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Posthaemorrhagic hydrocephalus

Diagnosis, differential diagnosis, treatment, and long-term results

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Lorber, J., and Bhat, U. S. (1974). *Archives of Disease in Childhood*, **49**, 751. **Posthaemorrhagic hydrocephalus: diagnosis, differential diagnosis, treatment, and long-term results.** The clinical features, investigations, and results of treatment are described in a series of 47 infants with posthaemorrhagic hydrocephalus. 7 were unfit for treatment; 3 had medical (isosorbide) treatment alone, 2 of whom made a good recovery; the remaining 37 were surgically treated. 32 (68%) survived for 18 months to 16 years, but 12 of them are severely affected mentally and/or physically. The results can be considered satisfactory in 19 (40%), though there are only 9 (19%) without detectable sequelae. The most serious adverse prognostic features on admission were acute illness with active bleeding or neurological signs such as spasticity, fits, visual defects, or subdural effusions, and such infants did not recover without severe sequelae. The degree of hydrocephalus was also of prognostic value.

Since results could have been far better with better management, it is hoped that the publication of this series will lead to better care and prognosis.

Intracranial haemorrhage in the newborn is common, especially in the premature infant. For example, in a series of 12,640 consecutive births, 1% died of intracranial haemorrhage (Grönroft, 1954). Not all infants who suffer intracranial bleeding die. Some recover without sequelae while others survive with hydrocephalus or other neurological and intellectual deficits. Necropsy studies of those dying in the newborn period commonly show gross hydrocephalus, though clinically this is not always apparent (Russell, 1949; Amiel, 1964; Larroche, 1972). The hydrocephalus is caused by obstruction of the Sylvian aqueduct, the basal foramina, or the subarachnoid cisterns by blood clot or by aseptic meningitis (Russell, 1949; Larroche, 1972).

It is not known how many survivors develop clinical hydrocephalus, but 3% of 588 infants admitted with hydrocephalus to the Children's Hospital, Sheffield, between 1959 and 1963, were cases of posthaemorrhagic hydrocephalus; 17% of 110 infants with hydrocephalus not associated with spina bifida were posthaemorrhagic (Lorber and Bassi, 1965).

Intracranial haemorrhage may occur after birth as a result of accidental injury or child abuse or rarely as a result of a blood disease. If the haemorrhage occurs in infancy, hydrocephalus may develop concurrently or after subdural haematoma. Hydrocephalus developing after intracranial haemorrhage in older children is rare and none of our cases was older than 6 months.

Present investigation

This paper gives an account of 47 infants who were admitted between 1957 and 1972 under the care of one of us (J.L.)*. The relative rarity of the condition is illustrated by the fact that this represents an average of only 3 cases per year (range 0–7), though this Unit admitted approximately 150 new cases of infantile hydrocephalus of all types each year. The patients were referred from a large area serving a population of approximately 5 million.

Case material (Table I)

The case material consists of 47 children born between 1957 and June 1972. The survivors were observed for a period from 18 months to 16 years. 31 (66%) were boys, and 12 had a birthweight of less than 2500 g. This predominance of boys and low birthweight is usual.

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*Most were under joint care with Mr. R. B. Zachary and Mr. J. Lister who were in surgical charge of the patients.

TABLE I

Clinical features of intracranial haemorrhage related to results of treatment in 47 cases of posthaemorrhagic hydrocephalus

	Total		Alive							
			IQ 80–114			IQ 60–79 (no.)	IQ <60		Total	
	No.	%	Without physical handicap (no.)	With physical handicap (no.)	All (%)		No.	%	No.	%
Sex										
Male	31	66	6	6	38	1*	10*	32	23	74
Female	16	34	3	1	25	3	2	12	9	56
Birthweight										
<2500 g	12	25	3	1	33	2*	3	25	9	75
2500 g or more	35	75	6	6	34	2	9*	26	23	66
Onset of intracranial haemorrhage										
First dy	31	66	5	7	39	1	7*	23	20	65
2–28 dy	5	11	–	–		2	1		3	
1–6 mth	11	23	4	–	36	1*	4	36	9	82
Causes of intracranial injury										
Birth injury	28	60	5	5	36	1	6*	21	17	61
Blood dyscrasia	3		–	2		–	–		2	
Accidental	1		1	–		–	–		1	
Battering	4		–	–		1	3		4	
Unknown	11	23	3	–	27	2*	3	27	8	73

*One child with IQ 60–79 and one with IQ below 60 have no physical handicaps. All the others in these groups have neurological handicaps.

History of initial intracranial haemorrhage

(a) **Age of onset.** In 25 infants (52%) the diagnosis of intracranial haemorrhage was made on the first day of life and the history suggests that the symptoms began on the first day of life in 6 others. The onset of the intracranial haemorrhage was between the second and the 28th day in 5, between 1 and 3 months in 9, and over 3 months of age in 2 infants.

(b) **Causes and clinical features of the haemorrhage.** The main causes of the haemorrhage were birth injury in 28, blood dyscrasia (haemolytic and haemorrhagic disease of the newborn) in 3, accidental head injury in 1, and definite or suspected child abuse in 4. 3 of the 4 battered babies were among the oldest group. No reason for the haemorrhage was detected in 11 infants (Table I). When symptoms began in early infancy there was usually a history of difficult or obstructed labour, abnormal presentation, or traumatic forceps delivery. One infant was delivered head first into a lavatory pan, but in a sizeable minority the onset of symptoms was the first indication of intracranial haemorrhage.

