THE MONITORING OF INTRACRANIAL PRESSURE IN INFANTS AND CHILDREN

by

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CONTENTS

CHAPTER 12

GROUP 'A' (Patients investigated on account of hydrocephalus). CASE NUMBERS 1-87.

Case Number 1

Name Y.P.

Age 9 years 4 months

Method Left parietal Rickham reservoir

Medical Diagnostic Background

She was born 7.10.67, the first child of healthy parents, after a normal pregnancy. Delivery was by S.V.D. at 41 weeks gestation following a $6\frac{1}{2}$ hour labour. Birth weight was 6lb 7 oz and no resuscitation was required at birth. She was admitted to the Royal Hospital for Sick Children, Edinburgh at 8 hours of age with a lumbar myelomeningocoele. There was marked moulding of the head and a left parietal haematoma. Head circumference at birth was 35 cms and the anterior fontanelle was soft; sutures were not separated. Her neurological level in the lower limbs was $L_{4/5}$ bilaterally. She had her myelomeningocoele closed at 16 hours of age and over the following week there was a significant improvement in motor and sensory function in both lower limbs down to approximately the S_1 level. An air ventriculogram on 11.10.67 showed moderate dilatation of the lateral and third ventricles, a cavum septum pallicidum present and a cerebral mantle at the occiput of 1.5 cms, at the vertex of 2 cms and in the frontal region of 2.5 cms.
A Pudenz ventriculo-peritoneal shunt was inserted on 23.10.67. After this operation the valve functioned well and the intracranial pressure was controlled. Prior to admission on this occasion, she had had five shunt revisions for either proximal or distal block and during one of these admissions a lumbar air encephalogram was carried out in May 1968 which revealed a block of the aqueduct of Sylvius.

Temperature 36.2

Zero Upper cortical subarachnoid space.

Duration 12 hours

Indication

Feverish and unwell for 2 days with neck stiffness and two 'turns' where her parents thought she was pale, sweaty and disinterested.

Resting Ventricular Pressure

In excess of 30 mm Hg.

Stress Ventricular Pressure

45 mm Hg (sleep)

Result Raised ventricular pressure

Action

Shunt revision was undertaken; however, due to technical difficulties, it could not be replaced and pressure was controlled with intermittent 'tapping'. At definitive replacement later, it was found that the ventricular end perforators were severely obstructed with hard crust.

Cardiac/Respiratory Artefact

CR = 7.5 mm before CSF drainage awake.

CR = 5 mm after CSF drainage awake

CR = 15 mm while asleep with high pressure.

Ventricular Dilatation

Mild hydrocephalus (non-communicating).

Points of Interest

On this admission shunt malfunction was suspected and after the exclusion of meningitis, arrangements were made for shunt revision. However, within the next hour an erythematous rash developed on the extensor surfaces of upper and lower limbs followed immediately by impairment of her conscious state (Fig. 70). Pressure monitoring

and CSF removal were undertaken which resulted in an immediate improvement of her conscious state. Routine neurological observations had been normal until the impairment of consciousness, shortly after the appearance of the rash. The rash returned twice, accompanied by rises in ventricular pressure prior to operation. It was clear therefore that with no abnormality of the vital signs or any other indication that the pressure was rising precipitously until the rash appeared at about 25 mm Hg pressure level, and if she had been tapped only intermittently, a less favourable outcome may have ensued. The Fig. shows one of the 'plateau waves' before she went on to continuous open ventricular drainage for the night. Note the wide pulse pressure here. Again the vital signs were steady, and with intermittent open ventricular drainage early in the night, 3 of these 'plateau waves' occurred before continuous open drainage was undertaken for the remainder of the night. Following relief of CSF and reduction in pressure level, there is then a gradual increase in CSF pressure in this child, compared with the precipitous increase in CSF pressure which occurred at other times. (Fig. 71) shows some abnormal rhythms of ventricular pressure occurring in short bursts while the pressure is low after the first open ventricular drainage. At this time she was quiet, conscious, her colour had improved and observations were stable. 30 mls of CSF had been removed prior to this section of recording. The nature of these abnormal rhythms is unclear. They were not associated with movement or any other obvious patient activity.

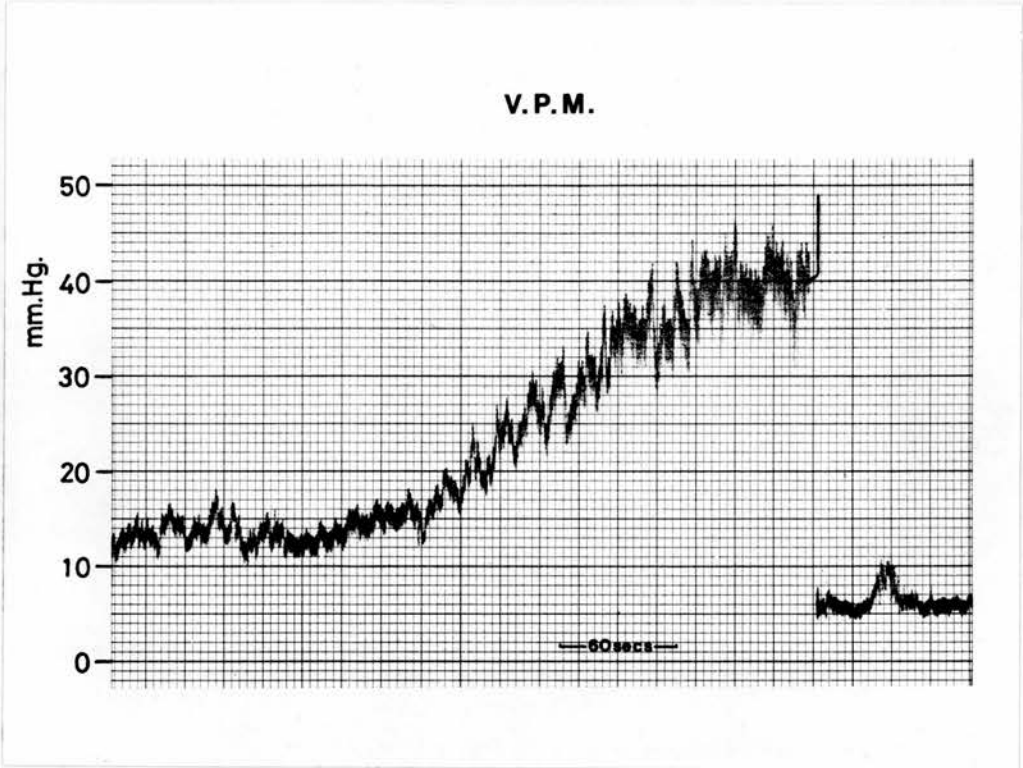


Fig. 70

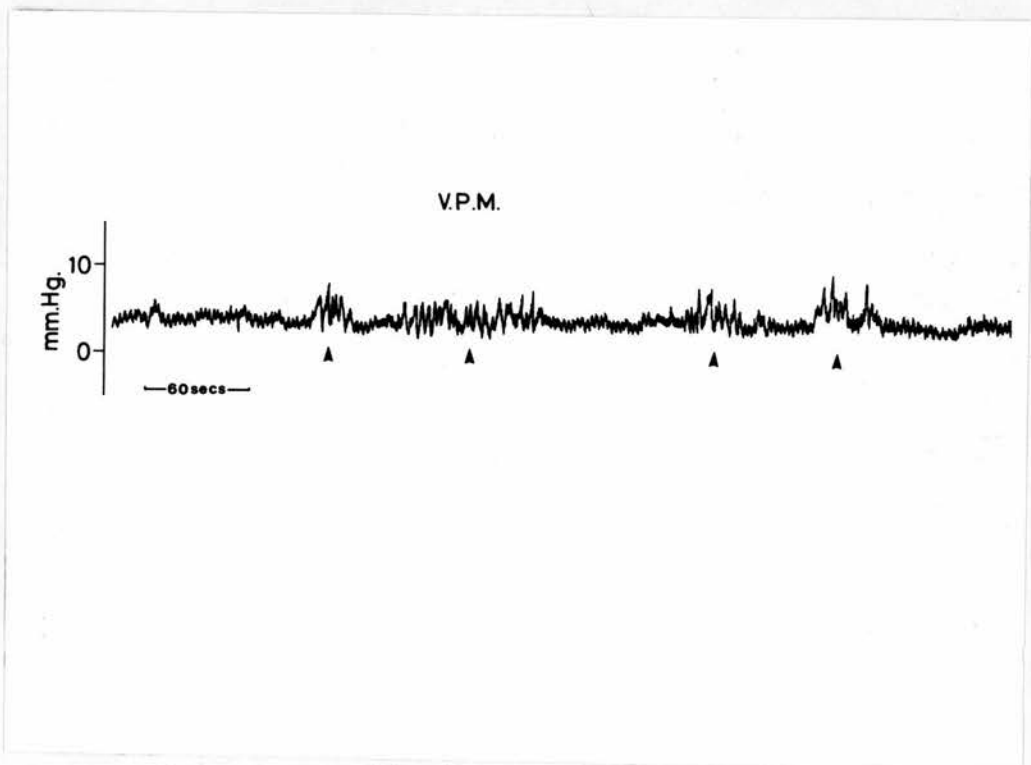


Fig. 71

Case Number 2

Name Y.P.

Age 10 years 10 months

Method Rickham reservoir

Medical Diagnostic Background

Myelomeningocoele (as before)

Temperature Normal

Zero

4" below the upper cortical subarachnoid space

Duration 1 hour

Indication

She presented with a 10 minute period of loss of consciousness the night before admission and a rash and headache associated with a stiff neck. She complained of a mild headache for one to two weeks.

Resting Ventricular Pressure

20 mm Hg.

Cerebral Perfusion Pressure

80 mm Hg initially.

Result

Raised ventricular pressure with no reduction of pressure on pumping the valve.

Action

Tapping the CSF reservoir until definitive revision.

Cardiac/Respiratory Artefact

R = 10 mm, C = 3 mm at a resting 20 mm Hg.

R = 5 mm, C = 2.5 mm at 13 mm Hg (5 mls of CSF released).

R = 4 mm, C = 2 mm at 11 mm Hg (10 mls of CSF released).

R = 4 mm, C = 1.5 mm at 7.5 mm Hg (15 mls of CSF released).

Pressure Recording

Her valve was pumped 10 times with no reduction in the level of ventricular pressure. At the outset the blood pressure was 120/80, ventricular pressure 20 mm Hg, i.e. cerebral perfusion pressure, CPP = 80 mm Hg with a pulse 120 per minute. After 5 mls of CSF released pulse was 120, BP 120/80, ventricular pressure 13, i.e. CPP = 87 mm Hg. A further 5 mls of CSF released, ventricular pressure = 11 mm Hg, BP 120/70, pulse 120, i.e. CPP = 84 mm Hg. A further 5 mls of CSF released, ventricular pressure = 7.5 mm Hg, pulse 132 per minute, BP 110/70, i.e. CPP = 82.5 mm Hg. From a clinical point of view it was established therefore that the valve was not reducing the level of ventricular pressure and the pressure was abnormally elevated. It was therefore necessary to proceed to a 'valvogram' with Dimer-X to further elucidate the site of blockage before operative revision.

Case Number	3
Name	A. McC.
Age	2 weeks
Method	Ventricular cannulation

Medical Diagnostic Background

This child was born on 5.11.76 in Edinburgh with a lumbo-sacral myelomeningocoele. Neurologically he had a bilateral L_4 motor level in the lower limbs and a sensory level of L_1 on the right and L_3 on the left. There was a large open defect measuring 5 x 6 cms and he also had a right reducable inguinal hernia and small hydrocoeles. His OFC at birth was 33.5 cms.

The spina bifida lesion was closed on 6.11.76 and the OFC on 8.11.76 was 33.8 cms. On 9.11.76 it was 34.2 cms and the fontanelle was rather

more tense. There was no vomiting and he was feeding well. On 11.11.76 the OFC was 34.5 cms, his fontanelle was unchanged and the fundi showed clear disc margins. He continued to feed well. On 16.11.76 his OFC was 35.2 cms and the anterior fontanelle was fuller. On 18.11.76 his pressure monitoring was carried out.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 22 hours

Indication — The assessment of suspected active neonatal hydrocephalus, (with clear disc margins, a full anterior fontanelle, strong A.T.N.R. and O.F.C. increasing).

Resting Ventricular Pressure

20 mm Hg

Result — Raised pressure

Action — Medium pressure ventriculo-peritoneal Pudenz shunt inserted on 20.11.76.

Cardiac/Respiratory Artefact

Very narrow tracing of variable width.

Head Circumference — 35.3 cms at VPM. 33.5 cms at birth.

Ventricular Dilatation/Cortical Mantle

Moderately large ventricles and a communicating hydrocephalus demonstrated 3.1.77.

Pressure Recording — (Fig. 72) shows three sections of ventricular pressure tracing before Dexamethasone (IV), 10 minutes and 60 minutes after, with little reduction in the elevated pressure level.

Follow Up — By 7.12.76 he needed a revision of the distal end of the shunt because it became entangled in the anterior abdominal wall. He subsequently developed a post-operative

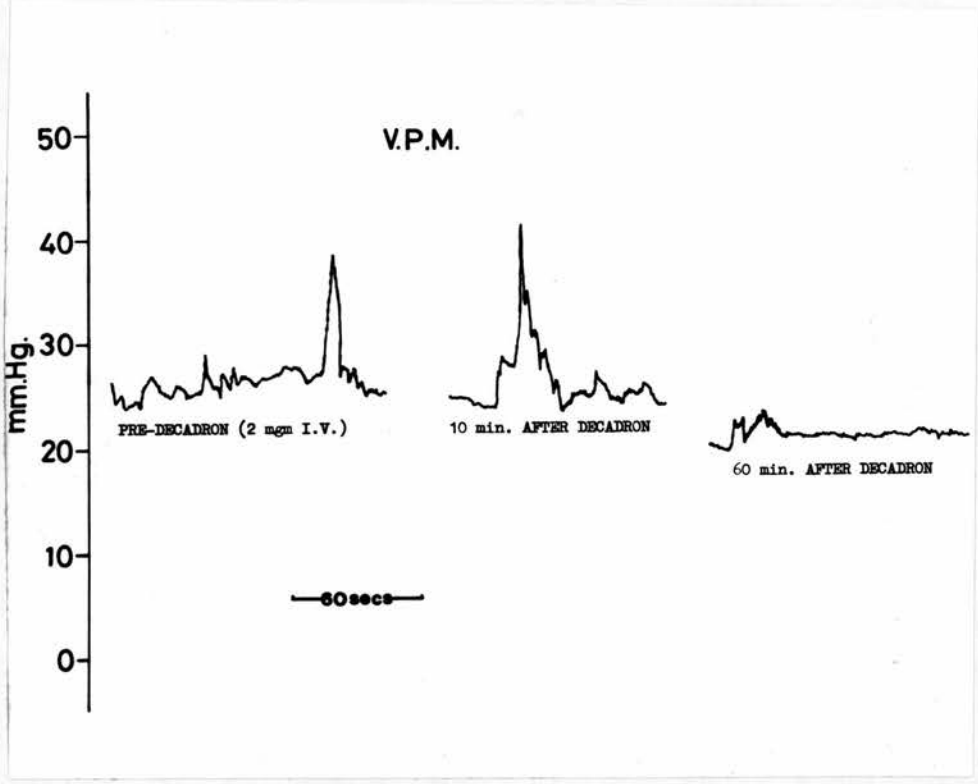

V.P.M.
50
40
30
20
10
0
mm.Hg.
PRE-DECADRON (2 mgm I.V.)
10 min. AFTER DECADRON
60 min. AFTER DECADRON
60secs

Fig.72

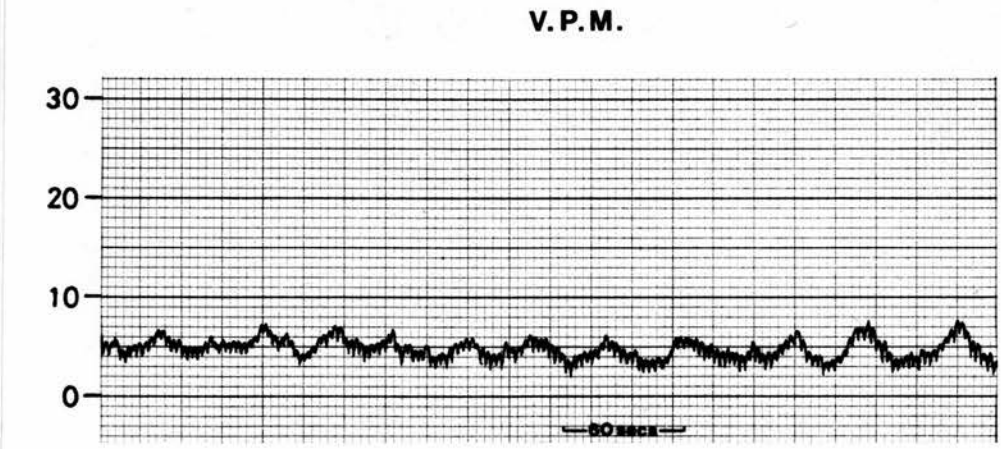

V.P.M.
30
20
10
0
60secs

Fig.73

ventriculitis and on 22.12.76 had a removal of his Pudenz shunt and insertion of a Rickham reservoir. A herniorrhaphy was done at this time. At a later operation on 3.2.77 a left ventriculo-peritoneal shunt was inserted and since that time he has progressed quite well but required one further shunt replacement in late 1977.

Case Number	4
Name	K.S.
Age	3 years 3 months
Method	Right parietal Rickham reservoir

Medical Diagnostic Background

She was born at term by Haig Ferguson's forceps delivery at a birth weight of 3.1 Kg. The baby's Apgar at birth was 2 at 1 minute, 6 at 5 minutes. She was intubated at 3 minutes and extubated at 7 minutes, but required re-intubation at 12 minutes for a further 12 minutes. Following this she was very irritable and a lumbar puncture showed xanthochromic CSF which was sterile on culture. Subdural taps were reported as negative on two occasions. Her general state improved but her OFC which was 34 cms at birth, increased to 36.9 cms by 3 weeks of age and at this time the anterior fontanelle was quite full. She remained irritable and was noted to be 'sunsetting'. At 3 weeks of age she was transferred to R.H.S.C. Neurological Unit and examination confirmed the elevated OFC, splaying of the sutures, no intracranial bruits and an absence of fixing. A slight divergent squint, reactive pupils and no facial weakness were also noted. A.T.N.R. was more marked on the right than on the left and tone appeared somewhat increased at this early age on the right side. When held suspended she was dystonic and had spontaneous Babinski responses. Other

systems were normal. A skull x-ray showed widened sutures, and an air encephalogram two days after admission showed posterior dilatation of the lateral ventricles and air in a dilated third ventricle. A lumbar air encephalogram indicated she had a 'communicating hydrocephalus'. On 16.10.74 a Rickham reservoir was inserted in the right parietal area and daily ventricular taps undertaken. A theco-peritoneal shunt was inserted on 28.10.74. On 13.11.74 she again came under pressure clinically and required ventricular tapping more than twice a day. Subsequently open ventricular drainage was instituted, contrast ventriculography was carried out and this now showed a non-communicating type of hydrocephalus.

It was decided to attempt to reconstitute the CSF pathway and a posterior fossa craniectomy was performed and a further splitting operation done. Subsequent to this she again came under pressure, until 18.12.74 when a left sided ventriculo-peritoneal shunt was inserted.

Her early diagnosis therefore is of birth asphyxia, hydrocephalus, a theco-peritoneal shunt followed by a ventriculo-peritoneal shunt because of non-communication. She slowly improved neurologically but remained hypotonic and ataxic with evidence of a left hemi-paresis. Her development has been slow and retarded to a degree.

Temperature — Normal

Zero — 3" above the upper cortical subarachnoid space.

Duration — 5 hours

Indication — She had become increasingly listless and tired over the previous 2 months and was more difficult to manage behaviourally. She had also developed a lump in the right subcostal

region and it was suggested that this may have been a pressure decompensation from her old theco-peritoneal shunt, if her CSF pathways had re-cannalised. The other alternative was an infective lesion associated with the remainder of her theco-peritoneal shunt which had not been previously removed. On admission her valve appeared to be taking longer to refill (45 seconds) and her usually poor appetite had worsened.

Resting Ventricular Pressure

6 mm Hg with the zero at the inter-ventricular foramina level, 4 mm Hg with the higher zero mentioned before. Her average resting pressure was 8 mm Hg with the zero at the upper level of the cortical subarachnoid space.

Stress Ventricular Pressure

36 mm Hg maximum peaks and 10.5 mm Hg peaks in sleep.

Result Confirmed a normal pressure.

Two days after this she was pyrexial with a blood leucocyte count of 24,500, 16 white cells in her CSF and an E.S.R. of 78. The lump had increased in her right subcostal region and was aspirated of 15 mls of pus. She was commenced on Ampicillin and Cloxacillin and on 27.12.77 an abscess was drained, the contents of which grew enterococci. On 12.1.78 the abscess cavity was re-opened and the theco-peritoneal shunt was dissected out and removed. The shunt end was located in the depths of the abscess cavity. The edges of the wound were excised to reduce the likelihood of recurrent infection and the wound was closed. Shortly after this she began feeding well, her weight was increasing, she was sleeping better, talking more fluently and her responses were more alert. When last seen in April 1978 she was progressing well with no evidence of any further infection or signs of raised intracranial pressure.

Cardiac/Respiratory Artefact

CR = 0.5 mm resting, CR = 1 mm in sleep.

Points of Interest The ventricular pressure monitoring on this occasion decided that the pressure was normal and that the swelling was not due to raised intracranial pressure. It also meant that with her present ventriculo-peritoneal shunt, no revision of this was necessary.

Theco-peritoneal shunts generally have a number of disadvantages compared to ventriculo-peritoneal shunts and it would appear clinically that their period of usefulness is somewhat limited due to:

(a) the possibility of the child converting from a communicating to a non-communicating hydrocephalus due to an intercurrent C.N.S. infection,

(b) problems of bone infection,

(c) hyperlordosis above the site of insertion and more importantly,

(d) should pressure symptoms occur with a theco-peritoneal shunt without a Rickham reservoir or other entry to the ventricular system, dangerous pressure effects may ensue.

Fig. 73 shows the effect of sleep in a child whose ventricular pressure level is in the normal range.

It is interesting that moving the zero $1\frac{1}{2}$" - 2" vertically in this child made no difference to the measured ventricular pressure level.

Case Number 5

Name J.McG.

Age 1 day

Method Ventricular cannulation

Medical Diagnostic Background

This child was born after an elective caesarean section for cephalo-pelvic disproportion which was performed at 42 weeks gestation by dates. Mother was a 20 year old and primiparous. Her pregnancy was normal with no drug ingestion, irradiation or antenatal infections. A maternal aunt was said to suffer from spina bifida. Apgar was 9 at one minute.

OFC. at birth was 39 cms and there was an obvious hydrocephalus associated with a lumbo-sacral myelomeningocoele. Alpha-feto protein estimations on blood had been performed during pregnancy but the levels were said to be within the normal range.

The child's general condition on arrival was satisfactory. His gestational age by clinical criteria was 38 weeks. B.P. 100 systolic and pulse 140 per minute. There was some ballooning of the abdominal wall below the costal margins bilaterally. All the sutures were widely separated and the fontanelles were of normal tension. OFC was 39.5 cms. There was no 'sunsetting' or dilatation of the superficial scalp veins. An extensive thoraco-lumbar myelomeningocoele with a leaking membrane and an easily visible neural plaque extended from T_{11} to S_1. The neurological sensory level in the lower limbs was T_{10} and the motor level was T_{12}, there was some evidence of 'isolated cord' function.

X-ray report of the spine showed some widening of the inter-pedicular distances from T_{12} to S_5 and there was an associated lumbar kyphosis. Cranial lacunae were seen in the skull x-ray.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 2 hours

Indication — The assessment of active neonatal hydrocephalus prior to a decision regarding 'selection' for treatment.

Resting Ventricular Pressure

11 mm Hg.

Stress Ventricular Pressure

28 mm Hg.

Result — Raised ventricular pressure.

Action — Nil

Cardiac/Respiratory Artefact

Too minimal for measurement.

OFC — 39.5 cms

Ventricular Dilatation/Cortical Mantle

Marked ventricular dilatation present.

Points of Interest — It is not exceptional that children with marked ventricular dilatation and an obvious active hydrocephalus have relatively smaller ventricular pressure levels than older children. For this reason I suspect that the upper limit of ventricular pressure in neonates should not be considered to be in excess of 10 mm Hg.

Pressure Recordings — Simultaneous recordings of arterial pressure via umbilical artery and CSF pressure from ventricular cannulation were performed. Fig. 74 shows the effect of abdominal palpation, increasing the ventricular pressure and decreasing the systemic arterial pressure. In Fig. 75 the effect of gentle head compression is seen. Fig. 76 shows the effect of breathing 100% oxygen and as can be seen the ventricular pressure level increases.

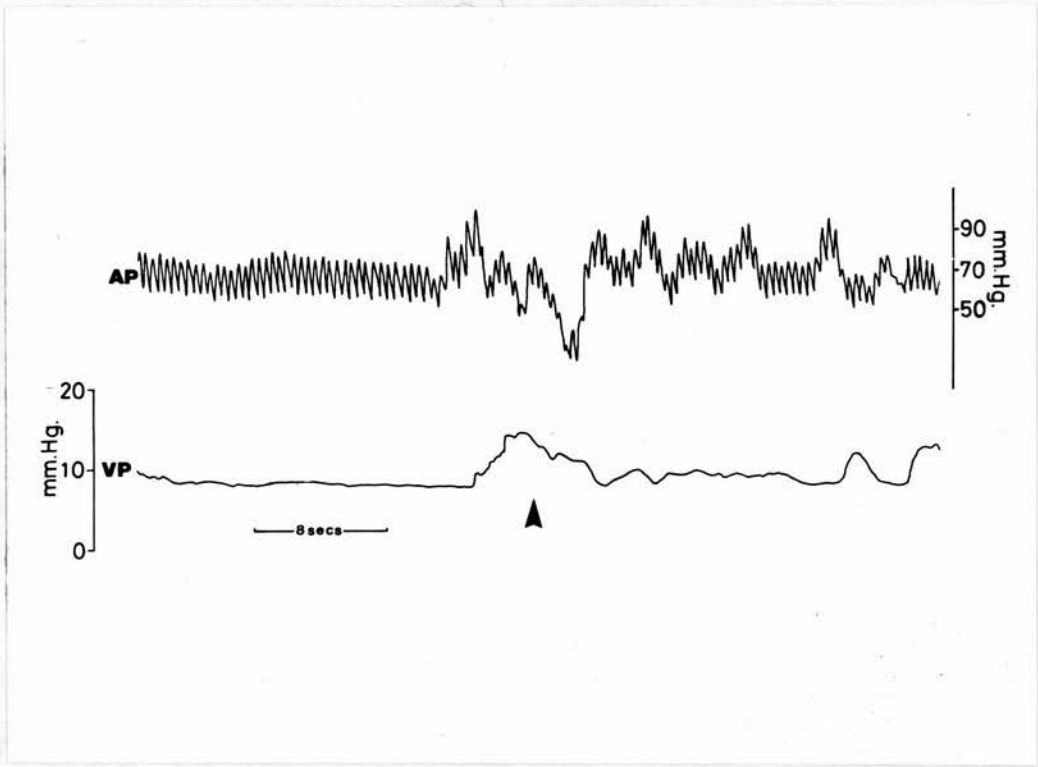

AP
90
70
50
mm.Hg.
20
10
0
mm.Hg.
VP
8 secs

Fig. 74

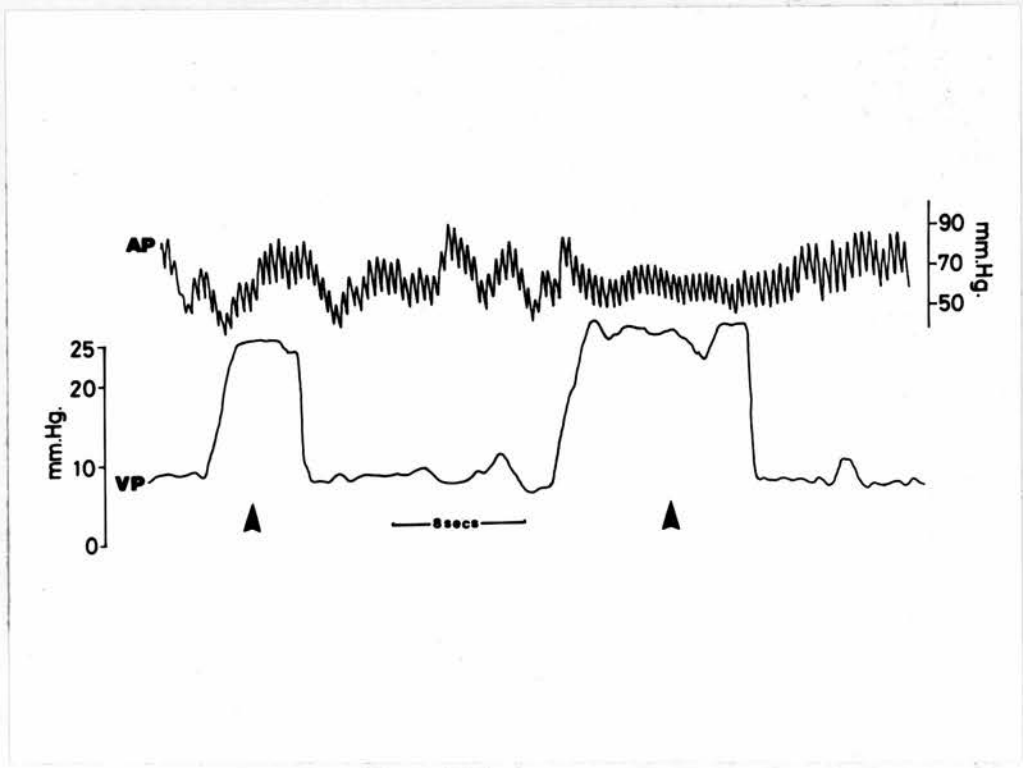

AP
90
70
50
mm.Hg.
25
20
10
0
mm.Hg.
VP
8 secs

Fig. 75

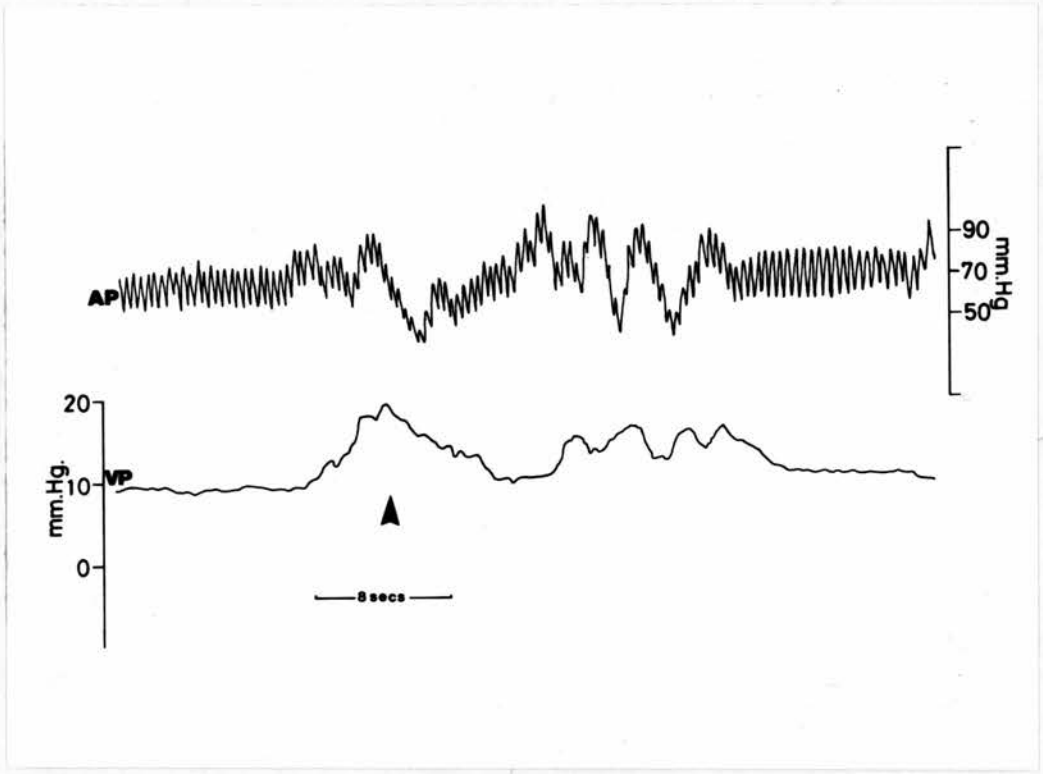

AP
90
70
50
mm.Hg.
20
10
0
mm.Hg.
VP
8 secs

Fig.76

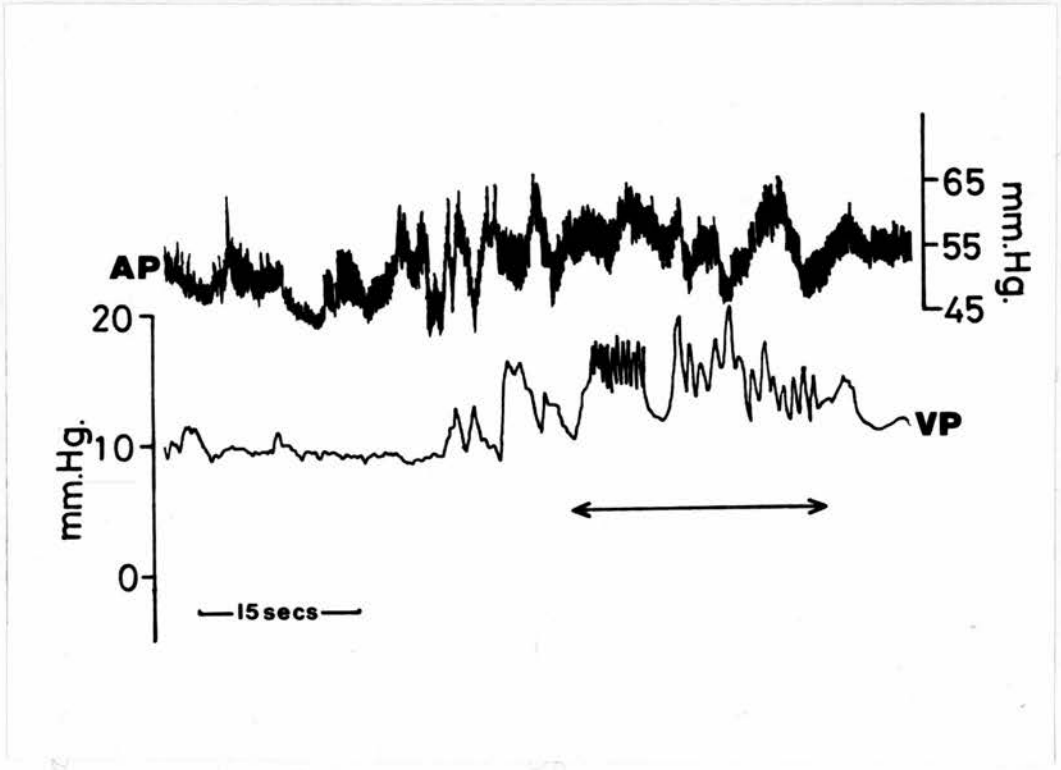

AP
65
55
45
mm.Hg.
20
10
0
mm.Hg.
VP
15 secs

Fig.77

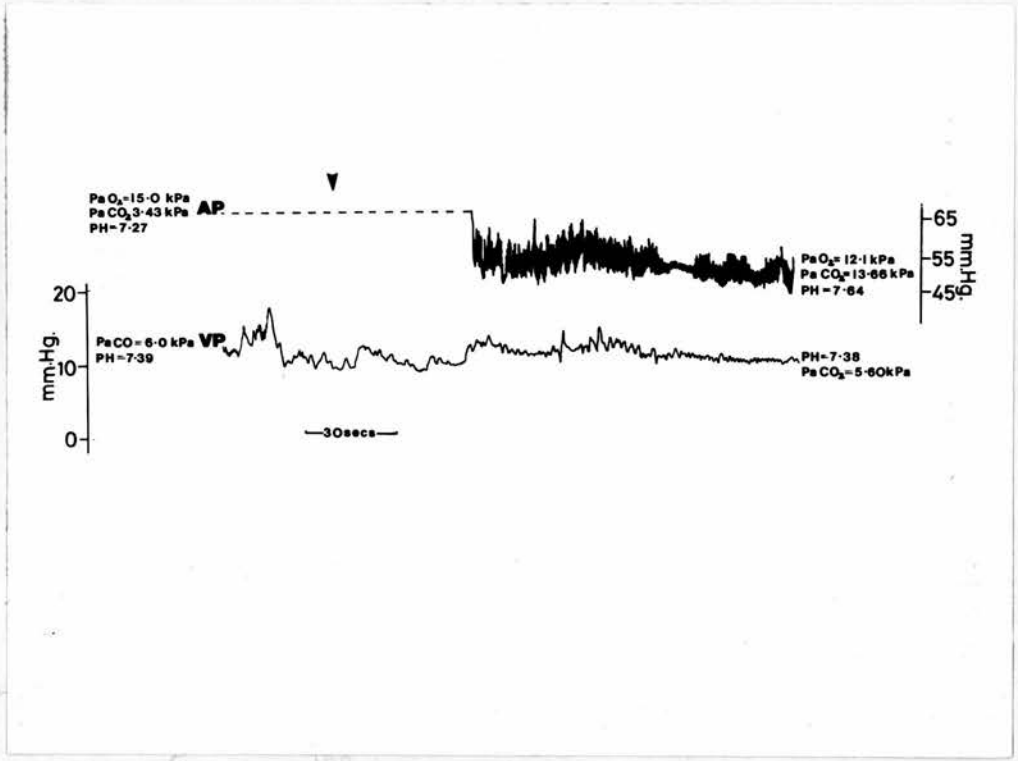

PaO2=15·0 kPa
PaCO2=3·43 kPa
PH=7·27
AP
PaO2= 12·1 kPa
PaCO2=13·66 kPa
PH=7·64
65
55
45
mmHg
20
10
0
mm.Hg.
PaCO=6·0 kPa
PH=7·39
VP
PH=7·38
PaCO2=5·60kPa
30secs

Fig. 78

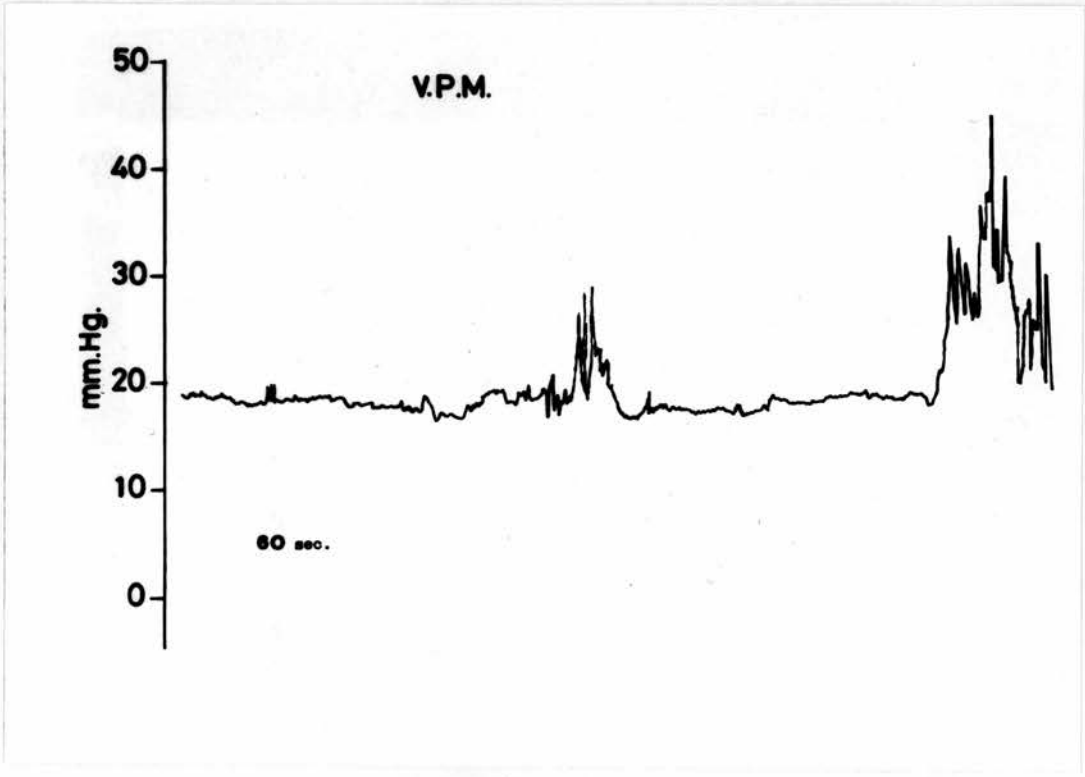

V.P.M.
50
40
30
20
10
0
mm.Hg.
60 sec.

Fig. 79

There may be a degree of re-breathing here. Fig. 77 shows the effect of depressing the anterior fontanelle, with its effect on pressure fluctuations in the ventricles, and simultaneous effect on the systemic arterial pressure. Lastly in Fig.78 the effect is seen of giving an intra-arterial bolus of 15 mls of sodium bicarbonate (8.3% w/v). pCO_2 and pH levels in the CSF do not change dramatically before and after this injection. Normally the CSF pH is 0.1 unit lower than pH of arterial blood because CO_2 diffuses quickly across the blood brain barrier whereas HCO_3 and H^+ are slower. The effect however on the systemic arterial pressure is one of over-correction in the blood gases and a reduction in the systemic pressure level. It must be remembered that it was necessary to sample a small amount of CSF to obtain these estimations in both cases, before and after administration. However, less than 1 ml was sampled on each occasion and this would not be expected to make a measurable difference to the resting ventricular pressure level. Pressure on the myelomeningocoele resulted in a similar increase in ventricular and arterial pressure as Queckenstedt's and eyeball pressure.

Case Number 6

Name S.W.

Age 1 day

Method Ventricular cannulation through the anterior fontanelle with a 20 gauge $1\frac{1}{2}$" steel needle.

Medical Diagnostic Background

The child was born by lower segment caesarean section for failure to progress and maternal hypertension. Apgar was 9 at 5 minutes. BW was 3.62 Kg. The child was noted to have gross hydrocephalus at birth and was transferred to R.H.S.C. for further management.

Mother was reasonably well during pregnancy with no infections, irradiations or medications. She was age 22 and a primagravida. Initial examination showed an OFC of 46 cms, an anterior fontanelle of 8 cms laterally; it was tense and the scalp veins were distended. The fontanelle merged with the metopic suture which was $2\frac{1}{2}$ cms separated. All the other sutures were markedly separated. There was a plagiocephaly with the cranial bones separated by CSF 'blowouts'. There was a large hypertrichosis tuft at the occiput measuring 2 x 3 cms and was oval in shape with a flat haemangioma surrounding this of 7 x 7 cms. The skull did not transilluminate. There were no cranial bruits, the child was mildly 'sunsetting' but there was a full range of eye movements and pupils were reacting to light. The fundi showed atrophic discs with marked venous congestion but no choroiditis. There was a bilateral grasp, equivocal A.T.N.R.'s and S.T.N.R.'s but a traction response and no decerebration posturing. The child rooted, sucked, had a symmetrically weak moro, placed, toe grasped and monosynaptic reflexes were 2+ in the upper limbs and 1+ in the lower limbs with extensor plantar responses. The spine showed no pits, dimples or hairy tufts. There was normal flexor tone with a full range of voluntary movements in the limbs and an anal and superficial abdominal reflex. The child was conscious and the vital signs, respiration, pulse, temperature and colour were satisfactory. Gestation was assessed clinically as term. Other systems, chest, cardiovascular etc. were normal. There were 3 vessels in the cord, the liver was tipped and the palate was intact. A skull x-ray showed gross cortical thinning, no calcification and a normal sella. Spinal x-rays showed no occult spina bifida. Although it would seem fairly obvious with this degree of hydrocephalus that treatment would not be undertaken, it was important to investigate

fully to arrive at an aetiological diagnosis as mother was primiparous. A detailed family history revealed no significant neurological conditions.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — Half an hour

Indication — Investigation of severe neonatal hydrocephalus and the ventricular pressure studies were carried out immediately prior to a pneumo-ventriculogram.

Resting Ventricular Pressure

18 mm Hg.

Stress Ventricular Pressure

A maximum peak of 45 mm Hg on crying.

Result — Raised ventricular pressure

Action — Nil from the point of view of management.

Cardiac/Respiratory Artefacts

Too minimal for accurate measurement.

OFC — 46 cms

Ventricular Dilatation/Cortical Mantle

Gross ventricular dilatation of lateral and third ventricles. No air in the fourth ventricle and a cortical mantle of 4 mm on pneumo-ventriculogram; therefore aqueduct stenosis. A C.T. scan confirmed an aqueduct stenosis.

Points of Interest — Post-mortem revealed a severe congenital internal hydrocephalus due to aqueductal malformation but no other developmental malformations. In particular, there was no resistance to the flow into the right internal carotid artery. An injection of the right artery resulted in the escape of the fixative through the left, but on the injection of the left artery it did not flow freely through the right artery. This finding suggests some block to the upward flow

of fluid through the internal carotid artery. The precise site of this block was not identified but it was clear that neither vessel was totally occluded. The brain was swollen and there was substantial thinning of the cerebral cortex. The cerebellum and brain stem were relatively normal by comparison although there was some reduction in the vertical diameter of the posterior fossa with corresponding reduction in the vertical height of the cerebellum. The spinal cord showed no abnormality. The appearances of this baby were almost of an hydranencephaly, it was interesting that the baby had a normal brain stem and cord function, though with virtually no cortex and a gross hydrocephalus. I could not be sure that this was a sex-linked recessive aqueduct stenosis but all the usual virological screens for cyto-megalovirus, toxoplasma, rubella, mumps, chromosomes, parental OFC et. were all normal. Mother has since had a further child who was perfectly normal.

Pressure Recording It was interesting here again that the pressure recording resting level is not in excess of 20 mm Hg with a gross hydrocephalus. Note also that the tracing itself is particularly thin, probably related to the very thin cortical mantle, and that the head is really a 'bag of water' damping any cardio-respiratory impulses excessively. Relatively small head movements give rise to sizeable ventricular pressure deflections (Fig. 79). A further study of ventricular pressure with concomitant EEG analysis was performed, the findings of which were inconclusive and have not been included in this report. Figs. 80 & 81 show a posterior view of the child and the brain at post-mortem.

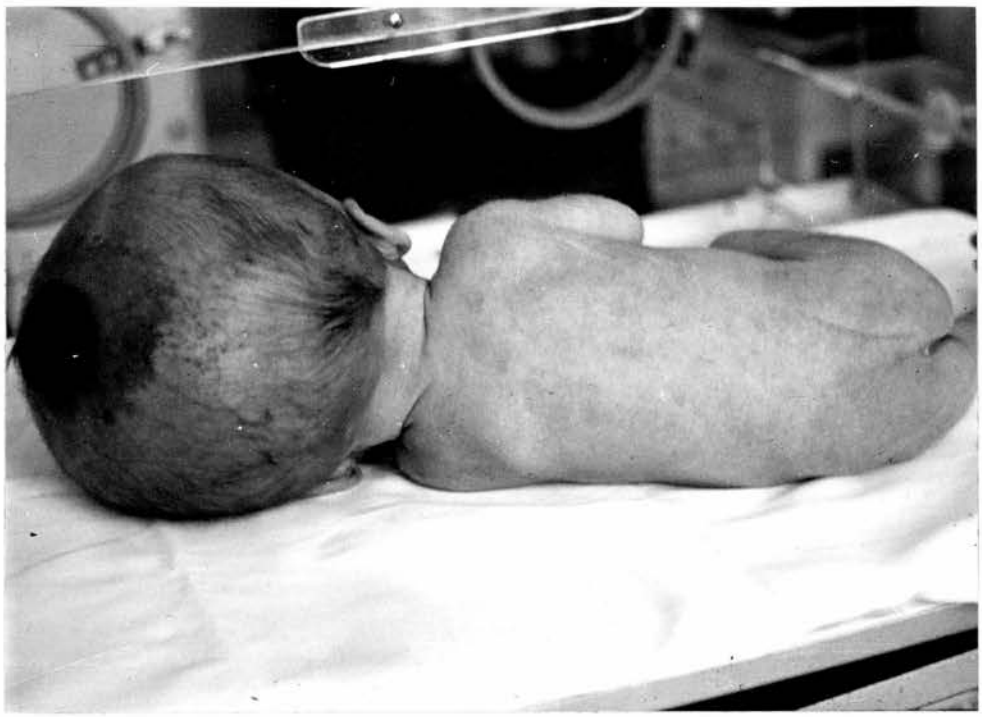


Fig.80

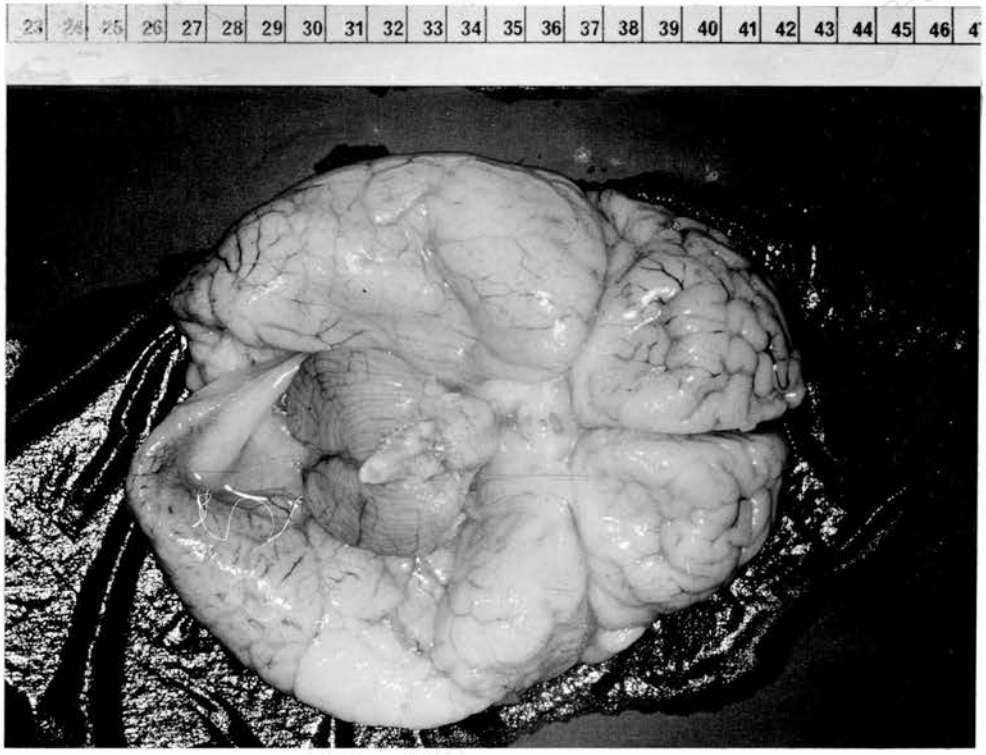


Fig.81

Case Number 7

Name M.F.

Age 5 months

Method Ventricular cannulation through post-auricular burr hole with 3.2 cm Teflon cannula.

Medical Diagnostic Background

The child was born to a 22 year old primiparous mother after a pregnancy that stopped at 30 weeks by spontaneous onset of labour. He was in good condition at birth and no resuscitation was required. BW was 1.59 Kg. He had a small sacral rachischisis but was managed conservatively because of his prematurity and inability to tolerate an anaesthetic at that time. The back lesion leaked CSF for a short time but then gradually epithelialised. His OFC increased by 3 cms during the initial period of observation and he was transferred for pressure studies and possible insertion of a valve. At the time of transfer his OFC was 33 cms, the fontanelle was full, he had a scaphocephalic shaped skull. There was some tendency towards dystonic posturing and good active movement down to the level of the ankles. Pressure studies were not undertaken immediately. At 2 months of age the sutures appeared slightly more splayed, the anterior fontanelle fuller and OFC 34 cms. An air ventriculogram at this time showed evidence of moderate dilatation of the ventricular system, an estimated cortical width of 2.5 cms at the vertex, having been 4 cms in an earlier study done shortly after birth.

He had a Spitz-Holter ventriculo-peritoneal shunt inserted and a low pressure valve.

He was next admitted at 4 monthsof age and although it was thought that the valve was working clinically, it was not thought to be as efficient as it should be, because, although he was contented, afebrile

and not irritable on admission, the scalp veins were rather distended and he was 'sunsetting'. He also had no control of his head at this time and persistent primitive reflexes and a left divergent strabismus were present. There was no evidence of papilloedema on fundoscopy.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 3 hours

Indication — The persistence of signs of raised pressure with a valve in situ, i.e. the question of competence of a single valve, although clinically it appeared normal.

Resting Ventricular Pressure

13 mm Hg.

Stress Ventricular Pressure

75 mm Hg on peaks on coughing and crying.

Result

The valve did not appear to be functioning effectively and there was no reduction in the level of ventricular pressure measured, on pumping the valve, until it had been pumped some 60 times and then it resulted in a 5 mm Hg drop in pressure only.

Action — Accordingly the Spitz Holter was removed and a Pudenz ventriculo-peritoneal shunt was inserted.

Cardiac/Respiratory Artefact

CR = 10 mm in sleep.

CR = 2.5 mm awake.

OFC — 45 cms

Ventricular Dilatation/Cortical Mantle

Severe ventricular dilatation with a cortical mantle of 10 mm.

Pressure Recordings — Fig. 82 shows the effect of awakening from sleep in this child with a gradual increase in ventricular pressure. Removal of 2 boluses of CSF, 19 mls in total, effectively reduces the ventricular pressure to the normal range. Pumping the valve 60 times caused a drop in CSF pressure of 5 mm Hg., the pressure then took 19 minutes to return to the pre-pumping level.

Follow Up — At follow up his OFC has progressed at an acceptable rate, and he has remained generally well with his new valve in situ.

Case Number	8
Name	A.L.
Age	$22\frac{1}{2}$ months
Method	Right sided Rickham reservoir

Medical Diagnostic Background

She was born with a myelomeningocoele which was operated on shortly after birth. Not long after this a ventriculo-peritoneal shunt was inserted. The shunt required refashioning at the age of 3 weeks and again at the age of 2 years when the peritoneal end was revised. She is epileptic and has had a number of major convulsions.

Temperature	$37^{\circ}C$ at the outset, $35.9^{\circ}C$ at the conclusion.
Zero	Inter-ventricular foramina level.
Duration	$2\frac{1}{2}$ hours

Indication — She had been admitted with constipation, loss of upward conjugate gaze and a non-tender distension of the peritoneal end of her tubing. At operation the distal end was found to be blocked. Two days later she was somewhat sleepy, had vomited twice, the anterior fontanelle was not tense but her valve was not clinically emptying. The indication therefore for pressure monitoring is a post-operative check on her intracranial pressure because of a

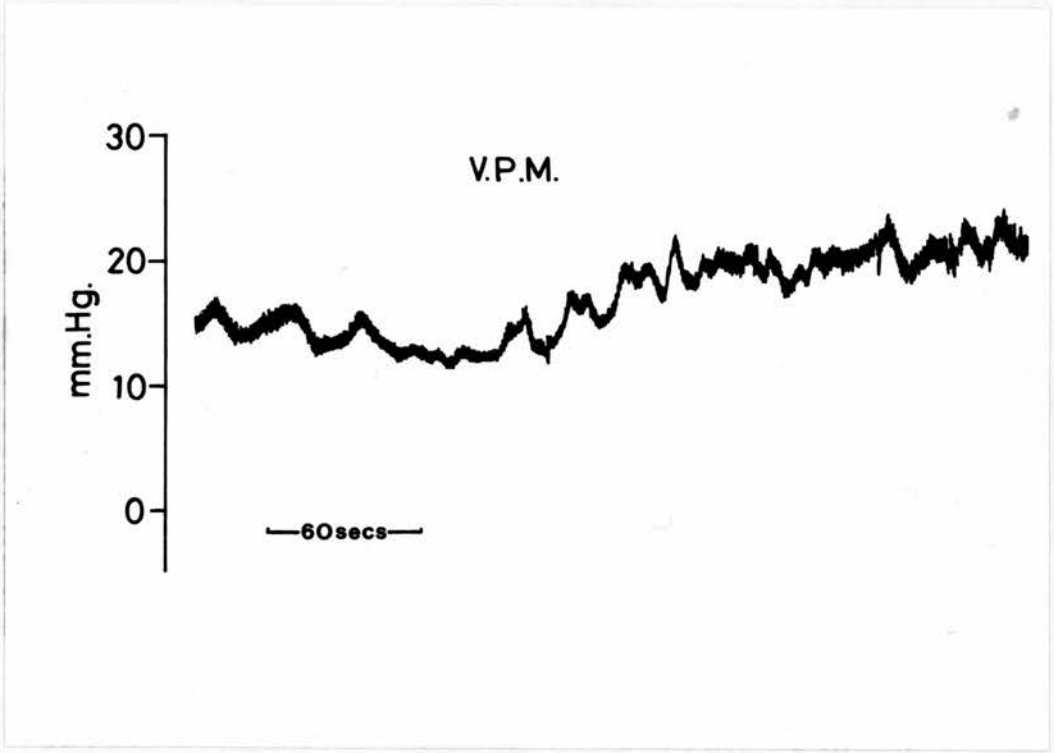

V.P.M.
30
20
10
0
mm.Hg.
60secs

Fig.82

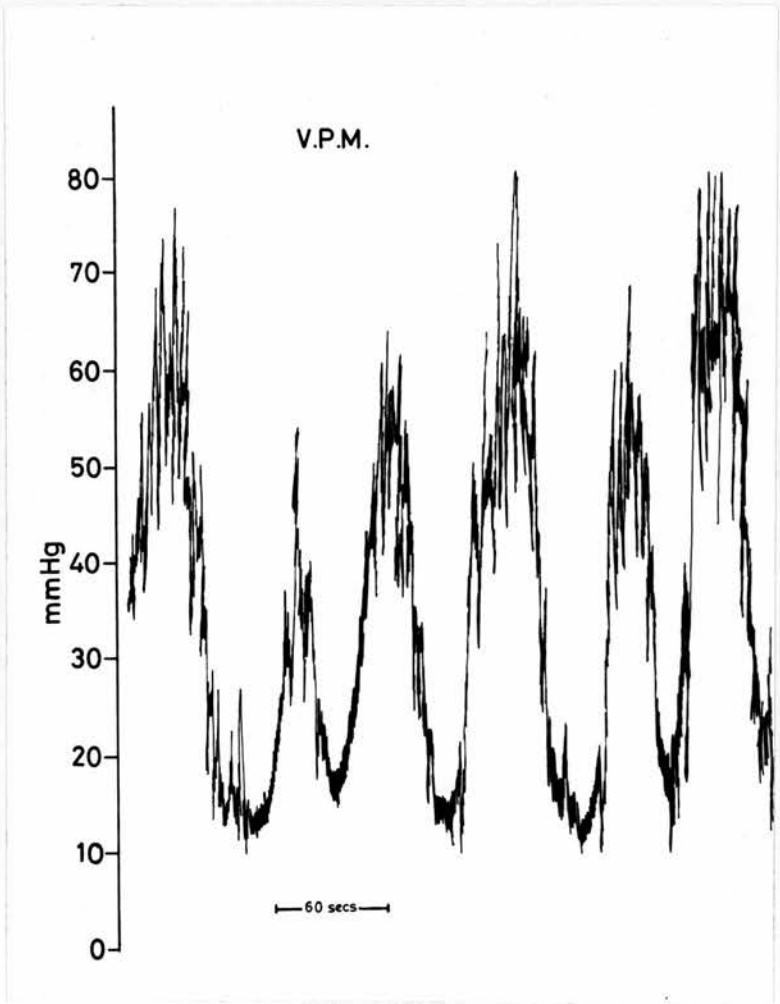

V.P.M.
80
70
60
50
40
30
20
10
0
mmHg
60 secs

Fig.83

clinical suspicion of early malfunction.

