SYSTEMATIC VENTRICULOGRAPHIC STUDIES IN INFANTS BORN WITH MENINGOMYELOCELE AND ENCEPHALOCELE*

THE INCIDENCE AND DEVELOPMENT OF HYDROCEPHALUS

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Increasing experience and the introduction of new techniques in the treatment of congenital hydrocephalus demand its earliest possible recognition and knowledge of its natural history. By far the commonest type of congenital hydrocephalus is the one associated with a meningomyelocele or encephalocele. The presence of hydrocephalus in such infants should be suspected at birth, simply because the association is so common.

Present Investigation

An investigation was started on January 1, 1959, in the Children's Hospital, Sheffield. Its first purpose was to determine the incidence and degree of hydrocephalus in infants who were born with meningomyelocele or encephalocele, and to relate this incidence to various clinical features. The second purpose was to determine what effect, if any, early operation on the meningomyelocele would have on the development of hydrocephalus, and to determine the prognosis for those who had no hydrocephalus at the first examination. Its third purpose was to include those infants, who were found to have a hydrocephalus, in a controlled therapeutic trial. This paper deals with the results of the first two aspects of the investigations.

Methods

Infants with meningomyelocele or encephalocele were subjected to full clinical examination. The main method of investigation was ventriculography, which was often associated with dye penetration studies and sometimes with pneumoencephalography. The term 'hydrocephalus' in this paper refers to the radiological diagnosis of a pathologically distended ventricular system on ventriculography irrespective of any other feature. Ventriculo-

graphy was performed through the right side of the anterior fontanelle. The depth of the cerebral cortex was measured by using a graduated needle for ventricular puncture. The cerebrospinal fluid pressure was measured in millimetres of water, while the infant was lying placidly, with the help of a sweetened dummy. This was followed by an injection of 20-40 ml. of sterile air. Radiographs were then taken in different positions of the head, including the head down position, to outline the whole ventricular system, as far as possible. Hydrocephalus was diagnosed if this system was unequivocally dilated, the angles of the lateral ventricles were rounded off, and the margins of the ventricles were convex. If hydrocephalus was found, it was arbitrarily graded into three groups. Grade I was the mildest, with considerable residual brain tissue, whereas in Grade III the cerebral cortex was, at the most, a few millimetres thick. Grade II fell between these two extremes (Fig. 1).

Case Material

This investigation is based on the study of 172 out of 214 infants who had been born with a meningomyelocele or an encephalocele, and who were admitted to the Sheffield Children's Hospital between January 1, 1959, and November 30, 1960 (Table 1). All were under 9 months of age on admission. There were 86 boys and 86 girls. Eighty-five infants were admitted on the first day of their life, 39 were between 2 and 7 days old, 25 were up to 8 weeks of age, and the last 23 were older (Table 2). These infants were admitted under the surgical care of Mr. Zachary, whose policy was to repair the lesion as soon as possible after admission. The excised segments were examined histologically by Dr. J. L. Emery. The diagnosis of meningomyelocele is based either on the presence of paralytic lesions in the segments involved by the spina bifida cystica or the presence of neural elements in the excised sac. In a few of the older infants the meningomyelocele had already been repaired at other hospitals, before they were transferred to us for the treatment of hydrocephalus. In 58 infants the meningomyelocele or encephalocele was repaired during the first day of life, in 60 between the

* Based on a paper delivered at a Joint Meeting of the Swedish and British Paediatric Association in Uppsala, in September 1959.

FIG. 1.—Diagrammatic representations of Grades 1, 2 and 3 hydrocephalus.

