THE NATURAL HISTORY OF HYDROCEPHALUS

DETAILED ANALYSIS OF 182 UNOPERATED CASES

BY

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Few sights are more distressing than a child with severe hydrocephalus associated with mental and physical defect. These cases, and clinical accounts based on highly selected series, are probably responsible for the depressing descriptions found in most standard textbooks. Earlier one of the authors (K.M.L.), presenting the preliminary results of an investigation on the natural history of hydrocephalus, suggested that the outlook in this condition was not without hope (Laurence, 1958). After further study those results have been reassessed and presented in greater detail.

Material

The investigation is based on the 239 cases of hydrocephalus under the age of 13 years, of whom 130 were males and 109 females. They were seen by Mr. Wylie McKissock at The Hospital for Sick Children, Great Ormond Street and the Atkinson Morley Hospital during the 20 years between January 1938 and December 1957 (Fig. 1). He advocated a conservative policy, but surgery was occasionally carried out by his assistants.

Operated Cases. Altogether 57 patients (29 males and 28 females) came to surgery (Table 1). Eight

cases were operated upon in other centres after the parents had sought further opinions. Removal of these 57 from the total did not greatly affect the composition of the series of unoperated cases on which this study is based, even though the group of operated cases is different in composition. Table 2a and b shows among the operated cases a large number of aqueduct stenosis, a condition for which surgery was thought to be more rewarding. Only five cases with spina bifida cystica and Arnold-Chiari malformation were operated upon for their hydrocephalus as these were regarded as hopeless.

Sixty-one operations of various types were carried out, including 19 Torkildsen's ventriculo-cisternostomies and 11 ventriculo-sinusostomies (Table 3). The indications for each procedure varied but with the exception of the lumbar intramedullary screw cannula operation (L. Walsh, 1953, personal communication) and a few of the Torkild-

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TABLE 1
THE SERIES

Cases	Numbers
Unoperated cases	182
Operated cases	57
at the Atkinson Morley	22
at The Hospital for Sick Children	27
elsewhere	8
Total number of cases followed up	239

FIG. 1.—Year of birth of the 239 hydrocephalic children under 13 years, both operated and unoperated.

TABLE 2A
COMPOSITION OF THE SERIES:
SITE OF BLOCK

Site of Block	Total Series		Natural History		Operated	
	No.	%	No.	%	No.	%
Aqueduct	59	25	23	40	36	20
Exit foramina ..	15	6	7	13	8	4
Basal cistern ..	100	42	22	38	78	43
Arnold-Chiari ..	39	15	5	9	34	19
Unknown	26	12	0	0	26	14
Total	239	100	57	100	182	100

TABLE 2B
COMPOSITION OF THE SERIES:
AETIOLOGY

Cause	Total		Natural History Series		Operated Cases	
	No.	%	No.	%	No.	%
Malformation ..	57	24	43	23	14	25
Trauma	76	31	59	32	17	30
Infection	48	20	42	22	6	10
Tumour	6	2	4	2	2	4
Unknown	52	23	34	21	18	31
Totals	239	100	182	100	57	100

TABLE 3
OPERATED CASES

Type of Operation	Number of Operations	Number of Cases Alive December 1957
Tentorrhaphy	3	2
Torkildsen's	19	10
Screw laminectomy ..	4	4
Sinuveniculostomy ..	11	8
Posterior fossa exploration or decompression	9	1
Removal of fourth ventricular membrane	4	0
Others	11	2
	61*	27†

* Four patients have had two separate operations.

† One patient has had two operations.

TABLE 4
DIAGNOSIS OF HYDROCEPHALUS
182 cases

Diagnosis	Number	Per cent.
Encephalography and other investigations ..	94	51
Ventricular punctures and dye studies ..	21	13
Skull radiograph and head measurements ..	31	17
Head measurements only	29	16
Autopsy only	6	3

sen's operations, all cases had progressive, though not advanced, disease, and at the time most were under 1 year old. On average the heads of the operated cases measured 2.33 in. (5.8 cm.) above the mean for the age when first seen, compared with 2.43 (6 cm.) in the unoperated series. It is of interest that of these 57, 31 (54%) had died by the time the investigation was concluded.

Unoperated Cases. The 182 patients (101 males and 81 females) that were not operated upon form the subject of this paper. These cases may still be considered representative of all those seen by Mr. Wylie McKissock, as there is no marked difference between these 182 patients and the total sample. The diagnosis of hydrocephalus rested on air encephalography, ventricular punctures and dye tests in addition to the less specialized investigations in 94 cases. In 69 only a radiograph of the skull or the serial head measurements were available. In six cases confirmation of the diagnosis was not available until autopsy (Table 4).

The series includes 36 cases of aqueduct and 78 basal cistern blocks, due in the main to perinatal 'trauma' (59 cases), and infection (42 cases). There are only 34 cases of spina bifida cystica with Arnold-Chiari malformation, largely because these infants were discouraged at the clinic, and because so many of them died early, before they were referred for treatment. The four cases of tumour represent the few diagnostic failures and were unexpected findings at autopsy. Cases of hydrocephalus following tuberculous meningitis were excluded, as these were considered quite a different problem. As a rule, the site of the block was easily found because of extensive investigation. The 14% of 'unknowns' represent the cases who were not investigated, through wartime conditions, or occasionally where the investigations proved unsatisfactory, or where the hydrocephalus was severe enough to appear completely hopeless. The aetiology was more difficult to establish, for often there was no definite history of meningitis or proven birth haemorrhage, and no associated spina bifida cystica nor post-mortem examination. Sometimes it was possible to categorize them on grounds of probability, using such pointers as the obstetric history and the time of onset, but in 19% even this proved impossible.

This group of hydrocephalics is only representative of cases seen at large children's hospitals to which they have been referred from other hospitals or paediatricians; it is not representative of this condition in the community as a whole as the severest cases are considered beyond help or die before being seen, and in the mildest cases the disease

may have already arrested spontaneously. Thus, in this series there is a comparatively small number of cases with spina bifida cystica which is by far the commonest cause of hydrocephalus in the community and carries a very high mortality largely from ascending meningitis.