Resting Ventricular Pressure

8 mm Hg.

Stress Ventricular Pressure

A maximum peak of 45 mm Hg on crying, and a maximum peak of 23 mm Hg during sleep.

Result On pumping her valve 40 times, a reduction in the level of ventricular pressure of 2 mm Hg occurred. This was unsatisfactory as was the level of the pressure recorded during sleep.

Action She was returned to theatre where a suture was removed from the distal end of the tubing which had interrupted the passage of free CSF drainage.

Cardiac/Respiratory Artefact

CR = 7.5 mm during sleep,

CR = 1.25 mm awake.

OFC 46 cms

Ventricular Dilatation/Cortical Mantle

A thin cortical mantle and gross ventricular dilatation was seen on the initial air ventriculogram but no further estimate of ventricular size was obtained on this occasion.

Case Number 9

Name A.L.

Age 2 years 7 months

Method Spinal needle into left sided (old shunt site) burr hole.

Medical Diagnostic Background

Myelomeningocoele with shunt in situ as before.

Temperature 37.5

Zero — As before

Duration — Half an hour

Indication — Following the last case, her next admission was on 4.8.77 when she had revision of the lower end of her ventriculo-peritoneal shunt as an urgent procedure. Pre-operatively she had been drowsy and vaso-constricted. The tube was repositioned in the peritoneal cavity and she was discharged home only to be readmitted with the complaint of abdominal pain, drawing up her knees and screaming. She was afebrile but miserable. Pressure monitoring was again carried out with peaks to 80 and 100 mm Hg. She became quite distressed as the pressure rose past 65 mm Hg (Fig. 83). She was subsequently tapped from the reservoir as necessary with an ECG monitor and close neurological observations. The next day she had a revision of the shunt but post-operatively developed a ventriculitis which necessitated therapy with Gentamycin and Cloxacillin. Gram-positive cocci and 68 white cells were found in the CSF at this stage. The valve felt increasingly tight and on 13.9.77 both the Rickham reservoir and the Pudenz shunt were removed. At the time of removal the CSF was under some tension. She was subsequently treated with intra-ventricular antibiotics and CSF. tapping via her right sided burr hole. The next day a pressure recording was undertaken, the indication being to estimate the level of ventricular pressure after all her shunt and reservoir had been removed, and to give an idea of how much CSF. to remove to control her pressure. It is important not to take too little and leave the patient with pressure symptoms, and not to remove too much and promote further production of CSF. necessitating even more frequent 'tapping'.

Resting Ventricular Pressure

40 mm Hg.

Stress Ventricular Pressure

In excess of 80 mm Hg on crying.

Result — Raised ventricular pressure.

Action — Removal of 15 mls CSF caused the pressure to fall to less than 10 mm Hg., i.e. in the normal range. Note that a CSF specimen at this stage was blood stained and unsuitable for accurate cell count, but was sterile on culture. She continued to have taps and intra-ventricular antibiotics until 18.9.77 when she had a further grand mal fit and three days later a left sided ventriculo-peritoneal shunt was inserted after which she made a satisfactory recovery. Incidentally, at the time of monitoring the pressure on 14.9.77 she was receiving intrathecal Gentamycin and Cloxacillin and parenteral Gentamycin, Cloxacillin and Septrin.

Cardiac/Respiratory Artefact

CR = 27 mm on stress.

C = 3 mm on stress.

At rest CR = 10 mm. After removal of 10 mls of CSF, CR = 6 mm.

After a further 4 mls of CSF removed, CR = 15 mm.

OFC — 48.5 cms.

Ventricular Dilatation/Cortical Mantle

Marked ventricular dilatation.

Pressure Recordings

Fig. 84 shows the effect of consecutive removal of 10 mls and 4 mls of CSF, respectively, on the level of ventricular pressure. Removal of the CSF resulted in her becoming much less irritable and quieter. The CSF removal was titrated against pressure. Following these two removals she then proceeded to sleep.

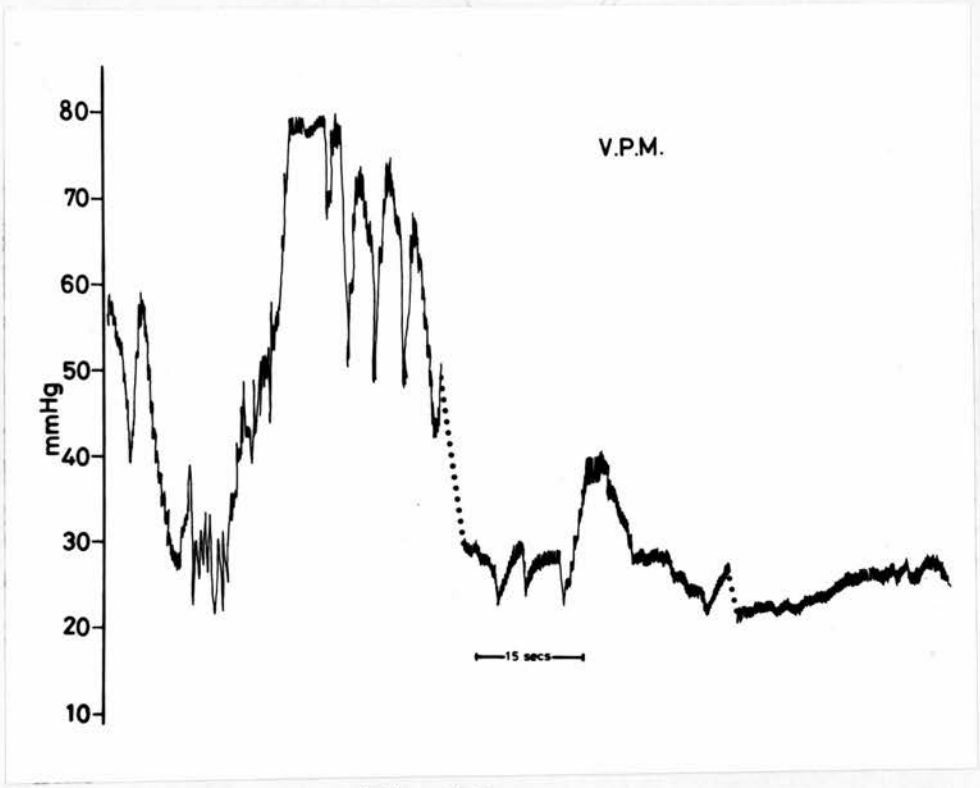


Fig. 84

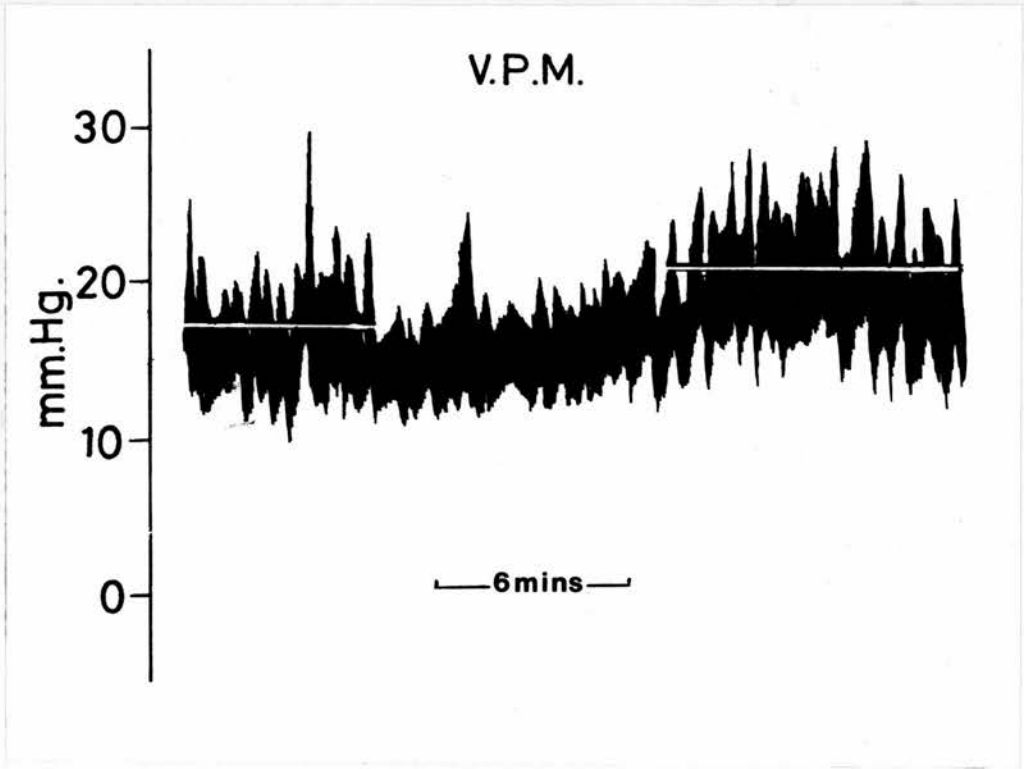


Fig. 85

Case Number 10

Name P. McG.

Age 8 years 10 months

Method Fontanelle cannula into right lateral ventricle via burr hole.

Medical Diagnostic Background

This child was referred at the age of 8 years with spina bifida and hydrocephalus for which he had a ventriculo-atrial shunt in situ. The proximal part of this had been removed in April 1975.

Temperature Normal

Zero Inter-ventricular foramina.

Duration 9 hours

Indication Severe headaches over the last few months, about one or two per week and associated with drowsiness. The parents noted that they seemed to settle with antibiotics on one or two occasions. Headaches occurred at no particular time of the day and they were associated only occasionally with vomiting. He had been having some difficulty in concentrating at school because of the headaches and had recently been intolerant of loud noise. On examination at a peripheral hospital it was felt that he had difficulty with fixation with the left eye and some tortuous vessels about the left disc. At presentation here he had a fairly severe headache in the frontal region (incidentally he had been on long term Septrin for recurrent urinary tract infections associated with his neurogenic bladder). Initial examination showed him to be in no real distress with a left lateral rectus weakness, marked nystagmus, especially on gaze to the right and some terminal nystagmus on gaze to the left. He had no loss of upward conjugate gaze. The indication therefore was chronic complaints of headaches associated with some drowsiness, no abnormality on clinical examination apart from mild eye signs.

Resting Ventricular Pressure

20 mm Hg.

Stress Ventricular Pressure

30 mm Hg peaks on sleep, 51 mm peaks on coughing.

Result Raised intracranial pressure.

Action It was planned to remove his old shunt and replace it with a new one. However he developed a fever of 38 - 38.5° after the procedure but was alert and with no neck stiffness. Throughout the period of minitoring he had been coughing frequently and these deflections on the ventricular pressure tracing give a clue to the aetiology. CSF was microscopically clear and sterile on culture and he was treated symptomatically. Subsequently he produced a classical morbilliform rash. Two weeks later he was afebrile but still having occasional headaches and three days later a ventriculo-peritoneal Pudenz shunt was inserted, the existing ventriculo-atrial shunt left in situ. Post-operatively he developed a staphylococcal ventriculitis with an acute bacterial septicaemia and pulmonary involvement. On examination he was delirious with inappropriate speech and disorientation and since he had the ventriculo-atrial shunt still present and had been on long term Septrin, there was a risk of masking bacterial endocarditis. However there was no splenomegaly or petechiae. He was therefore treated with a combination of Penicillin and Cloxacillin parenterally and intrathecally. Aspirin was also used to lessen the risk of superior vena caval thrombosis. He showed a gradual improvement and on 11.5.76 the remainder of his old shunt system and recent new system were removed and a completely new single system inserted. He was subsequently discharged.

Cardiac/Respiratory Artefact

C = 3 mm awake.

R = 13 mm awake.

CR = 17.5 mm in sleep.

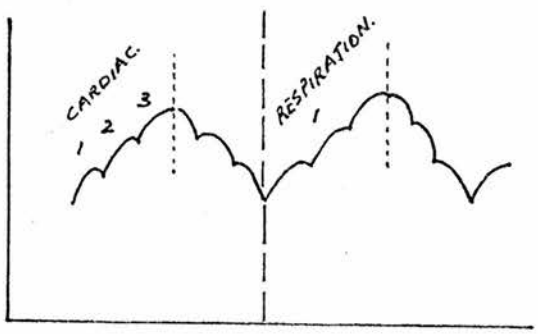
OFC 55 cms

Ventricular Dilatation/Cortical Mantle

Mild ventricular dilatation.

Pressure Recordings The frequent cough responses seen in this recording reached an amplitude of 24 mm Hg and were remarkably constant in amplitude throughout. Fig. 85 shows the degree of elevation of the tracing with sleep which was about 5 mm Hg in this case. At slower recorder speeds the sleep changes in this boy are seen in Fig. 86 and an attempt made to analyse the frequency of the positive pressure deflections (spikes). The timed values in seconds were 13, 21, 15, 10, 13, 27, 12, 8, 17, 17, 15, 19, 12, 10, 10, 15, 12, 6, 12, 9, 7, 9, 10, 14, 18, 27, 10, 10. There is no statistical progression here but an overall pattern of higher values then lower values and back to higher values. It was interesting that snoring begins at the first high value, i.e. 27 seconds. If one excludes the first 7 timed values, the remainder of these features are occurring at 12 seconds intervals, i.e. on an average 5 per minute. In Fig. 86 the arrow indicates movement and the subsequent awake state.

From an expanded fast speed tracing one can estimate the pulse and respiratory rate, e.g. a chart speed of 10 mm per second, the respiratory wave measured 6 mm, i.e. 25 per minute. The superimposed cardiac impulses can be counted a 3 cardiac beats per respiration, i.e. 75 per minute.



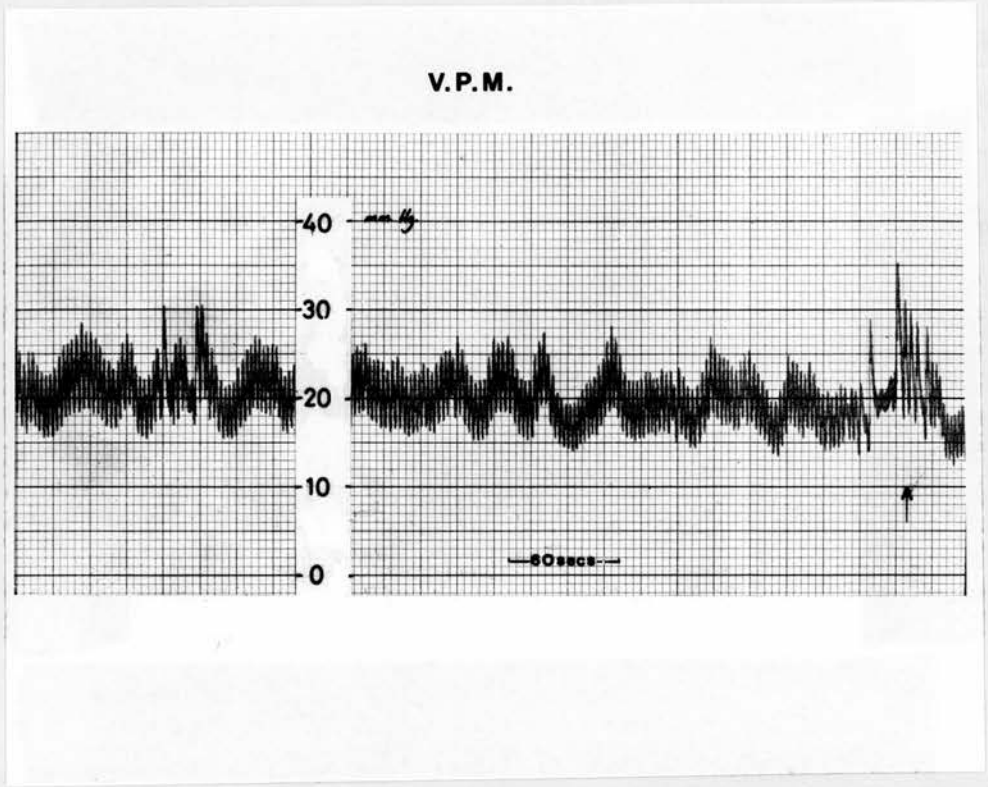


Fig.86

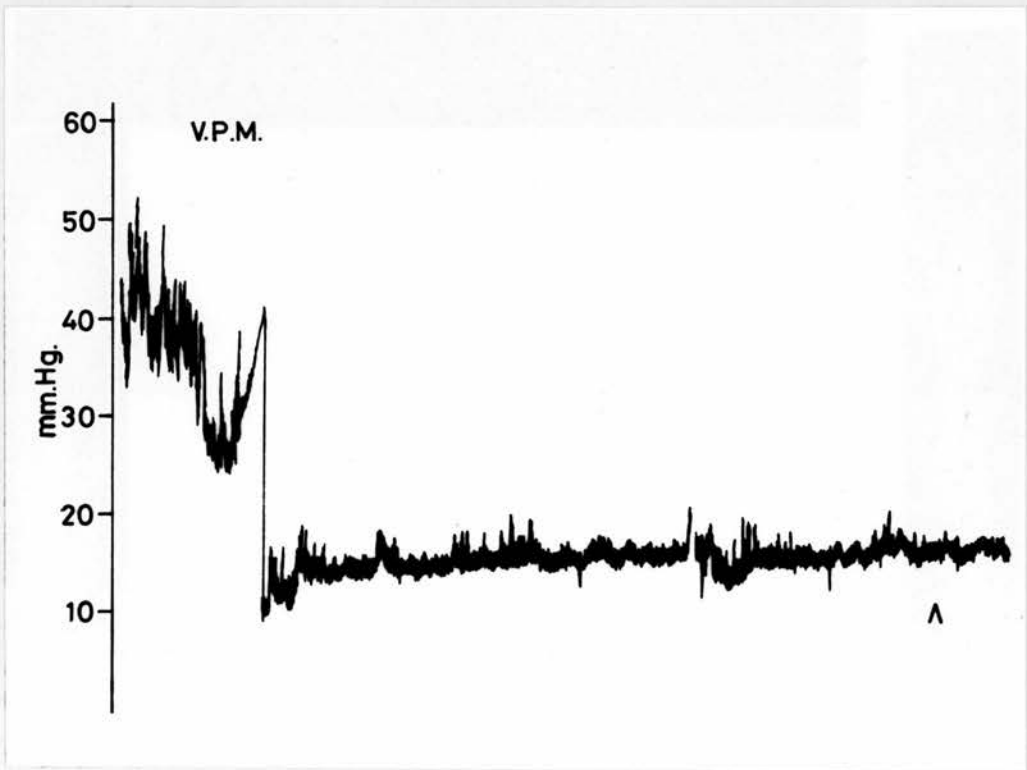


Fig.87

Case Number 11

Name P.McG.

Age 9 years

Method Right post-auricular burr hole.

Medical Diagnostic Background

Myelomeningocoele as before. Following his discharge on 27.5.76 he was readmitted on 29.6.76 with wretching, slurred speech and headache. He was again 'raving' with diminished consciousness but responding to commands. His valve cap was distended but was able to be depressed. These problems had commenced on the morning of admission, he had some slight neck stiffness but was apyrexial. A tap of 20 mls was said to show fluid not under pressure and although there were some red cells there were no white cells or organisms seen. That evening he deteriorated with dilating pupils, left lower motor neurone VII weakness and a marked left VI cranial nerve lesion. He was dysarthric with nasal escape, tachycardia and hypertensive, all suggesting a brain stem abnormality that could have been the result of a 'kinked stem' secondary to pressure. He had no fits and infection was excluded. The following morning he had his ventricular pressure monitored for 5 hours.

Temperature Normal

Zero As above

Duration 5 hours

Indication Evidence of brain stem dysfunction with a shunt in situ and absence of infection. Clinically there was the suspicion of valvular malfunction.

Resting Ventricular Pressure

12.5 mm Hg.

Stress Ventricular Pressure

Pre-tap peaks of 25 mm Hg. Post-tap peaks of 30 mm Hg.

Result Equivocally raised pressure, but on pumping the valve no reduction in ventricular pressure level occurred. Therefore there was not a marked increase in ventricular CSF. pressure, but a malfunctioning valve and evidence of abnormal brain stem function.

Action A valvogram confirmed occlusion at the proximal limb of the shunt with no dye seen in the ventricles. Ventricular taps were continued and eventually open ventricular drainage prior to a further shunt revision was performed. Post-operatively his valve was clinically working, squinting was less and there was now no meningism and he was responding to simple instructions.

Cardiac/Respiratory Artefact

CR = 5 mm pre-tap.

CR = 0.5 mm post-tap.

OFC 55 cms

Points of Interest When he presented with evidence of brain stem dysfunction, bloody CSF. was aspirated via his burr hole and although he may have had some evidence of a 'kinked stem' or abnormal stem vascularity, the possibility exists that an extension of intra-ventricular haemorrhage resulted in some blood clot about his brain stem. Furthermore, this episode of pressure resulted in a tonic, clonic fit lasting approximately 5 minutes with no focal signs for which he was treated with Valium and then Phenytoin prior to his open ventricular drainage.

Pressure Recording At the outset of the monitoring his BP was 120/65 and ventricular pressure about 5 mm Hg, i.e. CPP = 87.5 mm Hg. Later in the record, at BP. 120/80 and ventricular pressure 15, his

CPP. = 85 mm Hg. It can be seen from Fig. 87 a good effect from removing 23 cc of CSF from the 3-way tap while he was asleep; this resulted in him awakening and the pressure can then be seen to be approximately 3 mm Hg. 10 minutes following this CSF removal, at a BP of 114/90 and a ventricular pressure of 6 mm Hg, his CPP = 96 mm Hg. There was a slow build up in the CSF pressure until at the point indicated by the arrow, some 30 minutes after the tap, the ventricular pressure was 7 mm Hg. So, although there was a good effect from CSF removal, no reduction in pressure occurred from pumping the valve.

Case Number	12
Name	P.H.
Age	2 years
Method	Rickham reservoir

Medical Diagnostic Background

Neonatal E. coli meningitis with resultant hydrocephalus, spastic diplegia and hemiparesis and a left parietal Spitz Holter valve attached to a Rickham reservoir. There was associated mental handicap and recurrent seizures with a right frontal focus on the EEG.

Temperature	37.5
Zero	Inter-ventricular foramina level
Duration	Half an hour
Indication	'Not feeling well' and pyrexial for 2

days prior to admission. CSF microscopically clear but an upper respiratory infection diagnosed and she was treated with Ampicillin. On presentation she was drowsy, afebrile, there was a slightly sticky Spitz Holter valve but nil else to find.

Resting Ventricular Pressure

14 mm Hg.

Stress Ventricular Pressure

26.5 mm Hg maximum and deep sleep, 6 mm Hg mean peak, 12 mm Hg in sleep.

Result Raised ventricular pressure.

Action Revision of the proximal end of the shunt at operation which confirmed grossly elevated pressure in the ventricles.

Cardiac/Respiratory Artefact

CR = 15 mm.

OFC 46.75 cms.

Ventricular Dilatation/Cortical Mantle

CT scan reported of huge ventricles with the tip of the valve abutting into brain tissue and a very thin cortex.

Points of Interest There was no doubt that her resting ventricular pressure was raised, however it was not as elevated as might have been expected considering the operative findings and one has therefore to question the competence here of the proximal end of the shunting system, i.e. the Rickham reservoir. The other factor which questions this is the considerably widened pulse artefact in deep sleep at a low pressure level, and I suspect that the mean level of pressure should have been higher, although the cardio-respiratory impulses were coming through (refer Chapter 2) (Fig. 88). This is supported by the CT. scan which showed the tip of the valve abutting into the brain tissue.

Case Number 13

Name L. McR.

Age 3 years

Method Left frontal Rickham reservoir.

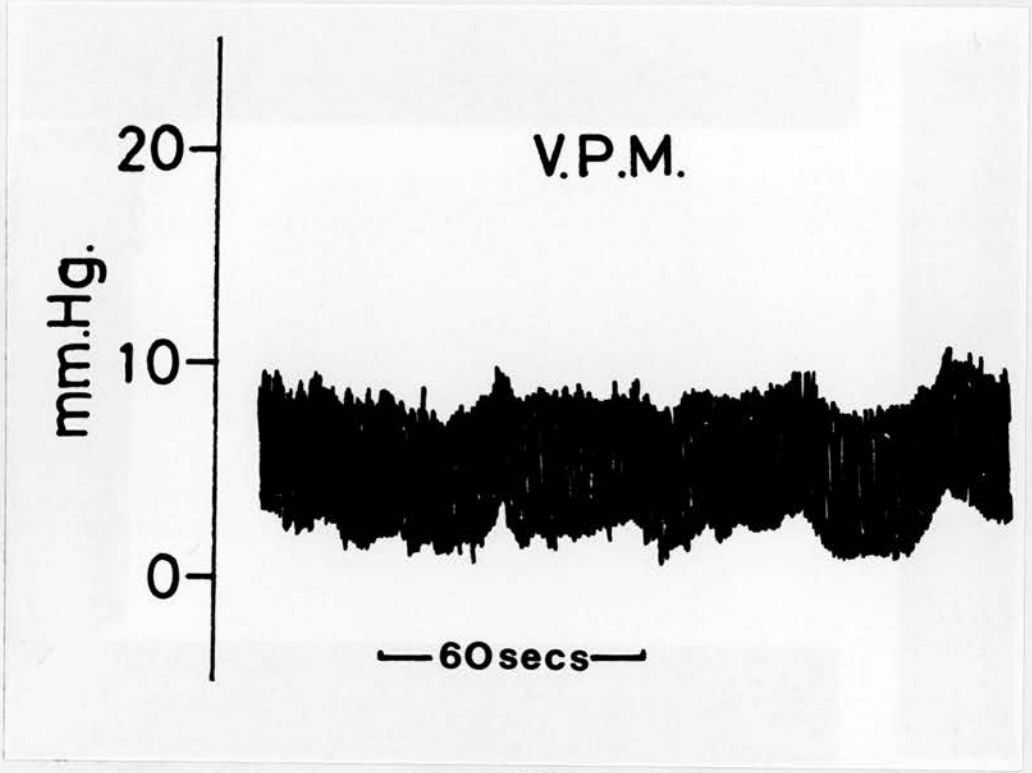


Fig. 88

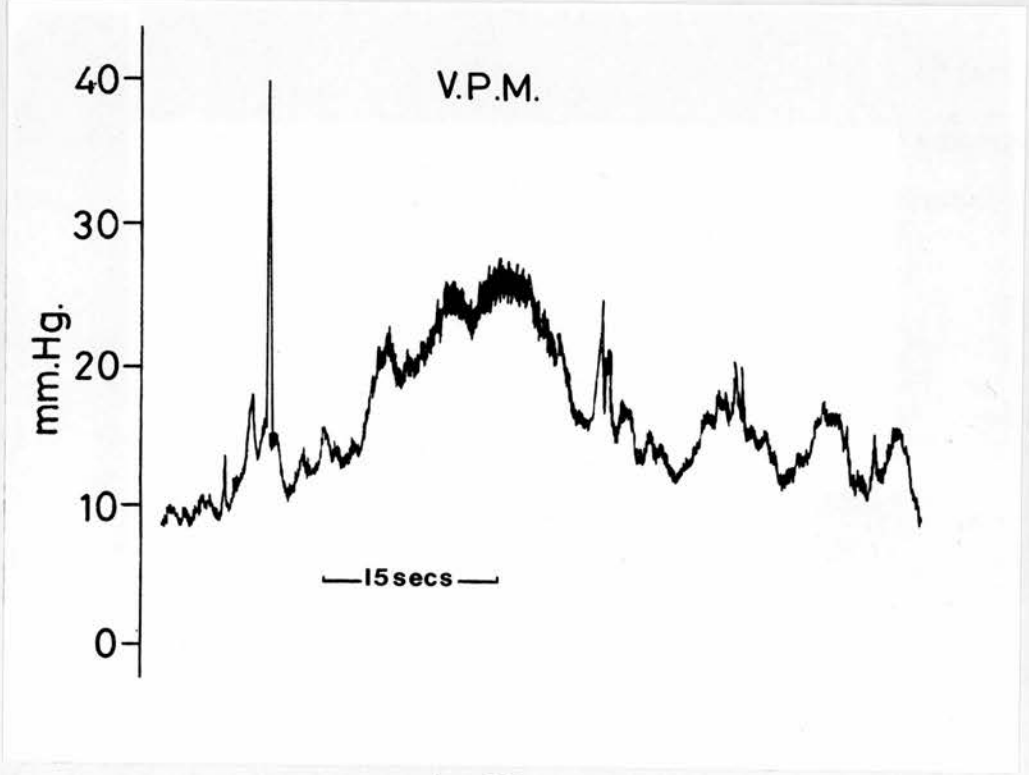


Fig. 89

Medical Diagnostic Background

Post-neonatal meningitis with residual hydrocephalus, a shunt inserted at $2\frac{1}{2}$ months of age and subsequently two revisions.

Temperature 37.7

Zero Upper cortical subarachnoid space.

Duration Half an hour

Indication The child had an ear infection 4 days prior to admission. On admission she was anorexic, irritable, she had a red fluctuant swelling over the anterior chest wall but no other clinical signs apart from irritability and her shunt seemed clinically to work. A tap of CSF showed 37 white cells. 6 hours later she had a convulsion; this may have been due to pressure or infection. She was therefore treated with intrathecal and intramuscular Gentamycin and Cloxacillin and the inflammation over her anterior chest wall subsided until 24.7.77 when she showed signs of raised intracranial pressure with vomiting, irritability, bradycardia, neck stiffness, lethargy and a weak cry. Therefore the indication was a blocked infected shunt.

Resting Ventricular Pressure

In excess of 50 mm Hg.

Stress Ventricular Pressure

Peaks to 70 mm Hg with a mean stress of 45 mm Hg.

Result Raised CSF ventricular pressure

Action She therefore had frequent taps of CSF until her shunt was removed two days later. Post-operatively she again came under raised intracranial pressure and needed open ventricular drainage and further revision two weeks later.

Cardiac/Respiratory Artefact

CR = 12.5 at 5 mm Hg.

CR = 42.5 at 14 mm Hg.

Pressure Recordings Although no resting baseline pressure was obtained initially because of persistent crying, 5 mls of CSF removal reduced the ventricular pressure to 6 mm Hg. This then gradually increased to 15 mm Hg when a further few mls of CSF were removed. That such a small quantity of CSF should result in relief of her symptoms questions whether she has very poor compliance or whether there was an element of brain swelling. However, no CT scan was obtained on this occasion.

Case Number 14

Name W.D.

Age 8 years

Method Left sided Pudenz flushing chamber.

Medical Diagnostic Background

Myelomeningocoele.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration Half an hour

Indication He presented with papilloedema, lateral nystagmus, a clinical distal block, intermittent vomiting and neck stiffness for 2 weeks. A valvogram showed an intermittent distal block and the proximal catheter had crossed the midline. CSF was clear and there was a slight swelling about the cap of the valve. The present shunt had been in situ for 3 years. So the indication was one of a symptomatic child with evidence of valve malfunction clinically.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

26 mm Hg peaks.

Cerebral Perfusion Pressure 80 mm Hg.

Result Raised ventricular pressure.

Action Shunt revision. At operation a partial blockage was found of the proximal catheter, it was blocked with fibrinous material. The distal limb appeared patent but the shunt was completely replaced.

Cardiac/Respiratory Artefact

7.5 mm at 15 mm Hg.

Ventricular Dilatation/Cortical Mantle

No recent estimate was available. However an air encephalogram in the first year of life showed moderate dilatation of the lateral and third ventricles. The cortical mantle was 2.5 cm at the vertex.

Case Number 15

Name I.F.

Age 2 weeks

Method Ventricular cannulation through anterior fontanelle.

Medical Diagnostic Background

Lumbar meningocoele.

Temperature 37.2

Zero $1\frac{1}{2}$" below upper cortical subarachnoid space.

Duration 1 hour

Indication The assessment of active neonatal hydrocephalus as a result of a spina bifida lesion. The pressure monitoring was done prior to a pneumo-ventriculogram. (There was an increasing OFC, scalp venous distension and tension of the anterior fontanelle.)

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

42 mm Hg maximum.

Result — Raised ventricular pressure

Action — Daily ventricular taps prior to a ventriculo-peritoneal shunt insertion.

Cardiac/Respiratory Artefact

CR = 17.5 mm on stress.

CR = 2.5 mm at rest.

OFC — 35 cms at birth. 38.5 cms at the time of VPM.

Ventricular Dilatation/Cortical Mantle

Moderately large ventricular distension with a cortical mantle of 16 mm at the vertex.

Case Number — 16

Name — M.R.

Age — 5 years

Method — Left frontal Ommaya reservoir

Medical Diagnostic Background

This child was born by caesarean section at 38 weeks gestation. BW. was 3.85 Kg and mother was a 32 year old para II. Presentation had been by the breech and the child's OFC at birth was 37 cms. Initial examination appeared otherwise normal. By 12 days of age the OFC was 38.5 cms and he was feeding well. He was next seen at the Child Welfare Clinic at age 9 weeks when his mother commented that he tended to 'roll his eyes downwards' and had done this for the previous 3 weeks. He was also jittery to

loud noises and his left leg was said to 'twitch' occasionally. On this examination it was noted that he was grossly hydrocephalic with an OFC of 48.5 cms, the sutures were widely splayed, his fontanelle was tense and there were dilated scalp vessels. He was 'sunsetting', he had brisk tendon reflexes and a generalised increase in tone. He had no papilloedema but he did have nystagmus. A skull x-ray at this time showed a large posterior fossa and an increase in the AP diameter. Other routine investigations were normal. A lumbar air encephalogram showed a gross degree of communicating hydrocephalus and subsequently a Pudenz ventriculo-peritoneal shunt was inserted. At this operation a preliminary ventriculoscopy was carried out to exclude a choroid plexus hypersecreting adenoma. Following this he was transferred to the care of a physician in Luxemburg. Two subsequent shunt revisions were done.

He was next seen at nearly 5 years of age when he was admitted for further assessment of his hydrocephalus.

Temperature 36.75

Zero The level of the anterior horn of the lateral ventricle.

Duration 16 hours

Indication All had been reasonably well since the previous revision, except that over the last 3 months he had complained of intermittent headache and episodes of vomiting. On several days he went through the same sequence of events. On getting up in the morning he would be well but during the day would lose his appetite and by the evening complain of headache and then vomit. There were 2 episodes of prolonged headaches lasting several days. There was no associated disturbance of affect and no diminution of neurological function. Recent developmental assessments had shown that he was an active child, attending play groups and infant school.

On examination at this time his Pudenz flushing device was slow to fill, but there were no deficits in his cranial nerve function and in particular, no evidence of papilloedema or neck stiffness. Accordingly a CT. scan was carried out which demonstrated a large left sided, well circumscribed lesion which probably represented a porencephalic cyst in relationship to the roof of the left lateral ventricle. This appeared to distort the trigone and occipital area of the left lateral ventricular system, but did not cross the midline. Under the same anaesthetic, a left frontal burr hole was made, ventriculoscopy undertaken and insertion of an Ommaya reservoir. No gross abnormality was seen on ventriculoscopy. The catheter was therefore placed in the cystic cavity and he was transferred for ventricular pressure recordings. The indication therefore was of a symptomatic child with vomiting and headaches, no clinical signs but a known congenital hydrocephalus with a shunt in situ, who had a porencephalic cyst related to the roof of the left lateral ventricle. The pressure monitoring was to find out if he needed modification of his existing shunt, or insertion of a shunt system into the cyst itself.

Resting Ventricular Pressure

3 mm Hg.

Stress Ventricular Pressure

A maximum peak of 53 mm Hg on crying, 57 mm Hg on coughing and maximum sleep peaks to 28 mm Hg.

Result Normal pressure within the cystic cavity, therefore no cysto-peritoneal anastomosis or further left sided ventriculo-peritoneal anastomosis needed to be undertaken. The suspicion that this was not freely communicating with the rest of the ventricular system and that the cyst was under tension, having been a shift of the midline structures to the right, was

incorrect. No further operative intervention was undertaken.

Cardiac/Respiratory Artefact

C = 1.25 mm at rest. R = 3.75 mm at rest.

Ventricular Dilatation/Cortical Mantle

The right lateral ventricle was normal in size and thus the right sided shunt assumed to be working effectively.

Pressure Recordings

Fig. 89 shows a section of the ventricular pressure tracing when the child complained of a headache and at this time the pressure transiently rose, with a corresponding increase in the cardio-respiratory artefact. The nature of this increase is unclear, but almost certainly pathological. Depression of the valve in situ caused no reduction in the ventricular pressure, however, the resting level was only 3 mm Hg. It might have been considered that depression of the valve would result in negative recorded pressures. However this did not happen.

Case Number	17
Name	M.R.
Age	5 years
Method	A twin recording simultaneously from Ommaya reservoir and Pudenz flushing device on the opposite side.

Medical Diagnostic Background

As above.

Temperature	Normal
Zero	Zero reference points differed by no more than 2".
Duration	3 hours
Indication	Because of the asymmetry of the ventricular

system and the porencephalic system on one side, and no real depression of the measured ventricular pressure level in the cystic cavity on pumping the valve, a simultaneous bilateral recording was undertaken.

Resting Ventricular Pressure

9.5 mm Hg from the Ommaya reservoir in the cystic cavity, and 11.5 mm Hg from the Pudenz flushing device in the normal sized right ventricle.

Stress Ventricular Pressure

48 mm Hg maximum stress. The two sides were very comparable.

Result Normal pressures from both sides with only a minimal difference between the two. There was however a slight damping of the tracing from the Ommaya reservoir (Fig. 90).

Action Nil

Cardiac/Respiratory Artefact

From Pudenz - C = 5 mm, R = 7.5 mm.

From Ommaya - C = 2.5 mm, R = 6.25 mm.

Pressure Recordings

The Fig. 90 shows 'stress deflections' initially which are associated with the child's using a bedpan.

Case Number 18

Name C.S.

Age 12 years

Method Right frontal Rickham reservoir

Medical Diagnostic Background

This boy was very severely mentally retarded with grand mal epilepsy since the age of 5 months. At the age of 5 years he had a theco-peritoneal shunt inserted for a communicating hydrocephalus diagnosed by a lumbar air encephalogram. In the past his anti-

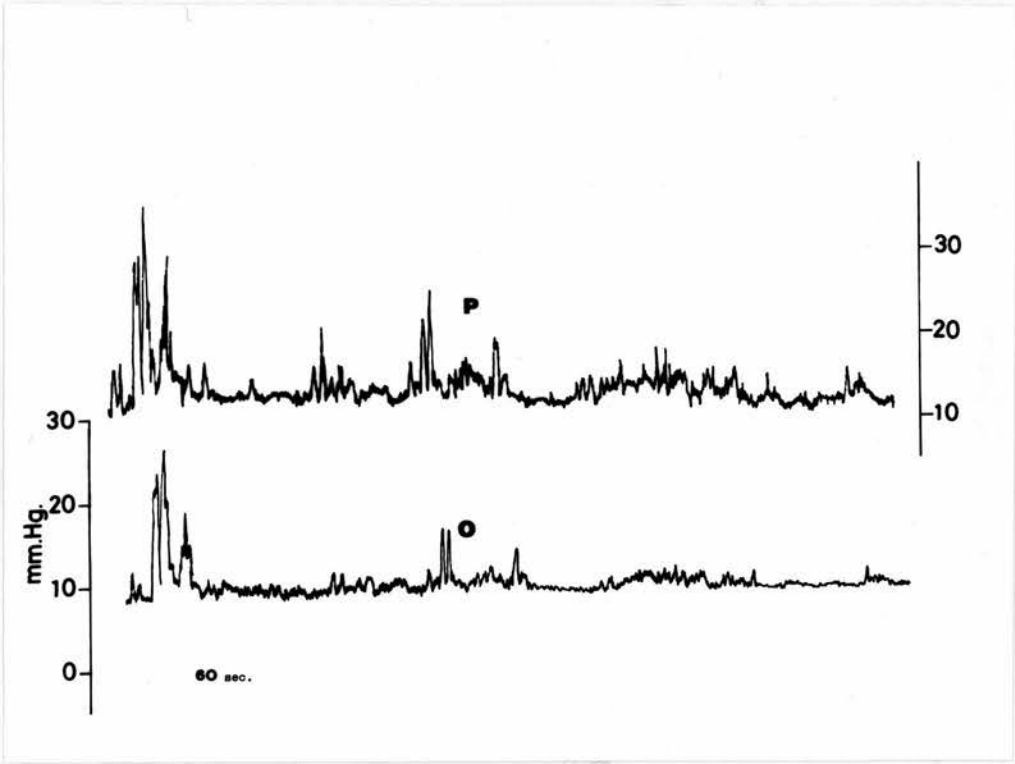


Fig.90

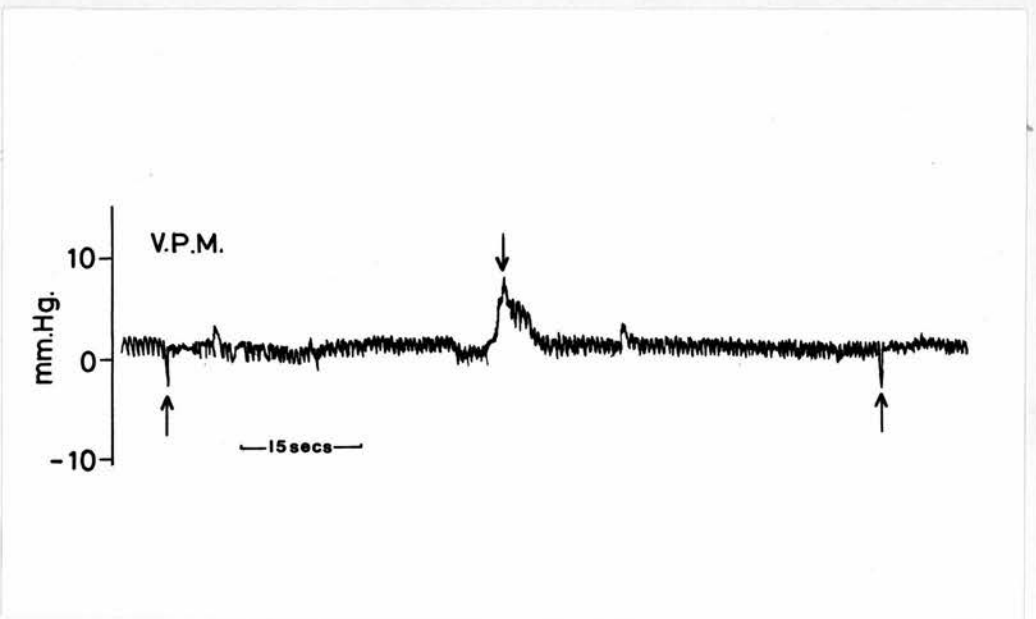


Fig. 91

epileptic regime has consisted of Phenobarbitone, Phenytoin, Ospolot, Diazepam, Primodone and Epilim. The parents noticed a recent deterioration 18 months prior to the monitoring following a head injury at school, and over the last 3 months he had severe uncontrolled epilepsy. His gait deteriorated, he began to stagger and fall. He was admitted on this occasion with status epilepticus and resistant to benzodiazepines. He was treated with intravenous Pentothal and Paraldehyde (IM. and rectally).

Temperature 36.5

Zero Upper cortical subarachnoid space

Duration One hour

Indication An elective Rickham reservoir was inserted, so that pressure monitoring could be carried out to determine if his recent neurological deterioration was due to raised intracranial pressure. Secondly, CSF was needed at the time to exclude infection.

Resting Pressure 3 mm Hg

Stress Ventricular Pressure

18 mm Hg maximum on head movements.

Result Normal pressure

Action Nil with regard to intracranial pressure.

Cardiac/Respiratory Artefact

CR = 10 mm at rest.

OFC 58.5 cms at the time of monitoring.

Pressure Recordings Throughout the recording, an example of which is shown in Fig. 91 , he had numerous petit mal fits lasting about 8 seconds. They consisted mostly of minimal eye deviation, and in between them he would speak. There are frequent yawns also as shown by the negative pressure deflections, indicated by the arrows, and this

is no doubt an epileptic manifestation. Sometimes the yawns are immediately pre-fit and sometimes they are immediately at the conclusion of a fit. On occasions these fits were associated with very slight hand movements, and minimal retraction of the neck. However, mostly they were true petit mal fits.

Case Number	19
Name	J.W.
Age	$2\frac{1}{2}$ months
Method	Fontanelle cannulation

Medical Diagnostic Background

Prematurity, BW. 1.474 Kg with an intra-ventricular haemorrhage and septicaemia, urinary tract infection and subarachnoid haemorrhage.

Temperature	Normal
Zero	Inter-ventricular foramina level.
Duration	Half an hour
Indication	The assessment of active neonatal hydrocephalus (there was a rapid increase in OFC over a period of days and 'sunsetting').

Resting Ventricular Pressure

14 mm Hg.

Stress Ventricular Pressure

46 mm Hg maximum.

Result	Raised ventricular pressure
Action	Immediate action, 'tapping' CSF.

Pre-tap level 14 mm Hg. After 5 mls CSF removed, ventricular pressure = 12.5 mm Hg. After a further 25 mls CSF removed, ventricular pressure 9 mm Hg. She then had a Rickham

ventriculostomy reservoir inserted so that tapping could be instituted twice daily.

Cardiac/Respiratory Artefact

CR = 12.5 mm pre-tap, CR = 7.5 mm post-tap.

Ventricular Dilatation/Cortical Mantle

Lumbar air encephalogram the day after VPM. showed an external hydrocephalus on the right and a free, communicating hydrocephalus.

Case Number	20
Name	J.W.
Age	11 weeks
Method	Anterior placed Rickham reservoir

Medical Diagnostic Background

Prematurity, intra-ventricular haemorrhage, etc. (as before).

Temperature	Normal
Zero	Upper cortical subarachnoid space.
Duration	Half an hour

Indication — She had been having daily taps since her Rickham was inserted and this recording was on the occasion of one of these taps. It had been 24 hours since she was previously tapped of CSF. The anterior fontanelle was not bulging.

Resting Ventricular Pressure

10.5 mm Hg pre-tap, 4.5 mm Hg post-tap.

Stress Ventricular Pressure

47 mm Hg maximum.

Result — This is a borderline ventricular pressure level, however she was becoming symptomatic every 24 hours, requiring CSF release.

Action — A ventriculo-peritoneal shunt was

inserted 2 weeks later, since when she has remained very well.

OFC 42 cms 5 days after this monitoring.

Points of Interest In retrospect, we possibly should have continued with daily tapping for a longer period, hoping for spontaneous arrest of her hydrocephalus.

Case Number 21

Name M.W.

Age 4 weeks

Method Ventricular cannulation through anterior fontanelle.

Medical Diagnostic Background

She was born with an occipital meningo-encephalocoele with a large arterio-venous malformation which was evident when the encephalocoele was removed surgically.

Temperature 37.2

Zero Inter-ventricular foramina level

Duration Half an hour

Indication The assessment of active neonatal hydrocephalus (distension of her anterior fontanelle, superficial scalp veins and OFC).

Resting Ventricular Pressure

19 mm Hg.

Stress Ventricular Pressure

56 mm Hg maximum.

Result Raised ventricular pressure.

Action Ventricular taps of CSF followed by a ventriculo-peritoneal shunt.

Cardiac/Respiratory Artefact

Very narrow immeasurable amplitude.

OFC 32 cms at birth, 37.7 cms at time of VPM.

Ventricular Dilatation/Cortical Mantle

Ventricular air encephalogram 2 days prior to VPM showed gross distension of the lateral and third ventricles, but not the fourth ventricle. Therefore, a non-communicating hydrocephalus with a cortical mantle of 18 mm.

Follow Up She has done remarkably well developmentally and visually without any seizures. She has required one shunt revision in her first 12 months.

Case Number 22

Name J.P.

Age 5 years 11 months

Method Right frontal Rickham

Medical Diagnostic Background

Myelomeningocoele with a right sided ventriculo-atrial shunt in situ when she first presented. It was revised once to a right sided ventriculo-peritoneal shunt.

Temperature Normal

Zero 2" above cortical subarachnoid space.

Duration Half an hour

Indication On presentation she had vague symptoms of intermittent vomiting and fundal changes on examination with some blurring and loss of the physiological cupping, She subsequently required frequent CSF removals from her Rickham reservoir and on this occasion she was grossly sweaty with grade 4 fundal changes

and had been experiencing daily headaches. At the time of presentation she had no Rickham reservoir in situ but a Spitz Holter valve, therefore she electively had a Rickham ventriculostomy reservoir inserted followed by pressure monitoring.

Resting Ventricular Pressure — 60 mm Hg pre-tap

Stress Ventricular Pressure — 60 mm Hg pre-tap

Result — Markedly elevated ventricular pressure.

Action — Revision of distal end of ventriculo-peritoneal shunt. Subsequently she required a further distal revision 2 days later as the shunt still continued to work unsatisfactorily.

Cardiac/Respiratory Artefact

C = 2.5 R = 5 at 5 mm Hg.

C = 12.5 R = 22.5 at 20 mm Hg post-tap.

Ventricular Dilatation/Cortical Mantle

No estimate of this on this occasion.

Points of Interest — This child responds to raised ventricular pressure with gross sweating on each occasion, she also has dilated pupils which are poorly responsive to light.
High excursions of pressure initially required 15-20 mls of CSF. to be released immediately. However, she remained grossly sweaty with normal BP. and a pulse of 96/minute. She was fully conscious. Shortly after she required a further 10 mls CSF.to be released then the ventricular pressure began to drop to acceptable limits and gradually reached 5 mm Hg.

Case Number — 23

Name — K.L.

Age — 8 months

Method — Right frontal Rickham reservoir

Medical Diagnostic Background

She was a first born child to parents, father 35, mother 28, after an uneventful pregnancy. BW. 7lbs 5 oz at 10 days after term. Labour was induced by ARM. and a Syntocinin drip. It lasted 5 hours and delivery was by the vertex. She was discharged on the eighth day and re-admitted shortly after with a right parotid swelling. At that time she was noted to have some unusual features with micronathia, coarse facies, slightly high arched palate, a large head, large hands and feet and widely spaced nipples. She was rather 'pot bellied', a little hypotonic but with peripheral reflexes. Chromosome estimation showed a pericentric inversion in number 9 chromosome. When this investigation was performed on mother, it was found she had the same inversion of number 9 chromosome but normal facies, intelligence etc. Father's chromosomes were a normal 46 XY karyotype. She was treated with Cloxacillin and Ampicillin for a staph.pyogenes abscess over the right parotid and sub-mandibular region. A TSH. result confirmed a minor degree of hypothyroidism and her height, weight and OFC were between the 30th and 50th percentiles. Bone age was also retarded and TSH was shown to rise from 5.38 basal to a maximum of 34.3, 40 minutes after an injection of thyrotropic releasing factor (TRF). There was also some evidence of psychomotor delay and the opinion was that she had a degree of compensated hypothyroidism and should be supplemented with Thyroxine in view of the clinical features. At 6 months of age her OFC was noted to be rising, perhaps too rapidly, it was then 46 cms, well above the 90th percentile. A hemi-syndrome became evident at 7 months of age and x-ray of the skull showed splaying of the sutures which was detectable clinically. A CT. scan at 8 months of age showed some moderately severe dilatation of both lateral ventricles, the right lateral ventricle somewhat

larger than the left and minimal displacement of the ventricular system to the right. The third ventricle was not grossly enlarged but probably a little more so than normal. It was not displaced, the fourth ventricle was not too clearly identified but was seen to be small. No parenchymal brain lesion could be distinguished. The basal cisternal region was satisfactorily displayed and there was no evidence of any neoplasm within the basal cysterns. The Sylvian fissure sulci were quite large as were the sulci in the inter-frontal regions. The appearance indicated some generalised atrophic condition of the brain with some focal features referring to the right side.

Numerous other investigations including cortisols, calcium phosphate, alkaline phosphatase, metabolic screening, amino acids, mucopoly-saccharides and gangliosides were all negative. A growth hormone study showed abnormal values with a partial deficiency which was an odd result in view of her exaggerated growth. So she presented as a very complex picture and appeared to be developing cerebral gigantism, secondary hypothalamic dysfunction with an abnormal growth hormone response. She also had cerebral palsy with left hemiplegia and evidence of definite brain damage with hydrocephalus and cerebral atrophy. It was thought that this dilatation of the ventricles, atrophy and the asymmetry may be related to some birth problems and with some bleeding into the subarachnoid space rather than a chromosomal disorder. Because of the hydrocephalus and increase in head size, an elective Rickham reservoir was inserted both to monitor her pressure and also as an entry to her ventricular system if acute pressure problems developed.

Temperature	Normal
Zero	Anterior horn of right lateral ventricle.

Duration 3 hours

Indication Inversion of number 9 chromosome, odd facies, secondary hypothyroidism and evidence of hydrocephalus and brain damage on CT. scan with rapid head growth.

Resting Ventricular Pressure

7.5 mm Hg.

Stress Ventricular Pressure

37 mm Hg maximum. 21 mm Hg peaked in sleep.

Result Probable normal ventricular pressure.

Action For serial OFC measurements, and it was decided if she became symptomatic with upper respiratory infections etc. to have a further period of ventricular pressure monitoring at that time.

Cardiac/Respiratory Artefact

CR = 15 mm at rest. CR = 20 mm maximum in sleep.

OFC 46.8 cms.

Ventricular Dilatation/Cortical Mantle

Scan result as above.

Follow Up By one year of age her OFC had levelled off and instead of crossing all the percentiles they were now beginning to follow the normal rate of growth, and there had been no further evidence of raised intracranial pressure. At 19 months of age she was reported to be sleeping poorly, irritable and the question of raised pressure was again raised. She was admitted for a further period of monitoring which showed similar results to those reported previously. Her Thyroxine was subsequently ceased at 22 months of age and since then her TSH. level has remained at the upper border of normal and she has remained well.

Repeated growth hormone estimations show that there was a suppression of growth hormone secretion to a level below 2 with glucose and this is not suggestive of 'gigantism'. An LH level was normal at 1.2 u/l but an FSH was elevated which is usually an indicator of ovarian dysgenesis, but also possibly of pituitary or hypothalamic hyposecretion. She has required follow up care for her hemi-syndrome.

A few drops of CSF were initially lost at commencement of this recording and this often happens. Upward and downward deviation of the eyes voluntarily makes no difference to the level of ventricular pressure. It is known that thyroid malfunction, hypoparathyroidism, Tay-Sachs disease, Maple syrup urine disease etc. can result in a degree of megalencephaly and may produce transient signs of raised intracranial pressure, in my opinion. Although this child has definite evidence of a hydrocephalus and brain damage, there was also an abnormality of her endocrine system. A recent description (Addy 1977) of a child with congenital hypothyroidism of pituitary origin, who was large from birth, was associated with no abnormal neurological findings and had a normal CT scan.

Case Number	24
Name	K.L.
Age	19 months
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

As before.

Temperature	Normal
Zero	Inter-ventricular foramina level.

Duration 3 hours

Indication Irritability, poor sleeping with known hydrocephalus, not shunted and an estimate of sleep pressure was necessary.

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

In early sleep she peaked to a maximum of 23 mm Hg, for a very short time.

Result Although the sleep changes are probably not normal, it was not considered advisable to shunt her at this time.

Cardiac/Respiratory Artefact

CR = 4.5 mm at rest. CR = 8 mm in early sleep. CR = 5 mm in deep sleep.

Points of Interest This child obviously will be followed up with serial CT scans to ensure that if insidious ventricular dilatation is occurring during sleep, before neurological deterioration is evident, an estimate of changing cortical width can be obtained.

Case Number 25

Name R.G.

Age $7\frac{1}{2}$ months

Method Ventricular catheters through burr hole.

Medical Diagnostic Background

She was born with a partial sacral agenesis, a lumbar myelomeningocoele with motor and sensory activity to S_4 on the right and L_5 on the left with a 'mixed reflex' pattern. Closure of the myelomeningocoele was undertaken within the first 24 hours. Her

OFC on admission was 37.5 cms and her anterior fontanelle was tense. An air encephalogram at 3 weeks of age showed a communicating hydrocephalus with a large ventricular system and herniation of the cerebellar tonsils. At $3\frac{1}{2}$ weeks of age the posterior fossa was decompressed and a temporary ventricular drain inserted. Two days later a ventriculo-peritoneal system was inserted. She had a further two revisions subsequently.

Temperature — Normal

Zero — Inter-ventricular foramina

Duration — 4 hours

Indication — Irritability of 24 hours standing.

Scalp vein distension. No vomiting and a rapid increase in OFC which was now 52.5 cms. The anterior fontanelle was full and tense, sutures were separated and there was some blurring of her disc margins with slight loss of conjugate upward eye movement.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

53 mm Hg maximum and 16 mm Hg in sleep.

Action — At operation the shunt was found to be working normally, and it was thought therefore that she had either intermittent obstruction, possibly by the tip of the ventricular catheter abutting into the ventricular wall. This causes the pressure to increase and the ventricles to distend, freeing the catheter tip until adequate CSF shunting again collapses the ventricles etc. Accordingly a shorted ventricular drain and lower pressure device was inserted. It then functioned satisfactorily.

Cardiac/Respiratory Artefact

CR = 10 mm at 10 mm Hg.

OFC 52.5 cms

Ventricular Dilatation/Cortical Mantle

Gross symmetrical ventricular dilatation with cerebellar herniation on air encephalogram 5 months later with a cortical mantle of 5 mm at the vertex.

Points of Interest The pressure prior to CSF relief in this patient was 30-40 mm Hg at times peaking to 50 mm Hg with a BP at the time of 88/55, that is CPP = 36.5 mm Hg.

Pressure waves in sleep in this child are not dramatically high after chloral although the resting level of ventricular pressure is elevated. There was a drop in pressure of about 5 mm Hg after chloral. The duration of REM sleep waves after chloral in this tracing was about $8\frac{1}{3}$ minutes and they began $7\frac{2}{3}$ minutes after the chloral was administered.

Case Number 26

Name C.J.

Age 2 weeks (38 weeks gestation by dates)

Method Ventricular cannulation through anterior fontanelle.

