Primary Congenital Hydrocephalus

Long-term Results of Controlled Therapeutic Trial

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Holter's invention of the non-return valve and its use in ventriculo-caval drainage operations has been a revolutionary step in the treatment of hydro-cephalus. The valves effectively control intra-cranial pressure and prevent the back flow of blood from the vascular system. This technique has been in use in this country since January 1958. In our unit 48 such operations were carried out during 1958, using the Holter valve. Having acquired technical experience and confidence in the operation, we decided to assess its value and limitations, and to define the indications for and the timing of this procedure by fully controlled therapeutic trials. In this paper we describe one such trial, in which all the patients suffered from uncomplicated congenital hydrocephalus. The second trial covering the treatment of congenital hydrocephalus in infants who were born with meningomyelocele will be described in a separate publication.

Material and Methods

During 1959 and 1960 30 infants (19 boys and 11 girls) under 1 year of age were admitted with primary hydro-cephalus. The hydrocephalus, as far as could be ascertained, was due neither to intrauterine infections (e.g. toxoplasmosis) nor to postnatal cerebral trauma or infection. There was no associated spina bifida cystica, or other complex multiple congenital deformities. All were under 1 year of age on admission and 11 were under 2 months, but 6 were over 6 months of age (Table I).

After the diagnosis of hydrocephalus was established by pneumoencephalography or ventriculography, they were allocated by random selection to one of two groups. One group was treated by operation immediately after the diagnosis was made, and the other group acted as controls; they were not operated on at least until it became clear that conservative treatment had failed. There were 5 patients who were not allocated to either group, because they already had such advanced hydro-cephalus that immediate operative treatment appeared essential.

The degree of hydrocephalus was determined by ventriculography (Lorber, 1961). Among 12 patients allocated to the treated group, 2 infants had Grade 2 hydrocephalus, with a cerebral mantle measuring between 15 and 35 mm., and 10 had Grade 3 hydro-cephalus, with a cerebral mantle of less than 15 mm. Among the 13 controls there were 8 infants with Grade 2, and 5 with Grade 3 hydrocephalus. Thus the treated group was accidentally weighted with more severe or more advanced cases. This adverse weighting is probably related to the age on admission. The 'controls' were admitted at an earlier age (9 under 3 months against 4 in the treated group). Had they come a little later, their hydrocephalus would have been more advanced, as indeed was demonstrated by the precipitous rise in their head circumference after inclusion in the trial.

Finally, in the 5 not included in the controlled investigation the cerebral mantle was extremely thin.

Admission of new cases to the trial ceased at the end of 1960, because intermediate analysis of the results already indicated a trend which led to a change in policy.

All children were examined at regular intervals at 6, 12, 18, and 24 months of age, and then at each birthday. Developmental assessments were made at each visit by one of us (J.L.), and independent assessment was made by educational psychologists immediately before their fifth birthday, to assess them for scholastic purposes. Their assessment continued subsequently, and school

TABLE I
Primary Congenital Hydrocephalus, 1959/60

	Number	30
Age on Admission (mth.)		
<1		5
1		6
2–3		6
4–5		7
6–11		6
Grade of Hydrocephalus		
1		0
2		10
3		20
Sex		
Boys		19
Girls		11

reports were obtained about their progress. During 1966 and 1967 they had psychometric assessment by Mr. J. Parsons, research psychologist at the Unit.

Physical examination included a full neurological assessment, with special reference to the appearance of the optic fundi. Tests of visual acuity and of visual field were undertaken when this was indicated.

The Holter shunt was examined for its patency by palpation of the 'pump' behind the ear, and by radiography for the position of the lower end of the tube in the right auricle or in the main veins entering it. If the tube could be compressed easily with the finger and filled up rapidly when the finger was removed, it was considered to be working satisfactorily.

Results

The results presented are those that applied in October 1967. As a controlled therapeutic investigation, this study failed in one respect. As time went on, all but 2 of the 13 infants allocated to the control group had to be operated on because of rapidly increasing head circumference or other signs of neurological deterioration. In fact, it appears retrospectively that some of those who were operated on at a later stage should probably have been operated on sooner.

In the control group the interval between the allocation into the trial and the operation was between 4 weeks and 4 months in 7 infants; it was 12 months, 16 months, 18 months, and 3 years, respectively, in one each (Table II).

Two operated children had a shunt in position for so short a time that in some respects these could be considered as untreated cases.

The first was operated on at 20 months of age but developed septicaemia with colonization of the valve. The shunt was removed within 3 weeks and was not reinserted until he was 7 years of age.

The second was first seen and operated on at 11 months of age, when he was already severely retarded. He reacted so badly with high fever to the procedure that the shunt had to be removed 6 weeks later. A new shunt was inserted 6 years later, without complication, but he remains profoundly retarded.

There are only 2 control children who have not had a shunt to date. Observation and treatment of large groups of older hydrocephalic children suggest, moreover, that it is by no means certain that the hydrocephalus even in these 2 is truly arrested. It may well be that their intracranial pressure is still higher than normal. Though at present they are well, with average intelligence and with no serious neurological handicap, deterioration could set in at any time, requiring reinvestigation and possibly operative treatment. Such sequence of events has been commonly observed in hydrocephalic children of an older age-group (Lorber, to be published).

As there is no true control group left, the results obtained in the entire group of 30 children will be analysed together.

Mortality (Table III). Up to the end of October 1967, 5 children (17%) had died. Death was directly due to the hydrocephalus or its treatment in 3, probably due to it in 1, and almost certainly was unconnected with it in the fifth.

The youngest (Case 1) died at 8 months of age. The skin over the Holter valve broke down, the valve became exposed, and the baby developed meningitis due to *Staphylococcus aureus*. (The whole shunting system had to be removed and the baby died from the combined effects of meningitis and untreated hydrocephalus.)