The clinical features of the haemorrhage are shown in Table II. A fracture of the skull was found in one baby in whom child abuse was not suspected. The diagnosis was confirmed either by lumbar or ventricular puncture. All had heavily bloodstained CSF associated with xanthochromia.

Sources and age on admission

The infants were referred by 24 paediatricians and 2 general practitioners; they came from 27 different hospitals and in 2 cases from home. A few were admitted during the initial intracranial haemorrhage, but the majority were seen when there was established and often severe hydrocephalus. Table III gives their ages on admission. The 4 who were over 6 months old had all been seen earlier for treatment of a preceding subdural haematoma or other intracranial haemorrhages.

Presenting symptoms and signs of hydrocephalus

Every patient had an abnormally rapid rate of head growth. In 5 infants this was not recognized and they

TABLE II

Clinical manifestations of cerebral haemorrhage

	Alive	Dead	Total	
			No.	%
Convulsions	19	7	26	55
Irritability, tense fontanelle, meningism	11	8	19	40
Vomiting	5	6	11	23
Other*	7	8	15	32

*Cyanosis, apnoeic attacks, asphyxia, squint, facial palsy, spasticity, drowsiness, jaundice, melaena.

TABLE III

Age on admission with hydrocephalus

Age	Alive	Dead	Total	
			No.	%
Up to 28 dy	8	5	13	27
1 to 3 mth	7	8	15	32
3 to 6 mth	13	2	15	32
Later*	4	–	4	9

*3 of these had primary subdural haematoma.

were referred for miscellaneous symptoms. All 42 whose abnormally large heads were recognized before admission were sent as cases of hydrocephalus, though the haemorrhagic origin of this was not always recognized. 5 had been treated for presumed meningitis in the newborn period. There were 3 definite and 1 suspected cases of child abuse. 8 were admitted during a convulsion and 11 others were acutely ill; 11 of those 19 died. Altogether 12 had a recent history of convulsions.

All infants had an excessive head circumference and in 21 it was between 4 and 10 cm above the 90th centile. 32 children had one or more abnormal neurological signs (Table IV). Spastic paralysis of various types was noted in 12 patients. At least 14 failed to follow light or other objects, or had nystagmus or irregular eye movements. Pallor of the discs was not considered of pathological significance at this age. Among a variety of other signs were isolated cranial nerve palsies and cyanosis, and in several cases there was abnormal skull transillumination.

Diagnostic investigations

Infants admitted with a provisional diagnosis of hydrocephalus were investigated by *x*-ray of the skull (and of the rest of the skeleton, if deliberate injury was suspected); by subdural and ventricular taps, with direct monitoring of the resting CSF pressure and analysis of the ventricular and lumbar CSF (and of subdural fluid, if found); and by pneumoventriculography or encephalography. In some of the earlier cases dye studies were carried out to determine whether the hydrocephalus was communicating or not. In the last 2 years echoencephalography was used to determine ventricular size. More recently selective positive contrast ventriculography (Dimer—X) was introduced, particularly for the study of the third and fourth ventricles.

TABLE IV

Main neurological and ventriculographic features on admission with hydrocephalus related to prognosis

	Total		Alive							
			IQ 80–114			IQ 60–79‡ (no.)	IQ <60‡		All living	
	No.	%	Without physical handicap (no.)	With physical handicap (no.)	All %		No.	%	No.	%
*Neurological features**										
(1) Acutely ill with recent haemorrhage	8	17	–	–	0	1§	4	50	5	62
(2) Subdural effusion	8	17	–	–	0	–	6	75	6	75
(3) Visual defects, nystagmus	14	30	–	4	29	–	8	57	12	86
(4) Spasticity	12	25	–	–	0	–	7	58	7	58
(5) Fits	12	25	–	1	8	–	3	25	4	33
(6) Others†	8	17	–	3		–	1		4	50
(7) Any one or more of 1–6	32	68	–	6	19	1§	12	38	19	59
(8) None, other than hydrocephalus	15	32	9	1	67	3	—		13	87
Cerebral mantle on AVG										
Less than 5 mm	7	15	–	1		–	1		2	29
5–15 mm	28	60	5	4	32	3	7		19	68
Over 15 mm	12	25	4	2	50	1	4		11	92
Porencephalic cerebral cysts	11	23	1	4	45	–	4		9	82
All patients	47	100	9	7	34	4	12	26	32	68

*The figures do not add up to 47 because many infants had more than one of these signs.
†Cranial nerve palsies, cyanosis, transilluminating skull.
‡One in each group has no physical sequelae.
§Has very severe physical sequelae.

Diagnosis

Intracranial haemorrhage was considered to be the cause of the hydrocephalus if the head size was normal at birth, and there was clear-cut clinical evidence in the newborn period of major intracranial haemorrhage confirmed by the finding of heavily bloodstained lumbar or ventricular CSF. The diagnosis was made in the older infants if the head was of normal size before an injury occurred.

Differential diagnosis

(a) **Meningitis.** Difficulty was sometimes experienced if before admission to us an infant was suspected of having had meningitis and was treated for it. Such infants presented with symptoms consistent either with meningitis or intracranial haemorrhage. Their lumbar CSF was bloodstained and sterile. Sometimes there was a relative excess of white cells in relation to red cells, but this cannot be accepted as evidence of meningitis because subarachnoid haemorrhage causes temporary pleocytosis. Necropsy finding of bloodstained ventricular ependyma or other evidence of haemorrhage confirmed the diagnosis.

