THE NATURAL HISTORY OF SPINA BIFIDA CYSTICA

DETAILED ANALYSIS OF 407 CASES

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Although spina bifida cystica has been accurately documented as far back as the 17th century and was well known to physicians many centuries before that, little real interest was taken in this malformation until the latter end of the last century, when the Clinical Society of London published a report on its natural course and some aspects of its treatment (Marsh, Gould, Clutton and Parker, 1885). With the gradual elimination of the infective and nutritional diseases, malformations in general and spina bifida cystica in particular have assumed greater importance in recent years. The latter account for about 1 in 430 births in England and Wales as a whole (Nash, 1963), but as many as 1 in 200 births in some areas (Laurence and David, 1963). Most of the more recently published series of cases of spina bifida cystica have either been based on small numbers (Harnack and Kirsten, 1959; Klein, Delègue and Engel, 1960; Mletzko, 1960; Regenstreif, 1961), or have been approached from a largely surgical point of view (Ingraham, Swan, Hamlin, Lowrey, Matson and Scott, 1944; Macnab, 1957; Klug and Tzonos, 1961; Wilson and Llewellyn, 1962; Guthkelch, 1962; Sharrard, Zachary, Lorber and Bruce, 1963). No real attempt seems to have been made to evaluate the natural history of the disease, though Norman (1949) followed up and reviewed all the cases seen at The Hospital for Sick Children, Great Ormond Street, London, from 1938 to 1947, that is in the years before antibiotics were in use generally, in order to get some idea of the natural course.

The present paper is a detailed account of the examination and follow-up of the infants with spina bifida cystica and cranium bifidum cysticum, born in the years 1947 to 1956. Brief summaries of some of the results have already been published elsewhere (Laurence, 1959c, 1960). An attempt has been made here to analyse this series in a similar

way to that used for an investigation on the natural history of hydrocephalus (Laurence, 1958b; Laurence and Coates, 1962).

To define terms, it is necessary to review briefly the embryology and pathology of spina bifida.

Embryology and Pathology

The central nervous system develops from the medullary ridge posteriorly. This infolds to form a tube, beginning in the mid-dorsal region, extending cranially and caudally and is complete by the end of the fourth week. The last parts to close at either end, the neuropores, disappear by the end of the fourth week (Hamilton, Boyd and Mossman, 1952). In the meantime, the mesoderm completely surrounds the neural tube and separates the neuroectoderm from the ectoderm. The mesoderm segments into somites which develop into the vertebral column. The meninges are formed from the mesenchyme at the time of the separation of the neural tube from the ectoderm.

In all forms of spina bifida, some part of this process is abnormal (Fig. 1). Spina bifida occulta, which is the least serious, is an abnormality of the mesodermal elements with non-fusion of the laminae, the spinal cord and the meninges remaining in their normal position. In some cases, however, both ectoderm and neuroderm participate in the anomaly as is evidenced by skin malformations and the occasional cord abnormalities that are encountered.

Spina bifida cystica is a generic term which also includes relatively flat lesions. Here the abnormality is more serious and may be conveniently subdivided into meningoceles and myeloceles. In meningoceles, in addition to the spina bifida, the meninges, both dura and arachnoid, herniate out of the spinal canal with the cord remaining in its normal position. The meninges may be exposed

FIG. 1.—Diagrammatic representation of the normal anatomy of the spinal column and of spina bifida.

on the surface as a thin membranous sac, or they may be covered by full-thickness skin and sometimes a lipoma. The surrounding skin may show malformations. There are quite often associated anomalies of the spinal cord and occasionally of the brain.

Myelomeningoceles, myelocystoceles, hydro-myelocoeles, syringomyelocoeles, meningo-myelocoeles and localized rachischisis, are all essentially the same lesion. They all have the same embryological basis; little benefit can therefore be derived from this subdivision, and they may thus be regarded as myelocoeles. In these lesions, in addition to the bony deformity, the neural plate has failed to close over locally to form a tube, and the cord tissue, often with some degenerative changes, lies exposed on the surfaces. At birth, this is generally surrounded by a bluish, semi-transparent membrane representing the meninges. Nerve roots, both sensory and motor, leave the ventral surface of the neural plate and traverse the sponge-like 'sub-arachnoid' space to gain the root canals. The myelocele is often flat at birth, only later becoming cystic with the accumulation of fluid. Cranial to the lesion, the spinal cord often shows abnormalities such as hydromyelia, diastematomyelia, with or without a fibrous, cartilaginous or bony spur arising from one of the affected vertebral bodies, or shows cysts, syringomyelia or gliosis. There may be associated lipomata, dermoids or teratomata. Below the lesion, when this is situated high-up on the spinal axis, the spinal cord may re-form into a relatively normal structure.

On occasion, even without active treatment, the myelocele epithelializes over from the surrounding skin without much inflammatory fibrosis (Cameron,

1956). More usually, the raw neural plate becomes infected with the ensuing fibrosis and scarring that leads to the typical scarred, thickened, fundus seen in the older children (Laurence, 1963). This generally results in much damage to the spinal cord tissue. Damage may also be caused to the cord tissue by stretching from the accumulation of fluid or sometimes of pus in the subarachnoid space beneath it, which generally occurs soon after birth (Nash, 1963) and which leads to the typical cystic lesion. Chemical changes resulting from drying or overhydration or the use of various substances to prevent infection may also cause damage. The three factors of infection, stretching and biochemical alteration, probably account for the deterioration in the neurological state.