A comparison of deaths for each age group reported by the Registrar-General, and deaths found in the series (Table 5 and Fig. 2) shows that the two rates become comparable only after the age of 3 months, so confirming that many of the severest cases are not included in the series. After that age the hospital cases are representative of the disease in the community. The unoperated cases from the Neurosurgical Unit represented about 3% of all the hydrocephalic infants in England and Wales surviving beyond 3 months.

Of the patients, 67% were first seen at the clinic when under 1 year old, including 31% who were less than 3 months old. Only 8% were over 4 years at their first consultation. When re-examined, usually many years later, only 45% including the nine who were still progressive were below school age (Fig. 3).

Methods

The full and systematic hospital case notes were scrutinized, with particular attention to family and obstetric histories, results of investigations and progress of the condition. Skull radiographs and encephalograms were re-examined.

Most of the patients had to be traced as many had moved home several times on account of war upheaval and post-war resettlement. The methods used in tracing the patients were described in the *Lancet* (Laurence, 1959a), and all but three were found. One of these, an illegitimate child, was adopted in 1943, and further attempts at tracing were thought undesirable. The second was placed in a nursing home in 1942 from which he was later removed, and tracing proved impossible as the case notes were destroyed in 1947 and because the child bore a common fore- and surname. The last patient returned to Ireland with his parents in 1945, who are then believed to have emigrated to Australia.

From the various reports a picture of the progress of the condition was built up. With a few exceptions, all the traced survivors were re-examined by one of us (K.M.L.) at The Hospital for Sick Children or at the institution where they were being cared for, with special attention to the central nervous system and vision. Physical disabilities were graded according to the severity of the handicap (see Table 11).

Intelligence tests were made (by S.C.) on most of those with natural arrest over 18 months. Children who were too handicapped to be brought to The

TABLE 5
COMPARISON OF HOSPITAL AND REGISTRAR-GENERAL'S DEATHS FROM HYDROCEPHALUS

Age	Number of Deaths		Ratio
	Series	Registrar-General*	
Months			
0-2	8	260	1 : 32.50
3-5	20	65	1 : 3.25
6-8	12	49	1 : 4.08
9-11	10	35	1 : 3.50
Years			
1-2	14	61	1 : 4.36
2-5	14	53	1 : 3.79
5-10	7	23	1 : 3.29
10-15	3	7	} 1 : 3.50
15-20	1	7	
20-30	0	11	

* Total of the Registrar-General's returns for England and Wales for 1952, 1953 and 1955.

Hospital for Sick Children or who were living abroad, had tests performed locally by educational psychologists. For some of the very grossly retarded children only intelligence assessments were made, since full intelligence tests were not considered justifiable. The type of test employed at The Hospital for Sick Children, whether the Revised Stanford-Binet, the Merrill-Palmer or the Griffiths Mental Development test, depended upon the mental age of the patient (Table 13).

FIG. 2.—A comparison between the deaths occurring in each age group in the series and those reported by the Registrar-General. The deaths have been expressed as the number of deaths occurring in a full year. For the Registrar-General's deaths an average has been worked out based on the reports for 1952, 1953 and 1955.

FIG. 3.—Age at re-examination. The nine progressive cases all under 5 years are indicated.

FIG. 4.—Life chart, prepared from the data on the life table. This can be used to compute the life expectancy of an 'average' hydrocephalic at any age after 3 months (see text).

TABLE 6
RESULTS
(182 cases)

Result	Numbers	Per Cent.
Alive:		
Hydrocephalus arrested	81	46.2
Untraced (presumed alive)	3	
Still progressive	9	5
Dead	89	48.8

Only a physical examination was performed on patients where the condition was still progressive.

Results

Survival. Of the 182 unoperated cases, 81 (38 males and 43 females) were found alive and well at the end of 1958 with the hydrocephalus spontaneously arrested. Three were presumed alive as their disease too had arrested spontaneously before they disappeared. In nine children, then still under 5 years, the condition was still progressive, and the remaining 89 cases (63 males and 38 females) had died (Table 6).

If no account is taken of the effective number of patients at risk at any one time, then this result might suggest that 46% of hydrocephalus cases referred to hospital could be expected to arrest without surgery and thus, like many hospital statistics, give a false picture of the prognosis. When, on the other hand, the life expectancy suggested from this series is computed on actuarial principles using a life table (Table 7 and Fig. 4), the chance of a hydrocephalic child, seen soon after birth, reaching adult life is 20% or 23%, depending on the method of calculation employed. After three months the hospital figures compare with the Registrar-General's so the 'average' hydrocephalic

TABLE 7
LIFE TABLE

Age	Effective Number At Risk	Number Who Died	Mortality/1,000	Loss	Mortality/Month/1,000	Life Table
Months						
0	9.5	0	0	0	0	1,000
1-2	37.5	8	213.5	213.3	107	1,000
3-5	66.5	20	300.8	236.7	100	786.7
6-8	79	12	151.9	83.5	51	550.0
9-11	81	10	123.4	57.6	41	466.5
Years						
1-2	86.5	14	161.8	65.8	13.5	406.9
2-5	76	14	184.2	62.8	5.1	341.1
5-10	51	7	137.3	38.2	2.3	275.3
10-15	29.5	3	101.7	24.4	1.7	240.1
15-20	13.5	1	74.1	16.0	1.2	216.3
						200.3

in the population, who has survived to 3 months, has a 26% chance of reaching adult life without surgery, and if he is already between 1 and 2 years, he has a 50% chance. It must be emphasized that this is an average and that hydrocephalus is a condition with separate aetiologies, all having different characteristics and prognoses.

Obstetric Factors. Factors intervene which lessen the value of retrospective studies in obstetric histories. Attempts are made to rationalize the unfortunate child's condition; some important events are deliberately suppressed and insignificant ones magnified. Pregnancy and delivery histories were normal in less than half, 47% or 87 cases (Table 8), and of these, 24 cases of hydrocephalus followed meningitis, 17 cases were due to spina bifida cystica and Arnold-Chiari malformation with no obvious hydrocephalus at birth, and 22 cases were of uncertain aetiology. Only 10 were 'traumatic'.

In the abnormal obstetric histories, there was an undue number of potentially traumatic deliveries, i.e. 63 cases. Of these, there were 18 with labour prolonged beyond 18 hours after regular contractions were established; 10 were breech deliveries often aided by forceps, and after long labour; 28 were delivered by the vertex with forceps; four were precipitate labours, and three had repeated version before being delivered normally.