The next (Case 2), according to age at death, was a boy who died at home at 2 years 10 months of age. He was not operated on until he was 12 months of

TABLE II
Reason for and Timing of Delayed Operative Treatment in Control Cases

Case No.	Grade	Age (mth.) on Allocation	Age (mth.) at Operation	Reason for Operation
6	2	2	3	Precipitous increase of head circumference
7	3	1	3¾	Precipitous increase of head circumference
8	2	5¾	9¼	Precipitous increase of head circumference
9	3	4½	5½	Precipitous increase of head circumference
10	2	1	3½	Precipitous increase of head circumference
11	3	3	5	Precipitous increase of head circumference
12	3	3¼	7	Precipitous increase of head circumference
2	3	2	12	Precipitous increase of head circumference and mental and physical deterioration
13	2	¼	18	Precipitous increase of head circumference and mental and physical deterioration
14	2	4	20	Progressive increase in head circumference, removed after 3 wk.; reinserted 5 yr. later
15	2	1	3¼ yr.	Very big head; hemiplegia; papilloedema; vomiting

TABLE III
Deaths

Case No.	Sex	Grade	Age at Death (yr.)	Cause
1	F	3	8/12	Staphylococcal meningitis resulting from exposed valve which had to be removed; necropsy
2	M	3	2 10/12	Not sure; at home; extreme hydrocephalus; severe retardation, blindness, spasticity
3	M	3	5	Blocked shunt; removal; meningitis; acute intracranial hypertension; necropsy
4	F	2	6	Bronchopneumonia; necropsy
5	M	3	7	Detachment and blockage of ventricular catheter; intraventricular haemorrhage; necropsy

age. By then his head circumference was 58 cm. and he was profoundly retarded and blind. The operation arrested the growth of his head, but did not help him otherwise. (It is not known whether his shunt became blocked before he died.)

The third child (Case 3), who was mentally normal with a moderate spastic paraplegia, died as a result of a blocked shunt at 5 years of age. The blockage could not be overcome by 3 operations in quick succession. He developed streptococcal meningitis. External drainage of CSF did not control his intracranial tension, and though his meningitis cleared up he died as a result of acute cerebellar coning.

The fourth child (Case 4) died at home at the age of 6 years. When last seen she was retarded but well, and her head was of normal size. She died of pneumonia. The necropsy report specified that the shunting system was in complete order.

The fifth and last child (Case 5) who died had extreme hydrocephalus which was first operated on at 8 weeks of age. His development had been retarded throughout the 7 years of his life. At 16 months of age blockage of the upper catheter was relieved by the insertion of a new one. At 7 years his ventricular catheter became detached and had slipped into the lateral cerebral ventricle. A new catheter was inserted, but much bleeding took place, both into the wound and the lateral ventricle, requiring several transfusions. He developed bilateral hemiplegia, and it was apparent that the new shunting system also became blocked. 5 further operations were performed within 12 days but to no avail. Necropsy showed that a large blood clot occupied his distended cerebral ventricles.

Survivors (Table IV). There are 25 survivors, who are now 7 to 9 years of age. All but one attend our clinic regularly; one child moved to the London area, but postal contact is maintained with him.

These 25 children are classified into 4 groups:

(a) 9 normal children, (b) 4 normal children but with large heads, (c) 8 children with normal intellect but with neurological lesions, and (d) 4 children with intellectual as well as neurological sequelae.

(a) *Normal children.* There are 9 children who are physically and mentally normal and whose head circumference is at or below the 90th centile. Their IQ is between 80 and 128. All have been operated on, but in 2, who were controls, the operation was delayed up to 30 and 37 weeks of age (Case 8 and Case 12, Table II). There are only 2 among these 9 who did not require further operations for their hydrocephalus.

The other 7 had to have further operations to deal with problems related to the shunt. In spite of his hydrocephalus, one boy (Case 16) (Fig. 1a and b) now 8 years of age, has an IQ of 128. His head circumference rose to 42 cm. by the time he was operated on at 5 weeks. Subsequently the rate of growth of his head slowed down and by 18 months of age his head circumference fell below the 90th centile. After two blockages due to thrombosis of

TABLE IV
Primary Congenital Hydrocephalus, 1959/60 Series: Assessment October 1967. Age of Survivors: 7–9 Years

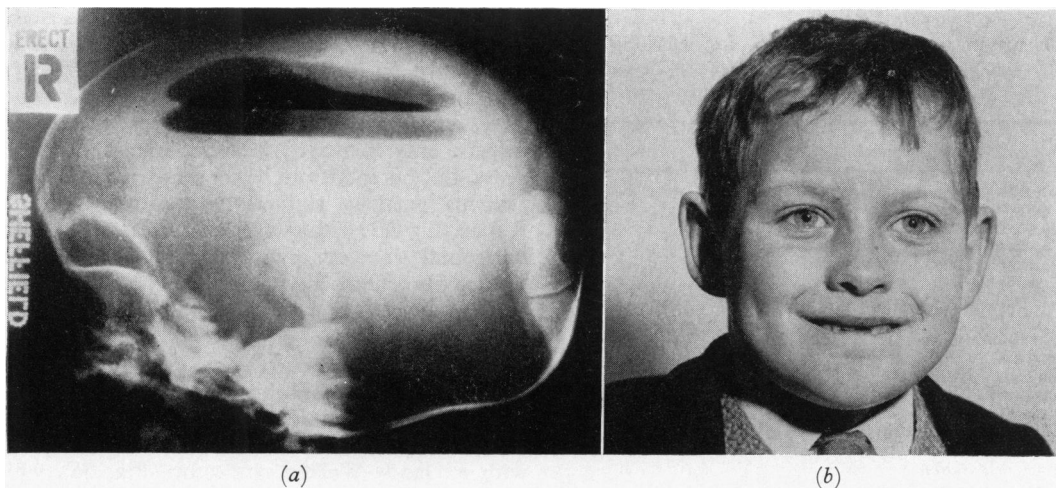| Neurological Condition	Mentally Normal	Educationally Subnormal	Ament
Normal head circumference 55 cm. or less	9	—	—
Normal head circumference 56–62 cm.	4	—	—
Varying handicaps (blindness, paraplegia, etc.)	8	3	1
Total	21	3	1