Subjects and Methods

This investigation was based on 426 cases of spina bifida cystica, born during the 10 years between January 1, 1947 and December 31, 1956, seen by the paediatricians and surgeons at The Hospital for Sick Children, Great Ormond Street, and the Westminster Children's Hospital, Vincent Square, London, before December 31, 1957 (Table 1). Cases of spina bifida occulta were specifically excluded and also four stillborn infants as these would not be of interest in this connexion. Seven

TABLE 1
THE SERIES—426 CASES

Cases	No.
Cases Included:	407
Meningocele	39
Myelocele	368
Cases Excluded:	19
Stillbirths	4
Not traced	8
Insufficient data	7
Total	426

others had to be discarded as the hospital notes were insufficiently complete to be of value, there being either an unsatisfactory history or no record of the neurological or pathological findings. Eight further cases, not traced during the follow-up, were also excluded. This left altogether 407 cases for analysis, which included about 80 cases analysed from a different viewpoint by Macnab (1957), 14 reported by Forrest, Laurence and Macnab (1957) in connexion with ventriculo-subdural shunt operations and 17 investigated by the author (Laurence, 1958b; Laurence and Coates, 1962). Most of the cases were drawn from London and the Home Counties, though a few came from the remainder of the British Isles and a very few came from overseas. The majority of the patients were seen in early infancy because of the spinal lesion, thus 35% attended within one month of birth and altogether 63% by 3 months (Table 2). Many

TABLE 2
AGE AT FIRST ATTENDANCE
(407 Cases)

Age	No. of Cases	Percentage	
1 day	35	35	65
-1 wk.	26		
-4 wks.	80		
-2 mths.	71		
-3 mths.	45		
-4 mths.	19	21	
-5 mths.	17		
-6 mths.	21		
-8 mths.	24		
-11 mths.	13		
-18 mths.	21	14	
-24 mths.	10		
-3 yrs.	12		
-4 yrs.	9		
-5 yrs.	3		
>5 yrs.	1		

of the remainder were seen at a later age, usually because of one of the complications, but only 14% were over 1 year old. The increasing number of cases seen each year towards the end of the investigation reflects the growing interest that is being taken in the serious malformations in general and spina bifida cystica in particular (Fig. 2).

This group of spina bifida cystica is representative of the cases seen at any large children's hospital to which cases are referred by outside paediatricians or from other hospitals. It is not representative of the condition in the community as a whole as the severest cases, often considered beyond help, are not referred, or die before they can be admitted. A comparison of the deaths for each age-group, reported by the Registrar-General (General Register Office, 1949-1957), and the deaths found in the series show that the progressive ratio of deaths for each month of life becomes comparable only after the age of 3 months, thus confirming that most of the severest cases never reached the two hospitals (Table 3 and Fig. 3). From the similarity of the two

FIG. 2.—Year of first attendance for the 407 cases of spina bifida cystica.

TABLE 3
COMPARISON OF HOSPITAL AND REGISTRAR-GENERAL'S
DEATH STATISTICS FROM SPINA BIFIDA FOR 1947-1956
INCLUSIVE

Age	Number of Deaths		Ratio of Hospital to Registrar-General
	Hospital	Registrar-General	
Mths.			
0-1	11	5,755	1 : 523
1-2	33	1,581	1 : 48
3-5	45	796	1 : 18
6-8	25	353*	1 : 14
9-11	23	223*	1 : 10
Yrs.			
1-2	30	243	1 : 8
2-3	13	92	1 : 7
4-9	5	66	1 : 12
10-15	0	34	—
Total	185	9,143	

* Estimate only, as for 1947-1950 inclusive only 6-11 months are published by the Registrar-General.

curves after 3 months it can be inferred that the series then becomes representative of the disease in the community, and it represents about one-twelfth of all the cases of spina bifida cystica in England and Wales surviving beyond the age of 3 months in the 10 years concerned. A rather greater proportion of the cases was probably seen towards the end of the period.

FIG. 3.—A comparison of the Hospital and Registrar-General's death records for the years 1947 to 1956 inclusive. The deaths have been expressed as deaths occurring per month and the hospital deaths have been multiplied ten times. Logarithmic scales have been used.

FIG. 4.—The position of the lesion in 407 cases of spina bifida cystica. Cr, cranial; Ce, cervical; D, dorsal; DL, dorsi-lumbar; L, lumbar; LS, lumbosacral; S, sacral.

The site of the lesion was similar to that reported in most other investigations, with just half of all the cases having the lesion in the lumbar or lumbosacral region (Fig. 4). There were 61 cases with the very large dorsi-lumbar lesions, all myeloceles, and 33 cases with cranium bifidum cysticum. Five cases of cervical spina bifida cystica were in fact encephaloceles in that they contained brain tissue or part of the ventricular system and should really have been considered as such. Thirty-nine meningoceles were included in the series, giving an incidence of about one meningocele to every 10 myeloceles, which is rather lower than is usually reported. The distribution of these meningoceles is, however, rather different: 31% of the latter were situated in the cranial or cervical regions, while only 9% of the myeloceles were in this part of the cerebrospinal axis.