(b) **Subdural haematoma.** In 3 infants an erroneous diagnosis of subdural haematoma was made before admission because bloodstained or xanthochromic CSF was found near to the surface when 'subdural' taps were attempted. Often prolonged daily taps were carried out under the impression that the subdural spaces were being tapped. Fluid will be found very near to the surface if the ventricular system is grossly dilated, or if as a result of earlier taps porencephalic cysts have developed (Lorber and Grainger, 1963). The correct diagnosis is established by needling, then injection of air, followed by *x*-rays. If fluid is found near the inner table of the skull a sample should be obtained. Then the needle should be slowly advanced further. If the needle is in the subdural space, then the flow of fluid will cease when the needle enters brain substance: by advancing the needle further the ventricle will be reached. The CSF should be separately collected and analysed. The fluids obtained are likely to be very different: subdural fluid will contain gross excess of protein, from several hundred mg to over 2 g/100 ml, while the ventricular fluid may be normal or nearly so. When the needle is in a grossly dilated ventricle near the surface, advancing it to any depth will result in a continuous dripping of fluid of identical composition. Air should be injected into the space found near the surface on one side only. If one is dealing with a subdural effusion, the air will spread over the hemisphere and will usually be confined to one side If one is dealing with gross hydrocephalus or porencephalic cysts, the air will outline both ventricles as well as the cyst(s) (Fig. 1a, b). If the superficial fluid is in the subdural space, then after having taken *x*-ray views, air can be injected into the ventricle as well, and so the entire anatomical situation can be readily shown (Fig. 2a, b).

Subdural effusion can be present with or without associated hydrocephalus. Several cases initially

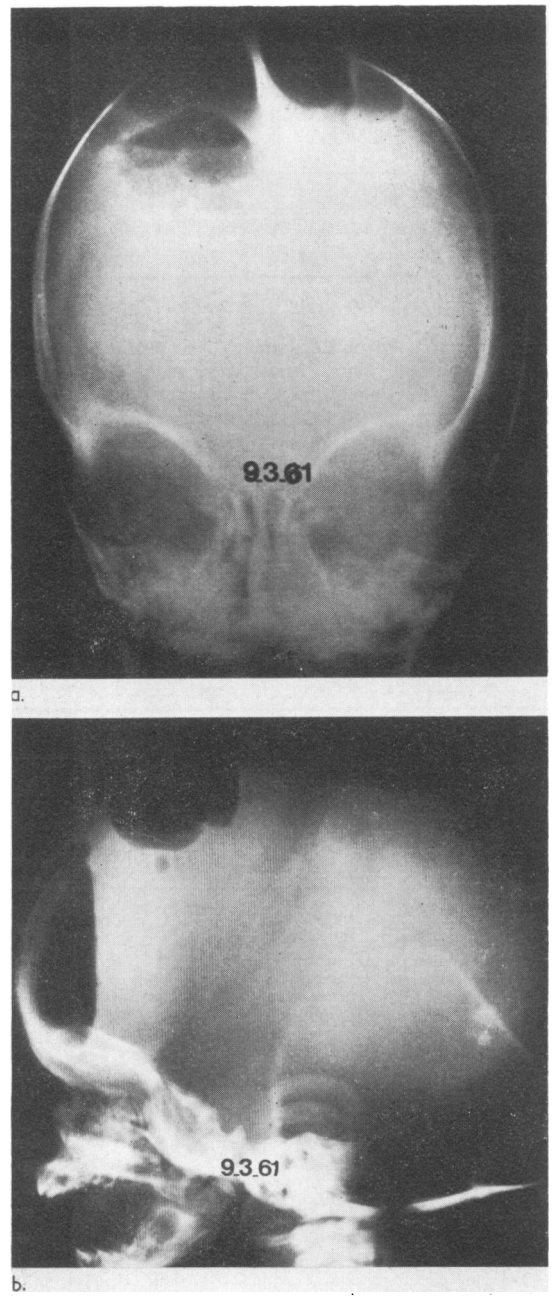

FIG. 1a and b.—*Air ventriculogram of posthaemorrhagic hydrocephalus with bilateral porencephalic cysts resulting from ventricular punctures through the anterior fontanelle. Xanthochromic fluid found near the surface could easily be attributed to subdural effusion. Note strict localization of the cyst in the brow-up lateral view (Fig. 1b).*

