1995). The speed of deterioration of patients awaiting transplant can be an additional concern to patients and their families.

Post-transplant, there is the possibility of organ rejection, but also upper respiratory tract infection specific to CF, since CF changes persist in the mucous above the transplant site. In many cases, quality of life has been shown to improve dramatically after surgery and one study showed improved social and emotional well-being on the Nottingham Health Profile (Caine 1991). Five generic quality of life instruments were administered before and after lung transplantation to patients in the Netherlands, (Busschbach et al , 1994). The authors concluded that the improvements in quality of life post transplant were comparable to those after a heart transplant. However, the scales used could have lacked specificity and failed to adequately assess the patient's subjective perspective of their functioning (see section 4).

1.6 Fertility and pregnancy

Although puberty is delayed by approximately 2 years, sexual development is otherwise normal in both sexes (Reiter et al 1981). The large majority of male patients (about 98%) are infertile due to obstruction of the vas deferens (Brugman 1984), however recent advances may mean that in vitro fertilisation is feasible if enough of the patient's sperm can be obtained.

However, infertility can remain a difficult issue for parents to discuss with their sons and the timing of this can be crucial. Madge and Carr (1996) found that 62% of their sample of parents felt infertility should be discussed openly in a clinic setting, and 24% were not sure. 78% felt that it was the joint responsibility of parents and the CF

team to discuss these issues. The period just prior to transfer from paediatric to adult CF centres was deemed the most appropriate time for this discussion. The age for transfer can vary however from 16 to early twenties, and not all patients attend a specialist paediatric centre.

Fertility in females appears to be normal with loss of periods occurring occasionally, this is generally secondary and related to poor lung function and nutritional status (Stead 1987). Recent data concerning 38 pregnancies in 25 women with CF who had relatively advanced lung disease indicated that pregnancy was well tolerated by the majority and there were low rates of neonatal mortality and no congenital malformations (Canney et al 1991). A longitudinal study concluded that the “physiological impact of pregnancy... on women with CF does not exert an independent and negative effect on her mortality or pulmonary status”. Women with insulin dependent diabetes were found to be the only subgroup of women at risk of pulmonary decline two years after pregnancy (Stacey et al 1996).

Despite these recent findings, prevailing clinical practice is for medical staff to generally advise against pregnancy in women with poor respiratory function and low weight who may not gain adequate weight in pregnancy and could be at risk of mortality during the stress of labour (Koch and Lanng 1995).

It seems likely that reproductive and sexual issues will place an additional strain on adolescents and young adults with CF, that is, over and above the usual pressures of adolescence. This could affect psychosexual functioning and impair the ability to form and maintain intimate relationships. Researchers such as Sawyer (1996) call for further needs assessment in this area and an increase in timely communication by specially trained health professionals. The present study aims to assess some of these needs in a sample of adult CF patients.

1.7. Service delivery

The context of service delivery may also be vital to outcome. Treatment has been found to be most effective when it has been delivered by a specialist team, familiar with the patient over a number of years. The variety of skill mix in these teams is also a considerable strength. They can include physiotherapists, dieticians, pharmacists, social workers and psychologists as well as nurses and physicians. Life expectancy has been shown to be better if services are delivered by such specialist clinics (Nielson and Schiotz 1982; Warwick 1982). Shared responsibility between centralised clinics and outlying areas can take place, so that even when a patient lives a long distance away from the centre, annual assessments can be carried out in the clinic.

Patient satisfaction with medical care has recently been assessed by Pownceby (1996) and found to be related to several features; feeling that the doctor understood how treatment affected their lives; feeling able to discuss treatment difficulties with her or him; and being given the right amount of information in a way that they could understand. The first two of these factors were also found to be related to increased treatment adherence (Pownceby 1997). However, there are likely to be other factors also mediating compliance (See 2.7).

2. PSYCHOLOGICAL ASPECTS OF CYSTIC FIBROSIS

The experiences of patients with CF highlight two important areas of interest in psychological medicine. Firstly the response of the individual to a chronic disabling illness, and secondly, the effects on the adult CF population as a whole. (Aspin 1991) The overall impact of the illness is likely to be determined by the complex interaction between physical symptoms and social and psychological factors.

2.1 The prevalence of stress and distress

Compared to the large bulk of literature about the psychosocial functioning of children and adolescents with CF, there still remains relatively little about adults (Lask 1996). Although the evidence is mixed from studies of psychosocial effects in children with CF, a major meta-analysis of the relevant research concluded that children with CF are as well-adjusted as healthy children and children with other chronic diseases (Cowan et al 1985). Kashani et al (1988) have suggested that the seeming lack of psychopathology in children could result from the fact that they are receiving appropriate care and attention from health professionals and family which could have a protective effect. Alternatively, younger patients may express psychological distress through less direct means than older patients (Pearson et al 1991).

Variables such as age, gender, family functioning and disease severity have been proposed to effect adjustment. Psychological problems in children with CF have been seen to vary with age with no significant behavioural problems in pre-schoolers (Cowan et al 1985) to a rate of 23% of psychiatric disorder in 6-11 year olds compared with a rate of 15% of their siblings (Simmons et al 1987). There has been some evidence that adults with CF also have a higher rate of emotional disorder compared to adolescents (Cowan 1984). Psychosocial factors certainly deserve consideration as individuals with CF enter adulthood. To date, studies of distress in adults have focused on various factors associated with the disease itself which could constitute a stressor on the individual. An early study by Boyle et al (1976) in a small sample of patients over the age of 18, noted four main areas of stress: altered physical appearance, strained interpersonal relationships, social isolation and increased awareness of the future.

Strauss and Wellisch (1981) also identified four aspects of having CF that bothered adult patients. In order of frequency, these were: cough 66%, thinness 57%, fatigue 47% and gastrointestinal or bowel symptoms 38%. Interestingly, the subjects in this study consistently underrated the severity of their symptoms compared to physician

ratings. A similar finding by Abbott et al (1996) indicated that patient's perceptions of severity remain stable over time despite decline on measures of physical functioning. This suggests that stressors do not always constitute a strain as perceived by the individual and that there are likely to be other mediating factors.

This is further illustrated by the conflicting evidence from studies of generalised emotional distress. In the Strauss and Wellisch study (1981) 43% of their sample reported feeling depressed occasionally or frequently with 86% indicating they rarely felt able to express feelings of anger. Contrary to this Moise et al (1987) interviewed 59% of an adult CF out-patient population and found no increase in either emotional discomfort or emotional alienation from others compared to test norms on the psychological screening inventory. Slightly lower self-esteem was found in a quarter of this sample. In the Cowan et al (1984) study postal questionnaires were used in a large sample of 191 patients over the age of 16. They found higher reported rates of distress at a rate of 12% in males and 30% in females aged 16-19 with the frequency of distress doubling over the age of 20. One reason put forward for the observed sex difference is the poorer physical prognosis for females with CF (Bywater 1981).

Although this last study would indicate that there is a substantial amount of distress amongst adults with CF, the reliability of using postal questionnaires alone and uncontrolled studies has been questioned (Aspin 1991). More recently a survey by Gee et al (1996) of anxiety and depression in 90 adults with CF and 106 controls showed there to be no significant difference between these two groups. Borderline anxiety was found to be quite common in both the populations at around 25% as identified on the Hospital anxiety and depression scale (HADS Zigmond and Snaith 1983). This study also provides some delineation of specific psychological disorders since previous research has generally not attempted to classify "distress".

There have been few evaluations of psychological interventions to treat stress or distress in the adult CF population. One such study by Keller et al (1985) assessed the effects of a six-week stress management and lifestyle modification programme on 18

adults with CF . In the pre-treatment phase, this sample had scored as being in the above average range for anxiety when compared to test norms for their age group. Post-treatment, they showed significant reductions in state and trait anxiety as well as reduced autonomic arousal on objective measures. The authors called for further larger scale studies of this nature and evaluations of effective components of treatment. There is some evidence from the health psychology literature that goal-setting approaches can be more effective in enabling coping with chronic illness than emotion focused counselling which tends to focus on the expression of negative affect (Wilkinson 1995).

2.2 Self Esteem

A further indicator of general well-being which has been considered is self-esteem. Apart from the Moise et al study (1987) there have been consistent reports of high self-esteem in adults with CF relative to controls . Simmonds et al (1985) illustrated this using the Tennessee Self-concept scale and Cowan et al (1984) found CF patients to be less self-critical than controls. An Australian study (Sawyer et al 1995) used the Offer Self-image Questionnaire for adolescents in a sample of 49 male and female CF patients. They noted that growth and pubertal development were delayed in both sexes and females, but not males, exhibited significantly lower self-concept than healthy peers. This may suggest that self-esteem is a general concept, but it has several dimensions within it and the above studies have employed different questionnaires measuring different but related constructs.

2.3. Body Image

One such construct which has received little attention in the prevailing literature is body image. Given the later maturational development of adolescents with CF and the problems with feeding noted in younger children with CF (Crist et al 1992) this could be a particularly relevant dimension of self-concept to investigate. CF patients also have to contend with the long term physical effects of the disease such as a

“barrel chest” or cushingoid swollen face if they take long-term steroid medication. Some other medications can also cause changes in skin condition. Any findings about body-image in this group have tended to be as a by-product of other investigations, for example from the few prevalence studies of eating disorders in CF patients.

Eating disorders could conceivably result from the arrested maturation and changes in eating and body composition associated with CF. A study by Steiner et al (1990) found that female adolescents with Anorexia Nervosa scored significantly higher than age matched CF subjects on body dissatisfaction, drive for thinness, and eating disordered behaviour . While Pumeriega et al (1990) reported on a series of 13 adolescents with an atypical eating disorder, Berry et al (1975) concluded that nutritional disorders were secondary to the disease. In another study (Jelalian et al 1996) CF subjects certainly showed a lack of eating disordered attitudes and behaviour in a comparison to their healthy peers. 15 females with CF aged 8 to 15 were more positive about their current weight than the comparison group. They also had less knowledge about the fat content of food and had a higher daily caloric intake. The females in the CF group were at a significantly lower weight percentile than their healthy peers and wished to weigh an average of 2 lbs more than their current weight while comparison girls wished to weigh an average of 8 lbs less. This may suggest CF females at a lower weight could be protected from the usual social pressures to diet and achieve slimness.

There have been very few studies of eating disorders in adults with CF. Pearson et al (1991) found that the prevalence of eating disorders was higher in adolescents than in adults with CF although the reverse was true for prevalence of anxiety and depression. Body Image in adults with CF was also investigated as part of an extensive study of sexual adaptation by Coffman (1984) who found that about half of their sample of male and female CF subjects acknowledged feeling unattractive. Typically subjects listed thinness, clubbed fingers, short stature and stained teeth as being the source of this . The contrast healthy control group nearly all reported feeling attractive, their only concern when present was of being overweight.

The scarce literature in this area..would suggest that attitudes towards eating are qualitatively different in a group of patients with an eating disorder and those with CF although this requires further investigation. Body image may also be characterised by less disparagement and drive for thinness in the CF group, and areas of body dissatisfaction may be more specific to CF.

2.4 Sexual functioning

In the Coffman et al study (1984) 48 young adults with CF were compared to a similarly aged group of healthy control subjects using interviews and questionnaires. Another previously studied group of married CF patients were also used as a comparison group. Single female patients with CF were found to have started to date later, less frequently, had less sexual desire and more sexual problems than the control subjects. Single male CF patients seemed to have better adaptation than females in all these areas . However 80% of these patients of a marrying age had not married, their general health scores were poorer than married patients and they had been diagnosed at a significantly younger age. There had been no link found in any group between severity of disease and sexual health.

One possible explanation for why single patients with CF had not married is the finding that they had experienced 3 times as many major losses as the married group in terms of sibling deaths, parental deaths and divorce. It was hypothesised that this might interfere with the forming of intimate relationships in later life, but it was difficult to establish these links in this study.

The relationship of perceived personal attractiveness to sexual adaptation is also not clear since both males and females with CF had a poorer body-image than controls in the Coffman et al (1984) study, but only females' adaptation was generally worse.

The authors proposed that body-image is only one factor in conjunction with self-esteem and separation-individuation problems which might predict deficiencies in sexual desire.

2.5 Psychological adaptation in the family

The diagnosis of CF can have devastating effect on the whole family. Various stages of adaptation for the family have been delineated, for example by Bluebond-Langner (1991). She describes strategies adopted by families to cope with 6 periods of the disease (see fig. 1)

STAGE	FAMILY RESPONSE
1. The first year following diagnosis	Routine for CF tasks developed Processing and compartmentalising information that has been given
2. Years up to first exacerbation	Avoidance of reminders of CF Redefining what is normal Reconceptualizing the future
3. Time between the first exacerbation and increasing hospitalisation	An intensification of all these strategies. Reassessment of family priorities
4. The period of increasing complications	Family coping strategies become challenged A changed, bleaker view of the future is projected Siblings become aware of chronic and terminal nature of CF.
5. The period of increasing deterioration	Family can no longer avoid reminders of CF They are confronted with changed view of the future, and family priorities change again.
6. The terminal phase	

When considering the impact of CF on a family, the stage of the disease is not always considered in research (Lask 1995). The literature generally suggests that families use coping strategies of minimisation, distancing and denial although some studies have pointed to the strength and flexibility of these families (Lewis and Kahn 1982). The

divorce rate among parents of a CF child is also no different from the national average as shown in a meta-analysis by Begleiter (1976); however lack of communication in CF families has been reported by Falkman (1977). It is likely that the opposing findings result from a lack of controlled studies and possible selection bias such as including only stable families in research (De Wet and Cywes 1984). Findings about the effects of CF on the family should therefore be interpreted with caution.

2.6 Adaptation in the individual

Due to the fact that CF has only recently come to be viewed as an illness of adulthood, there remains a dearth of research about the process of individual psychological adaptation as distinct from that of the family. The literature in health psychology concerning adjustment to physical disability may offer a framework which could be applied to individuals with CF.

Disability has been defined by the World Health Organisation(1980) as “a loss of function resulting from physical impairment” and as such, it is also distinct from the concept of handicap which is defined as “a perceived social or occupational disadvantage resulting from disability”.

Factors affecting the process of adaptation to a disability are; age of acquisition, insidious versus traumatic onset, stability of the condition and prognosis, severity of disability and degree of dependency, the presence or absence of pain and the frequency of periods of associated illness (Wilkinson 1995). Defining adjustment may also involve “personality-based” factors (such as the individual’s responses to disability) and “social context” factors such as the strength of social networks and an individual’s control over resources. These have been the two main trends in psychological research (Russell 1981).

If adjustment is a process, then this is likely to begin at the time of diagnosis of CF whatever age this is. Psychoanalytic views of adjustment liken disability to loss or

bereavement (Kreuger 1984). It is postulated that the individual will pass through a number of stages of shock, retreat or denial; grief/mourning and depression, hostility and anger, and adjustment. Stage theorists would argue that the disabled person has to pass through these in order to reach an acceptance of their condition and counselling can facilitate this process. As yet research in CF has not confirmed these stages exist or how they fit with the disease process.

A model which has greater applicability than the psychoanalytic bereavement model is that proposed by Erikson (1963) who has described eight developmental stages through the life cycle. For adolescents he described conflicts of Identity Vs Confusion, and for young adults ; Intimacy Vs Isolation then further into adulthood, Generativity Vs Stagnation. Erikson proposes that successful resolution of these conflicts at each stage is important for psychological well-being. This can be a useful framework to use when considering psychological adjustment in any population and for adults with CF, issues of autonomy (or identity), intimacy, and work and reproductive issues (generativity) are likely to be of particular salience. The present study therefore aims to assess these aspects and other factors which may impact upon them, for example body image and self-esteem and their relationship to issues of autonomy and generativity.

In health psychological research it is often correlates of adjustment such as coping style or strength of social support which are measured. While these are important it may also be advantageous to obtain an indication of the degree to which an individual has accepted their illness or disability. Self-report measures such as the acceptance of illness scale (Felton et al 1984) which address this directly are now available and this is included in the present study.

2.7 Severity, coping and compliance.

The degree to which an individual perceives themselves to be disabled can be mediated by coping style. Suls and Fletcher (1985) have described two defence

reactions exhibited by the individual threatened with an illness. The first is an avoidance response sometimes called denial, or repression. The purpose of this is to minimise threat by focusing attention away from the disease. The individual may show little interest in information, changes in the disease and may be unlikely to take any action. The alternative defence reaction is to attend to, or monitor the disease. These patients follow any changes in the disease process closely; they may be aware of recent research developments and talk openly about their disability and fears about the future.

Some psychological research has suggested that avoidance is a better defence strategy in the short term (e.g. Wolf et al 1964); however, later in a disease process thinking about emotional distress may be necessary in order to promote change in attitudes and behaviour. Thus in the long-term, individuals using attention as a response fared better both physically and emotionally (Suls and Fletcher 1985).

The response of CF sufferers is somewhat paradoxical since they tend to have less psychological distress, higher self-esteem and better physician-rated adaptation when they employ an avoidance strategy (Moise et al 1987). Strauss and Wellisch (1981) also found a "prominent use of denial" and an "achievement orientation" among CF sufferers. Shepherd (1990) also found that CF sufferers perceived themselves as "less severe" than their peers with CF which could be described as a minimisation strategy. At the same time other findings (Boyle 1976) suggest that CF patients reported considerable concern about their future, so it could be that the denial response is limited to certain aspects of the disease and is not universally adopted by all CF patients. The direction of causality between coping style and adaptation has also not been established by this research; it could be that those are better adapted physically are more able to use avoidance strategies.

Just as disease severity has been shown to have little relationship to psychological distress (Sensky 1990), so lung function and coping status have been found to be unrelated (Lask 1995). Therefore it seems likely that coping style has a more potent

influence on psychological functioning than disease severity. Two studies suggest that type of coping strategy used may be linked to compliance with medical treatments. Pinkerton et al (1985) found that those CF patients who showed less understanding of, and interest in, their illness had a greater frequency of hospital admissions. Caijkowski and Koocher, (1987) have pointed to the need for strict criteria for assessing compliance. In a study by these authors employing just such criteria for 40 adolescents with CF, they found that compliers were more optimistic about their health, had more interest in it and believed their actions made a difference. Using an attention strategy for coping with CF may lead to greater compliance but not necessarily less psychological distress as it would appear that anxiety may be raised during this process of monitoring hence the lack of correlation between physical severity and psychological distress.

This has been borne out in a recent study of health perceptions and treatment adherence in adults with CF by Abbott et al, (1996). It was found that worrying about having CF and the perception of having little personal control over the illness facilitated treatment adherence to physiotherapy, pancreatic enzyme and vitamin regimens. Patients tended to perceive exercise therapy as different ^{from} to other forms of treatment, since no anxiety was related to adhering to it. These findings illustrate the need for researchers to consider the differential effects of coping in response to particular treatments. At present the literature in the area of coping and compliance does not distinguish between worrying as an adaptive coping strategy or anxiety as a form of distress this requires further delineation.

2.8 Bereavement Issues

Many adults with CF were told as children that they were not likely to live long. This could mean until puberty, into their teens or possibly until the age of 20 at best. However many of these adults have lived beyond the expected life span, and this is an area that has only recently received some attention in the literature (Tracy 1996). This may have implications for how a patient plans for the future, their compliance with

treatment (as opposed to “living for today”), and regrets about things or projects which they have not undertaken due to their belief that their life span was limited.

It may be important to identify these issues in order to counsel patients at any age and also to prepare them for their own death if this is imminent. Many studies have indicated that people generally desire to know if their death is impending (Kalish and Reynolds 1976). As many as 80% of dying patients know they are dying and would like to talk about it (Crammond 1970). In another study by Lock (1990), patients did not complain about the information that was given to them but about the fact that doctors did not communicate that they cared for the patient. Further research is required about how these issues affect CF patients.

Other bereavement issues may concern loss of a sibling or other friends through CF. One study assessing the long-term effects of sibling death from CF in healthy adults found that anxiety, depression and survivor guilt were highest for those subjects who had been aged 13-17 at the time of their sibling’s death (Fanos and Nickerson 1991). It may be that this is also the case for surviving siblings with CF; this is an area which merits further investigation. Bereavement issues generally were addressed in the present study during interviews with CF subjects.

3. SOCIAL FUNCTIONING

3.1 Social Support

There is now growing evidence that personal adjustment as well as social behaviour can be linked to a person’s access to support from others (Brown and Harris 1978; Henderson et al , 1981). Particular definitions of social support have varied and this has been reflected in the large number of measures available. The term social support can refer to structural aspects such as type of support and how it is organised. More recent research has focussed on functional aspects of social support, and the ways in which different types of support serve the individual.

A study of structural aspects of social support by (Revicki and May ,1985) found that family members only, but not peers, mediated the relationship between occupational stress and depressive symptomatology in physicians. In attempting to define the relationship between functions of social support, Cohen and Wills (1985) defined them as follows: Esteem support; being valued by others; Informational support; necessary information which may be gained from social contacts, instrumental support-financial or material support; social companionship; support derived from spending time with others in leisure or other activities.

Another important distinction has been made between perceived social support and received support. For instance, Sandler and Barrera (1984) compared the relation of several support measures they had developed with measures of anxiety, depression , somatization and an index of psychological disorder. They found that these symptomatology scales were unrelated to measures of network size or received support. However greater frequency of symptomatology on all measures was related to low perceived support. Appraisal of support as being low was also the strongest predictor (of the three dimensions of social support assessed) of mortality risk in a study of elderly men and women (Blazer 1982). It is hardly surprising that appraisal is such an important component of social support given the individual differences in needs for social contacts. Two reviews by Sarason et al (1986 and 1987) have compared different available indices for measuring social support. The authors found that one common factor assessed by all measures was the degree to which an individual perceived themselves as being accepted, loved and involved in relationships in which communication is open. Individuals who had such relationships were less depressed and lonely than those who did not. The authors suggested that this central function of social support is an extension of the function of attachment in childhood as described by Bowlby (1969, 1980). Bowlby hypothesised that acceptance, affection and affirmation of personal worth by the care-giver can lead to secure attachment in the child.

Lieberman, (1977) and Sroufe and Fleeson (1986) have demonstrated that securely attached children are able to explore the environment and engage in activities outside of themselves (e.g. games and tasks) more easily than insecurely attached children. It is easy to understand how this process could be applied to adults, however it remains unclear as to whether it is only adults who have been securely attached as children who perceive themselves as adequately supported by others.