FIG. 1.—*Case 16. (a) Ventriculogram at 35 days of age, showing Grade 3 hydrocephalus. (b) Boy at 8 years of age. IQ 128.*

the jugular vein, the shunt had to be converted to a ventriculo-peritoneal shunt at 3¼ years.

Two children in this group were very advanced cases who were outside the controlled trial. One had a CSF pressure exceeding 400 mm. and the cortical depth on ventriculography was shown to be less than 10 mm., when he was 33 days old (Case 17) (Fig. 2a). A ventriculo-caval shunt was performed when he was 35 days old. This required no further attention. The boy is now 8, and is in excellent health (Fig. 2b). His IQ is 85. His head circumference

is 50 cm. The other is a 9-year-old girl now with an IQ of 109, and top of her class in several subjects.

(b) *4 normal children with large heads.* There are 4 other children with normal intelligence, who have no neurological sequelae, but whose head circumference is still outside the 90th centile. These include one child whose shunt had to be removed on account of bacterial colonization. He remained well for several years, but as a result of a fall in his IQ he was reinvestigated at 7 years of age. His
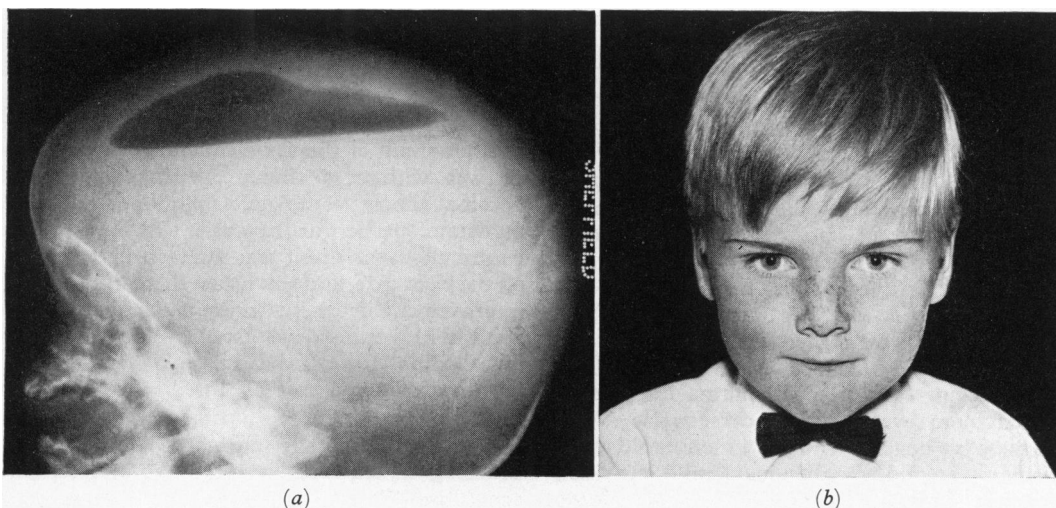

FIG. 2.—*Case 17. (a) Ventriculogram at 33 days of age, showing extreme hydrocephalus with cerebral mantle of less than 10 mm. (b) Boy at 8 years of age. IQ 85.*

TABLE V

Primary Congenital Hydrocephalus, 1959/60;
Neurological Sequelae in 12 Survivors (aged
7–9 years) at October 1967

Sequelae		No.	
Spasmus nutans	.. ..	1	
Visual defects	.. ..	8	
Optic atrophy only, good vision	.. .. ..		2
Optic atrophy, slight loss of vision	.. ..		1
Optic atrophy, severe loss of vision	.. ..		3
Optic atrophy, blind	.. ..		2
Deafness (unilateral)	.. ..	1	
Spasticity	.. ..	11	
One hand only	.. ..		1
Paraparesis	.. ..		6
Hemiplegia	.. ..		2
Quadriparesis	.. ..		1
Quadriplegia	.. ..		1
Convulsions (with hypsarrhythmia)	.. .. ..	1	
Craniosynostosis	.. ..	1	
Sexual precocity (girls)	.. ..	2	

CSF pressure exceeded 500 mm. H_2O. His cerebral mantle was only 7 mm. deep. A new ventriculo-caval shunt was inserted without complications. He is the only one in this series whose shunt could be removed, without disastrous sequelae, probably because it was only in place for a very short time. One child has not been operated on. His head circumference is 62 cm.—one of the biggest in the whole group.

(c) *8 children with normal intelligence but with neurological sequelae* (Table V). There are 8 further children whose intellectual development is within the normal range, but who have varying handicaps because of the neurological damage resulting from the hydrocephalus or the complication of its treatment. Their intelligence quotient is in the lower ranges of normal, between 80 and 100. Multiple handicaps (e.g. blindness with spasticity) make accurate estimation difficult and probably unnecessary.

There are 3 with slight disability consisting of spasmus nutans in one, a slightly spastic, clumsy hand in the second, and unilateral deafness plus slight spastic paraparesis in the third, not sufficient though to keep him from walking and going to ordinary school. A fourth child also has slight spastic weakness of her legs but she also has pale optic discs and impaired vision.