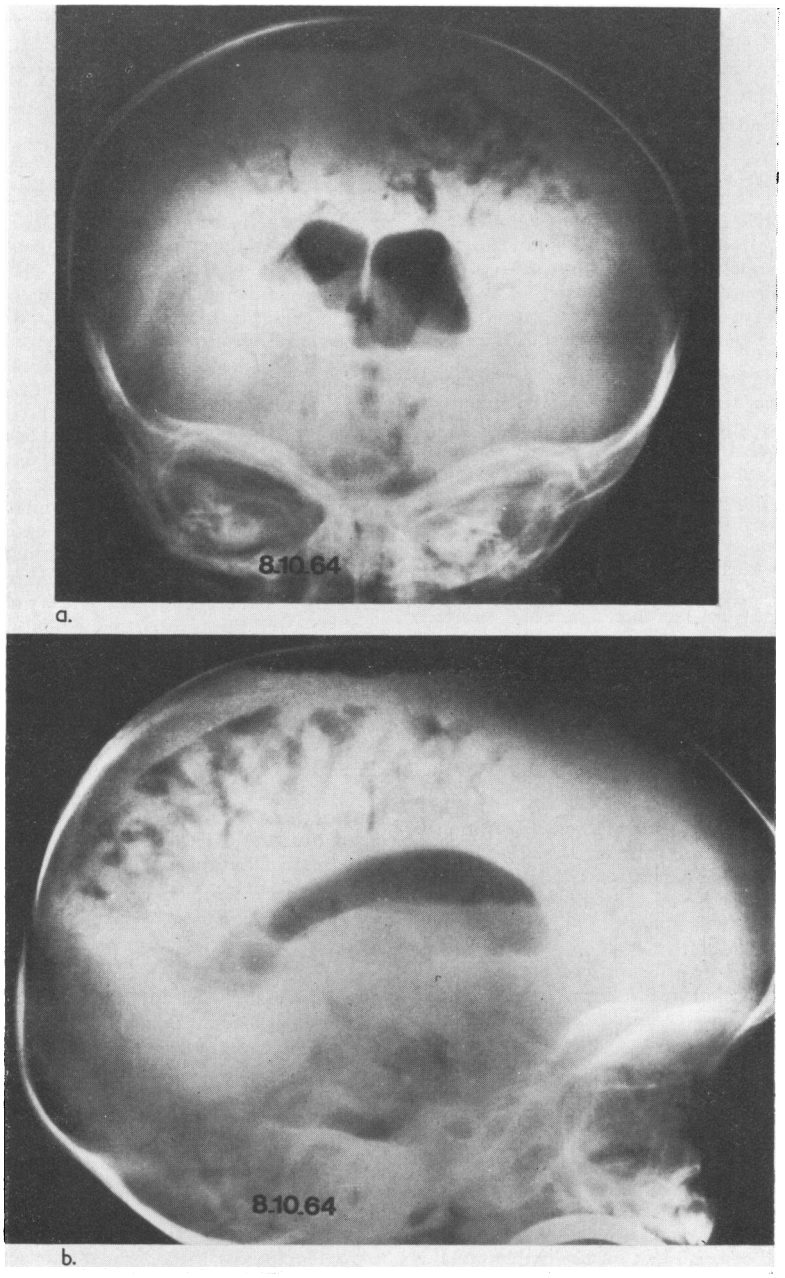

FIG. 2a and b.—*Lumbar air encephalogram plus air injection into the right subdural space in a child with subdural haematoma without hydrocephalus, at this stage. Erect view. Note that the ventricles are of normal shape and size. The subarachnoid air does not reach up to the inner table of the skull on either side. That space is filled with subdural fluid. Air injected into this on the right does not communicate with the left side, as the falx is a barrier between the two sides.*

diagnosed as hydrocephalus were proved to have a gross subdural haematoma without ventricular dilatation and are not included in this series. In other cases subdural effusions were associated with hydrocephalus either on admission or later and so are included in our case material.

(c) **Congenital hydrocephalus.** Congenital hydrocephalus of prenatal origin can be mistaken for post-haemorrhagic hydrocephalus. Delivery of an infant with an abnormally large head may easily result in intracranial haemorrhage; and this may aggravate an already existing hydrocephalus. Only one patient is included in this series whose head circumference at birth, related to weight, was over the 90th centile. This infant had an undilated ventricular system on his first ventriculogram.

Degree of hydrocephalus. The residual thickness of the cerebral mantle was measured in the pneumoventriculogram from the inner table of the skull to the roof of the lateral ventricle, at the site of the anterior fontanelle in the lateral view, and from the apex of the air bubble in the anteroposterior view. The average measurement of these AP and lateral views was taken. In most instances the cerebral mantle at the occiput was much thinner. In most cases over 75% of the supratentorial cranial contents consisted of CSF.

The cerebral mantle was less than 5 mm in 7, between 5 and 15 mm in 28, and between 16 and 25 mm in 11. In one it was 26 mm. This was an older infant in whom this measurement still represented gross ventricular dilatation. Porencephalic cavities were noted in 11 infants, but their incidence was undoubtedly higher because such cavities are often not visible when the cerebral mantle is very thin (Lorber and Grainger, 1963) (Table IV).

The hydrocephalus was shown to be communicating in 21 cases and noncommunicating in 21 cases; in the remaining 5 tests were not carried out to determine this point. The ventricular CSF was still frankly bloody with fresh or old blood in 26 infants. The CSF pressure was excessive in most infants.

Subdural haematoma or effusion was present as the main finding on admission in 8 infants. 3 had cranial injury at birth; the others acquired their lesions later, including 2 (or possibly 3) cases of child abuse.

Treatment

Before treatment for the hydrocephalus 5 infants received treatment for unproven meningitis: 8 were treated for proven subdural effusions with repeated subdural taps followed by a subdurocardiac shunt in 4 children. These shunts were removed later. 3 infants received treatment for other conditions.

The main method of treatment for the hydrocephalus consisted of a ventriculocardiac shunt, using the Holter type of valve, whenever this was indicated. Altogether 37 children were so treated, including 3 who had isosorbide therapy before operation (Lorber, 1972, 1973). 3 others were treated with isosorbide alone and 7 received no therapy (Table V).

TABLE V
Treatment for hydrocephalus

	Alive	Dead	Total
None	3	4	7
Isosorbide alone	2	1	3
Isosorbide followed by shunt	3	—	3
Shunt alone	24	10	34

Seven untreated patients. All 7 untreated infants were profoundly affected; 2 died early in infancy with active intracranial haemorrhage. Another died of his hydrocephalus at 4 years of age after a vegetative existence. The fourth had a subdural haematoma which after repeated taps was treated with a subdurocardiac shunt. This became infected and was removed and he died 2 months later after convulsions.