Researchers such as Sarason et al (1987) recommend that social support is conceptualised as a developmental personality characteristic rather than a social provision alone. These authors advise caution in generalising social support findings across age groups and socio economic groups since these latter factors also play a part. Procidano and Heller (1983) and Cutrona (1986) have pointed out that an individual's support networks are likely to change over time. Women of low socio-economic status have also been shown to be less able to make use of social support than others (Eckenrode 1983). Women have also tended to endorse more social support items on measures than men (Sarason et al 1987). This last finding may have been an artefact of measures used in that females were more familiar than men with the supportive situations described in the scale. Finally, it is likely that people who are experiencing a life event will score more highly on received support measures (Sarason et al 1987) due to the mobilisation of their support network at a time of crisis. This could conceivably also be the case for CF patients during acute exacerbations and periods of hospitalisation.

3.2 The family as a source of support in CF

The initial milieu for social support is usually the family. This context has received considerable attention in the CF literature. The effects of CF on the family were detailed in 2.5. How the family may impact on CF is also important as this has been shown to be linked to compliance (Patterson, 1985). Often family processes are hypothesised to mediate adjustment to illness via a transactional stress and coping

model where the CF is viewed as a chronic stressor to which the family have to adapt (Robert et al 1996). Recent research has shown that parents' perceptions of stresses associated with rearing a CF child are related to their mental health and that mothers but not fathers have significantly poorer mental health than the general population (Nagy and Ungerer 1990). Similar findings were reported by Quittner et al (1992) who found role strain was related to depression in mothers but not fathers of CF children.

The impact of CF on the family and general family dysfunction have been found to be related to severity but not duration of the illness in CF patients aged 7 to 15 years (Pameriega et al 1993). These negative effects can be mediated through paternal and family supportiveness. (Nagy and Ungerer, 1990). This familial stress is generally expressed through emotional and behavioural disturbances in the child with family processes either ameliorating or exacerbating their development into depressive symptomatology (Pameriega et al , 1993) .This lends support to the transactional model between family and CF.

Less conclusive findings were reported by Geiss et al (1992) who noted that there was no "characteristic pattern of psychopathology or adjustment problems" in children with CF or their families. Higher levels of perceived compliance were associated with less frequent maternal social contacts which could suggest that it is the quality of support from a parent (usually the mother) which might influence compliance. For example, frequent but controlling interactions from a mother with regards to physiotherapy may result in low compliance and the child taking little responsibility for their treatment (Johnson et al, 1982). There have been few studies to date which consider the role of social support in compliance in an adult CF population. The "coming of age survey" of young adults with CF (Pownceby 1996) found that higher levels of adherence were reported in families which were more organised. Shared responsibility between the parent and child for treatments also increased adherence.

A comparison of CF families with families of anorexic and healthy controls demonstrated that families of anorexia nervosa sufferers were more dysfunctional than those of controls and CF patients. CF and anorexic families did have higher levels of expressed emotion and emotional over-involvement than controls. Over-involvement correlated with illness severity (Blair et al 1995). The majority of CF adolescents and young adults in this study showed good psychological health, and their families showed as good problem-solving ability as control families (Blair et al 1994).

These findings point to the specific patterns of family functioning and how this may or may not relate to adjustment in the individual. Little is yet known about the role of the family in social support networks of adults with CF. One of the few studies in this area (Rothlisberger 1993) points to the likelihood of the continued importance of the family in providing support. CF adults in this study also reported slightly less perceived support than healthy controls but greater perceived support than depressed subjects. Given the higher rate of family over-involvement relating to illness severity in CF (Blair et al 1995), it could be hypothesised that adults with CF who may face an age-related decline in their health may simultaneously have to struggle to achieve autonomy compared to their healthy peers. Demographic data lends partial support to this argument.

3.3 Autonomy, marriage and significant others.

Demographic data from two U.K. studies would suggest that there is a reduced level of autonomy and rate of marriage in adults with CF. Penketh et al (1987) reported a low rate of 13% of marriage among their sample with the majority of patients still living at home with their parents. However the mean age in this sample was 23 which could be considered low. Another postal survey of adults with CF in the UK conducted by Walters et al (1993) found that a total of 34% of this population were married compared with 61% of the general population. Women were also significantly more likely to be married or co-habiting than men with 44% of women and 26% of

men in such relationships. CF adults continued to reside in 52% of family homes as a non-dependent child as opposed to 11 % in the general population. These findings suggest reduced autonomy and forming of intimate relationships in adult CF patients.

A U.S. study by Shepherd et al , (1990) reached different conclusions in a controlled study of 37 CF adults. They found no difference between the groups in terms of marital status, income, or number of dependants. The CF group did have a large majority of male respondents. The CF group who were not married or cohabiting were more likely to live alone or with their parents whereas the comparison group were more likely to share with flatmates or friends. Autonomy was also assessed by degree of financial independence, and although the numbers of subjects who were self-supporting were the same for both groups, those CF patients who were not, were 5 times as likely to be financially-supported solely by someone else. This led the authors to conclude that there is a subset of CF patients (about 30%) who have reduced autonomy. Interestingly, as with findings in other areas level of autonomy was not found to be related to physical functioning.

The quality of social support either from the family or significant others is also important. This was assessed by Shepherd et al (1990) using an objective measure of frequency of contacts (density) and general satisfaction with given family situations. While this showed no differences between the two groups of subjects, there was no measure of perceived practical support or disparity between actual and received social support from friends as well as family. Shepherd et al (1990) also found that single adults with CF reported higher dissatisfaction with their sex lives than single healthy controls, this could reflect the disparity between their wish for intimacy and their actual single situation. Alternatively it could indicate the presence of sexual dysfunction or a disturbance of body image and may therefore require further detailed assessment.

While objective, demographic data suggests that there may be difficulties with autonomy and intimacy in adults with CF little is known about the process of forming

intimate relationships or friendships in this group. There could conceivably be particularly specific aspects of this process which are important such as when to disclose they have CF, how much to tell others, what is the patient's experience of people's responses to disclosure. When and how this process occurs could also largely depend on self-perception and individual adjustment. The present study investigated this important process of forming relationships during interviews with the CF subjects.

3.4 Education and employment

In adolescents with CF employment and education was found to be no different from healthy controls (Smith 1983). Later studies have generally found that while many adult CF patients function well, they are still less likely to be in employment than their peers (Blair et al, 1994; Shepherd et al, 1990).

Penketh et al, (1987) reported that 78% of their sample were in full-time education or full or part-time employment with only a minority giving reasons for not working as being ill-health. Only 17% felt that their schooling had been seriously curtailed and 51% of patients had left school with some GCE or CSE passes and 12% went on to further education. However, a controlled comparison by Walters et al (1993) found that only 54% of their responders were in full-time employment, and about half of these had less than two weeks sick leave a year. Half of those who were unemployed gave ill-health as the reason and severity of disease was also linked to unemployment. People with CF had been less successful than their healthy peers at achieving 'O' level qualifications, but more successful at achieving 'A' level or higher qualifications. Achievement of any qualification increased employment prospects regardless of disease severity.

Walters et al (1993) also found that there were a significantly higher number of CF adults in non-manual occupations when compared to their healthy peers (and the social class of their family of origin was controlled for). Social class has been found to

have an independent effect on mortality in CF despite the fact that it is assessed by occupational status as manual or non-manual (which could be said to be determined by CF itself) (Britton, 1989). The author suggests that this effect could come about through lack of resources to visit hospitals since a disproportionately high number of patients (50%) attending clinics have been found to be in social classes 1 or 2 as coded by father's occupation (Penketh et al 1987). Resources for the provision of other dietary supplements may be limited for some patients, or other factors such as increased parental smoking or poor quality of housing could affect survival. It is therefore important that researchers address these effects of social class and consider demographic information of the CF population under investigation. In the present study social class was considered and assessed by geographic region of residence as opposed to occupation since the higher reported rate of non-manual occupations in CF populations could have been a confounding factor.

3.5 Social perceptions of CF and Stigma

Discrimination is something which is rarely assessed in CF adults but as in other groups with a perceived disability, this can occur. Perception of disability as socially or individually defined can affect adjustment as discussed in 2.6.

Walters et al (1993) reported on discrimination against adults with mild and moderate CF who revealed to employers that they had the illness. Acquiring mortgages, and life insurance is another area in which CF patients are likely to experience difficulty and this could militate against their achieving independent living away from their parents. Despite the improved prognosis for CF patients, health professionals and the public perceive the quality of life for children with CF to be poor, and poorer than that expected for children with Down syndrome (Marteau et al 1996). Different rates across populations of terminations for pregnancies affected by CF may be explained by these social perceptions. Rates of terminations have been seen to vary from 30% in families with a CF child already (Jedlickakohler et al , 1994) to 100% in population based screening programmes.

Attempting to overcome socially constructed disability could be one explanation for the minimisation coping strategy employed by some CF patients and the fact that they and their close companions perceive their disease as far less severe than physicians (Abbott et al 1996). Although some research has demonstrated the stigmatising effects of CF gene carrier-status in the general population (Evers-Kiebooms et al 1994, Denayer et al , 1996), little research effort has been directed towards investigating the stigmatising effects of having CF and the process by which social perceptions can be resisted (perhaps through minimisation strategies) or internalised . It is possible that stigma, if it is internalised, could be linked to a poorer psychological adjustment and a feeling of “being different”. Therefore social and professional perceptions of the disability associated with CF may need to be challenged. Quality of life research could be particularly useful in aiding this process. By comparing CF patients with a sample from the general population, the present study aims to explore the many similarities as well as possible differences which may arise between the two groups.

4. QUALITY OF LIFE

A decrease in mortality rates generally has promoted health researchers to consider ways of reducing morbidity. This has meant that the focus has changed to consider assessment not only of the impact of disease and disability, but quality of life too and how this might change over time (Skevington et al 1997). Such measures are essential to evaluate the recipient's perceptions of various treatments. While objective physical measures and demographic data can provide useful information about functioning, it is often the individual's own perceptions (as discussed previously) which can determine their satisfaction with treatments. This could also have implications for the individual's overall adjustment and adherence to treatments, and provide a useful parameter of outcome measurement. Quality of life measurement also provides a specific format for periodic appraisal by policy-makers, and could enhance clinician's understanding of the specific meanings an individual attaches to their illness.

Quality of life (QoL) has become a popular area for research, but researchers have defined the term in different ways (Bos 1991). QoL has been conceived as being part of a purely social context (Burt et al 1978). Such objective approaches to QoL assessment have assumed that quantifiable indicators such as income, the physical environment and deprivation are valid measures of QoL. However there are no finite methods for assessing these variables (Evans 1994) which are often affected by cultural and ideological factors and are therefore not “value-free” .

In contrast to the emphasis on the social context , QoL has been studied from the individual perspective. Sartorius (1987) defined it in terms of “the distance between a person’s position and his or her goals” thus emphasising the importance of individual appraisal. Both positive and negative affect are components of this psychological domain (Reich and Zautra 1981) and further dichotomies between physical and spiritual factors have also been proposed (Liu 1974). Some synthesis of these different conceptualisations of QoL as social and psychological have occurred more recently, for example Powers and Goode (in Goode, 1990) asserted that QoL is “primarily a product of relationships between people in each life setting” . Aaronson et al (1991) have also defined QoL in terms of physical, psychological and social functioning.

Most QoL measures have been developed within the area of health research. Generally the need to assess a wide number of domains has been emphasised (Fitzpatrick et al 1992) and while most measures are multidimensional, they are still limited to certain dimensions (Morrow et al, 1992). For example, Mercier (1992) in a review of the Cancer literature found that the following themes recurred: i) the status of daily physical activities including professional and domestic duties; ii) the frequency of physical and psychological complaints (e.g. pain, anxiety and depression) ; iii) the ability to maintain one’s sexual functioning; and iv) the subjective feeling of well-being. While these may all be important areas to assess in “ill” individuals, social and emotional support have often been neglected in research. Successful adaptation to a chronic illness and perceived health status are also important determinants of well-being for these individuals (Badura and Waltz, 1984).

4.1 Measures of health-related QoL

Existing instruments used to measure QoL can be split into generic and disease-specific questionnaires. Generic measures purport to be applicable across a wide range of types and severity of diseases and can also be used in healthy individuals (Fitzpatrick et al, 1992). Generic measures can be applied to a broader range of diseases and in a greater number of settings than more specific measures. They can usefully compare the relative benefits of different treatments or relative impact of different diseases and allow for detection of unexpected effects in research by employing a broader focus (Fletcher et al , 1992) However, some generic measures are impractical to administer due to their length, and may also be less sensitive to identifying disease-specific factors such as treatment (Bowling, 1995) or the effects of health care (Fletcher et al , 1992).

Disease-specific QoL measures are aimed at measuring responsiveness, that is, clinically important changes (Patrick and Deyo, 1989) for a narrow range of diseases. They have the advantage that they are short, sensitive to small changes but disadvantages include the lack of comparability of results from other disease groups and the possibility of missing effects in dimensions which are not included.

In many cases it has not yet been established whether specific QoL measures give significant information about incremental gains beyond what is provided by generic instruments.(Kaplan, 1985). For this reason, Bowling (1995) and others have called for comparison of generic and specific measures within the same disease group whenever possible. This would also be the most effective way of collecting accurate data. When a specific measure of QoL is not available then supplementary approaches could be used such as interviewing .

Until very recently there was no questionnaire that fully measured subjective quality of life. Existing health status measures that are based on domains such as social or

physical functioning are not applicable to the well population which makes controlled comparison difficult between ill and healthy persons (Patrick and Erickson , 1993). There is no “gold standard” basic measure questionnaire to which specific measures could be compared. The Short form / RAND-36 (SF-36) (Ware 1993) is one generic QoL questionnaire which is widely used in clinical research, but it measures objective rather than subjective health status.

Since there is a strong need for a universal definition of QoL and an instrument which can be used cross-culturally the World health organisation (WHO) initiated a project called “The Assessment of QoL in Health Care” with the aim of developing a generic instrument measuring subjective QoL . The WHO quality of life assessment instrument (WHOQOL) has now been developed and is considered to be applicable to well populations, can be employed cross-culturally and consists of global QoL items. This should be the starting point of all QoL measures according to Tuchler (1992).

4.2 Quality of Life in CF

Conclusions about QoL in CF have been drawn from limited, objective, data. For example demographic and survival data is not sufficient in itself to conclude that there has been an “improvement in quality of life” (Penketh et al 1987). For while this may be plausible given advances in treatments in CF, there is still a lack of research evidence which measures QoL from the subjective perspective in adults with CF.

A recent study by Congleton et al (1996) assessed QoL in 240 CF patients over the age of 16. The authors used the Nottingham Health Profile NHP a generic instrument and six additional questions which were specific to Cystic Fibrosis. A sex difference was noted, with men describing more distress/disability in the dimensions of sleep, emotion and social isolation in the older age group. There were no age differences in the female group, but mean scores for the CF group as a whole were comparable to other groups with minor, non-acute conditions when compared to published norms.

The NHP (Hunt et al 1990) has been criticised for focussing exclusively on problems and does not reflect the positive aspects of QoL. Furthermore without pilot studies it is difficult to construct specific questions pertaining to QoL in CF without the danger of excluding important domains.

The Congleton et al (1996) study also found correlations between some of the QoL dimensions and lung function (FEV1), breathlessness, and time spent on home treatment. Dodd et al (1996) also recently concluded that “perceived changes in chest disease mirror changes in the general quality of life in adults with CF”. This was based on their study of changes in lung function and QoL rating during a course of intravenous antibiotics. QoL or “general life satisfaction” was assessed on a 10 point scale. While global questions may be important for easy administration during clinical trials it is advisable to incorporate a standardised multidimensional measure (Bowling 1995) in order to establish the validity of a new brief rating scale.

5 . SUMMARY AND STUDY AIMS

The physical course of CF is unpredictable and there are likely to be many individual differences both in patient’s clinical presentation and their response to treatments. Despite this, comprehensive controlled QoL research can help to identify the many similarities in perceived QoL which may exist between the CF group and their healthy peers. Such similarities would serve to further the understanding of the process of the individual’s adjustment to illness, and lend weight to the continued research into new medications to prolong survival. The present study aims to include just such a comprehensive QoL assessment using a multidimensional measure including both the subject’s perceptions of their QoL and objective measures to compliment these.

The role of psychological factors in compliance and physical outcome has proved to be a popular area of interest to medical researchers generally , and yet it remains a complex one due to the presence of possible mediating factors such as coping style . There have been no clear links found between severity of illness and psychological

distress (Sensky 1990), however, social factors such as social support have received little attention as possible moderating variables. Indeed the only known psychosocial associations linked to outcome have been between social class and mortality (Britton 1989) and between unemployment and greater severity of CF (Walters et al 1993). The present study addresses these issues by including demographic information about the CF group and exploring the moderating impact of social support between psychological adjustment and severity.

There have been few studies in CF populations which have classified “distress”. There have also been few controlled studies of psychological factors (Aspin 1991) or studies which draw on qualitative as well as quantitative data. The present study differentiates between anxiety and depression and includes additional measures of body image and self-esteem. The current study is controlled and includes extensive qualitative data from interviews. The interview format is particularly useful for obtaining information about sensitive issues such as bereavement, intimate relationships and stigmatisation in CF all of which have been identified as important areas for research (Tracy, 1996; Sawyer 1995; Marteau et al , 1996).

Finally, the present study also utilises a direct measure of individual adjustment or acceptance of illness. Previous studies have tended to focus on the CF family’s adjustment or correlates of adjustment in the individual such as coping style. A direct measure allows easy comparison with other variables which may also be associated with adjustment; for example, age and age-related “tasks” of adulthood (such as employment) may be particularly important factors when considering the Eriksonian model of stages of individual adjustment through the life span (Erikson, 1963).

6. HYPOTHESES

Between the CF and control Groups

- 1) On the basis of previous findings it is predicted that there will be no significant differences between the two groups on psychological measures of anxiety, depression or self-esteem.
- 2) There may be differences in type of body-image concerns between the two groups with the CF group expressing more disease-specific concerns with their appearance.
- 3) The CF group will have more dependency on medication, less satisfaction with employment, and more problems with mobility than the control group on the QoL measure.
- 4) The CF group will have spent fewer years in education and be more likely to be unemployed through ill-health than the control group.

General aims

To explore any differences which may be apparent in social support, and relationships and living situations between the groups, and any other differences which may arise in the other QoL domains.

Within the CF group

- 1) It is predicted that there will be a positive association between acceptance of illness and general satisfaction with QoL, positive feelings and employment status.
- 2) It is predicted that there will be a relationship between deprivation category (social class) and severity of illness. No link will be found between severity of illness and

anxiety, depression or self-esteem. There will be a link between QoL facets and severity of illness.

3) A relationship will be present between acceptance of illness and degree of practical and emotional support and between social support measures and severity.

4) There will be no gender differences found on any measures.

General Aims

Further analysis of the relationships between psychological and QoL variables will be conducted.

An exploratory analysis of the interview data will be carried-out in order to identify themes and patterns of responses.

METHOD

1. Design

This study is cross-sectional and therefore measures were taken at one point in time. Self-report questionnaires were administered to subjects in each of the experimental and control groups. In addition, semi-structured interviews were carried out with a subset of the CF subjects in order to obtain qualitative as well as quantitative data.

Subjects

1.1 Experimental Group

This consisted of 30 adult out-patients who attended the Cystic Fibrosis Centre, Edinburgh. Subjects who were included were aged 18 or over, due to the adult focus of the research. All subjects were resident in the East of Scotland and travelled to the Edinburgh clinic for periodic reviews by the CF team. Once ethical approval had been obtained for the study, all clinic attenders aged 18 and over were sent an information sheet about the study and a consent-form (see appendix). They were asked to reply within two weeks and to indicate whether they were willing to be interviewed or to complete questionnaires by post. A reminder letter was sent after two weeks to elicit further responses.

Of the 61 clinic attenders who were contacted, 20 agreed to be interviewed and complete questionnaires (one subject later dropped-out) and 11 agreed to fill-in questionnaires only. 9 subjects indicated that they did not wish to take part and a further 21 failed to respond. The remaining 30 subjects in the experimental group consisted of 15 males and 15 females, with 11 females and 8 males who agreed to be interviewed.

1.2 Control Group

Control subjects were matched with the experimental group for age, sex and deprivation category (Carstairs and Morris 1991). Deprivation categories range from 1 (affluent) to 7 (deprived) and have been developed to correspond with every postcode in Scotland as a way of assessing morbidity, mortality and inequalities in health.

The majority of control subjects were recruited from a general medical practice in the East of Edinburgh. This practice was chosen for its wide range of postcode regions within its catchment area. With the agreement of the GPs and practice manager, a search was conducted on the practice database, by postcode area (corresponding to deprivation category) sex and date of birth. The health status of the subject was ascertained from medical records. In order to fulfil criteria to be “healthy” controls, subjects could have no chronic illnesses, no hospital admissions or referrals to a specialist in the previous year.

18 suitable control subjects were identified and were sent information by post. These subjects were asked to contact the investigator within 14 days if they did not wish to participate in the study. Consent was gained after this period by the investigator telephoning subjects, 17 of whom agreed to receive and complete the questionnaires by post. The remaining 13 control subjects were recruited via hospital staff from three Edinburgh hospitals when the investigator also distributed information sheets about the study. The health status of these controls was assessed through informal questioning using the same criteria as previously. A similar time period was also allowed for these staff to give their consent.

2. Measures

2.1 WHOQOL

The WHOQOL is designed as an international multidimensional quality of life instrument (WHO, 1992, 1993) It is a self-report questionnaire which is easy to administer, it is made up of items which are tied to a particular facet, with facets grouped into a number of domains. There is a five point Likert response scale for each of the items. It is designed to assess the quality of life of people with chronic diseases, disabled people and “healthy” individuals (WHO Group, 1994).

The WHOQOL group consists of international experts in health selected from various fields. It was this group who defined facets as being “a behaviour, state of being, a capacity or potential, or a subjective perception or experience”. A series of focus groups were held with health professionals and seriously ill individuals in order to generate facets and item weightings (Skevington et al 1997). Finally 24 facets of QoL were agreed, each with four items and four additional items relating to general QoL, giving a total of 100 questions. These facets are tied to 6 domains; Physical health, level of independence, psychological health, social relationships, environment, spirituality/religion/personal beliefs.

The reliability and validity of the WHOQOL-100 has been established in field studies (WHO, 1995), Cronbach alpha values demonstrate good internal consistency for the facets with a range of 0.65 to 0.93. Since items are scored by facet and domain, rather than summated from the whole questionnaire, it is possible to use selected facets. One study of QoL in psoriasis patients (WHO, 1995) showed that 12 facets could be used with high levels of reliability and validity when compared to existing questionnaires. The present study used the following 13 facets: Overall QoL, energy and fatigue, sleep and rest, positive feelings, mobility, activities of daily living, dependence on medication or

treatments, working capacity, sexual activity, financial resources, health and social care: availability and quality, participation in new opportunities for recreation, transport.