In some instances it is a matter of conjecture whether the delivery was traumatic because the head was disproportionately large, or whether hydrocephalus developed because of the traumatic delivery, for occasionally no accurate information concerning the head size at birth is available. There were, in addition, eight sets of twins, six more than expected, and 20 premature births all of which are liable to birth trauma. Of these 28 cases, 15 did indeed appear to have sustained some degree of perinatal trauma. The twins afforded an interesting group

as in each case only one child was affected (see later).

Five cases of postmaturity (more than 42 weeks) were included, but the usual proportion of toxæmias and caesarean sections was recorded. Hydramnios was noted in only two cases, and only three children were illegitimate. A family history of major central nervous system malformation appears to have been present in three instances.

Age of Onset. Slight head enlargement, some fullness of the fontanelles, with separation of the sutures, slight or intermittent downward deflection of the orbital axis, all early signs of hydrocephalus, are not easily recognized, and there may be considerable ventricular dilatation before the classical clinical features become manifest. The hydrocephalus itself may appear so gradually that those in daily contact with the infant do not become aware of the condition until it is well advanced. Generally early diagnosis is made only when expected or specifically looked for, or when some astute observer sees the baby for the first time or intermittently. Thus times of onset given may well be later than the real onset of the condition.

As hydrocephalus has a number of different causes, it is diverse in character, with the age of onset varying according to the aetiology. When due entirely to a malformation, it may be obvious at birth, and give rise to difficulties in delivery. In spina bifida cystica with Arnold-Chiari malformation this is not necessarily so, though they may well have some ventricular dilatation (Lorber, 1961), but they develop severe hydrocephalus often following ascending infection from the spina bifida sac. All the cases in the series of congenital aqueduct stenosis had obvious hydrocephalus by the end of the first month, and nearly all of the spina bifida cysticas and Arnold-Chiari malformations developed their hydrocephalus before the age of 6 months, when

TABLE 8
OBSTETRIC FACTORS
(182 cases)

Feature	No. of Cases	Percentage	Usual Percentage Reported
Caesarean section	9	5	6.2 (Marshall and Cox, 1950)
Toxaemia	18	10	3.5-18 (Lewis, 1956)
Postmaturity (42 weeks +)	5	3	14.1 (Gibson and McKeown, 1950)
Prematurity (46 weeks or less)	20	12	7 (Douglas, 1950)
Twins pregnancy	8	5	1.2
Other abnormalities	63	37	
Forceps	28	15	16-17 (Jeffcoate, 1953)
Long labour	18	10	3.2 (MacRae, 1949)
Breech	10	6	2.2 (Moore and Steptoe, 1943)
Precipitate	4	3	—
Versions only	3	2	—
Normal pregnancy and delivery	87	48	—

FIG. 5.—Age of onset broken down according to the aetiology of the hydrocephalus. In 11 cases insufficient information was available.

the sac is usually fully epithelialized and infection is excluded (Fig. 5). The exceptions were two cases of spina bifida cystica, both with comparatively

mild ventricular dilatation, who almost certainly had hydrocephalus earlier than the age at which it was recognized.

When hydrocephalus originated from perinatal trauma, the head usually remained normal in size for the first two to three weeks, and began to enlarge during the next two months. One child who developed hydrocephalus following a head injury at 18 months was included in the series.

There is a much wider scatter in the time of onset with the post-meningitic cases, for here hydrocephalus generally follows within a few weeks of the infection.

Natural Arrest. Natural arrest is a gradual process. When carefully read and charted head measurements taken at three consecutive monthly intervals remain the same, after a gradual slowing down of growth rate, then arrest is reasonably certain. When this coincides with noticeable improvement in the child's general condition, a reduction in the fontanelle tension and a return of the orbital axes to normal, and when also there is a commencement of rapid physical as well as mental development, natural arrest is very likely to have occurred. If careful head measurements are continued it will be observed that these remain the same until the infant has grown sufficiently for the head to become normal, relative to the body, before the head growth resumes (Fig. 6A). When this does resume it will be at the normal rate. In other cases, especially where the head has become very large, growth never resumes (Fig. 6B). In some cases, head growth resumes well before the normal head-body relation has been restored, but then at a rate parallel to normal growth (Fig. 6C). An actual reduction in the head size, as is often seen after surgical procedures, does not occur in spontaneous arrest.

In 47 cases in this series, arrest diagnosed on the basis of serial head measurements took place usually between the ages of 9 months and 2 years. In six

FIG. 6.—Types of skull growth after natural arrest:

The 90th percentile growth curve is shown on each graph and the examples drawn in as interrupted lines with an arrow indicating the time by which arrest is thought to have taken place.

- The rapid head enlargement was followed by 14 months of cessation of skull growth, by which time the measurement was below the 90th percentile. Slight growth resumed but the head remained below the 90%. The I.Q. was 85.
- Very rapid skull growth is followed by a complete cessation of growth. The head remained well above the 90th percentile and growth never resumed. E.B. had an I.Q. of 59 and C.R. of 101.
- Rapid head growth followed by natural arrest when the head was considerably above the 90th percentile. Head growth then continued at a rate parallel to the 90th percentile line. F.M. had an I.Q. of 113 and P.K. of 88.

cases the disease arrested earlier, while in 27 others the condition smouldered on (Fig. 7). It appears that no one aetiological group seems to arrest at an earlier time than another. Factors causing this spontaneous arrest are unknown, but obviously an equilibrium between formation and absorption of the cerebrospinal fluid must have developed. In some cases a diminution in the amount of fluid formed probably occurs, for the choroid plexi, regarded by most authorities as the seat of formation of at least part of the cerebrospinal fluid, are often small or fibrosed at autopsy (Laurence, 1959b). In others, the extracerebral cerebrospinal fluid pathways may have become partially patent again. This equilibrium seems to remain precarious and may be disturbed by a number of factors, including ventricular or lumbar puncture as carried out for encephalography, or sometimes by minor head injury.

Recrudescence of acute progressive hydrocephalus can occur without any apparent precipitating cause (see next section).