Three living untreated children are 10 and 11 years of age. One had massive intracranial haemorrhage leading to multiple cerebral cavities but only with moderate low-pressure hydrocephalus. He is microcephalic, profoundly retarded with spastic quadriplegia, fits, and blindness. The second had an extensive right-sided subdural effusion which communicated with his ventricles. After repeated aspirations his head stopped growing at an excessive rate. He is profoundly retarded, has left-sided hemiplegia, and is blind and subject to fits. The last child was a 'battered baby' who was repeatedly admitted for treatment of a subdural haematoma. Initially taps were successful, but he relapsed and a subdurocardiac shunt was inserted. Later this became blocked and was removed, followed by external drainage. This led to infection of the subdural space. He recovered on antibiotic therapy. His head is not enlarged but he is profoundly retarded, hemiplegic, and is in institutional care. None of these survivors had 'progressive' hydrocephalus and their gross sequelae are due to massive damage to their brain from the intracranial haemorrhage.

Three children treated with isosorbide alone. 6 infants were treated with isosorbide (Lorber, 1972, 1973). One, an acutely ill infant, died quickly with active intracranial haemorrhage. 2 infants responded well. One of them, a premature infant had severe melaena and intracranial haemorrhage. Hb fell to 7 g/100 ml by 72 hours of age. Her head circumference increased precipitously by 55 mm in 3 weeks (normal 18 mm) and her cerebral mantle was only 13 mm thick. Her head freely transilluminated. During isosorbide therapy her head stopped growing and afterwards grew at a normal rate. At first she was spastic, retarded, and did not appreciate light. Later gradual recovery of vision took place together with accelerated intellectual development. At 2½ years of age she had left-sided spastic hemiparesis, but was able to walk with one hand held. She could see and readily pick up tiny objects with her right hand but she still held her head tilted to one side. Her head circumference of 50 cm was normal. Her vocabulary was ahead of her chronological age.

The third child treated with isosorbide was asphyxiated and had fits soon after birth, the CSF being heavily bloodstained. Head circumference increased by 21 mm in 16 days (normal 12 mm). Cerebral mantle was 22 mm. During 28 days of isosorbide therapy head circumference increased by only 7 mm (expected 17 mm). After stopping therapy his head grew a little faster than normal, but his fontanelle remained soft. At 18 months he is a normal child apart from a squint, and has a normal head circumference.

The other 3 children treated with isosorbide did not respond well and as soon as they became fit for surgery were operated on. All were severely affected; one was treated before admission by repeated taps for presumed subdural effusions, but he had extreme hydrocephalus and not subdural fluid. The other 2 were battered babies who did badly. All 3 are alive.

Thirty-seven patients treated with ventriculocardiac shunt.

First operations. 37 patients (79%) were surgically treated. A ventriculocardiac shunt of the Holter type was used in nearly every case. In 6 patients, after blockage of the lower catheter this was converted into a ventriculoperitoneal shunt.

Approximately one-third were operated on under 3 months of age and only 2 were operated on after 6 months. Both these had their traumatic subdural effusions treated before hydrocephalus developed requiring operation. The timing of the first operation partly depended on the age of the infant on admission and partly on his condition, especially that of the CSF and the rate of head growth. Considerable and presumably necessary delay occurred while waiting for the CSF to clear of blood and for the protein content to diminish. At times the intracranial tension was so high that it was decided to operate in spite of a grossly abnormal CSF. This always resulted in disaster. The shunt system became infected and had to be removed and the infants died (as they would have done in any event).

Unreasonably long delay occurred in 5 infants because of late referral or because it was hoped that operation

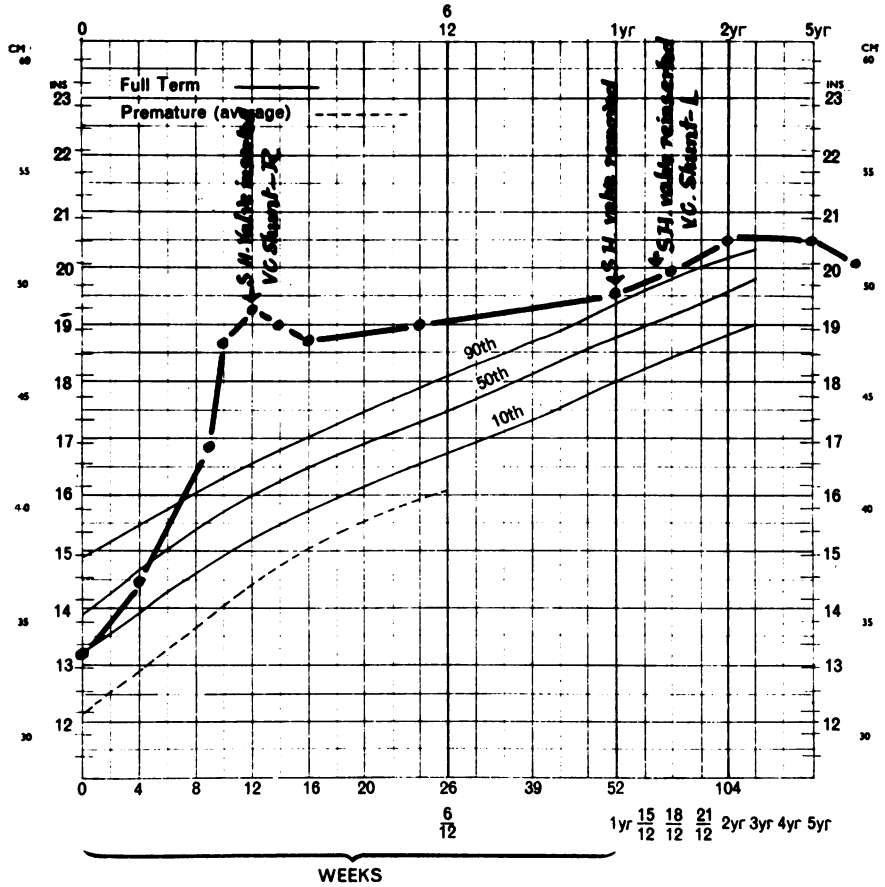

FIG. 3.—*Head circumference chart, showing undue delay in shunt therapy because of late referral. Bad result. Severely retarded child with physical sequelae.*

could be successfully avoided. This never happened in this series until isosorbide became available. 4 of the 5 whose shunt treatment was so delayed either died or survived with severe sequelae which might have been prevented (Fig. 3). In 3 infants the hydrocephalus appeared arrested at 3 to 5 weeks of age, but some 3 months later their heads started to grow suddenly, so that they eventually had extremely large heads (Fig. 4).