Facets were selected on the basis of their relevance to patients with CF . Some facets were excluded from the WHOQOL such as personal relationships, self-esteem and body image, which were to be assessed by separate questionnaires in order to gain greater detail in these particular areas. Scores were calculated for each facet .

2.2 Significant Others Scale (SOS) (B)

This measure of social support was developed by Power and Champion (1988) and assesses a number of dimensions of social support. It includes important factors such as structural and functional aspects of social support (Cohen and Wills 1985). The SOS version B asks the subject to list the seven most important people in their lives, thereby assessing network structure and the amount of perceived support is rated on a seven point scale.

Another important distinction which is addressed by the SOS is that between the practical and emotional function of support (Barrera and Ainlay, 1983). Also the SOS allows consideration of the quality of an individual's relationships by assessing the discrepancy between actual and ideal support. Four summary support scores are yielded by the SOS these are : actual Vs ideal x emotional Vs practical. Power et al (1988) indicate that the SOS shows satisfactory validity and reliability.

The scale is available in two versions A and B. Version A stipulates a subset of significant others (Mother, spouse etc.) whereas B allows the subject to nominate important individuals to them. The present study used version B in order to allow CF subjects to nominate a wide range of people (such as health professionals) if they wished.

2.3 Acceptance of Illness Scale (AIS)

Felton et al (1984) developed the AIS since there were very few existing measures which directly assessed adjustment to illness. The majority of instruments in this area assess correlates or outcomes of adjustment such as social functioning or mood. The AIS was based on Linkowski's (1971) scale to measure disability and aims to assess the degree to which individuals can accept their illness without experiencing negative feelings. It is a short scale with eight items, and respondents rate their agreement or disagreement to a number of statements about their illness.

The authors have reported high internal consistency of the scale and reasonable test-retest reliability and construct validity (Felton and Revenson 1984). There are no normative data available but the authors have presented data from a sample of 151 people with one of four chronic illnesses (hypertension, diabetes, cancer or arthritis). Means and standard deviations for each item in this sample are therefore available for comparison with data from the current study. Due to the specific emphasis of this scale on "illness" it was not considered appropriate to administer this questionnaire to the "healthy" control subjects.

2.4 The Hospital Anxiety and Depression Scale (HADS)

This is a fourteen-item scale developed by Zigmond and Snaith (1983) to measure state anxiety and depression. It has the advantage of being short, easy to administer and was designed for use in out-patient clinics to detect clinical levels of anxiety and depression independent of physical symptomatology.

The HADS can also be used to gauge severity of anxiety and depression as four score ranges have been classified; normal, mild, moderate and severe. Validity and reliability of this measure have been established in a study of medical out-patients (Zigmond and

Snaith 1983) and in a sample of cancer patients (Moorey et al 1991). These authors also reported on its high rate of internal consistency as assessed by Cronbach's alpha which was 0.93 for anxiety and 0.9 for depression. It may be a particularly useful measure in the current study for detecting differing levels of mood states independent of the symptoms of CF.

2.5 Rosenberg Self-Esteem Scale

This is the most widely used measure of self-esteem and was developed by Rosenberg in 1965 and reprinted in 1989. It consists of 10 items to which the subject responds on a four point scale of agreement.

There has been little research evaluating the psychometric properties of this scale (Bowling, 1995), however norms from the general adult population are available for comparison. The advantage of this scale is that it directly measures perceived self-esteem as opposed to the more general construct of self-concept. This is preferable in the current study since comparisons will be conducted between self-esteem and other dimensions of self-concept such as body image.

2.6 Body Image Questionnaire

This short scale was developed for the study by the investigator (see appendix). There are few existing scales which assess body image separately from body disparagement and include the concept of perceived attractiveness. To assess body image in the present study, a general questionnaire was required that still allowed respondents to state specific body image concerns which were likely to be different for the control and experimental groups.

The questionnaire was based on the four item body-image facet of the WHOQOL. One item about acceptance of bodily appearance was removed and substituted with a question about perceived attractiveness to others. The remaining three questions covered general satisfaction with appearance, aspects of their appearance which the respondent wished to change and feeling inhibited by their appearance. The extent of the subject's agreement or satisfaction with each item was assessed on a 5 point Likert- type scale, giving total scores ranging from 4-20, with 20 indicating a positive body image.

Respondents were asked to comment as fully as possible on specific aspects of their appearance they may wish to change and on situations in which they had felt inhibited by their appearance. Internal consistency of this scale was shown to be high when calculated using SPSS for windows; (Cronbach's alpha = 0.806).

2.7 Other data collected

A general information sheet was also included with the other questionnaires to gather demographic data such as years spent in education, occupation, expectations of working in the next year, who the subject lives with and their marital/ relationship status. Post code and corresponding deprivation category were also calculated for the experimental group, and control subjects were matched to this.

Age at diagnosis (when known) and other measures of physical functioning, were collected from the medical notes for all CF patients over 18 including non-responders. This was in order to assess whether the experimental group was representative of the patient sample as a whole on these parameters. Lung function is commonly measured by forced expiratory volume at one second (FEV1) and this is considered to be one of the more stable markers for adult patients with moderate to severe CF (Ramsey and Marshall 1995). Each CF patient is assigned a predicted FEV1 based on their gender, age and height. FEV1 % predicted is an indicator of the capacity at which an individual is

functioning, for example at 60% of their predicted FEV1. In the current study, the best FEV1 score in the previous 6 months was used to calculate FEV1% predicted, since often lung function can deteriorate with infections, so a higher FEV1 would indicate a stable period.

Other physical measures which were collected were body mass index. This consists of; weight in kilograms divided by height in metres squared, and gives some indication of the nutritional status of CF patients . A clinical rating of severity was also carried out by a Senior Registrar and a nurse from the CF team for the thirty subjects in the experimental group only. Limitations of time meant that no inter-rater reliability could be checked. This was called the Shwachman score (Shwachman, 1965) and was developed for clinical use, it classifies CF patients into severe, moderate, mild, good, excellent depending on their total score. The final rating is based on the sum of scores across four parameters; general activity, physical examination, nutrition and chest x-ray. It is not an objective measure since it relies on the perception of the health professionals involved and may also include “hidden” factors such as compliance with treatments. For these reasons it was considered to be a useful adjunct to the objective physical measures used in the present study.

3. Procedure

Subjects in the experimental group who indicated that they wished to complete questionnaires only, were sent them by post with a stamped addressed envelope. All control subjects recruited from the GP practice followed the same procedure, controls who were hospital staff generally returned questionnaires in person.

The 19 CF subjects who agreed to be interviewed had supplied their telephone number. They were contacted by the investigator to arrange a time and location (in the hospital or in their own home) at their convenience. At this point the possibility of tape-recording the interview was mentioned again and only one subject had any objection to this. The

majority of patients preferred to attend the hospital for their interview, particularly those living locally or those who already had clinic visits arranged. Interviews in a subject's home were conducted in four cases and two subjects were in-patients at the time of their interview but were still willing to take part.

Interview conditions were constant in every setting. Interviews took place in a private room with no interruptions and lasted approximately 40 minutes to one hour.

Questionnaires were administered to subjects prior to interview so as to reduce the bias between these responses and those of subjects who were not interviewed. The investigator did not read medical notes or obtain any other prior information about the subjects in order not to influence the course of the interview. Before the interview, all subjects were informed again about the purpose of the study, about the role of the investigator as being separate from the CF team. Confidentiality was also assured and subjects were told under what circumstances information would be fed-back to the CF team (i.e. If they were exhibiting signs of significant distress, or scored as being so on the questionnaires).

3.1 The Interview

This was a semi-structured interview in which the investigator wished to cover a range of issues identified from the literature. An interview schedule was constructed, but this was flexible in format in order to allow the investigator to easily adapt it to each subject's experiences. Sample questions from the interview are also contained in the appendix. The schedule was as follows:

General information

- family background

-education -

employment history (how the above were affected by CF)

Diagnosis

- age and impact on individual/ family
- stages of adjustment, e.g. Teenage years
- siblings with CF?

Relationships

- stigma, disclosing to others, how and when - problems with employers
- Who have been the most/least helpful people
- forming intimate relationships, (fertility issues/ having children)
- strain on relationships
- networks (involvement with others with CF or CF-related activities)

Quality of life

- most helpful treatments
- treatments (or regimens) which negatively affect QoL, compliance.
- activities avoided or prevented from doing due to CF
- how does CF affect QoL generally
- financial concerns, benefits, transport.
- obtaining mortgages, insurance
- autonomy and independence

Bereavement Issues

- worries about the future, about themselves or dependants
- spiritual beliefs/ other coping strategies
- who can they talk to about dying?
- degree of acceptance of the future generally

Information

- satisfaction with medical care
- how informed do they feel about CF?
- how much information do they want? -coping style
- how could services be improved?

4. Data Analysis

Data were analysed using SPSS for windows. The comparison of means between the two groups consisted of t-tests and non parametric tests. Pearson's r correlation and other non-parametric statistics were carried out for bivariate analysis within the CF group. Finally a linear regression analysis was employed to examine the variance in two important variables in the CF group.

RESULTS

1. Demographic Data

Demographic details are presented in table 1. Since the two groups were matched for age and deprivation category, there were no differences between them on these variables. Control subjects matched to CF subjects who had missing deprivation categories were coded as category 3. The frequency of deprivation categories is illustrated in figure 1, showing the majority of subjects came from middle class categories 2 and 3 with none from highly deprived areas (categories 6 and 7).

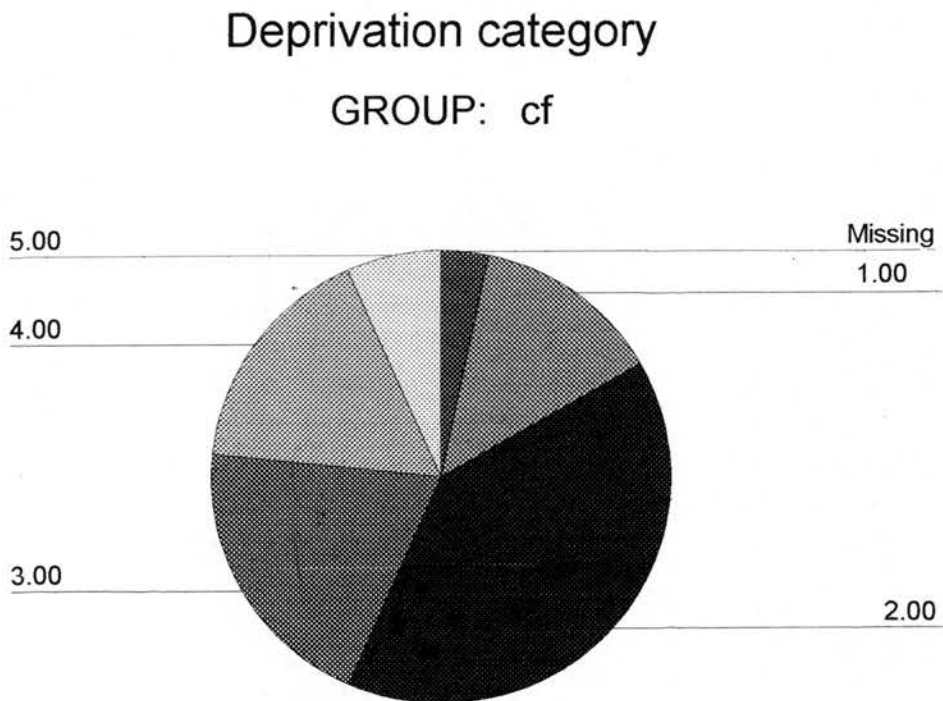


Fig.1



Demographic data

Table 1.

Var	CF group N=30					Control group N=30					t test / Wilcoxon test	
	N	Mean	SD	Min	Max	N	Mean	SD	Min	Max	t / Z	p
Age		29.17	8.18	18	49		29.7	7.7	17	48	t=0.28	n.s.
Female	15					15						
Male	15					15						
Deprivation Category		2.62		1	5		2.57	0.94	1	5	Z=0.05	n.s.
Years in** Education		6.73	2.2	4	12		8.63	3.15	4	14	t=2.72	0.009

*p<0.05; **p<0.01

The CF group spent significantly less years in education from age 11 than the controls (t=2.72, p=.0009) with the modal number of years being 5 for the CF group and 6 for the controls (see figures 2 and 3). Years spent in education was also negatively correlated with deprivation category within the CF group (Pearsons r correlation=-0.41 p<0.05), indicating that those subjects living in more deprived areas had spent less years in education.

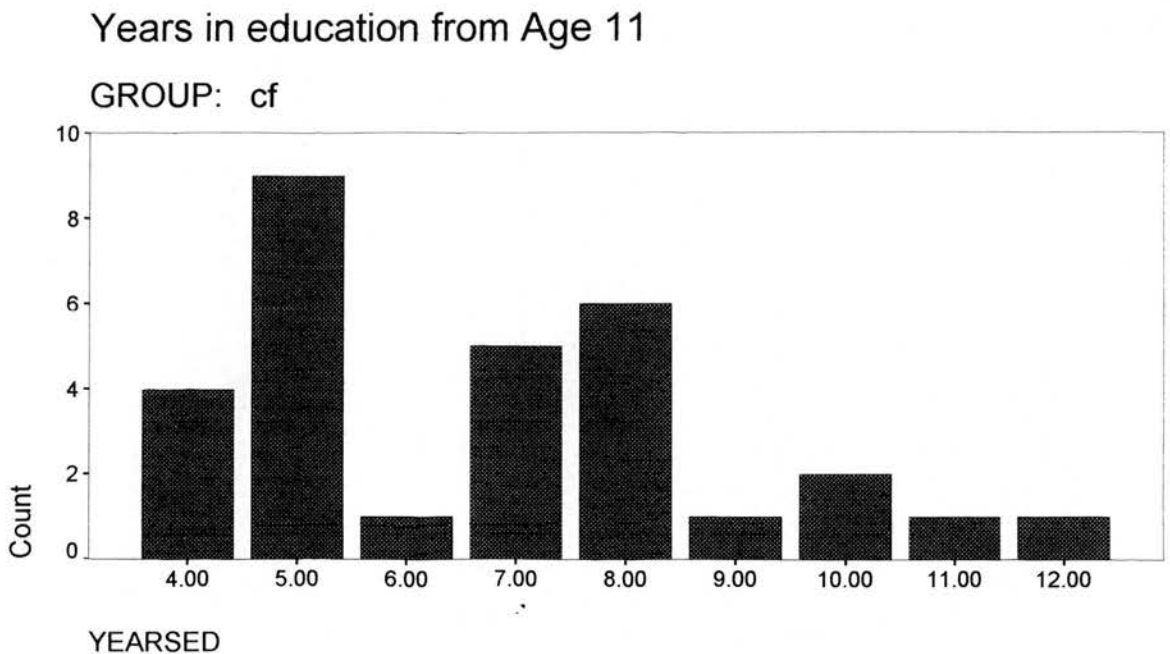


Fig.2

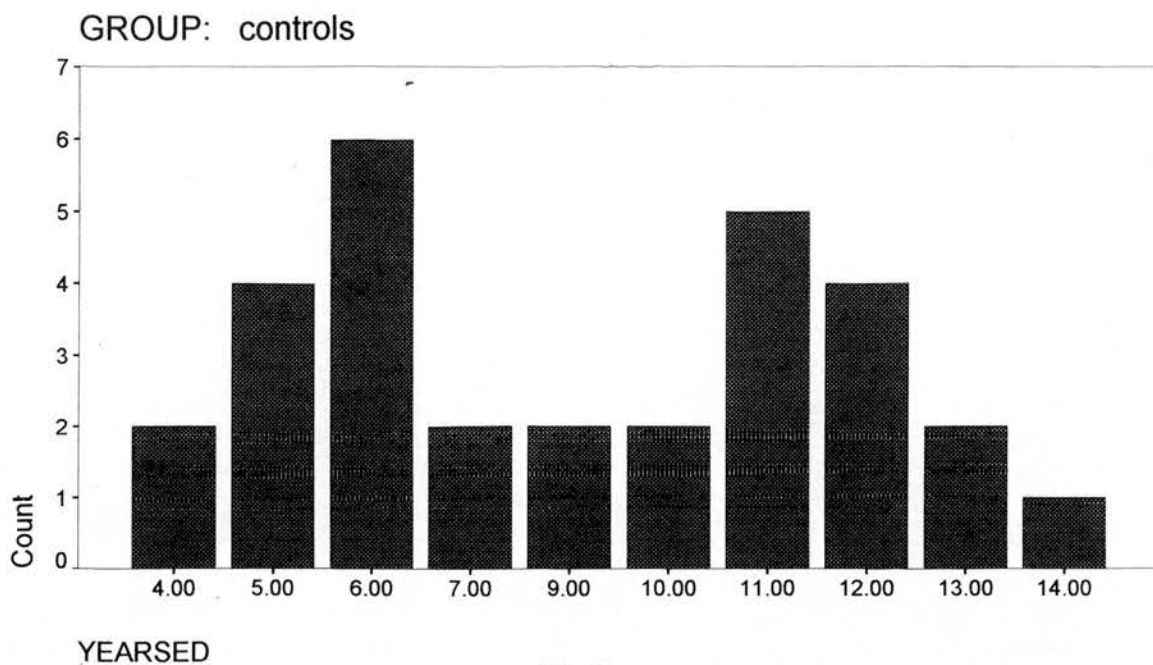


Fig.3

Age at diagnosis is illustrated in figure 4 for the CF group. In the majority of cases CF had been diagnosed in the first few months after birth and usually by the age of one with another sub-group being diagnosed in their early teens. Those who were diagnosed later were living in less deprived areas (Pearsons r correlation=-0.43, $p < 0.05$).

Age at diagnosis

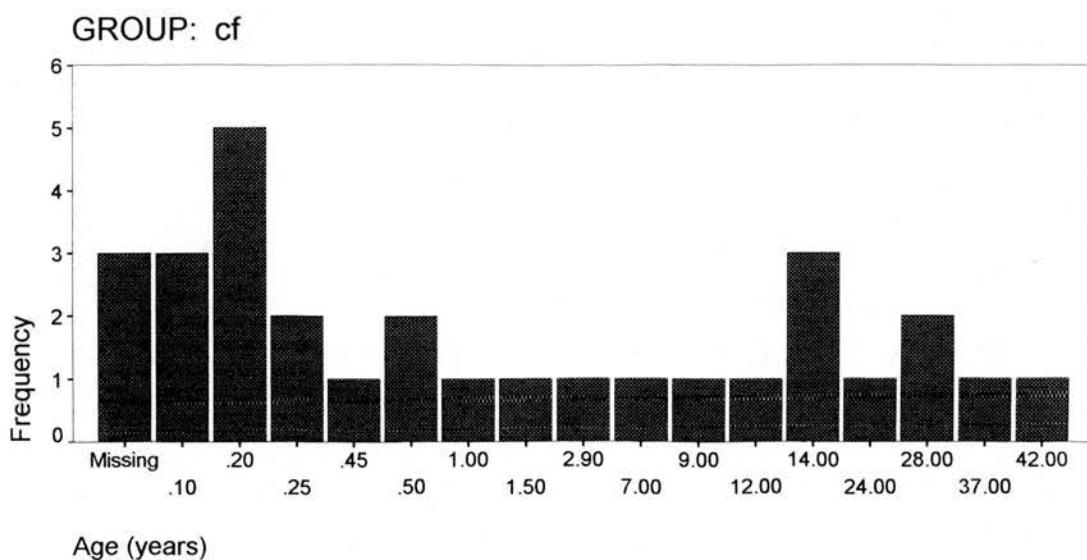


Fig.4

Figures 5 and 6 show working status in the CF and control groups with 40% unemployment due to ill health only in the CF group. The category “unemployed through personal choice” includes waiting to return to higher education, and choosing to look after children at home. The employed category includes those in higher education and voluntary work. These categories were collapsed to three for statistical analysis which indicated that employment status differed significantly between the groups (Chi square=10.34, p=0.001) with CF subjects less likely to be employed at the time of the study than the control group.

Of those who were working, six CF subjects were employed in indoor manual work, and one in outdoor manual work. In the control group, three subjects worked in indoor manual jobs and two worked in active outdoor employment.

Work status in the next year relates to the subjects’ expectation to work in the next year. In both groups, the percentage of subjects expecting to work increased to 56.7% in the CF group and 90% in the control group again giving a statistically significant between groups difference (Chi square=8.52, p=0.004) (see table 3).

Working Status

GROUP: CF

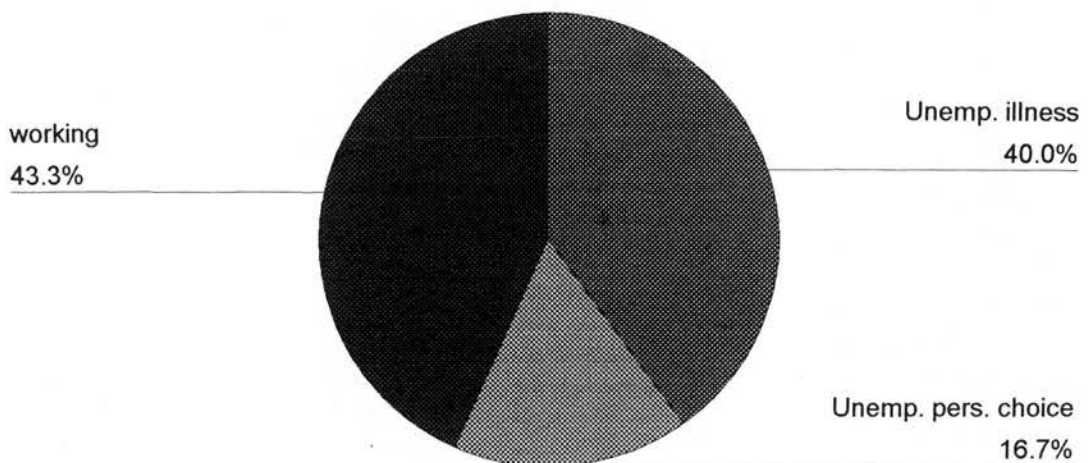


fig.5

Working Status

GROUP: Controls

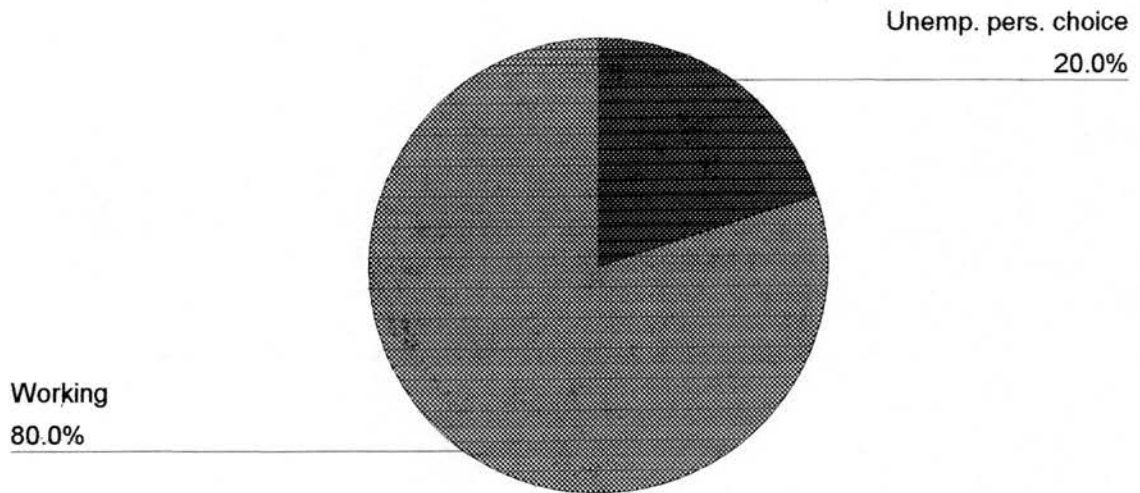


Fig. 6

The CF and control groups had identical numbers of single subjects and those in a relationship. 10% of CF subjects were separated or divorced and there were none in the control group. This percentage were re-coded as single people for analysis, with two other categories: married or cohabiting, and being in a relationship. Between the two groups, there were no significant differences in the proportion of subjects within these categories. (See figures 7 and 8 and table 3.)