The clinical notes, which were usually full and systematic, were scrutinized, paying special attention to the neurological state at birth and subsequent development, operation details, the onset and progress of clinical hydrocephalus and the appearance of other complications. Where the spinal lesion was operated upon, the pathological material or the histological sections were, whenever possible, examined personally, but in some cases reports only had to suffice.

No attempt has been made to investigate family incidence of spina bifida cystica and other malformations.

It is the author's experience that unless this family history is specifically and extensively asked about after previous warning, so that the parents can make their own inquiries from their relatives, it is unlikely that full information will be obtained. Any conclusions drawn from incomplete data would be misleading.

The 407 cases were followed up and traced during the spring and early summer of 1958, using the methods described in the *Lancet* (Laurence, 1959a). For statistical purposes, it was assumed that the date of this follow-up was June 30, 1958, and thus the youngest surviving child was 1½ years and the eldest just 11½ years of age. The majority of the survivors were re-examined by the author personally, or by the physicians and surgeons at one of the two hospitals. In the remaining patients, most of whom were living too far away or could not be moved, detailed reports about their condition from the local clinicians were thought sufficient.

Results

Survival. Of the 407 cases of the whole series, 222 children, including all those with a meningocele, have survived (Table 4). This suggests a survival

TABLE 4
SURVIVAL: 407 CASES

	Alive	Dead	Total
Meningoceles	39	0	39
Myeloceles	183	185	368
Total	222 (55%)	185 (45%)	407

of 55%. Most hospital statistics are indeed presented in this way, which gives quite a false impression of the prognosis. When account is taken of the number of actual patients in the series at risk at any one moment with the life expectancy computed on actuarial principles, using the life table (Table 5) and a life chart (Fig. 5), a very different result is obtained; it is found that just over 35% of the sort of cases that reach a large children's hospital survive until the age of 12 years. Bearing in mind that for the first three months this series of cases is not representative of the condition in the community, it is possible to forecast the survival of a case who has already survived beyond three months. Thus, it would be reasonable to predict that a 4-month-old child has a 51% chance and a 1-year-old child a 77% chance of surviving to the age of 12 years.

Meningoceles. The series includes 39 cases of meningocele (20 male and 19 female). The diagnosis of this lesion is difficult on purely clinical

TABLE 5
LIFE TABLE OF SPINA BIFIDA CYSTICA
(407 cases)

Age (mths.)	Deaths	No. of Cases at Risk*	Deaths/1,000/mth.	Survivors of Original 100 (Life Table)	Chance of Survival Up to 12 Years
0-	11	85.1	129	100.00	0.35
1-	12	159.9	75	87.07	0.41
2-	21	201.4	104	80.54	0.44
3-	11	217.1	51	72.14	0.49
4-	13	223.1	58	68.48	0.52
5-	21	225.2	93	64.49	0.55
6-	12	225.5	53	58.48	0.60
7-	6	227.0	26	55.36	0.64
8-	7	226.5	31	53.90	0.66
9-	9	224.0	40	52.24	0.68
10-	10	220.5	45	50.14	0.71
11-	4	216.0	19	47.86	0.74
12-	6	212.5	28	46.98	0.75
13-	4	212.0	19	45.65	0.77
14-	0	214.5	0	44.79	0.79
15-	3	215.5	14	44.79	0.79
16-	4	214.5	19	44.17	0.80
17-	2	216.0	9	43.34	0.82
18-	5	214.5	23	42.94	0.82
19-	0	211.0	0	41.94	0.84
20-	1	210.0	5	41.44	0.84
21-	2	209.0	10	41.74	0.85
22-	1	207.5	5	41.34	0.86
23-	2	203.0	10	41.14	0.86
24-47	13	168.7	77	40.74	0.87
48-119	5	83.8	60	37.60	0.94
120 and over	0	15.5	0	35.35	1.00
				35.35	

* The number of cases at risk is calculated assuming that the intake of recruits, the deaths and those leaving are all distributed uniformly in each of the age intervals.

grounds and many more were thought to have been meningoceles until operation or even histology revealed the lesion to be a myelocele. Of the cases, 22 were examples of fluctuant swelling covered with full-thickness skin and four of these were associated with a lipoma as well, making the 'sac' appear larger than it actually was. Only 17 presented as the typical thin-walled membranous, transilluminable lesion. These membranous lesions tended to be situated in the upper part of the neural axis with 11 cranial, cervical or dorsal in position, while the other type was more evenly distributed.

All the 39 cases had survived and 24 showed no neurological abnormality. Four more had minor disabilities such as slight equinovarus (two), congenital ptosis (one) or slight imbalance (one). Six were slightly or moderately incapacitated, with a bad gait (four), precipitous micturition (one) or partial paralysis of one hand (one).