Revisions. Shunt blockages and infections. 17 patients (46%) underwent 49 further operations because of blockages or infections of the shunt. 1 patient had 8 revisions (Table VI). Many children died of unrelieved shunt complications. Retrospective analysis indicates that in 3 survivors unnecessarily prolonged delay occurred before revision, which resulted in serious consequences for the child, including permanent total blindness in an otherwise normal boy, and in severe mental deterioration in 2 others (Fig. 5).

TABLE VI
Revisions

Revisions	Alive	Dead	Total	
			No.	%
None	12	8	20	54
1	4	1	5	14
2	5	–	5	14
3	1	–	1	2
4–8	5	1	6	16
Total with revisions	15	2	17	46
Total no. of revisions	43	6*	49	
Per years of observation	1 in 3·0	1 in 4·0	1 in 3·2	
Unreasonable delay in revision with disaster	3	–	3	

*8 more were blocked or infected—not revised. Died.

Follow-up assessment

Follow-up consisted of outpatient supervision at ages 3, 6, 9, 18, and 24 months, and thereafter at annual intervals. Neurological examination, including inspection of the fundi, was carried out. Under 4 years

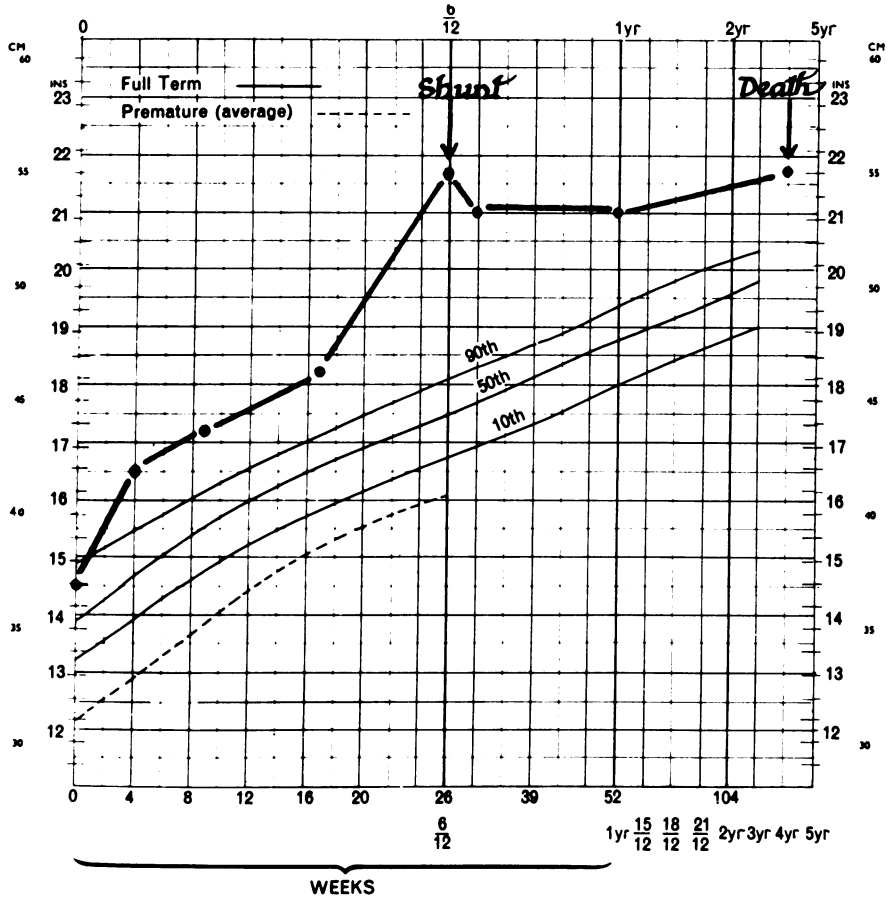

FIG. 4.—*Head circumference chart. Apparent spontaneous arrest of abnormal head growth followed by renewed activity of hydrocephalus. Late referral, as child was not seen often enough during the critical period. This child eventually died.*