TABLE 1

214 CONSECUTIVE CASES OF MENINGOMYELOCELE AND ENCEPHALOCELE UNDER 9 MONTHS OF AGE
(Admitted January 1959–November 1960)

	No. of Cases
<i>Ventriculogram not performed:</i>	
Died before or soon after operation on meningocele	30
Recent admissions, awaiting ventriculography	4
Survived, over 1 year of age, no apparent hydrocephalus	8
Total	42 (20%)
<i>Ventriculogram performed:</i>	
No hydrocephalus	30
Hydrocephalus	142
Total	172 (80%)

TABLE 2

172 CASES OF MENINGOMYELOCELE AND ENCEPHALOCELE

	Age			
	1st Day of Life	2–7 Days	1–8 Weeks	8 Weeks +
On admission	85	39	25	23
Meningomyelocele repaired*	58	60	26	18
Ventriculogram	—	4	126	42

* Ten were not operated on.

second and seventh days, in 26 during the next seven weeks, and in 18 at a later date. In 10 infants no attempt was made to repair the meningocele (Table 2).

Ventriculography was performed in 17 infants before the repair of the meningocele, in 80 within three weeks and in 75 more than three weeks after the operation (Table 3). Ventriculography was performed in 55% either before or at most three weeks after operation. Longer delays were usually due to late referral, the meningocele having been repaired elsewhere, or to post-operative illness, including meningitis, which delayed these investigations. In seven infants ventriculography was again performed on a later occasion.

Forty-two other infants were admitted into hospital during the same period with spina bifida cystica; 30 of these died shortly after admission, and were never fit for ventricular studies. Eight were discharged from hospital soon after operation on their spina bifida cystica. None of these had paralytic lesions and there were no neural elements in the excised sac. They were all seen later and did not develop clinical hydrocephalus. These infants had a simple meningocele. Ventriculography was not ethically justifiable. Finally, four recent admissions have not yet reached the stage of ventriculography. These 42 infants are not included in this study.

Results

Ventriculography showed that 30 (17%) infants had either normal or only doubtfully enlarged ventricular systems and 142 (83%) infants had hydrocephalus (Table 4). Of these the hydrocephalus was trivial (Grade 1) in six, but the large majority had more serious ventricular dilatation, this being extreme in 51 (30% of 172 cases). Table 3 shows that the ventriculograms on infants who had no hydrocephalus were performed about the same interval after the surgical repair (average 25 days) as in those with hydrocephalus (average 29 days). The latter figure does not include 16 infants whose meningocele was repaired after ventriculography, or not at all.

Head Circumference Related to Hydrocephalus.

Ventriculograms were performed on these infants irrespective of whether the size of their head was large or not at the time, and irrespective of whether there was fullness or bulging of the fontanelle, separation of the sutures or other signs of hydrocephalus. Table 5 shows that the head circumference of the 30 infants who had no hydrocephalus at the time of the ventriculography was below the 10th percentile at the time of examination in 20%, it was within the normal range* in 63% and was slightly above the 90th percentile in 17%. In contrast to this, 61% of the infants with hydro-

* For the purpose of this investigation the head circumference is regarded as normal if it falls within the 10th and 90th percentile.

TABLE 3

TIMING OF FIRST VENTRICULOGRAPH IN RELATION TO REPAIR OF MENINGOMYELOCELE

Result of Ventriculogram	All Cases	Ventriculography						Aver- age
		Before Repair		0-21 Days After Repair		22 Days or More After Repair		
		No.	%	No.	%	No.	%	
Normal Hydrocephalus	30 142	1 16	3 11	18 62	60 44	11 64	37 45	25 29*
Total	172	17	10	80	47	75	43	

* Excluding 16 infants whose meningomyelocele was not repaired.

cephalus had an enlarged and often considerably enlarged head at this stage. It is more interesting to note, however, that the head circumference was within the normal range in 48 of the 142 hydrocephalic infants (34%) and in seven babies it was even below the normal in size, although the ventriculogram showed a considerable degree of hydrocephalus (Figs 2 and 3). The same data show that of the 92 infants whose head circumference was over the 90th percentile 87 (95%) had hydrocephalus. It is important to note, however, that of the 80 whose head circumference at this stage was normal or below the normal range, 55 (69%) had ventriculographic evidence of hydrocephalus (Table 6). It is clear from these figures that although an abnormally large head almost always indicates hydrocephalus, a normal or small head in infants with meningomyelocele or encephalocele certainly does not exclude hydrocephalus which is present in almost two-thirds of such cases.