In six cases, all with cranial cervical or upper dorsal meningoceles, hydrocephalus was present, but this had arrested spontaneously. In four this was minimal or very mild, in one it was moderate and in only one was it severe: in both the latter

FIG. 5.—Life chart of 407 cases prepared from data on the life tables. The chart can be used to compute the life expectancy of an 'average' case of spina bifida at any age between 3 months and 12 years (see text).

two it was due to an aqueduct stenosis. It is of interest that 11 of the 17 cases of the thin-walled meningoceles had hydrocephalus or physical disabilities, while only five of the 22 with the lesions covered with normal skin had these complications which were all mild.

Myeloceles. The series included 368 myeloceles, of whom 163 were male and 205 female, giving a sex ratio of about 1 to 1.26.

SURVIVAL. 183 patients survived up to, or beyond, the end of June 1958, giving a crude survival of almost 50% of the myeloceles (Table 4), with a sex ratio of the survivors of about 1 to 1.08, male to female. The ratio of male to female deaths is 1 to 1.15 and is similar to the 1 to 1.14 recorded by the Registrar-General for the years 1947 to 1956 inclusive. Although the sex ratios of the survivors and those who died seem to be very different, this difference appears not to be statistically significant.

A life table for the myeloceles shows that 29% of the sort of cases that are referred to a large children's hospital survive to the age of 12 years, instead of 35.4% of the whole series (Table 6). Thus, the chances of a 6-month-old infant with a myelocele reaching 12 years of age are 54%, instead of 60% for the whole series. However, most of the features of myeloceles, such as the type and extent of paralysis, the incidence of hydrocephalus and the chances of survival, are determined to a large extent

by position of the lesion. Although superficially the mortality appears to vary considerably according to the site of the lesion, the only groups that are at all statistically different from the whole series are those where the myelocele is situated in the dorsi-lumbar and in the sacral region (Fig. 6). The dorsi-lumbar myeloceles, which are generally very extensive lesions and tend to leak or burst, carry a poor prognosis, this being significant at the 1%

FIG. 6.—Mortality of the 368 encephaloceles and myeloceles at the various sites along the spinal axis.

Cr, cranial; Ce, cervical; D, dorsal; DL, dorsi-lumbar; L, lumbar; LS, lumbosacral; S, sacral.

TABLE 6
LIFE TABLE OF MYELOCELES ONLY: 368 Cases

Age (mths.)	Deaths	No. of Cases at Risk*	Deaths/1,000/mth.	Survivors of Original 100 (Life Table)	Chance of Survival Up to 12 Years
0-	11	76.5	144	100.00	0.29
1-	12	141.4	85	85.63	0.34
2-	21	177.4	118	78.36	0.37
3-	11	191.0	58	69.09	0.43
4-	13	195.0	67	65.20	0.45
5-	21	194.7	108	60.77	0.48
6-	12	192.6	62	54.21	0.54
7-	6	193.0	31	50.84	0.58
8-	7	192.5	36	49.26	0.60
9-	9	189.0	48	47.46	0.61
10-	10	184.5	54	45.20	0.65
11-	4	180.0	22	42.76	0.69
12-	6	176.5	34	41.81	0.70
13-	4	175.5	23	90.38	0.73
14-	0	177.5	0	39.46	0.74
15-	3	178.5	17	39.46	0.74
16-	4	177.5	23	38.80	0.76
17-	2	179.0	11	37.93	0.77
18-	5	177.5	28	37.50	0.78
19-	0	174.0	0	36.45	0.81
20-	1	173.0	6	36.45	0.81
21-	2	173.0	12	36.24	0.81
22-	1	169.5	6	35.82	0.82
23-	2	166.0	12	35.60	0.83
24-47	13	135.7	4	35.18	0.84
48-119	5	65.8	1	31.81	0.92
Over 120	0	12.0	0	29.39	1.0000
				29.39	

* The number of cases at risk is calculated assuming that the intake of recruits, the deaths and those leaving are all distributed uniformly in each of the age intervals.

level. On the other hand, the sacral lesions have a relatively good prognosis, probably because in many the sacs are covered with whole-thickness normal skin. The p value here is 0.05. Indeed, all 20 cases with sacral and the 10 with lumbosacral lesions where the skin was normal had survived. This is partly because these patients are not prone to ascending meningitis and fewer are born with or develop hydrocephalus. The lumbosacral and sacral lesions between them have a significantly better prognosis ($p = 0.02$).

PARALYSIS. A detailed discussion of the neurological features of spina bifida cystica is beyond

the scope of this paper and paralysis will be discussed in general terms only.

The cases were graded according to the severity of the sphincter or skeletal muscle paralysis. In the moderate group, the sphincter control was generally achieved, though sometimes with difficulty and often rather late, and locomotion was possible without undue assistance. The severe cases generally suffered permanent sphincter paralysis and were unable to walk unaided or only with the greatest difficulty. No grading was possible in respect to loss of sensation, as this was generally insufficiently recorded or determined.

FIG. 7.—The incidence and severity of muscular and sphincter paralysis in the 368 encephaloceles and myeloceles in the various positions along the cerebrospinal axis. For both sphincter and skeletal muscle paralysis the chart shows heterogeneity between the various groups of very high significance. ($\chi^2_{12} = 91.6$ and $\chi^2_{12} = 88.8$ respectively.) Cr, cranial; Ce, cervical; D, dorsal; DL, dorsi-lumbar; L, lumbar; LS, lumbosacral; S, sacral.