Time and Cause of Death. Two-thirds of the 89 cases in the series who died did so before the age of 18 months, but 24 died after the age of 2 years, and 11 did so after their fifth birthday (Fig. 8). Apart from the cases of spina bifida cystica succumbing in the early months, no significant linkage of age and aetiology or cause can be deduced, though there is a very slight tendency for the cases of aqueduct blocks to die earlier.

Decision regarding the actual cause of 36 deaths was based on autopsies, some with bacteriological and histological investigation. A report of the terminal illness, often detailed, was obtained in another 36 without autopsy, but for the remaining 17 only the death certificate was available.

'Hydrocephalus' in the form of cardio-respiratory

TABLE 9
CAUSE OF DEATH

Cause	Numbers
Hydrocephalus ..	46
Acute hydrocephalus following investigation ..	7
Infection ..	16
Following investigation	4
Other related ..	22
Intraventricular haemorrhage	2
Chest infection	16
Tumour extension	3
After operation for spina bifida	1
Unrelated (gastro-enteritis)	2
Uncertain (probably hydrocephalus)	3
Total	89

FIG. 7.—Age at arrest of all cases with natural arrest. In 10 cases insufficient information was available to make an accurate assessment.

failure due to compression of the vital centres often together with hyperpyrexia was the cause of death in just over half the cases (46) (Table 9). Generally this followed a steadily progressive deterioration. On the other hand, in seven cases rapidly fatal acute hydrocephalus was precipitated by ventriculography, when previously the condition was only

FIG. 8.—Age at death divided up according to the site of block that caused the hydrocephalus.

FIG. 9.—Cause of death divided up according to both the site and the aetiology of the blockage (see text).

slowly progressive, and in two children, in whom the condition had become arrested spontaneously nine and 10 years previously, death took place suddenly after a brief period of progressive unconsciousness without any apparent precipitating cause. In two more the disease became fatally progressive again after remaining quiescent for seven and six years, again without apparent reason (Table 10).

Meningitis caused the immediate death of 16,

TABLE 10
CAUSE OF DEATH IN CHILDREN 2 YEARS AND OVER
(24 cases)

Cause	Numbers
Hydrocephalus	16
Intracranial hypertension after arrest	2
Progressive hydrocephalus after arrest	2
Progressive hydrocephalus	10
Following ventriculography	2
Infection	4
From spina bifida cystica	2
Following ventriculography	2
Other related	4
Tumour extension	1
After operation for spina bifida	1
Bronchopneumonia	2

of which 10 had developed ascending infection from a spina bifida cystica. A recrudescence of a meningitis due to an unusual organism occurred in one case after three years of freedom from obvious infection; another was never adequately cleared of her infection; and four cases developed acute meningitis after ventriculography, three because they required continuous ventricular drainage.

Most important amongst the related causes of death was bronchopneumonia as a terminal event. The two examples of intraventricular haemorrhage were due to a bleeding diathesis in one case, and to an unfortunate ventricular puncture in the other. Two cases of gastro-enteritis must be classed as unrelated though the children would probably not have succumbed if progressive hydrocephalus had not been present as well. In three cases the exact cause of death remained uncertain (though they probably belonged to the 'hydrocephalus' group), as the information from the death certificates about their terminal condition could not be amplified from other sources.

It will be seen from Fig. 9 that there appears to be little relation between the site of the obstruction and the cause of death except that none of the cases showing aqueduct block died of infection and that a large proportion of the Arnold-Chiari malformation succumbed to infection. As far as aetiology is concerned, only one of the 32 'post-traumatic' cases died of the same complication.

Condition of the Survivors with Arrest

Physical Disability. The patients were carefully examined and the physical disabilities were divided into four groups according to the severity (Table 11).

Group 1 included all the cases who were physically normal or who had only a slight or intermittent

TABLE 11
PHYSICAL HANDICAP

Category	Examples of Disabilities	No. of Cases
Normal Group 1	None Slight squint only Brisk reflexes only	26 (12 males) (14 females)
Slightly handicapped Group 2	Slight spastic Unsteady gait Impairment of vision Slight imbalance	16 (10 males) (6 females)
Severely handicapped Group 3	Sphincter paralysis Paralysis of legs Flexion deformities Unable to walk Severe incoordination Blindness	23 (9 males) (14 females)
Incapacitated Group 4	Confined to bed, Spastic quadriplegia Can just feed himself	16 (7 males) (9 females)

squint or who had brisk reflexes. There were 26 such children and they included a large proportion of cases of reasonable intelligence, with basal cistern or aqueduct blocks, but none with spina bifida cystica (Fig. 10).

Group 2 consisted of 16 cases with comparatively slight physical disabilities which in themselves would not always necessitate education in special schools. Disabilities such as slight spasticity, unsteady gait, some impairment of vision or slight incontinence are included. As with the remaining two groups, all types of hydrocephalus, including the Arnold-Chiari malformation are found amongst them.

There were 23 severely handicapped children in Group 3, all of whom required education in special schools or institutions. Cases with sphincter and limb paralysis and flexion deformities were amongst these, as well as children who were unable to walk because of severe incoordination. Some were blind.

The last group, the completely incapacitated, consisted of 16 cases largely with very low Intelligence Quotients, who were either in institutions or confined to bed at home and had to have home tuition where this was practicable. Many of these had spastic quadriplegia and some were unable to feed themselves.

On the whole little difficulty was experienced in deciding into which group a patient belonged, except in groups 2 and 3. Whenever there was doubt, the child was placed in the lower group. Fig. 15 suggests that the grouping appears to have been correct, as the line linking the average Intelligence Quotients shows no marked deviation from the straight.

Vision. Although blindness is stressed in the textbooks as a common complication of hydrocephalus, severe impairment of vision was found in comparatively few of the survivors. Vision was regarded as normal in 34 patients, while in a further 25 no comment was made about it (Table 12). In a further four no assessment was possible on account of extreme mental retardation. Only six children were actually blind and a further five had defective vision. It is of interest that 12 were thought to be blind in infancy while the disease was progressive. Probably this belief arose from a combination of the lack of interest in surroundings and lack of response to stimuli (including visual) common in severely ill children, and the findings of pale discs, normal in the very young, mistakenly thought to be due to optic atrophy. In fact only five of these 12 children were later found to be blind or to have a severely defective vision. One of them, indeed,

FIG. 10.—Physical disability in 81 cases divided according to both site and the aetiology of the blockage (see text).

became the school's champion marksman, while another was earning her living making costume jewellery.