Site of Meningomyelocele and Encephalocele and Incidence of Paraplegia Related to Hydrocephalus.

One infant had an anterior encephalocele (protruding through a cleft palate), and 15 had occipital

TABLE 4

RESULTS OF VENTRICULOGRAPHY IN 172 INFANTS WITH MENINGOMYELOCELE

(January 1959–November 1960)

	No.	%
No hydrocephalus ..	30	17
Hydrocephalus { Grade 1 ..	6	3
{ Grade 2 ..	85	50
{ Grade 3 ..	51	30

TABLE 5

HEAD CIRCUMFERENCE AT TIME OF FIRST VENTRICULOGRAPH

Result of Ventriculogram	All Cases	Percentile					
		Below 10th		10th–90th		Over 90th	
		No.	%	No.	%	No.	%
No hydrocephalus ..	30	6	20	19	63	5	17
Hydrocephalus ..	142	7	5	48	34	87	61
Treated ..	61	2		22	37	37	
Control ..	54	3		21	39	30	
Advanced ..	27	2		5	19	20	

encephaloceles. The meningomyelocele was cervical in two, thoracic in 10 and sacral in 15. The lumbar segment was involved in 129 infants (75%). These included 48 thoraco-lumbar, 51 lumbar and 30 lumbo-sacral meningomyeloceles (Table 7). The site of the encephalocele or meningomyelocele

TABLE 6

HEAD CIRCUMFERENCE AT TIME OF VENTRICULOGRAPHY RELATED TO INCIDENCE OF HYDROCEPHALUS

Percentile	Total	No Hydrocephalus	Hydrocephalus
Below 10th	13	6	7
10–90th ..	67	19	48
Over 90th ..	92	5	87
			55 (69%)
			87 (95%)

TABLE 7

RELATION OF SITE OF MENINGOMYELOCELE OR ENCEPHALOCELE TO INCIDENCE OF HYDROCEPHALUS ON VENTRICULOGRAPHY

(January 1959–November 1960)

Site of Meningomyelocele or Encephalocele	All Cases		Hydrocephalus		No Hydrocephalus	
	No.	%	No.	%	No.	%
Palatal	1		—		1	
Occipital	15		11		4	
Cervical	2		—		2	
Thoracic	10		7		3	
Sacral	15		9		6	
Thoraco-lumbar	48		46	96	2	
Lumbar	51		44	86	7	
Lumbo-sacral	30		25	83	5	
All	172	100	142	83	30	17

FIG. 2a and b.—Clinical photographs of an infant whose head circumference was normal and who showed no other clinical evidence of hydrocephalus.

FIG. 2c and d.—Ventriculograms of the same infant on the same day, showing Grade 2 hydrocephalus and craniolacunia.

had an important relationship to the incidence of hydrocephalus. The incidence of hydrocephalus according to the site of the lesion was as follows: thoraco-lumbar 96% (46 of 48), lumbar 86% (44 of 51), lumbo-sacral 83% (25 out of 30), occipital 73% (11 out of 15), thoracic 70% (seven out of 10)

TABLE 8
SITE OF MENINGOMYELOCELE AND ENCEPHALOCELE
AND INCIDENCE OF PARAPLEGIA
(January 1959–November 1960)

Site	Paraplegia		No Paraplegia		Total No.
	No.	%	No.	%	
Lumbar (including thoraco-lumbar and lumbo-sacral)	108	84	21	16	129
All other sites.. ..	9	21	34	79	43
All	117	68	55	32	172

and sacral 60% (nine out of 15). When the meningomyelocele involved the lumbar region the incidence of hydrocephalus was 89%, as compared with an incidence of 63% when only other sites were involved.