The remaining 4 children with normal intelligence have much more severe neurological sequelae. The first of these is a boy of 8 who was 6 months old when he was first seen and who then had a mild left-sided hemiparesis. A pneumo-ventriculogram disclosed a pressure of over 400 mm. CSF and there was only 10 mm. of cortex. Such thin cortex at 6 months of

age indicated far more severe hydrocephalus than such a figure would mean in early infancy, as was also shown by his head circumference of 52·5 cm. His fundi were very pale. After operation 5 days later, his progress was good, and his head circumference only increased 2·5 cm. over the next 7½ years, but his moderate hemiparesis persisted. He remains partially sighted, with visual acuity of R 6/60, L 6/36 (with glasses). He has considerable horizontal constant nystagmus and dead white discs. He attends a school for the partially sighted. When he was 4 he had some convulsions, but has had none since.

In the other 3, neurological signs developed or increased after the Holter valve was inserted for treatment of their hydrocephalus. A boy, now 7 years old (Case 6), had very severe hydrocephalus with a Dandy-Walker syndrome (Fig. 3a), which was demonstrated at 8 weeks of age, with a residual cortex of only 5 mm. His head circumference increased at an alarming rate to 51 cm. by the time he was operated on at 12 weeks. Since then it remained stationary. His sutures underwent premature fusion and he developed craniostenosis (Fig. 3b) with proptosis. At 2 years of age his optic discs were still normal and he had good vision, though there was progressive fall in his intellectual development to a low IQ of about 50. Coronal craniectomy was performed when he was 3¾ years. This did not control his headaches. His shunt must have been blocked and by 4¼ years he had papilloedema, with much proptosis. He required temporary tarsorraphy to save his eyesight. His shunt was found to be blocked with a ventricular pressure of over 400 mm. CSF. The Holter valve was removed and a new one was inserted on the left side. 8 months later this system became blocked both at the ventricular and the jugular ends and it had to be removed, without being able to establish another shunt at the same time. For over 2 years he was without a shunt. During this time he became almost completely blind and developed moderate spastic quadriparesis. At 7 years a new ventriculo-caval shunt was successfully performed on the right side, and since then there has been some improvement in his spasticity and much improvement in his mental development. He was accepted for a school for blind children, where the headmaster considered that his intelligence was within the normal range.

The third mentally normal child with severe neurological sequelae was first seen at 1 month of age (Case 15). She was in good condition, and though her head circumference was outside the normal from birth, initially it only grew at an
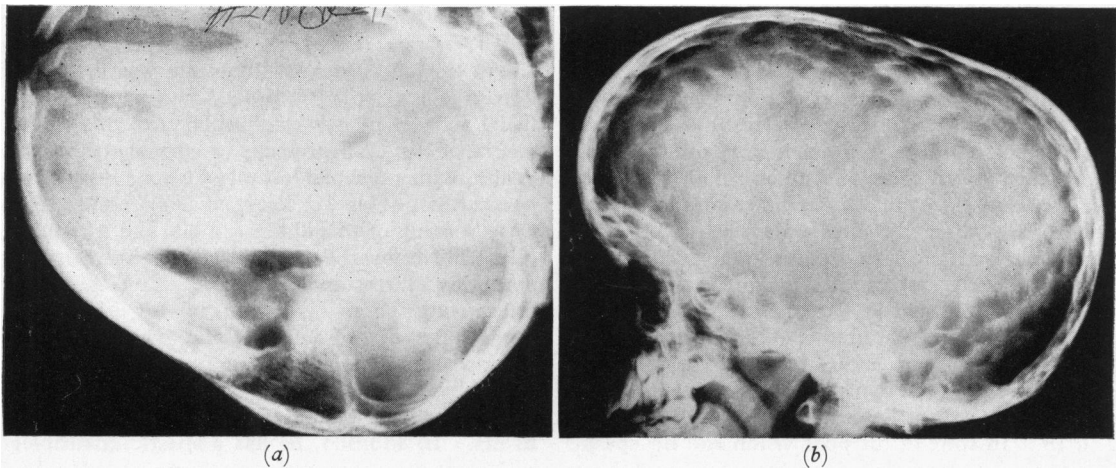
FIG. 3.—*Case 6.* (*a*) *Ventriculogram at 8 weeks of age. Brow-down position, showing elevation of the posterior horn of the lateral ventricles, due to very large fourth ventricle (not shown here). Dandy-Walker syndrome. Note operative result of coronal craniectomy.* (*b*) *Craniostenosis with premature fusion of sutures. Note large head with pronounced 'silver beating'.*

approximately normal rate and kept 2 cm. above the 90th centile. She had a Grade 2 hydrocephalus with only a moderate rise in CSF pressure to 200 mm. She was in the control group. From the fifth month onward her head started to grow rapidly and from 45 cm. at 18 weeks it grew to 55 cm. by 12 months. She remained well, with normal mental development, and the growth of her head slowed down again. It grew only 3·5 cm. in the next 2 years. She only just started to walk at 3 years of age, when she sustained a head injury, became ill with vomiting, and when seen at 3 years and 2 months of age, her head circumference was 64 cm. She had gross papilloedema with retinal exudates, could no longer sit, and had spastic quadriparesis. It was at this stage that a ventriculo-caval shunt was performed with much clinical improvement, but her spasticity increased, especially in the left side. The papilloedema was followed by pallor of both optic discs, but her vision remained good. In spite of all this, she attended an ordinary infants' school from 5 years of age onwards. She did not return to the clinic for 2 years. Meanwhile, secondary sexual characteristics began to appear at 7¼ years, together with an impressive pubertal spurt of growth. When next seen at 8 years of age she was well, but her shunt was obviously blocked. She still had a left-sided spastic hemiparesis and bilateral optic atrophy. Her height of 145 cm. was 23 cm. above the average. She attained full adult development and menarche occurred at 8 years 1 month.