Medical Diagnostic Background

The child was born with a sacral myelomeningocoele with a neurological level of S_1 motor and S_2 sensory. Born at 36 weeks gestation with neonatal apnoea and required endotracheal intubation and a 'head box'. Operation was performed at 24 hours of age and the child developed a post-operative ventriculitis. Antibiotics were ceased one day after the pressure monitoring.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 1 hour

Indication The assessment of active neonatal hydrocephalus (a rapidly increasing OFC, full anterior fontanelle with sutural separation, early 'sunsetting' and lethargic to feed).

Resting Ventricular Pressure

10 mm Hg.

Stress Ventricular Pressure

27 mm Hg maximum.

Result Normal level of ventricular pressure.

Action Conservative management with serial OFC measurements and at follow up the OFC was on the 75th percentile and then paralleled the percentiles, with no further evidence of raised intracranial pressure.

Cardiac/Respiratory Artefact

CR = 2.5 mm at rest, CR = 5 mm in sleep.

Ventricular Dilatation/Cortical Mantle

Air ventriculogram showed moderately severe ventricular dilatation with a cortical mantle of 20.5 mm at the vertex.

Points of Interest This child with a reasonable cortical mantle, and ventricular pressure levels in the normal range, was therefore spared a CSF shunting device. The pattern of expansion of the head in this child follows a not unusual one of compensation-decompensation-compensation etc. That is a step-wise increase in OFC with time. Over the first 63 days (i.e. 9 weeks) the OFC increased by 7.1 cms from an initial OFC of 32.5 cm. An average daily increase of 0.112 cm. The normal increase should be about 0.5 cm per week (i.e. 0.071 cm per day), or 4.5 cm over this period of 9 weeks. Therefore, the excessive growth of the head above normal limits in this child over the first 9 weeks of life amounted to 2.6 cm.

If the first 23 days are examined, the OFC increased by 3 cm (i.e. 1.3 cm above normal) and the child showed distended scalp veins and lost interest in feeding. For the next 20 days the OFC increased by only 1.9 cm (i.e. only 0.48 cm above normal) and during this time there was improvement in the feeding and subsidence of the scalp vein distension. For the last 20 days of frequent OFC measurements the OFC increased some 2.2 cm and again the scalp veins became prominent (i.e. 0.78 cm above normal rate). So although the values are not extreme, it does illustrate the change in the day to day accommodation and lack of accommodation of the newborn to raised intracranial pressure.

Pressure Recordings A 'saw tooth' pattern of ventricular pressure occurred during sleep in this child and it is significant that he has recently had a CSF infection.

Case Number	27
Name	G.F.
Age	19½ months
Method	Short Huber needle into right frontal Rickham reservoir.

Medical Diagnostic Background

He was the result of a second pregnancy of a 28 year old mother, the first child, the result of a previous marriage, was normal. Pregnancy was uneventful apart from mild eclamptic toxaemia towards term. He was induced at 39 weeks and delivered by S.V.D., after a 3 hour labour and he did not require resuscitation. BW was 3.48 Kg. There was no family history of spina bifida. On examination there was a lumbo-sacral myelomeningocoele extending from L_4 to S_3. His OFC at the time was 33.9 cms and the anterior fontanelle measured 2 x 2 cms. There was good voluntary motor power down to L_4 bilaterally with some

evidence of reflex activity in the S_{2-5} segments. Within the first 24 hours of life the myelomeningocoele was excised and post-operatively he made good progress apart from some generalised twitching associated with hypocalcaemia. 4 days post-operatively he developed a wound infection which resulted in a ventriculitis. By the age of 16 days the OFC was increasing at a rate only marginally above normal. The anterior fontanelle were slightly full and there was some scalp vein distension. Primitive pressure recording at that time was said to show no gross increase in pressure and an air ventriculogram showed a normal ventricular system and a good cerebral mantle with only minimal dilatation of the temporal horns of the lateral ventricles. At 1 month of age he was discharged and at this stage his OFC was 37.4 cms.

At 17 months of age his OFC was increasing at a slightly exaggerated rate and it was decided to electively insert a Rickham reservoir so that he could have pressure studies done. His OFC at this time was 55 cms, having been 34.1 cms at birth. It appeared to have crossed the 90th percentile at about 6 weeks of age, but fairly slowly. Now there was some distension of his scalp veins and his sutures appeared fused. He had a sixth nerve palsy. 19 months seems to be a critical time from the point of view of pressure symptoms occurring because up until now the OFC had been increasing but there had been no other signs. He now, however, had some distension of the veins and was squinting.

Temperature	Normal
Zero	Upper cortical subarachnoid space
Duration	2 hours
Indication	Accelerating OFC and other minor signs.

Resting Ventricular Pressure

9 mm Hg.

Stress Ventricular Pressure

70 mm Hg maximum peak awake, 19 mm Hg peaks in sleep.

Result Equivocal range ventricular pressure

Action No CSF shunt was inserted and he was then seen weekly for review with OFC measurements.

Follow Up Up to 2 months later his OFC was still 55 cms, i.e. there was no increase. 5 months later it had increased o.4 cms and 14 months later it had increased to 55.5 cms.

Cardiac/Respiratory Artefact

C = 5 mm. R = 7.5 mm in the awake state.

C = 6.25 mm. R = 10 mm in early sleep.

OFC 55 cms

Ventricular Dilatation/Cortical Mantle

Initial air ventriculogram had shown normal sized ventricles with minimal dilatation of the temporal horns. A CT scan was carried out as part of his follow up having decided not to insert a CSF shunt.

Result A very gross hydrocephalus involving both lateral ventricles and the third ventricle, the fourth ventricle was demonstrated and was not grossly enlarged. There was no displacement of the ventricular system. There was also a right anterior horn porencephalic cyst and some enlargement of the sulci over the vertex bilaterally indicating some cortical atrophy as well as hydrocephalus.

A further CT scan was carried out as conservative management was continued. The ventricular system is normal in size, shape and position. The third and fourth ventricles are also normal. There is no abnormality of cerebral parenchymal density and the prominence

of the cerebral sulci in the sylvian fissure regions previously reported is very slight and probably not of significance, i.e. the appearances are normal.

Points of Interest His subsequent development, although there is still a little developmental delay with his speech, has been very encouraging and there have been no further signs of raised intracranial pressure. There is a dramatic improvement in his CT scan and without pressure monitoring, one may have been tempted to insert a CSF shunt at the time of the first CT report. With all the attendant morbidity and mortality problems associated with shunting devices, one could not have predicted as good an outcome as the conservative management has produced.

Pressure Recordings

Fig. 92 shows the effect of sleep at fast chart speeds.
The frequency of peaked sleep values in one section of this recording was 46, 32, 28, 30, 38, 10, 26, 32, 30, 24 seconds.

Case Number	28
Name	T.W.
Age	12 years
Method	Frontal Rickham reservoir

Medical Diagnostic Background

He was born with a lumbar myelomeningocoele but no evidence of hydrocephalus. The lesion was repaired and 7 days later the OFC was definitely increasing and an air ventriculogram showed a block at the aqueduct of Sylvius. 7 days later a Spitz-Holter valve was inserted between the right ventricle and the right internal jugular vein. He had a spastic paraplegia clinically with a motor and sensory level of L_3.

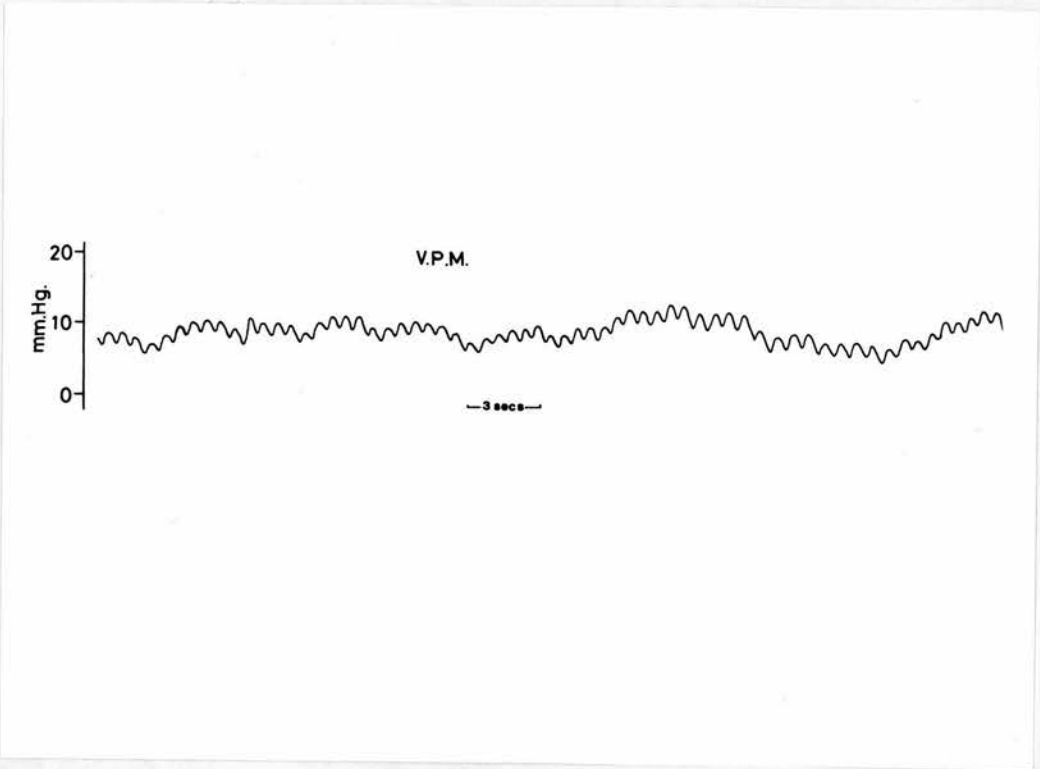

V.P.M.
20
10
0
mm.Hg.
3 secs

Fig. 92

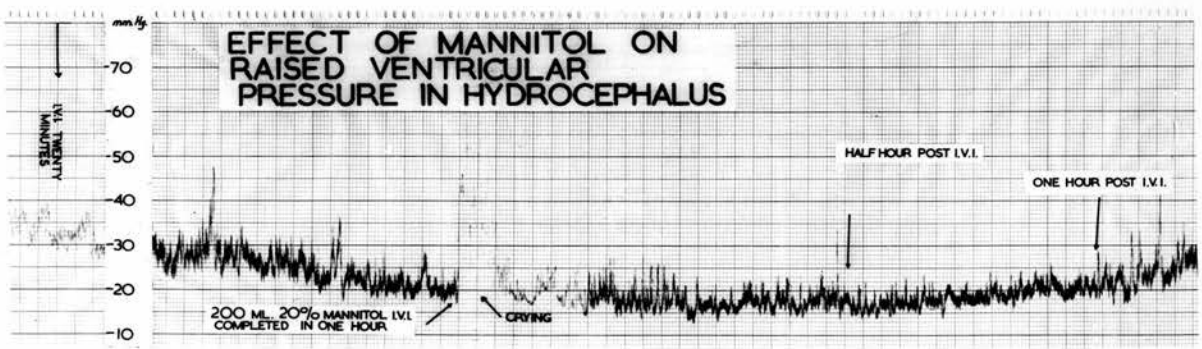

EFFECT OF MANNITOL ON
RAISED VENTRICULAR
PRESSURE IN HYDROCEPHALUS
mm.Hg
70
60
50
40
30
20
10
I.V.I. TWENTY MINUTES
HALF HOUR POST I.V.I.
ONE HOUR POST I.V.I.
200 ML. 20% MANNITOL I.V.I. COMPLETED IN ONE HOUR
CRYING

Fig. 93

He presented with a convulsion at the age of $6\frac{1}{2}$ years and because the clinical impression was that the catheter was blocked and that he had 'arrested hydrocephalus', he had a complete removal of his shunt. His OFC at this stage was 61 cms. Subsequently his ataxia appeared to become slightly worse and at the age of 12 years his OFC was 62.5 cms. Then urinary incontinence by day and night became a problem. An air myelogram showed the air did not enter the subarachnoid space within the cranial cavity but it did appear as though the foramen magnum was large and the tonsils were low and probably adhered to the cervical cord. Air did not enter the ventricular system. Myodil studies then revealed a deformity of the lumbar theca at L_{4-5} level with a partial occlusion on the dorsal aspect of L_4 level. A central bar of tissue was identified which appeared to be continuous with the cord shadow, and represented a 'tethered cord'. Ventriculography at that time showed a gross hydrocephalus present and the contrast entered an enlarged third ventricle and passed down the aqueduct into the fourth ventricle which was seen to be low in position. A small amount of contrast was seen down towards the obex but did not appear to enter a hydromyelic cavity. Pneumo-ventriculography at the same time showed filling defects in the right lateral ventricle, possibly due to adhesions and they were possibly multi-located. A Rickham reservoir was therefore electively inserted and pressure monitoring carried out.

Temperature — Normal

Duration — 7 hours

Indication — Admitted with a clinically blocked drainage system, thought to have an arrested hydrocephalus, but with some increasing ataxia and deterioration in bladder function.

Resting Ventricular Pressure

7.5 mm Hg.

Stress Ventricular Pressure

38 mm Hg peaks on bilateral Queckenstedt's, 32 mm Hg maximum peaks in sleep.

Result — Normal awake pressure but equivocal pressures during early sleep.

Action — No insertion of shunt was undertaken and since the pressure values during sleep were equivocal, follow up C.T. scans were to be performed at regular intervals.

Points of Interest

This child who was thought to have an arrested hydrocephalus, increasing ataxia and optic atrophy, could have been experiencing pressure transmitted through the obex, resulting in some loss of spinal cord function. However, myelography has shown an alternative possible explanation.

Cardiac/Respiratory Artefact

CR = 17 mm in sleep peaks. CR = 6.25 mm awake.

OFC — 63 cms

Ventricular Dilatation/Cortical Mantle

Gross communicating hydrocephalus at the time of VPM.

Case Number — 29

Name — S.M.

Age — 10 years

Method — Ventricular cannulation through burr hole.

Medical Diagnostic Background

He was born by lower segment caesarean section at 32 weeks because of P.E.T. and A.P.H. and he had an intracranial haemorrhage at 24 hours and developed an internal and external hydrocephalus. Accordingly he had

a shunt inserted and this has required several revisions. Now he has a right hemiparesis and mild spastic diplegia associated with his hydrocephalus and a moderate degree of mental handicap. Over active behavioural problems have been managed with Ritalin.

Temperature — Normal

Zero — Inter-ventricular foramina level

Duration — $3\frac{1}{4}$ hours

Indication — Vomiting for 6 weeks, once a day, mostly p.c. and occasional headaches associated with the vomiting. There was no physical abnormality detected apart from his long standing neurological signs, and, in particular, no evidence clinically of raised intracranial pressure. The right and left sided Spitz-Holter valves were functioning adequately but there was some evidence of an upper respiratory infection. Fundi were bilaterally normal.

Resting Ventricular Pressure

38 mm Hg.

Stress Ventricular Pressure

72 mm Hg maximum in sleep.

Result — Raised ventricular pressure

Action — Aspiration of 20 mls of CSF dropped the level of ventricular pressure, which then slowly built up over the next 2 hours. Shunt revision was undertaken and insertion of a Rickham reservoir. Post-operatively there was no vomiting or headaches and he has remained well since then.

Cardiac/Respiratory Artefact

17.5 mm in sleep at 75 mm Hg. CR = 5 mm at 30 mm Hg.

CR post-tap = 5 mm.

Ventricular Dilatation/Cortical Mantle

No CT scan or air pictures at this time.

Points of Interest At operation the peritoneal tube was found to be out of the peritoneum and the end was completely occluded by fibrosis.

Pressure Recordings Fig. 93 shows the effect of a slow diminution in the level of V.F.P. after Mannitol. Prior to Mannitol the CPP was 82 mm Hg with maximum decrease in ventricular pressure. At $1\frac{1}{2}$ hours after the commencement of IV Mannitol the CPP = 90 mm Hg; not a dramatic improvement in the CPP and certainly not an effective method of reducing intracranial pressure in hydrocephalus in the emergency situation. Sleep in this child results in huge excursions of ventricular pressure associated with minor myoclonic twitching of the fingers, lips and eyelids. As the ventricular pressure level rose facial flushing occurred and continuous nystagmus was more evident with increases in ventricular pressure.

It is interesting that with markedly elevated ventricular pressure in this child, he should present symptomatic but with absolutely no signs referrable to raised ventricular pressure.

Case Number	30
Name	G.McM.
Age	9 years 4 months
Method	Ventricular cannulation

Medical Diagnostic Background

Myelomeningocoele with hydrocephalus, mental retardation, epilepsy and 'precocious puberty'.

Temperature	Normal
Zero	Inter-ventricular foramina level
Duration	2 hours
Indication	Irritable and anorexic for 1 week

associated with headaches and not sleeping well. A valvogram showed

a partial distal block. This was performed because of some sluggishness of the valve. It settled however over the next few days.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

42 mm Hg maximum.

Result — Normal ventricular pressure

Action — No action with regard to ventricular pressure. However she subsequently displayed a downhill course with no signs of pressure or infection and on exploration of her valve 12 days after this monitoring, it was found to be working normally.

Cardiac/Respiratory Artefact

CR = 1.25 mm awake. CR = 2.5 mm asleep.

Ventricular Dilation/Cortical Mantle

An air encephalogram at the time showed asymmetrical ventricular dilatation with displacement of the lateral wall of the left ventricle but free communication.

Points of Interest — Removal of 30 mls of CSF and replacement of 30 ccs of air resulted in identical ventricular pressure levels being recorded both before CSF removal and after insertion of air.

Case Number — 31

Name — S.R.

Age — 5 years

Method — Ventricular cannulation through left frontal valve.

Medical Diagnostic Background

Congenital communicating hydrocephalus, mental retardation, epilepsy, deaf mutism. He had a theco-peritoneal shunt inserted in infancy and this was converted to a ventriculo-peritoneal shunt at 3 years of age. He subsequently had a number of shunt revisions for both proximal and distal blockages.

Temperature — Normal

Zero — Upper cortical subarachnoid space.

Duration — $1\frac{1}{2}$ hours

Indication — Presented with a clinical proximal block of his valve, irritability for 1 day, squinting of the right eye and generally a 'glazed' appearance, crying intermittently and 'not himself'.

Resting Ventricular Pressure

40 mm Hg.

Stress Ventricular Pressure

73 mm peaked on crying.

Result — Raised ventricular pressure due to a blocked valve.

Action — The blockage was confirmed at operation the following day.

Cardiac/Respiratory Artefact

CR = 10 mm post-tap (12 ccs CSF)

CR = 7.5 mm post tap (30 ccs CSF)

CR = 45 mm at 40 mm Hg.

Ventricular Dilatation/Cortical Mantle

An air encephalogram showed slight ventricular dilatation, a minimal increase in third ventricular size and the aqueduct and the fourth ventricle not well outlined. There was atrophy of the cerebellum and cerebrum.

Case Number 32

Name S.R.

Age 5 years 8 months

Method Left frontal Rickham reservoir

Medical Diagnostic Background

Mental retardation, optic atrophy, mutism, congenital hydrocephalus as before.

Temperature 36.5

Zero Upper cortical subarachnoid space.

Duration 9 hours

Indication Presenting complaints from his supervisors were feeling 'off colour' for 1 week, slightly lethargic and anorexic on the day of admission. He was restless, constipated and ataxic.

Resting Ventricular Pressure

25 mm Hg awake.

Stress Ventricular Pressure

Early sleep maximum 70 mm Hg peaks, 3 mm Hg post tap, 13 mm Hg post tap and early sleep.

Result Raised ventricular pressure and confirmation of a non-functioning valve.

Action Medium pressure Pudenz system replaced the following day.

Cardiac/Respiratory Artefact

R = 60 mm, C = 17.5 mm in early sleep.

R = 7.5 mm, C = 2.5 mm post tap.

CR = 32.5 mm pre tap at a level of 25 mm Hg pressure.

Ventricular Dilatation/Cortical Mantle

As before.

Pressure Recordings Most of these illustrated are accompanied by a respiratory tracing obtained from pressure recordings via a nasal catheter and are simultaneous with the ventricular pressure recording. Fig. 94 shows in the awake state, rhythmical swings of ventricular pressure extending from 10-50 mm Hg with superimposed sections of ECG. tracing obtained at the same time, showing a pulse in the range 90-120 per minute at the peak of these fluctuations and a pulse of 60 per minute at the base. In early sleep in Fig. 95 one can see elevations of the ventricular pressure in excess of 60 mm Hg and at the points where the ventricular pressure drops, a period of apnoea ensues. This is remarkably constant and occurs recurrently.

After a tap of 20 cc of CSF. Fig. 96 shows a level of ventricular pressure now at 7 mm Hg and in the normal range with a rhythmical normal respiratory pattern. Note that in all these figures, the two tracings are out of phase by 5 mm due to pen position. This is a good example of the cardio-respiratory artefact superimposed upon the ventricular pressure level. About an hour after the CSF. removal, the child became sleepy again and Fig. 97 again shows the rhythmical oscillations in early sleep but at a much lower level now. During these oscillations, one can see on this Figure, two points where the respiratory trace shows a downward deflection and this is illustrated by the arrow heads.

It is difficult to know if they represent a 'sigh' or a normally occurring valsalva. They tend to occur mostly on the downward stroke of the ventricular pressure oscillation in 'active' sleep and appear to either promote further oscillations or to progress to lower ventricular pressure levels in 'quiet' sleep.

There was no rebound increase in ventricular pressure level one hour following this tap.

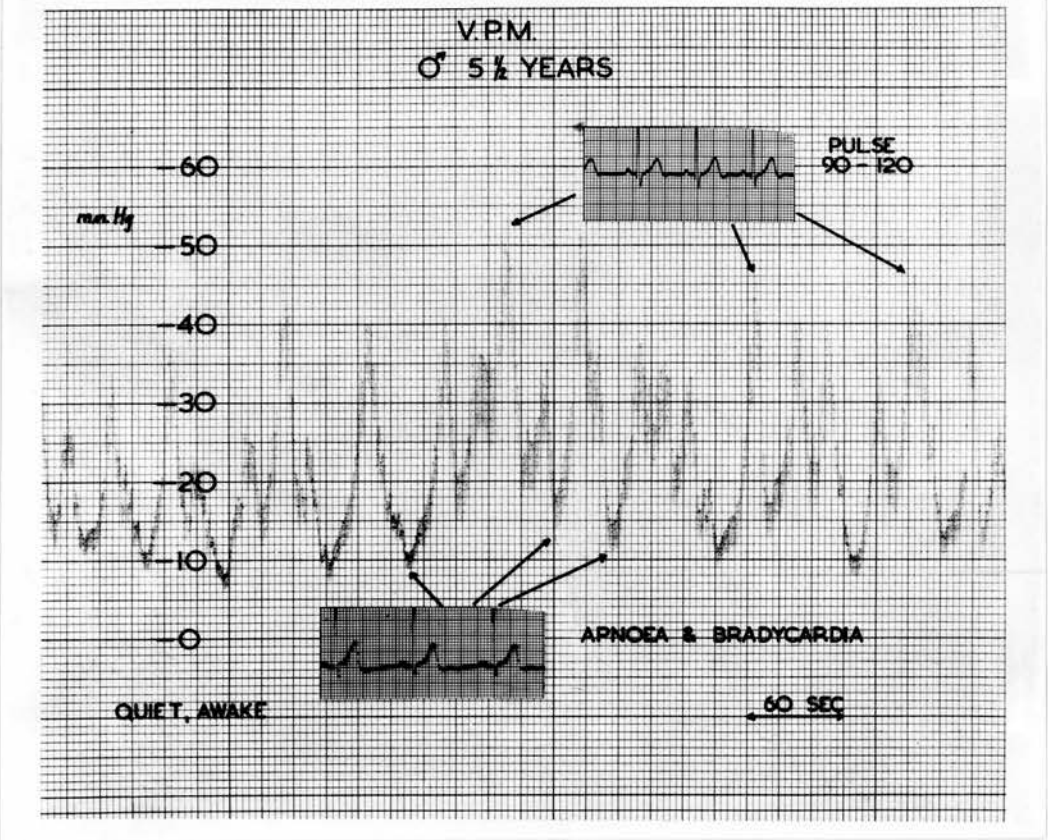

V.P.M.
♂ 5½ YEARS
PULSE
90 - 120
mm. Hg
60
50
40
30
20
10
0
APNOEA & BRADYCARDIA
QUIET, AWAKE
60 SEC

Fig. 94

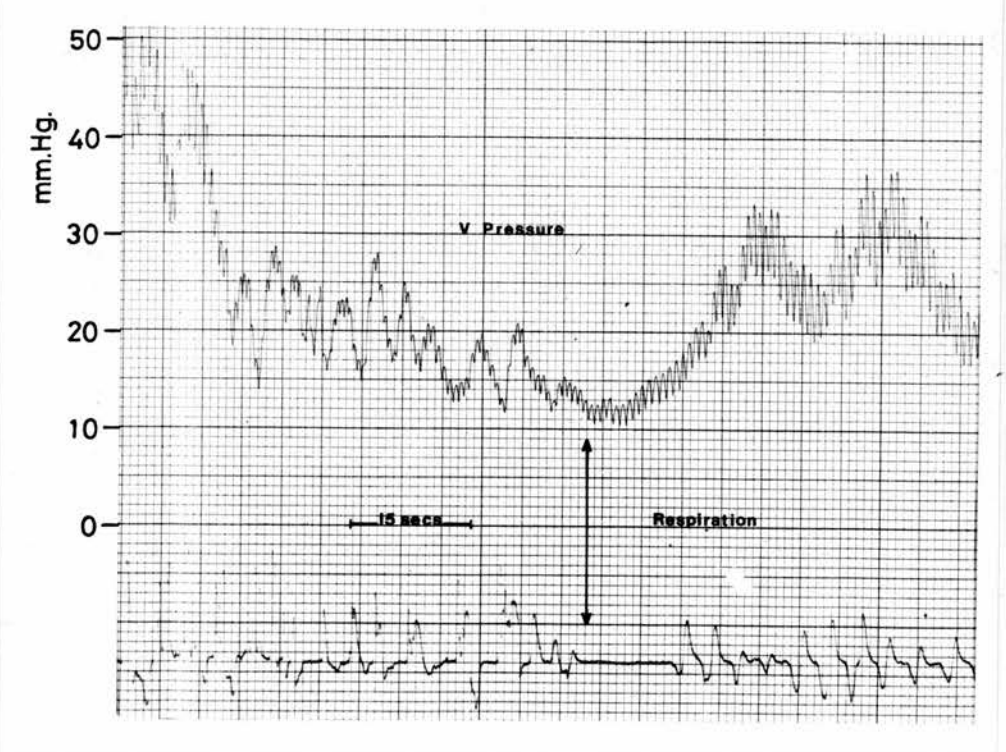

mm.Hg.
50
40
30
20
10
0
V Pressure
15 secs
Respiration

Fig. 95

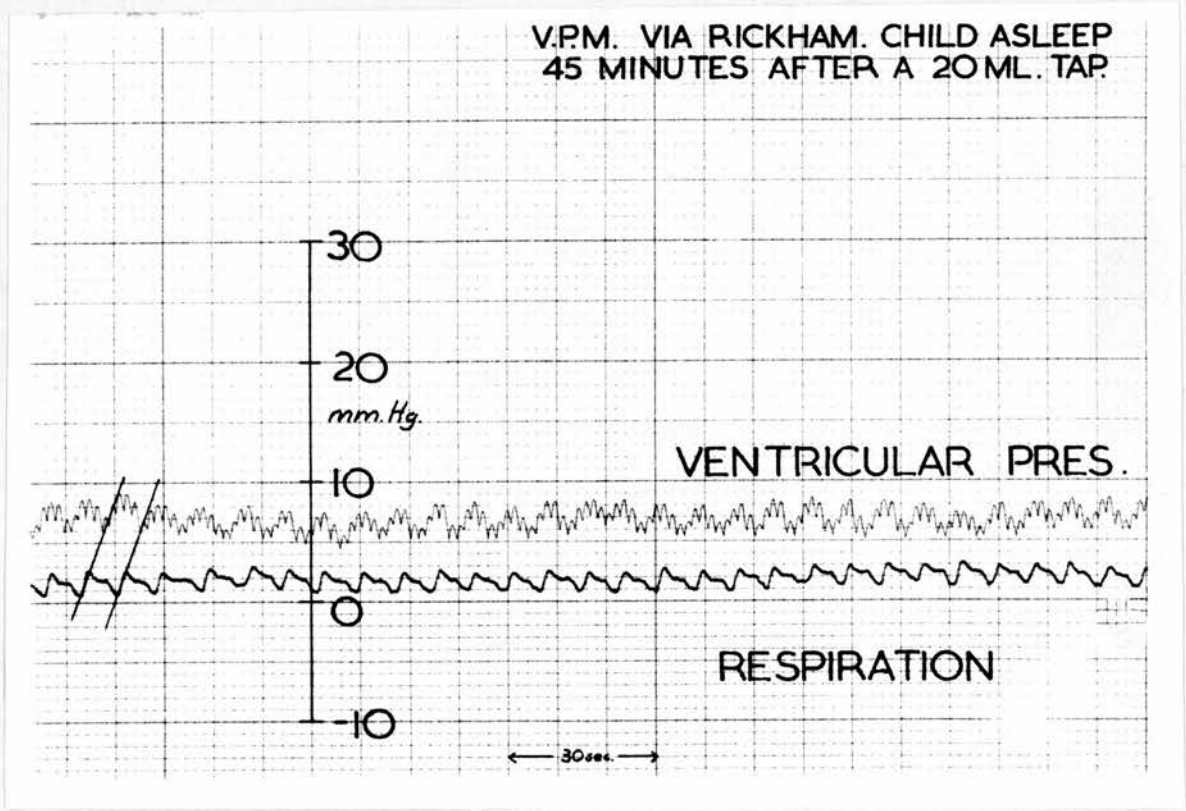

V.P.M. VIA RICKHAM. CHILD ASLEEP
45 MINUTES AFTER A 20 ML. TAP.
30
20
mm.Hg.
10
0
-10
VENTRICULAR PRES.
RESPIRATION
30 sec.

Fig.96

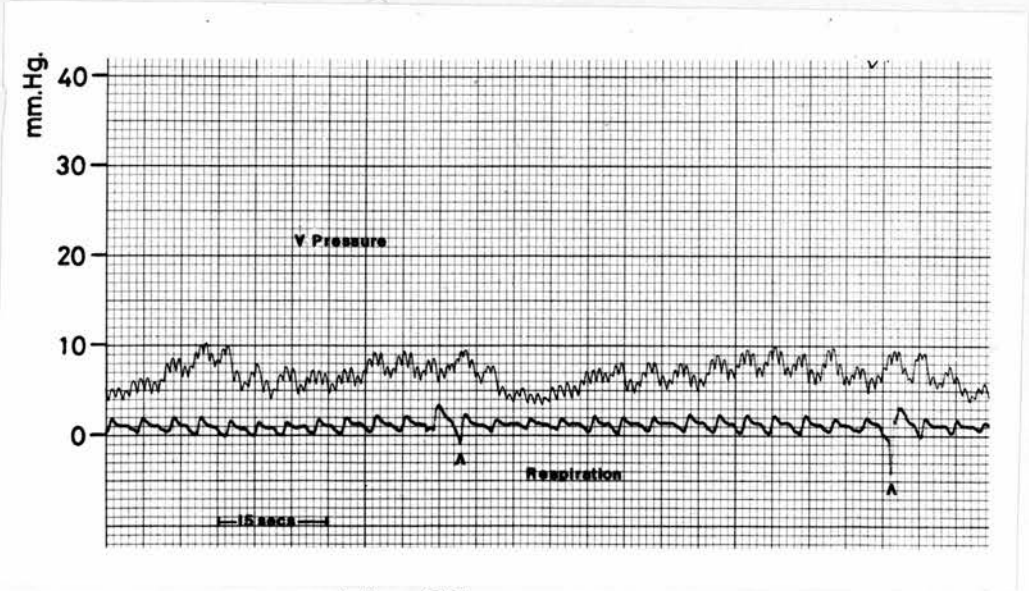

mm.Hg.
40
30
20
10
0
V Pressure
Respiration
A
A
15 secs

Fig.97

Case Number 33

Name C.M.

Age 2 years 10 months

Method Rickham reservoir

Medical Diagnostic Background

An $L_{3/4}$ level myelomeningocoele with a right sided Pudenz shunt and left sided Rickham in situ.

Temperature Normal

Zero Upper cortical subarachnoid space

Duration 1 hour

Indication The child had a seizure while being bathed. Subsequent examination raised the possibility of the valve having a proximal block. She was irritable and afebrile and had a hemiplegia of some long standing. The indication therefore was a clinical block and a fit that may have been due to raised intracranial pressure.

Resting Ventricular Pressure

4 mm Hg.

Stress Ventricular Pressure

45 mm Hg in peaks.

Result Normal ventricular pressure

Action Despite the pressure recording, it was considered that there was a clinical proximal block and that the valve should be revised. This was done the following day and at operation the proximal catheter was found to be blocked. The results of the ventricular pressure monitoring were not acted upon on this occasion. It seems likely that if she was an active hydrocephalic, such a degree of blockage would have resulted in a much more elevated level of ventricular pressure. It should be pointed out

that this was one of the earliest pressure monitorings, done in this series, and the degree of clinical usefulness at this time had not been established.

Cardiac/Respiratory Artefact

C = 3 mm awake

CR = 3.75 mm awake.

OFC 47 cms.

Ventricular Dilatation/Cortical Mantle

No recent air picture or CT. scan. Moderate ventricular dilatation and a reasonable cortical mantle were noted on AEG. 2 years earlier.

Case Number 34

Name C.M.

Age 2 years $10\frac{1}{2}$ months

Method Rickham reservoir

Medical Diagnostic Background

Myelomeningocoele, hemiplegia (as before).

Temperature 37

Zero Upper cortical subarachnoid space as before.

Duration 3 hours

Indication Readmitted shortly after previous discharge with irritability, mild drowsiness, constipation and a sluggish valve.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

45 mm Hg maximum on crying

22 mm Hg maximum in sleep.

Result — Normal ventricular pressure and on depressing the valve it appeared to reduce pressure effectively.

Action — Discharged with no operative intervention.

Cardiac/Respiratory Artefact

CR = 3 mm

OFC — 47 cms.

Ventricular Dilatation/Cortical Mantle

As before

Pressure Recordings — A limited saline infusion procedure was undertaken to estimate the cerebral compliance. Practically all that can be done prophylactically in hydrocephalic states if the compliance is poor, is to insert a low pressure shunting device. There are a number of hazards associated with compliance testing and in the absence of normative data, no other child in this series has undergone saline infusion tests.

Case Number — 35

Name — D.F.

Age — $3\frac{1}{2}$ years

Method — Fontanelle cannulation through right parietal burr hole.

Medical Diagnostic Background

Cervical meningocoele, cortical dysplasia and epilepsy, mental handicap, double hemiplegia, and with a left sided ventriculo-peritoneal Pudenz system. Medication consisted of Phenobarbitone and Phenytoin.

Temperature — Normal

Zero — Inter-ventricular foramina level

Duration $4\frac{1}{2}$ hours

Indication She was admitted in status epilepticus. A 'sticky valve' had been noted for the previous two weeks. On admission she was unconscious and given IV Mannitol, Diazepam and Dexamethasone. She has neurological evidence of 'asphyxial brain damage' and is cortically blind. The indication therefore was to differentiate whether pressure was causing her fits or whether the fits were appearing de novo or due to infection in her CSF.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

11 mm Hg maximum on Queckenstedt's.

Result Normal level ventricular pressure and valve apparently functioning satisfactorily.

Action No action taken for her intracranial pressure (apart from osmotic diuretics) and she has remained well since.

Cardiac/Respiratory Artefact

C = 2 mm, R = 3.75 mm.

Ventricular Dilatation/Cortical Mantle

No recent estimate of ventricular size was available.

Pressure Recordings By depressing the valve a number of times, one could create negative intra-ventricular pressures but obviously this was not encouraged because of the likelihood of collapsing the ventricles or inducing subdural haematomata. The CPP, when her ventricular pressure was negative (i.e. 5 mm Hg) was 95 mm Hg. It took 10 minutes for the pressure to return to the normal positive range. Fig. 98 shows the effect of hypoxia on the VEP sufficient to result in colour change. There is an increase in the mean level of

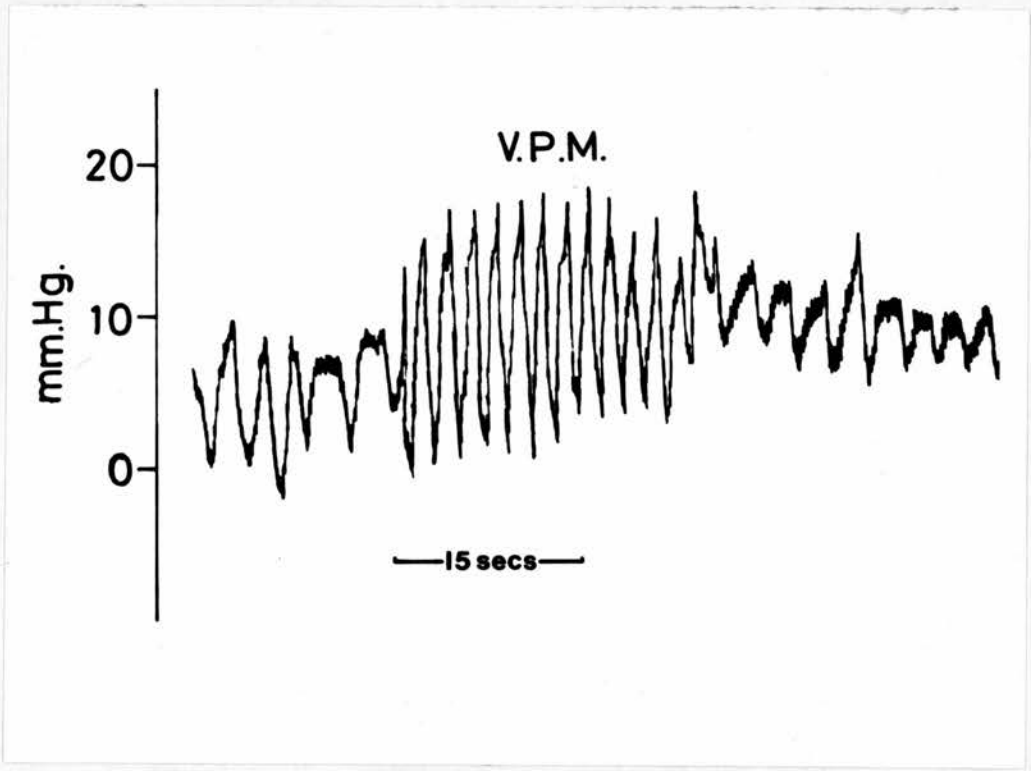


Fig. 98

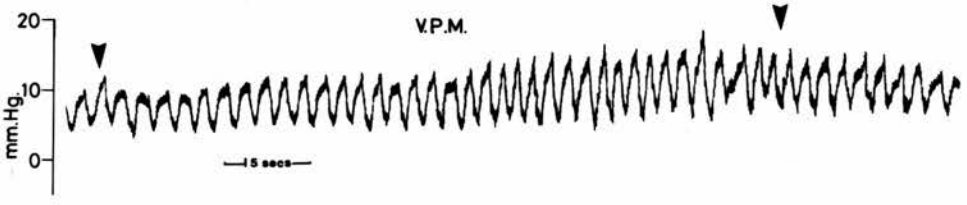


Fig. 99

ventricular pressure, but more marked was the greater width of the pulsation. Fig. 99 shows the effect of administering 6 litres of oxygen via a mask (the first arrow) with a resultant slight elevation of the base line of ventricular pressure. Again there is an increased amplitude of the ventricular pressure wave, with obviously some degree of re-breathing occurring. Also an element of emotional excitation may have contributed to the slight elevation of pressure. This is not a hyper-ventilation response but rather normal ventilation, with an increased F_iO_2. Shining bright lights into the patient's eyes made no difference to the level of ventricular pressure. Right sided Queckenstedt's raised the pressure from 3.5 mm Hg to 7.5 mm Hg on the right and 4.5 mm Hg to 6.5 mm Hg with left sided Queckenstedt's. Different movements of the head and neck initially created different respiratory patterns on the ventricular pressure tracing but this settled quickly.

Case Number 36

Name A.B.

Age 1 month

Method Ventricular cannulation through anterior fontanelle.

Medical Diagnostic Background

He was born by caesarean section because of disproportion and a previous caesarean section. Although he was generally a big baby, a large head was noted at birth and there was a relatively rapid rise in OFC from 38 cms to 43.5 cms in the early postnatal period. Apart from prominent scalp veins, examination and early progress was satisfactory. At 4 weeks of age he was admitted with a large tense anterior fontanelle, an OFC of 44 cms and distended scalp veins.

CT scan at this stage showed a considerable degree of dilatation of the lateral ventricles, most marked posteriorly. There was no displacement or deformity of the lateral ventricles. The third ventricle which was also distended was in a normal midline position. The fourth ventricle was small and in a normal position. The appearances suggested hydrocephalus due to aqueduct occlusion. Positive contrast ventriculography was carried out and this confirmed complete obstruction at the junction of the upper and middle thirds of the aqueduct of Sylvius. The lower end of the filled part of the aqueduct had a smooth rounded margin and this should be recalled as being in contra distinction to the 'rat tail' deformity of occlusion and raises the remote possibility of diaphragmatic occlusion which could be amenable to direct rupture. This possibility was considered extremely remote and should be only reconsidered if there are considerable shunt problems. Furthermore, at this age, no direct attack on the occlusion is usually attempted. Father had a rather large head when at school and this led to the possibility of a sex-linked inherited hydrocephalus due to aqueduct occlusion, but the evidence was scanty and it was felt inappropriate to pursue this as both parents are second cousins.

Temperature — Normal

Zero — Inter-ventricular foramina level

Duration — 2 hours

Indication — The assessment of active neonatal hydrocephalus confirmed as being aqueduct occlusion and although not symptomatic, he had signs (an increasing OFC, distended scalp veins, full fontanelle).

Resting Ventricular Pressure

12 mm Hg.

Stress Ventricular Pressure

45 mm Hg maximum peaks on struggling and 15.5 mm Hg sleep peaks.

Result — Marginally raised ventricular pressure.

Action — He therefore had a standard right sided ventriculo-peritoneal anastomosis with a Pudenz pump and valve inserted. Post-operatively his scalp vein distension resolved and his fontanelle became normotensive.

Cardiac/Respiratory Artefact

At rest CR = 1.5 mm. In sleep CR = 2 mm.

OFC — 38 cms at birth, 44 cms at time of VPM.

(All percentiles, height, weight, and OFC were above the 90th percentile, however, the OFC was increasing faster than the others).

Ventricular Dilatation/Cortical Mantle

CT. scan showed considerable degree of dilatation of lateral ventricles and third ventricle.

Points of Interest

With father's OFC of 60 cms and this child's development of hydrocephalus and a possibility of other 'large heads' on the paternal side of the family, it becomes very tempting to diagnose sex-linked inheritance. However, when the preceding members of the family have not required treatment and they are all of normal intelligence etc., genetic counselling in this situation is extremely difficult and in my experience this is the most common situation that arises where one suspects sex-linked inheritance but is unable to prove it definitively.

It is further evidence that children who have active neonatal hydrocephalus often have much lower recorded ventricular pressure levels than one expects. Again in this tracing, the sleep wave forms are occurring but like all neonates, the maximum sleep values are not high.

Case Number 37

Name A.B.

Age 6 months

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Congenital aqueduct occlusion with ventriculo-peritoneal shunt in as before.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 2 hours

Indication Pallor and irritability for 10 days. A 'doughy' feel to his valve and an increase in his scalp venous distension.

Resting Ventricular Pressure

18 mm Hg.

Stress Ventricular Pressure

57 mm Hg maximum sleep peaked to 35 mm Hg.

Result Raised ventricular pressure and on pumping the valve (Fig. 100) six times as indicated by the arrows, no reduction ensued.

Action His valve was revised and a right frontal Rickham reservoir inserted at that time.

Cardiac/Respiratory Artefact

CR = 4 mm at rest awake maximum

CR = 14 mm in sleep.

Follow Up He has remained well since with adequate neurological development with an OFC of 48 cms at 7 months of age, an OFC of 48 cms at 9 months of age and an OFC of 50.7 cms at 15

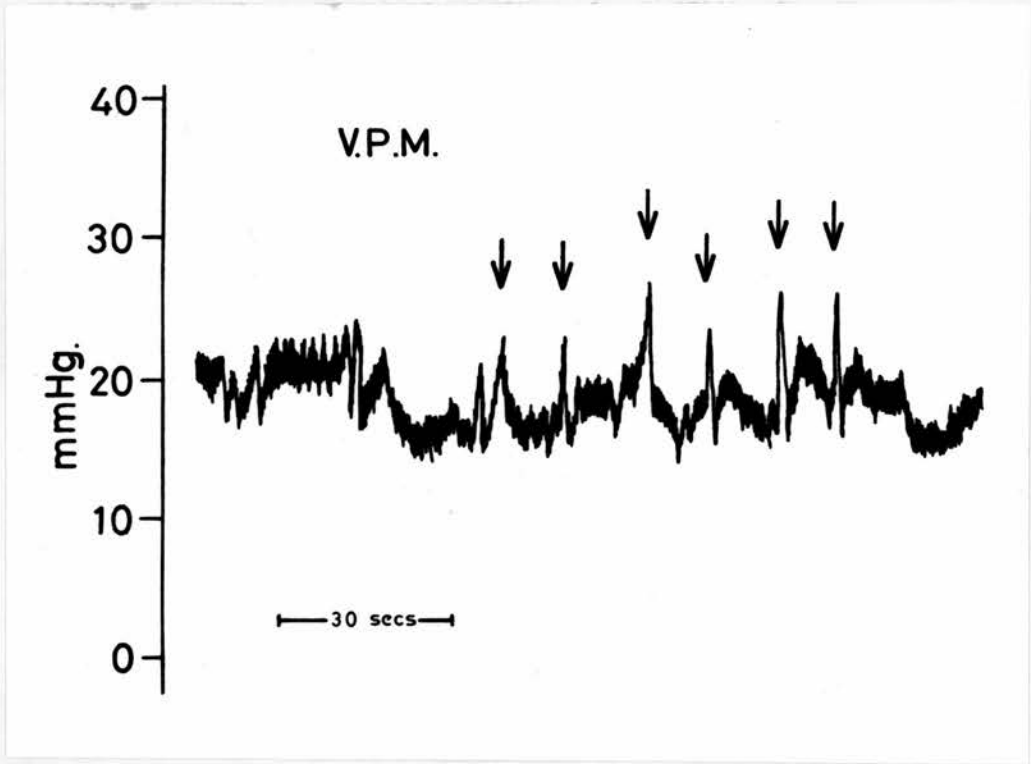


Fig.100

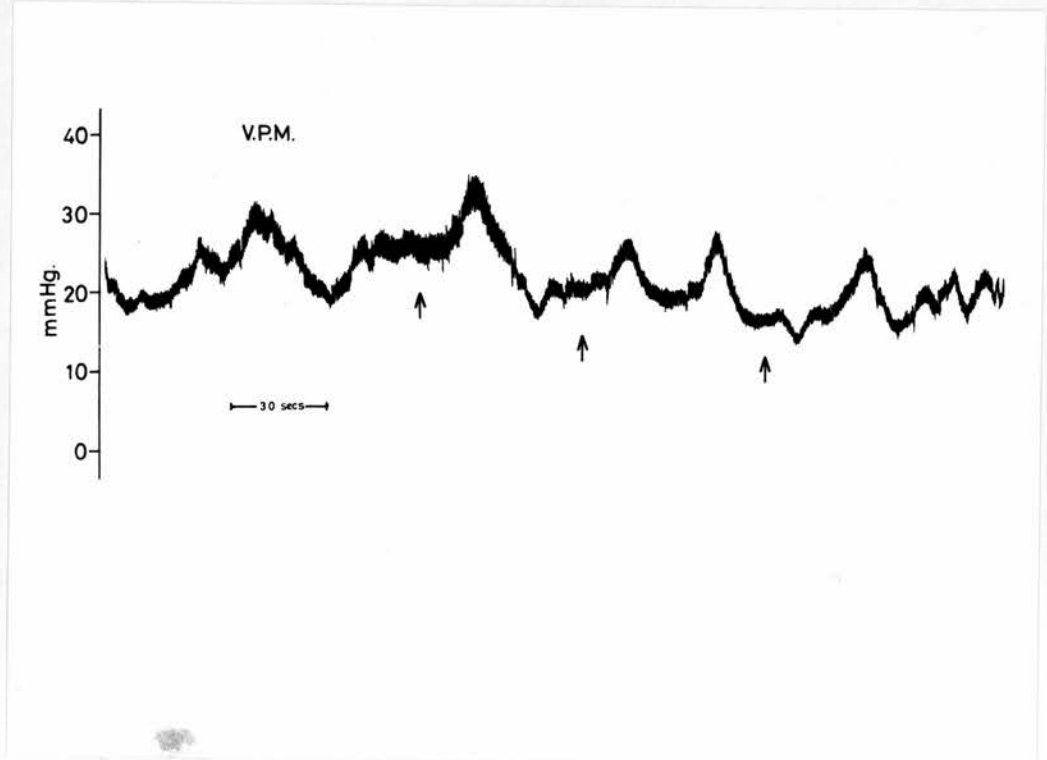


Fig.101

months of age and an OFC of 51.5 cms at 18 months of age.

Pressure Recordings The sleep levels now are quite marked compared with the previous reported ones. It can be seen in Fig. 101 during early sleep that there are periods indicated by the arrows when the ventricular pressure remains at the same horizontal level for a short period of time. From these horizontal periods the ventricular pressure appears to either continue with higher fluctuations or continue the fluctuations at a lower ventricular pressure level corresponding to the previously mentioned 'sighs' on the respiratory tracing.

Case Number	38
Name	A.B.
Age	12 years
Method	Right sided Pudenz flushing device.

Medical Diagnostic Background

He was born with a hydrocephalus and lumbo-sacral myelomeningocoele. Delivery was by forceps. He was the second twin to a mother who was well during pregnancy and whose other baby was perfectly normal. No drugs were taken during pregnancy; forceps delivery was carried out because of cephalo-pelvic disproportion. There was no neurological or familial disease in the family. The OFC at birth was 36.25 cms and $2\frac{1}{2}$ months later was 41.25 cms. The antero-posterior dimension was only slightly above normal. In 1971 he had an emergency revision of his ventriculo-atrial shunt to a ventriculo-peritoneal shunt and it then continued to function satisfactorily with no particular problems. He is of generally low IQ.

Temperature	Normal
Zero	Inter-ventricular foramina level.

Duration 1 hour

Indication He had been 'off colour' for two weeks. The school nurse noticed that his valve chamber had been much fuller than before, and he had some intermittent vomiting over one week. A valvogram showed a good flow proximally and initially a poor flow distally, but on pumping the chamber, dye was seen to flow into the abdominal cavity, i.e. the valvogram appeared to relieve any distal obstruction. However, the following day, the pumping chamber increased in size and pressure studies were carried out.

Resting Ventricular Pressure

2 mm Hg.

Stress Ventricular Pressure

30 mm Hg maximum.

Cerebral Perfusion Pressure

204 mm Hg.

Result Normal level ventricular pressure but the valve could not be demonstrated to be functioning effectively although the pressure was low.

Action No action re his shunt.

Cardiac/Respiratory Artefact

CR = 2 mm at rest.

Ventricular Dilatation/Cortical Mantle

There was no recent CT scan or AEG although 5 years earlier his cortical mantle was 13 mm.

Points of Interest

Having excluded CSF infection on admission, his symptoms disappeared after a valvogram and pumping his valve. Therefore it seems likely that the valvogram and pumping relieved an intermittent obstruction. He has remained well since.

By exerting pressure over the distal limb of his shunt 5 or 7 minutes, in the mastoid region, there was an increase in the level of ventricular pressure from 2-4.5 mm Hg and this remained so until pressure over the distal limb was released.

Case Number	39
Name	K.A.
Age	8 years
Method	Right frontal Rickham

Medical Diagnostic Background

He was born at 42 weeks gestation by S.V.D. with a lumbar myelomeningocoele. O.F.C. at birth was 33.8 cms and early surgical closure of the myelomeningocoele was undertaken. Congenital malformations of the dorso-lumbar area with hemivertebrae and a bifid lumbar spine were obvious on x-ray. At 6 days of age he had an A.E.G. which showed markedly dilated lateral ventricles. No air was seen in the third ventricle, the aqueduct, the fourth ventricle or the subarachnoid space. A reservoir was inserted on the 7th day of age and following this he developed 'hypocalcaemic fits' and had intermittent taps of C.S.F. from his Rickham to control his pressure. At 3 weeks of age a Pudenz valve was introduced. He has subsequently had a number of shunt revisions.

Temperature	Normal
Zero	Upper cortical subarachnoid space.
Duration	7 hours

Indication Gross deterioration in walking and in lower limb function generally, over 2 weeks. He had lost muscle power and now required considerable help with mobility. He had also had a spate of fits, although 'minor motor' fits had been a feature of his earlier state, and he was presently on Epanutin. There was no

change in bladder or bowel functioning. At this time his valve was 'sluggish' to feel, but there was no other evidence of raised intracranial pressure. It was known from a myelogram that there was a suggestion of a diastematomyelia with a bony buttress and scoliosis in the thoraco-lumbar spine. Total myelography was performed on this occasion under general anaesthetic which showed the cervical, dorsal and lumbar subarachnoid spaces all somewhat expanded, suggesting an overall accumulation of C.S.F. within the subarachnoid space of the spinal cord. There was no evidence of obstruction although the flow pattern of the myodil was distorted at the dorso-lumbar junction. It was decided therefore to investigate his hydrocephalus, as a reasonable spinal cause had not been found. The Rickham reservoir was therefore electively inserted, so that his pressure could be measured.

Resting Ventricular Pressure

3.5 mm Hg.

Stress Ventricular Pressure

58 mm Hg maximum on coughing.

Result — Normal level ventricular pressure.

Action — No action re C.S.F. dynamics.

Cardiac/Respiratory Artefact

CR = 2 mm.

Ventricular Dilatation/Cortical Mantle

A C.T. scan carried out one week prior to the pressure monitoring showed the midline structures undisplaced and the ventricular system was within normal size limits. Contrast studies showed a symmetrical ventricular system with no entry of contrast into the posterior third ventricle, aqueduct or fourth ventricle. The appearances suggested that the aqueduct had undergone atresia due

to effective shunting and, in fact, contrast could be seen entering the shunt system.

Points of Interest His E.E.G. was clearly indicative of cortical damage, with random spikes, small spikes appearing on both sides but mainly on the right where they are sometimes central and sometimes occipital. There was therefore no definite cause for his deterioration in walking at that stage, apart from an increase in weight and size, possibly associated with a vascular myelopathy. There was certainly no evidence of raised intracranial pressure transmitted through the obex and no evidence of 'tethering', diastematomyelia or syrinx.

Pressure Recordings He had a grand mal fit while undergoing V.P.M. (see Chapter 7).

Case Number	40
Name	K.A.
Age	8 years
Method	Right frontal Rickham

Medical Diagnostic Background

Myelomeningocoele as before.

Temperature	Normal
Zero	Upper cortical subarachnoid space.
Duration	$5\frac{1}{2}$ hours

Indication He was re-monitored because of a deterioration in the lower limbs, as, during the previous V.P.M., he had a grand mal seizure.

Resting Ventricular Pressure

3 mm Hg.

Stress Ventricular Pressure

80 mm Hg maximum.

Cerebral Perfusion Pressure

In 'quiet' sleep CR = 3.25 mm. In 'active' sleep (spikes) CR = 6.25 mm. Awake CR = 1 mm.

Ventricular Dilatation/Cortical Mantle

Normal sized ventricular system with aqueduct atresia due to effective shunting as before.

Pressure Recordings From a ventricular pressure level of 1.5 mm Hg in the awake state, he then falls asleep, and in the early sleep his pressure rises to a peak of 12 mm Hg. While the ventricular pressure is rising the B.P. is now 100/40, that is C.P.P. = 60 mm Hg transiently. There is therefore a decrease in his cerebral perfusion pressure in the early stage of sleep. Later in the recording during a further rise of ventricular pressure during 'active' sleep, the ventricular pressure level reaches 17 mm Hg and further on still in 'quiet' sleep, when the ventricular pressure is 2.5-3 mm Hg, his B.P. is 88/40, i.e. C.P.P. = 61 mm Hg.

Points of Interest This confirmed that there was no evidence of pressure occurring above the level of the foramen magnum, and furthermore, there was no evidence of the spinal pathology demonstrable on myelography. Some vascular compromise resulting in a degree of myelopathy associated with his increased growth may have been responsible.

Case Number	41
Name	A.W.
Age	12 years
Method	Rickham reservoir

Medical Diagnostic Background

He was born at 33 weeks gestation, weighing 2 lbs 11 ozs and as a

result at 9 months of age, was thought to have an ataxic diplegia. At 2 years he was thought to be bilaterally deaf and have a sixth cranial palsy. His L.A.E.G. showed communicating hydrocephalus and a theco-peritoneal shunt was inserted which required a revision at 3 years of age.

Temperature — Normal

Zero — Inter-ventricular foramina

Duration — 25 hours

Indication — He was admitted with occipital headaches and craniostenosis diagnosed on a C.T. scan. Three weeks prior to V.P.M. he had bilateral, wide relaxing craniotomies performed to decompress the brain. This left a central sagittal bar from which a segment of bone was also removed to allow antero-posterior enlargement (bone marrow seemed fatty, and was confirmed by histology). Following the operation he became very irritable and complained of headaches, and a repeat scan 8 days prior to V.P.M. showed dilatation of the ventricles, whereas before they had been small. Accordingly a Rickham reservoir was inserted, so that he could have intracranial pressure monitoring. At the time of V.P.M. he had some neck stiffness and the margins of his optic discs were blurred.

Resting Ventricular Pressure

53 mm Hg pre tap. (After removal of 3.5 mls C.S.F., ventricular pressure 43 mm Hg.)

Cerebral Perfusion Pressure

37 mm Hg.

Result — Very high ventricular pressure

Action — A medium pressure Pudenz system ventriculo-peritoneal shunt was inserted the following day, and although the ventricle was found at a depth of 6 cms, the C.S.F.

was obviously under considerable pressure.

Cardiac/Respiratory Artefact

16 mm during plateau.

OFC 53.5 cms

Ventricular Dilatation/Cortical Mantle

Cortical mantle of 6 cms and mild dilatation of the ventricles.

Points of Interest The skull x-ray had shown closure of all the sutures, but in particular, the sagittal and coronal sutures.