Cases with lumbar involvement are not only associated with a higher incidence of hydrocephalus, but usually they also have more serious neurological defects. The incidence of paraplegia in this group was 84% (108 out of 129), as compared with 21% in those 45 whose lesion did not involve the lumbar segments. Altogether 117 out of 172 infants had some partial or complete flaccid paralysis of their lower limbs (68%) (Table 8).

There was a close relationship between paraplegia and the incidence of hydrocephalus. Of 117 infants with paraplegia 109 (93%) had an associated hydrocephalus, as compared with an incidence of only

60% among 55 infants without paraplegia (Table 9).

If these two sets of circumstances are combined (Table 10) it is seen that if an infant had a lumbar meningocele with paraplegia, the incidence of an associated hydrocephalus was 96% (104 out of 108). If there was no paraplegia, or if the meningocele did not involve the lumbar segments, then the chance of hydrocephalus was much less. Of 64 such infants 38 (60%) had hydrocephalus.

Prognosis. In this section those 115 infants will be considered who had been admitted up to and including March 1960. The survivors have been observed for a minimum of eight and a maximum of 23 months at the time of this analysis in December 1960.

TABLE 9

RELATION OF PARAPLEGIA TO INCIDENCE OF HYDROCEPHALUS ON VENTRICULOGRAPHY

(January 1959–November 1960)

		All Cases		Hydrocephalus		No Hydrocephalus	
		No.	%	No.	%	No.	%
Paraplegia		117	68	109	93	8	7
No paraplegia		55	32	33	60	22	40
Total		172	100	142	83	30	17

FIG. 3a and b.—Clinical photographs of another infant without clinical evidence of hydrocephalus.

FIG. 3c and d.—Lateral views (one in the erect and one in the upside down position) of the same infant, on the same day, showing considerable dilatation of all parts of the lateral ventricles (Grade 2 hydrocephalus and craniolacunia).

TABLE 10

INCIDENCE OF HYDROCEPHALUS RELATED TO SITE OF MENINGOMYELOCELE OR ENCEPHALOCELE AND TO PARAPLEGIA

Site	Paraplegia	All Cases (No.)	Hydrocephalus	
			No.	%
Lumbar	+	108	104	96
Lumbar	—	21	11	52
Lumbar	+	9	5	63
All other sites	—	34	22	

(a) *Infants with No Hydrocephalus.* It was of considerable importance to watch the physical and mental development of those infants who did not have hydrocephalus at the time of the ventriculography. Of the 22 followed for eight months or more, none developed clinically detectable hydrocephalus. One infant died at 8 months of age, and all the others are 1 year old or more. The head circumference at 1 year of age was slightly over the 90th percentile in three, it was normal (10th to 90th percentile) in 11 and it was only slightly or moderately below the 10th percentile in five. One

of these infants had a most extensive thoracolumbar meningocele which could be only partly repaired. Nevertheless, she has no paraplegia, and did not develop hydrocephalus. At 1 year of age her head circumference was only 17½ in. (43.5 cm.) (Fig. 4a and b). She had a normal intelligence.

Finally, three infants with occipital encephaloceles became obviously microcephalic. One of these died in another hospital at 8 months of age with bronchopneumonia. The autopsy was not detailed, but no evidence of hydrocephalus was found. The head circumference of the other two extremely retarded infants was only 16 in. (40.5 cm.) at 12 months of age. One of these has very poor vision, and the developmental quotient of both is well below 50. Only these three infants with occipital encephalocele had an intellectual defect.

It would have been of great interest to repeat the ventriculogram in all 22 of these infants to prove that they did not develop hydrocephalus. In view of their good clinical progress this would have been unjustifiable. The ventriculograms, however, were repeated in three infants whose head circumference

FIG. 4a and b.—A 12-month-old infant whose extensive thoraco-lumbar meningocele could only partly be repaired, but who had a normal ventriculogram. No hydrocephalus had developed by 18 months of age.