A divergent squint was a more constant finding (12 cases), but only in two cases was this a marked disability. In five patients this squint was mild and was the only eye defect.

Psychological Investigations. It was possible to obtain assessments of intellectual status in all 81 surviving cases and in all but nine this assessment was based on a test. Most of the tests were carried out by one of us (S.C.) at The Hospital for Sick Children; the remainder, on children who lived abroad or could not be brought to the hospital, were carried out by local educational and clinical psychologists (Table 13).

The figures in this and later Tables differ from those given in the original paper, since 10 children have now been tested on whom previously only estimates were made, since they were at that time too young. It might be of interest to note that the average estimated I.Q. of these 10 children, whose

TABLE 12
VISION
(81 cases with natural arrest)

Normal vision	34	} 59
Vision not remarked upon	25	
Slight squint only	5	
Severe squint	2	
Vision defective	5	
Blind	6	
No assessment possible	4	

TABLE 13
TYPE OF INTELLIGENCE TEST USED

Test Used	Where Tested	Number	Per Cent.
Revised Stanford-Binet	Hospital Sick Children	28	35
Revised Stanford-Binet	Elsewhere	12	15
Merrill-Palmer Scale	Hospital Sick Children	10	12
Griffiths' Mental Development Scale	Hospital Sick Children	18	22
Other tests (1)	Elsewhere	4	5
Estimates only		9	11
		81	100

Two of these four 'other' tests were the results of the 11+ intelligence tests; one was a special test for blind children; and the last was reported from an institution apparently on the basis of Gesell norms.

average chronological age (C.A.) at the conclusion of the investigation was 3·5 years, was 47 with a range 10-80; and their average tested I.Q. was 43 with a range 4-82.

All children of school level were tested on the Revised Stanford-Binet Intelligence Scale as being the most acceptable clinical instrument. The Block Design Sub-Test from the Wechsler Intelligence Scale for Children was given to 19 children. It would have been interesting and useful to have done the full-scale Wechsler, but since many patients came long distances, without personally gaining very much from the investigation, they could not

be asked to come again. The Merrill-Palmer Scale of Mental Tests was used for the mental age range 2 years to 5 years unless there was serious physical handicap which made such a predominantly performance scale unusable. In fact most of such children, being of low mental age, were more accurately assessed, as were the less physically handicapped also of low mental age, on the Griffiths Mental Development Scale. Reading tests were also given.

The Distribution of Intelligence. The results of all tests and estimates are summarized in Fig. 11 (on which a normal distribution of the same number of cases has been superimposed) and Table 14.

The average intelligence quotient was 68·95. The scores were not distributed normally about this mean: the standard deviation was 30·61. There was no relation between sex and intelligence.

Reading Test Results. Of the 28 children who were tested on the Revised Stanford-Binet test, 21 were tested on the Schonell Graded Word Reading Test; that is, all those of intelligence levels equivalent to school age, excluding seven on the borderline with mental ages of about 6. The average chronological age of these 21 children was 11 years 5 months; their average reading age

FIG. 11.—Distribution of intelligence test results on 81 cases. The distribution is not normal about the mean (of 68·95) and shows a standard deviation of 30·61. The superimposed line represents the distribution of the same number of test results in a 'normal' population.

TABLE 14
DISTRIBUTION OF INTELLIGENCE

Group	I.Q.	No. of Cases	Per Cent.	
Normal intelligence	> 85	31 (18 F. and 13 M.)	38	73
Educational subnormality	50-85	28 (12 F. and 16 M.)	35	
Ineducable	< 50	22 (13 F. and 9 M.)	27	

was 9 years 10 months. It is clear that the educable children were in fact being educated.

Relationships of Intelligence and Other Measures. Some of the differences in intelligence among these surviving 81 children will be the result of innate difference which would have been there in any case. An attempt was made to find out what caused the larger part of the differences. A relation between the intellectual outcome and other measurable differences was looked for but was not generally found.

Head circumference at the time of re-examination varied from less than 0.25 in. (0.63 cm.) to 11 in. (27.94 cm.) above the mean for the age; but there was no relation between the final size and the final intelligence quotient (Fig. 12). The child with one of the largest heads had an I.Q. of 92.

It was possible in 68 cases to make an estimate of the duration of the illness, i.e. the length of time the brain had been under raised pressure or the time during which the condition remained progressive. This varied from two months to eight

years, the average being two years (Fig. 13). There was no relation, linear or otherwise, between the intelligence quotient and the duration of the illness.

In 38 cases there were measurements of the thickness of the 'cortex' as found on air encephalography in most cases during the acute part of the illness. This varied from 0.5 cm. to 4.5 cm. the mean being 2.1 cm. There was no correlation between this measurement and the later intelligence quotient (Fig. 14). It is not possible to predict the later condition from the condition of the brain at the height of the pressure, for example the five cases with a thickness of less than 1 cm. had quotients from 60 to 85. This absence of a relation between cortical thickness and later intellectual level is paralleled by the absence of a relation between the E.E.G. findings and intracranial tension or thickness of the pallium, except that over the completely atrophic cortex the traces are of very low amplitude (Laurence and Pampiglione, 1960).

As described above, the children were assessed for degree of physical disability: 42 cases (52%) were in the higher groups 1 and 2, leaving 39 cases

Fig. 12.—Distribution of intelligence assessments plotted against head measurement above the mean for the age and sex in 81 cases. There appears to be no relation. A relation between I.Q. and physical defect is shown.

FIG. 13.—Distribution of intelligence assessments plotted against the duration of progressive hydrocephalus. No relation exists.

(48%) in groups 3 and 4. There was a high negative correlation between degree of physical handicap and intelligence: $r = -0.712$ (Fig. 15). The more physically handicapped were also the less intelligent. This relation was further investigated.

Clearly increasing age might lead to the overcoming of a physical handicap with the result that the relation between intelligence and physical handicap might disappear as the child got older. That

this was largely not so was demonstrated. There was a correlation between age and intelligence but it was significant only at the 5% and not at the 1% level: $r = -0.274$. There was no significant correlation between age and degree of physical disability, $r = 0.202$. Apparently patients do not overcome their physical handicaps as they get older. Using these figures the partial coefficient of correlation between degree of physical disability

FIG. 14.—Distribution of intelligence assessments plotted against thickness of the cortex. No relation is shown. There is also no relation between the physical state and the cortical thickness.