Relationship Status: CF Group

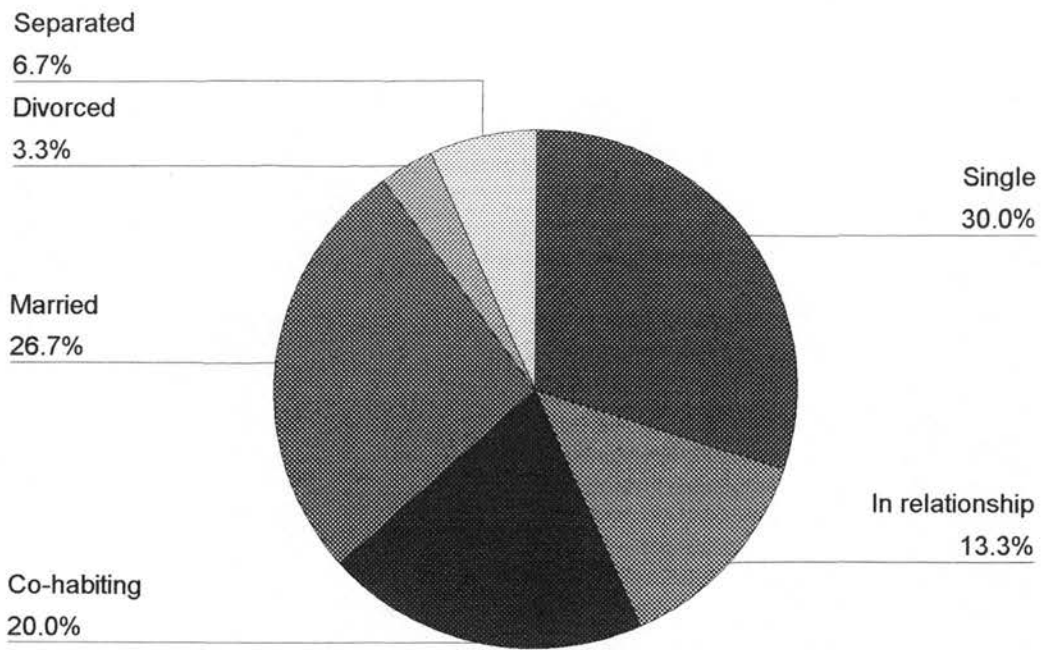


Fig. 7

Relationship Status: Control Group

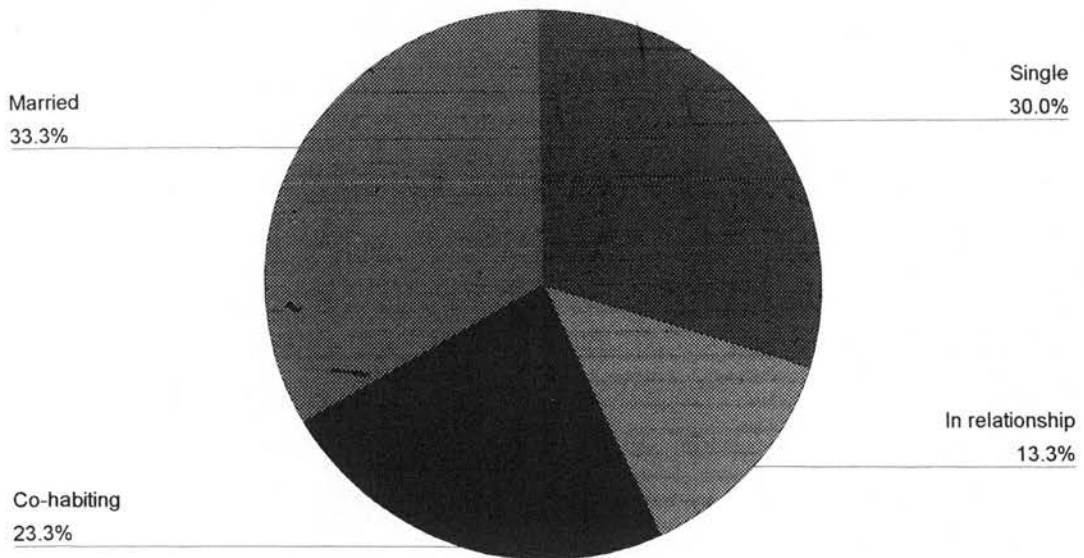


Fig. 8

Similarly there were no significant differences found between the two groups in terms of their living situations although figures 9 and 10 did suggest slightly differing frequencies. 20% of CF subjects lived with their parents as opposed to 10% in the control group and 20% lived alone and 13.3 % with flatmates as compared with 10% and 23.3% respectively in the control group.

Living Situation: CF Group

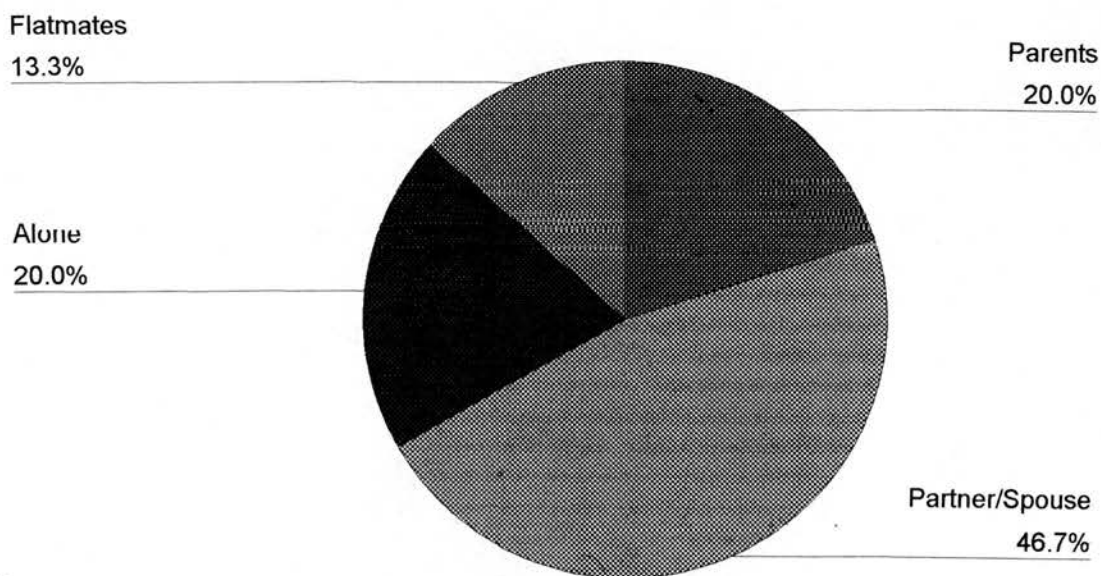


Fig. 9

Control Group

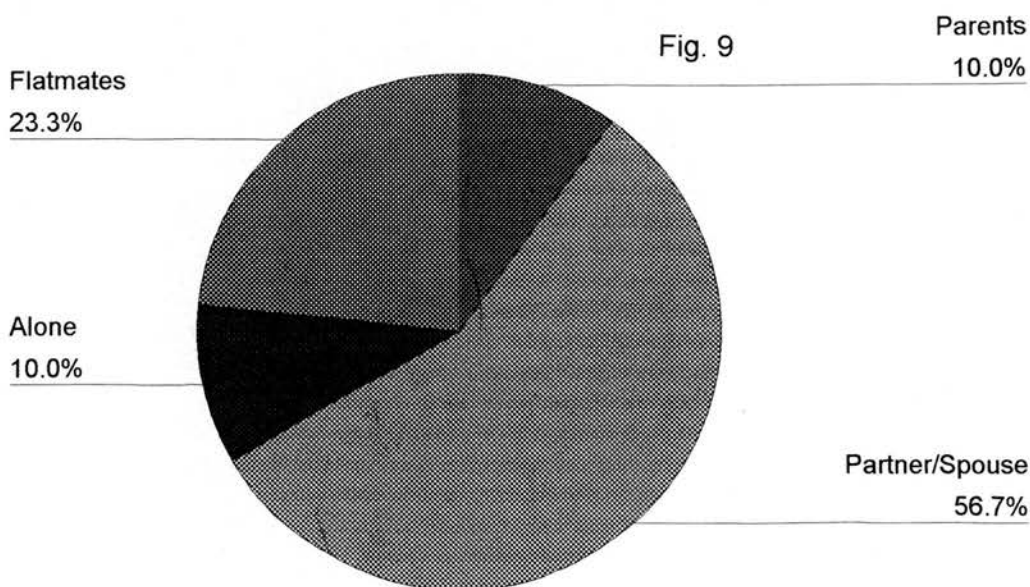


Fig. 10 57

1.1 Demographic data and measures of CF severity

FEV1 % predicted (forced expiratory volume at 1 second) the physical test indicating the percentage of lung capacity which is functioning, was found to be negatively correlated with age (Pearsons $r=-0.39$, $p<0.05$). Therefore lung function deteriorates with age. No other associations were found between physical measures of severity (Body mass index (BMI) and Shwachman clinical rating of severity) and demographic data (see table 7)

1.2 Non-participating CF subjects

The group of CF subjects who did not take part in the study (N=31) were compared with the participating CF group on a number of demographic and severity variables (see table 2). No significant between groups differences were evident suggesting that the experimental group were a representative sample from the CF population attending the clinic.

Comparison between CF experimental and non-participant groups (t-test and Wilcoxon test)

Table 2.

Var	CF group N=30		CF nonparticipants N=31			
	Mean	SD	Mean	SD	t / Z	p
Age	29.17	8.18	26.51	6.21	t=1.42	n.s
Deprivation Category	2.62	1.15	2.83	0.72	Z=1.12	n.s
Age at diagnosis	8.8	12.5	6.13	8.95	t=0.89	n.s
FEV1 % pred	60.11	22.73	63.00	25.4	t=0.46	n.s
BMI	21.49	8.18	21.09	2.29	t=0.75	n.s

2. Social Support

The Significant Others Questionnaire (SOS) measures actual emotional (EM) and practical support (PR), ideal emotional and practical support IEM and IPR and the discrepancies between actual and ideal supports (DEM and DPR). No significant between groups' differences were found on any of these measures except for actual emotional support (EM) which was higher in the CF group (Wilcoxon test $Z=2.06$, $p=0.039$) (see table 3)

Social support and employment: Between groups analyses (Wilcoxon test and Chi-square test)

Table 3.

Variable	CF group		Control group		Z	χ^2	P
	Mean	SD	Mean	SD			
Actual Emot. Support* (EM)	5.73	0.82	5.33	0.84	2.06	-	0.039
Actual Pract. Support (PR)	5.36	0.91	5.18	1.07	0.71	-	n.s.
Ideal Em. Support (IEM)	6.35	0.65	6.20	0.62	1.18	-	n.s.
Ideal Pract Support (IPR)	5.89	0.89	5.97	0.75	0.13	-	n.s.
Discrepancy (DEM)	0.61	0.59	0.88	0.72	1.39	-	n.s.
Discrepancy (DPR)	0.56	0.52	0.82	0.755	1.34	-	n.s.
Relationship Status	-	-	-	-	-	0.62	n.s.
Living Situation	-	-	-	-	-	2.86	n.s.
Work status ***	-	-	-	-	-	10.34	0.001
Work status in next yr.**	-	-	-	-	-	8.52	0.004

* $p < 0.05$; ** $p < 0.01$; *** $p < 0.001$.

A gender difference was also found in social support. Women with and without CF received significantly more practical support than men and they had higher levels of ideal practical support than their male counterparts in the control group. See table below:

Gender differences in practical social support

Table 4.

CF GROUP	MALE		FEMALE		Wilcoxon test	
	Mean	SD	Mean	SD	Z	p
Practical Support	4.98	0.90	5.75	0.77	2.18*	.029
Ideal Practical Support	5.56	1.05	6.21	0.57	1.76	n.s.
CONTROL GROUP	Mean	SD	Mean	SD	Z	p
Practical Support	4.7	0.97	5.66	0.96	2.40*	.016
Ideal Practical Support	5.6	0.71	6.34	0.61	2.95**	.003

*p <0.01; ** p<0.05

There were no other gender differences found in any of the data.

3. Psychological Factors

There were no significant between groups differences for anxiety or depression HADS scores. See (figures 11-14). In the CF group 20.5% (8) of the group scored above the cut-off for possible clinical anxiety in the borderline and moderate range, and 10% scored above the cut-off for depression but still remained in the borderline range (8-10). Control subjects scores were slightly more widely distributed with 1 subject in the moderately depressed range (11-15) and 3 subjects who scored as moderately anxious and one as severely so.

**Between groups analyses of psychological and QoL data
(t-tests)**

Table 5.

Variable	CF group		Control group		t	p
	Mean	SD	Mean	SD		
HAD anxiety	6.37	3.59	5.93	3.81	0.45	n.s.
HAD depression	3.03	2.80	2.4	2.50	0.93	n.s.
Self esteem	18.7	5.65	18.1	4.37	0.46	n.s.
Body image Q.4*	3.20	1.32	3.86	0.86	2.31	0.025
Body image total	13.23	4.17	14.8	2.54	1.76	n.s.
Energy	12.53	1.55	13.03	1.54	1.25	n.s.
Sleep*	10.63	1.16	11.27	1.14	2.13	0.037
Positive Feelings	13.8	3.03	14.5	2.39	1.09	n.s.
Mobility	12.23	1.41	11.8	1.16	1.30	n.s.
Activity	12.1	1.09	11.73	1.39	1.14	n.s.
Medication ***	11.23	3.72	5.23	2.43	7.40	0.0001
Work (capacity)***	14.10	4.79	17.83	2.42	3.81	0.0001
Sexual Satis.	11.48	2.78	12.2	2.41	1.02	n.s.
Finances	11.20	1.24	11.8	1.35	1.79	n.s.
Services	15.19	2.57	14.26	2.43	1.43	n.s.
Leisure	14.87	2.71	15.23	2.27	0.57	n.s.
Transport	11.23	1.00	11.73	2.08	1.18	n.s.
Overall QoL	14.53	3.60	15.80	3.30	1.42	n.s.

*p < 0.05; **p < 0.01; ***p < 0.001.

HADS Anxiety scores

GROUP: CF

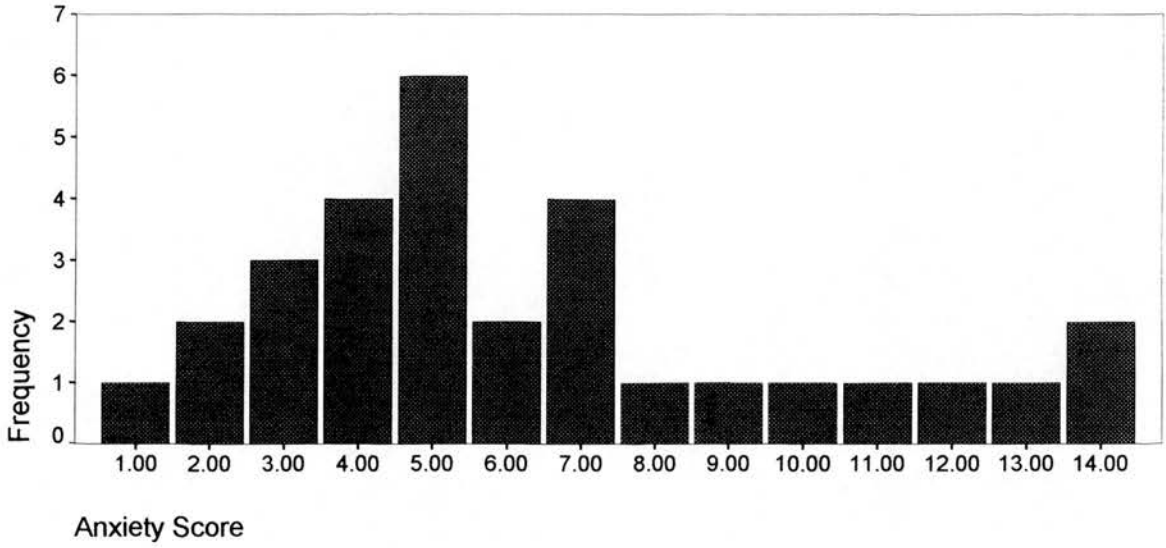


Fig. 11

GROUP: Controls

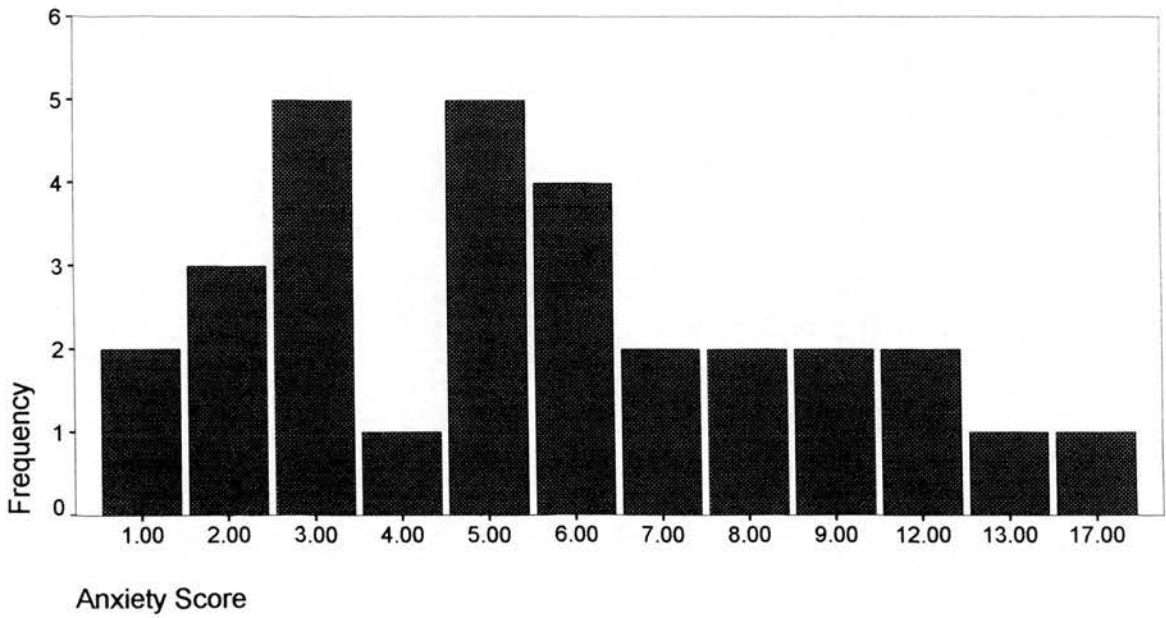


Fig. 12

HADS Depression Scores

GROUP: CF

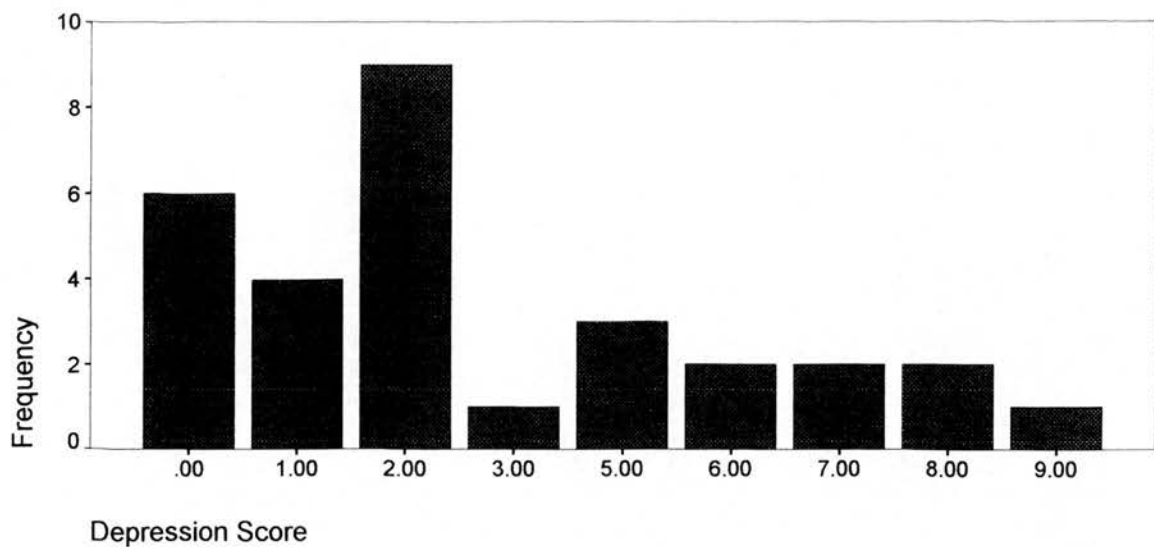


Fig. 13

GROUP: Controls

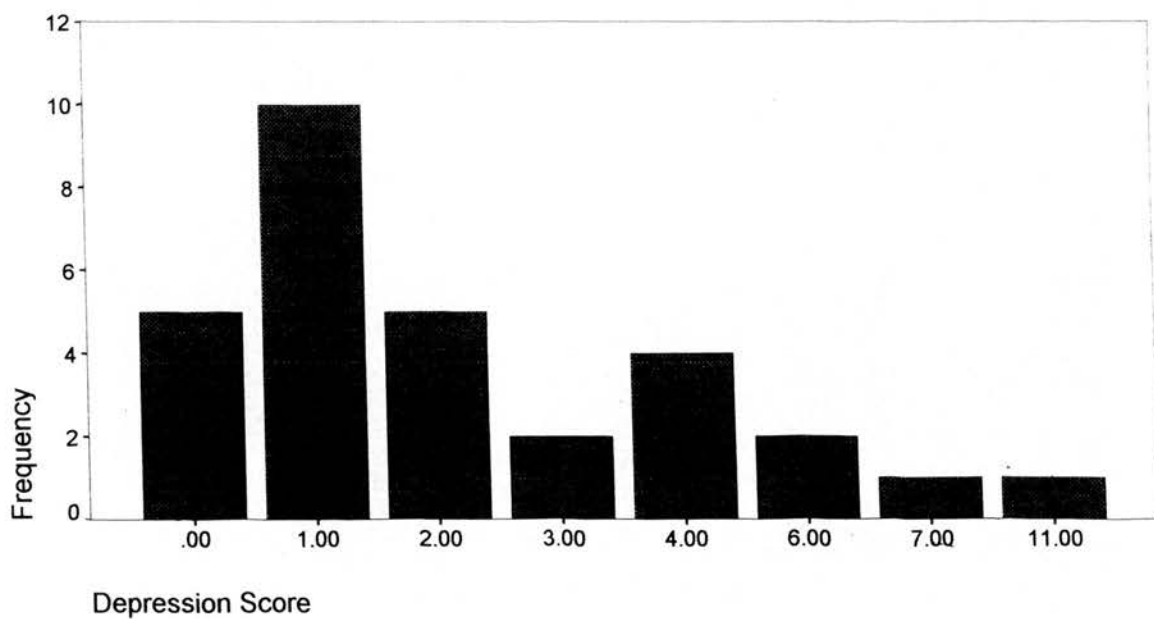


Fig. 14

Self esteem scores were also very similar for both groups with no significant differences found. The mean scores in both the CF and control groups were considerably lower than published norms by Rosenberg (1989) from a general population study (mean 34.73, standard deviation of 4.86) . This suggests that the study sample had higher self-esteem than the general population.