FIG. 5a and b.—Ventriculograms of an infant with a lumbo-sacral meningocele repaired when she was 1 day old. The ventriculograms were performed at 6 and 35 weeks of age respectively.

exceeded the 90th percentile. None of them showed hydrocephalus at the second examination. Fig. 5a and b shows the ventriculograms of such an infant with a lumbo-sacral meningocele which was repaired on the first day of life. The first ventriculogram was performed at 6 weeks, and the second at 35 weeks of age.

(b) *Infants with Hydrocephalus.* It is not within the scope of this paper to discuss in detail the outlook of the infants who had hydrocephalus. The natural history of their disease was influenced by various operative procedures, and more than half have been treated by a ventriculo-caval shunt. It is relevant, however, that 36 infants with hydrocephalus were not subjected to operative treatment immediately after ventriculography, but 11 were operated on later, because their head circumference showed a precipitous rise, well above the maximal normal range. There were only three infants in the 36 whose head circumference remained within the normal range (8%) and six whose head circum-

ference pattern rose only moderately above the 90th percentile line (16%). All these had only slight or moderate hydrocephalus on ventriculography. Most of the moderate and all the severe cases of hydrocephalus showed a gross or precipitate increase in head size, resulting in obvious clinical hydrocephalus in 25 of the 36 (70%) before they reached their first birthday (Table 11).

Discussion

Before this study there were reports of large series of cases of spina bifida cystica (meningocele and meningocele) and cranium bifidum (encephalocele) in which the site of the lesion was correlated with the incidence of hydrocephalus and in which the diagnosis of hydrocephalus was made either on clinical grounds or on autopsy evidence. The development of hydrocephalus either as a sequel to the operation on the meningocele or independent of such procedures has also been discussed at length.

TABLE 11
HEAD CIRCUMFERENCE PATTERN OF INFANTS AFTER VENTRICULOGRAPHY

	Total	Microcephalic	1st-9th Percentile	Normal Range 10th-90th Percentile	95th Percentile	Grossly Enlarged	Died Early
Infants with no hydrocephalus	22	3	5	11	3	—	—
Infants with hydrocephalus who were not operated upon	36	—	—	3	6	25	2
Grade 1	2	—	—	1	1	—	—
Grade 2	26	—	—	2	5	17	2
Grade 3	8	—	—	—	—	8	—

Ingraham and Matson (1954) studied 1,433 cases of spina bifida cystica and cranium bifidum in the course of some 20 years. Of these, 187 were cases of encephalocele, 396 were considered to be simple meningoceles and 754 were meningocele. The proportion of simple meningoceles (in which the sac does not contain neural elements) was much higher than in the present series. The very low incidence of meningoceles in our material corresponds well with the series of Laurence (1960) who found only 9% of cases of meningocele in a series of 407 cases of spina bifida cystica seen in London.

In this series the incidence of hydrocephalus, as shown by ventriculograms, was closely linked with the site of the meningocele. This observation is similar to that of Laurence, who estimated that the overall incidence of hydrocephalus in his series was 62% (based presumably on clinical signs and autopsy evidence). He too found that the incidence of hydrocephalus was highest when the meningocele involved the lumbar (80%) or thoracolumbar region (70%) and less common in purely sacral (50%) or thoracic (43%) lesions. Ingraham and Matson (1954) considered that internal hydrocephalus was probably present in some degree in almost all patients with the Arnold Chiari malformation and that the hydrocephalus was of the communicating variety.

There has been no conclusive evidence so far to show whether the hydrocephalus is an essential part of the developmental disorder, and is present at birth, or whether the hydrocephalus develops later during the first year of life possibly as a result of operative interference or of meningitis resulting from an ascending infection.