FIG. 15.—Relation between physical disability and intelligence assessments. There is a high negative correlation with the more physically handicapped being also the less intelligent. The line links the mean I.Q. of each group.

and intelligence, with age held constant, remains very much as it was when contaminated with age, namely: -0.699 . This is shown in Fig. 16. Most of the testing was verbal, especially in the patients with considerable physical disability, so this relation merely shows that if the brain is damaged at all it is likely to affect both motor development and intellectual development and that neither improve very much, if at all, with age.

Emotional Balance. It was possible to form a judgement on the emotional stability or balance of rather more than half the children. This classification into disturbed and normal does not mean that the disturbed were seriously neurotic or psychotic. The principal deviation from normality was excessive shyness, anxiety or nervousness, which led to withdrawal, inactivity and a difficulty in tackling problems. It is not surprising that damaged children should react in this way. It is perhaps more surprising that so many should be described as cheerful, energetic, friendly, enthusiastic, mature; all epithets that qualified a child for normality.

These judgements were made during the testing and therefore all cases tested elsewhere were excluded. There were also some children who were so young and so handicapped that either their whole personality seemed to be involved in the problem of movement or physical balance leaving nothing over for personal relationships or they were unable to express any feeling or relationship because they were so defective and immobile.

These judgements are tabulated in Table 15. It will be seen that the 44 children on whom judgements were possible are not representative of the sample as a whole. Of the 16 most physically handicapped children (group 4) only one was suitable for judging: of the 23 in group 3, 10 were unsuitable. The distribution of the 44 judged and the 37 omitted is significantly different ($\chi^2 = 19.58$, $p > 0.001$). The proportion of children showing no disturbance is 64% of those judged. But this is misleading. Probably we should be content with the figure of

FIG. 16.—Relation between age and intelligence assessments. The unbroken line represents the mean regression line. The regression line that has been calculated (broken line) is the average regression over the four disability groups (i.e. with disability held constant), and the slope of the regression is only nearly significantly different from zero. The I.Q. thus does not significantly rise with age nor is the physical disability overcome as the patient gets older.

35% of the whole sample as certainly normal, 20% as certainly disturbed, and 45% as unknown.

On the other hand Table 15 also shows that in those 44 children on whom a judgement was possible, there was no relation between the presence of disturbance and the degree of physical handicap ($\chi^2 = 0.89$). Such disturbance as was found could not be attributed to the physical handicap. Perhaps those severely handicapped children omitted from the judgement will not be so unhappy.

Brain Damage and Test Results. The sample of 81 children, all of whom had suffered some degree of brain insult with, as has been seen, effects of varying degrees of severity and permanence, was used to test various theories about test results in brain damaged children.

It is commonly found that the vocabulary score in many kinds of brain damaged adults, excluding arteriosclerosis, deteriorates less than scores on other kinds of tests, particularly tests involving spatial perception like the Koh's Block Design Test. It

TABLE 15
EMOTIONAL BALANCE AND PHYSICAL DISABILITY

Physical Grade	Whole Sample (81)		Omitted From Judgement (37)		Disturbed (44)		Normal (44)		Total Disturbed and Normal	
	No.	%	No.	%	No.	%	No.	%	No.	%
1	26	32	8	22	6	14	12	27	18	41
2	16	20	4	11	5	11	7	15	12	27
3	23	28	10	27	5	11	8	18	13	30
4	16	20	15	41	0	0	1	2	1	2
Totals	81		37		16	36	28	64	44	

TABLE 16

COMPARISON OF INTELLIGENCE QUOTIENT BETWEEN HYDROCEPHALIC AND NEUROTIC CHILDREN

	Intelligence Quotients	
	Hydrocephalics	Neurotics
Mean Binet I.Q.	83·95	83·70
Standard deviation	20·47	21·71
Mean age (years)	12·05	12·11
Standard deviation (years)	3·47	3·52

has seemed as if the definitions of words were remembered even when logical thinking was impaired. If the difference were due to remembering the same effect would not be expected with children. The 60-year-old paretic may retain the vocabulary ability he had when he was 20, or at least on that sub-test, and perhaps on the sub-test alone, he still scores like the average 20-year-old. But the 10-year-old child who had meningitis at 5 has in fact a 10-year-old vocabulary, but has difficulties in learning, especially when involving spatial perception and reasoning, which give the test picture we call brain damage. In other words, it seems that in many types of brain damage, as they affect children, the ability to acquire a vocabulary is retained even when the ability to reason is impaired.

That memory is not the reason for retention appears to be suggested by the clinical experience, not yet wholly supported by research, that the 10-year-old child who had meningitis at 5 has in fact a 10-year-old vocabulary, but has difficulties in learning, especially when involving spatial perception and reasoning, which give the test picture we call brain damage. In other words, it seems that in many types of brain damage, as they affect children, the ability to acquire a vocabulary is retained even when the ability to reason is impaired.

To test this theory an attempt was made to match those of the hydrocephalic children who were tested in the Revised Stanford-Binet Scale with a group of neurotic children. Further, a number was also given the Block Design Test from the Wechsler Intelligence Scale for Children.

It was possible to match 31 children. They were matched for sex, which is important in work on vocabulary (Dunsdon and Roberts, 1955; 1957) and for age and intelligence. The matching appeared to be accurate (Table 16).

Vocabulary quotients were calculated for these 62 children. The mental age equivalent of any particular vocabulary score was obtained, sometimes directly, sometimes graphically by interpolation. The quotient was calculated from this vocabulary age by dividing the chronological age in the usual way and in this way chronological age was eliminated from the comparison of the two groups.

It was expected that there would be no difference between the Binet and vocabulary quotients of the neurotics, but that there would be a difference in the

TABLE 17

COMPARISON OF MEAN VOCABULARY QUOTIENT BETWEEN HYDROCEPHALIC AND NEUROTIC CHILDREN

	Hydrocephalics	Differences	Neurotics
Mean vocabulary quotient N=31	90·00	3·9	86·10
Mean vocabulary quotient N=19	94·84	6·95 } 9·26 2·31 }	
Mean Binet quotient	87·89		
Mean Block Design quotient	85·58		

hydrocephalics. Since the Binet quotients had been made the same we were, therefore, investigating the difference between the mean vocabulary scores of the two groups.