Her CSF pressure was 370 mm. H₂O. Her blocked shunt was revised and the thrombosed

internal jugular vein was bypassed by putting the catheter into the external jugular vein and through into the right auricle.

The fourth and last child who is intellectually normal in spite of severe neurological handicap is a boy of 7½, whose Grade 3 hydrocephalus was operated on at 24 weeks. By then his head circumference was 52 cm. For the next 7 years his progress was uneventful, though there was some doubt about the efficient function of his shunt from 4 years onwards. His head circumference gradually increased to 62·5 cm. by 7½ years. He then developed measles from which he failed to make a quick recovery. He became drowsy, with headaches and vomiting, but was not returned to the clinic until some 7 weeks later. By then he had very little residual vision. He had bilateral secondary optic atrophy with tortuous retinal vessels. There was spastic weakness of both legs with bilateral ankle clonus.

The shunt was explored and was found to be completely blocked by a calcified thrombus in his internal jugular vein. It could not be removed, but was disconnected from the valve. A new tube was attached and put into the right auricle, through the external jugular vein. This operation was followed by sudden high fever and the development of a large abscess in his neck, from which β-haemolytic streptococci were grown. The abscess was drained and the ventriculo-cardiac shunt was disconnected by removal of the catheter from the external jugular vein and establishing external drainage via the ventricular catheter. The intra-

ventricular pressure was 380 mm. H$_2$O, under anaesthesia. A burr-hole was made on the left side of the skull, and through this soluble penicillin was injected daily into the cerebral ventricle to prevent ventriculitis which might have resulted from the abscess surrounding the ventricular shunt on the right side. With this and with antibiotic treatment ventriculitis was avoided, the infection cleared up, and 8 days after the first operation a left-sided ventriculo-peritoneal shunt was established. The whole shunting system on the right was removed. Meanwhile the spastic weakness of his legs had increased. After the first operation he had very little eyesight, but after the establishment of satisfactory drainage there was progressive improvement. Five months later his vision was R 6/36 and L 6/18. In spite of his poor vision and his spastic paraparesis he went back to his ordinary school. Recovery of vision after prolonged blindness has been described in a larger series of patients elsewhere (Lorber, 1967).

It is possible that these complications would have been avoided by attending to his shunt before he had any symptoms.

(d) *4 children with intellectual and neurological sequelae* (Table V). These 4 survivors are mentally and physically handicapped: 2 are educationally subnormal and 2 are even more severely retarded; 2 have a normal-sized head, the head circumference of the others is from 57 cm. to 60 cm. One of the moderately retarded children has a spastic paraparesis and bilateral optic atrophy, but his vision is good. The other, now 8, had a most severe degree of hydrocephalus, so that by 14 weeks her head circumference reached 57 cm. Her intraventricular pressure was over 400 mm. H$_2$O and her cerebral mantle was less than 5 mm. wide. A ventriculo-caval shunt at this time arrested the growth of her head, but her development was slow and her vision was very poor. She had searching nystagmus with bilateral optic atrophy. Two blockages of the shunt were corrected, but she developed moderate spastic weakness apart from her almost total blindness. Sexual precocity was another feature. Menarche occurred at 7 years and 5 months of age, and menstruation was regularly established by 8 years. At this age her head was still of the same size as when she was 14 weeks old.

The third child, a girl of 8, was normal until she was 7 years 10 months, when she developed intracranial hypertension as a result of jugular vein thrombosis. She was sent to a local hospital unfamiliar with her case. She had 5 operations in quick succession, and during this time her shunt system has been twice removed. This led to extreme illness and her life was in balance. Nevertheless, at the fifth procedure a new ventriculo-facial vein shunt was established, and this led to a return of her CSF pressure to normal and to good health, with a residual but rapidly improving spastic paraparesis. Her IQ dropped from above 100 to 62 as a result of this illness. This, and many other cases illustrate the grave dangers from rapidly mounting intracranial tension, if a previously functioning shunt is removed for any reason, beyond the age of infancy, and when the child's sutures are unable to separate further.

Finally, there is only one child who is profoundly retarded, who is unable to walk, and who is subject to fits. In addition, he has a spastic quadriplegia and has poor vision with optic atrophy. His EEG shows the pattern of hypsarrhythmia. This boy was 11 months old (the oldest in the series) when he first came to be investigated and treated. Though his hydrocephalus was not severe (Grade 2, pressure 220 mm. H$_2$O, head circumference 49 cm.) it was probably the association with hypsarrhythmia which caused his profound retardation. He had the typical history of developmental arrest at about 5 months of age, concurrent with the development of 'salaam' convulsions. He responded uniquely to the insertion of the ventriculo-caval shunt for he became extremely ill with high fever, which persisted over several weeks, without any ascertainable infective cause. CSF was normal, blood cultures were sterile, and there was no leucocytosis. There was no response to antibiotic treatment. Immediately on removal of the shunt 6 weeks after its insertion the fever ceased. No organisms were cultured from the components of the shunt. It was thought that possibly this was a hypersensitivity reaction to some component of the system. This boy remained without a shunt for just over 6 years. His head circumference rose to 60 cm. He was bedridden and kept in a long-term hospital. Even so, it was felt that a new shunt might save his sight and possibly benefit him in other ways, and a ventriculo-caval shunt was inserted without any febrile reaction, in February 1967. He improved considerably but it is too early to judge just how much benefit will accrue.