The reason for the original relaxing craniotomy, was that a Rickham reservoir was attempted to be inserted, but this failed due to a difficult puncture, with the small ventricles at that time. To postulate, the most likely sequence of events here is of a child with congenital hydrocephalus, treated with a theco-peritoneal shunt initially and then either from inadequate shunting, or as a result of some bony dysplasia (which caused the hydrocephalus in the first place) he developed a craniostenosis. He next displayed signs of pressure with occipital headaches. Whether this was due to a degree of aqueduct blockage as a result of an intercurrent infection, or whether due to transient upper respiratory infection causing an increased C.S.F. production, or due to a degree of brain swelling from whatever cause, there was certainly limited room for expansion of the intracranial contents because of the craniostenosis. Remember that sutures may spread with increased intracranial pressure up until about 12 years of age. It seems likely that this was due to a degree of brain swelling, particularly as the ventricles were not excessively small or collapsed, but the alternative explanation is that his theco-peritoneal shunt had blocked or was not draining adequately, and with an intercurrent infection, increased ventricular pressure resulted.

There was no adaptive mechanism such as C.S.F. displacement into the spinal subarachnoid space or room for expansion of the cranial cavity. Since the brain is incompressible, (nearly) when the relaxing craniotomies were performed, this created sutures and allowed for some of the pressure to be accommodated by swelling the brain. When a reservoir was inserted and the pressure monitored, it was found to be high and large amounts of C.S.F. removal resulted in very small reductions in pressure, and only for short periods which was further evidence to support a degree of brain swelling and, on looking at the operative notes, the ventricle was found at a depth of 6 cms which is certainly not gross hydrocephalus.

Insertion of a ventriculo-peritoneal shunt resulted in adequate compensation and an excellent recovery with no post-operative complications. At follow up, his mother reported that he was quite back to normal, if not performing a little better than he had been previously. There was indrawing of a small defect over the surgical sinus as seen in Fig. 102 .

Pressure Recordings Huge sleep 'plateaus' are seen in this child (Fig. 103). During these plateaus he was moaning, tossing and turning, and although the pressure was raised all the time, it was much more so in sleep. The plateaus were virtually continuous. Despite 80 mls of C.S.F. removal in four stages (15 ml, 15 ml, 30 ml, 20 ml), the pressure still remained elevated. Frequently throughout these recordings the nurses' observations written on the recorder paper at the time of plateaus included such comments as 'sleeping', 'settled', etc., that is, these pressure changes were occurring without any disturbance of vital signs or obvious problems to the nursing staff. Fig. 103

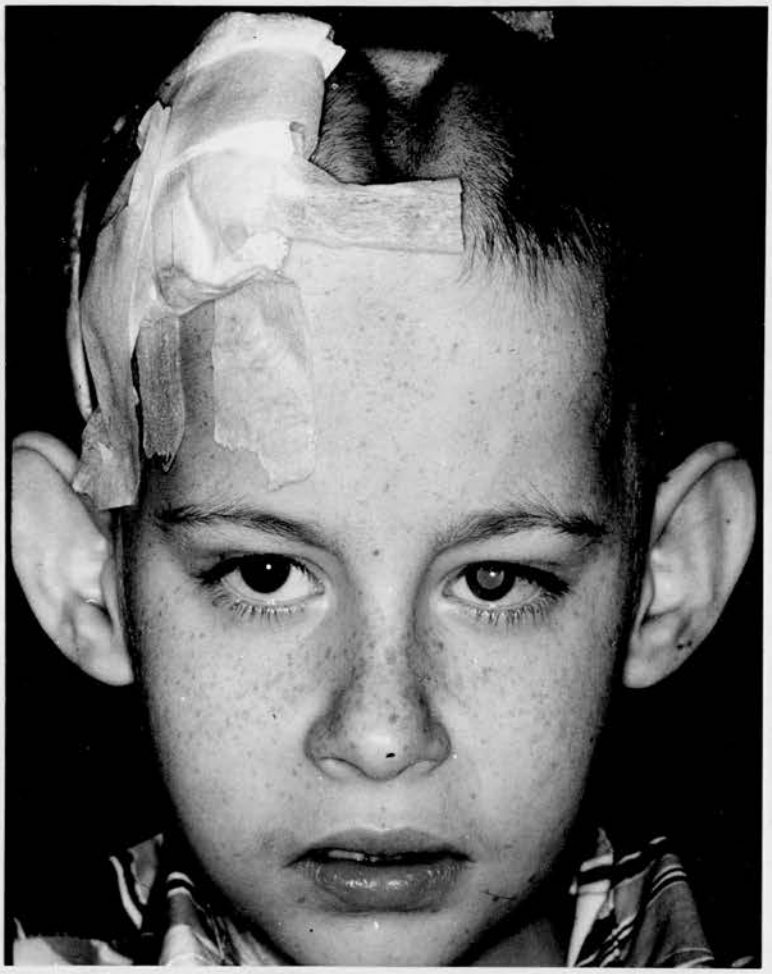


Fig.102

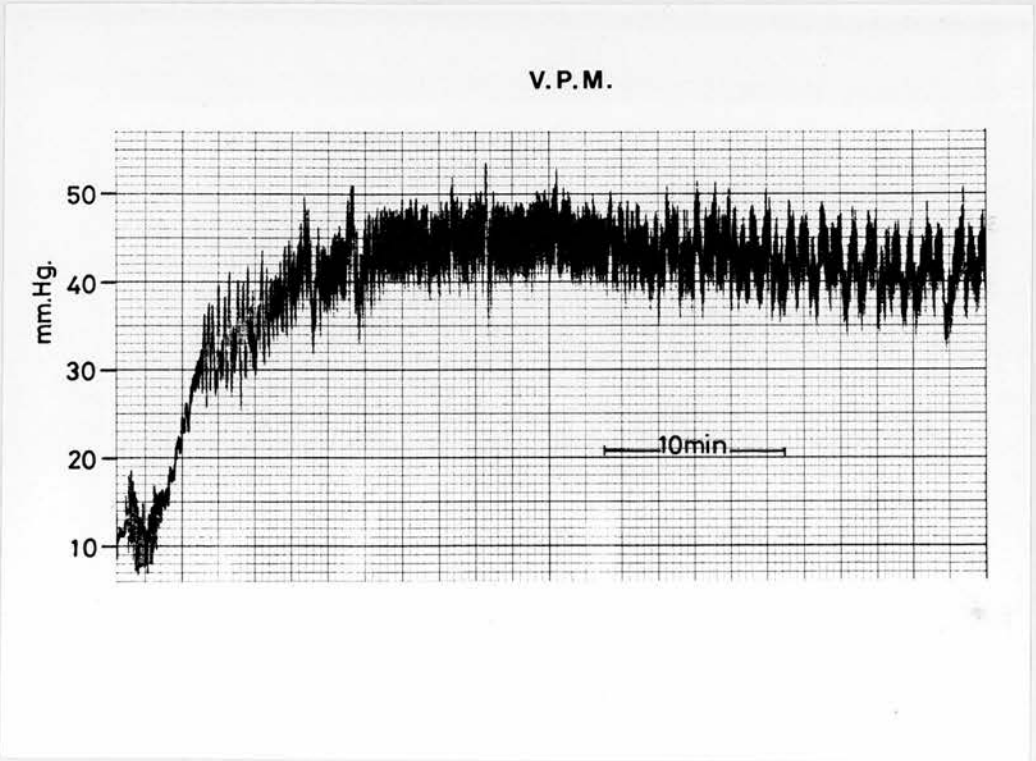


Fig.103

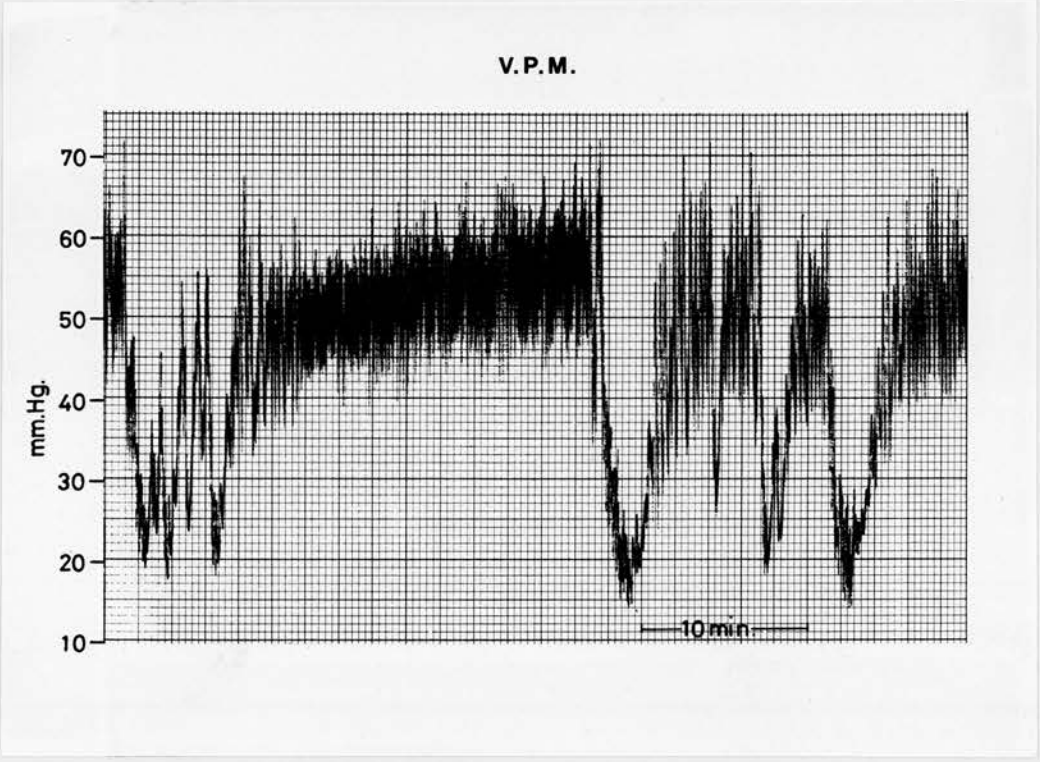


Fig.104

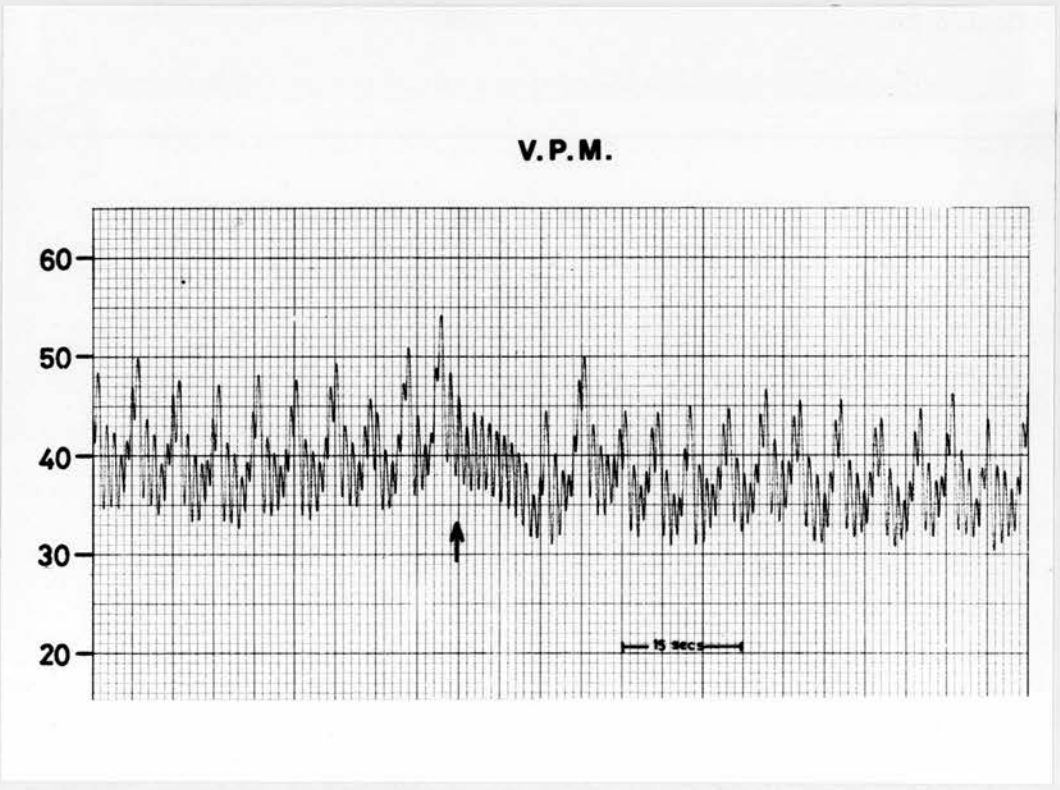


Fig.105

shows one continuous plateau which goes on for quite some long period. A further Fig. 104 shows shorter duration plateaus associated with a lot of moaning and disturbed sleep. Throughout these plateaus, he frequently scratched his nose or his face. With such a high level of pressure, a disturbed sleep is not unexpected.

Case Number 42

Name S.W.

Age 6 years $10\frac{1}{2}$ months

Method Fontanelle cannulation via burr hole.

Medical Diagnostic Background

Myelomeningocoele.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 4 hours

Indication For investigation of headaches of 3 weeks' duration, associated with nausea and vomiting. A Pudenz ventriculo-peritoneal shunt in situ, which was clinically acting normally, and his C.S.F. was microscopically clear.

Resting Ventricular Pressure

4 mm Hg.

Stress Ventricular Pressure

32 mm Hg maximum, 36 mm Hg peaks during sleep for 5-10 minute periods only.

Result Normal resting ventricular pressure. Equivocal levels during early sleep.

Action It was decided not to revise his shunt at this time.

Follow Up He has remained well since then, with no further evidence of raised intracranial pressure but requires follow up with regular C.T. scans.

Cardiac/Respiratory Artefacts

CR = 2.5 mm at rest. CR = 20 mm during sleep.

Ventricular Dilatation/Cortical Mantle

No recent estimate of ventricular size.

Pressure Recordings Again the V.P.M. showed well defined characteristic sleep changes.

Case Number 43

Name G.D.

Age 8 days

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Congenital aqueduct stenosis.

Temperature Normal

Zero Upper cortical subarachnoid space.

Duration 1 hour

Indication The assessment of suspected active neonatal hydrocephalus (increasing OFC, irritability, scalp vein distension associated with poor feeding and mild jitteriness).

Resting Ventricular Pressure

12.5 mm Hg.

Stress Ventricular Pressure

23 mm Hg maximum.

Result Raised ventricular pressure

Action Insertion of ventriculo-peritoneal shunt.

Cardiac/Respiratory Artefact

CR = 0.75 mm at rest

OFC 38.8 cms

Ventricular Dilatation/Cortical Mantle

Gross symmetrical dilatation of lateral and third ventricles.

Case Number 44

Name G.D.

Age 7 weeks

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Congenital aqueduct stenosis with right sided ventriculo-peritoneal shunt in situ.

Temperature Normal

Zero 2" below upper cortical subarachnoid space.

Duration 2 hours

Indication Despite shunting, she still had slight 'sunsetting' and the OFC had not dramatically improved, but clinically the shunt felt normal. A valvogram showed a poor position of the ventricular end of a long catheter, and some residual myodil in the proximal tip. Although C.S.F. flowed back, dye did not appear to be entering the ventricles.

The distal end of the shunt appeared normal. General observation of the child was satisfactory, and she was sent home for 3 days with instructions for mother to pump the valve 25 times, three times a day before meals, and she was readmitted for pressure monitoring to be carried out.

Resting Ventricular Pressure

4.5 mm Hg.

Stress Ventricular Pressure

12 mm Hg maximum peak in sleep.

Result — Normal ventricular pressure

Action — No action re pressure.

Cardiac/Respiratory Artefact

CR less than 1 mm at rest.

CR 15 mm during sleep.

OFC — 42.5 cms at time of this V.P.M.

(OFC at birth 36.4 cms. OFC at 48 hours of age 38 cms. OFC at the time of the ventriculo-peritoneal shunt insertion 38.8 cms).

Points of Interest — Two days following the V.P.M. a tap of C.S.F. revealed that she had a ventriculitis. Unfortunately, sampling of the C.S.F. at the conclusion of the V.P.M. for culture was not undertaken in this case, and it is an important facet of management in all cases now, that C.S.F. is sampled for microscopy and culture at the end of the procedure. However, on the 17th post-operative day, after the shunt was inserted, there was 10 leucocytes per cu.mm in the C.S.F. No action was taken on this in retrospect and the child then at discharge appeared to be having pressure symptoms. The C.S.F. was not examined microscopically until 2 days after the V.P.M. So although the possibility does exist, that infection complicated this procedure, an alternative possible explanation is that she had persisting minimal C.S.F. infection following her insertion of shunt, which was responsible for the persisting mild pressure symptoms.

Case Number 45

Name W.C.

Age 4 years 9 months

Method Left frontal Rickham reservoir

Medical Diagnostic Background

He was born after a normal pregnancy, delivery and labour. He weighed 8 lbs at birth. He cried immediately and required no resuscitation. However he was kept in a special care baby unit because of an enlarged head, and transferred for investigation after 3 weeks. During this time he had been vomiting, anorexic, constipated, irritable and was failing to gain weight. A theco-peritoneal shunt was inserted at that time. There was no significant family history. At the time of insertion of the shunt the OFC was 46 cms. The fontanelle was normotensive. There was generalised hypotonia, with an increase in the phasic reflexes. During the early neonatal period, he also had idiopathic hypo-clacaemia and a left inguinal hernia. The theco-peritoneal shunt was abandoned and a right sided ventriculo-peritoneal shunt was inserted at 10 months of age. This was revised at 1 year of age.

Temperature 36.5

Zero Inter-ventricular foramina level.

Duration 4 hours

Indication He was symptomatic but without any clinical signs. He was known to be of short stature and on the occasion of admission he had been unwell for 10 days with vomiting, unrelated to meals, associated on some occasions with pain in the right side of his neck. He had lost some weight during the previous two days and had been more drowsy than usual. A slightly sluggish

(refilling) valve had not changed markedly. There was no evidence of papilloedema, neck stiffness or any increase in the previously noted truncal ataxia. Parents reported that the episodes when he complained of headache, backache and pain in the neck were associated with sleepiness and intermittent pressure increases were suspected. Therefore an elective Rickham reservoir was inserted for the purposes of measuring his pressure.

Resting Ventricular Pressure

15 mm Hg awake.

Stress Ventricular Pressure

45 mm Hg maximum during sleep.

Result

Raised ventricular pressure.

Action A valvogram was performed which showed an encysted distal end of the shunt. He was therefore taken to theatre and a new right sided ventriculo-peritoneal shunt was inserted. At operation the peritoneal catheter was found to be short and was in fact lying in the abdominal wall. On exploration of the upper end of the ventricular tubing, which was known from a previous valvogram to be lying in the temporal horn, it was extremely reluctant to be removed and therefore the flushing device was removed and the ventricular catheter tied off and sutured to the side of the burr holes, so that it could not migrate into the ventricular system.

Cardiac/Respiratory Artefact

R = 50 mm in sleep. C = 25-30 mm.

R = 18 mm awake. C = 10 mm.

OFC 55 cms.

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed some dilatation of the ventricles.

Pressure Recordings

Fig. 105 shows the elevated pressure in this child during a section of sleep. At the point indicated by the arrow, there is a drop in the level of ventricular pressure, without any obvious respiratory component. The start of the decline in pressure before 'quiet' sleep commences without any clinically obvious respiratory change. It occurred about 10 minutes into sleep. Thereafter the level of ventricular pressure is minimally below that which preceded it. Fig. 106 shows at a faster recorder speed, the inspiratory (I) and the expiratory (E) components of the ventricular pressure tracing in this child.

Case Number	46
Name	I.C.
Age	10 years 7 months
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

She was born with a thoracic myelomeningocoele. She is moderately retarded and had a right sided Pudenz ventriculo-azygos shunt functioning well. She also suffers from fits with no increased frequency of occipital spikes on an E.E.G. During the fits which she experiences, she becomes transiently blind. She has had several complications pertaining to her shunt and is generally a little ataxic and has been monitored in the past in an attempt to decide whether her fits were a result of pressure or not. Her E.E.G.'s have shown a left occipital spike to be the most prominent feature. Otherwise the background tends to be of theta activity at 4-5 Hz with

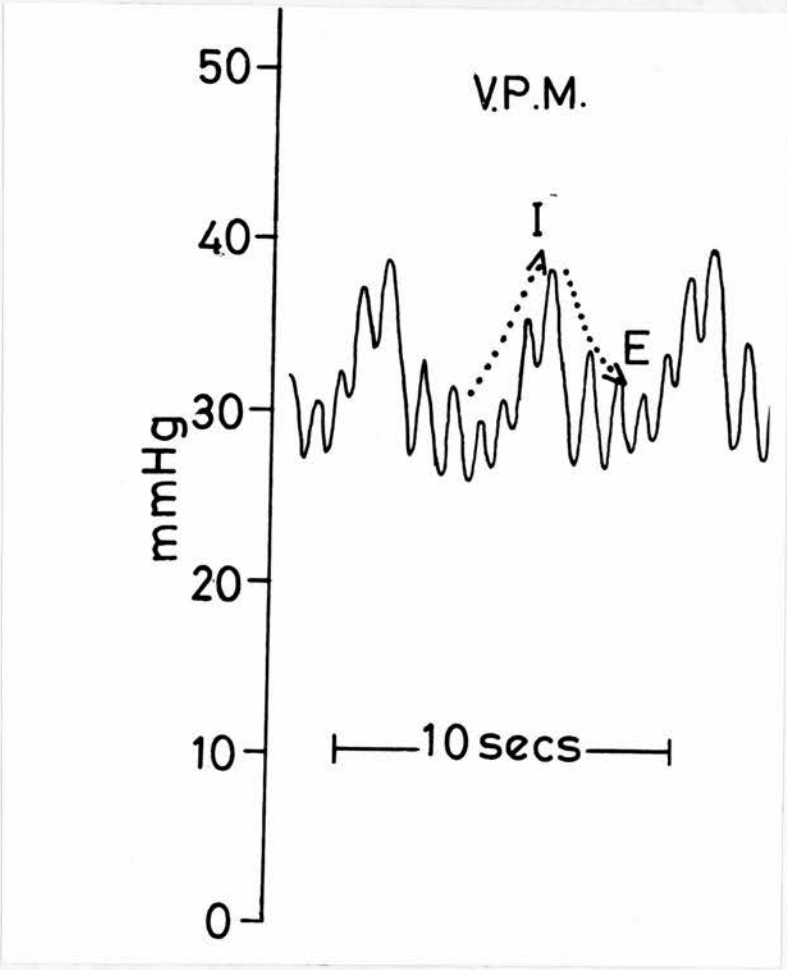


Fig.106

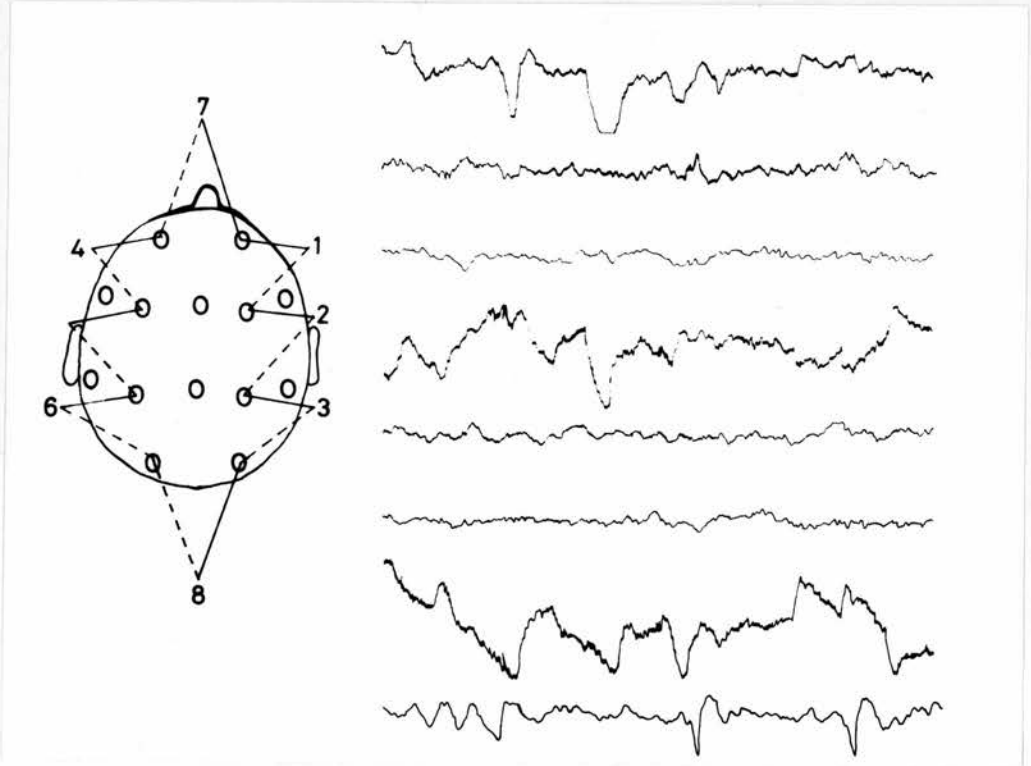


Fig.107

superimposed background fast activity. It is thought that this spike is probably rising from the medial surface of the occipital cortex and in the E.E.G. (Fig. 107) there is very prominent occipital spike activity. The overall pattern is curiously disorganised and the spike potentials are seen only in recordings in a transverse plain appearing to have a potential maximim on the right side, but probably, as before, a focus on the left. There is a slight asymmetry of the general background activity, most marked posteriorly which would be consistent with the earlier explanation of slightly diminished activity on the left. There is evidence of structural damage to one or other occipital lobes. This may have been due to posterior cerebral insufficiency during some of her acute pressure episodes in the past, or, in view of her high level lesion, associated cerebral dysplasia resulting in fits. I think the former is more likely in this child. She is managed on Epanutin and Epilim. During these attacks she becomes panicky and says to her mother 'not to leave her and to hold her tight'. The fit may abort at this stage. After an aura the seizure lasts for about 5 minutes. Sometimes there is a complete loss of vision.

Temperature	Normal
Zero	Upper cortical subarachnoid space
Duration	$8\frac{1}{2}$ hours
Indication	She was admitted with an increased

incidence of fits and a 'change in character' and there was confusion clinically whether her symptoms were due to fit control or intracranial pressure. She had been complaining of right frontal headaches. These were spreading to the occipital region and causing stiffness on bending her neck. Her fit frequency had been of the

order of 3-4 per day and her anticonvulsant medication had been increased. On the day of admission, a fit lasted about 2 minutes. She became akinetic, floppy, her eyes stared and she was incontinent. There was some post-ictal drowsiness. She had a poor appetite for the previous 3 weeks.

Resting Ventricular Pressure

16 mm Hg.

Stress Ventricular Pressure

69 mm maximum on crying. 27 mm maximum peaks during sleep.

Cerebral Perfusion Pressure

96 mm Hg at conclusion of monitoring.

Result Raised ventricular Pressure

Action Arrangements were made for revision of her shunt but since it was an atrially placed shunt, an echo cardiogram and E.C.G. were peformed first. The E.C.G. was normal and the echo showed a normal tricuspid valve and clinically there was no evidence of pulmonary hypertension. At removal of the atrial catheter through a right sided thoracotomy incision, the end of the shunt was decidedly blocked. During monitoring, before the shunt was removed, she was controlled by regular tapping from her reservoir.

Cardiac/Respiratory Artefact

CR = 2.5 mm at the lowest pressure.

CR = 21 mm at maximum stress.

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed considerable dilatation of both lateral ventricles. The dilatation was somewhat asymmetrical, the left lateral ventricle being larger than the right. There was not good

visualisation of the third and fourth ventricles.

Points of Interest The change in character mentioned was a withdrawal in demeanour compared with her previous good natured socialibility, and there was no obvious focus of infection on admission. The fundi had shown some choking of the discs and there was some limitation of eye movement, but most marked was her non-communicative state.

Case Number	47
Name	I.C.
Age	10 years 10 months
Method	Right frontal Rickham

Medical Diagnostic Background

Myelomeningocoele as before.

Temperature	36.5
Zero	1" above upper cortical subarachnoid space
Duration	1 hour

Indication She was admitted with vomiting and a spate of fitting having been well until the night before, when she had a grand mal seizure, vomiting, incontinence, followed by further fitting and then post-ictal dazing. She continued with copious vomiting overnight and was sweating profusely. She complained of a stiff neck. There had been no contact with any inter-current infection and her anticonvulsant medication had been taken regularly.

Resting Ventricular Pressure

25 mm Hg.

Cerebral Perfusion Pressure

At the outset 65 mm Hg with a B.P. of 120/60.

Result — Raised ventricular pressure

Action — After 5 mls of C.S.F. was removed the C.P.P. was now 80 mm Hg (Fig. 108). The pressure did not decrease on pumping her valve more than 3 mm Hg. A further 5 mls of C.S.F. was released, which improved the C.P.P. to 92 mm Hg, i.e. a successive improvement in the C.P.P.'s with consecutive C.S.F. releases. At the conclusion of V.P.M. the pulse rate was now 100 per minute compared to 80 per minute at the outset, and the pulse had become regular, losing the sinus arrythmia. A further 10 mls of C.S.F. was removed the next day, but by the following day she was asymptomatic and not needing further C.S.F. taps. One can only assume that this was an intermittent blockage or a transient block of the proximal atrial shunt which righted itself with pumping of the valve, or a slightly incompetent valve which can deal with only a fraction of the C.S.F. she was producing at that time. In view of all her previous problems and the fact that it was an atrially placed shunt, it was decided not to pursue this any further and although a further pressure recording was done before she was discharged, this was within normal limits, and, on this occasion, her valve appeared to be functioning adequately.

Ventricular Dilatation/Cortical Mantle

As before.

Case Number — 48

Name — L.M.

Age — 2 years 8 months

Method — Left parietal Rickham reservoir

Medical Diagnostic Background

Lumbar myelomeningocoele and hydrocephalus.

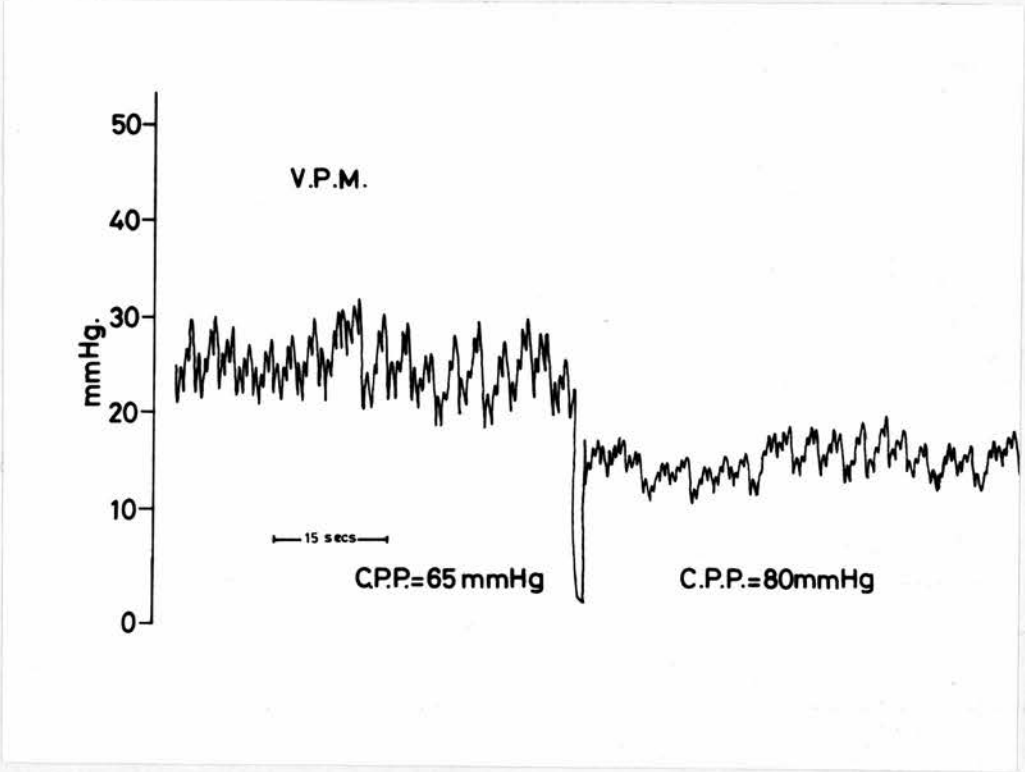


Fig.108

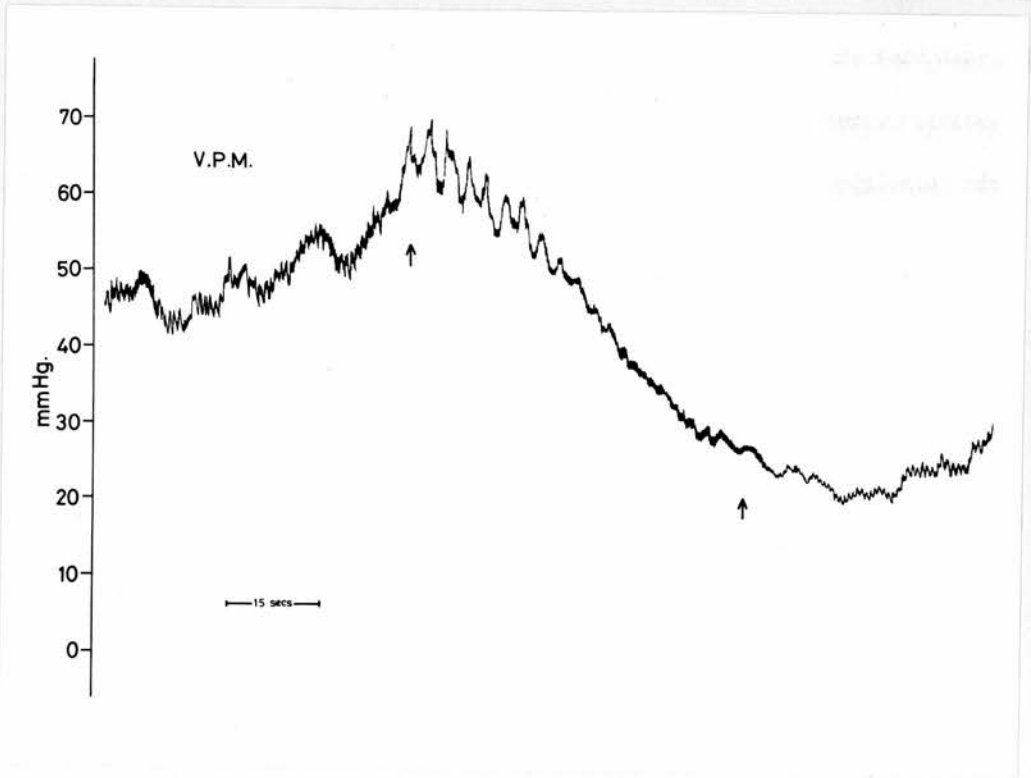


Fig.109

Temperature — Normal

Zero — Inter-ventricular foramina level

Duration — 4 hours

Indication — This child was admitted with a ventriculo-peritoneal shunt in situ, which was found to be leaking through an umbilical defect for the previous 6 weeks. This was revised and the tract excised. However she developed a ventriculitis following this and required the appropriate treatment, and the ventriculo-peritoneal shunt was removed and a Rickham reservoir inserted. It seemed evident that since the shunt had been leaking C.S.F., that it was required to control her pressure. However, in addition to the leaking C.S.F. from the umbilical lesion, she had a distensible swelling over her lumbar region, the site of her original spina bifida lesion, and this pressure monitoring was therefore not only to estimate the amount of C.S.F. required to tap, but to get an overall indication of her ventricular pressure over a period of time.

Resting Ventricular Pressure

12 mm Hg.

Stress Ventricular Pressure

34 mm Hg maximum on crying.

27 mm Hg maximum peaks during sleep.

Result — Raised ventricular pressure.

Action — She continued to be 'tapped' as necessary until a ventriculo-peritoneal shunt was re-inserted.

Cardiac/Respiratory Artefact

CR = 5 mm in sleep peaks. CR = 2.5 mm awake. C = 1.25 mm in sleep.

OFC — 49 cms (34.2 cms at birth)

Ventricular Dilatation/Cortical Mantle

Moderately severe communicating hydrocephalus. (It is interesting that her first lumbar air encephalogram showed a non-communicating hydrocephalus immediately after birth.)

Points of Interest At the conclusion of her pressure monitoring, she had developed a pyrexia, which subsequently proved to be due to chickenpox. She required 30 mls of C.S.F. released per day to remain well during this period.

Case Number 49

Name D.D.

Age 13 months

Method Right frontal Rickham reservoir

Medical Diagnostic Background

Lumbo-sacral myelomeningocoele with associated hydrocephalus.

Temperature Normal

Zero $1\frac{1}{2}$" above cortical subarachnoid space.

Duration 4 hours

Indication He was admitted with two episodes of vomiting and a valve which was clinically 'sluggish'.

Resting Ventricular Pressure

10 mm Hg.

Stress Ventricular Pressure

75 mm Hg maximum on crying.

28 mm Hg maximum peaks during 'active' sleep.

7 mm Hg during 'quiet' sleep.

Result Raised ventricular pressure and no reduction of pressure on pumping his valve.

Action — Operative revision of shunt showed the proximal end to be blocked.

Cardiac/Respiratory Artefact

R = 5 mm, C = 2 mm at rest.

R = 47 mm, C = 30 mm at maximum stress.

OFC — 46 cms.

Ventricular Dilatation/Cortical Mantle

No recent C.T. scan although initial A.E.G. showed mild ventricular dilatation 12 months earlier.

Case Number — 50

Name — D.D.

Age — 13 months

Method — Rickham reservoir

Medical Diagnostic Background

Lumbo-sacral myelomeningocoele with hydrocephalus.

Temperature — 37.2

Zero — Upper cortical subarachnoid space

Duration — $\frac{1}{2}$ hour

Indication — Following operative intervention two days earlier and renewal of the proximal end of the ventriculo-peritoneal shunt, he remained symptomatic with drowsiness and bradycardia and required 4 hourly tapping of C.S.F. Infection at this stage was excluded but C.S.F. was mildly blood stained.

Resting Ventricular Pressure

18 mm Hg.

Stress Ventricular Pressure

55 mm Hg maximum stress.

Result — Raised ventricular pressure and no effect on pumping the valve again on this occasion.

Action — Removal of 10 ccs of C.S.F. reduced the pressure to 5 mm Hg, that is in the normal range. Tapping was continued until the shunt was revised again 24 hours later. From this operation he made a good recovery.

Cardiac/Respiratory Artefact

C = 2 mm, R = 5 mm at 5 mm Hg.

C = 8 mm, R = 22 mm at 18 mm Hg.

OFC — 46 cms.

Case Number — 51

Name — E.M.

Age — 4 years 11 months

Method — Left frontal Rickham reservoir

Medical Diagnostic Background

Congenital hydrocephalus, mental retardation, ataxic diplegia and right hemiplegia.

Temperature — Normal

Zero — Upper cortical subarachnoid space.

Duration — $\frac{1}{2}$ hour

Indication — This girl presented with 'fits' having fallen at her home. She had a diminished conscious state, vomiting and irritability. On investigation she had a shift on her echo and a Rickham reservoir was inserted. Following this one week later she had a right sided ventriculo-peritoneal shunt revised. At that time she had evidence of midbrain compression from probably posterior territorial herniation with hemibalismic movements,

irritability, blindness and an element of flexor regidity. Raised intracranial pressure was suspected and pressure monitoring carried out.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

36 mm Hg maximum.

Result — Normal level ventricular pressure, although the recording was done over a short period of time. Pumping her valve resulted in a drop in ventricular pressure level.

Cardiac/Respiratory Artefact

CR = 22 mm resting.

OFC — 59.5 cms.

Points of Interest — It is difficult to know whether her midbrain signs were a result of chronic herniation and compartmentalised pressure, or whether they were due to a strep. viridans encephalitis which complicated her reservoir and shunt revision.

Case Number — 52

Name — M.M.

Age — 3 months

Method — Cannulation via anterior fontanelle

Medical Diagnostic Background

Congenital toxoplasmosis.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 3 hours

Indication The assessment of suspected active neonatal hydrocephalus (an abnormal increase in OFC, suturral separation, full anterior fontanelle, large areas of coroido-retinitis and a gradual loss of fixation and following).

Resting Ventricular Pressure

8.5 mm Hg.

Stress Ventricular Pressure

80 mm Hg maximum stress on crying.

Result Low or normal ventricular pressure

Cardiac/Respiratory Artefact

CR = 4 mm at rest.

OFC 45.5 cms.

Ventricular Dilatation/Cortical Mantle

Ventricular dilatation and aqueduct stenosis.

Points of Interest

In retrospect, this pressure monitoring was done for too short a period and it was carried out after a C.S.F. tap. Three days later, however, it was performed again and on this occasion the mean ventricular pressure was 22 mm Hg, and very high responses to stress, such as crying. Action on this occasion was open drainage and a ventriculo-peritoneal shunt. Since this he has developed epilepsy; no doubt a result of his congenital toxo-plasmosis.

This child was intolerant to low pressure levels and responded to it with vomiting and pallor.

A comparison was done of the ventricular pressure levels of this child with a fontanometer, in an attempt to quantitate some fontanometry recordings. The results were not conclusive and have not been included here.

Case Number 53

Name J.MacM.

Age 11 years

Method Ventricular cannulation via burr hole.

Medical Diagnostic Background

Myelomeningocoele and hydrocephalus.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration $3\frac{1}{2}$ hours

Indication Presentation with headaches and vomiting.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

50 mm Hg maximum.

Result Normal level ventricular pressure and the 'in situ' valve functioning. Discharged with an order to pump the valve 50 times three times a day.

Cardiac/Respiratory Artefact

CR = 5 mm at low pressure.

CR = 7.5 mm at high pressure in the awake state.

Ventricular Dilatation/Cortical Mantle

Moderately severe ventricular dilatation.

Points of Interest His presentation with headaches and vomiting was presumed to be due to an intercurrent infection.

Case Number	54
Name	N.B.
Age	9 years
Method	Rickham reservoir

Medical Diagnostic Background

This child was born with no family history of congenital malformations and following a normal pregnancy. She was an assisted breech delivery and weighed 6 lbs 5 ozs. She had an active neurological level of $L_{3/4}$ in the lower limbs, a result of a lumbar myelomeningocoele, which was closed within the first 12 hours. Within a week there was evidence of progressive hydrocephalus, the OFC at birth being 35.2 cms and at one week 37.5 cms. Because of a staph. ventriculitis and a persistent raised C.S.F. protein, a left sided Pudenz ventriculo-peritoneal shunt was not inserted until 7 months of age. This has required one revision.

Temperature	Normal
Zero	$1\frac{1}{2}$" above upper cortical subarachnoid space.
Duration	1 hour
Indication	A 3 week history of intermittent headaches

accompanied by nausea and vomiting on occasions. We did learn that one month prior to admission she had fallen at school and struck the left side of her head. At this time a lump came up above her valve chamber, but it subsequently subsided. The headaches and nausea continued however and she suffered two short lived grand mal fits. Examination suggested that she may have some hyperaemia to her fundi, and the cap of her Pudenz system was quite distended. The drainage tube from her shunt was difficult to feel in the mastoid area, suggesting that it had either become disconnected or buried in bone. C.S.F. sampling revealed microscopically clear and sterile fluid.

Resting Ventricular Pressure

20 mm Hg.

Stress Ventricular Pressure

Plateaus of 60 mm Hg level.

Result Raised ventricular pressure

Action A small amount of C.S.F. was drained, which returned the pressure to within normal limits.

Cardiac/Respiratory Artefact

C = 9 mm, R = 22 mm during plateaus.

Points of Interest At the revision operation, the shunt was found disconnected in the mastoid region. This is a vulnerable area from the point of view of head injury, in children with shunts. The subcutaneous catheter had fractures about 3" down from the Pudenz valve. Both ends were therefore removed and a new non-kinkable catheter was introduced and fixed in the previous site. Since that time her mother reports a great improvement in her mobility, and the teachers reported that her school performance was dramatically improved. In retrospect her mother thought that she had had these symptoms of chronic low grade pressure for some time and that one week prior to admission her behaviour had deteriorated markedly.

Pressure Recordings There was a massive increase in ventricular pressure shortly after this tracing was commenced, from 20-60 mm Hg, while the child remained conscious and showed no visible change in vital signs. Hyperventilation response (Fig. 109) commencing at the first arrow, ceased at the second arrow, reduces the pressure again to 20 mm Hg. But within a few seconds, the pressure began to rise again and within $1\frac{3}{4}$ minutes was back to a level of 45 mm Hg. At that time 5 mls of C.S.F. was released, which reduced the pressure to about 9 mm Hg. So although the hyperventilation is an effective short term treatment, it must be continued.

Case Number 55

Name M.M.

Age 6 months

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child suffered a neonatal intracranial haemorrhage and was hypertonic, unresponsive, opisthotonic and fitted in the neonatal period. An A.E.G. showed the third and fourth ventricles to be dilated, as well as the lateral ventricles. Air passed into the cisterna magna. Thereafter the OFC increased at a normal rate, and the child was thought to be spontaneously arresting. However, a rapid increase in OFC over 2 weeks necessitated this admission.

Temperature Normal

Zero Inter-ventricular foramina level

Duration $4\frac{1}{2}$ hours

Indication A rapid increase in OFC over 2 weeks, with vomiting and sleeping poorly. Examination revealed some distension in the scalp veins, slight 'sunsetting' and a full anterior fontanelle.

Resting Ventricular Pressure

27 mm Hg.

Stress Ventricular Pressure

80 mm Hg maximum.

Result Raised ventricular pressure

Action A ventriculo-peritoneal shunt and Rickham reservoir the following day.

Cardiac/Respiratory Artefact

CR = 10 mm in sleep (at 30 mm Hg).

CR = 6.25 mm (at 30 mm Hg) awake.

OFC 51 cms.

Ventricular Dilatation/Cortical Mantle

Moderately severe communicating hydrocephalus.

Pressure Recordings Fig. 110 shows 3 sections of a complete awake-sleep-awake cycle in this child. The upper figure begins with a treatment, then sustained elevation of pressure, and during this time there is rapid eye movement, lip movements, sucking, grinning and finger twitching. A 'sigh' tends to initiate 'quieter' sleep (the last negative deflection in the upper tracing). The middle tracing is of deep sleep and like the other two, lasts $7\frac{2}{3}$ minutes in this child. The lower tracing again shows sucking, lip movements, eye rolling, grinning etc.

Case Number 56

Name L.G.

Age 8 years 3 months

Method Ventricular cannulation via right posterior parietal Rickham.

Medical Diagnostic Background

Lumbo-sacral myelomeningocoele with hydrocephalus and a Pudenz ventriculo-peritoneal shunt in situ.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 7 hours

Indication Admitted with a leaking distended 'back lesion'. This was in the site of her original myelomeningocoele and acts as a 'safety valve' for her during periods of increased intracranial pressure. She therefore had pressure monitoring and a

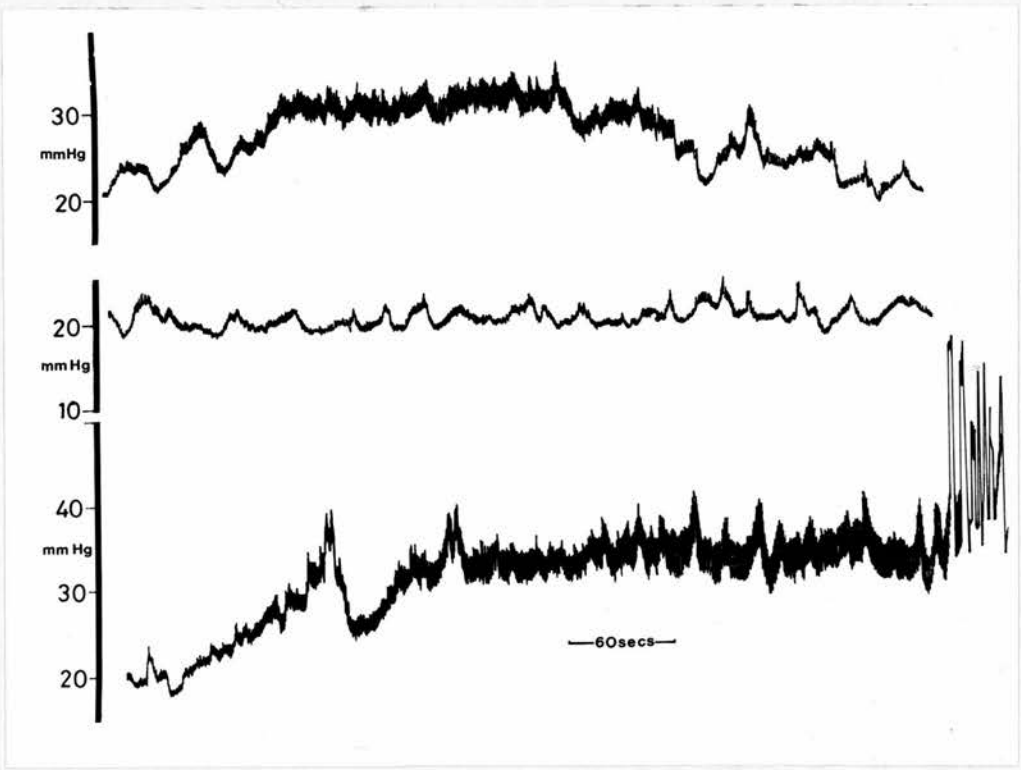


Fig.110

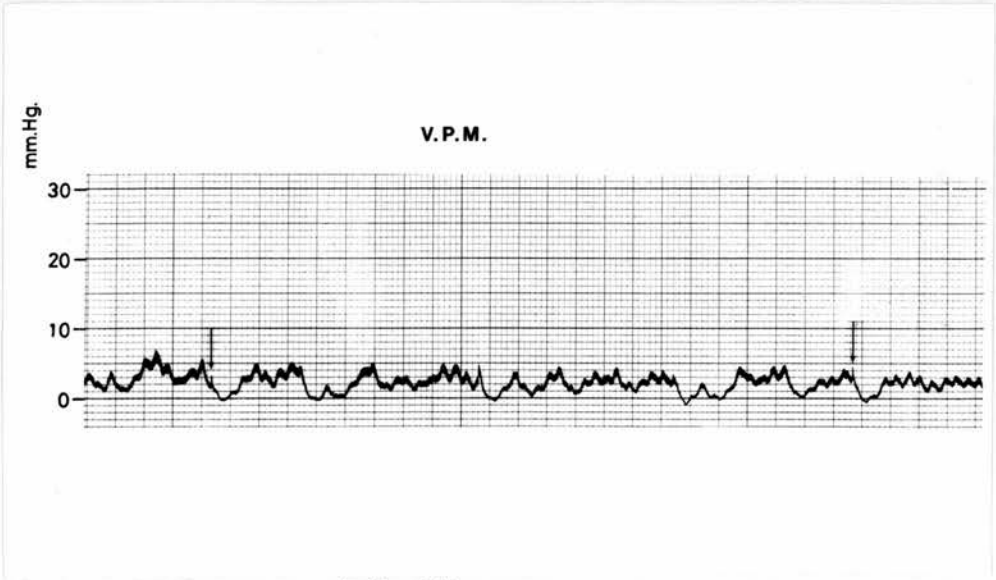


Fig.111

C.T. scan.

Resting Ventricular Pressure

28 mm Hg.

Stress Ventricular Pressure

72 mm Hg on crying. 50-55 mm Hg peaks in 'light' sleep.

Result Raised ventricular pressure

Action A shunt revision, and plastic repair of her 'leaking back lesion'.

Cardiac/Respiratory Artefact

CR = 13 mm awake.

CR = 17 mm asleep

Ventricular Dilatation/Cortical Mantle

Communicating hydrocephalus of moderate severity.

Points of Interest The fact that she is able to decompress herself by distension of the cystic lesion on her back means that she remains asymptomatic during periods of raised ventricular pressure. However, because of the leak from the lesion, she is at risk from the spinal meningitis.

Case Number 57

Name D.L.

Age 9 weeks

Method Right temporal Rickham reservoir.

Medical Diagnostic Background

This child developed E. coli meningitis, ventriculitis and septicaemia at 6 weeks of age.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 9 hours

Indication Following in the course of treatment of his ventriculitis, the OFC began rising. A Rickham reservoir was inserted so that antibiotics could be given intra-ventricularly, pressure monitoring carried out. At the time of monitoring, he had had no tap of C.S.F. or intra-ventricular antibiotics for 24 hours. However, the C.S.F. cell count 3 days earlier had shown 80 leucocytes per cu.mm. The indication here was therefore the assessment of suspected active neonatal, post-meningitic hydrocephalus.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

50 mm Hg. (During sleep maximum pressure of 22 mm Hg, after Phenobarbitone sleep peaks of 40 mm Hg.)

Result Low awake R.V.P., but moderately raised pressure during sleep.

Action He had a ventriculo-peritoneal shunt inserted.

Cardiac/Respiratory Artefact

C = 3.75 mm, R = 7.5 mm in sleep.

CR = 1 mm awake.

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed symmetrical dilatation of all four ventricles.

Points of Interest Although this child's resting ventricular pressure was within normal limits, he had been 'tapped' of the C.S.F. 24 hours previously.

An alternative management would have been to continue to tap his Rickham as necessary for up to even 6 weeks, and not to insert a ventriculo-peritoneal shunt at that early time.

It is interesting that he has had a number of shunt revisions for blockage and at least one episode of ventriculitis since that time.

Pressure Recordings Fig. 111 shows a section of light sleep with a distorted 'saw tooth' pattern which appears commonly in states of post-meningitic hydrocephalus, where prolonged periods of R.E.M. sleep appear to occur.

15 mgms of Phenobarbitone was given to this child during the course of his pressure monitoring, because he became restless, and this resulted in 'quiet sleep' pressure waves in 30 minutes. Later, as his R.E.M. phase appeared (some $2\frac{3}{4}$ hours after the Phenobarbitone), an elevation of ventricular pressure occurred (Fig.112) and this reached a mean level, at the arrow, of 28 mm Hg and was of short duration. During this R.E.M. period of sleep there was evidence of lip smacking. It is interesting that in the first Figure, prior to any sedation, sighing responses, indicated by the arrows, again characteristically appear on the downward slope of the rhythmical sleep-pressure oscillations.

Case Number 58

Name E.H.

Age 24 hours

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Lumbar myelomeningocoele with hydrocephalus at birth. She was also 'small for dates' and showed brain-stem fits in the first few days.

Temperature Normal

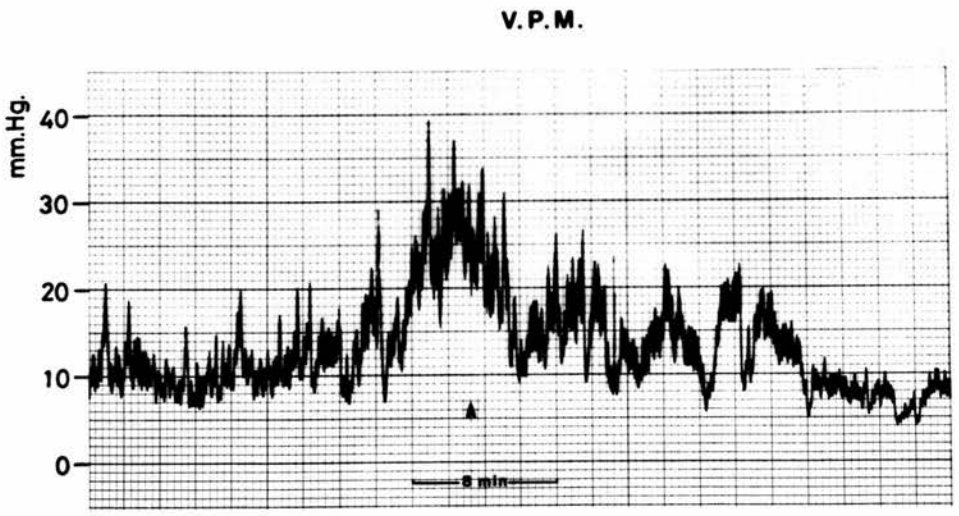


Fig.112

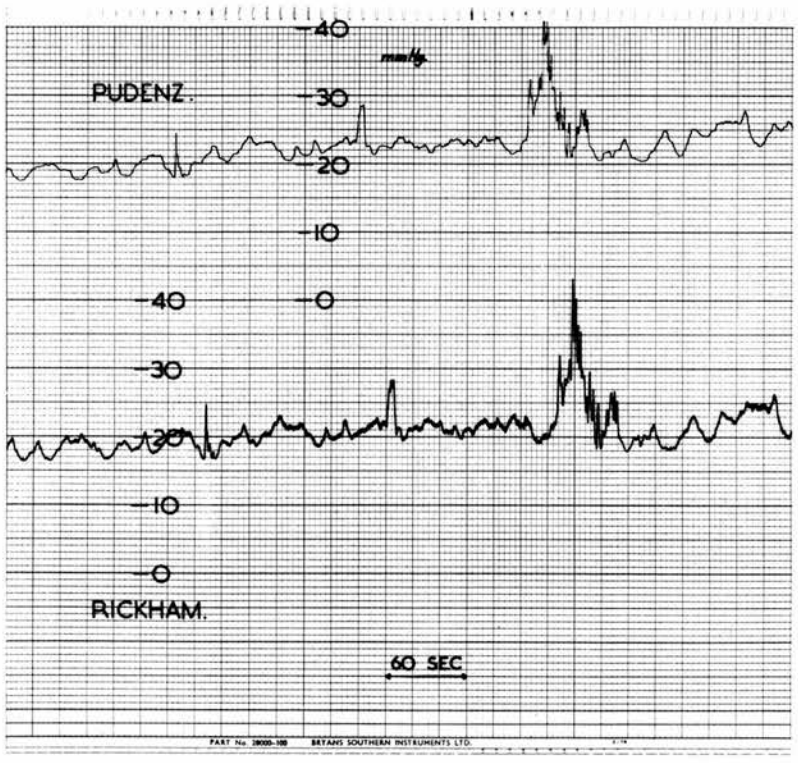


Fig.113

Zero — Inter-ventricular foramina level.

Duration — 1 hour

Indication — To assess the extent of hydrocephalus and raised intracranial pressure within the first 24 hours of life in a child with spina bifida, whose neurological level was borderline, re selection, i.e. a neurological level of L_2 bilaterally.

Resting Ventricular Pressure

4 mm Hg.

Stress Ventricular Pressure

24 mm Hg.

Result — Normal ventricular pressure

Action — No action re C.S.F. pressure

Cardiac/Respiratory Artefact

CR = 0.75 mm

OFC — 31 cms.

Ventricular Dilatation/Cortical Mantle

A.E.G. showed gross dilatation of all ventricles and a cortical mantle of 10 mm.

Follow Up — The pressure was low both because of the gross ventricular dilatation and because of the distensible leaking myelomeningocoele sac.

The child died shortly afterwards and at autopsy the main cerebral findings were of polymicrogyria, hydrocephalus and an associated cerebellar malformation.

Case Number — 59

Name — M. McC.

Age — 16 days

Method — Ventricular cannulation via anterior

fontanelle.

Medical Diagnostic Background

Myelomeningocoele with a neurological level of S_1 motor and sensory in the lower limbs.