It was expected that the vocabulary quotient of the 19 children who had done the Block Design Test would be higher than their Binet quotients and that in turn the Binet quotient would be higher than the Block Design Quotient.

All these expectations were fulfilled, but in no case was the size of the difference large enough to become statistically significant (Table 17). Thus there was a difference of 3·9 I.Q. points between the vocabulary scores of the hydrocephalics and the matched neurotics. In the same expected direction there was a difference of 6·95 I.Q. points between the vocabulary and full Binet quotients of the 19 cases who had done the Block Design Test. As was expected, these latter had average Block Design Scores 9·26 I.Q. points lower than their vocabulary scores, but even this is not statistically significant. In the 31 matching cases the hydrocephalics had 21 cases with vocabulary higher and 10 with it lower than the Binet quotient; the neurotics had 14 cases with vocabulary higher, 13 with it lower and four with it the same. Larger numbers might confirm these tentative indications which suggest that brain damaged children retain their ability to acquire a vocabulary. A vocabulary score alone may, therefore, be a misleadingly optimistic indication of such a child's educability.

These findings can be discussed in relation to the work of Walton and Black (1957) and White (1959) on the Modified Word Learning Test. Their demonstration that the ability to learn a new vocabulary is reduced in adult cases of organic brain damage suggests that the higher vocabulary scores in such cases are the result of selective memory. The failure to find the same loss of ability in brain damaged children, using these same tests, can be explained on the basis of our tentative results. Brain damaged children tend to retain the ability to acquire a vocabulary, and this ability is greater than their ability in general intelligence (in so far as the

Binet measures that quality) and even greater than their ability on a spatial reasoning test such as Block Design. We would, therefore, not expect any difference between brain damaged and normal children on a word learning test.

Twins. Eight cases are included in the series who are each one of twins. Of these, six were arrested, one was dead and one was still progressive at the close of the investigation (Table 18). In three of these both the affected and the normal twin were examined and tested.

It is proposed to study these and some of the others further and report upon them in due course. It is of interest here that in one pair the affected twin had an intelligence quotient of 125 and was being educated at a grammar school and the normal twin had an I.Q. of 135 (Fig. 17). The affected sister was unduly timid and dependent upon the normal twin and tended to have difficulties in solving her problems, which was out of keeping with her intelligence test results.

Prognosis

As already pointed out, the overall survival into adult life of the series computed on actuarial principles is between 20 and 23% (Fig. 4). This refers to all forms of hydrocephalus grouped together. As hydrocephalus is caused by various agencies, each group with its own pattern of disease, it would be of considerable interest to know the prognosis both for survival and disability when the series is broken down both according to the site of block and to aetiology. Unfortunately, when this is done the individual groups become too small for

Fig. 17.—A photograph of twins at 1 year 2 months. The twin on the left developed hydrocephalus at 3 months, was investigated at 9 months and arrested at 1 year. At 15 years both were at Grammar School; the normal twin had a skull circumference of 21 in. (53.34 cm.) and an I.Q. of 135 while the affected twin with a skull of 23½ in. (59.62 cm.) scored 125 and was physically normal. She was unexpectedly timid and dependent upon her sister.

statistical evaluation. It is possible to appreciate differences when the results are expressed graphically as in Figs. 18 and 19. When the cases, apart from the tumours, are considered according to aetiology, malformations and trauma carry the highest mortality. The former has, in addition, the highest morbidity (I.Q. of less than 85 or physical disability groups 3 and 4), because of the large proportion of cases of spina bifida cystica. A reasonable proportion of the group, when they survive, seems to do so with less disability.

TABLE 18
TWINS

Case	Sex	Obstetric	Neonatal	Affected Twin				Other Twin
				Aetiology	Age	Head Size (in.)	I.Q.	
J.B.	M	Caesarean	Resuscitation; in oxygen tent 2 weeks	Trauma	5 years	23½	20	Normal
B.M.	F	Normal	Normal; meningitis later	Infection	8 years	25½	78	100
D.G.	M	Toxaemia; post-mature birth	Resuscitation; oxygen tent 2 weeks	Trauma	3 years	24	68	80
M.S.	M	Breech and forceps	Cyanosis with convulsions	Trauma	3 years	20	20	Stillborn
D.S.	F	3 lb. 2 oz. (other twin 5 lb. 4 oz.)	Normal	Unknown	13 years	23½	125	135
P.H.	M	Breech	Normal; meningitis later	Infection	3 years	22	19	Normal
D.L.	M	Normal	Hydrocephalus noted immediately (other twin normal)	Unknown	Died at 15 years			Normal
T.C.	M	Normal	Both twins slow to suck	Unknown	Still progressive at 2 years			Normal

FIG. 18.—Prognosis regarding survival and handicap according to the site of the block (normal or near normal is I.Q. above 85 and physical disability groups 1 and 2).

Cases of infection, though they seem to have a lower mortality, have a large proportion of survivors who are physically or mentally severely handicapped. On the other hand those in whom the aetiology is unknown seem to have the best outlook both in respect to life and morbidity.

When the cases are divided according to the site of the block, excepting cases of exit foramina blocks of whom there were very few, those with aqueduct block and those with spina bifida cystica with Arnold-Chiari malformation carry the worst prognosis. On the other hand, the basal cistern blocks and those in whom the site of block was unknown have the best outlook with regard to life. Unfortunately an appreciable proportion of the survivors are handicapped.

Although the head measurement above the mean for the age when the patients first presented themselves was a little greater in those who subsequently died than in survivors, i.e. 2.55 in. (6.35 cm.) as against 2.31 in. (5.86 cm.), the scatter in the amount of head enlargement was too great for it to be of any prognostic value. The only relation of possible prognostic value that could be seen, was that all the 10 children whose heads measured more than 4.5 in. (11.43 cm.) above the mean when first seen, subsequently fell into the 'severely physically handicapped' or 'incapacitated' groups. On the other hand their I.Q. scatter was fairly even, between 101 and 104.