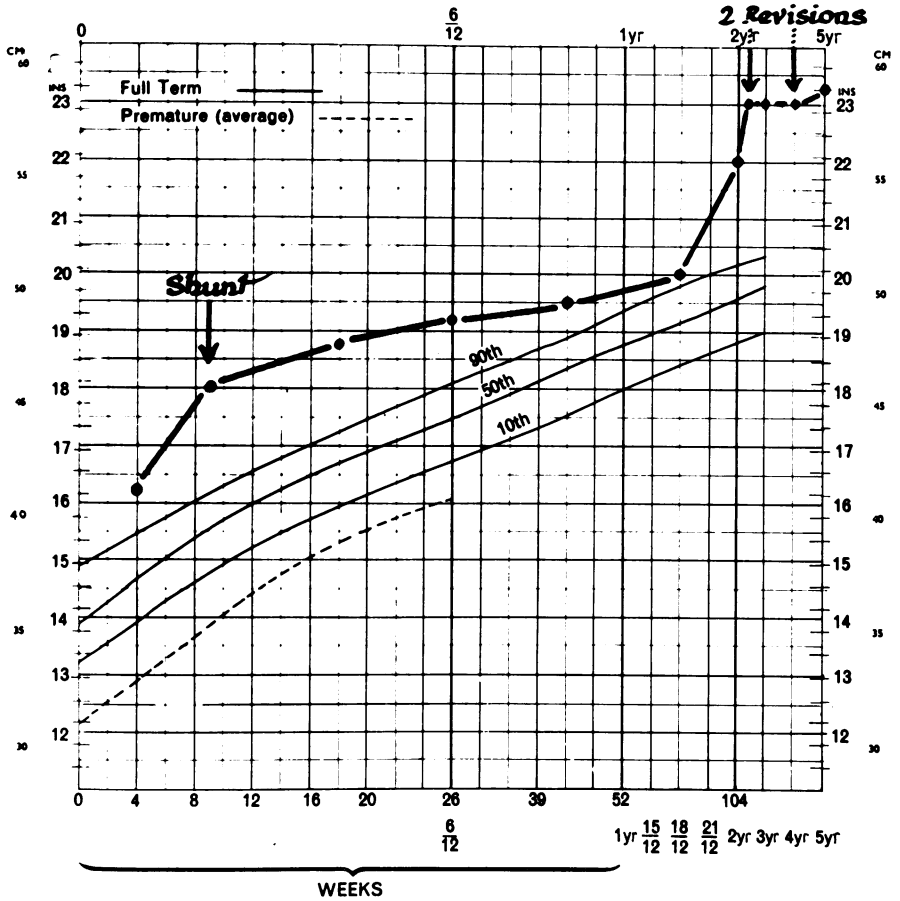

FIG. 5.—*Head circumference chart of a child whose blocked shunt was not recognized for over a year. He became severely retarded as a result.*

of age developmental assessment was made at the clinic. In older children formal intelligence tests were carried out by the education psychologists of the unit (Dr. J. Parsons and Mr. A. P. Lonton), using usually the Stanford-Binet and the Wechsler Intelligence Scale. Intellectual and psychological assessments were also made by the local education authority. Reports about scholastic progress were obtained from the head of the school. Postal communications were maintained with the parents if they left the district, but follow-up visits were often possible even from long distances. Medical reports were also obtained from colleagues who continued the care of children who left our district. No patient had been lost track of when the survey was analysed in December 1973. The age at follow-up is given in Table VII.

TABLE VII

Age at follow-up (32 survivors)

18 mth–5 yr	9
6 yr–10 yr	15
11 yr–16 yr	8

Results

Thirty-two survivors

(a) **Intellectual development** (Table VIII). The 7 untreated children and the 3 treated with isosorbide alone have already been detailed. 9 (19%) survive without physical sequelae and with normal intelligence (IQ 80–114) and 7 others with varied physical sequelae are also of normal intelligence (IQ 80–110). Altogether 16 (34%) survive with normal intelligence but only 9 (19%) have an IQ of 100 or more. Of 4 educationally subnormal (IQ 60–79) children, one has no physical handicap. He was a premature infant with a

TABLE VIII

Overall results

	No.		%
Normal, no shunt (IQ 80–114)	1	9	19
Normal, shunt (IQ 80–114)	8		
IQ 80–110, physically handicapped	7		
IQ 60–79	1	11	23
IQ 60–79, physically handicapped	3		
IQ <60	1		
		12	26
IQ <60, physically handicapped	11		
Dead	15		32
Total	47		100

TABLE IX

Principal physical defects in 32 survivors

	No.		%
Physical defects			
None	11		34
Fits	10		31
Hemiplegia, quadriplegia, paraplegia	12		38
Visual defects	11	47	34
Completely blind	4		13
Head circumference			
<10th centile	4		13
10th–90th centile	22		69
>90th centile	6		18

birthweight of 950 g. His low IQ of 60 is as likely to be due to his prematurity as to his hydrocephalus. 12 children (26%) are severely retarded with an IQ below 60. There is 1 severely retarded boy without physical sequelae; he displays autistic behaviour which may be the cause of his poor performance. 3 of the 11 profoundly retarded children had no treatment. Moderate to severe retardation was at least potentially preventable in 6 children. 2 of the moderately retarded and 3 of the severely retarded were referred extremely late for treatment (Fig. 3). Another child with a very low IQ made good progress till 18 months of age. After that his head circumference rapidly increased (Fig. 5), but as he had no other symptoms this was not at first recognized as a sign of blocked shunt though his head circumference reached 56 cm before the blocked lower catheter was revised.

Of the 32 survivors 50% have normal intelligence, 12% are educationally subnormal and 38% are grossly retarded.

(b) **Physical development.** 11 children (34%) are physically fit, including 1 who was treated with isosorbide alone. 21 (64%) are physically handicapped. Of the 15 dead children 13 were severely handicapped throughout their life.

The physical handicaps in 21 survivors vary from single and slight to combined gross defects. 10 (31%) are subject to fits (not due to blockage of the shunt); 12 (38%) have variable degrees of spastic hemiplegia, bilateral hemiplegia, or paraplegia; 11 (34%) have visual defects not amounting to blindness, and 4 (13%) are completely blind (Table IX).