Head circumference related to results (Table IV). As might be expected, children who at the time of their last assessment still had an abnormally large head fared worse. Only 4 out of 12 children whose head circumference is 56 cm. or more are intellectually and neurologically normal as

compared with 9 of 13 whose head is smaller than that. Conversely, only 1 of 13 with a normal-sized head is retarded and has physical handicaps as well, as compared with 3 out of 12 whose head is unduly large. Nevertheless, there are no surviving children with extreme enlargement of the head. The child with the largest head circumference of 64 cm. is a tall, attractive girl who goes to an ordinary school in spite of her handicaps (Case 15). There are 6 children whose head circumference is already 60 cm. or more, and hence it will always be above the normal. None of the children who had a ventriculo-caval shunt procedure performed at the first occasion, and who were able to retain it, belong to this group of 6.

Revisions (Tables VI and VII)

(a) *Number of children who required revision.* One of the principal risks and disadvantages of all shunting procedures, including the ventriculo-caval shunt using the Holter valve, is the need for repeated operative procedures, colloquially known as 'revisions', to deal with the many complications that arise.

Of the 28 children who had a primary ventriculo-caval shunt, 23 have already required one or more operative procedures to deal with such complications.

Altogether, 52 further operations were performed in these 23, or an average of over 2 per patient in an average of nearly 6 years after insertion of the shunt: 4 children have already had 4–7 further operations, often in quick succession.

(b) *Reasons for revision.* The reasons for the revisions are given in Table VIII, together with the steps taken. Trouble with the catheter in the jugular vein alone was the reason for further operations in 13 instances, usually due to blockage by thrombosis, with or without calcification. The symptoms and signs of such blockages are irritability, headache, vomiting, drowsiness, diminishing eyesight, development of or increase in spasticity, and convulsions. Usually, however, these symptoms take several weeks or even months to develop.

Blockage of the jugular catheter may be diagnosed even before there are obvious symptoms, and effective action can be taken to prevent illness, neurological sequelae, and intellectual deterioration. The signs differ in infants whose fontanelle is still open from those seen in older children. In infants an increased fontanelle tension in conjunction with continued or newly recurring abnormally rapid rate of growth of the skull, together with a feeling of fullness of the compressible silastic part of the valve,

TABLE VI

Primary Congenital Hydrocephalus, 1959/60 Series; 'Revisions'

Number at risk	28
No revision required to date	5
Number with 1 revision	12 ⎫
Number with 2 revisions	3 ⎬ 23
Number with 3 revisions	4 ⎪
Number with 4 to 7 revisions	4 ⎭
Total number of procedures	52
Average number per patient	2·2

TABLE VII

Primary Congenital Hydrocephalus, 1959/60 Series; 'Revisions'

Reason for Operation	Action Taken	No.
Jugular catheter thrombosis	Ventriculo-peritoneal shunt	5
Jugular catheter thrombosis (and calcification)	New catheter in external jugular vein	6
Short jugular catheter	Lengthened	1
Jugular catheter detached	Reconnected	1
Peritoneal shunt blocked	Freed	5
Ventricular catheter blocked	Replaced by new catheter	7
Ventricular catheter disconnected	Replaced by new catheter	1
Exploration for doubtful blockage	Replaced by new shunt	3
Blockage both ends of shunt	Removal of whole shunt, not immediately replaced (major error)	4
Meningitis septicaemia, persistent fever	Removal of whole shunt	3
Infection, ventricular haemorrhage	External drainage	4
Craniosynostosis	Craniectomy	1
Temporary absence of shunt, intracranial tension	New V-C or V-P shunt	7
Miscellaneous		4

TABLE VIII

Congenital Hydrocephalus, 1959/60, Intelligence Quotient of 25 Survivors

IQ							No. of Cases
120–129	..	..	..	..	..	..	2
90–119	..	..	..	..	..	..	12
70–89 ..	..	..	..	..	..	..	7
50–69 ..	..	..	..	..	..	..	2
<50 ..	..	..	..	..	..	..	2
Total ..	..	..	..	..	..	..	25

are the most important signs. In properly functioning valves this tube is normally full of CSF, but not under tension, and on pressure it readily empties and quickly refills from above. If the jugular catheter is blocked, the tension in the tube is high, it feels stiff, and cannot be readily compressed. In older children the head cannot grow rapidly any more and the mounting intracranial tension usually shows itself by tortuosity of the retinal vessels, their distension, and eventually papilloedema. All these signs may be present before the child has any subjective complaint. *The ophthalmoscope is therefore the most essential diagnostic tool in the routine examination of these children.* Blockage or impending blockage of the jugular catheter may also be diagnosed or suspected, if on radiography of the chest the tip of the radio-opaque catheter is seen to have risen out of the cardiac atrium and is at the level of the 2nd rib or even higher in the neck. Elective revision may be carried out where the tube has risen out of the atrium into the superior vena cava even if there are no symptoms, but this was the reason for operation in only one child in this series. The probable need for revision on this evidence has been recognized only more recently. Many more such revisions might have been done otherwise, anticipating and preventing the trouble which followed later.

Blockage of the jugular catheter is confirmed on the operation table. On detaching the distal catheter from the lower valve, no blood wells up from the jugular vein and none may be aspirated with a syringe. If the intraventricular pressure is measured, it is shown to be raised. The patency of the catheter in the cerebral ventricle is shown by free flow of CSF from the upper catheter (if detached from the valve), or by dripping CSF from the lower end of the valve. This observation also proves the patency of the valves themselves.

Blockage of the jugular catheter may be overcome by removing it and replacing it by a new longer catheter, inserted into the internal jugular vein at a slightly lower level in some cases; into the external jugular vein or some minor vein in the neck in other cases. With experience, an alternative vein can often be found, but if not, the shunt may be converted into a ventriculo-peritoneal shunt, draining the CSF into the peritoneal cavity. This was carried out on 5 occasions in this group of patients. The peritoneal end of the shunt may also get blocked by fibrin, omentum, or the formation of an enclosed cyst round the peritoneal 'button'. This occurred 5 times in this series. The symptoms and signs are very similar, except that the compressible tube between the valves may not be so stiff, and compression is easier because of the extra length of the tube and the larger dead space in the system.