Temperature 37

Zero Inter-ventricular foramina level

Duration Half an hour

Indication The assessment of suspected active neonatal hydrocephalus (a tense anterior fontanelle, scalp venous distension, an increasing OFC). He continued to feed well with no vomiting.

Resting Ventricular Pressure

22 mm Hg.

Result Raised ventricular pressure

Action Ventriculo-peritoneal shunt inserted.

Cardiac/Respiratory Artefact

CR = 1 mm at rest.

OFC 39.7 cms at the time of V.P.M.

35 cms at birth.

Ventricular Dilatation/Cortical Mantle

Moderate dilatation of the lateral and third ventricles, aqueduct obstruction and a cortical mantle of 17 mm at time of V.P.M.

Points of Interest It is interesting that this child had a short period of pressure monitoring, during the course of a fontanelle puncture 11 days previously and at that time the R.V.P. was 6.6 mm Hg, with a maximum stress on crying of 25 mm Hg, and a cortical mantle of 19.5 mm. It seems reasonable not to do invasive investigations too early, on active neonatal hydrocephalus, both because the hydrocephalus itself may arrest, and secondly, if the pressures are

TABLE 11

Case Number 59 59

Day	OFC	
1	35 cms	
2		
3	36	
4		
5	36.5	
6	36*	Ventricular pressure normal, anterior fontanelle tense.
7		
8	37.4	
9	37.5	
10	38	Signs decerebration with fisting, cycling etc.
11	38	
12		
13	38.4	
14	38.5	
15	39	
16	39.5	Scalp vein distension.
17	39.7	Ventricular pressure raised
18		
19	40	Anterior fontanelle full
20	40.7	Anterior fontanelle tense, veins distended, feeding well.
21		
22	41	Anterior fontanelle tense, veins distended.
23	41.1	'sunsetting' and ventricular 'tap'
24	41*	Anterior fontanelle full
25	41.5	Anterior fontanelle full
26		Shunt inserted.

An overall increase of 6.5 cms or mean increase of 2.5 mm per day.

An increase of 3.4 cms over 13 days

An increase of 3 cms over next 13 days

*may have been inter-observer error.

found to be within normal limits, and the child continues to be symptomatic, i.e. the OFC continues to increase, or he vomits, 'sunsets' etc., then the procedure will need to be repeated later. The optimal time is not clear, but when the head size is normal at birth, it seems reasonable to wait until definite relief of pressure by tapping is necessary, before measuring the pressure. The ideal management is discussed in Chapter II.

Relief of 17 mls of C.S.F. from this child resulted in a pressure drop to 5.5 mm Hg, that is, 1 cc of C.S.F. release was equivalent to lowering the ventricular pressure 1 mm Hg at this pressure level. Table 11 shows the incremental changes in OFC over the first 26 days of life in this child. The mean daily increase in OFC was 2.5 mm and the increase was continual and not in a step-wise fashion as in a previous case.

Case Number	60
Name	G.M.
Age	4 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was born by a normal vertex delivery with good 'apgars'. She weighed 1.90 Kg at an estimated gestational age of 24 weeks. Apart from mild jaundice in the neonatal period, there was no R.D.S. or apnoea to suggest the possibility of any intracranial bleed. At the time of discharge at 4 weeks her OFC had increased to just over + 2 standard deviations from the mean. From $2\frac{1}{2}$ to 3 months of age there had been a rapid increase in the head size, her OFC at 3 months of age then measuring 45.2 cms. She was irritable,

unresponsive, not fixing or following, she was slightly dystonic with brisk primitive and monosynaptic reflexes. She was also rather wasted, but there was no other system abnormality. C.S.F. was obtained from the ventricle at 15 mms, and an L.A.E.G. showed a gross communicating hydrocephalus. Accordingly, a Pudenz ventriculo-peritoneal shunt was inserted. Following this she still required the valve to be pumped.

Temperature 37.5

Zero Inter-ventricular foramina level.

Duration $3\frac{1}{2}$ hours.

Indication Persistent increase in the OFC and symptomatic, in that she was 'sunsetting' as much as before her operation and the fontanelles were tense. One might have expected her OFC to increase at a less rapid rate, but certainly did not expect 'sunsetting' and tension in her fontanelles.

Resting Ventricular Pressure

21 mm Hg.

Stress Ventricular Pressure

A maximum peak on crying of 55 mm Hg and maximum peaks during sleep of 33 mm Hg.

Result Raised ventricular pressure. On pumping the Pudenz valve, this produced a drop in ventricular pressure equivalent to 1 mm Hg for 10 pumps of the valve, i.e. it appeared to be inefficient. No obvious compartmental differences were seen when the pressure was measured from the Pudenz and the contralateral Rickham. This suggested a distal block of her catheter, which was revised, and following this her shunt worked well and did not require pumping. She was also more alert, less 'sunsetting', less dystonic and feeding well.

Ventricular Dilatation/Cortical Mantle

Severe communicating hydrocephalus, a cortical mantle of 15 mm at the vertex, and less than 5 cms at the occiput.

Points of Interest Having established that her pressure was raised, despite her shunt in situ, the possibility still existed that one ventricle only was being shunted, and thus her signs could have been attributable to that. Measuring the pressure on both sides eliminated this as a possible cause. Furthermore, being able to measure the pressure through the Pudenz, meant that the proximal limb was almost certainly patent, and therefore a distal revision was performed.

Pressure Recordings Early sleep recordings again showed a characteristic waking and dozing pattern with a fluctuating base line. But the amplitude of these sleep waves is certainly diminished, in cases of gross hydrocephalus, in neonates and young infants, compared to the older child. Again the early prolonged R.E.M. sleep is punctuated by sighs. Fig. 113 shows a twin recording from a Pudenz, and contralateral Rickham reservoir, at the same time. Similar pressure levels exclude compartmental block but there is a minimal damping effect on pulse and respiratory components in the tracing from the Pudenz compared to that from the Rickham.

Case Number	61
Name	T.A.S.
Age	2 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was born with severe bilateral cephalo-haematomas and a

large head. A C.T. scan showed a massive increase in the size of the ventricles. The cephalo-haematomas at the time of admission were rock hard and there was some calcification in the one on the left which measured 8 x 3 cms.

Temperature 37.1

Zero Inter-ventricular foramina level.

Duration 4 hours

Indication The assessment of active neonatal hydrocephalus with a large head, 'sunsetting' tense anterior fontanelle, increase in scalp venous distension and pallor of the optic discs. It is significant that this child was feeding by the breast extremely well and had no vomiting, and nursing observations were satisfactory. The pupils were sluggishly reactive and routine investigations such as subdural taps, antenatal virology etc. were normal.

Resting Ventricular Pressure

22 mm Hg.

Stress Ventricular Pressure

A maximum peak of 42 mm Hg was recorded on crying and a maximum peak of 34 mm Hg during sleep.

Cerebral Perfusion Pressure

75.5 mm Hg at the outset. 70 mm Hg during sleep.

Result Raised ventricular C.S.F. pressure.

Action Insertion of a C.S.F. shunting device, but initially 10 mls of C.S.F. were removed, which reduced the ventricular pressure to 13 mm Hg. A further 10 mls of C.S.F. removal dropped the ventricular pressure level to 10 mm Hg. With a further 5 mls of C.S.F. release it dropped to 9 mm Hg and with

a further 5 mls release, it remained at 9 mm Hg, that is, the ventricular pressure tended to reach a 'steady state' at the end of the recording when sufficient C.S.F. had been removed. The C.P.P. was then 101 mm Hg.

Cardiac/Respiratory Artefact

CR = 1.5 mm at rest. CR = 3 mm in sleep. After the first 10 mls of C.S.F. were removed, CR = 0.75 mm, thereafter CR too minute for measurement.

OFC 47.7 cms

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed gross dilatation of all four ventricles and no communication with the cephalo-haematoma.

Points of Interest

That the child should continue to feed particularly well and have no vomiting in the presence of markedly raised ventricular pressure was astonishing.

The increase in OFC initially in this child progressed in a step wise fashion, with a step wise increase in the signs as well, as previously mentioned.

It is well known that precipitate deliveries result in tearing of the Falx with intracranial haemorrhage, and subsequently raised intracranial pressure. Precipitate deliveries at the same time cause cephalo-haematomas frequently but these are not causally related to the intracranial pressure.

Congenital hydrocephalus however, results in cephalopelvic disproportion (which may cause 'overlaying of the sutures' and by an entrapment mechanism limiting C.S.F. absorption and aggravate I.C.P). Because of the large head, the cephalopelvic disproportion may cause cephalo-haematomata, which, when they are large, may limit expansion

of the skull and further exaggerate the I.C.P.

To speculate on a further mechanism: when cephalo-haematomata are large there is obviously a diminution in the superficial venous drainage from the head and neck and this induces R.I.C.P. in some individuals. The rationale of this is that R.I.C.P. results in scalp venous distension so that anything interfering with scalp vein distension (in the neonatal period) will limit the buffering available. (How much the distension of scalp veins contribute to this compensation in the newborn is unknown.)

This raises an important point because there are numerous babies born with varying sizes of cephalo-haematomas due to some sort of birth trauma, (and not as a result of a large head before birth), but I have not seen one case obviously result in hydrocephalus. However, no scans of ventricular size have been routinely done in these children to my knowledge, to detect minor degrees of ventricular dilatation.

I would suggest that in the normal baby, who, because of some birth trauma, develops cephalo-haematomas, the normal compensatory mechanisms are adequate to cope with any transient increase in I.C.P. occurring as a result of unilateral or bilateral cephalo-haematomas limiting skull expansion or interfering with venous drainage.

In the hydrocephalic baby, on the other hand, both because of the intra-uterine pressure before birth is greater than the I.C.P. (Schaller et al 1977) and because of cephalo-haematomas after birth, the pressure dynamics may be compromised even further. Therefore, with bi-parietal diameter and pelvimetry estimations done at term, the obstetrician should be persuaded to 'section' rather than do a 'trial of labour' on doubtful cases.

It would appear in this case that the congenital hydrocephalus with

cephalo-pelvic disproportion resulted in the cephalo-haematomata and I postulate that by limiting expansion of the skull, and by interfering with superficial venous drainage, the intracranial pressure was further exaggerated.

Case Number	62
Name	J.D.
Age	10 years
Method	Rickham reservoir

Medical Diagnostic Background

This child was born by S.V.D. at 10 days past term. The cord was about the neck twice and he was initially cyanosed and needed oxygen. Subsequently he was put into an incubator for several hours. His development thereafter was satisfactory, but at 5 months of age he developed meningitis (no details known). At 16 months he was noted at a baby clinic to have a large head and a C.S.F. shunt was inserted at 2 years of age. Since that time he has had 10 valve revisions and 4 burr holes. He was then lost to follow up from his previous place of supervision, and was referred on this occasion because of intermittent vomiting for $2\frac{1}{2}$ years! Investigations locally revealed no cause and the question of a blocked valve was raised.

Temperature	Normal
Zero	$1\frac{1}{2}$" above upper cortical subarachnoid space.
Duration	24 hours
Indication	Vomiting at irregular intervals. He could be sick for one week and then well for the next 2-3 weeks. The last episode was 2 weeks before he was admitted and the vomiting then was copious. He had also the complaint of intermittent headache,

approximately once every 2 weeks. They lasted 2-3 hours and were relieved by sleep. The headaches were not especially associated with vomiting or photophobia and tended to be frontal in nature and there was no aggravating phenomena or radiation. No visual disturbances, hearing problems or fits were reported. There was however a suggestion that his right hemiplegia was becoming a little worse. His mother reported some 'temper tantrums'. Examination revealed an OFC of 58.3 cms, but no other abnormality. Insertion of a Rickham reservoir was undertaken electively so that his pressure could be monitored. At the time of insertion, the frontal horn of the right lateral ventricle was entered at a depth of 3 cms. The picture was confusing because the skull x-ray also showed some calcification present on the surface of the brain, probably the result of old infection.

An E.E.G. at the time showed some moderate global abnormalities. The record was dominated by rather irregular theta activity, larger in amplitude on the right side, and widespread maximally anteriorly, and the asymmetry was less evident in the transverse derivations.

In summary, the indication was of vomiting irregularly for 2 years, frequent headaches, some behavioural and emotional deterioration and a worsening of his right hemiplegia.

Resting Ventricular Pressure

4 mm Hg in the awake state.

Stress Ventricular Pressure

Peak ventricular pressure was 42 mm Hg during sleep.

Result The sleep pressures were unsatisfactory. It was felt that he was experiencing intermittent pressure problems.

Action A reconstitution of a left sided ventriculo-atrial anastomosis into a ventriculo-peritoneal anastomosis using the

original low pressure Spitz Holter pump and valve, was undertaken.

Cardiac/Respiratory Artefact

Not applicable in this case as the recordings were performed on the 'alternative apparatus'.

OFC 58.3 cms

Follow up Following this valve revision, he developed a mild ventriculitis and accordingly had that shunt replaced by a Pudenz system. Again he presented with diplopia, frontal headaches, vomiting and 3 short 'fits' and required a further revision of the Pudenz system. Since that time he has remained very well.

Pressure Recordings Fig. 114 shows a full 24 hour record. The time markers at the top of this illustration are in 2 hourly intervals. One can see a fluctuating pressure level during sleep and a marked elevation prior to waking. At this time there is a drop in the heart rate. A mean pressure level of 15 mm Hg occurs during the first half of this tracing, but a mean of 20 mm Hg occurs prior to wakening. The maximum peak of pressure oscillations during sleep reached in excess of 42 mm Hg. Note also that the amplitude of the pressure fluctuations is greatest when the level of ventricular pressure is elevated. Fig. 115 and Fig. 116 were tracings obtained at paper speeds, faster than real time, and were obtained during early sleep when the pressure fluctuations oscillate, and it can be seen that the pressure fluctuations extend from 20-35 mm Hg at its maximum, and then begins to decrease. The timing of the 'peak' of these oscillations can also be seen to follow the T wave in the E.C.G. tracing (Bering 1955), that is, in diastole, and is consistently at its lowest during systole. At these speeds the E.E.G. becomes more difficult to interpret. Fig. 117 at an even faster paper speed again shows a maximum peak of the ventricular pressure waves occurring

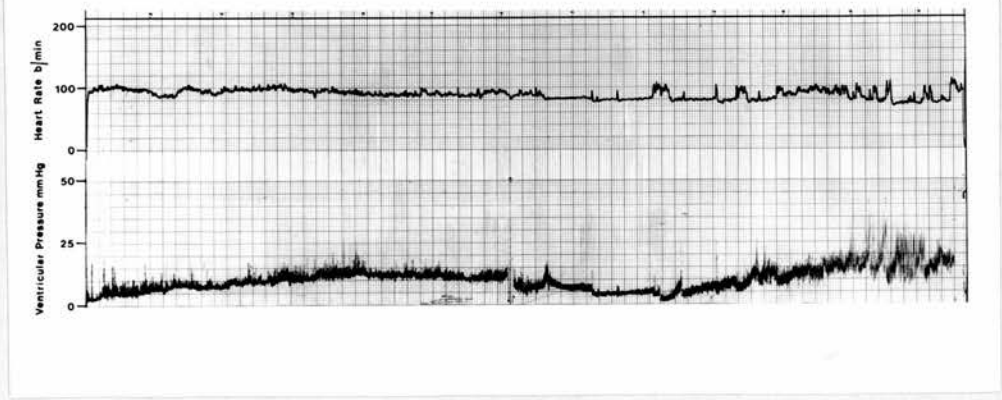


Fig.114

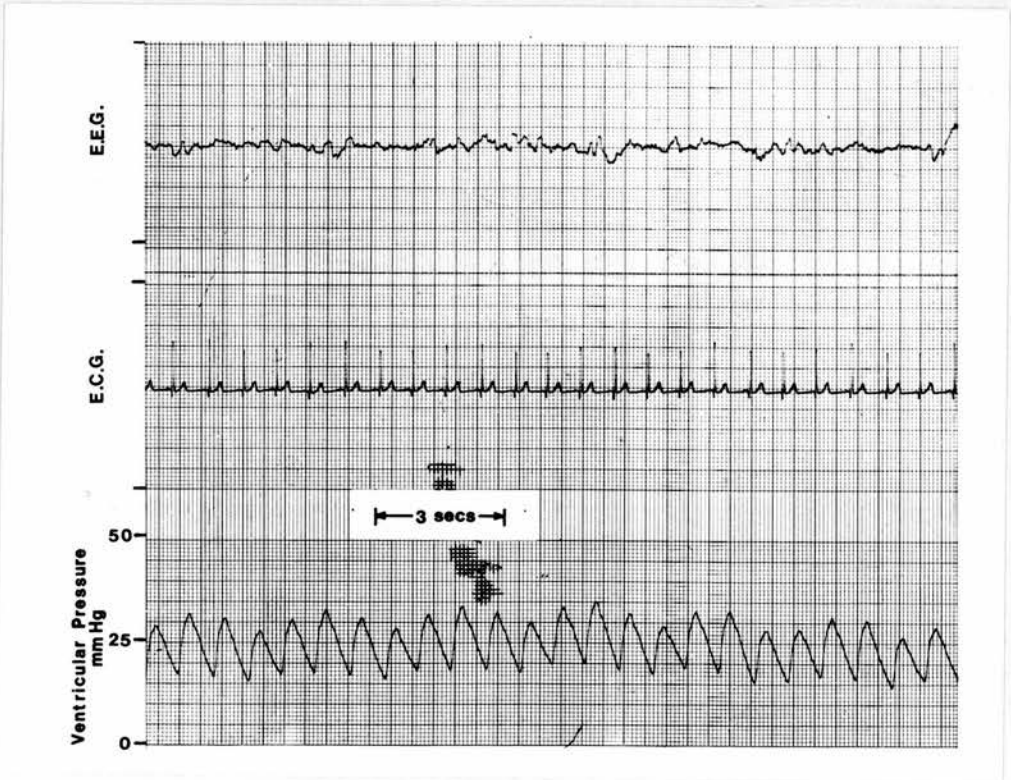


Fig.115

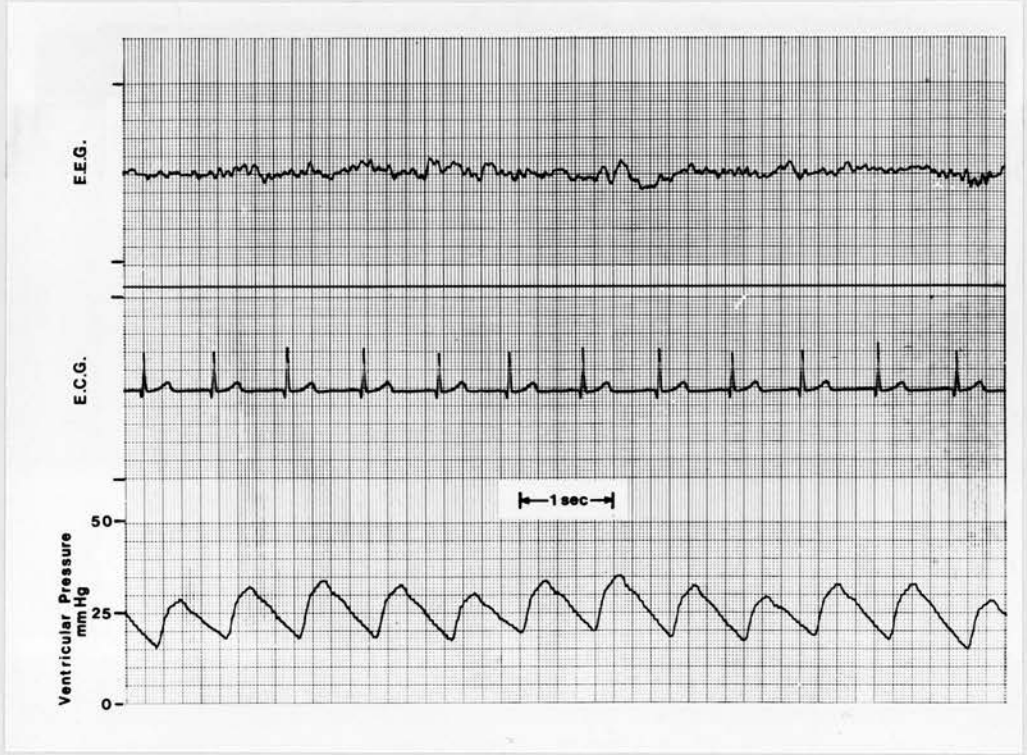


Fig.116

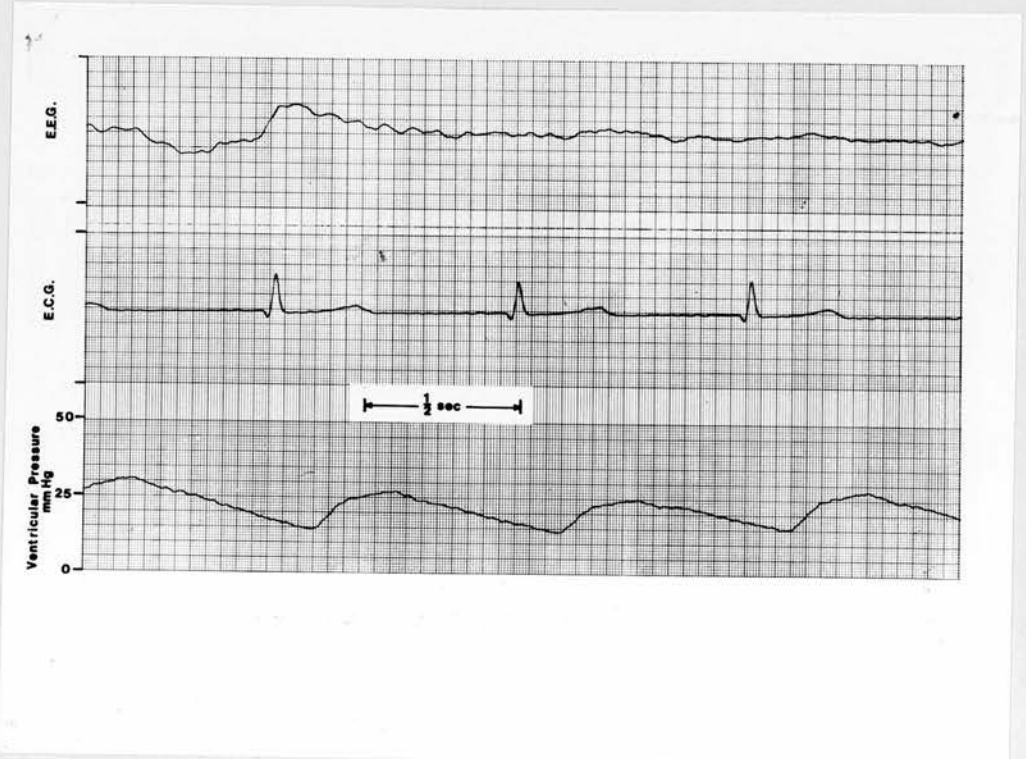


Fig.117

after the T waves of the E.C.G., and this must maximalise the venous outflow from the head, because in pathological conditions, such as an arteriovenous malformation of the hind brain, there is limited C.S.F. absorption into the sagittal sinus, when the pulse wave, at its maximum, does not occur during atrial diastole (De Lange and De Vlieger 1970). A further Fig. 118 is obtained also at faster than real time, when these pressure changes in sleep are at their lowest point. During this phase of early sleep, the child begins to snore, shows eye movements, myoclonic movements of his little finger, noisy respiration and lip smacking. It can be seen on this E.C.G. tracing that there is a slight irregularity, compared to the previous tracing. Where the pulse rate was 75 per minute, now at the lower level of pressure, the pulse rate is about 66 per minute. When one looks at an overall Fig. 119 of this early phase of sleep with the pressure oscillations, one can see in the E.E.G. tracing, at points indicated by the arrow, sleep spindles are occurring. Although they often occur at the height of the pressure wave, it is not a constant relationship. This section of tracing was obtained at about 10.40 p.m. not long after sleep had ensued. Towards the end of one of the periods of R.E.M. sleep, one can see in Fig. 120 apart from the pressure fluctuations, there is an E.C.G. tracing abnormality (compare with the previous respiratory tracing abnormalities during this phase of sleep) and it is associated with a prolongation of the falling ventricular pressure wave. A similar abnormality not shown here, occurs a few seconds later.

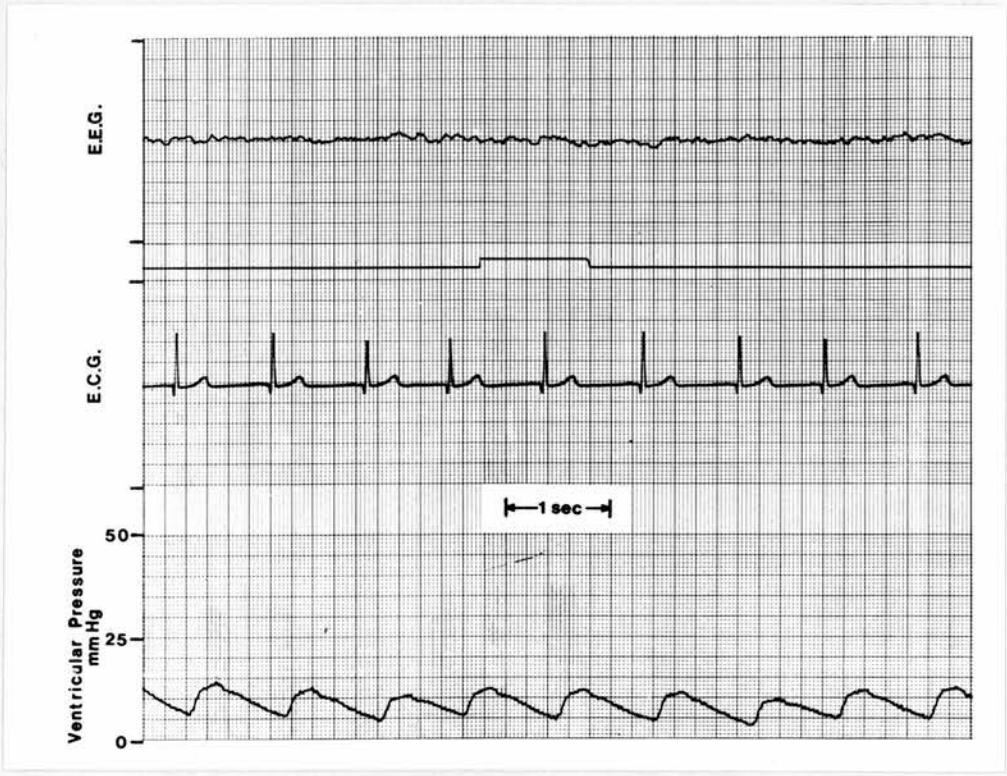


Fig.118

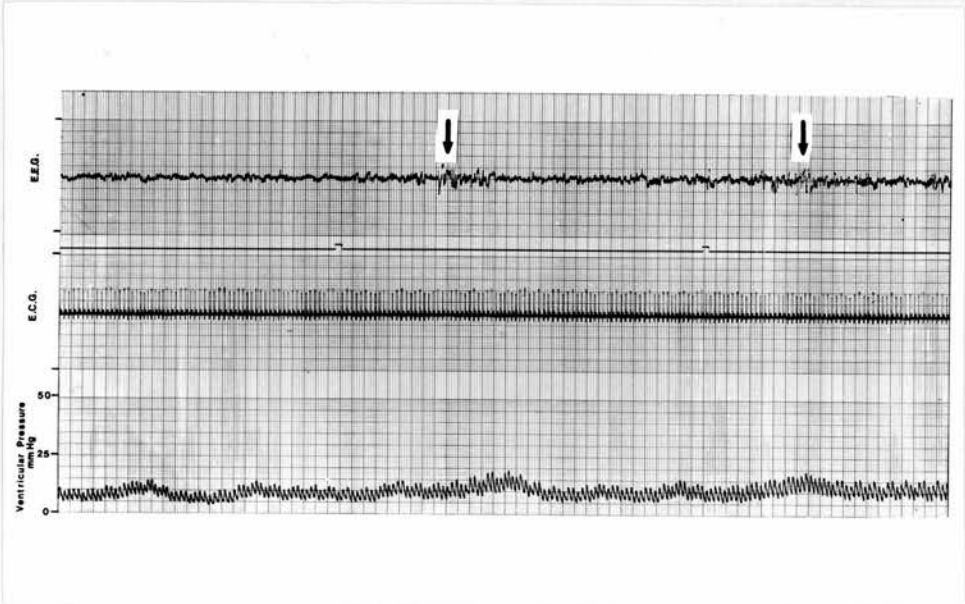


Fig.119

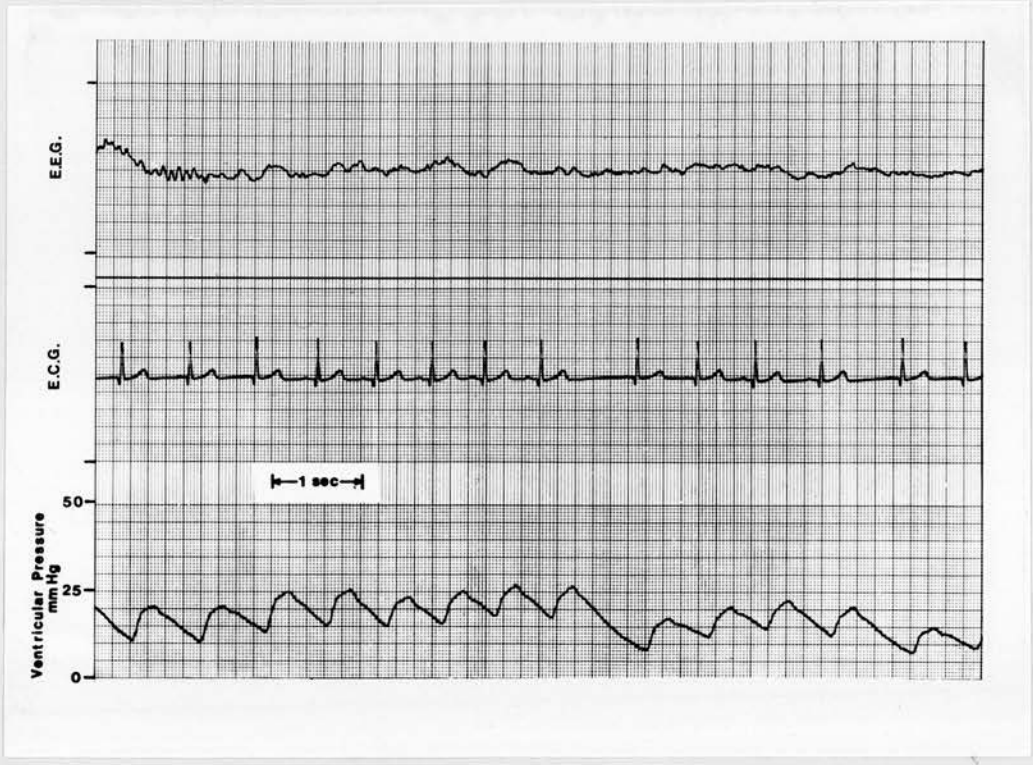


Fig.120

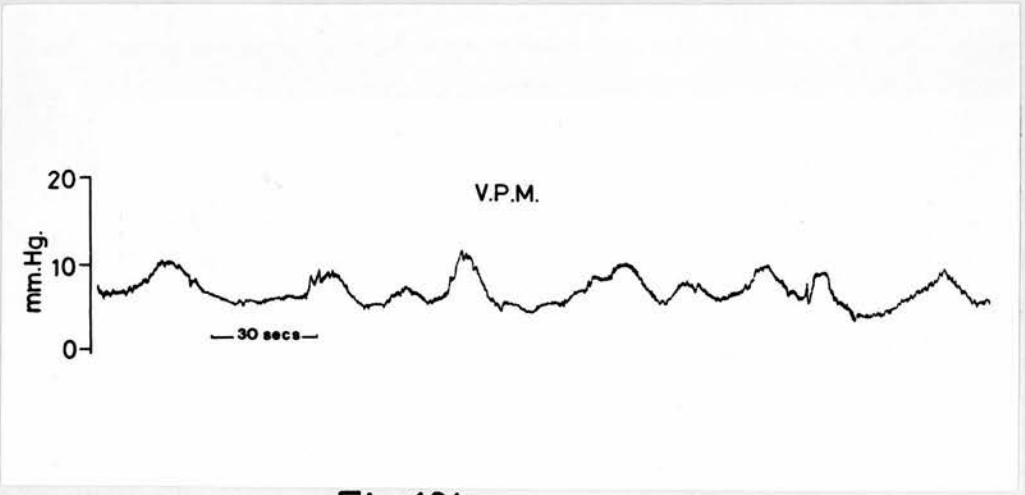


Fig.121

Case Number 63

Name M.G.

Age $9\frac{1}{2}$ years

Method Left frontal Rickham reservoir

Medical Diagnostic Background

This boy was diagnosed as subacute sclerosing panencephalitis and was admitted in stage 2, with continuous myoclonic status, impaired consciousness, with evidence of spasticity and involuntary movements. The diagnosis was confirmed by the E.E.G. changes, a first zone rise in colloidal gold, an increased gamma globulins in the C.S.F., a high measles titre in the C.S.F. and serum and brain biopsy with fluorescent antibody studies. At the time of biopsies he had a left frontal Rickham reservoir inserted so that a course of Ara-A 5 phosphate and Ara-C could be given intraventricularly, at the same time as a 10 day course of intravenous Ara-A.

Temperature Normal

Zero Upper cortical subarachnoid space.

Duration 12 hours

Indication He had developed a tense swelling over his biopsy sites, was hypoventilating with metronomic myoclonus, and becoming marasmic. He had no evidence of papilloedema or neck stiffness.

Resting Ventricular Pressure

30 mm Hg.

Result Raised ventricular pressure.

Action Frequent tapping, prior to a definitive ventriculo-peritoneal shunt being inserted.

Cardiac/Respiratory Artefact

C = 1.5 mm.

R = 5 mm when the ventricular pressure was greater than 30 mm Hg.

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed moderate dilatation of the ventricular system.

Points of Interest The hydrocephalus and raised ventricular pressure is not a consistent feature of S.S.P.E., and although the C.S.F. 'blow-out' over his biopsy sites were quite large, the biopsies themselves were full thickness and extended to the ventricle. It seems more likely in this child, that the origin of his hydrocephalus and pressure was from the intra-ventricular therapy resulting in a chemical arachnoiditis. It should also be mentioned that he developed a ventriculitis following the insertion of his reservoir. However, when his homeostasis had settled to his pre-operative state, a further course of Ara-C again resulted in increased myoclonic jerks, pronounced opisthotonous and decerebrate posturing, and C.S.F. samples showed that he had suffered an intra-ventricular haemorrhage.

Pressure Recordings Fig. 56 is seen in Chapter 7 showing the fluctuations of ventricular pressure at a mean raised level. These fluctuations correspond with the burst-suppression of the E.E.G. It is interesting that 50 mls of 20% Manitol, given intravenously, resulted in very little reduction of his ventricular pressure. Furthermore, pressure over the posterior swelling produced a deflection from 12 - 32 mm Hg (this was after he had been tapped regularly).

Case Number	64
Name	L.G.
Age	6 weeks
Method	Right parietal Rickham reservoir

Medical Diagnostic Background

Myelomeningocoele and hydrocephalus.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 1 hour

Indication — The assessment of suspected active neonatal hydrocephalus. After closure of the myelomeningocoele, she developed a wound infection necessitating treatment with antibiotics, for extension of the infection from the spinal to the ventricular system. Therefore a Rickham reservoir was inserted and pressure monitoring was carried out at this stage, when her OFC was increasing and she had developed neurogenic stridor from vocal chord paresis.

Resting Ventricular Pressure

25 mm Hg.

Stress Ventricular Pressure

Maximum peaks of 66 mm Hg.

Result — Raised ventricular pressure

Action — A ventriculo-peritoneal shunt was inserted.

Cardiac/Respiratory Artefact

CR = 8.75 mm awake and sucking.

CR = 3.75 mm at lowest pressure.

OFC — 37.5 cms at time of V.P.M.

33.5 cms at birth.

Ventricular Dilatation/Cortical Mantle

Moderately dilated ventricles and a cortical mantle of 3 cms at the vertex.

Case Number	65
Name	K.M.
Age	$5\frac{1}{2}$ months
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

This child was born at 37 weeks gestation by S.V.D. There had been 2 episodes of threatened abortion in early pregnancy but it was otherwise uneventful. The second stage of delivery was apparently quite rapid and there was a lot of bruising about the head. She also required phototherapy for neonatal jaundice. The only abnormality on neonatal examination was two umbilical vessels. At routine follow up, it was noticed that her OFC was increasing too rapidly. It had been 33 cms at birth but at 12 weeks of age, was 44 cms, i.e. it had moved from just above the tenth percentile to well above the 90th percentile. Routine investigations such as antenatal virology, chromosomes, subdural taps etc. were all negative. Other features of her examination at this time were marked suturral separation, a large anterior fontanelle which merged with the metopic suture, a large posterior fontanelle and head tilt to the left. A C.T. scan report read 'The lateral ventricles are grossly enlarged without displacement. The third ventricle is also enlarged and is in a normal midline position. The fourth ventricle is small. There is however a very large cisterna magna, clearly seen in communication. No parenchymal cerebral lesion is demonstrated and no surface lesion is seen. The appearances are those of a gross hydrocephalus. There may possibly be an element of aqueduct stenosis in this case'. A soluble iodine ventriculogram showed the same abnormalities and in view of this, it was decided to explore the posterior fossa and insert a Rickham reservoir. A posterior fossa

craniectomy was done and drainage of the asymmetric cisterna magna expansion on the right side and division of arachnoidal adhesions on the left side.

Indication — Congenital hydrocephalus with a large cisterna magna and adhesions in the posterior fossa which were divided, and it was now important to know if her pressure remained elevated and if shunting from the lateral ventricles would be required.

Temperature — Normal

Zero — Upper cortical subarachnoid space.

Duration — $2\frac{1}{2}$ hours

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

39 mm Hg maximum on crying.

Cerebral Perfusion Pressure

70 mm Hg.

Result — Normal level ventricular pressure.

Action — Close follow up with OFC measurements etc. She has done remarkably well with no signs of raised intracranial pressure and acceptable developmental progress.

Cardiac/Respiratory Artefact

CR = 0.75 mm awake.

CR = 1.25 mm asleep.

OFC — 44 cms.

Ventricular Dilatation/Cortical Mantle

Gross enlargement of the ventricle (scan report as before).

Pressure Recordings — Fig. 121 shows a recording, which is a

good example of light sleep, in a child with normal pressure. This child however took quite a long time to get into deep sleep, but at $4\frac{1}{2}$ months of age, there is still relatively more R.E.M. than non-R.E.M. sleep.

Case Number 66

Name F.C.

Age 9 days

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

The child was born with a myelomeningocoele, with a neurological level in the lower limbs of L_5 bilaterally. The lesion was closed shortly after birth and the OFC commenced increasing.

Temperature Normal

Zero Inter-ventricular foramina level

Duration $1\frac{1}{2}$ hours

Indication The assessment of active neonatal hydrocephalus (an increasing OFC, full anterior fontanelle).

OFC At birth 34 cms. At time of V.P.M. 35.8 cms.

Resting Ventricular Pressure

13 mm Hg.

Stress Ventricular Pressure

58 mm Hg maximum.

Result Equivocal ventricular pressure level.

Action Follow up, with twice weekly visits for OFC measurements. Following this, the OFC increased less rapidly and there were no other signs of pressure.

Cardiac/Respiratory Artefact

CR = 1.25 mm at rest.

Ventricular Dilatation/Cortical Mantle

Mild communicating ventricular dilatation.

Case Number	67
Name	F.G.
Age	$2\frac{1}{2}$ weeks
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Myelomeningocoele as before.

Temperature	Normal
Zero	Inter-ventricular foramina
Duration	4 hours
Indication	The assessment of suspected active neonatal hydrocephalus (a continuing increase in OFC but otherwise the child remained very well clinically).

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

58 mm Hg maximum peaks. 10 mm Hg in sleep.

Result	Normal ventricular pressure
Action	Close follow up and repeat C.T. scan.

Cardiac/Respiratory Artefact

CR = 0.75 mm awake.

CR = 2.5 mm asleep.

OFC	36.5 cms

Ventricular Dilatation/Cortical Mantle

A C.T. scan at 3 months of age showed the lateral ventricles considerably enlarged and the enlargement was fairly symmetrical. There was also considerable enlargement of the third ventricle but the fourth ventricle which was not too well displayed but was probably not grossly enlarged. No intrinsic brain lesion could be distinguished and there was some shrinkage of the cortex, particularly in the front half of the cranium and particularly in the inter-frontal region. The appearances indicated a moderately severe degree of hydrocephalus. At this time the OFC was 41.8 cms. At 4 months of age a C.T. scan was repeated and the appearances were virtually identical. At this point the OFC was 44 cms. At one year of age the OFC was 48.75 cms. When last seen at the age of 2 years 7 months, she was making splendid progress, with no neurological deterioration. At the time of V.P.M. the cortical mantle was 4 cms from an A.E.G. done at the time.

Points of Interest This child has obviously been spared a shunt without any neurological deterioration. However, it is important that follow up C.T. scans are performed, and if any diminution of the cortical mantle occurs over a period of time, then a reservoir should be inserted and further pressure monitoring carried out.

Case Number	68
Name	C.V.
Age	4 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This baby was born by lower segment caesarean section following a

normal pregnancy. Birth weight 3 Kg and the child was fostered from birth. He was admitted to hospital at 1 month of age with a history of irritability for the previous 2 days and vomiting every feed for 24 hours. The natural mother was a single Philippino woman and the caesarean section had been done because of disproportion and second stage delay. The OFC at birth was 33.5 cms. Foster parents had reported, apart from irritability and vomiting, that the child appeared to be 'very nervous' and 'jittery'.

On examination on admission the OFC was 40.5 cms, there was an asymmetrical enlargement of the head, the right temple region was distended more so than the left. The sutures were markedly separated, the anterior fontanelle measured 3 x 3 fingers breadth and was very full but not tense. The posterior fontanelle was 1 fingers breadth in diameter. There were no intracranial bruits or carotid bruits and no scalp venous distension. Trans-illumination showed a massive hydranencephaly or external hydrocephalus anteriorly, and fundoscopy revealed on the left an incomplete disc with whitened vessels similar to medullated nerve fibres. On the left, the fundal fields in the temporal region had a greenish irridescent pigmentation between the vessels. This was quite extensive. There was no evidence of choroiditis, choroidal sclerosis or negroid funal appearances. The anterior chamber, lens and iris were satisfactory. The baby was very 'jittery' and irritable and grossly dystonic with reduced popliteal antles, very brisk phasic reflexes, extensor plantar responses and bilateral clonus. The child could be lifted with finger traction alone. There was however symmetry of movement in the upper and lower limbs and the child sucked and cried strongly, had normal temperature, control etc.

An A.E.G. at the time showed the classical hydranencephaly with no cortical mantle and some brain tissue in the posterior fossa region. Investigations at this time showed normal calcium, magnesium, glucose, urea, electrolytes, osmolalities, C.S.F., glucose, protein, full blood count and on the skull x-ray there appeared convex swellings of both lateral aspects of the anterior fontanelle and the appearances suggested haematoma; there was wide suturral separation. There was no spinal lesion on x-ray. Chest x-ray was satisfactory. Antenatal virology was negative, C.S.F. was clear as was subdural fluid. Immunoglobulins were satisfactory. Subsequently the child had recurrent chest infections and developed a right inguinal hernia; a herniotomy was performed.

Temperature — Normal

Zero — Inter-ventricular foramina

Duration — 1 hour 20 minutes

Indication — The assessment of the child with suspected hydrocephalus, ocular abnormalities and hypertonicity bilaterally (Fig. 122), prior to a diagnosis of hydranencephaly being made.

Resting Ventricular Pressure

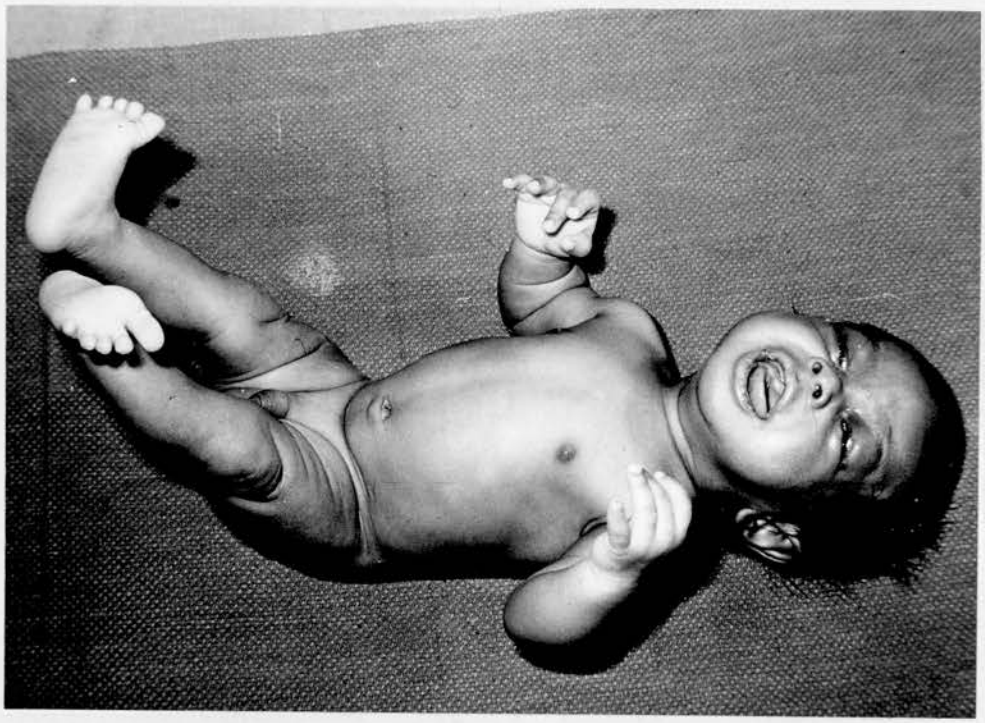
6.5 mm Hg.

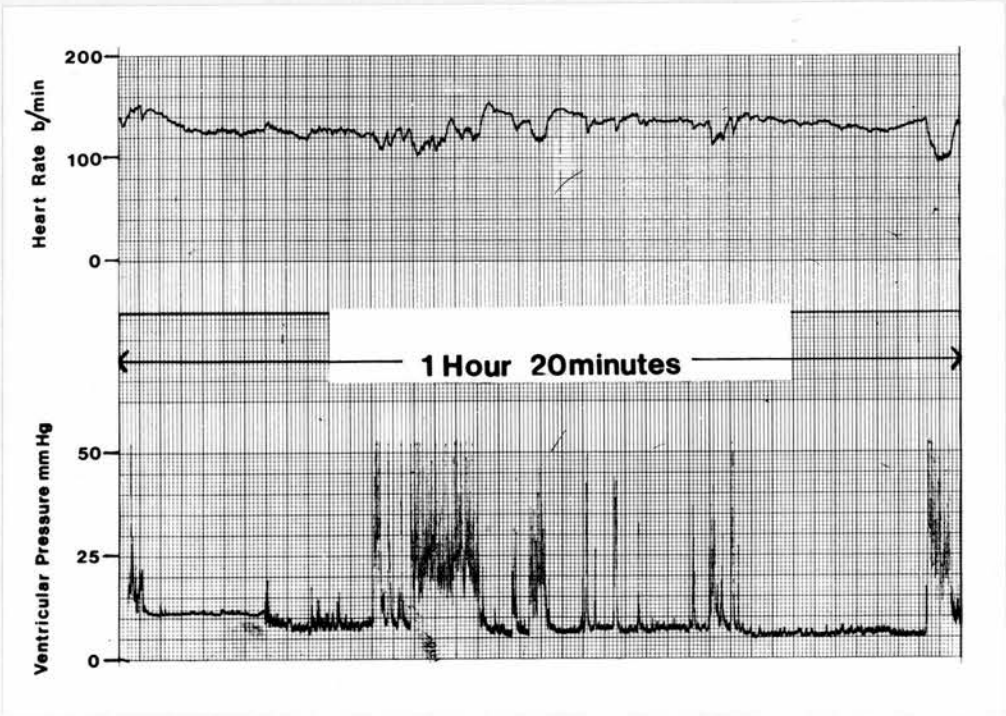
Stress Ventricular Pressure

70 mm Hg maximum.

Result — Normal level intracranial pressure as often in children with hydranencephaly there is no complete ventricular system and the surface of the remaining brain is the base of the ventricle, with some surface choroid plexus producing C.S.F.

Action — No action taken.




Heart Rate b/min
200
100
0
1 Hour 20minutes
Ventricular Pressure mmHg
50
25
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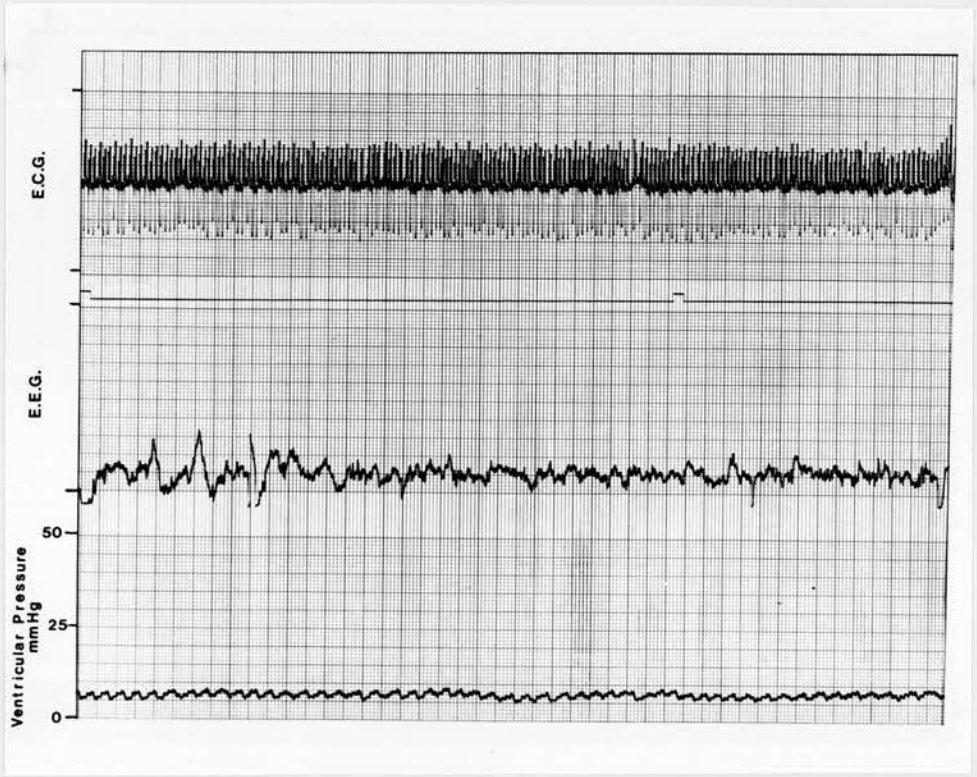


Fig.124

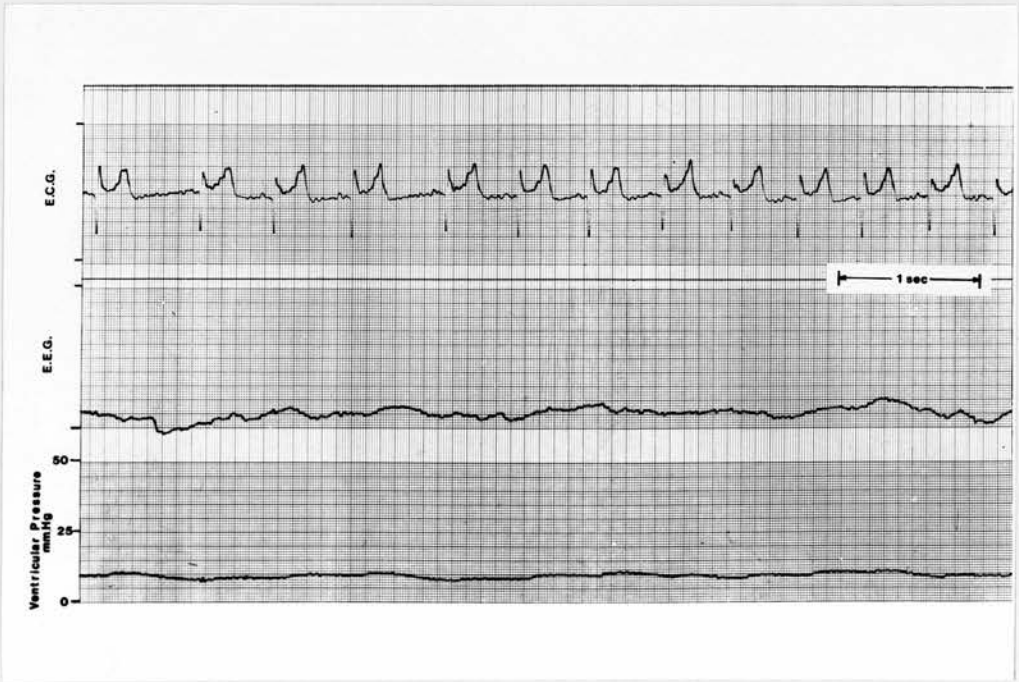


Fig.125

Cardiac/Respiratory Artefact

Undeterminable due to the method of monitoring.

Ventricular Dilatation/Cortical Mantle

Hydranencephaly.

Pressure Recordings Fig. 123 shows a tracing of ventricular pressure and heart rate. The resting pressure is within normal limits throughout, with the obvious elevation of pressure on crying etc. Fig. 124 shows simultaneous E.C.G., E.E.G. and pressure during a phase of light sleep, and, although the cardio-respiratory artefact is visible in the pressure tracing, no sizeable pressure oscillations occur in light sleep in this child. However, at an advanced recorder speed, Fig. 125, abnormalities of the E.C.G. tracing are obvious, not only the tall T waves, but also the short periods of asystole, which we have noted before, during a change from light to deep sleep. The E.E.G. on the right side was relatively flat and the left side showed larger slower waves, which became more slow with sleep. If the pressure fluctuations during early sleep that occurred normally are mediated by CO_2, then obviously one would need an intact cerebro-vascular tree for these to occur. In this child, it is not surprising with only a remnant of brain in the posterior fossa, and in intact brain stem, that they do not occur. C.S.F. for CO_2 estimations was obtained on two occasions throughout this tracing and levels of 4.47 and 4.54 were obtained (the normal range is 4.66 to 6). This may well be a dilutional effect, due to the large quantity of C.S.F.

Case Number	69
Name	N.R.
Age	11 months

Method — Right frontal Rickham reservoir

Medical Diagnostic Background

This child was born by S.V.D. one week before the E.D.D. and weighed 5 lbs $7\frac{1}{2}$ ozs. He was initially treated in an incubator because of cyanosis and hypothermia and subsequently tube fed. During the third month of pregnancy, mother had a severe attack of colitis which required admission to hospital and therapy with Salazopyrine, codeine phosphate, prednisolone and iron. The baby's OFC was noticed to be raised at about 6 weeks of age and it was followed up thereafter at a local peripheral hospital. At the time of referral, at 8 months of age, the OFC was 50 cms and he was clinically very well with no symptoms.

On examination the anterior fontanelle was almost closed, there was no papilloedema but the medial margins of both discs were blurred. There was no scalp vein distension but there was marked plagiocephaly.

Routine investigations including an E.E.G. were normal. The skull and spine x-ray were normal as was antenatal virology and toxoplasma titres etc. He accordingly had a C.T. scan and a Rickham reservoir electively inserted so that pressure could be measured. Unfortunately after inserting the Rickham, he developed a ventriculitis. When this had cleared, pressure monitoring was carried out.

Temperature	Normal
Zero	Upper cortical subarachnoid space.
Duration	$3\frac{1}{2}$ hours
Indication	The assessment of suspected active infantile hydrocephalus.

Resting Ventricular Pressure

3.5 mm Hg.

Stress Ventricular Pressure

65 mm Hg maximum peaks on crying. 27 mm Hg maximum peaks during sleep.

Result Normal level ventricular pressure awake, equivocal pressure changes during sleep.

Action A theco-peritoneal shunt was inserted.

Cardiac/Respiratory Artefact

R = 25 mm during sleep peaks, C = 12.5 mm in sleep.

CR = 6.25 mm awake.

CFC 50.8 cms at time of V.P.M.

Ventricular Dilatation/Cortical Mantle

A moderately symmetrical hydrocephalus with air passing over both cerebral cortices and a large subdural frontal collection in relation to the Rickham reservoir.

Pressure Recordings This child was given 5 mls of Trichloral at the outset of the V.P.M. and hence the pressure starts relatively low. He moves into an early phase of sleep quickly and this lasts for 5.8 minutes. He then enters into deep sleep. After a further 15 minutes in deep sleep, he then enters a light sleep phase again, with an elevation of pressure this time to a mean of 20 mm Hg peaking to 27 mm Hg. This phase of lighter sleep now lasts approximately 25 minutes. It would be interesting to know if sedation shortens the early light phase of sleep, as happened in this case. During the lighter phase of sleep, he is snoring lightly, stirring and grunting, smiling, eyes are half closed and half open with deep sighing respirations and occasional rubbing of the eyes, myoclonic jerks of his hands and facial

twitching. Fig. 126 shows this first period of 'light sleep' immediately after the Chloral. The frequency of the major fluctuations during 'active sleep' in this tracing was assessed as 1.46/minute or 1 wave every 41 seconds during the first R.E.M. phase, to 1 wave almost every 2 minutes, i.e. every 110 seconds in the second R.E.M. phase.

The rationale for insertion of a theco-peritoneal shunt in this child is that with an external collection, a theco-peritoneal shunt would be more likely to reduce the ventricular and the surface pressure than a pure ventriculo-peritoneal shunt, which may induce the aqueduct to close and allow continuing build up of the surface collection.

Case Number	70
Name	N.R.
Age	3 years
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

Congenital hydrocephalus with a theco-peritoneal shunt, as before.