Macnab (1958), discussing 200 cases (a part of the 407 reported by Laurence in 1960), found that 32% showed evidence of hydrocephalus at birth, in 16% the onset was between the 2nd and 3rd months and in 7% progressive hydrocephalus only became evident after about one year of observation. The criteria of the diagnosis of hydrocephalus were not stated, and it seems that the diagnosis was based on clinical evidence. Schaffer (1960) stated that many meningoceles were associated with hydrocephalus at the onset, while in some infants hydrocephalus made its appearance early and progressed speedily.

Our findings indicate that hydrocephalus is frequently present in those infants whose head is not enlarged at the time and who show none of the other clinical signs which are accepted as evidence of hydrocephalus. We found this in 55 (39%) out of 142 infants with hydrocephalus. A similar obser-

vation on a smaller scale was made 20 years ago by Shapiro and Tosti (1940) who found ventriculographic evidence of hydrocephalus in six consecutive infants before the operation on their meningoceles and whose head circumferences were normal at the time of the examination.

The ventriculograms in this series were performed on very young infants, either before or very soon after the repair of the meningocele or encephalocele. No hydrocephalus developed subsequently in any infant who showed no hydrocephalus at this examination, in spite of the repair of their meningocele or encephalocele. It has been shown that hydrocephalus, therefore, is an integral part of the clinical syndrome and is probably present right from the beginning in all those infants who subsequently develop clinical signs of the condition. In a very few cases the hydrocephalus remains subclinical throughout, but in the large majority this becomes manifest within the first two months of life. In view of these observations it is hardly necessary to discuss the mechanism of the development of the hydrocephalus following operative removal of the meningocele. Nevertheless, Ingraham and Matson (1954) may well be right in their judgment that in some cases of meningocele the associated Arnold-Chiari malformation in which the cerebrospinal fluid circulation is impaired only to a minor degree, may be disturbed by removal of the meningocele. The operation may precipitate a more severe degree of obstruction of the cerebrospinal fluid's pathway and this may be followed by acute or slowly progressive hydrocephalus. They consider that this is the cause of the hydrocephalus after surgery, and not the decrease in the available absorptive surface, resulting from the removal of the sac. I would like to modify this view by suggesting that even in these cases the hydrocephalus was already present at the time of the removal of the sac, but that its progression to clinical hydrocephalus might have been accelerated.

It is possible, of course, for hydrocephalus to develop in those infants who did not have hydrocephalus before, or whose hydrocephalus was of trivial degree, if they should develop a pyogenic meningitis. This unfortunately is a common complication of meningocele and may well occur in the mildest cases with disastrous results. In Macnab's (1958) series of 200 cases the incidence of meningitis was 18%. The meningitis usually occurs very early, and is often fatal. In this study, ventriculograms were either never performed in these infants or were done only after recovery from the meningitis, by which time evidence of gross hydrocephalus was usually available. Pyogenic

meningitis will, of course, also aggravate existing hydrocephalus. This has been shown in several infants in this series.

Summary and Conclusions

The following conclusions can be drawn from this investigation: (1) There is an 80% incidence of hydrocephalus in association with meningo-myelocele and encephalocele in infants. This hydrocephalus can usually be detected in the first few days or weeks of life, even if the size of the head is not larger than normal, and other signs of increased intracranial pressure are not present. (2) The incidence of hydrocephalus is far higher in infants whose meningo-myelocele involves the lumbar region, than when it involves other sites, and is far higher if there is associated paraplegia; 96% of infants with paraplegia and a lumbar meningo-myelocele, and 95% of infants whose head circumference was over the 90th percentile in the first few weeks of life had radiologically demonstrable hydrocephalus. (3) There is no evidence that repair of the meningo-myelocele in the early neonatal period influences the development of hydrocephalus. No infant

who did not have hydrocephalus at the first examination developed clinical or other evidence of hydrocephalus during a period of follow-up lasting up to 23 months. (4) It is desirable to perform routine ventriculography in babies with meningo-myelocele as soon as possible after birth. This is of great prognostic value and is useful as a guide to treatment.

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