Blockage or other problems connected with the ventricular catheter occurred in 12 instances, in addition to 4 others, in which both ends of the shunt became blocked. Such a blockage is more dangerous than a jugular catheter block, because the onset of symptoms is more sudden, it leads to more rapid increase in intracranial tension, and may lead to coma, convulsions, and death in a matter of hours in some cases. The signs are similar to those found in jugular blocks, except that the symptoms and the papilloedema develop much more quickly. The feel of the tube is different. Here no fluid can enter it from above, so it feels flat and empty, or remains empty after compression. A soft, empty tube, however, is not necessarily due to blockage of the ventricular catheter: it may merely mean that the pressure gradient, at the time, is such that no fluid need escape from the cerebral ventricles. In such cases the child is well, has no signs of intracranial hypertension, and no action need be taken. If there is clinical evidence of obstruction, and exploration shows that both upper and lower tubes are patent, the fault must lie in the valve itself. This is rare and did not occur in this series though we have seen it in more recent cases (Corkery and Zachary, 1967).

Removal of the shunt in the belief that it is no longer necessary is to be avoided as it may lead to the same sudden rise in intracranial tension as blockage of the system. It may even lead to death, for it is very difficult to reinsert a shunt after the cerebral ventricles become of near normal size as a result of satisfactory treatment up to that time. Diminution of the size of the ventricles is indeed one of the main causes of blockage of the ventricular catheter, because it becomes embedded in cerebral substances. Another cause is obstruction by the choroid plexus which is less likely if the ventricular catheter is passed well forwards towards the anterior horn of the lateral ventricle.

Various infections connected with the shunt necessitated its removal or exteriorization in 6 instances. Meningitis, colonization of the valve, or exposure of the valve by skin breakdown were responsible for the need for revision in this group. This was often followed, at a later date, by reinsertion of a new shunt after the infection was overcome. These are usually grave events, which may lead to death or serious neurological deterioration in the patient.

An unusual, but interesting and well-recognized complication of the operation is the development of craniosynostosis, leading to proptosis, visual defect,

and increased intracranial tension, which occurred in one patient in this series and who required craniectomy for this reason (Case 6).

At least 3 and probably 4 of the 5 children who died lost their life as a result of complications with the shunt, and in at least 8 others these complications led to physical or intellectual deterioration.

Schooling

(A) *Ordinary infants' and primary schools (18 children)*. Twelve of the 13 children with normal intelligence and who have no neurological sequelae attend the ordinary infants' and primary schools. The thirteenth is unsettled and moves from place to place for social reasons. Five handicapped children also attend ordinary schools in spite of very considerable difficulties in at least 2 children. It is the policy in our Unit to encourage children to go to an ordinary school, whenever possible.

(B) *Special schools and training centres (7 children)*. Only 2 children attend schools for the physically handicapped. Another goes to a school for the partially sighted, and the fourth goes to a Blind School. One child has home tuition, because there is no school for the physically handicapped near his home and his mother will not let him go far from home. The sixth child in this group is in a training centre for the severely retarded, and the last is resident in a home for defective children.

Intelligence (Table VIII). The IQ was assessed using the Terman-Merrill scale (Form L or M), or other special tests, where applicable, to blind and other handicapped children. As is seen in Table VIII, there are 2 children with an IQ of 120 or over, 12 have IQs ranging from 90–119, and 7 between 70–89. It is also apparent, however, that there are more children among the lower IQ ranges than would be expected in the general population. The four children with the higher IQ (109, 111, 120, and 128) were all treated 'early'. None of the 'controls' achieved an IQ of more than 101.

Discussion

Many series, some quite large, have now been published detailing the results of treatment of hydrocephalus using ventriculo-caval shunt operations, but in most of these, cases of hydrocephalus of various aetiology were combined together, and there has been no published series in which all patients had primary uncomplicated hydrocephalus of congenital origin. Neither has there been any series published in which concurrent controls were included in the protocol of the investigations.

Finally, in most series the period of follow-up was short and there was a great scatter of the period of observation from the time the operation was performed.

This therapeutic trial shows that in an unselected group of infants with 'primary' congenital hydrocephalus the progression of the untreated disease was consistently and often rapidly unfavourable. Operative interference became necessary in the large majority of those who were initially allotted to the 'control' group. In many the operation had to be performed because of a precipitous rise in head circumference, but in others even more serious neurological symptoms and signs developed, such as spastic weakness or paralysis of hemiplegic or paraplegic distribution, or the development of eye defects, such as papilloedema or optic atrophy associated with visual defect. Severe intellectual deterioration was another feature in untreated children, or in those whose shunt became ineffective, or in whom it had to be removed. The so-called 'spontaneous arrest', at least as far as it concerns congenital hydrocephalus unassociated with spina bifida, must be rare indeed. It is obvious that there is a limit of head growth, but progressive neurological damage tends to occur, and most, though not all such patients, end up with serious intellectual, visual, and neurological handicap. A study of older children, who were born before the current techniques of treatment became available or widely used, also showed that most of those who were considered to be 'arrested' cases inexorably progressed and became severely handicapped. Similar observations were made by Schick and Matson (1961).

It is essential to point out that *these remarks do not apply to many of those infants whose relatively mild hydrocephalus is associated with spina bifida cystica*, and in whom the hydrocephalus is often detected as a result of systematic ventriculography (Lorber, 1961). About one-third of infants born with spina bifida and hydrocephalus do better without surgical treatment of their hydrocephalus, because they do not run the risks of the late complications of the shunting procedures. This problem will be the subject of a separate report.