4 children are not severely handicapped. 1 has persistent nystagmus (? congenital) without loss of visual acuity. Another, a boy of 12, has nystagmus with optic atrophy and poor vision: his scholastic progress in a school for the partially sighted corresponds with his IQ of 104. A boy of 16 has nystagmus with poor vision as well as short fits in spite of therapy, but attends an ordinary school: his IQ is 98. The fourth is a girl of 8 years; she has slight spastic paraplegia but her IQ is only 69 and she attends a school for the educationally subnormal. All the rest are much more severely affected. Among the worst are the 4 battered children, all of whom are profoundly handicapped physically and mentally.

The head circumference of 22 children (69%) is within the normal range. 4 others are microcephalic and all 4 are retarded. 6 are macrocephalic, 3 of whom are retarded. No child has a very large head (Table IX).

Fifteen dead patients (Table X)

Fifteen patients died (32%). The 4 who were untreated and 1 who had isosorbide treatment only have already been described. 2 who were not severely affected before their death at 4 and 9 years of age, respectively, are noteworthy because their deaths were preventable. 1 had a large head but was in excellent condition up to 3½ years of age when he suffered a head injury. He made an apparently quick recovery, but 4 months later was admitted

TABLE X

Age at and causes of death of 15 patients

Age	
<7 mth	10
3½–4 yr	3
9–10 yr	2
Causes	
Original haemorrhage	2
Untreated hydrocephalus	3
Blocked shunt	5
Infected shunt	3
Gross prematurity, liver disease	2

moribund and died as a result of an unrelieved blocked shunt. The second was well, with an IQ of 88 and had attended an ordinary school, but after several shunt blockages she died after a further such occurrence. Necropsy revealed that her ventricles had returned to normal size. All the others were profoundly handicapped throughout their life.

Summary of results and prognostic factors

In summary, in 19 patients (40%) the results may be considered good or fair, but in 28 (60%), including 12 profoundly handicapped survivors, no success was achieved. Tables I to IV detail the various prognostic features of the infants, related to the initial intracranial bleeding and to the condition of the infant on admission for treatment of the hydrocephalus. The outstanding features of prognostic importance were the neurological condition of the infant on admission for treatment of the hydrocephalus and the degree of ventricular dilatation.

The 12 infants who were spastic, the 8 who were still acutely ill with intracranial haemorrhage, and the 8 who had subdural haematoma with hydrocephalus at this stage, were all failures.* Of the 12 who were still subject to convulsions when admitted with hydrocephalus, only 1 achieved a worthwhile result. Nystagmus or failure to follow light on admission did not have quite such adverse prognosis. 4 of 14 (29%) were considered to be fairly satisfactory, though all 4 still have some visual defect consisting of nystagmus in 1 and partial vision in 3.

Of the 32 infants with single or combined neurological defects on admission only 6 (19%) achieved acceptable results, while of 15 without neurological signs other than the hydrocephalus, 13 (87%) achieved this, including all 9 fully normal children. The mortality of those with abnormal neurological features was 41% as compared with 13% in those without neurological signs on admission. The only 2 children who died in the latter group were potentially good cases, but died of shunt complications, while all the 13 in the group with neurological signs were severely handicapped as long as they lived.

The other important prognostic feature was the residual cerebral mantle. 15 of the 16 with acceptable results had a cerebral mantle of more than 5 mm. There was a progressive increase in survival rate from 29% in those with a cerebral mantle of less than 5 mm to 68% when the mantle was 5–15 mm and to 92% with a mantle of over 15 mm.

*One child with subdural effusion developed mild hydrocephalus after the successful treatment of the effusion. He is normal (Fig. 2a, b).

Shunt complications did not necessarily prevent normal development. 1 normal child had 4 revisions by 10 years of age and 2 others had 2 revisions each. Nevertheless, shunt complications were relatively rare in children without sequelae. 4 of the 8 shunt-operated normal children had revisions, a total of 9 further operations during 64 observation years or 1 revision per 8 years. This is much less than the average for the whole group (Table VI).

Shunt complications, however, can also have grave results. There were 5 children who were in good condition before blockage of the shunt. 2 of them died, one lost his sight, and 2 became severely retarded due to this blockage, which was recognized too late or was not effectively relieved by operation.

Discussion

Intracranial haemorrhage in the newborn is of grave significance. Among those who survive the initial haemorrhage an unknown proportion will develop hydrocephalus. Medical treatment has proved to be useful in a proportion of those treated with isosorbide (Lorber, 1972, 1973). At present, operation is still the main treatment. Many infants will still die or survive with grave sequelae because of the extent of the intracranial haemorrhage and the cerebral damage it causes.

A serious practical problem is the decision whether and when to interfere surgically. Undue haste might either lead to disaster with shunt complications, especially infections, or lead to an unnecessary operation on a child who may recover without it. It is hoped that in future isosorbide or other drugs will allow a longer time for the blood to absorb without allowing excessive growth of the head or the development of neurological complications, so that surgical intervention can be done under safer conditions. Possibly a larger proportion will recover without operation.

If treatment is to be successful the patients should be referred to an experienced unit early, so that the best time for successful intervention is not missed. The fact that only one-fifth of our patients survived without detectable sequelae is largely due to delayed referral and mistaken management after referral, rather than to the basic disorder.

Since evidence that hydrocephalus is developing may not be apparent for several weeks after recovery from an intracranial haemorrhage, it is essential to see these infants at least every 2 weeks for assessment and measurement of their head circumference. Had this been done to many patients in this series considerable delay might have been avoided.

A large number of persons contributed to the care and investigation of these children including our medical and surgical colleagues in the hospital and in the School Health Service, colleagues who referred the patients, and Dr. J. Parsons and Mr. A. P. Lonton who carried out the intelligence tests.

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