Discussion

A clear knowledge of the natural history of this disease has become desirable in recent years since the development of more satisfactory surgical

FIG. 19.—Prognosis regarding survival and handicap according to aetiology (normal or near normal is I.Q. above 85 and physical disability groups 1 and 2).

methods of treatment. Two operations (Pudenz, Russell, Hurd and Shelden, 1957; Spitz, 1959), which claim about a 70% chance of arresting hydrocephalus, encourage a less conservative policy in treatment, and so now may be our last opportunity of observing a series of untreated cases. A similar investigation was recently reported briefly from Sweden (Hadenius, Hagberg, Hyttén-Bensch and Sjögren, 1962). Their results are similar to our own, though they seemed to have a great preponderance of boys amongst their survivors. The publication of a detailed analysis of their cases will be most valuable.

Spontaneous arrest in a proportion of patients is now generally accepted. The assertions of Schick and Matson (1961) that natural arrest in children under the age of 2 years (in cases other than Arnold-Chiari malformation) is extremely rare, is obviously not supported by the detailed analysis of our cases. The survivors with arrest, many of over 10 years' standing, included only a few cases of malformation (see Fig. 5). Some have mental and physical disabilities which show no correlation either to the size of the head or to the degree of ventricular dilatation. Indeed, a pallium of 0.5-1 cm. in thickness is quite compatible with average intelligence and little or no physical disability. It is of interest that the great French naturalist, Cuvier, of the early nineteenth century appears to have been a hydrocephalic in childhood, and was found to have grossly dilated ventricles at autopsy (Weber, 1902). The cause of much of the oft associated disability must be sought elsewhere than simply in the reduction in thickness of the pallium. Birth trauma often sufficient to produce haemorrhage

may be accompanied by diffuse extensive parenchymal brain damage, and meningitis, especially that occurring in infancy, may be accompanied by severe encephalitis and vascular lesions (Laurence, 1959b). In addition, malformations, particularly in cases of spina bifida cystica, are responsible for severe physical disabilities quite independent of any hydrocephalus. Many cases must have suffered brain damage from the original process that led to the development of hydrocephalus. This would explain the severe disabilities found in a large proportion of the cases who have only slightly enlarged heads and a pallium of relatively normal dimensions. Reduction in the thickness of the cortex which can undoubtedly occur very rapidly (Shurtleff and Foltz, 1959) is associated with some motor changes such as spasticity and incoordination, especially when the brain is still under raised pressure. Mental impairment can generally also be shown, but this is usually not nearly as great as that caused by the original brain insult. The fact that these children's performance in school and later in life belies the expectations suggested by the standard tests must be partly due to the effect of 'brain damage'. They are also affected in some other subtle way, for they are often unexpectedly dependent on others and unadaptable and shallow in their emotions. The comparative preservation of mental faculties in cases of quite gross thinning of the pallium is accounted for by the relative sparing of the cortical grey matter at the expense of the white. A similar explanation is offered to account for the relatively normal traces on electroencephalography (Pampiglione and Laurence, 1962).

Attempt has been made to forecast the eventual head size early in the condition (O'Neill, 1961) by means of 'control lines', but this method can hardly be used to predict the outcome of a disease with so many complicating factors. The patients remain at risk even after natural arrest. Sudden death may occur without warning, or the disease may again progress to a fatal termination. A rapid downward course can also be precipitated in arrested or slowly progressive cases by ventricular or lumbar punctures; reason enough to avoid meddlesome investigation.

Few will dispute the need for immediate resolute surgical attack in rapidly progressive cases, however young the subject. It is regrettable that the preliminary report (Laurence, 1958) has been interpreted by some as a condemnation of surgery for this condition and has been used by others to justify a permanent 'wait and see' policy. The aim must obviously be to arrest the disease as early as possible, in order to prevent further brain damage.

Surgery cannot undo the brain damage that has already occurred. Some increase in the thickness of the pallium does occur during the two or three months following successful surgery (Grotte and Lundberg, 1961), but it is most unlikely that this represents an actual regeneration of damaged brain tissue. In the very slowly progressive cases which are probably on the point of arresting and in those where arrest has already occurred caution must be exercised, for surgical interference may well upset the delicate balance achieved.

It is of interest that the survival of the 57 operated cases is very similar to the unoperated group. Procedures employed before the 'Spitz-Holter valve' era may not have altered the course of the disease, but with the more satisfactory operations now available, results better than 46% survival and 17% total survival with an I.Q. above 85 should be achieved (Rickham and Roberts, 1960). Whether the operations really do much more than help the patient over the acute stage of the disease and enable him to 'arrest naturally' and in the meanwhile reduce the danger of additional brain damage, poses a question. In some cases the valve is undoubtedly required to go on functioning indefinitely. Critical investigation of this point is urgently needed.

It is the aim to continue the study of these survivors and to re-examine and retest them when the youngest is 12 years old and the majority should have reached their twenties having found their rightful place in the community. In this way it should be possible to see how much arrested hydrocephalics remain at risk and how far the conclusions of this investigation prove to be correct.

Summary

Over a 20-year period 182 unoperated cases of hydrocephalus seen by one surgeon were followed up. Of these 81 cases (46%) were found to be alive with the hydrocephalus arrested spontaneously, nine remained progressive and 89 (49%) had died. Three remained untraced but were presumed alive.

The series is representative of the disease in the community after the age of 3 months, and the life expectancy (actuarial) of the survivors into adult life is 26%.

The age of onset is dependent upon the aetiology of the condition—the natural arrest occurs between 9 months and 2½ years, but the patients remain at risk.

Of the arrested survivors 73% were educable (I.Q. more than 50) and 38% had I.Q.s within the normal range (more than 85). Little or no physical

handicap was seen in the 39 patients and only 11 had severely defective vision or were blind.

No correlation was found between intelligence, head size, cortical thickness and duration of progressive disease. A high negative correlation existed between intelligence and physical disability, and neither improved with age.

Many of the children show the classical features of brain damage which is thought to be due more to the original brain insult than to cortical thinning. Emotional disturbances were relatively few.

It is concluded that surgery should be undertaken in rapidly progressive hydrocephalus in order to prevent further brain damage, but caution must be exercised in arresting or arrested cases.

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