The pattern of skeletal muscle and sphincter paralysis and of sensory loss depends on the level and the extent of the lesion (Sharrard, 1962). Thus, only one-third of encephaloceles and cervical myeloceles were associated with skeletal muscle involvement, and then only in the upper limbs, and then not severe. Only one of these cases, one with a lesion in the lower cervical region, had any sphincter paralysis at all. The dorsal, lumbar and lumbosacral lesions frequently had severe skeletal muscle involvement, while the purely sacral lesions

had a somewhat lower incidence. On the other hand, the lower the lesion on the spinal axis, the more prevalent became the sphincter involvement, until 90% of the cases with sacral lesions had some paralysis, 65% of them severe. There is striking statistical heterogeneity between the various groups (muscle paralysis $\chi^2 = 88.8$; sphincter paralysis $\chi^2 = 91.6$). Where records were sufficiently accurate, the level of anaesthesia correlated accurately with the level of the lesion (Fig. 7).

Only in lesions confined to the sacral regions was severe sphincter paralysis seen in the absence of any skeletal muscle involvement, while a very few cases with lesions elsewhere in the lower parts of the spinal axis escaped sphincter paralysis if they had also severe skeletal muscle involvement (Table 7). Apart from the cranial and cervical lesions, approximately a similar proportion of cases had both severe skeletal and sphincter paralysis.

Associated with skeletal muscle paralysis, paralytic congenital dislocation of the hip and talipes calcaneovarus, amongst others, were frequent. Various degrees of genu recurvatum, arthrogryposis and flexion deformities were also encountered. The sensory paralysis led to severe trophic ulceration in a number of patients, and vasomotor disturbances of the skin were frequent.

HYDROCEPHALUS. The hydrocephalus associated with spina bifida may be caused by a malformation of the brain, by an acquired lesion or by the one superimposed upon the other (Laurence, 1959b). Nearly all myeloceles are associated with some form of Arnold-Chiari malformation, except for a few of those where the spinal lesion is situated in the sacral region (Laurence, 1963). The Arnold-Chiari malformation does not, in itself, necessarily cause hydrocephalus, though plugging of the foramen magnum or blockage of the fourth ventricle exit foramina is sometimes offered in explanation. Severe overcrowding in the poorly developed posterior fossa of the skull may cause at least partial obliteration of the basal cisterns and may lead to

TABLE 7
NEUROLOGICAL INVOLVEMENT: 368 MYELOCELES

Type of Involvement	Percentage Affected				
	Site of Lesion				
	Cranial Cervical	Lumbo- sacral	Lumbar	Dorsi-lumbar Dorsal	Sacral
Severe muscle paralysis with normal sphincters ..	15	7	18	24	4
Severe sphincter paralysis with normal muscles ..	0	0	0	0	15
Severe muscle and sphincter paralysis ..	3	45	47	47	41
No muscle or sphincter paralysis ..	62	4	5	12	6
Severe muscle and sphincter paralysis with hydrocephalus	0	32	40	47	22
Total number of cases in each group ..	35	81	123	80	49

TABLE 8
HYDROCEPHALUS: 368 MYELOCELES

Group	Degree of Hydrocephalus	Alive	Dead	Total
1	Absent	97	3	100
2	Minimal (cortex > 3 cm.)	21	12	33
3	Moderate (cortex 1½–3 cm.)	32	48	80
4	Severe (cortex < 1½ cm.)	33	122	155

some hydrocephalus. There is also considerable congestion of the meninges, which eventually causes fibrosis, thus aggravating any blockage. Aqueduct malformations, in the form of forking, or more rarely congenital stenosis, are seen in about one-quarter to one-third of cases of myelocele at autopsy (Alvord, 1961). The Arnold-Chiari malformation and the aqueduct anomalies between them probably account for the hydrocephalus seen at birth in so many of these cases. Acquired post-inflammatory lesions resulting from ascending meningitis from the exposed medullary plate, are frequently superimposed upon the malformations, thus aggravating what may already be an embarrassed cerebrospinal fluid circulation. The post-inflammatory blocks of the foramina of Monro, the aqueduct of Sylvius and the exit foramina of the fourth ventricle, usually follow the grossest pyogenic infections and are often also associated with severe parenchymal brain damage. Milder infections, which are common and often recurrent in nature, are generally not associated with severe parenchymal brain damage and tend to result in basal cistern blocks and, less often, in acquired aqueduct stenosis, and sometimes both.

The cases were divided into four groups; Group 1, where hydrocephalus was clinically or radiologically absent, and Groups 2, 3 and 4 according to the severity of the disease. Group 2 included those in whom there was apparently only minimal hydrocephalus, either clinically or radiologically, with rounding off of the edges or convexity of the margins of the lateral ventricles and where the 'cortex' was more than 3 cm. thick in the superior parietal region. Group 3 included cases with moderate hydrocephalus clinically and radiologically or at autopsy, with the 'cortex' between 3 and 1.5 cm. in thickness, while in Group 4, the condition was severe, with a 'cortex' of less than 1.5 cm. in thickness and often measuring only a few millimetres.