Acceptance of illness measured only in the CF group was generally high with a mean total score of 31.5 out of a possible total score of 40. The standard deviation was 5.52 with a minimum score of 20 and maximum of 40. Acceptance of illness was slightly higher than norms published for 151 patients with a chronic illness (Felton and Revenson 1984) who reported a mean score of 28 with a standard deviation of 5.6.

Comparison of the two groups on the total score from the body image questionnaire revealed no overall significant differences, with 80% of the CF group and 86.7% of the controls scoring as being moderately or highly satisfied with their appearance. However, item four of the questionnaire relates to any changes which a subject might wish to make to their physical appearance and the CF group scored as wishing to make significantly more changes ($t=2.31$, $p=0.025$). Comments about these changes are analysed further in the qualitative analysis section.

Correlations between psychological measures were high for the CF group with acceptance of illness being positively correlated with body image total score (Pearsons r correlation= 0.38 $p<0.05$). Acceptance of illness was negatively correlated with self esteem score (-0.53 $p<0.01$) and with depression score(-0.55 , $p<0.01$). No relationship was evident between acceptance of illness and anxiety. These findings all indicate that a higher acceptance of illness is associated with less psychological distress.

4. Quality of Life Facets

As illustrated in table 5, of the thirteen QoL facets investigated, the CF and control groups differed significantly on three. CF subjects showed much greater dependence on medication, much less satisfaction with their working capacity and slightly less satisfaction with their sleep.

5. Correlations within the CF group

Between the psychological and QoL data there were several highly significant correlations (see table 6). Positive feelings and overall QoL were related to all measures of psychological well-being. Greater dependence on medication was only related to a lower acceptance of illness and not to other measures of distress. Less satisfaction with sleep, services and opportunities for leisure were also correlated with greater psychological distress and a lower acceptance of illness.

Reduced satisfaction with working capacity was related to depression, low self-esteem and a lower acceptance of illness. Satisfaction with working capacity was also related to employment status only in the CF group (Kruskal Wallis 1-way Anova=5.79 ; p=0.16). The data suggested that being in employment was related to greater satisfaction with working capacity. Being in employment also appeared to be associated with a higher acceptance of illness (Kruskal Wallis 1-way Anova=4.31; P=0.03).

**Significant Correlations Within the CF Group: QoL and Psychological Factors
(Pearsons r correlation)**

Table 6.

QoL Facets									
Variab. Groups		Pos. feelin.	Medi-cation.	Sex Satis.	Services	Leis	Work Satis.	Sleep	Overall Qol
Psychological	Anxiety	-.60**			-.57**	-.57**		-.43*	-.53**
	Dep	-.71**			-.58**	-.50**	-.40*	-.45**	-.60**
	SE	-.61**		-.48*	-.58**	-.52**	-.48**	-.55**	-.55**
	A.I.S.	.51**	-.53**		.38*	.46*	.52**	.49**	-.53**
	B.I.S.	.65**		.41*	.62**	.40*		.36*	.50**
Demographic	Age	-.64**			-.37*	-.38*			-.57**
	FEV1 % p.	.43*		.59**			.40*		.57**
Physical.	Shwac. Rating			.46*			.51**		.43*

* p< .05; ** p< .01

Key: A.I.S.=Acceptance of illness scale; B.I.S=Body image scale; FEV1% p.=FEV1% predicted; Dep=Depression score.

Satisfaction with sexual functioning was only associated with two psychological factors poorer self-esteem and body-image. Sexual satisfaction was related to relationship status in both groups with a slightly higher level of significance in the CF group (Kruskal-Wallis 1-way Anova=9.7 ; p=0.007) than in the control group (Kruskal -Wallis 1-way Anova=8.8 ; p=0.01). The data suggested that those subjects who were single experienced reduced sexual satisfaction as seen in figures 15 and 16.

RELATIONSHIP STATUS AND SATISFACTION WITH SEXUAL FUNCTIONING

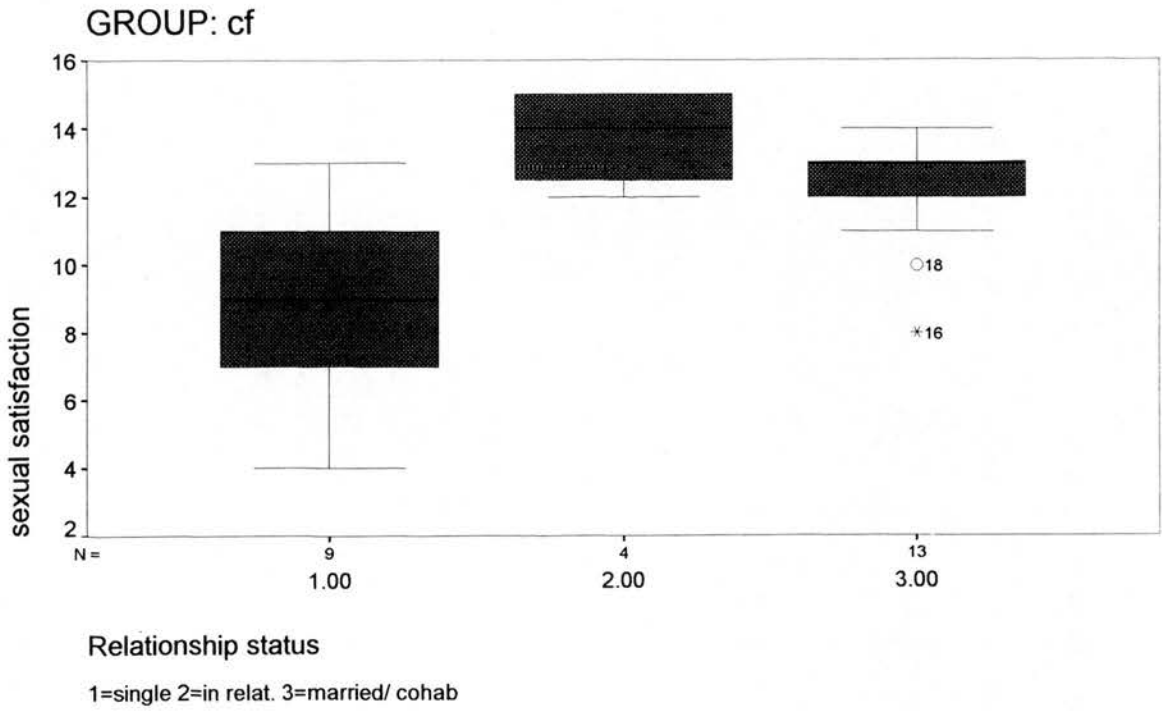


Fig. 15

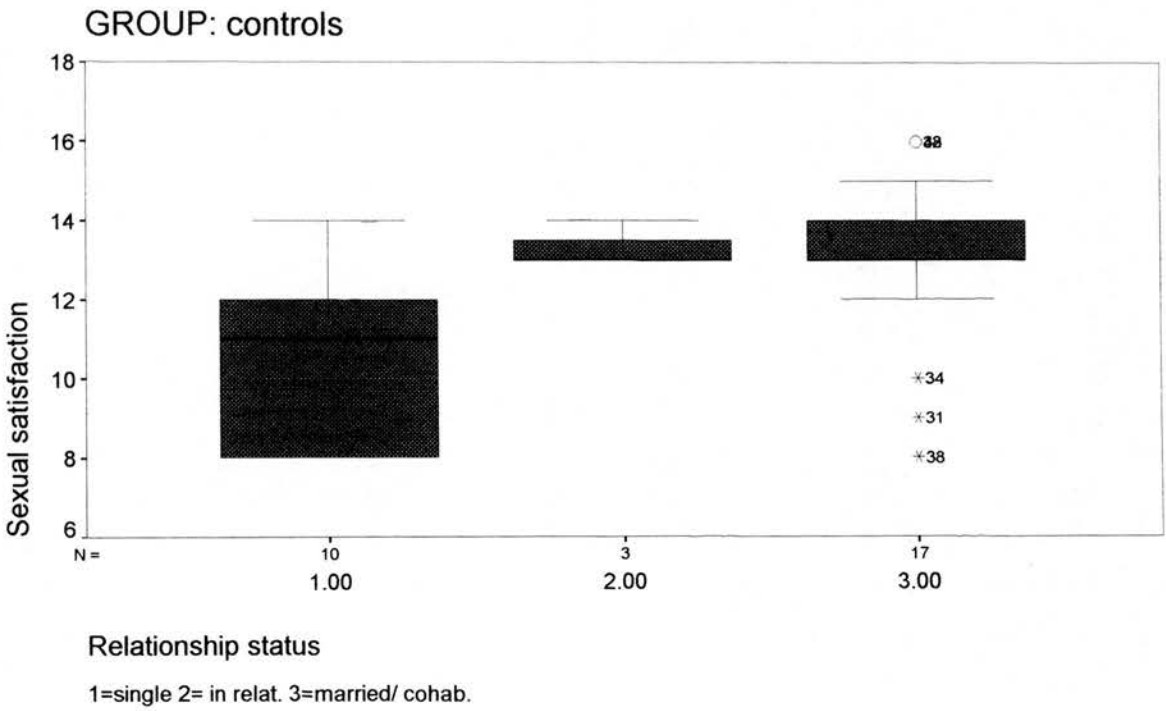


Fig. 16

Measures of physical functioning, FEV1 % predicted and Shwacman score were positively correlated to sexual functioning and working capacity and overall QoL. Poorer lung function was also associated with less acceptance of illness and a poorer body image (see table 7). Body mass index (BMI) was not associated to body image but was positively correlated to acceptance of illness .

**Significant Correlations within the CF group
Demographic, psychological and physical measures.
(Pearsons r correlation)**

Table 7

Variab. Type		Demographic Variables				Physical Meas.	
		Age Diag.	Yrs in Ed	Deprivat. category	Age	BMI	FEV1 % Pred.
Psychological	Anxiety				.43*		
	Dep				.46*		
	SE				.47**		
	A.I.S.				-.51**	.43*	.62**
	B.I.S.				-.53**		.41*
Demog.	Age	.62**					
	Depcat.	-.43*	-.41*				
Physic.	FEV1 % p.				-.39*	.49**	

*p < .05; **p < .01

Key: AIS=Acceptance of illness scale; B.I.S=Body image scale; Depcat=Deprivation Category; FEV1% p.=FEV1% predicted; Dep=Depression score.

Psychological distress increased with age; positive feelings and general QoL decreased with age; lung function (FEV1 % predicted) was poorer in older CF subjects. Age at

diagnosis was not related to any psychological or QoL data, but older CF subjects had been diagnosed later.

6. Predictors of Acceptance of Illness and Quality of Life Scores.

A hierarchical regression analysis was undertaken in order to determine which variables best predicted acceptance of illness and QoL. Table 9 summarizes the steps in the first analysis. Variables entered at each step were chosen for their correlational association with the dependent variable, in this case acceptance of illness. Age was the first demographic variable entered; the second step comprised of physical measures of severity, FEV1 % predicted and Body Mass Index (BMI); the third step consisted of psychological and QoL factors; depression and working capacity.

Hierarchical Regression of Acceptance of Illness scores with age, physical severity variables and depression and working capacity

Table 9.

		R ²	β	t	F	R ² Change	F
Step 1	Age	0.26	-0.50	3.02**	9.58**		
Step 2	FEV1 % p. BMI	0.47	0.44 0.11	2.476* 0.67 n.s	7.18***	0.21	5.15* p=.025
Step 3	Depression Work Sastis.	0.61	-0.25 0.25	1.6 n.s. 1.58 n.s.		0.14	4.31*
All predictors		0.52 (adj. R ²)			6.86***		

*p<0.05; **p<0.01; ***p<0.001; R²cumulative multiple R²

When all the variables were considered together, they accounted for 52% of the overall variance in acceptance of illness scores. Acceptance of illness was negatively related to age and positively related to FEV1 % predicted. The introduction of age at the first step ($R^2=0.26$) accounted for around 50% of the final variance (R^2 adjusted=0.52).

FEV1% predicted also contributed independently a further significant proportion of the explained variance, with BMI and variables in the third step (depression and working capacity) exerting no significant effect on the overall R^2 .

There was a significant change in R^2 between steps one and two and steps two and three. This suggests that groups of variables at each step made a significant combined contribution to the overall variance. Physical variables were stronger predictors than psychological factors.

Findings from a second regression analysis of the facet overall QoL are summarized in table 10 as follows:

**Hierarchical Regression of Overall QoL scores with Age, lung function,
Psychological variables and QoL facets**

Table 10.

		R ²	β	t	F	R ² Change	F
Step 1	Age	0.32	-0.57	3.62***	13.14***		
Step 2	FEV1 % predicted	0.44	0.42	2.30*	9.86***	0.12	5.79* p=.025
Step 3 (Psychological)	Depression Acceptance of illness	0.56	-0.41 -0.09	2.41 * 0.41 n.s.		0.12	3.41*
Step 4 (QoL Facets)	Leisure opportunit. Satisfact sex life Satis.services Work Satis.	0.80	0.16 0.21 0.02 0.06	1.15 n.s. 1.09 n.s. 0.09 n.s. 0.30 n.s.		0.24	6.3**
All predictors		0.70 (adj. R ²)			7.82***		

*p<0.05; **p<0.01; ***p<0.001; R²=cumulative multiple R2.

All the variables together accounted for 70% of the variance in overall QoL scores.

Age was found to be strongly related to overall quality of life. As before age was entered as step one and the R² value (0.32) accounted for around 42% of the overall variance (R² adjusted=0.70). FEV1 % predicted and depression also exerted a significant effect, but acceptance of illness did not. The four QoL facets individually made no significant contributions to the overall QoL, but together accounted for a significant change in R² between steps three and four, and were more strongly predictive than the other sets of variables. The other steps had also made a significant difference in change in R² which suggested again that physical variables made a greater contribution to predicting variance than psychological factors.

7. Qualitative Data

Body Image

Comments were collated from the body image questionnaire indicated that seventeen CF subjects reported CF specific aspects of their physical appearance with which they were dissatisfied. Three subjects reported a desire to lose weight or “tone-up” as did fifteen subjects in the control group. The remainder of subjects fifteen controls and ten CF subjects either did not comment or commented on other aspects of their appearance (e.g. thinning hair etc.)

CF specific aspects of body image were as follows: Hunched shoulders, wish to gain weight (in 10 cases), to be taller, skinny legs and arms, delayed in development, bad skin (due to medication), barrel chest, swollen face. Six respondents said they would avoid swimming or sunbathing because it would reveal their physique. Two subjects felt that their poor body image had affected their confidence in dating and socializing.

Interview data

The interview data was analyzed thematically for each respondent. Due to the constraints of time a full analysis of all the data was not conducted, but the most salient findings are presented as follows. The letter in brackets denotes the sex of the respondent:

Diagnosis

Four of the respondents had been diagnosed in their twenties or later. There were particular issues for them. Those diagnosed earlier had more issues relating to their prognosis, and often had older siblings with CF.

R28 (M) (Diagnosed at 5 mths) “ I feel lucky ‘cos later on more damage is done to the lungs”

Late Diagnosis

R23(F) "I used to smoke, I probably wouldn't have started if I'd known I had CF, but who knows when your a teenager"

R15(F) "Oh good I'm not going mad, there is something wrong with me , cancer and even AIDS had run through my mind"

R 15 (diagnosed age 25) "It would have been better for my health if I'd been diagnosed earlier but I don't think I would have wanted to know because I may have been molly-coddled more or scared to do things"

Late diagnosis also meant that fertility problems had been unexplained for two of the male respondents :

R12(M) "We'd been trying for a child for a long time...I put it down to tuberculosis"

Prognosis

Seven respondents who were diagnosed in childhood remember being given a short life expectancy which they have since outlived:

R22(F) "My parents were told that I wouldn't see the next birthday, then I wouldn't see the next, until I was 15 then they (the medical profession) gave up"

R25(M) "They tell you when you're little you won't make it through your teenage years...well I'm 24 what do you think of that!"

School

Only two respondents reported any serious disruption to their schooling through ill-health. Three female respondents were bullied at school directly because of their CF.

R28(M) "I still did P.E and had no time off school"

R28 (M) "Everyone knew, I always told everyone (at school) everyone just treated me normal"

R31(F) "I got bullied at school, I got called a spastic, my sister (who also had CF) always stood up for me, it made it easier, we knew we had that extra support"

R31(F) "As a teenager, people wrapped me up in cotton wool, my family and friends and even teachers, it wasn't fair to the other kids"

R9(F) "CF affected my schooling dreadfully, being diagnosed at 12, I'd just started a new school and I became quite ill, then I changed schools...it was a complete disaster area"

Disclosure

The majority of respondents reported being able to disclose more to people as they have grown older. Three respondents gave a talk about C.F. to their classmates and found this a very positive experience both in terms of responses from classmates and increasing their own confidence to disclose. Reactions to disclosure had generally been positive.

R13(M) "Until 3 years ago I never told anyone I had it, If people used to ask before I used to play it down fob them off, but I've had no bad reactions from people I have told....I was more worried people would start doing things for me"

R15(F) "I don't talk about it much, my close friends know, but I don't tell them if I'm going to the clinic"

R19(F) "In secondary school I did a talk on CF and it was good..some people at school became quite protective"

R14(M) "It's something I used to hide (CF) at primary and secondary school for fear of not being accepted"

R8 (F) "When I was young I was paranoid about people knowing (about CF.) ,at school I was very fit and active, I only started telling people when I became ill"

R21(M) "It was none of their business so I wouldn't tell anyone...I was that paranoid I learnt to swallow my tablets in a oner"

Disclosure and intimate relationships

Four respondents had had negative responses from prospective partners regarding their CF. Respondents who were married had often employed indirect means of disclosure about their CF or had known their partner for a long time.

R9(F) “Now, I wouldn’t tell anyone unless I was dead sure of them, and if I did tell someone too soon and it did break-up, I’d assume that it would be because of the CF...people have been quite accepting of it”

R25(M) “I’ve had a couple of girlfriends react like they don’t want anything to do with this (CF) so I just go out and get another”

R17(F) “I think if you’re going to be serious with them you have to tell them right away..before I went out with S we were friends anyway and he knew I had CF”

R8(F) “I wrote a letter to my husband before we got married to explain about the CF”

R9(F) “I find relationships very difficult when you start them ... you’ve got to choose a time”(to tell your partner about CF)

Employment

Of the respondents who were not working due to ill health , the majority wished they were able to and described the effects of unemployment.

R17(F) “I get quite emotional about not being able to go out to work, well I could, but there’s all the colds and chest infections”

R13(M) “If I had to give up working full-time I would start worrying, I’d miss the salary and activity”

R25(M) “It does get me down (not working) I knew there’d be a stage I’d have to give up working...I’m an outdoor person, I couldn’t be stuck in an office all day”

R25(M) “Quite a lot of CF folk don’t have careers or jobs because they’ve been told that” (i.e. they won’t live long)

R16(M) “I get depressed like anyone else, the worst thing about not working is the lack of company..if you’re stuck at home all day you get lonely”

R9(F) “I find not fitting- in hard because you’re not working”

Employers reactions

Five respondents had experienced negative reactions from prospective employers who they told about C.F. Those who were working had found employers to be quite flexible if they needed time off for hospital appointments:

R16(M) "I probably could have kept working in the office , but they wanted to get rid of bodies"

R19(F) "I applied to several places, and in one job I went for an interview and a s soon as |I started coughing they asked if I smoked and I said "no" and as soon as I said I had CF they didn't want to know"

R19(F) " I told them (present employers) right from the start, if you're honest then, there can't be any comeback on you"

R 23(F) "I wouldn't tell them at work I had it, otherwise I wouldn't have got my job, so I didn't let on"

R28 (M) "It's better to tell them (employers) right away, the company I used to work for were great about it, time off for clinics and everything!"

Relationships

Two respondents believed that having C.F. had placed a strain on their marriages which had ended, mainly relating to disagreement about whether to try to have children.

R21(M) "To start with I thought of trying to meet a lassie with CF, the same gene as me, it would be easier because the two of yous have to do this, take that..."

R13(M) "My ex wife wouldn't speak about it (CF) tried to deny it , her personality just wasn't strong enough to deal with CF"

R22(F) (re. husband, now separated) "He always wanted children right from the start, I decided to be sterilized but R (husband) became very grumpy on me"

R14(M) "I'm single , and I always have at the back of my mind "CF " and this is probably the major reason why I haven't married...plus my self confidence is zero and I think" "why would she want to talk to me".