It is clear, however, that the infants in this series whose hydrocephalus was not of an extreme degree when surgical treatment was first undertaken did better than those who were referred very late or in whom operation was delayed. Nevertheless, infants with even the most extreme degree of hydrocephalus, as long as it had not been untreated for many months, could be treated fully successfully. 2 among 5 who were extremely severe cases are mentally and physically normal at 8 and 9 years,

respectively. There are several other children treated in our Unit, who made similar satisfactory progress in spite of such extreme hydrocephalus that their head freely transilluminated before operation. There is a much younger child in our series, whose head had to be punctured during labour to allow delivery, and yet he is intellectually normal. Even the most severe degree of hydrocephalus is no contraindication to active treatment, though, of course, not all of such subjects will be normal individuals, and many will be severely retarded in spite of early treatment (Lorber, 1968).

The results obtained in this series were not optimal and it is easy to see, looking back on the case histories, that different action in several instances might well have led to better results. This, indeed, is one of the objects of a prospective study, which was carried out. Even so, the results here reported compare favourably with the very few other reports in which untreated cases of similar composition were studied: Yashon, Jane, and Sugar (1965) reported on 47 untreated cases of hydrocephalus. Half of these died at an average age of 3 years. Of the survivors, only 9 were 'well' and only 4 (8%) had IQs ranging from 82–101. Only 63 (35%) of Hagberg and Sjörgen's (1966) 180 untreated patients survived to 6 to 24 years of age. Of these, only 23 (13%) had no neurological sequelae, and 23 (13%) survived with normal intelligence. Though it is not possible to discern clearly the results in this type of hydrocephalus in Laurence and Coates' review (1962) in which all types of unoperated cases of hydrocephalus were analysed, it is again apparent that only about one-fifth survived with normal intelligence. These figures compare with an 83% prolonged survival rate in the current series, among whom 21 children or 70% of all cases, and 84% of the survivors have normal intelligence. In this series the range of intelligence quotient in children treated early was higher than in those whose surgical treatment was delayed. The 4 children with the highest IQ are in this group. There are no other comparable series of treated or untreated cases reported, in which cases of uncomplicated hydrocephalus can be separated from those associated with spina bifida or other complications.

Nevertheless, the high incidence of complications, the need for further operative procedures, and the high late mortality resulting from these are indications for careful study of the optimal timing of primary operation and of the value of 'elective' revision procedures at a time when the child has no symptoms of increased intracranial pressure or suggestion of a blocked system. It is not known whether the existing shunts will be satisfactory or necessary for these patients' continued good progress. It is not known how many will reach adult life in good health and able to compete with other individuals in normal circumstances. It is, however, evident, that though the technique of the current surgical treatment is the best that can be offered today, search for better methods, and if possible without the necessity of indwelling foreign substances, should be continued.

Summary

Thirty consecutive cases of primary congenital infantile hydrocephalus unassociated with spina bifida cystica are reported. All were admitted and studied during their first year of life (24 under 6 months of age) and all were followed up to 7–9 years of age. A controlled therapeutic trial was carried out, in which infants were allocated by random sampling to immediate operative treatment, using the technique of ventriculo-caval shunting and the Holter valve. All but 2 of the control cases had to be operated on later because of the rapid progression of their condition and the development of intellectual and neurological deterioration.

There were 5 deaths, at least 3 and probably 4 being due to complications of the operative procedures. In at least 8 others, complications due to the 'shunt' led to serious intellectual or neurological deterioration in those who did well before blockage or infection of the shunt.

Nevertheless, the high survival figure of 83% and the fact that 84% of the survivors are of normal intellect indicate the advance made in this field of treatment. Half of the survivors have normal-sized heads, and there are none with really gross cranial enlargement. Large heads in hydrocephalic children are now avoidable with early treatment.

There are 13 children with varying neurological handicaps of which visual defects of varying degrees and upper motor neurone lesions of varying distribution and severity are the most important. An interesting feature is sexual precocity in 2 girls. Convulsions are extremely rare.

Further advances in treatment are to be sought by earlier referral to specialized teams dealing with these conditions; by the more accurate timing of operative interference; by the elective revision of existing shunts before they cease functioning effectively and the early recognition of such defects. Among more distant objectives are techniques of surgical treatment in which the permanent insertion of foreign substances can be avoided.

Meanwhile, if adequately treated, most children with uncomplicated hydrocephalus should survive, at least for many years, with normal-sized head,

normal intellect, good vision, and little or no neurological handicap.

REFERENCES

Corkery, J. J., and Zachary, R. B. (1967). Increased resistance developing in Holter valves. *Lancet*, **2**, 1331.

Hagberg, B., and Sjörgen, I. (1966). The chronic brain syndrome of infantile hydrocephalus. *Amer. J. Dis. Child.*, **112**, 189.

Laurence, K. M., and Coates, S. (1962). The natural history of hydrocephalus. Detailed analysis of 182 unoperated cases. *Arch. Dis. Childh.*, **37**, 345.

Lorber, J. (1961). Systematic ventriculographic studies in infants born with meningomyelocele and encephalocele. The incidence and development of hydrocephalus. *ibid.*, **36**, 381.

—— (1967). Recovery of vision following prolonged blindness in children with hydrocephalus or following pyogenic meningitis. *Clin. Pediat. (Philad.)*, **6**, 699.

—— (1968). The results of early treatment of extreme hydrocephalus. *Develop. Med. Child. Neurol.*, Suppl. In the press.

Schick, R. W., and Matson, D. D. (1961). What is arrested hydrocephalus? *J. Pediat.*, **58**, 791.

Yashon, D., Jane, J. A., and Sugar, O. (1965). The course of severe untreated infantile hydrocephalus. Prognostic significance of the cerebral mantle. *J. Neurosurg.*, **23**, 509.