In 155 cases the condition was advanced, and in a further 80, moderately severe (Table 8). Only 21% of the former and 40% of the latter were

TABLE 9
DIAGNOSIS OF HYDROCEPHALUS: 368 MYELOCELES

	Alive	Dead	Total
<i>Diagnosis Confirmed:</i>			
Contrast radiography	48	59	107
Ventricular puncture	14	9	23
Autopsy only	—	66	66
	62	134	196
<i>Diagnosis Not Confirmed:</i>			
No hydrocephalus	86	2	88
Hydrocephalus present—mild ..	15	4	19
moderate ..	15	17	32
severe ..	5	28	33
	121	51	172

alive at the time of the follow-up. In 33 the condition was mild with 64% alive, and in 100 there was apparently no hydrocephalus and 97% of these had survived. Lorber (1961) has shown, however, that the purely clinical evaluation of the degree of hydrocephalus associated with spina bifida cystica is extremely difficult. Considerable hydrocephalus may be present with a normal-sized or even a small skull. Although clinical signs are usually present in progressive hydrocephalus, these may be absent if it is arrested or only slightly progressive. Therefore, of the 172 cases where the hydrocephalus was not confirmed by special investigations or autopsy, only in the 65 cases where the hydrocephalus was thought to be moderate or severe is there any doubt about the condition (Table 9). It is likely, however, that the incidence and severity in the other 107 is greater and the onset earlier than is suggested by the charts. 64% of all the myeloceles had hydrocephalus of Groups 3 and 4 severity, but altogether at least 73% had some hydrocephalus. These figures do not take into account the incidence or severity of hydrocephalus with lesions in the various positions along the cerebrospinal axis. Although encephaloceles were associated with hydrocephalus at more than the average frequency, this appeared to be often less severe than in lesions lower down (Fig. 8). The extensive lesions in the dorsi-lumbar region had the highest incidence of moderate or severe hydrocephalus (89%), while in the purely dorsal myeloceles, associated hydrocephalus was somewhat less frequent but when present it was severe. On the other hand, cases with the lesions involving the sacral spine commonly escaped hydrocephalus, with only about 40% of those with the lesion confined to the sacral spine having Groups 3 and 4 hydrocephalus. There is marked heterogeneity of degree and incidence of hydrocephalus between the various groups. This appears to be highly significant ($\chi^2 = 55.9$).

FIG. 8.—The incidence and degree of hydrocephalus in 368 encephaloceles and myeloceles in the various positions along the spinal axis. The chart shows heterogeneity between the various groups of high significance. ($\chi^2_8 = 55.9$.) Cr, cranial; Ce, cervical; D, dorsal; DL, dorsi-lumbar; L, lumbar; LS, lumbosacral; S, sacral.

The remaining comments about hydrocephalus will concern only those with moderate or severe ventricular dilatation (Groups 3 and 4), since, in the 33 cases of Group 2, the mild hydrocephalus was not a major factor in the disease.

Hydrocephalus was clinically present or suspected at birth in 64 cases, and in a further 78 it became obvious during the first month (Fig. 9). In many

FIG. 9.—Age of onset of hydrocephalus in 235 encephaloceles and myeloceles who had either moderate or severe hydrocephalus. In 23 the age of onset was not known precisely.

of these 78 and in some where hydrocephalus manifested itself subsequently, it is likely that ventricular dilatation was also present at birth. In only 15 cases was the clinical onset after the third month and in only one after the sixth month. This is what might be anticipated if the hydrocephalus is due to a malformation or to ascending infection from the myelocele, as the latter generally epithelializes by the fourth month, thus, as a rule, excluding infection. The sac, however, remains liable to injury.

The 53 operations for the relief of hydrocephalus, all carried out before the introduction of the more satisfactory ventriculo-cardiac shunts with Holter and Pudenz-Hayer valves, were performed by surgeons with widely differing approaches using 11 different operations, none of which proved really effective in this series. The largest group of the children (21 cases) had ventriculo-subdural anastomoses (Forrest *et al.*, 1957), but 10 had spinoureteric anastomoses (Matson, 1953). Seven had ventriculo-pharyngeostomies (Nosik, 1950), five had ventriculo-peritoneal shunts (Scott, Wycis, Murtagh and Reyes, 1955) and the remaining 10 had a variety of procedures (Table 10). In one, the disease had become arrested, apparently as the result of a ventriculo-subdural operation, and in two others, where the hydrocephalus was arrested following a ventriculo-peritoneal and a ventriculo-subdural operation, death occurred subsequently from uraemia and ascending meningitis respectively; 38 other operated cases died with the hydrocephalus still progressive. Of the 13 patients who were still alive at the end of June 1958, five had still progressive hydrocephalus; in one already referred to, it arrested following operation; and in seven the hydrocephalus became arrested spontaneously later.