R19(F) "I also stuck with him (ex boyfriend) because my CF was starting to get worse and I thought "no-one else is going to go out with me when it's this bad"

R19(F) "Now I don't know if I'd want to get into another relationship because if something happened (and she became very ill) I d feel guilty for hurting them"

R31(F) "When I was 16 I met my husband, and that changed my life...the relationship between husband and wife plays a great part"

R5(F) "He (husband) can run a house, I could go into hospital tomorrow and I wouldn't have to worry about a thing"

R16(M) "We've never really talked about it (with wife) she keeps it to herself, she's quite difficult to talk to really"

Death and worries about the future

All respondents were willing to talk about death, four of them had made practical provision in the event of their death.

R19(F) "My dad's going to help me to do my will, practical things like that...it helps to have someone...I like to be in control of things, I've planned my funeral and everything, I want everyone to be in bright colours, nothing morbid"

R18(F) "If I start talking about it (death) I start thinking about it, I get goose pimples, I can't walk past a graveyard without getting the shivers...I couldn't talk about it openly"

Six respondents made a statement to the effect of the following:

R15(F) "Obviously it does bother me that you could die , but you could get run over tomorrow"

Five respondents expressed a similar concern to that below:

R19(F) "I'm not worried about dying, how I die worries me, being in pain, I don't want to feel it"

Talking about death

Most respondents said they found it difficult to discuss death with their family or partner:

R31(F) "N (husband) doesn't like me speaking about death, he says we'll cross that bridge when we come to it"

R15(F) "I did talk to the doctors 'cos I'd hate to think if they knew you were going to die and they didn't tell you"

R12(M) "If I was going to die in a weeks time I'd want to know so I could have a bloody good party"

R5(F) "People are so taboo about it (death) but some people just go sooner than others"

Bereavement, the effects of sibling and friend's deaths from CF

Four respondents had experienced the death of a sibling from C.F. when they were old enough to recall the impact. Four others had had older siblings who had died in infancy.

R31(F) "My sister died 6 years ago (of CF) that's had a big impact on my life...will I get to 16? I was 12 when she died"

R5(F) "Just after my brother died I thought I would take worse, but then you always think "nothing will happen to me"

R19(F) "I've lost a lot of friends (with CF) into double figures, you think "why did they go, they weren't even ill, and you can't ask "who's next?" when you're the only one left"

Transplant

Only three of the nineteen respondents said they would not consider having a transplant. Two respondents were on the waiting list for transplant and one had received a transplant.

R18(F) "I've always let them know that if I ever need a transplant then not to bother, I couldn't go about with someone else's heart and lungs"

R19(F) "I'd rather die in the process of trying (to get a transplant)."

R12(M) "No I wouldn't have a transplant, your only guaranteed about 2 years, and I'd have to put my family through it....I believe when your bodies finished it's finished"

R25(M) "I've been down (South for a transplant) once, it was a false alarm, they checked the lungs and found they were no good, it was a bit of a downer...a relief too"

Fertility

Two male respondents had had children by donor sperm. One other male was in the process of attempting an IVF program.

R28 (M) "My mum told me, at first I was thinking about it a lot, but then it didn't really bother me, you can adopt...should have someone at the clinic to tell you more details about it (fertility)"

R21(M) "Other lassies in the past have said they were pregnant by me, but it was only when I came to the clinic here (in Edinburgh) that they told me there was a 92% chance I was infertile"

R13(M) "6-7 years ago it was pointed out to me at the clinic....should have been brought up long before, but I never asked and no-one ever said"

R11(M) "Having a family I had problems with that, (had a baby by donor) at first it's a bit upsetting, but once it's done it's O.K."

Pregnancy

Four female respondents had had children. Two others admitted feeling upset about not being able to (due to their poor lung function), and one respondent was in the process of trying to conceive.

R26(F) "I was advised against having a child, the junior doctor was appalled when I went into labour" (daughter now aged 19)

R22(F) "I never considered having children, initially because I didn't like them, later I came into contact with women (with CF) who'd had children and it had been detrimental to their health to say the least, three of them are now dead"

Most helpful people

All respondents cited a mixture of helpful individuals. Predominantly family, friends, partners and health professionals at the Edinburgh clinic.

R19(F) "I'm much more confident, I have such a good network of friends, it depends who's around you"

Social support and compliance

Ten respondents commented on the importance of practical and emotional support in helping to motivate them with treatment:

R28 (M) "My dad does them (Intravenous antibiotics) it does help a bit, I'm a bit lazy"

R13(M) "I tend to rely a lot on remembering from others , especially now with the insulin..if I lived on my own I'd be a wee bit worse"

R19(F) "You get bored of the treatment, but there's so many people there to motivate you and say" just do it!"

R12(M) "My wife's carried me through it..I've said that on more than one occasion, you need someone to push you"

Treatment: Motivation and Compliance

The majority of respondents considered that a high level of motivation was required in order to comply with treatments. The treatment in which adherence and motivation appeared to be the lowest was physiotherapy .

R5(F) "Having cancer, you can sit back and let it happen, but with CF you have to work hard"

R15(F) "I don't do physio., I've started trying to do it, and I know I should stop smoking"

R9(F) "Your motivation's not always there and it is bloody hard!"

R5(F) (Transplant patient) "If I went into rejection I would worry...I don't think I'd fight as hard if it happened again you get weary"

R13(M) "You tell clinic things to keep them off your back, but they generally know how fit you are...If I feel worse I do more physio.."

Best and Worst treatments

Intravenous antibiotics (IVs) were listed as some of the "worst" treatments by four respondents who simultaneously acknowledged their benefits. DNase was believed to be beneficial by two patients, and port-o-caths were also considered to be very helpful in increasing quality of life.

R9(F) "I find the IV course stressful, and I don't think people appreciate that it is quite so exhausting..but it makes you feel better, you've got no choice or you'd be dead otherwise"

R25(M) "The DNA drug (DNase) if I hadn't been put on that I'd be dead by now"

R12(M) "I say to a lot of folk, I wish I had cancer instead of this, they can cure cancer and give you radiotherapy"

R11(M) "IV's have been really helpful"

R16(M) "My inner ear's been damaged by all the drugs I've been taking, and that's the worst thing that's happened to me, but if it wasn't for the drugs I wouldn't be here"

R21(M) "Physio., I hate doing it....my mates come round and they ask you to do something and you say "no I can't" I have to do my physio., take tablets and have a meal....by the time you've done all that 3 hours could have gone by"

R 13 "Diabetes has been the greatest problem at work last 6 months"

R16(M) "Since I've had a port-o-cath, it's helped a lot, I can treat myself at home"

R12(M) "The treatments are a bloody nuisance, I just want to get on with my life,...my life's not my own I'm in the hands of everyone else.."

Link between psychological and physical factors

Five respondents asserted that they believed that psychological factors played an important part in their health.

R26(F) "The only reason I'm alive today is because I'm bloody-minded and stubborn"

R17(F) "You just feel down and you're not doing anything, and if you're not active then your secretions get thicker and then you can't move them, then you have to do more physio..."

R31(F) "Happiness plays a great part in keeping yourself well, apart from the physio. and the tablets"

R9(F) "Over the years I've met people with CF that had it quite mildly, they don't survive, and one of the biggest things is they don't have the power of their minds, they mentally give up, it makes me 100 times more determined"

Self perception

All 19 respondents considered themselves to be less severe in relation to other people with CF.

R23(F) "I see one or two (other patients) at the clinic and I thank my stars it's not me"

R5(F) "I really am one of the lucky ones, if I could have changed things and had a longer life and not had my daughter, I wouldn't change anything"

R25(M) "If I get a new set of lungs I'll be a healthy person, everything else works perfectly"

R15(F) "I may be thinking these (worrying) thoughts about CF and someone else will be thinking thoughts about something else so you don't know whether you're actually any different from "normal" people"

R12(M) "It rules my life (CF) it's a bloody nuisance (coming to hospital a lot because recently diagnosed)"

R12(M) "I think I'm lucky because I've had a lot more time than my sister"

Worst aspects of CF and what it has prevented people from doing

Five respondents felt that CF had not prevented them from doing anything. The remainder had mixed views about it, three respondents believed it made it difficult for them to make future plans:

R23(F) "It doesn't stop me doing anything, infact it makes me enjoy my life better every minute 'cos you never know.."

R16(M) "It does restrict me a bit, holidays, you have to plan everything, you can't just get up and do it"

R22(F) "I used to think"oh I've got CF I couldn't possibly live on my own ", I've always used it as an excuse for things I've not done...but there's no reason for my CF to be a hindrance"

R5(F) "I couldn't clean my own house when I was in a wheelchair, not being in control was hard

R16(M) "It's restricted my sport, I've had to pack in my golf"

R13(M) "I've been all over the world, it's never stopped me"

Avoidance of activities / environments

Four respondents reported avoiding people or places because of “bugs” :

R21(M) “I try to block these thoughts out but there could be germs in this room that could kill me”

R8(F) “It infuriates me that other people smoke in public, I won’t go into pubs”

R9(F) “I’m at the stage, I’d like to take a gun to anyone with colds or flu, I do find that difficult...I don’t like going to cinemas in the winter”

Involvement with other CF individuals

Four respondents had one or two particular friends with CF, the remainder said that they did not have any, for various reasons:

R16(M) “We’re ships that pass in the night (other people with CF) we have to keep apart anyway because of infection”

R9(F) “I’ve never been in to mixing with lots of CF people, and it’s paid off because now that bug (cepacia) is around”

R14(M) “I wouldn’t go to the extent of being friends with them (other CF patients) I’ve got the disease and that’s enough for me”

Smoking behaviour

Three respondents currently smoked and two had done in the past. Four of these had been diagnosed late:

R21(M) “I do a lot of things I shouldn’t do which don’t help like smoking..but you’ve got one chance only, I live from day to day so why give-up?”

CF in the media and public awareness

The majority of respondents felt that the public were generally not well aware of CF, but that there had been more recent media coverage. Respondents generally thought it was not that important for there to be greater public awareness:

R 23 “People that do know about CF only know the really bad bits I think they are becoming more aware”

R26(F) “ I don’t want to be associated with C.F. “sufferers” that image is demoralizing”

R9(F) “I don’t like when it’s (programs about CF) on TV they bring out the worst cases”

Life insurance, mortgages and transport

Six respondents with CF had experienced difficulty obtaining a mortgage or life insurance. Only one female respondent cited this as a concern. Two respondents had lied about their health , and three others had managed to obtain insurance or a mortgage before they were diagnosed. Satisfaction with benefits and transport was high and clinic staff had helped some respondents to claim these.

R23(F) “but I can’t get insurance, not even my name on the house...I just think what a cheek they think I’m not going to live very long”

R19(F) “The mobility car makes such a difference”

R12(M) “I always look ahead, we’re well insured, I had them all before I was diagnosed”

R21(M) “I can’t buy houses and go abroad, all my mates do that.. I canne get life insurance and I start thinking about these things”

Satisfaction with medical care

There was generally a high level of satisfaction with medical care, particularly with the clinic in Edinburgh. Three respondents felt there should be someone outside of the clinical team to deal with emotional difficulties. Four respondents commented that they have known more than their GP or a doctor in their local hospital about CF:

R16(M) “All the medical staff are good, but here (Edinburgh clinic) they’re right on top of the situation”

R9(F) “I think you need more support emotionally, sometimes you can talk to the nurses but you can’t talk to family”

R21(M) "If I speak to a Doctor and I don't like his tone, if he speaks to me like a bairn then I'll walk out the door...I'd rather someone advised me"

R17(F) "The CF nurses are brilliant you can just pick-up a phone and you know they're there"

R18(F) "I was able to tell the doctor (a GP) more than she could tell me (about CF) I couldn't believe a doctor could be so ignorant"

R19(F) "I've often thought there should be someone to help with work, who can help us to get in there, somebody outwith the clinic"

Information

Generally most respondents (except three) said they preferred to be told information about interventions being carried out. They thought that the clinic staff were sensitive to this and did not conceal information about their physical state.

R25(M) "They (the clinic staff) wouldn't keep anything from you, healthwise I like to know why I take things"

R8(F) "I hate coming to the clinic because I never know what they're going to tell me"

R19(F) "I like to know everything, it's my body and my illness, I like to know if I'm going up or down"

R14(M) "I'd really rather not know the results, unless they have to do this or that"

Family responses and independence

Six respondents reported that they had one parent who was "overprotective". Two had been forced to move home after they had become ill again and had found this difficult.

R28 (M) "I'd rather live with someone than live on my own, my mum and dad have been brilliant, and my sisters"

R31(F) "When I started driving (mobility car), that gave me a lot of confidence, not having mum and dad drive me around"

R18(F) "Leave me alone, I want a life of my own, my dad's still like that (i.e. overprotective) my brothers aren't anymore"

R12(M) "my parents divorced just after my sister died (of CF) I think it split them up"

R14(M) "I've had to move back to live with my parents, because I had no where else to go really...my mum wants to molly-coddle me, and was delighted when I moved back...which is exactly what I don't want...but she's coped well over the years"

R12(M) "Dad didn't say much, he's one of the old brigade like, he was upset but didn't talk about it (sister's death) none of us do, and I suppose I'm in the same category"

R19(F) "We're all close, some families with CF it can make them or break them, in our family it brought us a lot closer"

Coping Strategies

The following quotations illustrate the range of coping strategies employed:

R21(M) "I wasn't a person to talk about it, because if you talk about it then you must be thinking about it and then you start worrying"

R21(M) "There's a lot of things in my head, my brother's a heroine addict and the fact that he's got perfectly good health and throws it down the toilet really winds me up"

R25(M) "Being here (hospital) for 8 weeks isn't a joy but it isn't a prison sentence either"

R15(F) "I don't ever think" oh I've got CF "...and maybe that's why I don't do what they say (breathing exercises)"

R19(F) "I'm O.K. about it now (sister's death) I find talking about it helped me"

R12(M) "I suppose I bury my head in the sand, but it's not going to go away"

R25(M) "Some people just sit in and think about their health all the time, I just thought about being happy and went out"

Adjustment process

Three respondents referred to a period of rebellion usually in their teens which was associated with a lack of adherence to treatment: Three older respondents talked

about a period of serious illness from which they recovered and lead to a greater acceptance of their CF:

R13(M) "I've never felt "Why me?" I've always looked on the bright side"

R12(M) "Why me?.. I wouldn't impose it on anyone else like, I've got to live with it"

R13(M) "I just decided one day oh sod it! I'm going to enjoy this (his life)"

R9(F) "I did go through that rebellious stage in my teens, there were heaps of tablets I never took"

Attitudes to screening / medical procedures

Very few respondents voiced particular opinions about screening except for one.

Three other females with children said they would not have had a child if their partner had been a CF carrier too.

R22(F) "If there'd been a test I would have hoped she'd (mother) have had a termination, as it is I'm in my thirties and have a good QoL, but if I hadn't ever been here, no-one would have missed me"

Religion / spiritual beliefs

Only one of the respondents reported any religious or spiritual beliefs.

Being a parent.

Two of the mothers with CF were concerned that their children were worried about them and insecure. They were uncertain how much to explain to their children about CF and the future, they also had practical worries:

R23(F) "I've often thought about what would happen if anything happened to me with them being so young (the children) because my husband wouldn't be able to cope"

R17(F) "How do I explain to my son that I'm going to die, do I tell him or do I not? It's hard to think about"

R5(F) "I think that she thinks (daughter) that I'll be fine forever more and I make that out to her because she's too young to have all these problems"

Discussion

1. Comparisons of the CF group with their healthy peers

The majority of between groups hypotheses were supported. Firstly as predicted, no statistically significant differences were found between the CF and control groups on measures of anxiety, depression or self-esteem.

Data from the HADS indicated that depression and anxiety scores were similar to published norms for 573 people with cancer (Moorey et al 1991). The authors reported that 27% of their sample scored above the cut-off for anxiety and 8.7% for depression compared with 20.5% and 10% respectively in the present study. These findings are in accordance to those of Gee et al (1996) who found similar levels of anxiety (25%) in the CF and control groups on the HADS and no significant differences in depression scores. This had been one of the few studies of the prevalence of anxiety and depression in CF which employed a control group. One other previous study had also found a lack of generalised psychological distress (Moise et al 1987).

It would appear that the level of generalised anxiety is common in the general and CF populations. The HADS appears to have been a reliable screening instrument for both ill and well groups given the consistency of results from the three studies above.

Previous research regarding CF and self-esteem has yielded reports of high self-esteem in the CF group relative to controls (Simmonds et al , 1985; Cowan et al , 1984). The present study similarly found self-esteem to be relatively high in both study groups (when compared with norms) with the CF group exhibiting no higher level of self-esteem than controls. Contrary to previous findings, (Sawyer et al , 1995) females were not found to have a lower self-esteem than males.

The Rosenberg self-esteem scale has not been used with a CF population before. The published norms suggest that its scores are negatively skewed towards low self-esteem (Rosenberg, 1989) in the general population, although this does not appear to be the case in the present study. There is no cut-off score for the scale, however the inclusion of the body image questionnaire offers some means of comparison since the total scale score and each of its four component scores were found to be highly correlated with self-esteem scores in the CF group. This would suggest that the two scales were measuring related aspects of overall self-concept.

The second hypothesis that body image concerns would be more disease specific for the CF group was also borne out. Although there was no statistically significant differences between the two groups on overall body image score, the CF group did wish to make significantly more changes to their appearance. One third of the CF group commented that they wished to gain weight compared with 50 % of the control group who stated they wished to lose weight which is in line with previous findings related to adolescent girls with CF (Jelalian et al , 1996). There was a sub-group of three CF subjects who desired to lose weight all of whom were pancreatic sufficient and therefore described themselves as “looking normal”.

Apart from low weight and short stature the types of CF-specific body-image concerns which were reported in the present study had little in common with those described in previous studies (Coffman, 1984; Strauss and Wellisch , 1981). This illustrates the diversity of these concerns in the CF population.

In terms of quality of life,(QoL) it was predicted that the CF group would have greater dependency on medication than the control group which was supported as was the fact that that CF subjects have significantly less satisfaction with their employment capacity. Satisfaction with employment capacity was found to be significantly related to actual working status in the CF group. There is no previous research to draw on which examines CF individuals' subjective satisfaction with these areas of QoL. Comparison with published norms for the WHOQOL from a sample of

224 subjects with a variety of chronic illnesses in Bath (WHOQOL Group 1995) indicates that there was no significant differences with the CF group on the facet, overall QoL (Mean=13.58; SD=3.55).

Surprisingly, there were no between groups differences in mobility as predicted. Comparison with the Bath norms for a well population (N=157; Mean mobility=18.40; SD= 2.03) suggest that the present Scottish control group differ considerably in terms of their satisfaction with mobility. This may explain why there were no between groups differences found in the present study. When considering other related areas to mobility such as ability to perform daily activities, (Bath norms for ill group; Mean= 14.36; SD=3.99) , it appears that the CF group are not significantly more disabled in these areas than others with a chronic illness or the general population.

The CF group were found to have spent an average of 2 years less in education from age 11 than control subjects as predicted in the fourth between groups hypothesis. They were also significantly more likely to be unemployed due to ill-health. The early finding of Smith (1983) that there were no differences in terms of employment and education between CF adolescents and their peers was therefore not replicated. Walters et al (1993) reported that CF subjects were less successful than their peers at gaining 'O'levels, but more successful at achieving 'A'levels. Although the present study did not consider qualifications, it is unlikely that the point at which qualifications are gained would affect the overall time spent in education given the wide range of subject's ages in the study.

The bulk of controlled studies suggests that people with CF are less likely to be in employment than their peers (Shepherd et al ,1990; Blair et al ,1994). Walters et al (1993) found 54% of their responders in full-time employment which is comparable to the present finding of 43.3%. 40% of CF responders who were not working gave ill-health as the reason in the current study compared with around 23% in the Walters et al study. One explanation for this could be that the present study sample is smaller

and subjects may have more severe health problems at a specialist CF clinic than previously investigated large populations such as that surveyed by Walters et al (1993).

1.1 Other findings between the two groups

Other demographic findings between the two groups indicated that there was no higher rate of non-manual occupations amongst employed CF subjects contrary to previous findings by Walters et al (1993). The rate of manual occupations was low in both groups.

A U.S. study by Shepherd et al (1990) found that those CF respondents who were not married or cohabiting were more likely to live alone or with a parent whereas the control group were more likely to live with flatmates or friends. This finding was exactly replicated in the present study as was the finding that there were no significant between groups differences in terms of relationship status. Previous large sample studies (Penketh, 1987; Walters et al, 1993) had reported low rates of marriage in this group and high rates of respondents living with their parents, 52% in the latter study compared with 20% in the current study. These studies had not included a control group.

The fact that there were 10% of CF subjects who were divorced or separated and none in the control group may have reflected some strain within marriage specific to CF. This was partially supported by two divorced respondents who were interviewed, where the divorce related in both cases to disagreements over whether to have children . It is not possible to generalise from these findings, but this is an area which may merit further investigation.

Coffman et al (1984) in a large survey of sexual adaptation in CF reported on poorer sexual adaptation in single females with CF. Although the present study assessed sexual satisfaction rather than adaptation, no gender differences or between-groups

differences were present. However relationship status in both the CF and control groups was significantly related to sexual satisfaction. The data suggested that being single was related to less satisfaction and this was a stronger trend in the CF group. This was also found by Shepherd et al (1990).

Psychological factors of low self-esteem and poorer body image were also associated with lower sexual satisfaction in the CF group as were lung function, Shwacman score, dependence on medication and satisfaction with work capacity. In the control group, sexual satisfaction was only related to overall QoL and positive feelings. This suggests that reasons for dissatisfaction with sexual functioning are likely to be more complex and multifactorial in the CF group, and the additional link with physical severity may place an added strain on single CF respondents. This could possibly dissuade them from embarking on a relationship and heighten the discrepancy between desired intimacy and their actual single status. This is discussed further in relation to qualitative data (see 2.2).

In terms of other aspects of QoL, the CF group scored as being significantly less satisfied with their sleep than the control group. This is a finding that had been reported previously by Congleton et al (1996) using the Nottingham Health Profile in one of the few quality of life assessments of CF patients. This only held true for men whereas there were no gender differences in any QoL facets in the present study. Reduced sleep satisfaction was found to be correlated with low self-esteem, anxiety, depression and less acceptance of illness in the CF group but not in the control group. Severity ratings were not related to sleep satisfaction in the CF group either, which suggests that sleep difficulties may be an indirect expression of psychological distress.