Temperature	Normal
Zero	1" below upper cortical subarachnoid space.
Duration	1 hour

Indication — This child had been asymptomatic from the point of view of raised intracranial pressure, but a movable mass was palpated below the left costal margin at a routine clinic visit. There was no obvious focus of infection, his OFC was 55.5 cms and an anti-staph. titre was significantly raised at 6 units per ml. Full investigations including renal investigations, tuberculin testing, Australia antigen, hepatitis A titres, antinuclear factor, anti-

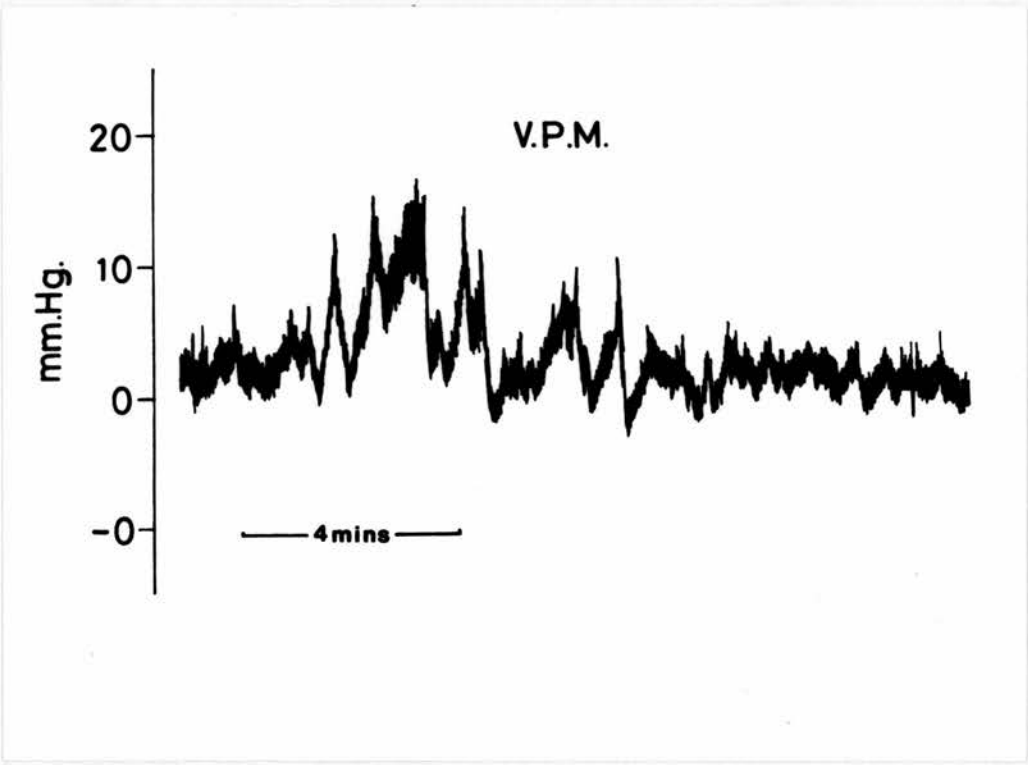


Fig.126

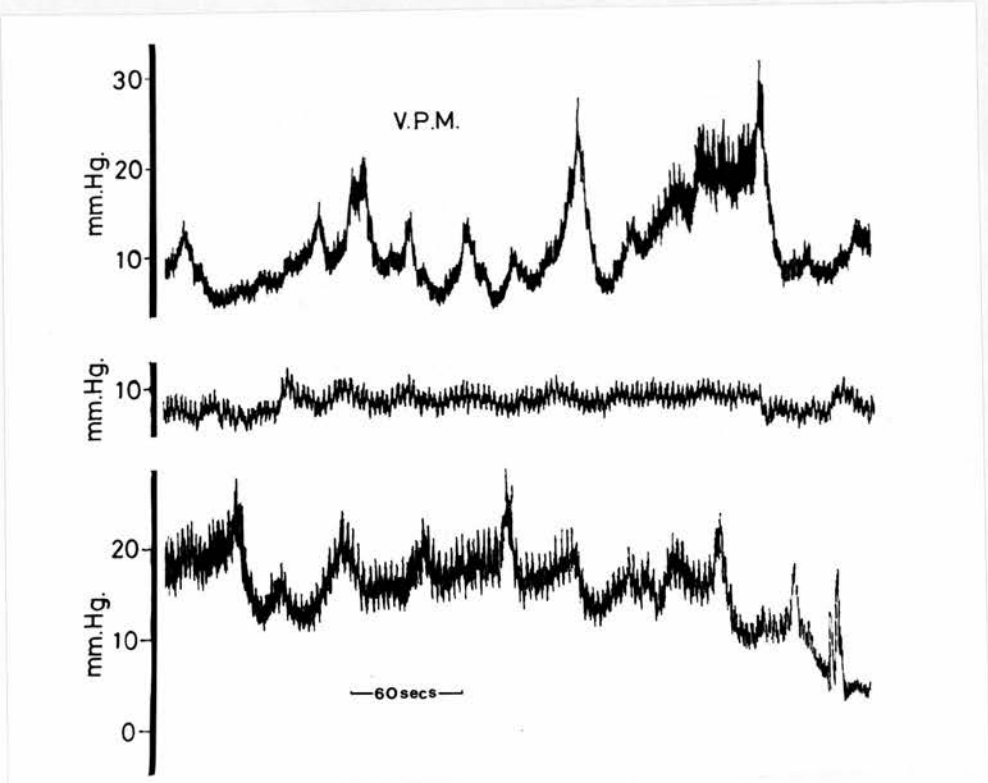


Fig.127

mitochondrial and antismooth muscle antibodies etc. were all normal. An ultrasound scan of the area confirmed that this was spleen, but the worry was that it may have been a lipomatous mass, or an abscess related to his theco-peritoneal shunt. Therefore, to test the effectiveness of his theco-peritoneal shunt, a V.P.M. was carried out.

Resting Ventricular Pressure

4 mm Hg.

Stress Ventricular Pressure

50 mm Hg maximum on crying, 24 mm Hg maximum during sleep.

Result — Similar to previous estimation.

Action — Nil with regard to C.S.F. pressure

Cardiac/Respiratory Artefact

CR = 5 mm in sleep, 2.5 mm awake.

OFC — 55.5 cms

Points of Interest — It was interesting that examination wise, his scalp veins distend very readily, with any stress such as crying etc. This short V.P.M. initially showed inappropriate responses, despite the apparatus functioning normally, and a presumed partial blockage of the Rickham reservoir or infected C.S.F. was suspected. However, with two successive gentle aspirations of half ml of C.S.F. from the Rickham reservoir, the flow improved and eventually the responses to crying etc. became more appropriate. He fell asleep for a short period and during this time similar sleep waves were observed.

It is unusual to get even a partial block of a Rickham reservoir, and if blockage does occur, it is most important in the child with a theco-peritoneal shunt in situ, because it means that if non-communication occurs, or there is infection of the C.S.F. spaces,

then diagnosis and treatment cannot be undertaken rapidly.

Case Number	71
Name	B.G.
Age	6 months
Method	Right parietal Rickham reservoir

Medical Diagnostic Background

This child was born by forceps delivery after labour was induced at term because of maternal hypertension. Mother was well during pregnancy. Birth weight was 8 lbs 5 ozs. Resuscitation was required with an endotracheal tube for 2 minutes, Apgar scores were 4 at 1 minute and 7 at 5 at 5 minutes. Intravenous sodium bicarbonate was also given. Thereafter there were no real problems, apart from mild jaundice which did not require phototherapy.
He breast fed well and was discharged home along with his mother at 7 days. Subsequently he was slow to acquire proper head control. In the family history mother had one previous miscarriage during the first trimester of her first pregnancy. Both parents have cousins with epilepsy and a maternal grandmother's last pregnancy resulted in a stillbirth at term and she was told that this baby had a 'large head'. A maternal cousin also had a baby who was followed up for a time on account of a 'large head'.
This child had been followed up because of an OFC above the percentiles. On examination his OFC measured 47.5 cms and there was no dilatation of superficial scalp veins and no loss of upward conjugate gaze. Fundal examination did not show any papilloedema. The anterior fontanelle was not under pressure. All four limbs moved symmetrically without any persistence of primitive reflexes. Tone was equivocally generally increased, but he had very poor

head control and marked lag on pulling to sit from the supine position. He had a preference for assuming an extended posture with head held well back. His weight lay exactly on the mean for age. His length was between the mean and tenth percentile line and his OFC was well above the 90th percentile for age, although it has been so for the past 12 weeks.

At 3 months of age the TSH and T_4 estimations were at a borderline elevated level but on this admission both were now in the normal range. A C.T. scan showed moderate severe dilatation of both lateral ventricles and the third ventricle. The fourth ventricle was not grossly enlarged and there was no displacement of the ventricular system. The sulci on both sides were enlarged and the appearances were those of a generalised cerebral atrophy. There was no alteration in the appearance of the cerebral parenchyma.

A 'degenerative enzyme' screen showed activities within the normal range.

Temperature	Normal
Zero	Posterior horn of lateral ventricles.
Duration	5 hours
Indication	The indication was therefore of a child with a large head (above the percentiles) tending to assume an opisthotonic position and with considerable head lag.

Resting Ventricular Pressure

8 mm Hg awake.

Stress Ventricular Pressure

Persistent crying resulted in maximum peaks on stress of 74 mm Hg and sleep waves of 11 mm Hg.

Result	Normal level ventricular pressure.
Action	No C.S.F. shunting device, but close

developmental follow up and C.T. scanning follow up.

OFC 47.5 cms

Ventricular Dilatation/Cortical Mantle

Moderate dilatation of ventricles without displacement.

Generalised cerebral atrophy.

Cardiac/Respiratory Artefact

CR = 2 mm awake.

CR = 3 mm asleep.

Points of Interest At follow up this boy is showing psychomotor retardation which is patchy and effects posture more than it does general manipulation and interest in his surroundings. So he has some atrophic cerebral condition, possibly familial, which has resulted in a large head, retardation and he must be at risk from epilepsy as well.

Case Number 72

Name G.M.

Age 4 years

Method Right sided Pudenz shunt

Medical Diagnostic Background

She was the first child of a healthy 24 year old mother. Pregnancy was normal and the child was born at 38 weeks gestation and birth was assisted by forceps because of delay in the second stage of labour. The neonatal period was uneventful. A district nurse noticed the large size of the child's head and she was admitted at 9 weeks of age. Examination at that time showed an OFC of 46 cms, both the fontanelle were tense and she had moderate 'sunsetting' of the eyes and was irritable. At that time L.A.E.G. showed no air entering the ventricular system. However, a myodil ventriculogram showed that

myodil passed through the aqueduct to the fourth ventricle and then to the subarachnoid space indicating a communicating hydrocephalus. Accordingly a right sided ventriculo-atrial shunt was carried out. At 4 years of age she was admitted again because a recent chest x-ray revealed the distal end of the shunt between the 3rd and 4th thoracic vertebrae. It was therefore necessary to consider lengthening this shunt and she was admitted for this and for pressure monitoring.

Temperature — Normal

Zero — Inter-ventricular foramina level

Duration — 3 hours

Indication — A question of converting a ventriculo-atrial shunt to a ventriculo-peritoneal shunt or removing it entirely, because of the unsatisfactory position of the lower end of the ventriculo-atrial shunt in the large neck vessels. The shunt was felt to be functioning normally.

Resting Ventricular Pressure

12 mm Hg.

Stress Ventricular Pressure

58 mm Hg maximum. 32 mm Hg maximum during sleep.

Result — Resting level of ventricular pressure acceptable but sleep pressures unacceptable. She therefore had a revision of her shunt system. At operation the shunt lower end was found to be firmly adherent to the innominate vein. The Pudenz tubing was seen to lie in a separate endothelialised compartment and at the time there did not seem any danger of internal jugular or innominate thrombosis and the system appeared to be allowing C.S.F. to flow down it, as judged by pumping on the flushing device. It was decided therefore to leave the whole system as it was.

Cardiac/Respiratory Artefact

CR = 7.5 mm asleep.

CR = 6.25 mm awake.

CR = 15 mm maximum sleep spikes.

Ventricular Dilatation/Cortical Mantle

No recent C.T. scan or A.E.G.

Pressure Recordings

Fig. 127 shows a complete sleep cycle segments from this child. The upper tracing shows the period going off to sleep, the middle tracing during deep sleep and the lower tracing at the awakening phase.

The pressure response to early sleep reached a maximum peak of 32 mm Hg. However, in the same child, when given 25 mgms Ketamine, the maximum response was 57 mm Hg. The time interval for the first elevation to occur with natural sleep was about 11 minutes, compared to 6 minutes following the Ketamine. Another feature of the early sleep in this child was quiet chewing movements. A good deal of minor twitching of the right hand tended to occur in the awakening phase, and occasionally left hand and body movements, and in this child particularly, these myoclonic jerks were seen to coincide with the peaks of this pressure wave.

Points of Interest It would appear then that this child was shunt dependent and still needed a C.S.F. shunting device.

Case Number	73
Name	D.A.
Age	10 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was the result of mother's third pregnancy, but the second successful one. Hydramnios was noted at 36 weeks and an elective caesarean section at 38 weeks gestation was carried out for a 'contracted' pelvis. The baby's large head was noted from birth, the OFC being 37 cms. The Apgar score was 5 at 3 minutes and the baby was noted to be floppy and daily OFC measurements were carried out which showed no increase over the first week. Regular follow up revealed the OFC continuing to run at +2 standard deviations. He had titubation of the head and showed signs of generalised hypotonia and dystonic extension of the arms when suspended. The reflexes were brisk and he showed features of ataxia and upper limb dystonia consistent with early hydrocephalus. Apart from fundoscopy showing rather pale discs, no other abnormality was noted. Routine investigations were normal and he was admitted on this occasion with an OFC which was now well above two standard deviations from the mean and scalp veins were slightly full.

Temperature	Normal
Zero	Inter-ventricular foramina level
Duration	1 hour
Indication	The assessment of infantile hydrocephalus.

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

During sleep he had peaks to 15 mm Hg.

Result	Probably normal level of ventricular pressure.
Action	Follow up OFC measurements, etc.

Cardiac/Respiratory Artefact

C = 5 mm. R = 12.5 mm.

OFC 51 cms

Ventricular Dilatation/Cortical Mantle

Communicating hydrocephalus with blockage over the left cerebral cortex.

Follow Up He has been seen a number of times since that admission and his head growth has paralleled the percentiles. He has had no active pressure problems or symptoms. He has, however, been found to have only partial hearing.

Case Number 74

Name A.B.

Age 4 months

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was admitted to the Infectious Diseases Unit with suspected meningitis at the age of 3 weeks. He had been born at term and weighed 7 lbs 6 ozs. Birth was by S.V.D. and there were no postnatal problems. The history on first admission was that for 10 days he had become progressively more drowsy and not showing interest in his feeding. On admission the main findings were 'twitching' of the right side and a full fontanelle. Subdural taps and lumbar puncture confirmed that he had bilateral subdural haematomas and a subarachnoid haemorrhage. The haematomas were tapped for several days with considerable improvement. A coagulation screen was negative as was a full skeletal x-ray survey. By 1 month of age an abnormal increase in his OFC occurred,

he was transferred for further investigations and management. At this stage his OFC was 40 cms, there was diminished movements of the left arm and a high pitched cry. He was generally hypotonic and had considerable head lag and no evidence of fixing or following. An echo encephalogram was midline and a C.T. scan showed a most peculiar picture with bilateral massive subdural collections with C.S.F. and blood clot mixed, but the midline appeared preserved. His OFC then progressed at a normal rate and he was allowed home. The following day he was readmitted with further bruising, irritability and he became extremely shocked and required urgent resuscitation. An L.A.E.G. showed a large collection of air in the subdural space and a gross hydrocephalus. The intention at this stage was to insert a theco-peritoneal shunt to drain both hydrocephalus and subdural collections. However, at the time of bilateral frontal burr holes, there were no subdural collections seen but the presence of gross hydrocephalus and accordingly a low pressure Pudenz ventriculo-peritoneal shunt was inserted.

Again he was allowed home and again he returned with bruising and some time later developed a ventriculitis and a massive intra-ventricular haemorrhage. There were numerous homeostatic defects with persistent hypernatremia, low blood urea, low serum osmolarity due to an inappropriate A.D.H. secretion. He also had 'brain stem fits'.

The diagnosis here is one of a battered child, initially with subdural haematomas and a gross hydrocephalus. It was during the recovery phase from his intra-ventricular haemorrhage that an encephalogram was performed and pressure measured to test the patency of his valve. The C.S.F. was blood stained during these measurements.

Temperature Normal

Zero — Inter-ventricular foramina level.

Duration — 1 hour

Resting Ventricular Pressure

4 mm Hg.

Stress Ventricular Pressure

32 mm Hg maximum.

Result — Normal level ventricular pressure.

Action — Nil

Cardiac/Respiratory Artefact

CR = 1.5 mm at rest.

OFC — 47.5 cms

Ventricular Dilatation/Cortical Mantle

Severe communicating hydrocephalus.

Pressure Recordings — Removal of 20 ccs of C.S.F. dropped the pressure from a resting pressure of 4 mm to - 3 mm Hg. This was followed by an injection of 20 ccs of air, which raised the pressure to 5.5 mm Hg.

Case Number — 75

Name — A.B.

Age — 5 months

Method — Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Battered baby syndrome, bilateral subdural haematomas and communicating hydrocephalus as before.

Temperature — Normal

Zero — Inter-ventricular foramina

Duration — 1 hour

Indication — There had been improvement in his homeostasis with an increasing sodium level on fludrocortisone daily and his shunt clinically felt normal. However, he was continually irritable and still having minor brain stem fits (Fig. 128).

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

50 mm Hg maximum on crying.

Result — Normal level ventricular pressure

Action — Nil

Cardiac/Respiratory Artefact

CR = 2 mm at rest.

OFC — 48 cms.

Ventricular Dilatation/Cortical Mantle

As before

Points of Interest

Since he was thought to be having small brain stem fits (going blank and swallowing), a concomitant E.E.G. was arranged for this pressure recording, but throughout a lengthy recording, only one electrical abnormality occurred and no changes in ventricular pressure were associated with it.

Case Number — 76

Name — C.N.

Age — 5 years

Method — Right parietal Ommaya reservoir

Medical Diagnostic Background

This child had a ventriculo-peritoneal shunt inserted for hydrocephalus at the age of 11 months. He had been perfectly well since

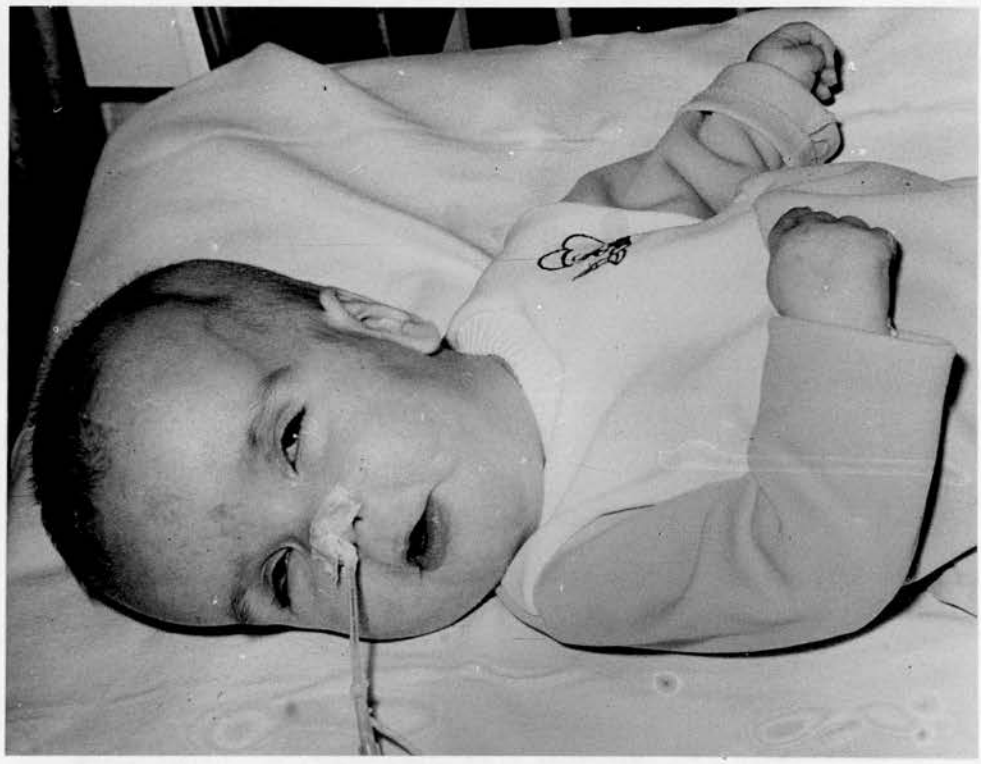


Fig.128

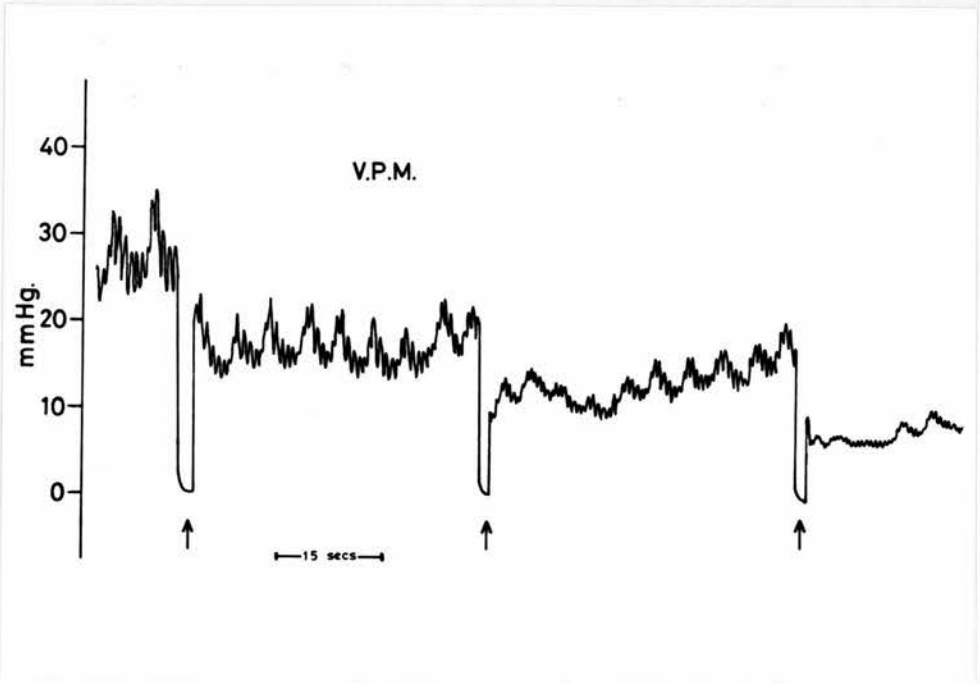


Fig.129

then but in the previous six months his parents had noted that the area around the valve was 'spongy'. Since it was thought that his hydrocephalus had been clinically arrested, he had his V.P. shunt removed two days prior to V.P.M.; the peritoneal end was found blocked and was removed. The ventricular end was left in situ.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 3 hours

Indication — To decide if his hydrocephalus was arrested sufficiently, to leave him without a C.S.F. shunt.

Resting Ventricular Pressure

10 mm Hg.

Stress Ventricular Pressure

60 mm Hg maximum peak on coughing. In early sleep maximum peaks of the pressure tracing reached 30 mm Hg for 4 minutes only and then he settled to 5 mm Hg level in deep sleep.

Result — Acceptable resting ventricular pressure but questionable values for very short durations during light sleep.

Action — No C.S.F. shunting system was reinserted, but it was planned for him to have regular follow up measurements of OFC, fundoscopy, height, weight, a C.T. scan in 3 months' time with serial C.T. scans at 6 monthly intervals after that. Also detailed assessment of hand function.

OFC — 54.7 cms

Ventricular Dilatation/Cortical Mantle

A C.T. scan performed 3 months after the monitoring, showed the lateral ventricles were moderately dilated without displacement. There was minor dilatation of the third and fourth ventricles and the appearances were little changed from those seen at the time of

monitoring and did not suggest progressive hydrocephalus.

Points of Interest At follow up 5 months later his mother reported that his balance had improved considerably; he was now coming downstairs without holding onto the railing. He was riding a tricycle and was attempting a 'two wheeler' with stabilisers.

A baseline psychometric assessment shortly after admission using the Stanford Binet Test found he was in the well below average range and possible special school level. Functioning in all areas was at a $3\frac{1}{2}$ - 4 year level for a chronological age of 5.

A repeat psychometric assessment 4 months after his V.P.S., by a different tester, showed him still to be in the well below average range in ability. There was some fine inco-ordination in drawing and writing was poor, and graded exercises in tracing were advised. A speech and hearing report one month after monitoring found that both comprehension and expression were certainly behind his chronological age and that his speech was dysarthric. Accordingly this aspect was to be reviewed to make sure it was not deteriorating. When this was repeated 5 months after his operation, articulation was normal apart from minor development areas, such as 'r' for 'l'.

Comprehension, however, on the Reynell Level Comprehension Scale A found him functioning at 4.10 - 4.11 years at a chronological age of 5 years 5 months. His expressive level was 4.7 - 4.9 years.

He remains without any signs of raised intracranial pressure and 7 months after his shunt removal, he was continuing to do very well. There was no deterioration in any sphere and most definite improvement in his balance and hand control. His behaviour and sleep were still satisfactory.

I have refused permission for an anaesthetic for dental extraction to be done outwith the hospital on this boy, and feel that expert anaesthesia, which does not raise the intracranial pressure, is a necessary precaution.

A repeat in his hand function test in one year after V.P.M. showed favounable that during that period he had not suffered any deterioration and indeed had made developmental progress.

Case Number	77
Name	S.G.
Age	16 months
Method	Rickham reservoir

Medical Diagnostic Background

This child was admitted at the age of 12 days. Mother had suffered hypertension during pregnancy and he was delivered by S.V.D. at 39 weeks. His birth weight was 3.53 Kg. and he was born with a congenital limb abnormality. He was discharged home after 48 hours and subsequently suffered some mild hypocalcaemia with jitteriness. On investigating this, it was found that he had an E. coli meningitis and this was associated with minor convulsions. He developed a secondary hydrocephalus with signs of pressure including dystonia and irritability and required ventricular taps. An L.A.E.G. at that time showed evidence of a non-communicating hydrocephalus and at 2½ months of age he had a ventriculo-peritoneal shunt inserted.

He remained quite well then, until 9 months of age when he was admitted with irritability, anorexia and clinically his ventriculo-peritoneal shunt was not working. His OFC at this time was 45 cms. During this admission he had a reconstitution of the right sided Pudenz ventriculo-peritoneal shunt and insertion of a Rickham

reservoir.

He was next admitted having been 'off his food', had a 'cold', vomited once, but had otherwise been well and his mother had felt that the valve was not working properly. Therefore pressure monitoring was carried out.

Temperature — Normal

Zero — Upper cortical subarachnoid space.

Duration — $2\frac{1}{2}$ hours

Indication — Proximal block clinically and on valvogram.

Resting Ventricular Pressure

8 mm Hg.

Results — Normal level ventricular pressure awake. The valve was non-functioning.

Action — He was discharged with a diagnosis of a non-functioning shunt but the pressure could still go up during infections.

OFC — 45.5 cms

Follow Up — He remained symptom free. In retrospect, unfortunately no prolonged sleep tracing was done on this child.

Case Number — 78

Name — S.G.

Age — 2 years 8 months

Method — Left parietal Rickham reservoir

Medical Diagnostic Background

Post-meningitic hydrocephalus, assumed to be arrested over the last two years, and symptomless during that period.

Temperature — Normal

Zero — $1\frac{1}{2}$" above the upper cortical subarachnoid space.

Duration — Half an hour

Indication — This child was admitted with an haemophilus upper respiratory infection and produced signs of acute pressure. He needed frequent tapping of C.S.F. from his reservoir.

Resting Ventricular Pressure

13 mm Hg pre-tap.

Result — Raised ventricular pressure

Action — Shunt revision

Cardiac/Respiratory Artefact

C = 18 mm, R = 26 mm pre-tap.

C = 6 mm, R = 19 mm after 5 ml tap of C.S.F.

C = 4 mm, R = 11 mm after 10 mls C.S.F. tapped.

C = 2 mm, R = 6 mm after 15 mls C.S.F. tapped.

Pressure Recordings

Note in Fig. 129 the changes in the height of ventricular pressure with successive 5 ml releases of C.S.F. 30 mm Hg pre-tap, 17 mm Hg after 5 mls, 14 mm Hg after 10 mls and 8.5 mm Hg after 15 mls removed.

Points of Interest

It may well be that because no sleep tracing was done on the first monitoring two years earlier that we did not suspect that this child was at risk from wuch elevated levels of ventricular pressure. On the other hand, it may be that because he had haemophilus, which has a known neurotropic effect, this may have resulted in a degree of cerebritis and encephalitis, with a resultant increase in level

of intracranial pressure. It seems likely in this case that the effect of the haemophilus is one of increasing C.S.F. production, because the pressure tracing responds very well to release of C.S.F. If the haemophilus had affected mainly brain substance we would expect less effectiveness with release of C.S.F.

Case Number 79

Name W.B.

Age 18 years

Method Right frontal Rickham reservoir (electively inserted).

Medical Diagnostic Background

Myelomeningocoele with hydrocephalus and chronic renal failure.

Temperature Normal

Zero 1" above upper cortical subarachnoid space.

Duration 22 hours

Indication To distinguish if the fundal changes which she had were due to raised intracranial pressure insidiously appearing, or whether they were due to intracranial hypertension from renal involvement.

Resting Ventricular Pressure

7.5 mm Hg.

Stress Ventricular Pressure

Maximum peaks in sleep to 20 mm Hg.

Result Normal level ventricular pressure.

Action Nil re C.S.F. pressure

Cardiac/Respiratory Artefact

CR = 1.5 - 2 mm at rest.

Pressure Recordings The pressure recordings in this girl show long periods of sleep with rhythmical oscillations occurring 1 every 18 seconds, that is 3.3 per minute (Fig.130).

Case Number	80
Name	C.P.
Age	3 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Normal pregnancy and no history of birth injury. Congenital hydrocephalus.

Temperature	Normal
Zero	Inter-ventricular foramina level.
Duration	2 hours
Indication	The assessment of suspected active infantile hydrocephalus with a rapid increase in OFC over the previous 2 weeks, an increase in scalp venous distension, tension of the fontanelle and no irritability until the day of monitoring. There had been no vomiting and vital signs were normal (father had a 'large head' but was also of large build).

Resting Ventricular Pressure

22.5 mm Hg awake.

Stress Ventricular Pressure

34 mm Hg maximum and during sleep mean of 25 mm Hg with sleep peaks to 30 mm Hg.

Result	Raised ventricular pressure.
Action	C.S.F. shunt

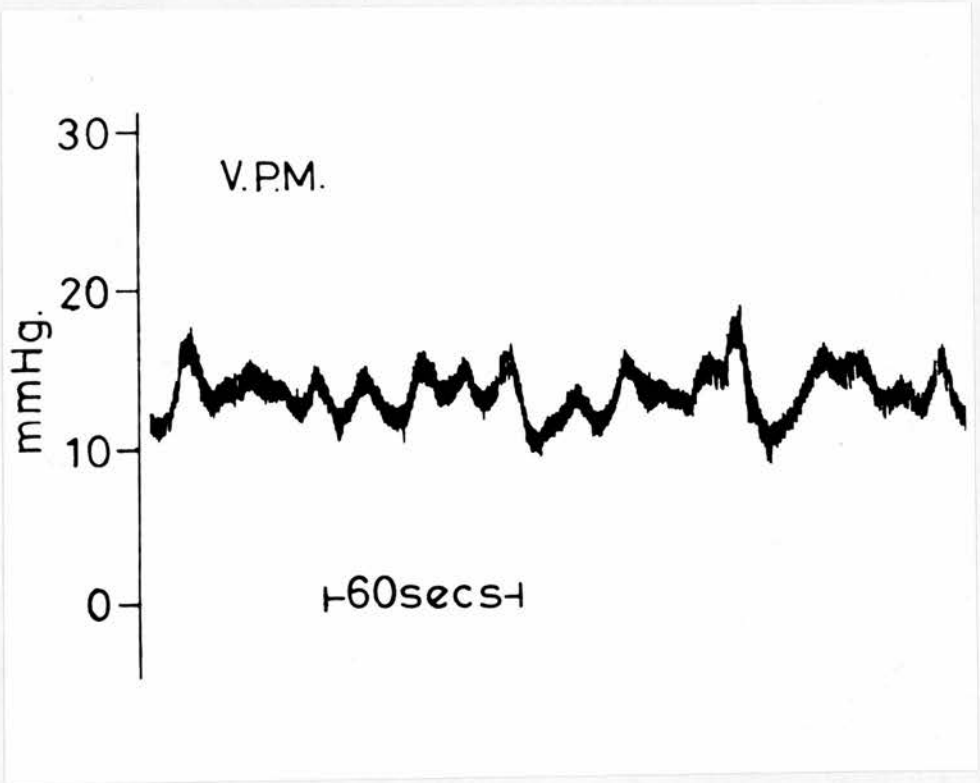


Fig.130

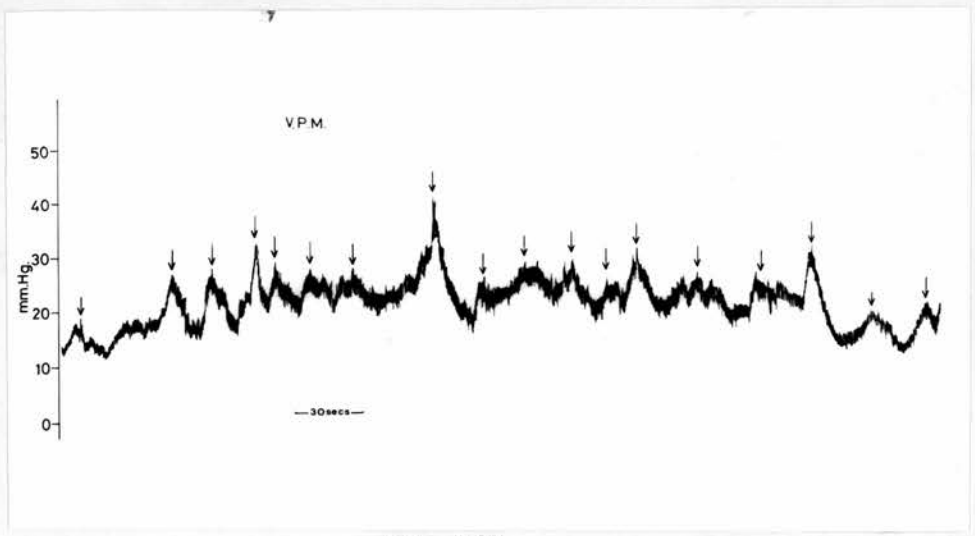


Fig.131

Cardiac/Respiratory Artefact

CR = 3 mm in sleep.

CR = 1 mm awake.

Ventricular Dilatation/Cortical Mantle

A C.T. scan showed gross enlargement of ventricles with a cortical mantle of 3.75 mm.

Pressure Recordings

Pressure	C.S.F. release	C.R.A.
23 mm Hg.	0	2 mm
18 mm Hg	5 ml	1.5 mm
15 mm Hg	10 ml	1.5 mm
12 mm Hg	10 ml	1 mm
10 mm Hg	10 ml	0.75 mm
9.5 mm Hg	10 ml	-

i.e. withdrawal of 45 mls of C.S.F., reduced the ventricular pressure by 13.5 mm Hg. On average 1 ml C.S.F. = 0.3 mm Hg for this patient over this pressure range.

Case Number 81

Name D.R.

Age 4 months

Method Right frontal Rickham reservoir

Medical Diagnostic Background

This child was admitted to hospital having had a convulsion lasting 4 minutes and at the time was noted to have a large head and tense fontanelle. Subdural taps revealed the presence of a large subdural haematoma and fundoscopy revealed multiple retinal haemorrhages. Other injuries noted were fractures of the 6th, 7th, 9th, 10th and 11th ribs on the left side. Subsequently alternate daily subdural taps were

performed and a C.T. scan showed symmetrical subdural collections and moderate symmetrical hydrocephalus. Again subdural taps revealed 14 mls of blood stained fluid from the right side and 13 mls from the left. Therefore burr holes for external drainage were carried out in theatre. The tubes of the external drainage were removed after 3 days and he gradually improved. A repeat C.T. scan showed a subdural hygroma in both frontal regions was not clearly as evident as before. The plan was that in babies with subdural haematomas and cranial disproportion, to put in a Rickham reservoir and after external drainage of the subdural haematoma, to pressure monitor, and if the pressure is raised, to insert a shunt.

Temperature 36.9

Zero Upper cortical subarachnoid space.

Duration 8 hours

Indication Assessment of secondary hydrocephalus following bilateral subdural haematomas.

Resting Ventricular Pressure

9 mm Hg.

Stress Ventricular Pressure

40 mm Hg maximum stress (crying) and 40 mm Hg maximum during sleep.

Results Acceptable awake levels of pressure but elevated levels during sleep.

Action C.S.F. shunt

Cardiac/Respiratory Artefact

CR = 8.75 mm asleep.

CR = 2.5 mm at rest.

Ventricular Dilatation/Cortical Mantle

Severe symmetrical ventricular dilatation.

Pressure Recordings This pressure recording contained a good deal of sleep. Fig. 131 shows a typical sleep elevation and fluctuations (indicated by the arrows). In this section the frequency of these fluctuations was 42, 18, 19, 9, 15, 20, 36, 21, 19, 21, 15, 14, 20, 7, 27, 25, 27 and 23 seconds.

Analysis of these frequencies again shows no statistical progression. However, the intervals tend to progress from long intervals to short intervals to long intervals etc.

It is interesting that in this child crying and stress elicit a response of 14 mm Hg maximum and this is exactly the same level as that obtained during sleep recordings.

Case Number	82
Name	M.C.
Age	1 month

Medical Diagnostic Background

This child was born by lower segment caesarean section for 'failure to progress' and at birth was noted to have a cervical spina bifida lesion. B.W. was 4.01 Kg and Apgars were satisfactory. The baby was generally hypotonic and had an OFC of 34.5 cms. The cervical myelomeningocoele measured 2 x 3 cms and at the base was covered with a thick membrane. There was some right arm weakness. The following day the lesion was excised and the baby made a good post-operative recovery. However, the OFC rapidly increased.

Temperature	Normal
Zero	Inter-ventricular foramina level.
Duration	4 hours
Indication	The assessment of active neonatal hydrocephalus (with retraction of the neck, 'sunsetting', increasing

OFC and tense anterior fontanelle, since the back operation).

Resting Ventricular Pressure

14 mm Hg.

Stress Ventricular Pressure

64 mm Hg maximum on crying.

Result — Raised ventricular pressure.

Action — A ventriculo-peritoneal shunt was inserted at 18 days of age. An attempt was made to insert a Rickham reservoir through a frontal burr hole, but, because of the mobility of the frontal bone, having decompressed his hydrocephalus, this was unsuccessful.

Cardiac/Respiratory Artefact

0.75 mm at 15 mm Hg.

OFC — 38 cms

Ventricular Dilatation/Cortical Mantle

A.E.G. at the time of V.P.M. showed dilated lateral and third ventricles, but no air was seen in the aqueduct or fourth ventricle or subarachnoid space. The cortical mantle was 3 cms at the vertex.

Pressure Recordings — A tap of 30 mls of C.S.F. reduced the pressure from 15 mm Hg to 8 mm Hg. There was a considerable improvement in 'tidal volume' using a pneumotachygraph after tapping 30 mls of C.S.F. compared to the 'tidal volume' in the high pressure, pre-tap state. This appeared to be independent of the frequency of respiration.

Case Number — 83

Name — K.L.

Age — 4 months

Method — Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was admitted at 3 months of age. She had a very stormy neonatal period, a result of recurrent ventriculitis and her prognosis at the time of transfer was thought to be particularly poor. She was born to a 31 year old para 1 + 0 mother who had been well during pregnancy. Antenatal screening had shown hydrocephalus and labour was induced at an estimated 32 weeks gestation by Prostaglandin E combined with Pethidine and Phenergan. The first stage of labour lasted approximately 5 hours and the second stage 9 minutes. This baby had not been expected to survive but at birth cried spontaneously and showed satisfactory Apgars of 9 at 1 minute and 5 minutes respectively. B.W. was 2.12 Kg and the OFC was 34 cms with a wide anterior fontanelle and separated sutures. There were no other external stigmata of neurological disease. Gestational assessment placed her at around 34 weeks. She was tube fed, initially with success, and had mild jaundice with a peak Bilirubin of 176 umol/l and treated with phototherapy.

Over the first 10 days of life there was progressive increase in OFC and by the 12th day this measured 35½ cms with some scalp venous distension and a full tense anterior fontanèlle. Accordingly a right sided ventriculo-peritoneal Pudenz anastomosis was carried out. A brain biopsy was also taken at the time of surgery. Post-operatively she had some abdominal distension and 5 days post-operatively she developed a fever with tonic convulsions, bilateral 6th nerve palsy and marked papilloedema. Ventricular tap revealed a ventriculitis and this was associated with a septicaemia. Her shunt was removed and intrathecal antibiotics continued. However, signs of infection persisted and an A.E.G. showed a grossly dilated left lateral ventricle and the right was not seen. The possibility

of a cyst on this side was raised. On admission her OFC was 39 cms, her crown-heel length was 52 cms and weight 3.8 Kgs. She had a low grade fever and fontanelle were slightly full but not bulging. She was a little irritable and was uniformly hypotonic. Phasic reflexes on the left side were slightly increased compared to the right. 24 hours after admission her fontanelle became very tense and she vomited. A ventricular tap was carried out which showed a persistent ventriculitis and she required daily tapping for control of her C.S.F. pressure. At the end of 10 days of intrathecal Gentamycin and Cloxacillin, there was no cellular increase and no growth on culture. Therefore a V.P.M. was carried out.

Temperature — Normal

Duration — 1 hour

Indication — Recurrent or persistent ventriculitis and signs of raised intracranial pressure (lethargy and an increase in scalp venous distension).

Zero — Inter-ventricular foramina level.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure.

Result — Raised C.S.F. ventricular pressure.

Action — Immediate relief of 30 ccs of C.S.F. reduced the pressure to 8 mm Hg. 15 cc of air was inserted which increased the pressure from 8 to 10 mm Hg. Later a left sided ventriculo-peritoneal shunt was inserted and a right frontal Rickham reservoir to avoid unnecessary puncture of the cortex.

Cardiac/Respiratory Artefact

CR = 1 mm at rest.

OFC — 39 cms

Ventricular Dilatation/Cortical Mantle

Gross bilateral ventricular enlargement and no obvious communication with the 4th ventricle, indicating a probable aqueduct stenosis. Cortical mantle on the right measured 5.6 mm and on the left 13.8 mm.

Points of Interest The Rickham reservoir was inserted both for the immediate and for the future interests of this child. Recurrent ventricular tapping, apart from increasing the likelihood of infection often results in puncture porencephaly and intra-ventricular haemorrhage. No doubt a result of tearing of a friable eppendyma, especially when infection has complicated the case.

Case Number	84
Name	R.C.
Age	2 years 3 months
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

This child was born following an uncomplicated pregnancy and straightforward S.V.D. Neonatal progress was satisfactory. He was seen in hospital 6 days after mother was diagnosed as suffering from pulmonary tuberculosis. The child was then 7 months of age. At the time he was drowsy, ill looking, fretful and had a shrill cry. He had some neck retraction and the anterior fontanelle was wider than expected. OFC was on the 50th percentile and he had a slight left facial weakness and evidence of a left hemiparesis. He was proven to have a tuberculous meningitis with a markedly raised protein in the C.S.F. with 2.45 gm/l and a glucose depressed at 0.4 gm/l. He was treated with Rifampicin, Isoniazid and Prednisolone orally. Over the first two weeks there was, if anything, slight deterioration in his clinical progress with the left hemiplegia becoming more

obvious and repeated C.S.F. examination showing a rising protein. While the OFC remained constant, the level of consciousness began to deteriorate and it seemed likely that his vision was impaired and possibly his hearing. He also suffered some brain stem seizures which were treated with Nitrazepam. Accordingly he was given Hydrocortisone intrathecally and a C.T. scan confirmed that he had marked hydrocephalus. Four weeks after admission he had a Rickham reservoir inserted and thereafter daily removals of C.S.F. and installation of Streptomycin. Following this his progress was more satisfactory, he became less irritable and ceased to have seizures. A further C.T. scan, 2 months after admission, showed that his hydrocephalus persisted. An E.E.G. showed moderately severe generalised abnormalities. As his C.S.F. became normal, a ventriculo-peritoneal shunt was inserted using a Pudenz pump.

Three months after admission a chest x-ray showed no abnormality. He continued to improve, but was left with a residual left hemiplegia and a left ventriculo-peritoneal drain, functioning well. At the time of discharge he was receiving Rifampicin, Isoniazid and Pyridoxine.

He was next admitted at the age of 2 years 3 months with a history of vomiting three times, sporadically drowsy, irritable and crying. The only other feature of note was that three weeks before admission he had fallen out of his cot, but at this time did not appear to have hurt himself.

Examination suggested a right homonomous hemianopia and because of technical problems, it was difficult to assess his shunt function properly, although it was felt that he was not under gross pressure. He was therefore transferred for the assessment of intracranial pressure.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — $7\frac{1}{2}$ hours

Indication — To decide if his vomiting, irritability and drowsiness were a feature of raised intracranial pressure and shunt malfunction.

Resting Ventricular Pressure

5 mm Hg.

Stress Ventricular Pressure

Sleep peaks of 38 mm Hg and 6.5 mm Hg in deep sleep.

Result — Normal awake ventricular pressure level, but during the lighter phases of sleep, the levels became unacceptable reaching a peak of 37 to 38 mm Hg as shown by the arrow in Fig. 132. The mean level of pressure at this time was 24 mm Hg and the maximum time that the pressure peaks exceeded 20 mm Hg was about 6 minutes. This tracing commences with the child crying himself to sleep, then in early sleep we see a nice symmetrical pattern of 'active sleep' lasting in all 14 minutes, with waves occurring at 40 and 60 second intervals. Following this he progresses into deep sleep, which lasts $26\frac{2}{3}$ minutes. A section of this is shown in Fig. 133 with an acceptable level of ventricular pressure. He then passes into a lighter phase of sleep again, $16\frac{2}{3}$ minutes, with characteristic wave forms, and again into a deep sleep phase of $25\frac{2}{3}$ minutes.

Approximately one third of this boy's sleep is in 'light' or 'active' sleep and two thirds spent in 'deep' or 'quiet' sleep. The awakening phase, lasting 19 minutes, shows characteristic wave forms and in Fig. 134 there are unusual sequences, indicated by the arrows, then there is no respiratory component. This is prior to waking and it appears as though they are forerunners to the sleep spikes prior to

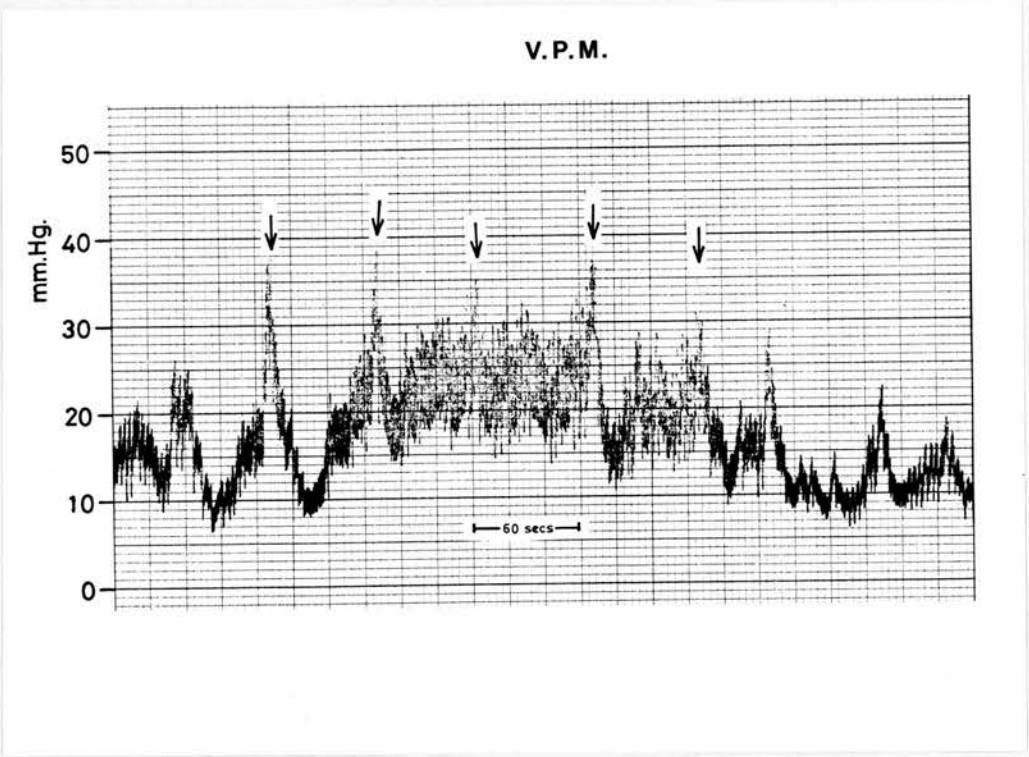


Fig.132

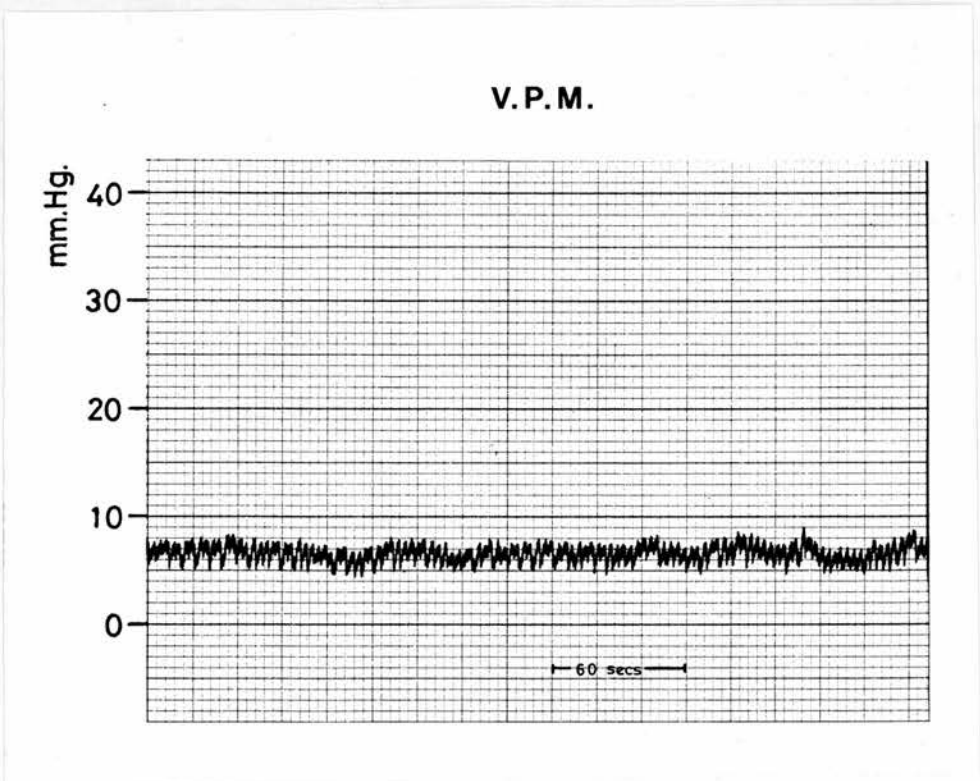


Fig.133

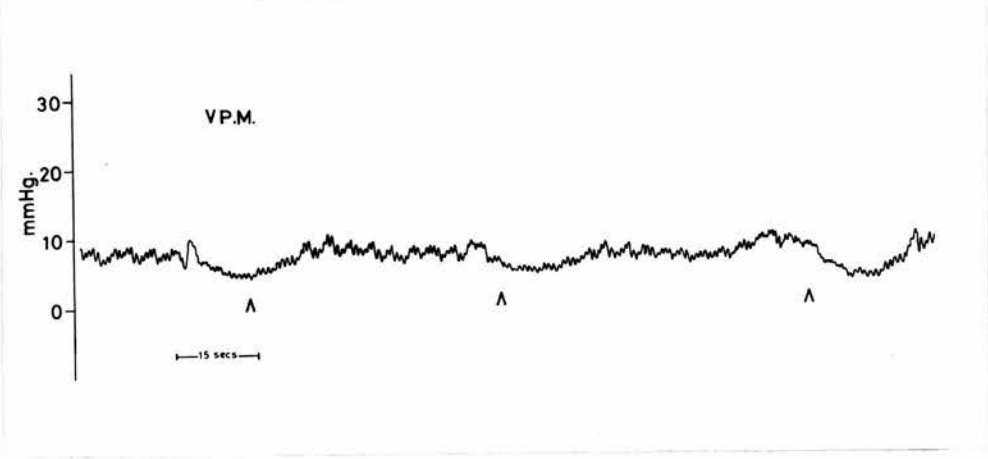


Fig.134

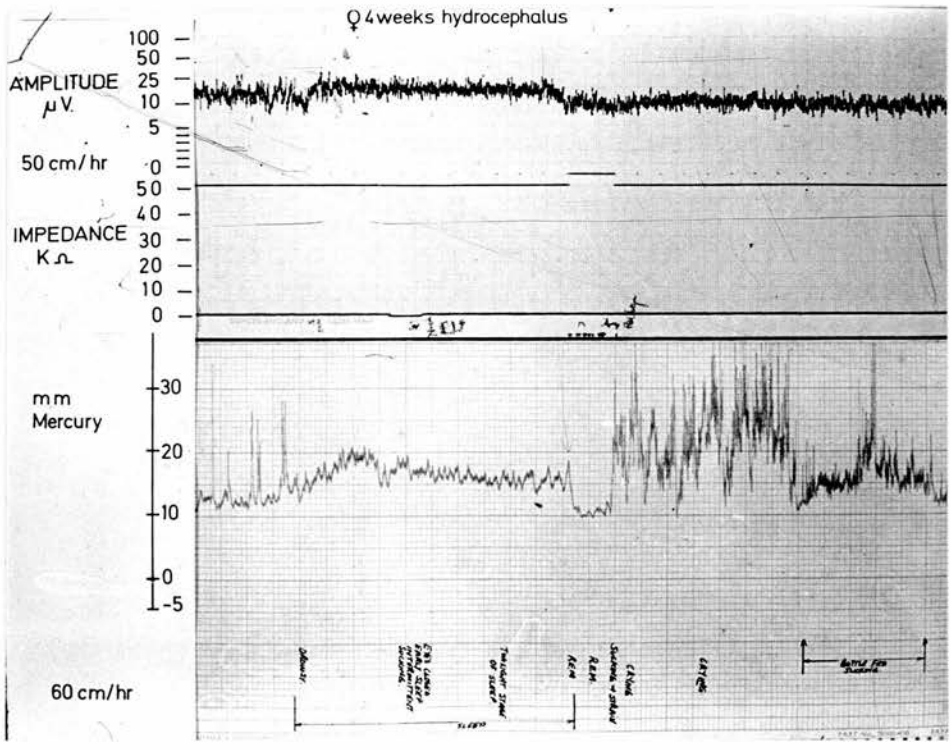


Fig.135

awakening. The spikes probably are a result of an increase in CO_2 resulting in an increasing cerebral blood flow to provide substrate for an increased cerebral metabolism, to begin active thought and to initiate wakefulness.

Action It was decided therefore not to reconstitute or revise his shunt, but to undertake close follow up observation of this child, partly because he was post-infective communicating hydrocephalus, and partly because the awake pressure was at an acceptable level, and only unacceptable for very short periods during sleep. Therefore long term psychometric assessment, detailed hand function assessments and C.T. scans to detect any insidious ventricular dilatation that might occur during a period of time, during sleep.

Cardiac/Respiratory Artefact

CR = 15 mm in sleep.

CR = 2 mm at normal pressure levels.

Follow Up Physiotherapy assessment three days after V.P.M. found his left arm alternated between dystonia with flexion and supination. He had shoulder abduction in equilibrial reactions, and was beginning to use his left arm as a prop. There was no grasp reflex and no voluntary grasp. There was extension of the fingers but questionable index finger isolation and there was no hand regard. Voluntary supination was present.

Functionally he watched where articles went when they dropped but did not eat a biscuit. He did not clap hands and did not drink from a cup and had no proximal fly swatting movements.

The opinion was that the left hand was about a $3\frac{1}{2}$ months functional level and he would probably greatly benefit from intensive physiotherapy.

He was next seen 7 months after his V.P.M. and compared to his previous assessment there has been no deterioration. Neither had there been any improvement in his upper limb development, in that he was still functioning at the 6 month level at an age of 2 years 10 months and he still had proximal arm movement only. He was unable to take weight on his left arm to use it as a prop, and he did not have any lateral protection.

A further physiotherapy assessment 9½ months after V.P.M. showed him functioning at 12-14 months level. The ranges of movement were reasonable throughout, with some limitation of left hip flexion and abduction. He was just beginning to achieve independent standing and could attempt to take a few steps unsupported. His left hand function test (Minns et al 1977) remained poor.

A Clinical Phychologist established a base line at 2 years 4 months with a view to future follow up. An overall impression at that time was that of a child of 15 months age level. This assessment was just after his V.P.M.

A C.T. scan was performed 7 months after his V.P.M. and this showed there was considerable dilatation of the lateral ventricles and the third ventricle. The fourth ventricle was quite small in comparison with the remainder of the ventricular system, suggesting that there may now be some impairment of conduction through the aqueduct. The right lateral ventricle was very slightly larger than the left, but there was no displacement. No gross abnormality was seen in the basal cisternal region. There was no undue collection of fluid on the cerebral surface but a small area of density around the insertion of the right posterior parietal shunt tube, suggesting an area of necrosis at that point.

An Occupation Therapy functional assessment 9½ months after his V.P.M.

showed him overall to be functioning at a level of about 12-15 months. At 3 years of age he was functioning at about 15-16 months in all things. There had been no deterioration. However, his mother felt that if she touched the site of the valve he cringed and he had an extravasation of C.S.F. in the sub-galeal space. There had been no irritability or screaming attacks and no suggestion of raised pressure other than the extravasation. Post-meningitic hydrocephaluses are not necessarily shunt dependent in the long term unless there is a secondary aqueduct stenosis. It is therefore worthwhile at this time doing a further V.P.M. and C.T. scan to exclude C.S.F. infection.

Case Number	85
Name	E.W.
Age	1 months
Method	Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

This child was born by caesarean section at 39 weeks gestation because of foetal distress and a previous caesarean section. This baby was small at birth, B.W. 2540 grams, a length of 46 cms and an OFC of 33.5 cms. However, a large pedunculated occipital meningoencephalocoele was evident, 10 cms in diameter. Examination was otherwise normal. Accordingly this was removed at 3 days of age, but post-operatively the child became pyrexial with a tense fontanelle and developed a wound infection which progressed to a septicaemia and staph. albus ventriculitis. Treatment was undertaken with intrathecal and systemic antibiotics and regular ventricular taps. The OFC throughout this time was increasing slowly.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — $2\frac{1}{2}$ hours

Indication — The assessment of active neonatal hydrocephalus. An A.E.G. was done at the same time as the V.P.M.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

40 mm Hg.

Result — Raised ventricular pressure.

Action — Continued tapping of C.S.F. until a Pudenz ventriculo-peritoneal system was inserted 3 days later.

Cardiac/Respiratory Artefact

Nil.

Ventricular Dilatation/Cortical Mantle

Gross ventricular dilatation and air in the basal cisterns.

Cortical mantle 8 mm.

Pressure Recordings — Fig. 135 shows a concomitant recording of cerebral function monitor (the upper tracing) and ventricular pressure (lower tracing). It can be seen that during the 'active' phase of sleep, there is a rise in the ventricular pressure level and a similar change in the base line of the summated E.E.G. tracing.