Altogether only 65 with Groups 3 and 4 hydrocephalus were still alive at the follow-up. In 18, including the five who have had operative attacks upon the hydrocephalus, the condition was still progressive but the hydrocephalus had become arrested spontaneously in 53 cases, including the seven who previously had unsuccessful operations (Table 11). The arrest occurred most commonly between the ages of 9 months and 1 year and 9 months (Fig. 10). Various factors, such as cessation of head growth, reduction in the fontanelle tension and improvement in the general condition of the child were taken into account when making the judgement. However, as it is a gradual process, the time is difficult to gauge accurately and the age given at which arrest took place can only be an approximation. The distribution of lesions along the cerebrospinal axis, among the arrested cases,

TABLE 10
SURGICAL TREATMENT OF HYDROCEPHALUS WITH SPINA BIFIDA CYSTICA
(53 cases)

Type of Operation	Alive		Dead		Total
	Operation Satisfactory	Operation Not Satisfactory	Operation Satisfactory	Operation Not Satisfactory	
Ventriculo-subdural	1	9	1	10	21
Spino-ureteric	—	1	—	9	10
Ventriculo-pharyngeal	—	—	—	7	7
Ventriculo-peritoneal	—	1	1	3	5
Ventriculo-ureteric	—	1	—	2	3
Posterior fossa decompression	—	—	—	2	2
Third ventriculotomy	—	—	—	1	1
Torkildsen's	—	—	—	1	1
Ventriculo-sinusotomy	—	—	—	1	1
Ligation of carotid arteries	—	—	—	1	1
Diathermy excision of choroid plexus	—	—	—	1	1
	1	12*	2	38	53

* Of those whose operation was not successful but are still alive, seven arrested spontaneously subsequently and five still had progressive hydrocephalus at follow-up.

TABLE 11
HYDROCEPHALUS: FATE OF 235 CASES OF
GROUPS 3 AND 4

	Nos.
<i>Dead</i>	170
Progressive hydrocephalus (including 38 operated upon)	161
Arrested hydrocephalus, but died later (including two with successful operations)	9
<i>Alive</i>	65
Spontaneous arrest (including seven who had unsuccessful operation)	46
Successful operation	1
Hydrocephalus still progressive (including five with unsuccessful operations)	18

was about the same as that in the series as a whole. Also, a similar proportion of the survivors of Groups 3 and 4 hydrocephalus had become arrested, i.e. 28 and 26 cases respectively.

Of the 47 arrested survivors an estimate of intelligence had been obtained in 39, including 12 who have had a formal I.Q. examination. The eight cases for whom no estimate was obtained were all children under the age of 3 years. Half fell within the normal I.Q. range with several well above

TABLE 12
INTELLIGENCE OF ARRESTED SURVIVORS:
47 MYELOCELES

Intelligence Quotient	Degree of Hydrocephalus		Total
	Group 3	Group 4	
0-49	4	4	8
50-84	4	8	12
>85	14	5	19
No estimate	4	4	8
Total	26	21	47

average intelligence and in only eight was there severe retardation (Table 12). Although the relation between I.Q. and the degree of hydrocephalus was not a close one, as in another recent investigation (Laurence and Coates, 1962), almost three-quarters of those with normal intelligence had Group 3 hydrocephalus. Among those with normal intelligence there appeared to be a slight preponderance of cases with lesions in the lumbar or lumbosacral region, though this is by no means statistically significant. In this context it is of interest that, of the 15 survivors with an encephalocele, which included those in whom the hydrocephalus had not yet become arrested as well as those without significant hydrocephalus, eight were thought to be very severely retarded.

Unfortunately, arrest of the hydrocephalus is no

FIG. 10.—Age of spontaneous arrest of 53 cases of Group 3 and 4 hydrocephalus. Seven cases died subsequently (see text). One case, not included in this chart, arrested following operation at 1 year 3 months.

guarantee of survival in these cases. Not only may hydrocephalus associated with spina bifida cystica become progressive once more without any apparent predisposing cause, as in other forms of hydrocephalus, but these patients are at risk to a much greater extent from other causes. Ascending meningitis may develop as it did in two of this series at 9 months and 2½ years. Three who had a 'cosmetic' operation on their sac at 3 years, 5 years and 6 years, died of acute intracranial hypertension within a short time of the operation; two further patients died of renal failure at 2½ and 2½ years.

LEAKING AND BURSTING OF SPINAL LESION. In a considerable number of cases of myelocoele, the sac burst dramatically (34 cases) or else leaked continuously or intermittently (102 cases). This occurred most commonly in the dorsi-lumbar, lumbar and lumbosacral spina bifida, where this complication arose in almost half the cases (43%), while at the upper or lower end of the cerebrospinal axis, the incidence was much lower (18% and 26% respectively). The occurrence of severe hydrocephalus was rather higher with this complication than in the myelocoeles as a group (55% instead of 42%) and only 15% escaped hydrocephalus altogether. The mortality was also rather raised—70% in those that leaked and 61% in those that burst—by far the commonest cause of death being meningitis. On the face of it, one might expect the sudden rupture of the sac to be the more lethal event, but most of the cases in which the sac burst were dealt with as surgical emergencies, while a considerable proportion of those with leaks were treated expectantly.

SURGICAL TREATMENT OF SPINAL LESION. Some features of interest emerge from a brief analysis of the 160 who were operated on. As they were operated upon by a number of different surgeons with different operating policies, using different techniques, it is not possible to draw any conclusions from this series on these points. A little under half of the cases were operated upon, except for the sacral lesions and the encephalocoeles, where 28% and 84% respectively were so treated. This is probably because almost half of the former had normal skin covering the lesion and because the latter present difficult nursing problems when the lesion is large. Only 13 cases were treated on the day of birth and just over half (87 cases) were operated upon after 3 months. Several of those receiving early operation were dealt with elsewhere and were later referred because of one of the complications of the condition.