The CF group scored as receiving significantly more actual emotional support than their healthy peers. This is consistent with reports of people who experience a life event or crisis (Sarason et al, 1987) and an illness could be conceived as being a chronic stressful event, for which individuals are able to mobilise greater emotional support. The mean scores for all subscales of the Significant others scale were most

similar to people with Parkinson's disease (McCarthy, unpublished data) . Perceived support in the CF group was therefore greater than for other groups according to published norms (e.g. depressed patients, Power et al, 1988).

The control group did not differ from the CF group on any of the other subscales of social support. A gender difference was found which revealed that females received more practical support than males in the CF and control groups. They also had higher levels of ideal practical support than males but only in the control group. This seems to rule out the possibility that females with CF receive special treatment compared to their male counterparts. It is more likely that this finding reflects cultural factors and social roles for men and women, or reporting bias. It has been noted by (Sarason et al 1987) that females generally endorsed more social support items on a questionnaire.

Due to the constraints of time, the present study did not include a detailed analysis of social networks and density. The interview data with some CF individuals did provide a guide to which were the important supportive situations and significant others (see 2.2).

To sum up, the lack of between groups findings relating to living situations and relationships generally suggest that CF subjects in the present study were achieving a high degree of autonomy and intimacy, equal to that of their peers. This is in line with recent controlled studies as is the fact that the CF group do not differ on measures of psychological distress. As predicted, the CF group have more CF specific body image concerns, less years spent in education and greater unemployment through ill-health than controls. Hypotheses related to differences in QoL were also upheld except for differences in mobility which were not present. Unforeseen findings revealed that the CF group experienced less satisfaction with sleep, received more emotional support and were slightly more likely to be dissatisfied with their sex lives if they were single than the control group. These findings had also been documented in the previous literature.

2. Findings within the CF group

Acceptance of illness in the CF group was high with a mean score higher than published norms for a group with chronic illnesses (Felton et al , 1984). While this could suggest particularly good adjustment in the CF group, it is possible to question the sensitivity of this measure to all aspects of individual adjustment since it is not a multidimensional measure, and therefore it may be important to include other validating data, for example from interviews. Nevertheless, the range of scores on the questionnaire in the present study were considered sufficient to indicate patterns in the data and relationships with other variables.

The first hypothesis within the CF group was supported. It had been predicted that there would be a positive association between acceptance of illness, overall QoL; positive feelings and employment status. Satisfaction with working capacity was also found to be strongly correlated with acceptance of illness, but did not explain enough variance to achieve individual statistical significance in a regression model with acceptance of illness. This suggests that there is no strong causal relationship between individual adjustment to having CF and working capacity.

From the bivariate correlations between variables, it would appear that acceptance of illness and overall QoL and positive feelings may be measuring similar constructs. This was not found to be entirely the case since acceptance of illness made no significant contribution to explaining the variance in overall QoL when regression analysis was carried out. This implies that acceptance of illness and Overall QoL scores reflect important separate dimensions in the CF population under investigation.

The second hypothesis had predicted a relationship between deprivation category and severity of illness in line with findings by Britton (1989) that lower socio-economic status is related to increased mortality in CF. This was not borne out in the present study, since no relationship was found between physical measures of severity and deprivation category. Obviously it is necessary to differentiate between severity and

mortality, but it is usually the case in CF that lung function (FEV1% predicted) declines before death so it could be assumed to be an appropriate measure for the present study . However the relatively small sample size and the fact that the subjects were drawn from a specialist clinic which did not include patients from the two lowest deprivation categories could have influenced these findings. Recent work on health inequalities (Wilkinson, 1996) suggests that mortality rates from all forms of disease increase as a function of income share and social class.

Interestingly lower deprivation category (higher socio-economic status) was associated with a later age of diagnosis. This could suggest that infants born into more well-off households are healthier for longer and therefore CF is not detected as early. It is also possible that this is related to demographic factors such as family size, with a greater number of older siblings affected with CF in lower socio-economic groups increasing early detection in these families. Certainly this would have to be examined in a larger cohort of patients in order to draw any definite conclusions.

No link was found between severity of illness (as measured by FEV1 % predicted, BMI and Shwacman rating) and anxiety, depression or self-esteem. This was in accordance with the hypothesis based on previous findings of a lack of relationship between psychological factors and severity (Sensky 1990). There was a link however between acceptance of illness and measures of severity (except Shwacman rating) . FEV1 % predicted was found to be a significant independent predictor of acceptance of illness in a hierarchical regression analysis, and a better predictor than body mass index. This suggests that severity of CF directly affects an individual's adjustment , but that severity has little direct impact on psychological distress. One possible exception to this could come from the relationship observed between FEV1 % predicted and body-image which suggests that while BMI, actual weight and shape does not affect the CF individual's perception of their physical appearance, their general severity may .

Severity was also related to overall quality of life, to positive feelings and satisfaction with sexual functioning and work capacity. Previous research by Shepherd et al, (1990) also found that those individuals who were unemployed tended to be so due to ill health. Coffman et al (1984) did not find any correlation between severity of disease and sexual functioning, but it is one which may be indirect, in that it may be moderated by relationship status as discussed above.

FEV1 % predicted was also a significant predictor of overall QoL in a hierarchical regression. Therefore, as hypothesized there was a clear link established between QoL and a measure of physical severity. A tentative association between lung function and time spent on home treatment had previously been noted by Congleton et al 1996, and Dodd et al (1996) had found a link between lung function and general life satisfaction on an unvalidated global rating scale. The evidence from the present study therefore lends weight to a growing body of literature in this area.

Since there has been considerable previous research relating social support within the family to compliance in young people with CF (Pownceby 1996; Patterson, 1985) it was hypothesized that this would continue into adulthood. In the present study it was proposed that practical and emotional support from family and significant others may moderate the severity of illness and help to increase adjustment. This hypothesis was not supported by the quantitative data in the study since no relationships were found between social support subscale scores and any other variables.

If the qualitative interview data was examined, around 50% of the respondents reported that a significant other, usually a partner, gave them help with their home treatments. This took the form of reminding them, "nagging", motivating them and actually performing physiotherapy or intravenous regimens. These respondents were all adamant that this type of support was indispensable to them. It would appear that the Significant Others Scale was not sensitive to these CF-specific types of resources which were qualitatively different to either practical or emotional support. It is therefore important to consider the specific nature and function of this support in

future research as it seems likely that CF adults are transferring the role of their parents and families of origin to their partners. It is possible that this could play an important part in compliance and therefore in severity and adjustment.

The final hypothesis stated that there would be no gender differences found on any of the measures. This was partially supported since the only gender differences found were that females received greater practical support, but this finding was not specific to females with CF.

2.1 Other findings within the CF group

All psychological variables were found to be correlated negatively with certain QoL facets; these were: positive feelings, overall QoL, satisfaction with services and opportunities for leisure. Satisfaction with these was seen to decrease as anxiety and depression increased and was also associated with acceptance of illness, self-esteem and body-image. While this emphasises the importance of psychological well-being, it was found that these factors differed in their significance to QoL. In a regression analysis, depression but not acceptance of illness was a significant predictor of overall QoL. Other QoL facets when combined were also significant predictors.

Age emerged as one of the most important demographic factors since it was not only associated with increased severity of CF, but increased distress on all psychological measures and reduced QoL on the four facets discussed above.

Age was also the single strongest predictor of adjustment to illness, explaining 26% of the overall variance. Similarly it explained 32% of the overall variance in general QoL. This finding indicates that age has a strong effect which is independent of the increasing severity of CF with age. This is a finding which has been noted in comparisons of psychological distress between children, adolescents and adults with CF. These studies already suggested that emotional disorders appeared to increase

with age in CF (Cowan, 1984; Simmons et al , 1987) . In comparison, QoL in a healthy elderly population was shown to be better than that of a younger group (Brown et al , 1994) .In the present study ageing was only related to an increase in depression, less positive feelings and less satisfaction with leisure opportunities in the control group. These correlations were also weaker than in the CF group. Ageing in the CF group was therefore more strongly associated with a greater number of negative aspects affecting QoL than in the control group.

Although lung function (FEV1 % predicted) is associated with this age related decline and a decrease in acceptance of illness, the independent effect of age suggests that it may be partly the individual's appraisal of their life situation which is important. It is possible that people with CF may have regrets about unfulfilled ambitions, possibly related to their reduced working capacity or relationship status if they are single. This could be considered to relate to Erikson's life stage model of adjustment (Erikson, 1963) and the stage of generativity Vs stagnation (in work and reproductive matters) in later adulthood. Perhaps they feel less optimistic about the future as they become older and weary with constant battling against their worsening condition. The interview data provides some further insight into these possible explanations.

2.2 Interview data: Further findings specific to CF

The impact of diagnosis

Previously in the literature, it has been proposed that individuals diagnosed later may initiate a search for information (Wideman 1996) This was certainly reported by the four respondents who were diagnosed late. For them there were also a mixture of relief and shock which was expressed at knowing finally what was wrong with them. The idea that there may have been some advantages to not being diagnosed early was also expressed by two of these respondents, that they may have had a better quality of life not knowing they had CF. This was balanced with a concern for their health and lifestyle which may have been different if they had known they had CF; in this latter

regard, factors such as smoking and infertility were mentioned. Although the quantitative data did not point to a link between acceptance of illness and age at diagnosis it is likely that those diagnosed later may still require considerable support and information at this time

Disclosure and relationships

Around one third of respondents recalled being told, or knowing their parents were told they had a short life-expectancy which they have since outlived. The effects of this may be as one of the respondents said, not bothering to have a career, or embarking on a relationship. There was some support for this argument from the data about relationships. The idea of not wishing to hurt a partner by being ill or dying was prevalent, as was the notion of warning partners about CF before becoming involved with them.

Disclosure about having CF was an area which was also closely linked to that of intimate relationships. Several respondents had commented that they found indirect means of disclosing CF to a serious partner, for example via a third party, or giving them a book. Others had always believed it was best to tell a partner from the start of a relationship, and some concealed it from partners with whom they were not seriously involved. Several respondents had experienced negative reactions from prospective partners and had differed in their strategies for coping with this. Some had clearly been effected by others' negative responses and this had a detrimental effect on their confidence and subsequent relationship status. Others had merely shrugged off any discrimination by others. It therefore seems likely that coping style and personality factors have a moderating influence on whether or not distress is experienced.

This suggests that the issue of forming and maintaining relationships is a difficult one for the CF population. While they may not be significantly different to their healthy

peers in terms of actual relationship status, they seem to differ qualitatively. Indeed the finding that single CF patients are less satisfied with their sex lives than single control subjects could reflect the sub-group of CF individuals who are not involved in a relationship but would like to be, and are prevented from doing so because of factors specific to CF. The importance of love and intimacy was stressed by more than one respondent in line with the finding by Sarason et al (1987) of the protective effects of this type of relationship.

Partners and spouses were also mentioned frequently as being among the most “helpful” people to CF subjects both in terms of emotional and practical support. Parents were also cited as being helpful, although in some cases (around a quarter of respondents) they were thought to be overprotective. This is something which has previously been reported (Blair et al, 1995); however, further comparison with a control group would be necessary in order to assess the significance of this finding.

A fear of “not being accepted” certainly prevented some individuals from disclosing their CF when they were younger. This tendency wanes as people age which may reflect the process of adjustment described by Kreuger (1984) and Erikson (1963), to pass through a phase of denial in order to achieve a task of adolescence by developing an identity which is consistent with that of one’s peers. Certainly a sub-group of CF respondents described a phase of denial of their illness associated with rebellion, lack of compliance and low rates of disclosure . Some respondents described presenting a talk to their peers at school about CF which not only helped obtain positive feedback, but allayed their own fears about not being accepted .

Discrimination and stigma

In terms of discrimination from wider society, about one quarter of respondents had experienced this from prospective employers and had either failed to get jobs or had concealed their CF once they were in employment. Generally these respondents had all gone on to obtain employment and some employers were thought to be extremely

supportive and flexible . Such positive attributes of employers were not considered in the previous literature (Walters et al 1993). This author also reported on discrimination by prospective employers against adults with mild to moderate CF.

Six of the respondents had felt discriminated against with regard to obtaining a mortgage or life insurance. Some of these individuals felt strongly that this reinforced their feeling of “being different” from their peers. If this is taken with employers reactions and at a micro-level individual’s reactions, it could be argued that discrimination is experienced by a number of people with CF in one form or another. This reflects the finding of Marteau et al (1996) that health professionals viewed the QoL of individuals to be poor, so it is hardly surprising that non-medical institutions and even the media will share this view.

The quantitative QoL data indicated that those facets which might reflect the degree to which an individual is disabled, such as satisfaction with mobility and activities of daily living, demonstrated that CF individuals experienced no difference in these areas compared to their healthy peers. It could be argued that the CF group are therefore not perceiving themselves as disabled so society should not either. From comments made about discrimination by individuals, it seems possible that some CF subjects internalised stigma with deleterious consequences, and others did not and this largely depended on coping style.

The recent segregation of CF patients with a resistant form of infection called Cepacia (Smith et al ,1992) could lead to a sense of stigmatisation. Most of the respondents understood the need for segregation. Fear of catching Cepacia was high for three respondents who also tended to avoid environments where they might come into contact with other viruses or infections. Only four respondents reported presently having other friends with CF. Reasons for not having other friends with CF , when given, included the desire to avoid infection. There was also the sense of not wishing to choose friends simply on the basis of them having CF, and for the majority of respondents, they did not seem to want CF to become their entire identity.

Employment

Satisfaction with working capacity decreased with poorer lung function. Employment was an area which was particularly important to respondents. From comments which were made, the consequences of not working through ill health had ramifications both in terms of social contact and psychological well-being . This was also partly supported by the quantitative data since dissatisfaction with work capacity was linked to higher rates of depression and lower self-esteem.

Dying and bereavement

All respondents were able to discuss death and fears about the future during the interview. Several coping strategies were evident and appeared to reflect differing degrees of acceptance. There were only three respondents who said they tried not to think or talk about death and used denial as a strategy. There was a subgroup who admitted they were bothered about death but had what could be described as a minimization strategy; “ that you could be run over by a bus tomorrow”. Another cluster of respondents voiced worries about the manner of their death; although as a group they appeared well adjusted to the idea of their mortality they are perhaps a group for whom reassurance, and information about medical procedures and pain control could be helpful. The last sub-group appeared to be totally adjusted to the idea of dying and had made practical plans for this eventuality. This group consisted of two respondents awaiting transplant and one who had already received a transplant.

The relationship between type of coping strategy used, severity of CF and acceptance of dying was quite striking. It appeared that the severity of illness had partly dictated adjustment to dying, something which is contrary to the quantitative data which suggests that increased severity of CF is associated with a poorer acceptance of illness. If adjustment to illness and to dying are part of the same process, then it is

possible that this inconsistency reflects a reporting bias in that willingness to talk about fears about the future is the same as willingness to admit to a lower level of acceptance of illness. Those subjects employing denial or minimization strategies may over-report their acceptance of illness as part of their overall strategy. This would be consistent with the fact that acceptance of illness was higher in the CF group than published norms of other groups with chronic illnesses (Felton et al, 1984) and could reflect a more prominent use of denial and minimization strategies in the CF population than previously noted (Strauss and Wellisch, 1981).

In keeping with previous findings (Kalish and Reynolds, 1976) the majority of CF patients said they would wish to know if they were dying or not. They also described the importance of having someone outside of their family to discuss these issues with or to help them to make practical plans for the future.

The impact of sibling's and friend's deaths from CF was not explicitly one of survivor guilt as noted previously by (Fanos and Nickerson, 1991). It seemed to engender a fear or realisation of their own prognosis in surviving siblings or friends. Confronting bereavement issues at an early age particularly for those with siblings who had died appeared to bring about an earlier acceptance of their own death in these individuals. This is something that could not be generalised from the small numbers in the interview group, but may require further investigation.

Attitudes to transplant

The period of awaiting transplant was one which was indeed reported to be stressful by those respondents who had experienced this, as previously reported by Madden (1995). The idea of receiving a transplant did not appeal to three of the respondents who interestingly appeared also to use a higher degree of denial regarding death generally. This could suggest that willingness to consider a transplant is related to coping style and acceptance of dying. This may also require further research .

Fertility and pregnancy

It was evident from comments made by some male respondents regarding fertility that there had in the past been no clear protocol for discussion of these issues with them, a problem which was noted previously by (Sawyer, 1996). The inability to firmly predict physical outcome in pregnancy clearly makes advising female patients a difficult task. The subjects who were interviewed reflected the diversity of outcome since one had had a transplant and two others had remained healthy since having children. Several respondents who had had personal contact with females who had become ill or died post-partum had decided themselves not to have children. Adjustment to infertility or not having children was not closely examined in the present study, but individual differences in this were apparent and would merit further investigation.

The satisfaction with medical services was high in the respondents who were interviewed, and no different to the general population from the analysis of quantitative data. Satisfaction with the Edinburgh clinic was high particularly in relation to accessibility of staff, information-giving, and confidence in the effectiveness of treatments. The first two of these factors have also been noted by Pownceby et al (1996) to be important to patients and to facilitate adherence to treatment. Several respondents stated that the specialist CF clinic was far better than services they had received elsewhere, which has been generally accepted, (Nielson and Schiotz 1982; Warwick 1982).

Treatment and compliance

Treatments were perceived by respondents to have advantages and disadvantages. Pownceby et al (1996) had described how compliance with physiotherapy may be poor in about 25% of patients. This certainly appeared to be the case in the present study although no direct measure of compliance was used. A number of respondents did comment on the time-consuming nature of physiotherapy and its lack of perceived

immediate benefit for those with milder CF which may also have influenced rates of adherence.

This weighing-up process of the relative costs and benefits of each treatment in relation to perceived quality of life seemed to take place for most respondents. They were able to simultaneously describe drawbacks and advantages of treatments. Those who used a coping strategy of denial appeared not to engage in this process to the same extent and found any treatment regimen to be “a reminder” of their CF and therefore to be an unwelcome intrusion. Conclusions about the relationship between these types of coping strategies and compliance can not be drawn from the present study, but may be important areas for future research.

Coping style

The consistent finding in the literature (Shepherd, 1990) that CF individuals rate themselves as less severe than others with CF was strongly exhibited by all respondents. This could reflect a coping strategy of minimization, but given its pervasiveness, it could be argued that it served a positive function. Interestingly comments reflecting the use of denial as a coping strategy illustrated that these respondents were aware of this, for example; “I suppose I bury my head in the sand, but it’s not going to go away”. Beyond the fact that some respondents (the minority of those interviewed) used a strategy of denial, there were no other clear categories of coping strategy which emerged. The remaining comments reflected the generally positive and philosophical outlook of most of the respondents some of whom felt that they had always been able to accept their CF, and that it was just part of their nature to do so.

Models of Adjustment

The retrospective nature of examining the adjustment process means that comments made about adjustment in the past have to be treated with caution. Apart from there being some support for the stage of denial as proposed by Kreuger (1984) there appeared to be little evidence to support the psychoanalytic bereavement model of

individual adjustment. However, Erikson's model (1963) may be useful in considering at what stage in the life-span the adjustment process began. For example a person who was diagnosed late as having CF may already have had a family, and have married and achieved tasks of adulthood and therefore the impact of diagnosis is low. However, if diagnosis takes place late for an individual who has not had children or achieved the tasks of adulthood they wished, then the impact of diagnosis could be greater. This certainly seemed to be the case for some of the later-diagnosed respondents.

For those diagnosed early, it is possible that the process of adjustment is assimilated into the life-stages. If there are no obstacles to achieving tasks through the life-span, then adjustment to CF may also be smooth without the tendency to engage in denial or expressions of anger and sadness. If there is some disruption to this process such as an exacerbation of the illness during teenage years, or a negative experience with peers, then adjustment may be affected at this stage with psychological and behavioural consequences. These models of adjustment would certainly require further investigation.

3.What are adjustment and quality of life in CF? : A summary

For the purposes of quantitative analysis in the present study, adjustment was considered to be measured by the acceptance of illness scale . This does not however reflect the complexity of adjustment as it is revealed by the qualitative data. On the basis of the regression analysis, it could be concluded that age and lung function were the most important predictors of adjustment, with psychological factors and satisfaction with work capacity playing a less significant role. This still leaves the question of defining the nature of the correlational relationship between psychological variables and acceptance of illness.

Some clues as to this link are suggested by taking an integrated view of the quantitative and self-report data. Some respondents who employed a strategy of

denial could be considered to have a poorer long-term adjustment to CF (Suls and Fletcher, 1985). They were found to have less acceptance of death, and were able to engage less in the weighing-up of the relative pros and cons of their treatments. There also appeared to be a link between degree of acceptance and ability to disclose about CF. Those who were unwilling to disclose for fear of not being accepted also appeared to experience greater concern about forming intimate relationships. The status of being single was also associated with less sexual satisfaction in the CF group. These findings together suggest that acceptance of illness or coping style may have an important moderating influence between the severity of CF and psychological distress and between other life events and stages and experienced distress. This would also explain the lack of direct association found between severity and other psychological factors.

While coping style could be considered to be a correlate of acceptance of illness, it may in future research be useful to include a direct measure of coping style. This would also allow the detection of any reporting bias which may be present as a result of coping style. For example, do older respondents with CF report less acceptance because they use less denial ?

The present study did not employ a measure of compliance which may have been useful. Contrary to the quantitative data, the interview data suggested strongly that specific aspects of social support aided compliance and could also have made a significant difference to overall QoL and adjustment. This is an area which certainly merits further investigation.

The quantitative data indicated that there was no significant differences between the CF group and their healthy peers in terms of psychological distress and most aspects of quality of life. Despite the fact that this was a multidimensional assessment, the quantitative data do not illuminate the aspects of QoL specifically affected by CF.

Within the CF group however, the regression analysis demonstrated the importance of age and physical factors as predictive of QoL, with depression and acceptance of illness contributing less to explaining the variance.

The specific types of situations relating to QoL in CF were identified from the interview data. The effects of unemployment may be much the same as in the general population therefore this was reflected in the reduced satisfaction with working capacity compared to controls. The specific experiences of discrimination at a societal level, by employers and financial institutions, and at an individual level, may well have impacted on QoL in the CF group. For those diagnosed late QoL may be perceived as better prior to the knowledge of having CF. In cases when prognosis is poorer than necessary, this could also have an impact on life decisions. Fertility issues may also require further needs assessment since this was also an area identified as being important by respondents.

The areas of bereavement, forming relationships, attitudes to treatment and factors influencing compliance (particularly social support) also have an impact on QoL and adjustment in CF. Issues of autonomy and relationships with family of origin in adulthood were not explored in detail in the present study. It is possible that moving back home following a deterioration in health may also affect QoL in older CF respondents and may require further research.