Case Number — 86

Name — E.W.

Age — $7\frac{1}{2}$ months

Method — Left temporal Pudenz flushing device.

Medical Diagnostic Background

She had a number of operations since the first pressure recording,

the first operation was removal of the occipital meningo-encephalocoele, the second was insertion of the Pudenz ventriculo-peritoneal shunt, the third was removal of the ventriculo-peritoneal shunt and a new left sided ventriculo-peritoneal shunt inserted, the fourth was revision of the left sided ventriculo-peritoneal shunt and the fifth was a further revision of the distal end of the left sided ventriculo-peritoneal shunt. On this occasion she was admitted irritable, anorexic with an upper respiratory infection and vomiting. A valvogram confirmed a distal block of her ventriculo-peritoneal shunt.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 3 hours

Indication — A blocked C.S.F. shunt with no clinical signs.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

77mm Hg maximum on crying, 14 mm Hg maximum peaks during sleep.

Result — Raised ventricular pressure and blocked C.S.F. shunt.

Action — The ventriculo-peritoneal shunt was reconstituted with a new distal limb.

Cardiac/Respiratory Artefact

R = 11.25 mm in early sleep.

C = 6.25 mm in early sleep.

Ventricular Dilatation/Cortical Mantle

Severe ventricular dilatation.

Pressure Recordings Fig. 136 shows a polygraphic recording with the ventricular pressure tracing at a slightly faster sleep speed than the other parameters, i.e. respiration, E.E.G., E.M.G., E.C.G. and E.O.G. (eye movements). This section of recording was taken during sleep and one can see eye movements occurring, sleep spindles and a slightly irregular respiratory tracing.

Case Number 87

Name E.W.

Age $10\frac{1}{2}$ months

Method Ventricular cannulation via anterior fontanelle.

Medical Diagnostic Background

Since the shunt revision following the previous V.P.M. she has had a further 5 operations related to her C.S.F. drainage. The most recent shunt was inserted, a ventriculo-atrial shunt, 4 days prior to this V.P.M.

Temperature Normal

Zero Inter-ventricular foramina level.

Duration 1 hour

Indication To test the effectiveness of the ventriculo-atrial shunt as the child clinically was still requiring to have ventricular taps. (She was symptomatic, in that she had a tense anterior fontanelle, her conscious state was slightly diminished and there was hippus and bradycardia).

Resting Ventricular Pressure

22 mm Hg.

Stress Ventricular Pressure

During sleep, peaks of ventricular pressure reached 36 mm Hg.

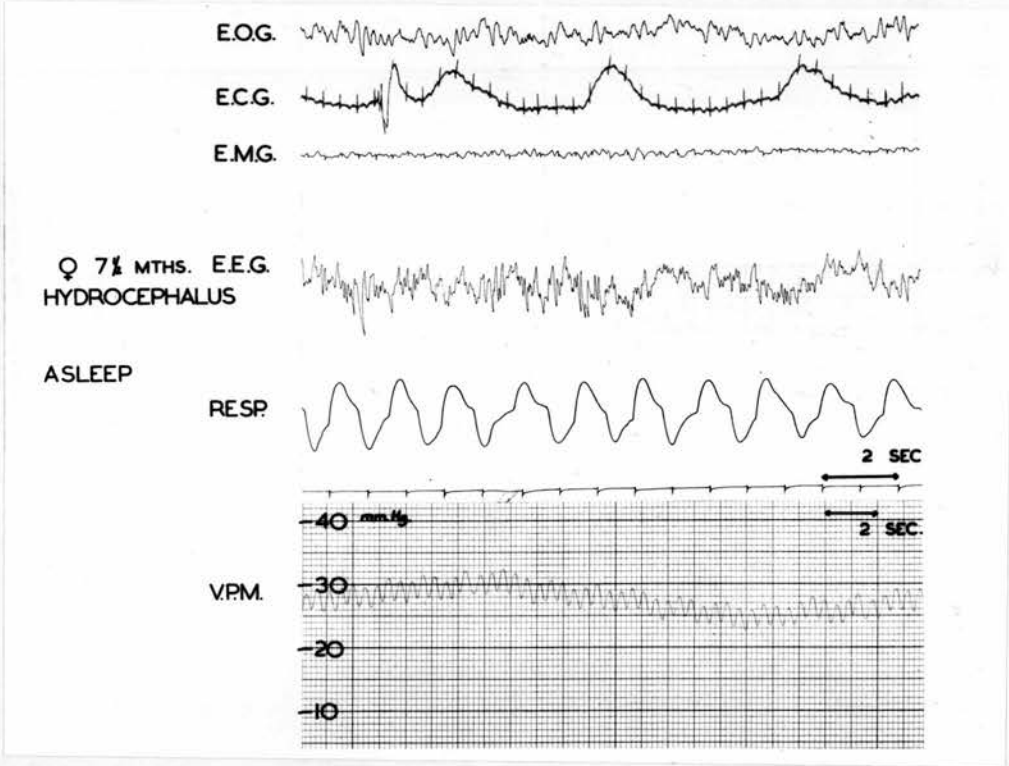


Fig.136

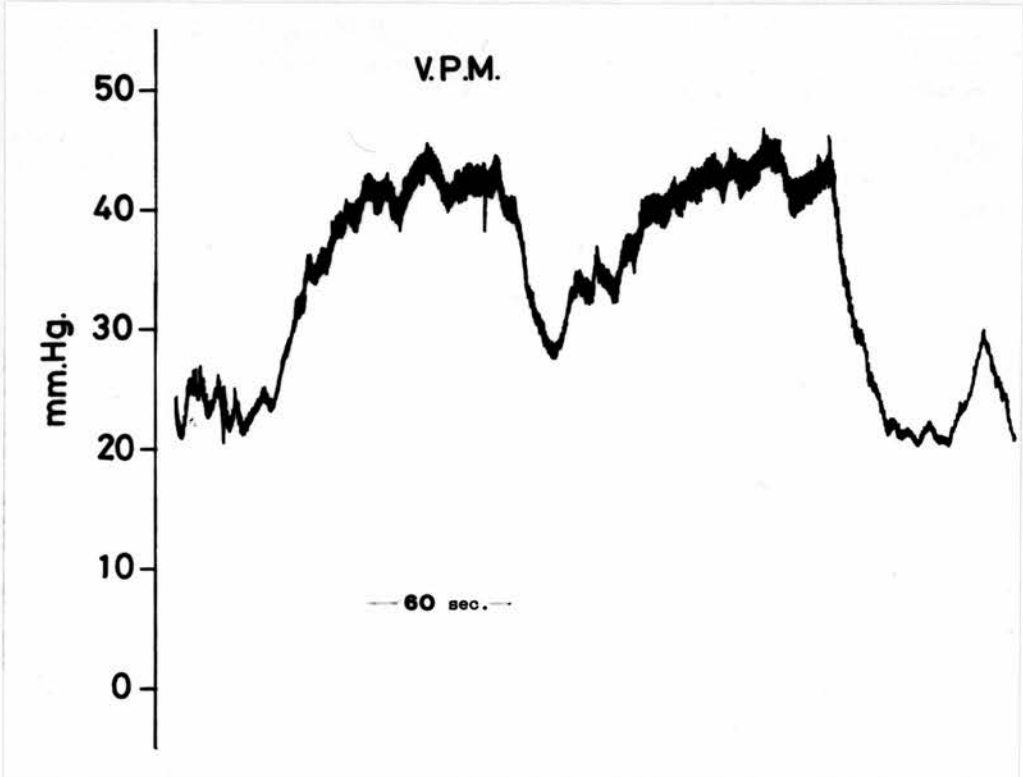


Fig.137

Result — Raised ventricular pressure.

Action — Revision of ventriculo-atrial shunt.

Cardiac/Respiratory Artefact

CR = 2.5 mm at rest.

CR = 12.5 mm during plateau waves.

Pressure Recordings — Fig. 137 shows an example of two minor or 'pre-plateau' waves which occurred during this tracing, while the child was quiet, lying supine with the eyes to the front. She was sleepy and on the descending slope of these plateaus she consistently made a 'grunt'. The smaller wave at the end of this tracing is immediately preceded by observable twitching of her closed eyes. There was a lot of eye blinking throughout this recording, and during the blinking phase the pressure descended. At the height of the pressure waves the child was quiet and immobile.

Later successive pumping of the valve abated the plateaus and brought the pressure back to the resting level of about 25 mm Hg, but it did not drop into the normal range. Therefore, the valve appeared to be only partically working. During this tracing the plateau wave lasts for 4 minutes, a further plateau wave occurring shortly after, lasted 3 minutes 20 seconds.

Points of Interest — This little girl who had some 16 operations in as many months, was developmentally retarded to a 5-6 month old level. She was neurologically intact from the point of view of her original lesion and her vision and hearing were thought to be normal, but she would no doubt be intellectually retarded due to the gross ventricular dilatation. On a few occasions the cortical mantle was measured as less than 1 cm.

It was noticeable that she had delayed healing of her wounds on many occasions. On some occasions this was due to systemic infections and

on others due to local wound infections. This was added to doubtless by some of the different types of shunt, for example the acuflow shunts which have a considerable amount of plastic material to lie beneath the skin resulting in some stretching and not ideal for rapid healing. Furthermore, when C.S.F. leaks through the wound this delays healing markedly. These facts plus her overall retardation in growth, prolonged hospitalisation etc. mean that her healing problems had probably been multifactorial.

We took the opportunity to investigate her healing ability in some detail and a skin biopsy was taken from her thigh. Although this was primarily for histology and histochemistry, it was also in the nature of an experimental wound and was photographed daily from the same distance. On this occasion the wound did not actually break down, the healing appeared to progress very well for the first 7–10 days. Thereafter the wound stretched, the scan became quite thin and transparent and erythematous and it remained a rather tenuous thin looking scar which could not have been put under any tension, three weeks and even up to eight weeks after the biopsy. It was interesting that in the family, father and one of father's maternal aunts, when even they have had lacerations, have taken a good deal longer to heal than normal.

The skin from the biopsy was sent to the Clinical Research Centre at Northwick Park Hospital. London, and it was found that her fibroblasts did not produce abnormal collagen types. It was also found that collagen synthesis was not different from that of normal controls. On light microscopy and electron microscopy no abnormalities were grossly detected.

Nutrition throughout this episode was adequate, although her growth percentiles were 3 to 4 standard deviations below the mean.

Her subcutaneous fat distribution was satisfactory. Results of numerous other investigations, not included here, were all within normal limits.

CHAPTER 13

GROUP 'B' (patients investigated on account of cerebral tumours). CASE NUMBERS 88-90.

Case Number 88

Name S.D.

Age 10 years 10 months

Method Right frontal Rickham reservoir

Medical Diagnostic Background

This girl had an extensive resection of a Grade 1 cystic astrocytoma of the left cerebellar hemisphere 7 years earlier. She was admitted on this occasion with an 8 week history of early morning headache and vomiting. She held her head deviated to the right, was unsteady on heel to toe walking, had some inco-ordination of her left arm and horizontal and vertical nystagmus. Accordingly a further posterior fossa exploration was carried out and a large astrocytic gliomatous cyst of the left cerebellar hemisphere was evacuated. The cyst wall and some tumour tissue was excised and the cyst was brought into communication with the fourth ventricle, and by a tube to the peritoneal cavity, that is, a cysto-peritoneal shunt. She developed a post-operative infection and a right frontal Rickham reservoir was inserted and the cysto-peritoneal shunt removed. Treatment at this stage consisted of intra-ventricular antibiotics. She was then transferred for pressure monitoring.

Temperature 37

Zero Level of the anterior horn for the lateral ventricle.

Duration 5 hours

Indication Having intrathecal drugs, but increasingly symptomatic with lethargy and increasing papilloedema and swelling at the site of her posterior fossa exploration.

Resting Ventricular Pressure

20 mm Hg.

Stress Ventricular Pressure

53 mm Hg maximum and in excess of 30 mm Hg in sleep.

Result Raised ventricular pressure

Action C.S.F. release and maintenance antibiotics and intravenous fluids until the C.S.F. was sterile, when she was transferred back for a Pudenz low pressure cysto-peritoneal anastomosis reconstitution to be carried out.

Cardiac/Respiratory Artefact

CR = 3 mm at 20 mm Hg awake

CR = 16 mm when higher pressure excursions occur.

Ventricular Dilatation/Cortical Mantle

A C.T. scan at this time showed an increase in the ventricular size, the fourth ventricle was large and there was no obvious obstruction and no increase in the cystic formation or abscess formation.

Pressure Recordings Relief of 10 mls of C.S.F. dropped the ventricular pressure level to 12 mm Hg from 30 mm Hg immediately. However, as the C.S.F. production increased, the pressure gradually increased again in excess of 20 mm Hg. At this stage 100 mls of a 20% solution of Mannitol was administered I.V. with only a gradual reduction in the level of ventricular pressure over a 22 minute period (Fig. 138) and following this the pressure is still at an unacceptably high level, although it had dropped from 40 mm Hg to 25 mm Hg. So, although Mannitol does have an effect, it is slow and probably related to associated brain swelling due to hypoxic ischaemic changes in the cortex at this stage. Infection was well under control by the time of this V.P.M.

Follow Up Following the new low pressure Pudenz cysto-peritoneal anastomosis, she was discharged, only to be readmitted 4 days later with progressive drowsiness, vomiting and

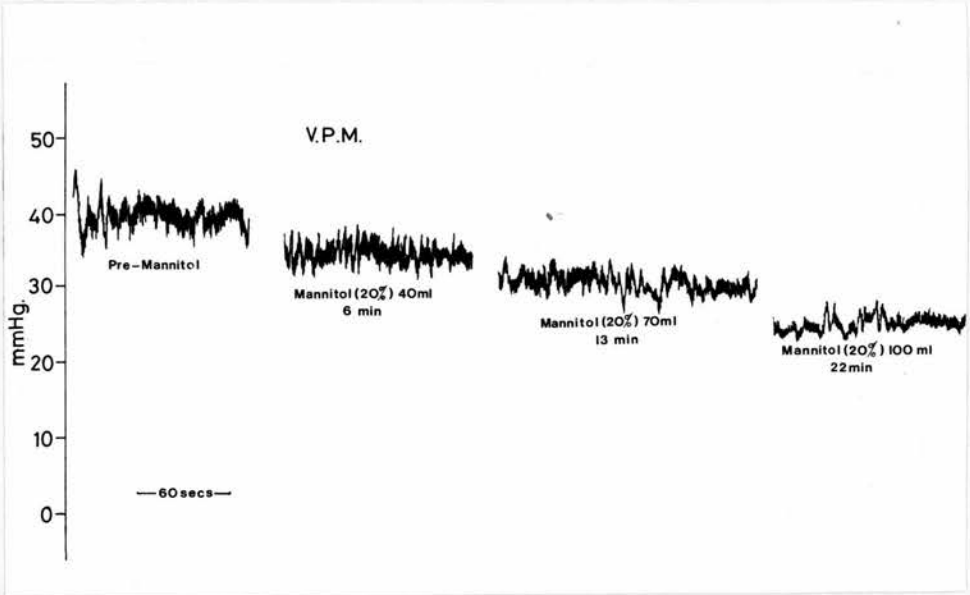


Fig. 138

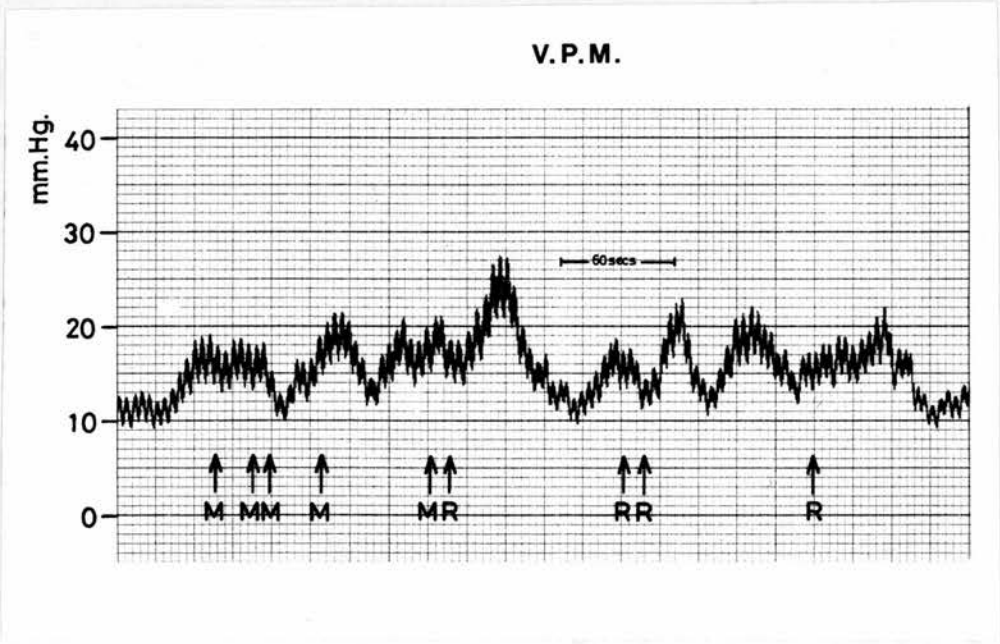


Fig. 139

headache.

C.S.F. examination and measurement of pressure on this occasion was normal. However there was progressive neurological deterioration and a C.T. scan was carried out which showed that although the posterior fossa related to her astrocytoma was draining adequately, there was no doubt that there had been a very definite increase in her hydrocephalus which involved all four ventricles, and therefore an emergency ventriculo-peritoneal anastomosis was performed with a Pudenz medium pressure system from the right lateral ventricle. Following this she settled quickly and was discharged. This is to be expected from posterior fossa space occupation even when the C.S.F. pathways remain clear there is interference with the large venous channels in this area which results in a back pressure and an increase in the choroidal arterial hydrostatic pressure. For Starling's law to remain effective, the hydrostatic and osmotic pressure across the choroidal capillaries must be in equilibrium with the hydrostatic and osmotic pressures in the choroidal interstitial space, and in the C.S.F. and therefore an increase in C.S.F. production occurs and early hydrocephalus with posterior fossa space occupation (Andeweg 1976). This increase in C.S.F. production and increase in ventricular size was not managed effectively with the one cysto-peritoneal anastomosis and required a second ventriculo-peritoneal anastomosis as well.

Case Number	89
Name	S.D.
Age	11 years
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

Posterior fossa cystic astrocytoma as before.

Temperature Normal

Zero Inter-ventricular foramina

Duration 10 hours

Indication A four week history of increasing headache, unsteadiness and a tendency to vomit, but on examination there was no doubt that both ventriculo-peritoneal and cysto-peritoneal flushing devices were clinically not working. At this stage her fundi were normal, pupils were dilated, equal but reacting. There was evidence of a sixth and seventh nerve paresis. Plantars were bilaterally extensor. There was marked ataxia of the upper limbs and lower limbs so that the indication was twin blocked shunts with increasing unsteadiness and nausea.

Resting Ventricular Pressure

20 mm Hg.

Stress Ventricular Pressure

62 mm Hg peaks in sleep. During this 'light' phase of sleep there was a rapid rise of ventricular pressure, with increasing irritability, lethargy, dysarthria and she awoke transiently and complained of headache.

Ventricular Dilatation/Cortical Mantle

See previous C.T. scan report.

Pressure Recordings The frequency of sleep oscillations is seen in Fig. 139 and the extracted numerical values for the frequency intervals were analysed. The 'M' and 'R' makings indicate the appearance of myoclonic movements and rapid eye movements respectively. Fig. 140 shows a plateau wave which occurs while lightly asleep, in a semiconscious state at point X and when she

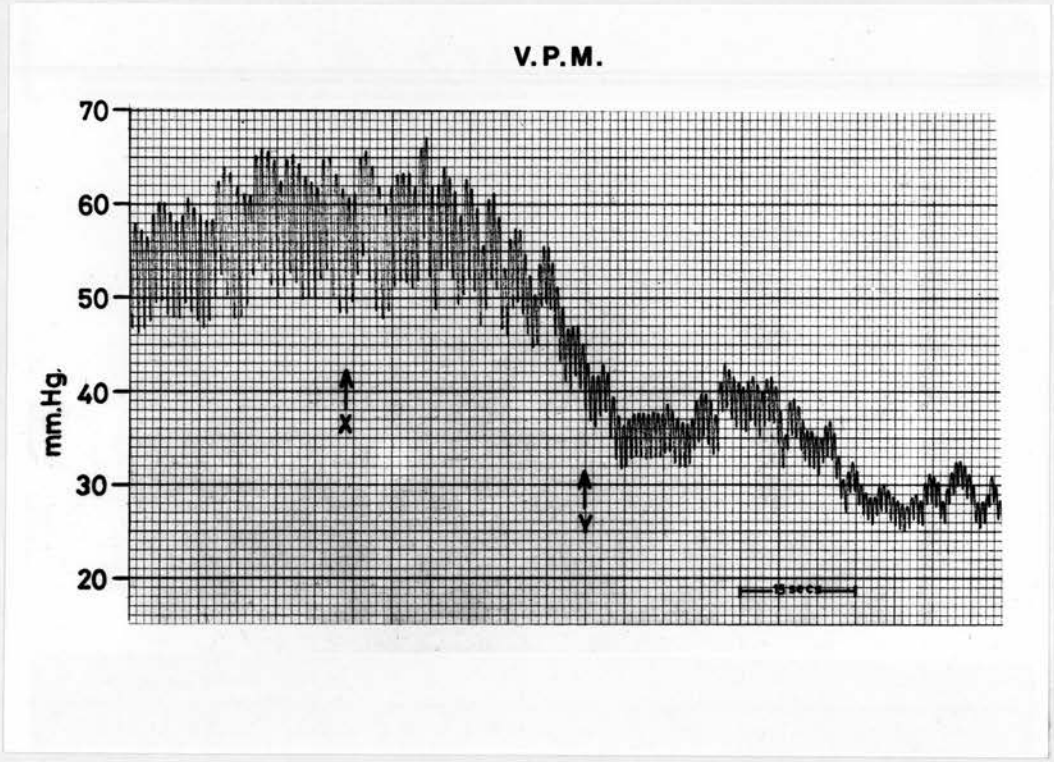


Fig. 140

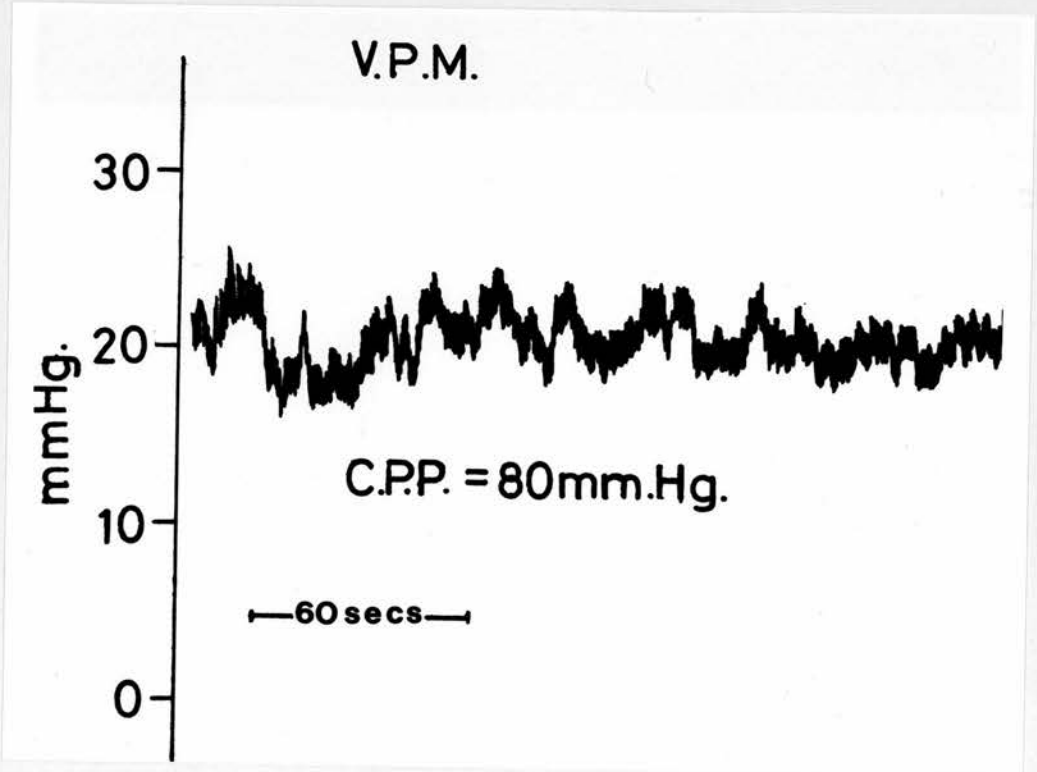


Fig. 141

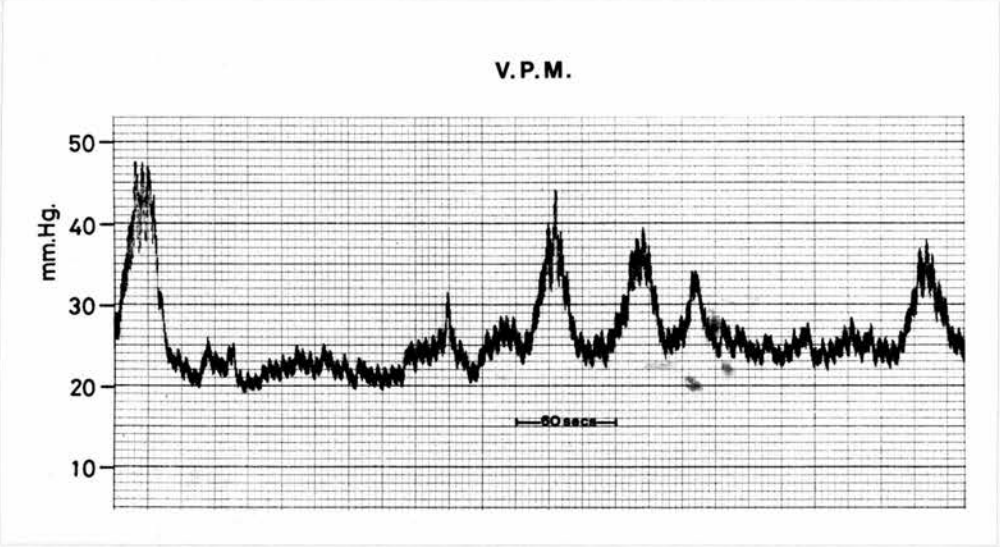


Fig. 142

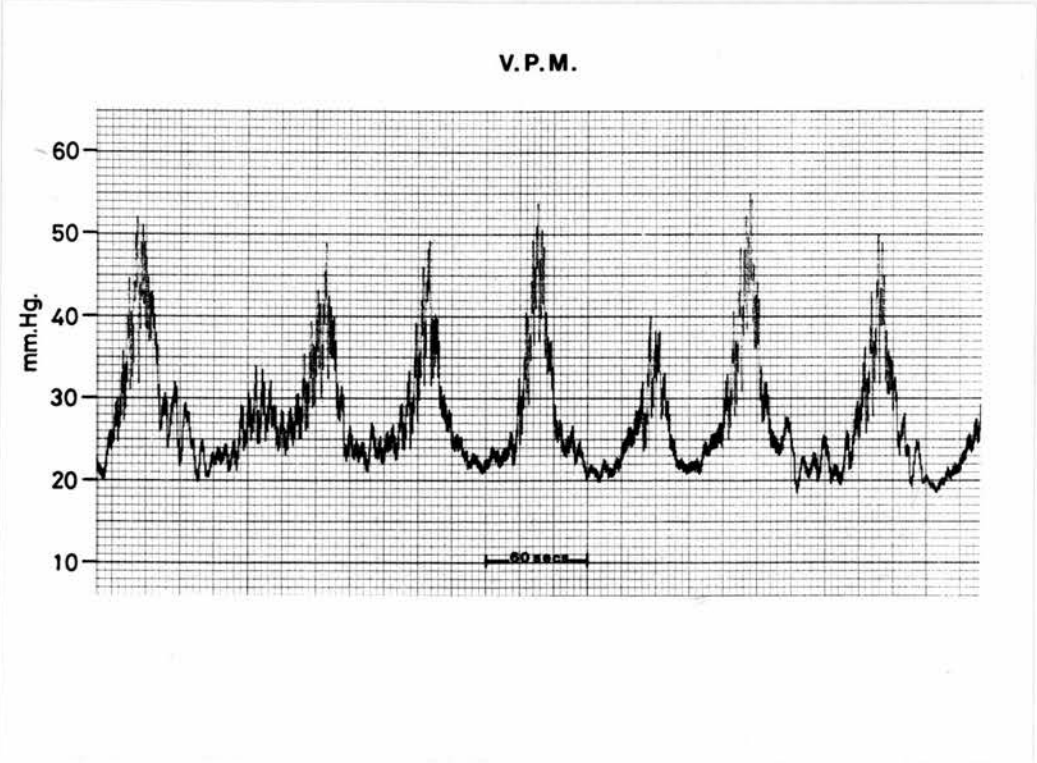


Fig. 143

awakens at point Y, with a continual drop in the level of ventricular pressure. She was complaining of pain about the posterior fossa exploration site at this time.

After this plateau wave, while awake, the C.P.P. was 80 mm Hg (Fig.141) and in a further example of 'light' sleep, Fig. 142 the C.P.P. was 70 mm Hg. Fig. 143 shows a third section of 'light' sleep with pressures ranging from 20 to 55 mm Hg, and on this occasion these frequency intervals look amazingly regular and appear to recur about every 60 seconds. If they do not appear at each 60 second interval and one is missed out, the amplitude of the succeeding pressure elevation is larger at the 2 minute interval. Following this section of trace, she eventually settled into deep sleep, still at a pressure level of about 20 mm Hg. Pumping of the flushing devices of the ventriculo-peritoneal shunt made only a small difference to her ventricular pressure and the cysto-peritoneal shunt was non-functioning.

Action She was taken to theatre when the ventriculo-peritoneal shunt (proximal end) was revised and then a further posterior fossa exploration was carried out, in order to replace the proximal end of the cysto-peritoneal anastomosis. At this time biopsy of the remaining tissue within the posterior fossa cyst was carried out and this demonstrated the presence of persistent tumour. She made a good recovery following this operation. One further period of pressure monitoring on this child was performed but has not been included in this series.

Case Number 90

Name G. McL.

Age 12 years

Method Right frontal Rickham reservoir

Medical Diagnostic Background

Posterior fossa astrocytoma; the child having daily radiotherapy. He had a ventriculo-peritoneal shunt and a reservoir in situ because of secondary hydrocephalus, inserted 3 weeks prior to this pressure monitoring.

Temperature 36.2

Zero 1" below the upper cortical subarachnoid space.

Duration 20 hours

Indication Unresolving papilloedema and visual disturbance, despite shunting.

Resting Ventricular Pressure

6.5 mm Hg.

Stress Ventricular Pressure

41 mm Hg. Maximum stress during a bowel movement.

Cerebral Perfusion Pressure

At the commencement of monitoring 83 mm Hg.

Result Normal ventricular pressure

Action Nil. It was considered his marked visual disturbance was secondary to infarction of the occipital lobes, as a result of posterior cerebral insufficiency.

Cardiac/Respiratory Artefact

CR = 0.75 mm at rest.

Pressure Recordings At the outset, ventricular pressure of 5 mm Hg and B.P. 116/60, i.e. C.P.P. = 83 mm Hg. Three and a half

hours later, B.P. 122/80, ventricular pressure 8 mm Hg.

C.P.P. 93 mm Hg.

Rather asynchronous pressure waves are seen in Fig. 144 while the patient is awake, quiet, supine with a normal mean level of ventricular pressure. They become quite synchronous and regular when he is in early sleep, Fig. 145 .

In this child a shift of the zero in the vertical plane of 1" was equivalent to 1 mm Hg difference in the recorded ventricular pressure.

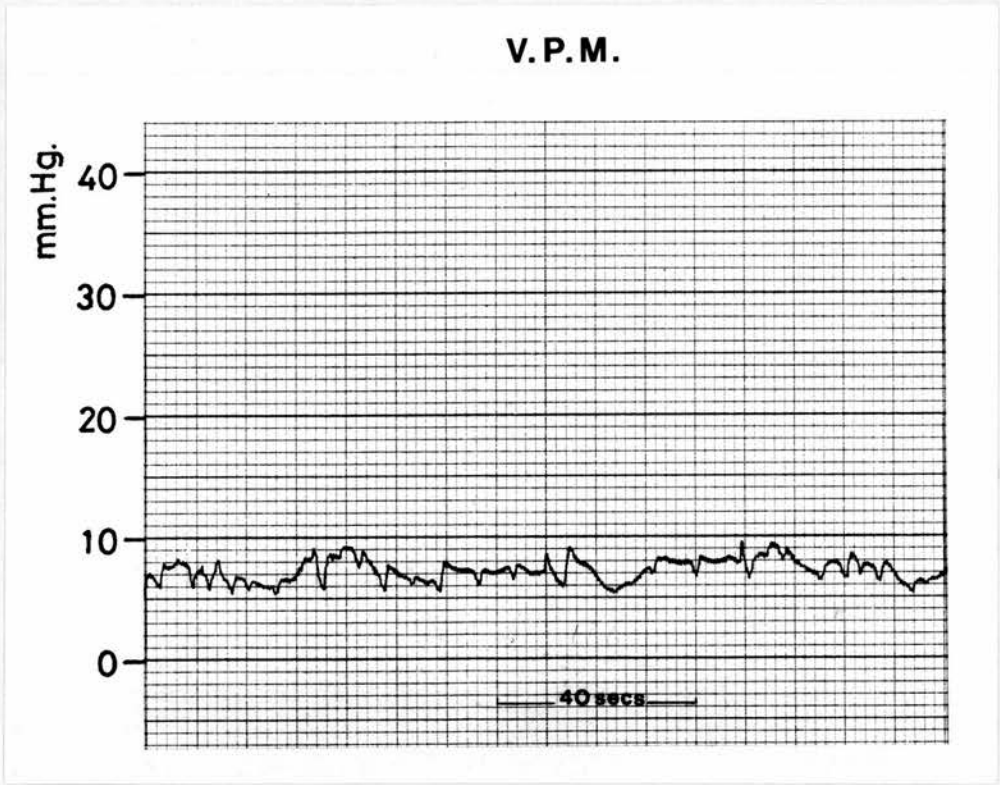


Fig. 144

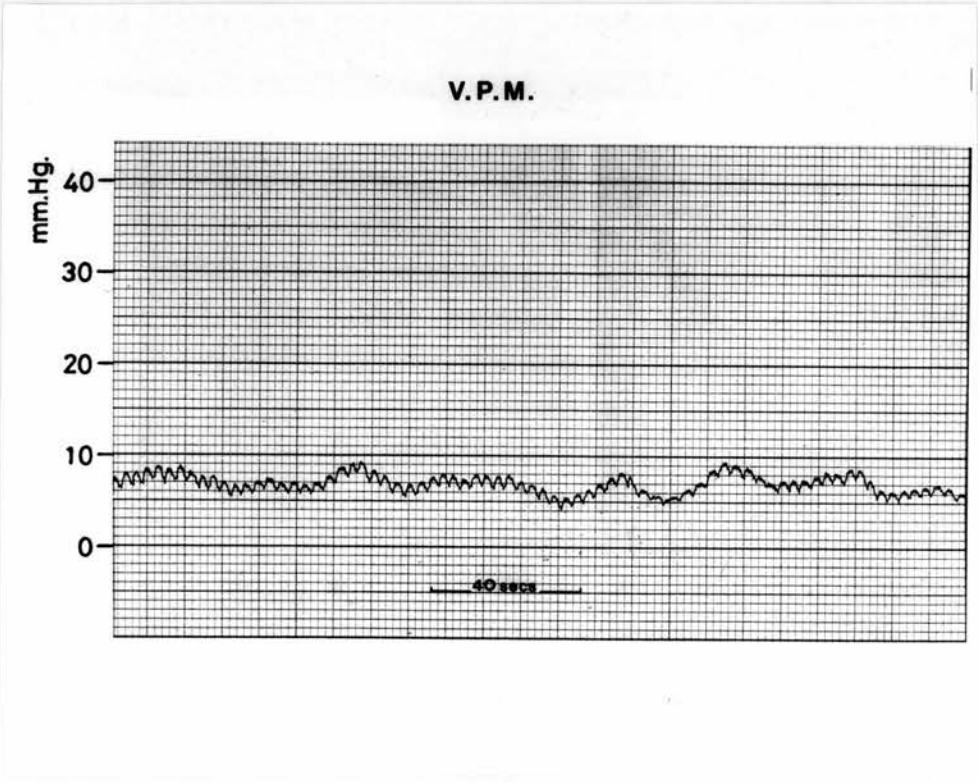


Fig. 145

CHAPTER 14

GROUP 'C' (patients investigated on account of brain swelling or enlargement). CASE NUMBERS 91-98

Case Number	91
Name	S.D.
Age	6 months
Method	Right frontal Rickham reservoir

Medical Diagnostic Background

This child was born by a caesarean section at term and weighed 2,900 grams. There were no perinatal problems. Because of craniostenosis with an oxycephalic skull, a maxillary hypoplasia and a syndactyly, he was labelled as Crouzon's Disease.

He was admitted at 12 days of age with a pyloric stenosis and a Ramstedt operation was carried out with an uneventful post-operative course. He was then admitted at 4 months of age because of a reported apnoeic episode. He had also had a number of 'turns' described by his mother, when he became quiet, his eyes opened very wide, he became rigid and then cyanosed. They lasted approximately 1 minute and in all he had some 10 of these.

Routine investigations including an E.E.G., glucose, calcium, magnesium, amino acids, E.C.G., chest and barium x-rays were all normal.

He was readmitted 2 weeks later with similar complaint of 'fits' as previously described. His OFC had increased normally to 41.3 cms, having been 31 cms shortly after birth. However his optic discs appeared a little pale and an E.E.G. on this occasion showed some spike activity in the right temporal region which became generalised. He was therefore commenced on Phenytoin and he had an elective Rickham reservoir inserted for monitoring of his intracranial pressure. A C.T. scan was also carried out. Unfortunately he developed a post-operative ventriculitis. The indication therefore for measuring this boy's pressure was 'fits' occurring in a child with cranial

synostosis where raised intracranial pressure can occur, as well as blindness and deafness because of a limited intracranial volume.

Temperature 37

Zero 2" below upper cortical subarachnoid space.

Duration 3 hours

Indication As before

Resting Ventricular Pressure

10 mm Hg.

Stress Ventricular Pressure

54mm Hg maximum stress. 17.5 mm Hg maximum peaks in sleep.

Result Normal awake ventricular pressure.

Action Nil re C.S.F. pressure.

Cardiac/Respiratory Artefact

CR = 3 mm at rest.

Ventricular Dilatation/Cortical Mantle

Brain and ventricular system normal although the sulci in both Sylvian fissures were perhaps larger than normal.

Pressure Recordings

Fig. 146 shows a section of ventricular pressure tracing during light sleep with an oscillating level of ventricular pressure. An insert containing 3 channels of E.E.G. recording and 1 channel of E.C.G. is included. The E.E.G. recording is at full gain and it can be seen that during these pressure fluctuations, 'sleep spindles' are occurring. One of the clinical features of lighter R.E.M. sleep in this child is that of 'tongue movements'. This patient spent quite a considerable time in R.E.M. sleep and it is important to remember that he had ventriculitis prior to this V.P.M. The relatively lower amplitude E.E.G. tracing is common in light sleep. The sleep spindles last approximately 2 seconds and can occur at any point on the ventricular pressure waves.

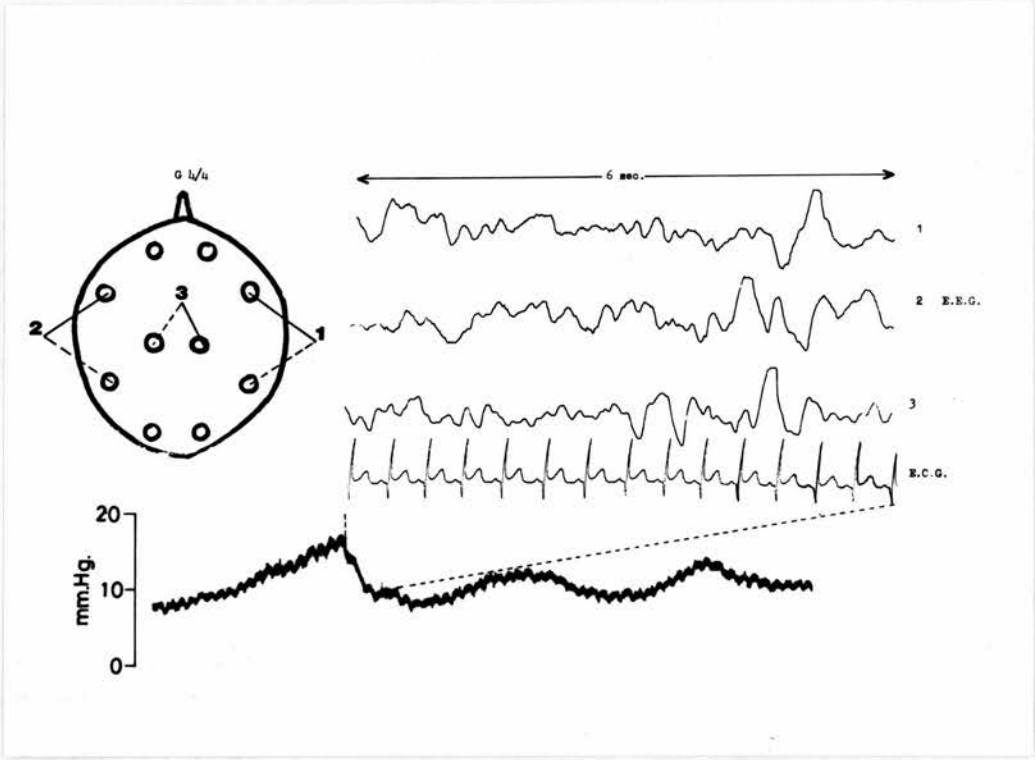


Fig. 146

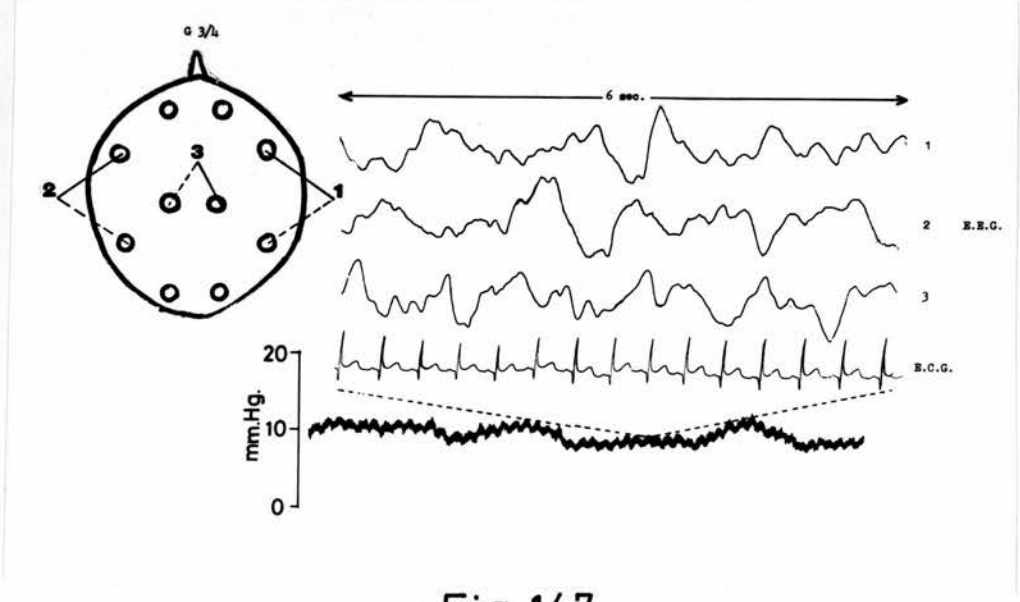


Fig. 147

Fig. 147 shows a similar combined diagram of ventricular pressure E.C.G. and E.E.G. during a deep or 'quiet' phase of sleep and characteristically the amplitude of the E.E.G. tracing is very much increased at this stage (only $\frac{3}{4}$ gain here) with no apparent spindles and no clinically detectable movements. The ventricular pressure is lowest at this time.

Fig. 148 shows a similar conglomerate recording following the previous two, when the child is awake and movements occurring. Here the E.E.G. is of low amplitude with very little activity and contrasts with the E.E.G. findings in the previous two.

Points of Interest At the time of monitoring this child's pressure, he was still on Gentamycin and Cloxacillin intrathecally. It is important to ensure in children with cranio-facial dysostosis that the head is continuing to grow, ie. that the OFC is increasing and that fusion of a number of sutures is not resulting in brain compression and raised intracranial pressure with papilloedema and increasing deafness. This child's fits do not seem to be a result of pressure but a result of an underlying abnormal brain.

Case Number	92
Name	E.I.
Age	3 years
Method	Ventricular cannulation

Medical Diagnostic Background

This boy had a complex cyanotic congenital heart disease with dextrocardia, a single ventricle and tricuspid atresia, transposition of great arteries, pulmonary valve and infundibular stenosis and a septal defect. He had been admitted for further assessment of his cardiac condition because of increasing cyanosis and

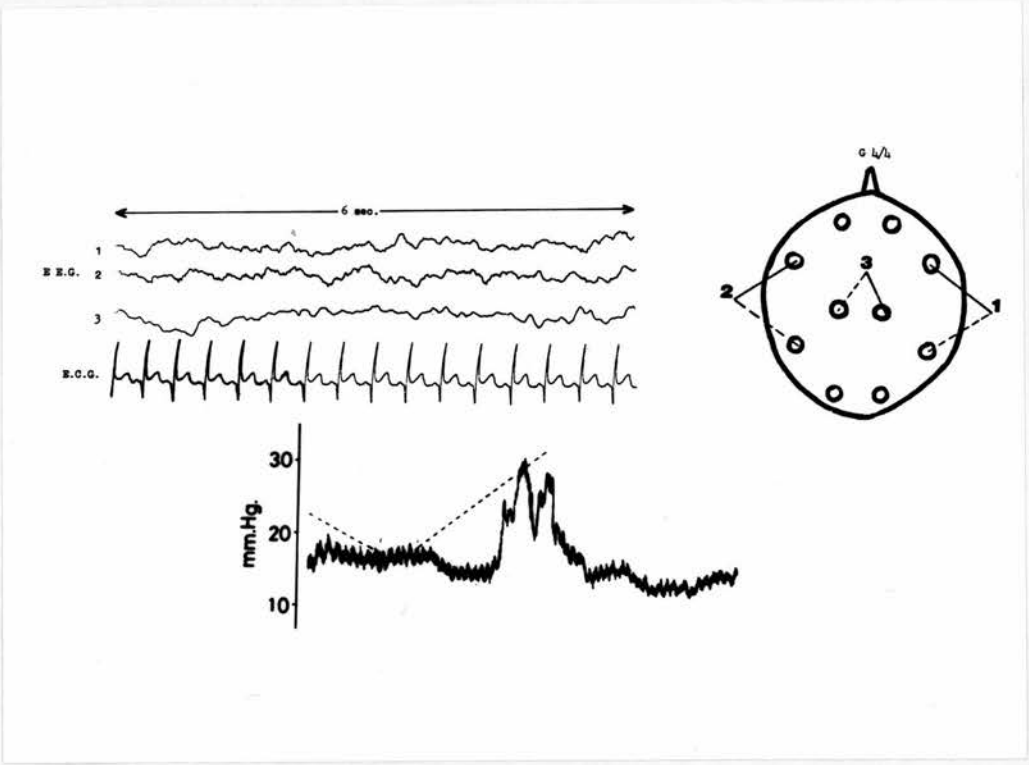


Fig.148

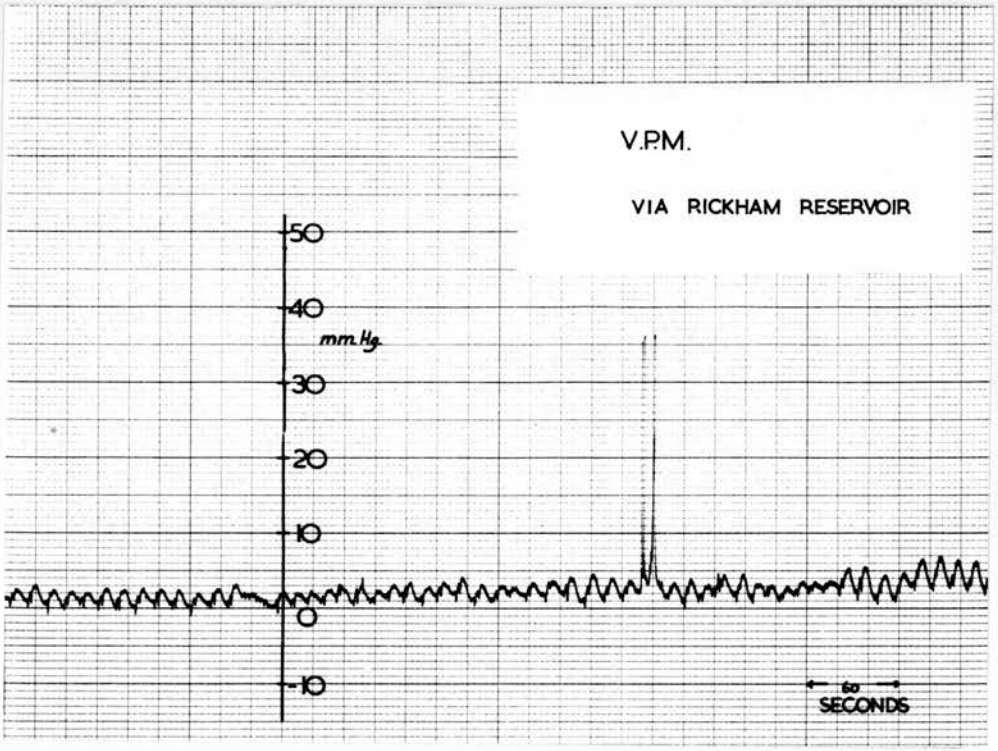


Fig.149

tiredness and intermittent chest pain. A decision was made to have a cardiac catheter for the purposes of deciding whether further palliative surgery or more radical surgery would be helpful.

The cardiac catheterisation procedure was uneventful apart from a very short period of bradycardia lasting no more than 45 seconds. However, on return to the ward he appeared still heavily sedated and only responded to painful stimuli. It seemed probable that he had been hypersensitive to the Pethidine compound mixture which is particularly likely to be the case in children with cyanotic congenital heart disease.

Later he had no lightening of his consciousness, pin point pupils and he developed intermittent decerebrate posturing. His respiration was sighing and despite Pethidine antagonists remained so. His anterior fontanelle was still widely patent and he had been thought previously to have had an arrested hydrocephalus.

He was given an intravenous infusion of Mannitol. He went on to have 'brain stem fits' and an increasing pyrexia. He became hypertensive and had systemic hypoperfusion. Accordingly ventricular cannulation was carried out through his rather wide anterior fontanelle.

Temperature	Elevated at 38.5
Zero	Inter-ventricular foramina
Duration	3 hours
Indication	Decerebration and unconsciousness, post-cardiac catheter.

Resting Ventricular Pressure

15 mm Hg.

Stress Ventricular Pressure

18 mm Hg.

Result Raised intracranial pressure

Action Open ventricular drainage, but despite this and symptomatic treatment of his brain stem fits, etc., his condition deteriorated and he died.

Cardiac/Respiratory Artefact

C = 1.25 mm at 15 mm Hg.

C = 5 mm at 35 mm Hg.

R = 6.25 mm at 15 mm Hg.

R = 16.25 mm at 35 mm Hg.

Post-Mortem Confirmed the complex cyanotic congenital heart disease. There was major infarction of the brain stem and occlusive lesions in the basilar arteries and branches to the brain stem. Massive infarction of the superior third of both cerebellar hemispheres and pons was evident. The brain itself weighed 1600 gm. and was abnormally large. A coronal section showed that there was only slight dilatation of the lateral and third ventricles.

Children with cyanotic problems during cardiac catheters, particularly during the injection of contrast media, which is viscus and may cause sludging within the circulation and spontaneous thrombosis. This is a recognised, although rare complication. A possibility of emboli being induced at the time of injection seemed unlikely clinically, because of the selective pattern of damage within the brain stem it would be unlikely to be an embolic process.

A number of factors probably contributed; his long standing arterial saturation of only 60%, his known long standing hydrocephalus which may have resulted in some stretching and slowing of the circulation at the base of the brain and any sluggishness in the circulation may have been accentuated by injection of contrast media which could have set the scene for the development of occlusive lesions with

ischaemia and infarction.

Case Number 93

Name G.H.

Age 5 years

Method Emergency Rickham reservoir

Medical Diagnostic Background

He was born following a pregnancy complicated only by a number of maternal urinary tract infections. At 41 weeks he was delivered by S.V.D. at a birth weight of 8lbs 4 ozs.

At the age of 6 weeks he was noticed by the local Child Welfare Clinic to have a 'large head' with a large full anterior fontanelle, a high pitched cry and to be generally a little floppy with a 'hairy patch' at the base of his spine.

Three weeks later his OFC was 45 cms (see OFC chart). The full anterior fontanelle measured 3.9 by 6 cms, 'sunsetting' was present and he was lethargic and irritable with a high pitched shrill cry, hydrocephalic facies, distended scalp veins and fundal venous congestion. He had poor head control, weak grasp and truncal responses, a positive S.T.N.R. and stronger A.T.N.R.'s, absent Moro and stepping reflexes and generalised hypotonia.

A detailed family tree showed that a 4 year old sibling also had a 'large head' as did father and paternal grandfather. The child's paternal aunt was a mongol.

At his first admission the results of investigations were: viral titres, especially Rubella were all less than 8, and encephalogram was midline; a skull x-ray was normal, bilateral subdural taps were negative, a spine x-ray showed a dorsal spina bifida and mild scoliosis to the left. An L.A.E.G. showed an external hydrocephalus and some

cerebral atrophy and a mildly dilated ventricular system. At this first admission a theco-peritoneal shunt was inserted and a right frontal burr hole was made.

From then until the age of 4 years he was followed up regularly and his OFC progressed. The maximum OFC appears on the OFC chart at the point at which the theco-peritoneal shunt was inserted. Subsequent OFC measurements were at 1 year 47.5 cms, 18 months 48.5 cms, 2 years 3 months 49.5 cms, 2 years 8 months 50 cms and 3 years 2 months 50.3 cms.

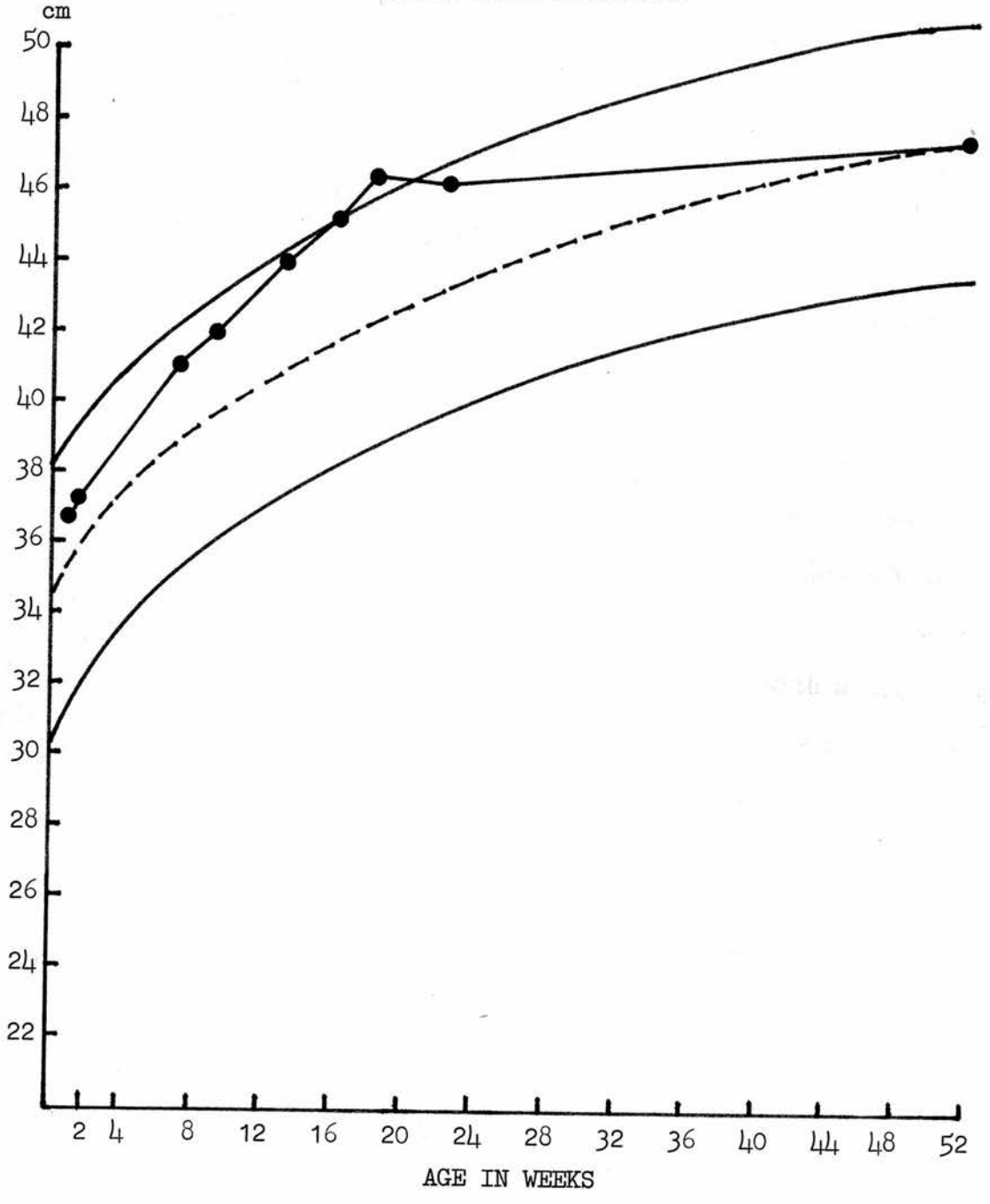
His motor functions were about 1 year retarded and he had a broad based stomping gait with a tendency to recurvatum of the left leg and slight dwarfing below the knee. Obviously a central gait problem of ataxic diplegia associated with his hydrocephalus.

His speech development was normal and it was noted earlier that he had occasionally dizzy spells associated with upper respiratory infections.

At the age of 4 years he was admitted with headaches, vomiting, lethargy, meningism and enlarged parotid glands. C.S.F. at the time showed 170 white cells and when repeated 3 days later showed 1,000 lymphocytes/cu.mm and a C.S.F. glucose of 55 mm%. A presumptive diagnosis of mumps meningitis was made from which he recovered spontaneously.