As the life expectancy of people with CF continues to increase, the findings of the present study indicate that QoL and acceptance of illness may decrease with age. Further development of the model presented in section 2.2, which integrates physical functioning and the clinical course of CF with psychosocial factors, would be particularly relevant at the present time. This may help in deciding at which point in the life-span services and psychological support could be most beneficial.

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APPENDIX

INFORMATION SHEET

Dear.....

RE: PSYCHOLOGICAL FACTORS AND QUALITY OF LIFE IN CYSTIC FIBROSIS STUDY

I am a Psychologist who is currently attached to the Western General Hospital. I am conducting research into which psychological factors are important to adults with Cystic Fibrosis. I am writing to all the clinic attenders at the Western General Hospital and would like to ask you if you would be willing to participate in this study.

This would involve filling- in questionnaires about your mood, the way you see yourself, and how much help you receive. There would also be a questionnaire about some of the things which could affect the quality of your life, such as sleep, and daily activities. There are five questionnaires in total which should not take more than 30 minutes to complete.

An interview with myself about you and your experience of Cystic Fibrosis would also be part of the research and could last about 40 minutes . I would like to tape record this interview with your consent, but this would be entirely optional. Any information given, either written or verbal would be treated in the strictest confidence and would only be available to myself as the sole researcher. Audio tapes of the interview would be destroyed at the end of the study.

The aim of this study would be to help medical staff and other researchers to understand the link between physical and psychological aspects of Cystic Fibrosis. This may , in turn influence and improve the way in which treatments and services are delivered to you.

Therefore I would appreciate your participation in this research. However, if you do not wish to take part then this would not affect any treatment you receive now or in the future.

If you agree to take part in this research then I will let your consultant at the Western General and your GP know of your decision.

If you require any further information about the study please do not hesitate to contact me on 0131 537 1834 or Cathy Liddle , Liaison Sister, Western General Hospital (who is independent of this study) on 0131 537 1762.

Please could you fill-in the enclosed reply slip and return it to me within fourteen days in the stamped-addressed envelope provided. If you wish to take part in this study, could you also sign both copies of the consent form and return one copy to me.

I look forward to hearing from you.

Yours sincerely

Joanne Dent
Psychologist

Enc.

REPLY SLIP

Please circle your response

I am willing to take part in this study, to be interviewed and complete questionnaires **YES** **NO**

If yes, please could you give your telephone number to arrange an appointment
(The interview could be at your home, or the Western General, whatever you prefer)

Daytime Tel:.....

Evening Tel:.....

I am unavailable for interview, but would still be willing to receive and complete questionnaires by post.

YES **NO**

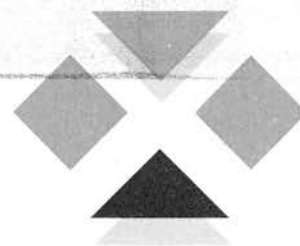
Signature:.....

Please print your name in block capitals.....

THANK YOU FOR YOUR CO-OPERATION

Consultants: *Dr G K Crompton*
Dr A P Greening
Dr J A Innes

Telephone: *Mobile 08313 10377*
Secretary 537 1783
Ward 537 1787



26th March 1997

INFORMATION SHEET

WESTERN
 GENERAL
 HOSPITALS
 NHS TRUST

EDINBURGH

Dear.....

RE: PSYCHOLOGICAL FACTORS AND QUALITY OF LIFE IN CYSTIC FIBROSIS STUDY

I am a Psychologist who is currently attached to the Western General Hospital in Edinburgh. I am conducting research into which psychological factors are important to adults with Cystic Fibrosis.

Most research projects require that subjects in the study, in this case, adults with Cystic Fibrosis, are compared to a group of healthy people from the general population. I am looking for about 30 volunteers to act as these control subjects, who are a similar age to people in the study. Your GP has given me permission to write and ask you if you would be willing to help in this research as one of these control subjects.

Cystic Fibrosis is a genetic disease which requires further research in order to understand its impact on the individual. It is also hoped that the results of this study will help health professionals in planning treatments and services for people with this disease.

As a control subject you would be asked to fill-in questionnaires about your mood, the way you see yourself, and how much help you receive from friends. There would also be a questionnaire about some of the things which could affect the quality of your life, such as sleep, and daily activities. There are five questionnaires in total which should not take more than 30 minutes to complete overall. All responses to questionnaires would be treated in the strictest confidence and would allow you to remain anonymous. Information would only be available to myself as sole researcher.

I would appreciate your participation in this research and will telephone you in ten days time to find out your decision. If you do not wish me to telephone you, please phone or write to me within this period. If you decide to take part I will then send you the questionnaires by post.

If you require any further information about the study please do not hesitate to contact me on **0131 537 1834** or Cathy Liddle, Liaison Sister, Western General Hospital (who is independent of this study) on 0131 537 1762.

Thankyou for your co-operation, I look forward to speaking with you..

Yours sincerely

Joanne Dent
 Psychologist

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LOTHIAN RESEARCH ETHICS COMMITTEE

STANDARD CONSENT FORM

Please read carefully and sign both copies, keep one copy and return the other with your reply-slip in the stamped-addressed envelope provided.

TITLE OF THE PROPOSED RESEARCH:

Psychological factors and quality of life in adults with Cystic Fibrosis

NAME OF INVESTIGATOR:

Joanne Dent, Psychologist

ADDRESS:

Department of Clinical Psychology, Western General Hospital, Crewe Road, Edinburgh

TELEPHONE:

0131 537 1834

FURTHER INFORMATION IS AVAILABLE FROM: (A person who is not involved in the study)

Cathy Liddle Tel: 0131 537 1762

LIST ANY DRUGS TO BE GIVEN IN THE STUDY EXPLAINING THEIR ACTION:

N.A.

GENERAL INFORMATION

Please state your age _____

Are you currently working? **YES / NO**
(Please circle the response)

If yes, what is your occupation? (Please state if you are a student)

Do you expect to be working in the next year? **YES / NO**

If yes, please indicate what work or study this will be

If no, please state reasons

Please state the number of years you have spent in full-time education since the age of 11

Your relationship status: please circle the one which applies to you:

Single

In a relationship
but not co-habiting

Co-habiting

Married

Separated

Divorced

Please state the relationship of those you live with to yourself : eg.Mother, brother, wife , live alone etc.

What is the occupation of the householder ? Please state if this is yourself

BODY IMAGE QUESTIONNAIRE

Please read the following questions carefully and put a tick in the box which corresponds to your answer. Please write as much as you can in the spaces provided.

1) How satisfied are you with your physical appearance?

Not at all	A little	A moderate amount	Quite alot	Very much
<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Comments _____

2) Do you think that other people find you physically attractive ?

Not at all	A little	A moderate amount	Quite alot	Very much
<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

3) Has the way you feel about you physical appearance ever prevented you from doing something you wanted to?

Not at all	A little	A moderate amount	Quite alot	Very much
<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Please specify: _____

4) Do you wish that you could change some aspect of your physical appearance?

Not at all	A little	A moderate amount	Quite alot	Very much
<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Please specify what aspect/s this would be if any: _____

WHOQOL

Instructions

This questionnaire asks how you feel about your quality of life, health, and other areas of your life. Please answer all the questions. If you are unsure about which response to give to a question, please choose the one that appears most appropriate. This can often be your first response.

Please keep in mind your standards, hopes, pleasures and concerns. We ask that you think about your life in the **last two weeks**.

For example, thinking about the last two weeks, a question might ask:

How much do you worry about your health?

Not at all	A little	A moderate amount	Very much	An extreme amount
1	2	3	4	5

You should circle the number that best fits how much you have worried about your health over the last two weeks. So you would circle the number 4 if you worried about your health "Very much", or circle number 1 if you have worried "Not at all" about your health. Please read each question, assess your feelings, and circle the number on the scale for each question that gives the best answer for you.

Thank you for your help

The following questions ask about **how much** you have experienced certain things in the last two weeks, for example, positive feelings such as happiness or contentment. If you have experienced these things an extreme amount circle the number next to "An extreme amount". If you have not experienced these things at all, circle the number next to "Not at all". You should circle one of the numbers in between if you wish to indicate your answer lies somewhere between "Not at all" and "Extremely". Questions refer to the last two weeks.

- F2.2(F2.1.3) How easily do you get tired?
- | | | | | |
|-----------------|---------------|-----------------|-----------|----------------|
| Not at all
1 | Slightly
2 | Moderately
3 | Very
4 | Extremely
5 |
|-----------------|---------------|-----------------|-----------|----------------|
- F2.4 (F2.2.4) How much are you bothered by fatigue?
- | | | | | |
|-----------------|---------------|-----------------|-----------|----------------|
| Not at all
1 | Slightly
2 | Moderately
3 | Very
4 | Extremely
5 |
|-----------------|---------------|-----------------|-----------|----------------|
- F3.2 (F4.1.3) Do you have any difficulties with sleeping?
- | | | | | |
|------------------|---------------|------------------------|----------------|------------------------|
| None at all
1 | A little
2 | A moderate amount
3 | Very much
4 | An extreme amount
5 |
|------------------|---------------|------------------------|----------------|------------------------|
- F3.4 (F4.2.3) How much do any sleep problems worry you?
- | | | | | |
|-----------------|---------------|------------------------|----------------|------------------------|
| Not at all
1 | A little
2 | A moderate amount
3 | Very much
4 | An extreme amount
5 |
|-----------------|---------------|------------------------|----------------|------------------------|
- F4.1 (F6.1.2) How much do you enjoy life?
- | | | | | |
|-----------------|---------------|------------------------|----------------|------------------------|
| Not at all
1 | A little
2 | A moderate amount
3 | Very much
4 | An extreme amount
5 |
|-----------------|---------------|------------------------|----------------|------------------------|

F4.3 (F6.1.4) How positive do you feel about the future?

Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
-----------------	---------------	-----------------	-----------	----------------

F4.4 (F6.1.6) How much do you experience positive feelings in your life?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F10.2 (F12.1.3) To what extent do you have difficulty in performing your routine activities?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F10.4 (F12.2.4) How much are you bothered by any limitations in performing everyday living activities?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F11.2 (F13.1.3) How much do you need any medication to function in your daily life?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F11.3 (F13.1.4) How much do you need any medical treatment to function in your daily life?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F11.4 (F13.2.2) To what extent does your quality of life depend on the use of medical substances or medical aids?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F15.2 (F3.1.2) How well are your sexual needs fulfilled?

Not at all 1	Slightly 2	Moderately 3	Very much 4	Extremely 5
-----------------	---------------	-----------------	----------------	----------------

F15.4 (F3.2.3) Are you bothered by any difficulties in your sex life?

Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
-----------------	---------------	-----------------	-----------	----------------

F18.2 (F23.1.5) Do you have financial difficulties?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F18.4 (F23.2.4) How much do you worry about money?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F19.1 (F24.1.1) How easily are you able to get good medical care?

Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
-----------------	---------------	-----------------	-----------	----------------

F21.3(F26.2.2)How much do you enjoy your free time?

Not at all 1	A little 2	Moderately 3	Very much 4	An extreme amount 5
-----------------	---------------	-----------------	----------------	------------------------

F23.2(F28.1.4)To what extent do you have problems with transport?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F23.4(F28.2.3)How much do difficulties with transport restrict your life?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

The following questions ask about **how completely** you experience or were able to do certain things in the last two weeks, for example activities of daily living such as washing, dressing or eating. If you have been able to do these things completely, circle the number next to "Completely". If you have not been able to do these things at all, circle the number next to "Not at all". You should circle one of the numbers in between if you wish to indicate your answer lies somewhere between "Not at all" and "Completely". Questions refer to the last two weeks.

F2.1(F2.1.1) Do you have enough energy for everyday life?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F10.1(F12.1.1)To what extent are you able to carry out your daily activities?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F11.1(F13.1.1)How dependent are you on medications?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F18.1(F23.1.1)Have you enough money to meet your needs?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F21.1(F26.1.2)To what extent do you have the opportunity for leisure activities?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F21.2(F26.1.3)How much are you able to relax and enjoy yourself?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F23.1(F28.1.2) To what extent do you have adequate means of transport?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

The following questions ask you to say how **satisfied, happy or good** you have felt about various aspects of your life over the last two weeks . For example, about your family life or the energy that you have. Decide how satisfied or dissatisfied you are with each aspect of your life and circle the number that best fits how you feel about this. Questions refer to the last two weeks.

G2 (G2.1) How satisfied are you with the quality of your life?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

G3 (G2.2) In general, how satisfied are you with your life?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

G4 (G2.3) How satisfied are you with your health?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F2.3 (F2.2.1) How satisfied are you with the energy that you have?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F3.3(F4.2.2) How satisfied are you with your sleep?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F10.3(F12.2.3) How satisfied are you with your ability to perform your daily living activities?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F15.3(F3.2.1) How satisfied are you with your sex life?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F18.3(F23.2.3) How satisfied are you with your financial situation?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F19.3(F24.2.1) How satisfied are you with your access to health services?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F19.4(F24.2.5) How satisfied are you with the social care services?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F21.4(F26.2.3) How satisfied are you with the way you spend your spare time?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F23.3(F28.2.2) How satisfied are you with your transport?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

G1(G1.1) How would you rate your quality of life?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

F15.1(F3.1.1) How would you rate your sex life?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

F3.1(F4.1.1) How well do you sleep?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

F19.2(F24.1.5)How would you rate the quality of social services available to you?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

The following question refers to **how often** you have felt or experienced certain things, for example the support of your family or friends or negative experiences such as feeling unsafe. If you have not experienced these things at all in the last two weeks, circle the number next to the response "never". If you have experienced these things, decide how often and circle the appropriate number. So for example if you have experienced pain all the time in the last two weeks circle the number next to "Always". Questions refer to the last two weeks.

F4.2 (F6.1.3) Do you generally feel content?

Never 1	Seldom 2	Quite often 3	Very often 4	Always 5
------------	-------------	------------------	-----------------	-------------

The following questions refer to any "**work**" that you do. Work here means any major activity that you do. This includes voluntary work, studying full-time, taking care of the home, taking care of children, paid work or unpaid work. So work, as it is used here, means the activities you feel take up a major part of your time and energy. Questions refer to the last two weeks.

F12.1 (F16.1.1)Are you able to work?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F12.2 (F16.1.2)Do you feel able to carry out your duties?

Not at all 1	A little 2	Moderately 3	Mostly 4	Completely 5
-----------------	---------------	-----------------	-------------	-----------------

F12.4(F16.2.1)How satisfied are you with your capacity for work?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

F12.3(F16.1.3)How would you rate your ability to work?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

The next few questions ask about **how well you were able to move around**, in the last two weeks. This refers to your physical ability to move your body in such a way as to allow you to move about and do the things you would like to do, as well as the things that you need to do.

F9.1(F11.1.1) How well are you able to get around?

Very poor 1	Poor 2	Neither poor nor good 3	Good 4	Very good 5
----------------	-----------	----------------------------	-----------	----------------

F9.3(F11.2.2) How much do any difficulties in mobility bother you?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F9.4(F11.2.3) To what extent do any difficulties in movement affect your way of life?

Not at all 1	A little 2	A moderate amount 3	Very much 4	An extreme amount 5
-----------------	---------------	------------------------	----------------	------------------------

F9.2(F11.2.1) How satisfied are you with your ability to move around?

Very dissatisfied 1	Dissatisfied 2	Neither satisfied nor dissatisfied 3	Satisfied 4	Very satisfied 5
------------------------	-------------------	--	----------------	---------------------

THANK YOU FOR YOUR HELP

ACCEPTANCE OF ILLNESS SCALE

Name:

Date: Record Number:

Instructions

Please respond to each of the following items by choosing a number from 1 to 5 on the scale adjacent to the item which you feel best describes you. Then **circle** the number you have chosen. There are no right answers to any of the questions.

1. I have a hard time adjusting to the limitations of my illness.

Strongly agree 1 2 3 4 5 Strongly disagree

2. Because of my health, I miss the things I like to do most.

Strongly agree 1 2 3 4 5 Strongly disagree

3. My illness makes me feel useless at times.

Strongly agree 1 2 3 4 5 Strongly disagree

4. Health problems make me more dependent on others than I want to be.

Strongly agree 1 2 3 4 5 Strongly disagree

5. My illness makes me a burden on my family and friends.

Strongly agree 1 2 3 4 5 Strongly disagree

6. My health does not make me feel inadequate.

Strongly agree 1 2 3 4 5 Strongly disagree

7. I will never be self-sufficient enough to make me happy.

Strongly agree 1 2 3 4 5 Strongly disagree

8. I think people are often uncomfortable being around me because of my illness.

Strongly agree 1 2 3 4 5 Strongly disagree

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SIGNIFICANT OTHERS SCALE (B)



Name:

Date: Record Number:

Instructions

Please list below up to seven people who may be important in Your life. Typical relationships include partner, mother, father, child, sibling, close friends, For each person please circle a number from 1 to 7 to show how well he or she provides the type of help that is listed.

The second part of each question asks you to rate how individuals would like things to be if they were exactly as they hoped for. As before, please put a circle around one number between 1 and 7 to show what the rating is.

Person 1 –

	Never		Sometimes		Always		
1 a) Can you trust, talk to frankly and share your feelings with this person?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
2 a) Can you lean on and turn to this person in times of difficulty? ..	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
3 a) Does he/she give you practical help?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
4 a) Can you spend time with him/her socially?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7

Person 2 –

1 a) Can you trust, talk to frankly and share your feelings with this person?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
2 a) Can you lean on and turn to this person in times of difficulty? ..	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
3 a) Does he/she give you practical help?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
4 a) Can you spend time with him/her socially?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7

Person 3 –

1 a) Can you trust, talk to frankly and share your feelings with this person?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
2 a) Can you lean on and turn to this person in times of difficulty?	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
3 a) Does he/she give you practical help?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7
4 a) Can you spend time with him/her socially?.....	1	2	3	4	5	6	7
b) What rating would your ideal be?.....	1	2	3	4	5	6	7

PLEASE CIRCLE ONE NUMBER ONLY FOR EACH QUESTION



Person 4 –		Never		Sometimes			Always	
1	a) Can you trust, talk to frankly and share your feelings with this person?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
2	a) Can you lean on and turn to this person in times of difficulty?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
3	a) Does he/she give you practical help?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
4	a) Can you spend time with him/her socially?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7

Person 5 –		Never		Sometimes			Always	
1	a) Can you trust, talk to frankly and share your feelings with this person?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
2	a) Can you lean on and turn to this person in times of difficulty?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
3	a) Does he/she give you practical help?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
4	a) Can you spend time with him/her socially?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7

Person 6 –		Never		Sometimes			Always	
1	a) Can you trust, talk to frankly and share your feelings with this person?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
2	a) Can you lean on and turn to this person in times of difficulty?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
3	a) Does he/she give you practical help?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
4	a) Can you spend time with him/her socially?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7

Person 7 –		Never		Sometimes			Always	
1	a) Can you trust, talk to frankly and share your feelings with this person?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
2	a) Can you lean on and turn to this person in times of difficulty?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
3	a) Does he/she give you practical help?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7
4	a) Can you spend time with him/her socially?	1	2	3	4	5	6	7
	b) What rating would your ideal be?	1	2	3	4	5	6	7

PLEASE CIRCLE ONE NUMBER ONLY FOR EACH QUESTION

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This measure is part of *Measures in Health Psychology: A User's Portfolio*, written and compiled by Professor John Weinman, Dr Stephen Wright and Professor Marie Johnston. Once the invoice has been paid, it may be photocopied for use **within the purchasing institution only**. Published by The NFER-NELSON Publishing Company Ltd, Darville House, 2 Oxford Road East, Windsor, Berkshire SL4 1DF, UK.

Code 4920 05 4



List of Abbreviations

- *FEV1 % predicted*

Forced expiratory volume at one second. This is a lung function test , percent predicted refers to the percentage of functioning lung capacity when compared with a predicted value based on age and weight for each person. The formula is as follows:

$$\frac{\text{Actual FEV1}}{\text{Predicted FEV1}} \times 100$$

- *BMI*

Body Mass Index. This is a formula which takes into account a person's height and weight, it is calculated as follows:

$$\frac{\text{Weight (in Kgs)}}{\text{Height (metres)}^2}$$

- *CF*

Cystic Fibrosis

- *Port*

Port-o-cath. This is a device inserted into the chest wall through which medications can be administered to take a rapid effect. It is often used by the patient themselves for home treatment instead of having to take medication intravenously .

- *IVs*

Intravenous line. This is usually used for antibiotic treatment, and can be administered in hospital or at home.

Sample Questions from the interviews with CF patients

These are examples of the types of questions which were asked during the interviews. They were not asked in this particular order and were flexible according to the respondent's replies. This is a guide to the type of structure the interview had:

General Information

"Please could you start by telling me a bit about yourself, where are you from?"

"Where did you go to school? ; did you enjoy school?"

"Do you have any brothers or sisters?"(to elicit those who have CF, who have died or are carriers etc.)

Diagnosis/prognosis

"When did you find out you had CF?; what do you remember about that time"

"What effect did CF have when you were growing up?"

(on schooling, relationships)

Autonomy, impact of CF on family

"How did your parents cope when they found out?"

"How do you get on with your parents?; do you see much of them now?"

"Who do you live with?"

Employment / daily activities

"Are you working at the moment? what do you do?"

"Has CF had any effect on your work...in what way?"

"What do you do now you're not working to keep yourself occupied?"

Disclosure/ relationships

"How much do you tell people about CF?"

"When would you tell them?"

"What sort of reactions have you had from people?"

"How did you tell your partner/spouse about CF?"

"Has having CF had any effect on your relationship with your partner/spouse?"

Fertility/pregnancy

"What about having children, is this something you've thought about?"

"How did you find out that you couldn't have children?"

Death/ worries about future

“Do you think about the future at all?”

“Do you have any particular worries about the future?”

“Who do you talk to about these worries?”

Quality of life

“What treatments have made the most difference to your quality of life?”

“What have been the worst things about having CF..what are the most difficult treatments?”

“Is there anything that CF prevents you from doing?”

Social support

“Who have been the most helpful people to you over the years?...in what way?”

Information/services

“Do you feel you know enough about CF?”

“Do you think that the services you receive could be better in any way?”

Coping strategies

“Do you have any particular religious or spiritual beliefs that have helped you?”

“What sorts of things have helped you cope with CF?”

“Was there ever a time when you haven't coped so well?”

Miscellaneous questions

“Are there any places or people that you avoid ?”

“What do you think about programmes on the television about CF?”

“Do you have any friends with CF?”

“Would you consider having a transplant if it was offered to you?”

“What do you say to your child about CF?”