At 5 years of age when followed up at Out Patients, it was noted that he was settling in well at school, he was above average intelligence and there were no behaviour problems. He was right handed but with a primitive grasp and some functional difficulties with his hands. He was still ataxic, particularly with upper respiratory tract infections and in retrospect his parents mentioned some 12 of these attacks associated with upper respiratory tract infections. His

O.F.C. CHART FOR MALES



O.F.C. at this time was 52 cms. He had extensor or plantar responses, hypotonia and brisk DTR'S, more so on the right, and ankle clonus.

He was then admitted as an emergency following 24 hours vomiting with a varying conscious state and a normal temperature (T = 37.2). His pulse rate was 100 per minute, he had small reactive pupils, bilateral abducens weakness, fundal venous congestion, upgoing plantar responses with very brisk reflexes and a positive Kernig's sign. The following morning he had a cardiac arrest and was treated with Dexamethasone, Mannitol, Sodium Bicarbonate and routine cardio-respiratory support. Following this he immediately had a right frontal Rickham reservoir inserted and post-operative ventricular pressure monitoring was carried out through the Rickham while in the Intensive Care Unit and treatment was continued with intravenous Epanutin, Ampicillin, Cloxacillin and Dexamethasone.

Fig. 149 shows the ventricular pressure immediately on arrival in the Intensive Care Unit after the Rickham reservoir was inserted. No doubt the loss of C.S.F. and osmotic diuretics resulted in this low pressure. Fig. 150 shows a dramatic rise in ventricular pressure without any change in the child's physical state. A classival plateau wave is seen and drainage of C.S.F. reduces the pressure temporarily. Fig. 151 shows the effects of drainage of C.S.F. to be minimally useful in controlling the C.S.F. pressure in this case and the arrows indicate where the C.S.F. was released. Fig. 152 shows the level of intracranial pressure immediately after one hour of open ventricular drainage. Fig. 153 shows a less dramatic rise of intracranial pressure which settles spontaneously without any medication or intervention. Fig. 154 shows the effect of Mannitol infusion in this child causing a rapid reduction in the level of

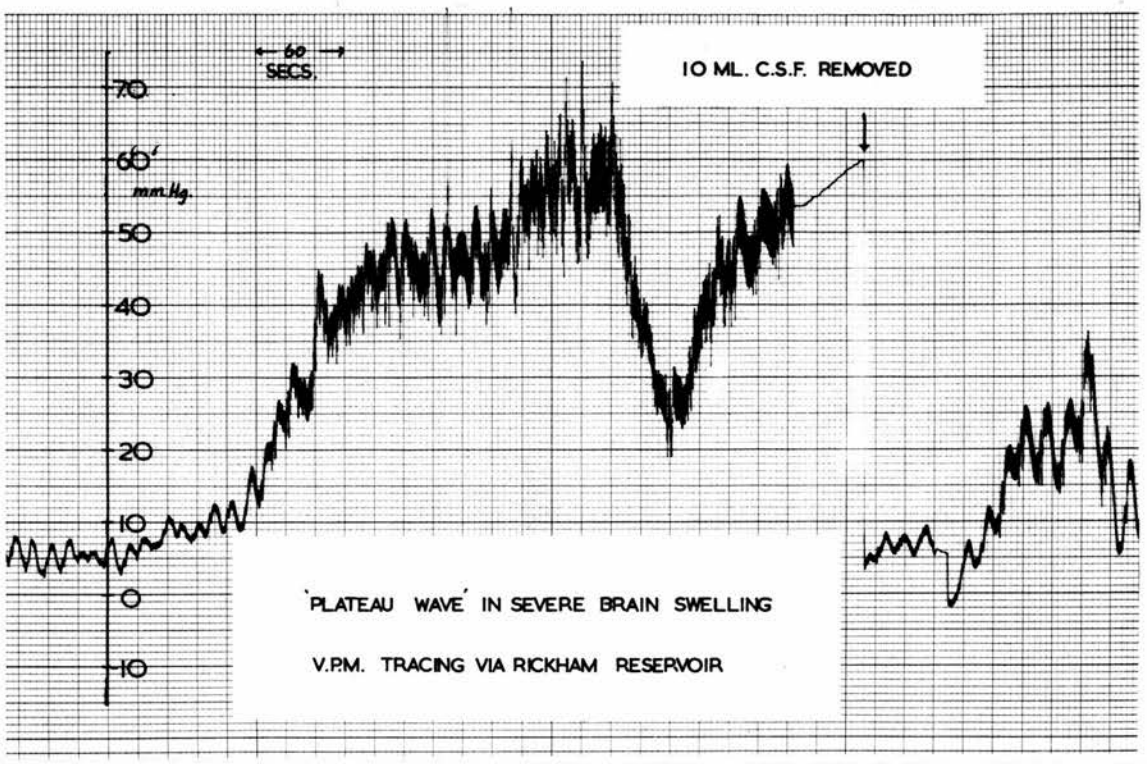


Fig. 150

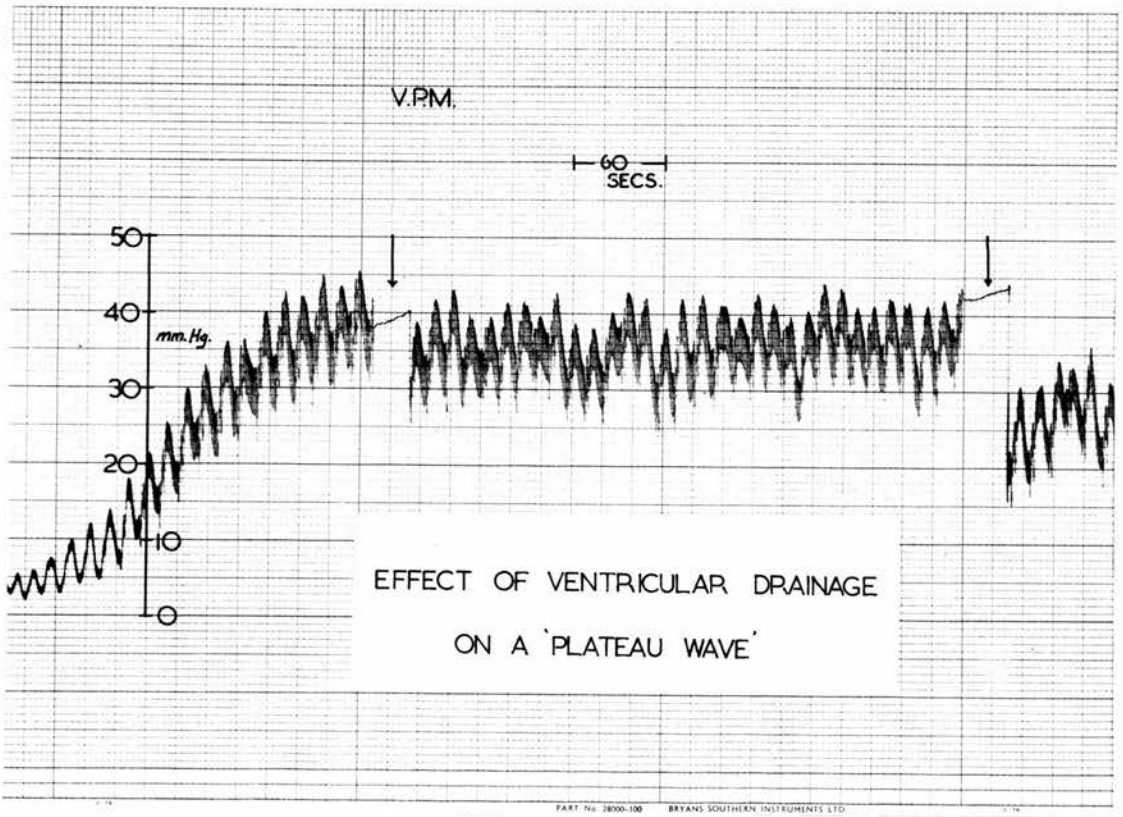


Fig. 151

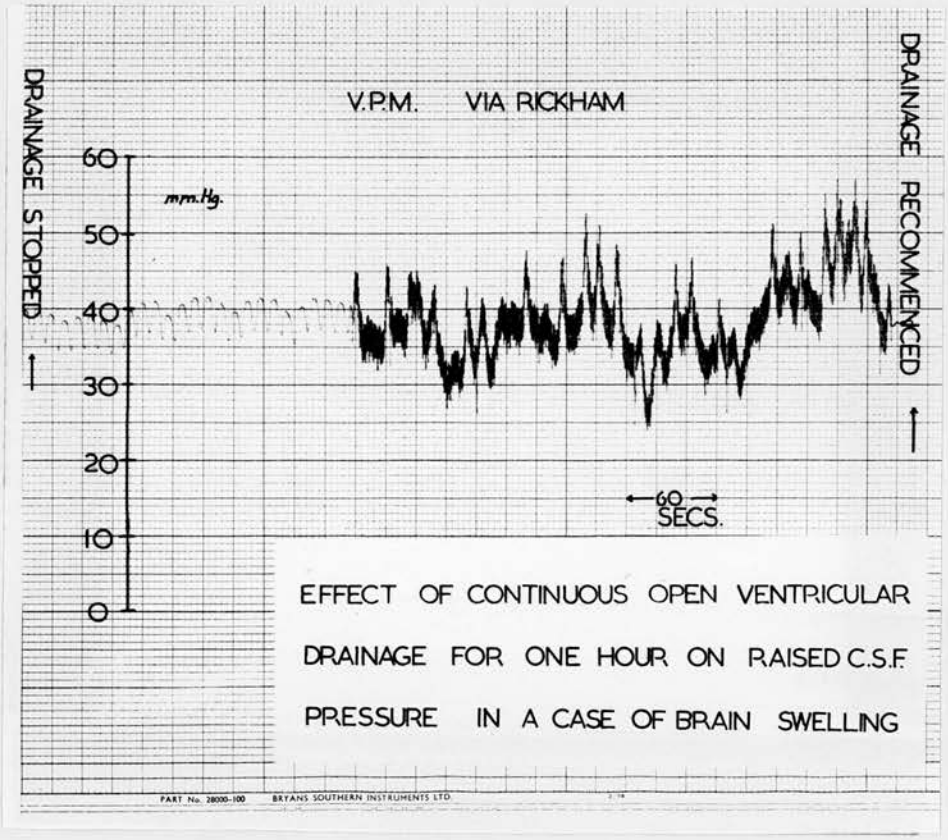


Fig.152

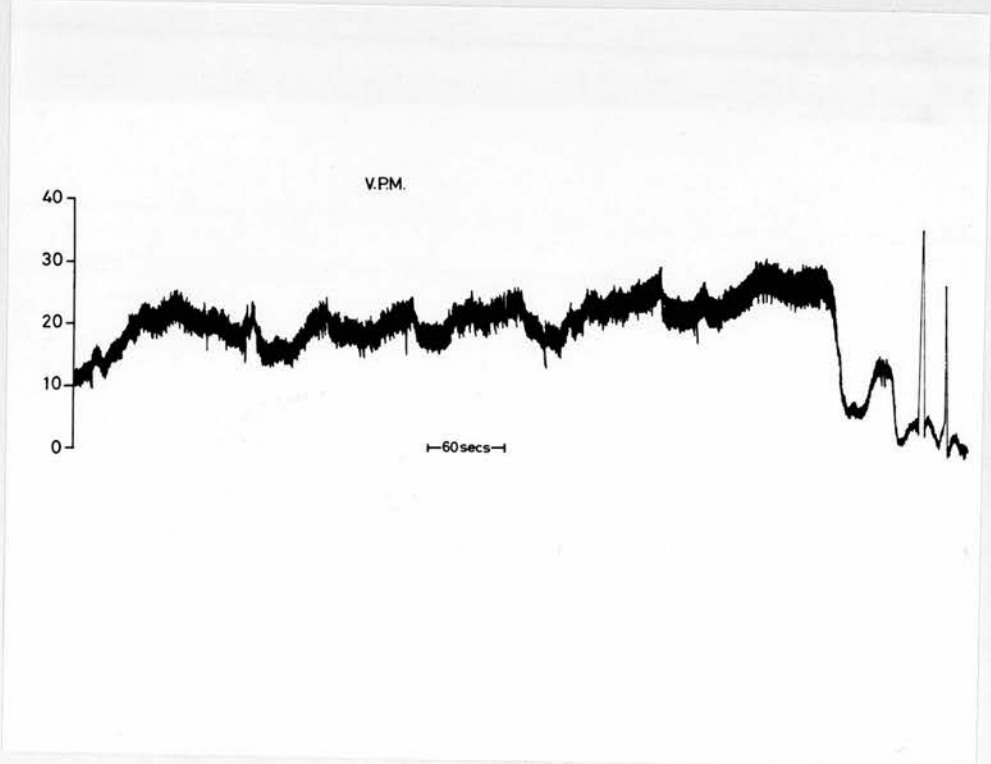


Fig.153

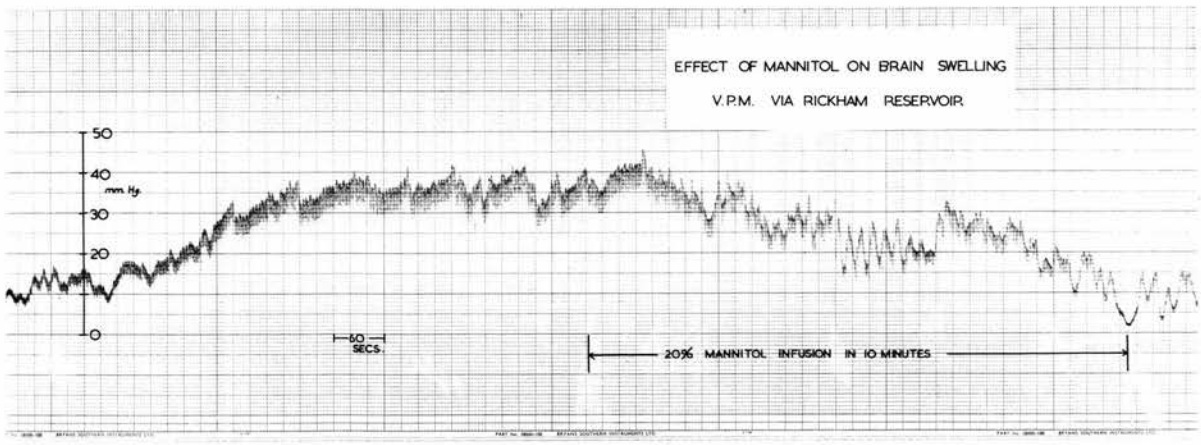


Fig.154

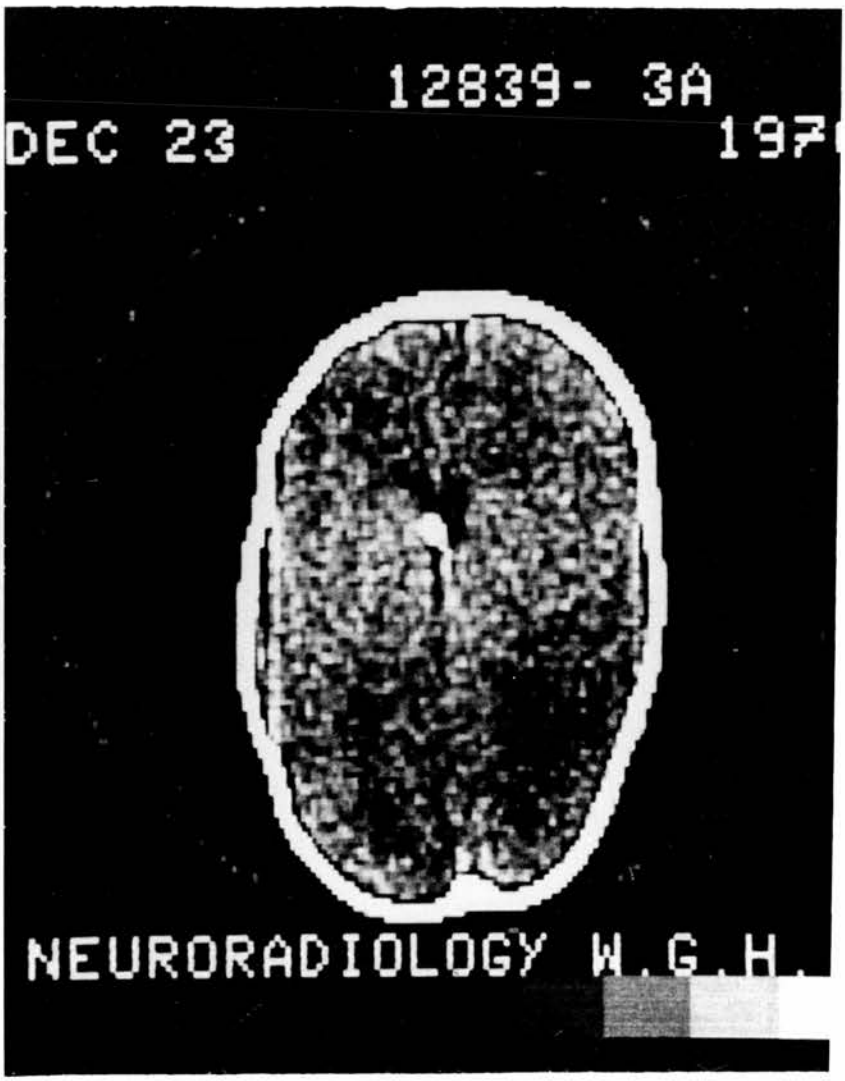


Fig.155

intracranial pressure. A 20% Mannitol infusion was run in, while the C.S.F. pressure was measured, thus the quantity of Mannitol could be titrated to the level of ventricular pressure. A C.T. scan (Fig.155) obtained during this admission shows a normal size ventricular system. The parents reported 24 similar episodes of headache, vomiting and head retraction associated with upper respiratory tract infections had always previously settled within 24 to 48 hours with no treatment. They also noted that he generally tended to sleep with his head slightly retracted. The massive increases in C.S.F. pressure levels were not associated with changes in the child's vital signs.

Zero — Upper cortical subarachnoid space.

Duration — 36 hours

Resting Ventricular Pressure

Variable but raised.

Stress Ventricular Pressure

A maximum of 74 mm Hg peaks during plateau waves.

Result — Raised intracranial pressure due to brain swelling. This was confirmed by C.T. scan and at operation.

Cardiac/Respiratory Artefact

CR = 26 mm at 35 mm Hg.

CR = 5 mm at 10 mm Hg.

C = 1.25 mm at 3 mm Hg.

R = 6.25 mm at 3 mm Hg.

OFC — 52 cms

Ventricular Dilatation/Cortical Mantle

Small ventricles on C.T. scan.

Summary — This child with hydrocephalus treated initially with a theco-peritoneal shunt and frontal burr hole subsequently had an episode of mumps meningitis at 4 years of age.

After this, minor episodes of raised intracranial pressure appeared to have occurred, each time associated with an upper respiratory tract infection and settled spontaneously on each occasion. A similar episode of raised intracranial pressure on this occasion resulted in a cardiac arrest with brain swelling. The possibility of acute brain swelling on each occasion being associated with inappropriate ADH secretion or a Reyes type syndrome has now been excluded. A number of important points are illustrated. Firstly, the advisability of having a Rickham reservoir or at least a burr hole, in all cases where a theco-peritoneal shunt is employed. Secondly, episodes of raised intracranial pressure which settle spontaneously and seem to be initiated by an upper respiratory tract infection. On this occasion there was on Coxachsie B neutralisation tests, a rising titre to Cox B_5 from 32 to 128. Thirdly, mumps meningitis may have converted him for a communicating to non-communicating hydrocephalus. Fourthly, open ventricular drainage through a Rickham reservoir had little lasting effect on the plateau waves, whereas a Mannitol infusion was effective within 10 minutes and without a rebound pressure rise. A number of these plateau waves occurred over a number of hours, while in the Intensive Care Unit and as mentioned before, at definitive shunting the lateral ventricle was entered at 4 cm and there was evidence of gross brain swelling.

Case Number	94
Name	G.H.
Age	6 years
Method	Rickham reservoir

Medical Diagnostic Background

As before.

Temperature Normal

Zero Upper cortical subarachnoid space.

Duration 1 hour

Indication He was admitted on this occasion having been well prior to the day of admission when he had complained of a headache in the morning. His neck had become slightly stiff, he had vomited, was anorexic and had a mild sore throat.
His temperature on admission was 37.5. His tonsis were large and 'injected' but there was no frank pus. He had marked neck rigidity and Kernig's sign was positive. Pupils were sluggish in response to light and he had hyperaemic fundal discs. So the indication was of a child with known episodic brain swelling with symptoms suggestive of raised intracranial pressure and an upper respiratory tract infection.

Resting Ventricular Pressure

7 mm Hg.

Stress Ventricular Pressure

14 mm Hg.

Result Normal level ventricular pressure

Action He was put to bed and an intravenous line was maintained running at maintenance rates. Mannitol was handy but was not used. He was given no other treatment.

Cardiac/Respiratory Artefact

CR = 3 mm at rest.

Points of Interest Viral titres obtained from the serum had shown a mumps titre of 32, adenovirus of 64, measles less than 16, herpes simplex less than 16 and respiratory syncitial virus less than 16. On this admission the serum specimen showed titres to mumps 16, adenovirus 16, measles 32, herpes simplex less than 16, parainfluenzae

less than 16, Coxiella burnetii less than 16.

His white cell count was 9,000 with a differential count of neutrophils 70%, lymphocytes 26%, monocytes 4% and ESR normal. Nose and throat swabs grew no organism and C.S.F. was microscopically clear and culturally sterile.

Urea and electrolytes at the time of this V.P.M., sodium 134 mmol/l, Potassium 5.8 mmol/l, Serum Co_2 20 mmol/l, Serum Urea 3.5 mmol/l; C.S.F. osmolality at the same time was 288 mosml/l. Serum osmolality was 279 mosml/l. Urine osmolalities over that first 48 hours were 1086 mosml/l.

He subsequently developed a classical morbilliform rash which necessitated transfer to an Infectious Diseases Unit.

Case Number	95
Name	G.H.
Age	6 years
Method	Rickham reservoir

Medical Diagnostic Background

As before.

Temperature	Normal
Zero	Upper cortical subarachnoid space.
Duration	8 hours
Indication	He had still marked neck stiffness and his valve was taking more than 2 minutes to refill. He showed horizontal nystagmus, a slightly red infected throat, extensor plantar responses, sluggish pupillary reactions and bradycardia. There was Cheyne-Stokes respirations.

Resting Ventricular Pressure

6 mm Hg.

Stress Ventricular Pressure

23 mm Hg.

Result — Normal level intracranial pressure.

Action — He was then put on Penicillin and over the next 48 hours became much more alert. His neck stiffness was less and he was discharged 4 days after admission.

Results of investigations now when he improved showed a decrease in his urine osmolality. No marked change in his electrolytes was noted and C.S.F. and serum osmolality were obtained on this occasion.

Pressure Recordings — Fig. 156 shows an example of the ventricular pressure tracings on this occasion towards the end of the 8 hour monitoring period. Slight fluctuations of pressure level occurred but the level was never very raised.

It is worth discussing at this stage the case of a girl (N.J.) not included in these pressure monitorings who developed obesity quite rapidly over a 6 month period from $2\frac{1}{2}$ to 3 years of age. She had been in contact with mumps and was thought to have suffered a mild mumps encephalitis. Over the subsequent years she had a number of emergency admissions with rapid acute on chronic weight grin, evidence of CO_2 retention with associated hypoventilation. This clinically fits some of the features of inappropriate ADH secretion with low plasma osmolalities, hyponatremia, and urinary osmolalities inappropriately high in comparison to the plasma. There was sometimes associated 'fits' and papilloedema. Certain of these episodes have definitely been triggered by infection e.g. measles, staph. pneumonia.

It was noteworthy that throughout most of her life, since the onset of obesity, her parents have noticed that she gains weight during the day and passes volumes of urine during the night.

There are similarities between this case and the child in Case Numbers

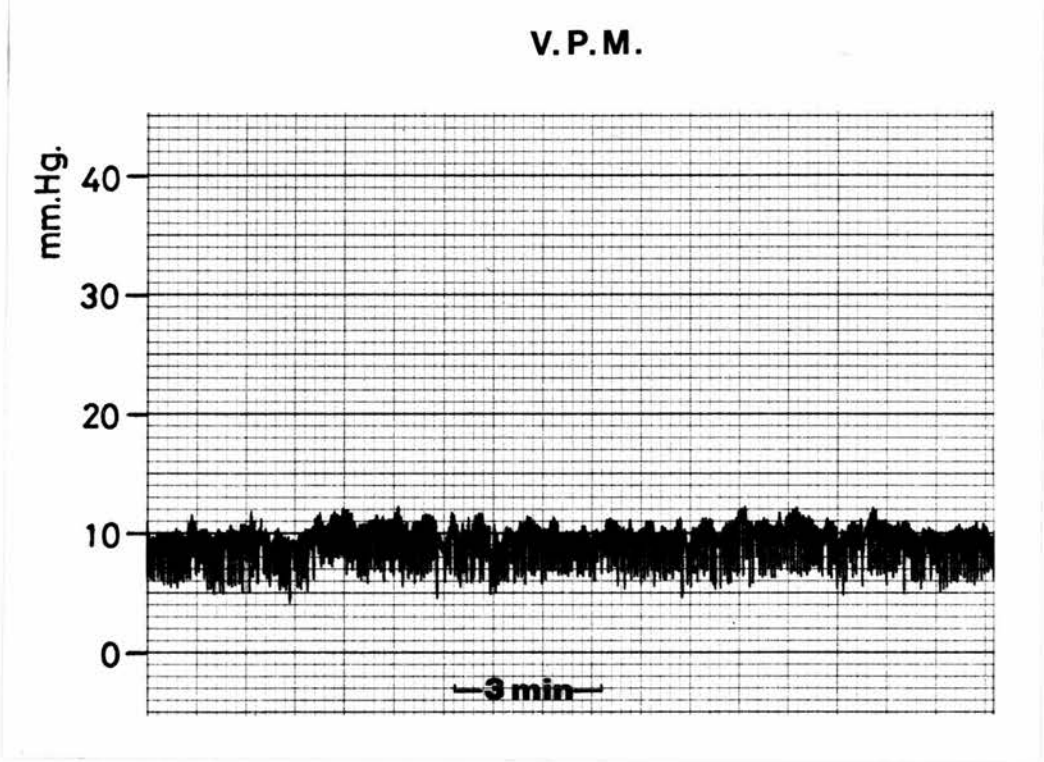


Fig.156

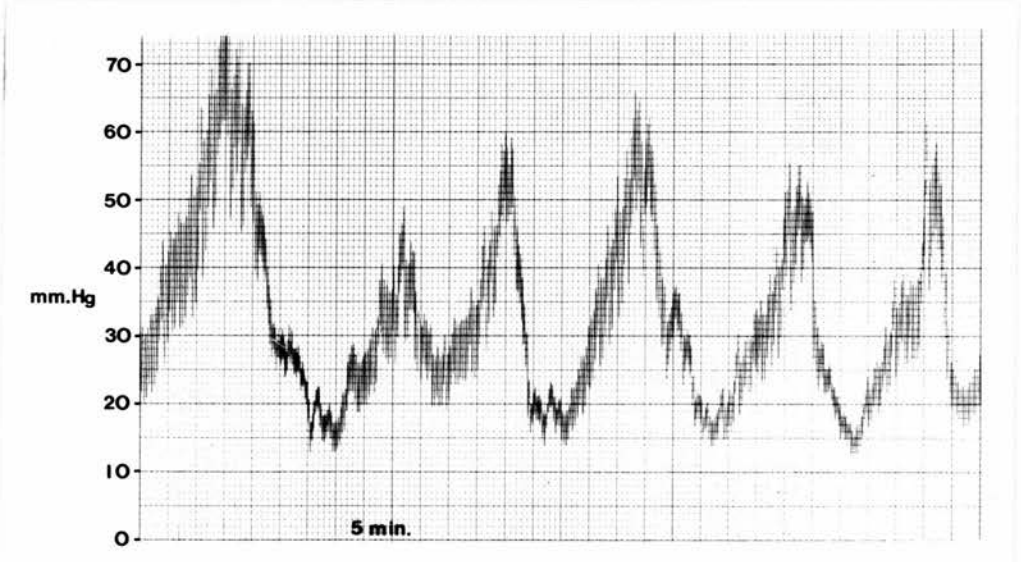


Fig.157

95, 96 and 97: one, a previous history of mumps from which the symptoms date: two, both have serum and urine osmolalities consistent with inappropriate ADH secretion (ADH levels have not been obtained): three, the attacks tend to be precipitated by an upper respiratory infection (neutrotropic viruses or bacteria) e.g. Coxachsie, measles, and hence the response to Penicillin in Case Number 97.

It is not unreasonable to suppose that since mumps affects most glandular structures within the body, resulting in parotitis, pancreatitis, oophoritis, orchitis etc., that the hypophysis could also be involved. I do not think this explanation tenable because

1. it does not explain selective involvement of the posterior pituitary (neurohypophysis) and in child G.H. no abnormalities have been found on TSH estimations etc.

2. it does not explain the chronic recurrent nature of the condition, one might have expected this, if it was related to direct invasion by mumps virus to run a course similar to other organ involvement.

To speculate, I think a possible explanation is that in cases of mumps complicated by say 'aseptic meningitis' a degree of adhesive meningitis or arachnoiditis may occur in the region of the third ventricle which then, at a later date, is affected either directly by neurotropic viruses or bacteria, or indirectly, similarly to the effect of cerebellitis following chicken-pox, resulting in involvement of the infundibulum and neurohypophysis and consequent inappropriate ADH secretion which causes water intoxication and in particular cerebral oedema with signs of raised intracranial pressure.

In case G.H. there is certainly evidence that arachnoiditis has occurred because following mumps, he converted from a communicating C.S.F. pathway to one of non-communication.

The roof of the third ventricle is thin ependyma and is closely applied to the under aspect of a double fold of pia mater and there is a

cavity in the floor of the third ventricle which projects into the infundibulum as a small recess. This in some animals is more than a recess and is a persistent space into the posterior lobe of the pituitary but in man merely points to derivation of the neurohypophysis from the floor of the third ventricle, i.e. there is extremely close proximity between the neurohypophysis and possible meningitis or ependymitis, furthermore inappropriate ADH is seen in neonates with meningitis in the acute situation.

A further child P.T. Case Number 96, has recurrent meningitis and it is significant (apart from his numerous other problems) that his parents report very significant polyuria, polydipsia, bulky stools etc. prior to his admission with raised intracranial pressure. This may be more related to diabetes insipidus occurring with brain (tentorial) shifts in a child who has had mucocent pressure problem from recurrent C.S.F. infection.

Case Number	96
Name	P.T.
Age	6 years 4 months
Method	Left frontal Rickham reservoir

Medical Diagnostic Background

This boy who has a Mondini defect (a congenital defect linking the inner ear with the C.S.F. spaces) suffers from recurrent meningitis and is deaf and mute. He also had discharge of C.S.F. fluid from the nose and was on long-term antibiotics. In the past he has had his middle ears explored and air injected into the middle ear clefts. Also microscopic examination of the middle ear looking for leaks and fluid, but nothing concrete has been found.

At 15 months of age he had meningitis associated with C.S.F.

rhinorrhoea. At 17 months he had insertion of a Rickham reservoir. At 18 months of age he had a bifrontal exploratory craniectomy and at 21 months of age he had a pneumonoccal meningitis. At 4 years of age he had further meningitis and was found to have a left sided hemiparesis and left sided torticollis. The findings on that occasion were thought to be due to recurrent meningitis.

A cavity has been seen at the time of positive contrast from a ventriculogram in the cervical cord at C3 and 4 level, i.e. a cervical syringomyelia and since 1978 the parents had noticed that the left arm had become more spastic and his balance had deteriorated, so that he now required support in walking.

Because of the findings in the ventriculogram it was decided to explore the posterior fossa and aspirate the cervical cord.

This was carried out and a cervical laminectomy performed. Some arachnoidal adhesions had formed in the region of the foramen magnum and were thought to be due to the previous recurrent meningitis. Both cerebellar tonsils were also seen below the foramen magnum. With a needle the cervical cord was punctured at C3 level and in spite of the consistency of the cord being normal, no fluid could be obtained and it was thought that the cavity had collapsed. The fourth ventricle was then opened and some muscle was plugged into the obex. Also a silicone tube was placed in the fourth ventricle to drain this into the subarachnoid space.

He was then admitted with convulsions which began in the face, spreading to the extremities, mainly in the left. C.S.F. was clear and of normal pressure. Plain skull films and echo encephalography showed no abnormality. There were further admissions with fits after this.

Seven months later he was admitted on Pheyntoin and Penicillin and the presenting complaint was that for the past month he had severe global headaches which wake him in the morning and then last until late in the afternoon. Most days he is affected, the headaches stopped suddenly. Strabismus was more marked on the right and he had been vomiting occasionally with the headaches. He was also noted to wet the bed when he had these headaches. On examination he was distressed complaining of a headache and, although the fundal veins were engorged, the discs were clearly outlined.

Temperature — Normal

Zero — Inter-ventricular foramina

Duration — 12 hours

Indication — As previously described, headaches associated with vomiting and polydipsia prior to the attack.

Resting Ventricular Pressure

16.5 mm Hg.

Stress Ventricular Pressure

80 mm Hg peaks in early sleep.

Result — Raised intracranial pressure

Action — At the time of monitoring there were gram-positive cocci (staph.albus) and 30 white cells in the C.S.F., necessitating intrathecal Cloxacillin and oral Flucloxacillin, i.e. there was a low grade meningitis at the time of monitoring. The action therefore consisted of antibiotics, plus tapping as necessary, though there were only small amounts of C.S.F. recovered from the Rickham on each occasion. Over the next 10 days he improved and was eventually discharged.

Cardiac/Respiratory Artefact

CR = 6 mm at rest.

CR = 50 mm in early sleep.

Ventricular Dilatation/Cortical Mantle

An L.A.E.G. was done which showed free communication to the fourth ventricle which showed the appearances of an obstructive arachnoiditis. The left lateral ventricle was moderately enlarged and contained a round filling defect in its middle part which appeared to be an arachnoidal cyst. The third ventricle was grossly enlarged and the aqueduct was not very clearly seen but was obviously patent and not grossly displaced. The fourth ventricle was fairly normal in appearance. At the time of this ventricular pressure monitoring, an E.E.G. was done, which showed alpha rhythms and no obvious focal or paroxysmal abnormality. He had a theco-peritoneal shunt in situ and a ventriculo-peritoneal shunt at the time of this V.P.M. A C.T. scan carried out 12 days prior to the V.P.M. demonstrated a normal ventricular size and no parenchymal abnormality.

Points of Interest It seems likely therefore that the infection resulting in a degree of cerebritis or brain swelling may well have been responsible for this boy's pressure waves Fig. 157 shows this child during early sleep with big swings of ventricular pressure extending off the recording paper. As the level of pressure reaches the peaks of these waves, he cries and awakens and each time he does so, the pressure then falls and this situation repeats itself and is a stumbling block to his progressing into more peaceful deep sleep. His mother reports that he has always been a very light sleeper. The section illustrated is but a small amount of the very protracted sleep changes which occurred in this child who had persisting C.S.F. infection. A further Fig. 158 is shown here after 7 mls of C.S.F. was aspirated from his Rickham, without the child stirring, resulting in the pressure dropping into the normal range allowing progression to deeper sleep patterns. During this recording, he had

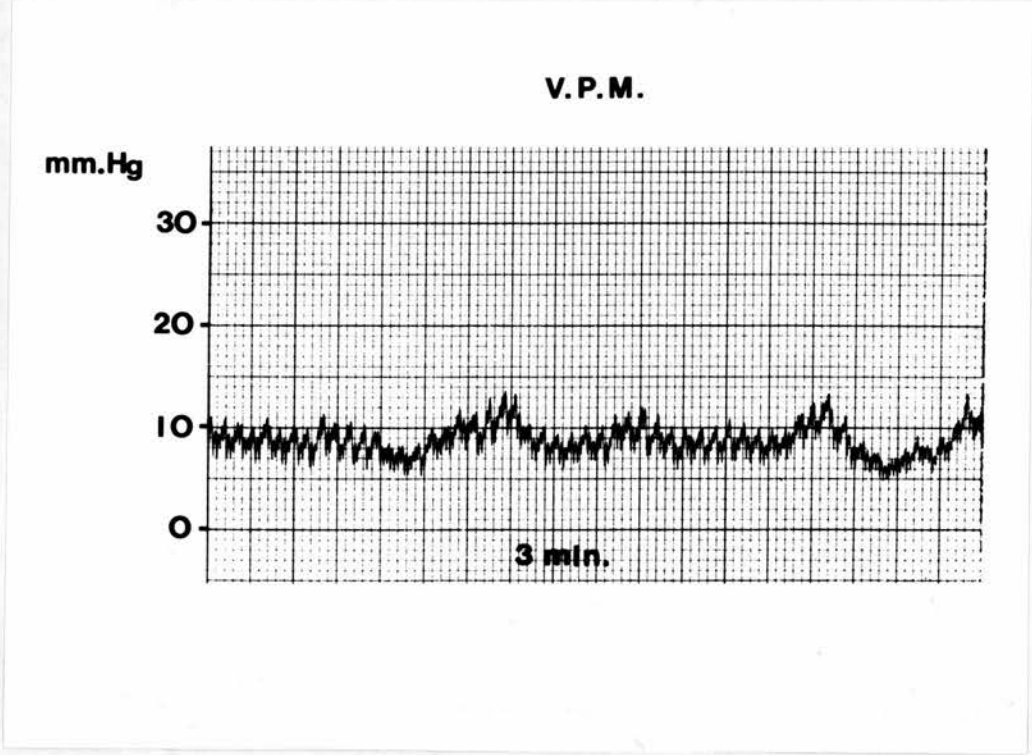


Fig.158

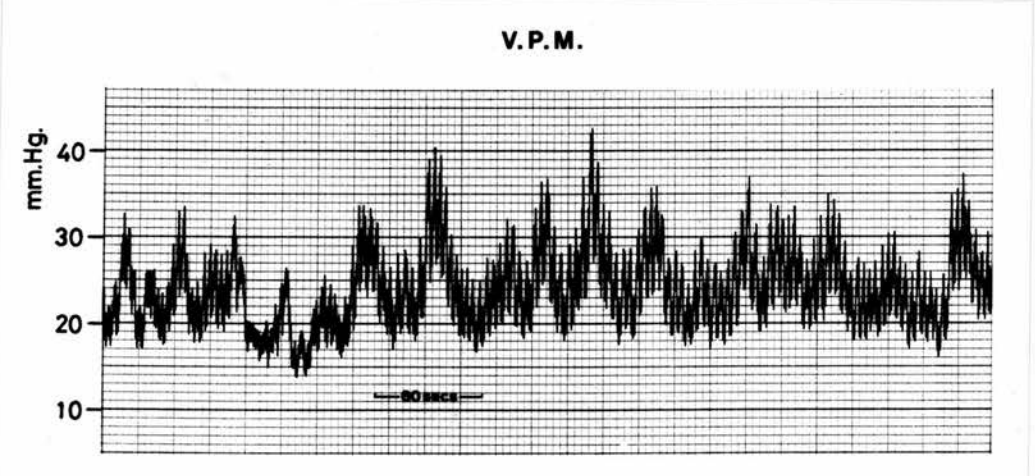


Fig.159

a major tonic clonic fit which is illustrated in Chapter 7, Vol. 1 of this book.

Case Number 97

Name P.T.

Age 6 years 7 months

Method Frontal Rickham reservoir

Medical Diagnostic Background

Mondini defect, recurrent meningitis, deafness etc. as before.

Temperature 36.5

Zero Upper cortical subarachnoid space.

Duration 21 hours

Indication He had been admitted 8 days before this admission with status epilepsy and discharged after 4 days. However at 6 a.m. on the day of this admission he started to complain of occipital headaches and abdominal pain and had been anorexic and vomited twice. The previous 3 days he had been girney and complained of a slightly sore head and tummy. He had very bulky stools and this, mother had mentioned prior to previous admissions, but no polyuria or polydipsia on this occasion.

Resting Ventricular Pressure

20 mm Hg.

Stress Ventricular Pressure

49 mm Hg maximum.

Result Raised intracranial pressure

Action C.S.F. was checked and found sterile and microscopically clear. He did have some neck stiffness and neck retraction and was Kernig's positive. His sodium was 132 and urea 7.4. After 5 cc of C.S.F. was released he still remained somewhat

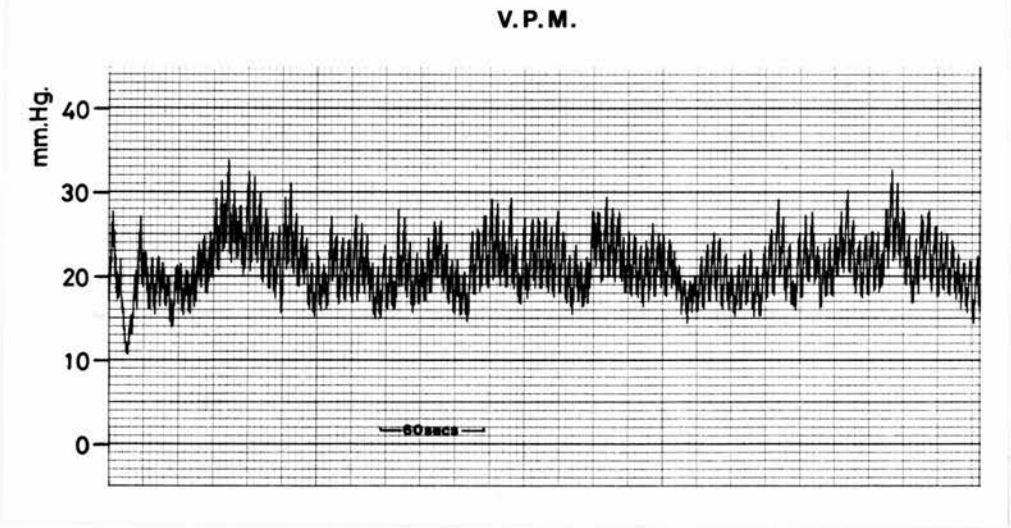


Fig.160

drowsy and irritable and Mannitol was kept close at hand.

Cardiac/Respiratory Artefact

CR = 23 mm at 25 mm Hg.

CR = 4 mm when ventricular pressure 3 mm Hg at the end of this recording.

Pressure Recordings Initially when the child went off to sleep on this occasion the pressure rose to peaks of 39 mm Hg. Removal of $2\frac{1}{2}$ mls of C.S.F. drops the pressure from 20 mm to 4 mm Hg instantly. Then there is a gradual rise to 25 mm Hg. Mannitol infusion (20%) is run in over an hour and the pressure dropped steadily to 15 mm Hg. By the time this V.P.M. was finished next morning he was well, had no headaches, no neck stiffness, was afebrile and happy. 75 mls of 20% Mannitol in all had been given the night before, which had brought his pressure down and it remained normal for the rest of the night. Fig.159 shows him on this occasion during a light phase of sleep. A similar Fig.160 is shown obtained while the child was quietly dozing during the afternoon depicting a mean level of ventricular pressure at 20 mm Hg.

Case Number	98
Name	P.T.
Age	7 years 2 months
Method	Frontal Rickham reservoir

Medical Diagnostic Background

Since previous recording he had decompressive craniotomies performed.

Temperature	Normal
Zero	$1\frac{1}{2}$" above upper cortical subarachnoid space.
Duration	2 hours

Indication On this occasion he had been vomiting on the way home from school for the last 3 weeks. He had lacked his usual energy and his gait had deteriorated during the last 10 days. A headache was reported a couple of days before admission lasting about 20 minutes. Frequency of micturition associated with a severe headache for 7 hours and some abdominal tenderness occurred 1 day before admission. Prior to all his previous episodes, he had polyuria, constipation, headaches and lethargy and it seems that this pattern had been repeated again on this occasion.

Examination remained much the same with his hemiplegic signs, and slight pallor of his optic discs. His shunt was a little 'sluggish'. A C.T. scan carried out was reported to be normal although the ventricles were thought to be smaller than usual. At the time of V.P.M. however, he was asymptomatic.

Resting Ventricular Pressure

3 mm Hg.

Stress Ventricular Pressure

25 mm Hg although reaching a maximum of 7.5 mm Hg during light sleep.

Result Normal level intracranial pressure.

Action Nil re C.S.F. It appeared that his decompression craniotomies, which had been performed 8 months previously, had been effective in minimising the results of brain swelling which he apparently has with these episodes. There had been bilateral fronto-parieto-temporal craniotomies to allow for transient intermittent rises in intracranial pressure and following this he was treated with reducing doses of Dexamethasone. At the same time as the decompression craniotomies he had a right sided Pudenz ventriculo-peritoneal shunt inserted, as it was felt that this would at least give him some small buffering during these episodes.

On one occasion when he was admitted to the Department of Neurosurgery at the time of bilateral decompressive craniotomies, post-operatively he was very drowsy and C.S.F. was aspirated via Rickham reservoir. He was placed on open ventricular drainage because of the high pressures and given Mannitol. Blood taken before the Mannitol showed a plasma osmolality of 259, sodium 125, potassium 3.5, chloride 100, CO_2 13.5 and urea 4. These electrolyte results would be compatible with a syndrome of excessive ADH as previously suggested. A further admission not a month after this one was because of a sore neck, irritability, crying, anorexia and abdominal discomfort and no C.S.F. infection. Electrolytes showed a sodium of 132, plasma osmolality 270. In view of this fluids were restricted. Two days after admission and fluid restriction, he was much brighter and almost his normal self.

CHAPTER 15

GROUP 'D' (patients investigated for miscellaneous reasons). CASE NUMBERS 99-100

Case Number 99

Name I.A.

Age 7 months

Method A fontanelle needle into the right subdural space.

Medical Diagnostic Background

This child was referred for pressure studies having been investigated with a C.T. scan when it was suspected that hydrocephalus was the cause for her 'failure to thrive'. The C.T. scan was normal but air encephalography showed a large subarachnoid space in the parietal region.

She was the third child of a 37 year old university lecturer and a 32 year old mother. Father is known to have pulmonary eosinophilia which causes 'asthma-like' attacks. There was no other significant family history.

She was noticed to be gaining little weight while still on breast milk from the age of 2 months onwards. Artificial milks did not make any difference to her weight gain and she vomited on and off throughout infancy. Investigations ruled out cystic fibrosis, coeliac disease, pyloric stenosis, renal abnormalities and although her height and weight were below the -2 standard deviations level, her OFC was between the 50th and 90th percentiles. Her fontanelle was pulsatile, scalp veins were sometimes distended and sometimes not. Her OFC was 38 cms but she was generally alert. The child had slight eczema, a 1 cm soft liver and at initial examination there was no 'sunsetting', no bruits and she could sit without support. She could reach, she had hand regard and tone and reflexes and peripheral nerves were all intact. She had normal blood films, chest x-rays, skull and bone age x-rays. Immunoglobulins, urinary investigations and stool

investigations were negative. Amino-acids osmolality, urea and electrolytes, calcium, phosphate, alkaline phosphatase, serum proteins etc. were all normal.

Pressure studies were carried out.

Temperature — Normal

Zero — Inter-ventricular foramina level.

Duration — 2 hours

Indication — Failure to thrive with suspected intermittent pressure symptoms.

Resting Ventricular Pressure

17 mm Hg.

Stress Ventricular Pressure

On crying reached a maximum peak of 60 mm Hg and a peak of 35 mm Hg in sleep.

Result — In view of the method of monitoring this pressure it was difficult to know how to interpret the results, but it was considered at the time not particularly elevated and no action was taken.

Cardiac/Respiratory Artefact

CR = 10 mm in the awake resting state.

OFC — 38 cms

Ventricular Dilatation/Cortical Mantle

Slight ventricular dilatation was noted.

Follow Up — It was considered in view of a high eosinophilia and rash in the presence of failure to thrive, that a milk allergy may have been responsible for at least part of her condition. Nutramogen resulted in a marked increase in weight. She then showed a catch up velocity in weight and has since progressed at an acceptable rate.

Pressure Recordings This tracing confirmed some recent work that the pressure in the cortical subarachnoid space is higher than that in the ventricles and is at variance with earlier reports.

Contrary to some reports in the literature, my clinical impression is that when the head is relatively large compared to the rest of the body, due to some systemic disease, signs of raised intracranial pressure such as scalp vein distension, may appear transiently.

Similarly, my clinical impression is that cases of megelencephaly do produce signs of raised intracranial pressure although they are often less severe and transient, e.g.a male child (D.N.) not included in this monitoring series who was born with a large head. Father and one or two other male members of the family tree had 'large heads' but the child's early neonatal period was uneventful and he was discharged at the usual time.

He was admitted for investigation at 16 weeks of age because of slow development, vomiting after feeds, irritability during the evenings and screaming attacks when nother pacified him. Examination revealed an OFC of 46.5 cm (i.e. + 4 standard deviations above the mean), irritability, hypotonia, marked head lag and a 'dysplasic body'. The anterior fontanelle was large and bulging, slight intermittent 'sunsetting' and his weight gain was unsatisfactory.

Investigations revealed that the lateral ventricles were virtually normal in size and the cerebral parenchyma was normal. Chromosomal analysis showed a translocation between the long arm of number 3 and the long arm of number 16 while parents had normal karyotypes. As well as the above he had an iron deficiency anaemia and a disaccharide intolerance with lactase deficiency on jejunal biopsy.

He appears therefore to have a familial megelencephaly possibly associated with his abnormal chromosome. However, there were undoubted

clinical features of raised intracranial pressure possibly related to the relatively large brain volume in relation to the small body size.

Case Number	100
Name	R.M.
Age	17 months
Method	Twin recording from Pudenz valve via

Huber needle and the cortical subarachnoid space via 19 gauge steel needle.

Medical Diagnostic Background

This child was admitted at the age of 5½ months with a history that he had been hit over the head by his 18 month old sister with a hairbrush. On examination at the time, there was a very large haematoma extending over the right side of the head. He was pale, had a heart beat of 140, blood pressure of 95/65 and an OFC of 44.8 cms with a soft anterior fontanelle. There were full eye movements, no retinal haemorrhages and no evidence of raised intracranial pressure. X-ray of the skull showed an extensive longitudinal fracture of the right side of the head with wide separation. There was also a healing fracture of the 6th left rib noted. An echo encephalogram was midline. Subdural taps were performed and 1 mm of bloody fluid was obtained from the left side. Not long after admission his haemoglobin fell dramatically despite a normal coagulation screen and he was noted to have a mild left facial weakness and a mild left hemiparesis. He fed slowly with occasional vomiting and was lethargic and irritable. An E.E.G. was rather featureless but showed some asymmetry.

After this episode he made excellent progress in a residential centre but at the age of 9 months his OFC was 44.6 cms and there was still a

large defect in the right parietal bone measuring approximately 9 x 2 cms but no bulging of the defect to suggest raised intracranial pressure. He had persistent, mild left hemiparetic signs. At the age of 15 months it was evident that he had had multiple non-accidental injuries and there was a large expanding fracture of the right parietal region. It was decided to perform air studies to assess the state of the underlying ventricle and obtain some impression of the C.S.F. pressure. This was to be followed by rib grafting and possibly an accompanying ventriculo-peritoneal shunt to diminish any expansile force beneath the graft.

An L.A.E.G. showed a marked asymmetry of the lateral ventricles, the right being considerably dilated, particularly in the posterior part, in association with the skull defect. The third ventricle was normal in size, shape and position. The sulci in the right parietal region were noted to be dilated and this was thought to indicate a significant degree of right sided atrophy in focal relationship to the previous injury. In the light of this information it was decided to insert a Rickham reservoir in the left frontal region so that pressure monitoring could be carried out to compare pressures on the right and left sides.

Temperature — Normal

Zero — Levels for the two recordings were not separated by a vertical distance of more than 2".

Duration — 2 hours

Indication — To decide (a) if there was any compartmentalisation and (b) if a shunt would be necessary to reduce the C.S.F. pressure beneath the expanding skull fracture.

Resting Ventricular Pressure

From the Rickham reservoir 15 mm Hg. From the posterior defect

8.5 mm Hg.

Stress Ventricular Pressure

From the Rickham reservoir 70-80 mm Hg peaks. From the posterior defect 60-70 mm Hg maximum peaks.

Result The pressure from the Rickham reservoir was in the equivocal range and that within the posterior defect at an acceptable level, but obviously a degree of asymmetry between the two sides.

Action He had a right posterior parietal craniectomy and 12 days later he had a theco-peritoneal anastomosis and rib cranioplasty. Since then a year later an acrylic cranioplasty was performed which has since needed re-fashioning once.

Cardiac/Respiratory Artefact

Firstly from the Rickham: R = 13.75, C = 5 mm.

CR = 10 mm asleep and CR = 5 mm awake.

Secondly from the posterior defect: R = 11.25 mm, C = 3 mm.

CR = 7.5 mm asleep and CR = 3.75 mm awake.

Ventricular Dilatation/Cortical Mantle

Marked asymmetry of the lateral ventricles, the right being considerably dilated, particularly in the posterior part in association with the defect. The third ventricle was normal in size, shape and position. Sulci in the right parietal region was noted to be dilated and this was thought to indicate a significant degree of right sided atrophy in focal relationship to the previous injury.

Points of Interest It is questionable whether inserting a C.S.F. shunt in this situation merely encourages further production of C.S.F. or whether inserting a shunt reduces the pressure long enough to allow healing to take place.

Pressure Recordings Fig. 161 shows the twin recordings from the Rickham reservoir (B) and simultaneously from the posterior defect (A). The vertical axis with degrees of pressure shown refer to the 'B' tracing. For the (A) tracing 0 mm Hg = 30 mm Hg shown and correspondingly 50 mm Hg = 80 mm Hg shown. One can see a slight damping of the tracing (A) compared to (B) with a more narrow cardio-respiratory oscillation.

Note that the tracings are 3.5 mm out of phase, this is artefactual and due to needle position, and is a necessary effect with this particular type of recorder. Also, that the reservoir was sampled of clear C.S.F. but the posterior defect side contained slightly blood stained C.S.F. Fig. 162 shows the simultaneous recording during light sleep.

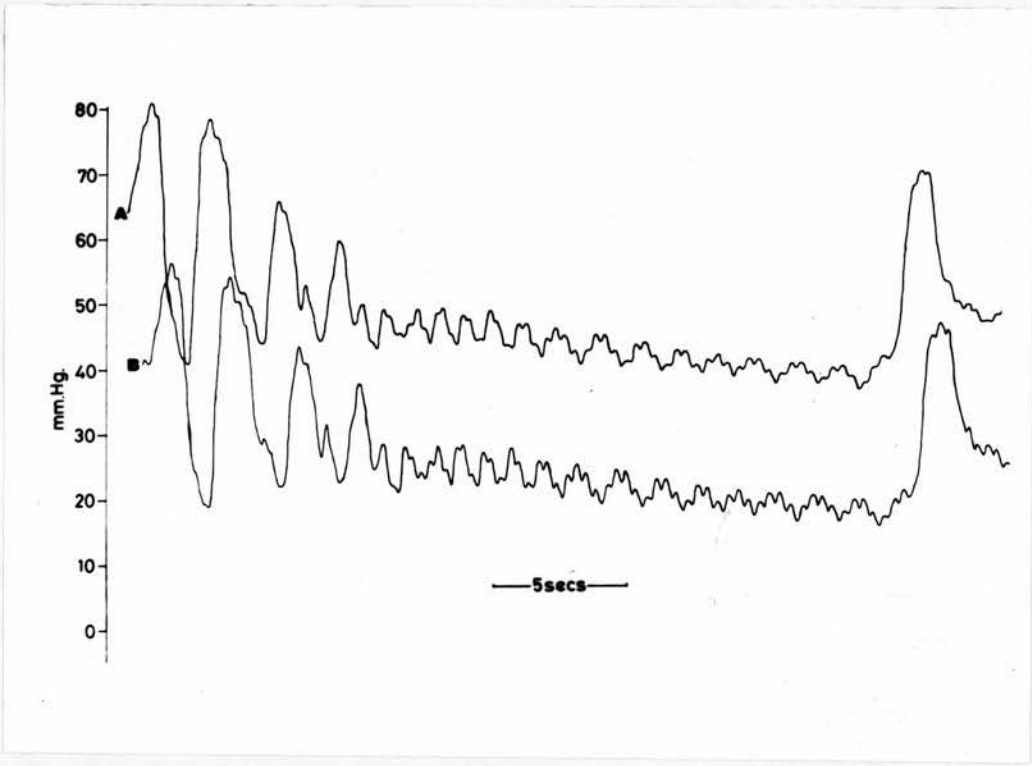


Fig.161

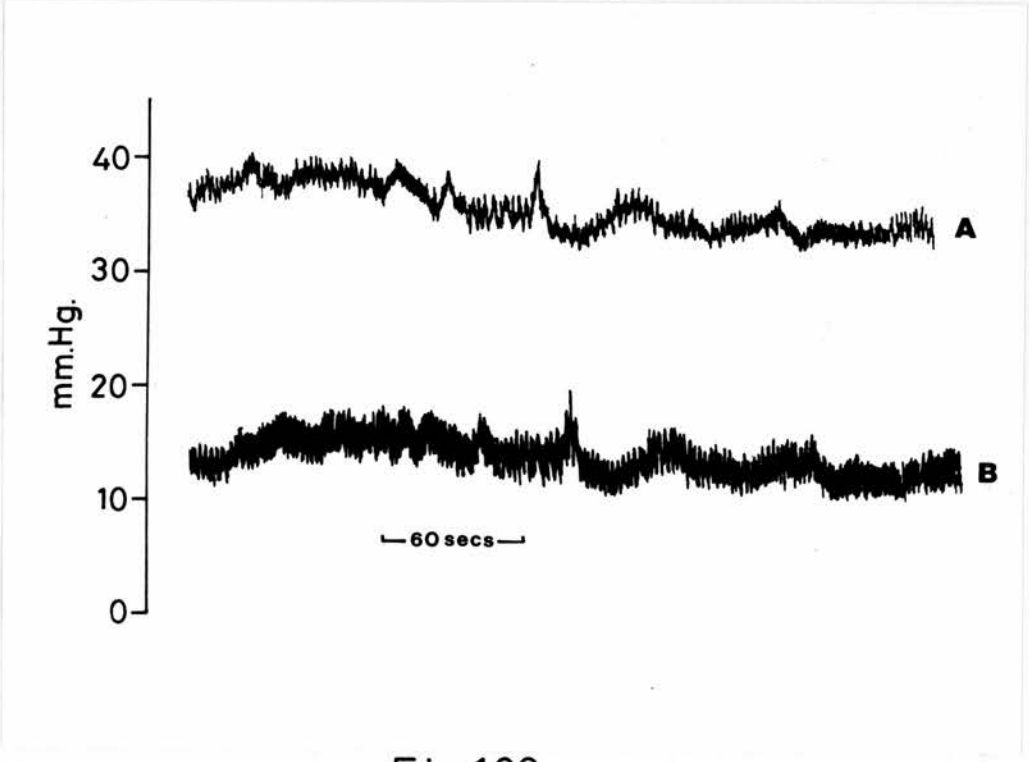


Fig.162