The mortality of the operated group was only about half that of the series as a whole (23% instead

of 50%). At the same time, this group contained a very much larger proportion of cases who had either mild hydrocephalus (Group 2) or none at all (63% instead of 36%). This was likely to have been due to selection for operation rather than a result of it.

The fear of hydrocephalus resulting from surgical interference has been largely allayed by others, but in this series eight cases developed hydrocephalus fairly acutely after the operation on the spina bifida when apparently none was present clinically before it. However, all but one of these were operated upon during the first month of life and it is very likely that all did, in fact, have at least some hydrocephalus before the operation. A comparison of the time of onset in the operated and unoperated groups shows that there are moderate differences that again are likely to be largely accounted for on the basis of selection for surgery (Fig. 11).

FIG. 11.—A graph showing the age of onset of hydrocephalus in the 160 cases that had an operation for a myelocoele or encephalocoele and 208 cases that have not.

Nine cases died directly as a result of their operation, three from intracranial infection and six from acute medullary compression. In others, the paralysis became more severe generally following a 'radical' removal of the sac, carried out for 'cosmetic' reasons.

CAUSE OF DEATH. Altogether 185 cases died and in 109 of these the cause of death was determined by autopsy. In 67 instances I either carried out the autopsy or examined the slides and specimens myself. In 42 other cases the cause of death was taken from reports of autopsies carried out by other pathologists. In the 76 cases in whom no autopsy

TABLE 13
CAUSE OF DEATH: 185 CASES

Cause of Death	Nos.
Hydrocephalus	59
Intracranial infection	83
Operative	15
Other related	25
Bronchopneumonia	17*
Renal failure or infection	8
Unrelated	3†

* 14 of these with progressive hydrocephalus should probably be regarded as hydrocephalic deaths.

† One case of 'inhaled vomit' and two of gastro-enteritis, all with progressive hydrocephalus, should probably also be regarded as hydrocephalic deaths.

was carried out, the cause of death was deduced from a detailed report of the terminal illness or taken from the death certificates.

Intracranial infection was the commonest single cause of death (Table 13) and was responsible for nearly all the deaths during the first three months and a very much smaller proportion of deaths after that age (Fig. 12). Hydrocephalus, which accounted for 59 cases, became an important cause after 3 months, but ceased to be a major factor after the age of 2 years. Although bronchopneumonia was the immediate cause in a further 17 instances, 14 of those who had rapidly progressive hydrocephalus should perhaps be regarded as hydrocephalic deaths. This also applies to three where the cause of death was

FIG. 12.—A histogram showing the relation between cause of death and age at death in the 185 cases.

'unrelated': two cases of 'gastro-enteritis' and one of 'inhaled vomit' that had also severe hydrocephalus. It is of interest that of the 185 cases that died, 182 had associated hydrocephalus, 170 of them falling into Groups 3 or 4. In 12 with only mild hydrocephalus (Group 2), it was unlikely to have been an important factor in the condition. Of these, six died of meningitis all in the early weeks of life, five of them after cerebrospinal fluid leakage or shortly after an operation on the sac; two died of acute medullary failure following operation; one died of renal failure and three of bronchopneumonia. The three with no ventricular dilatation died of renal failure (one case) and ascending meningitis (two cases).

Discussion

This series is presented not because of any greater rate of survival than in previous reported series, but because it may give some indication of the natural history of the condition. Indeed, it is very difficult to compare the survival in one series with that of another because of the inevitable differences of selection of cases and because the cases included in the series are then not discussed in the context of all those occurring in the community from which they have been drawn. These problems aside, straight survival figures, such as are normally given, do not mean a great deal and do not give a reliable indication of the prognosis, for reasons already stated. An urgent plea is made for survival data to be presented in the way they have been done in this paper, as such data from life tables and life charts can give comparable, more generally applicable survival figures. The only survival figures with which the data here presented might be compared are those of McKeown and Record, who investigated all the births in Birmingham from 1950 to 1952 inclusive (McKeown and Record, 1960). They found that of 63 cases, including stillbirths, of spina bifida cystica, without other malformations, 41% were still alive at 5 years of age, but that only 5% of the 103 cases with associated malformations (presumably mostly hydrocephalus) had survived the same length of time. Comparing the proportion of 3-month-old infants who then survived to 5 years of age or longer, 54% in the present series and 75% in that of McKeown and Record, it is seen that even when allowances are made, the two series are still not quite comparable. This suggests that it is extremely difficult to relate accurately, or even partially, selected groups of patients to the disease in the community.

The difficulty of diagnosing a meningocele with certainty is remarked on by several authors and is probably reflected by the great variation in the

incidence quoted, varying from 29% (Ingraham and Matson, 1954), and 28% (Fisher, Uihlein and Keith, 1952) and 27% (Schwidde, 1952) to 9% in the present investigation and about 4% in the Sheffield series (W. J. W. Sharrard, 1963, personal communication). The criteria for making the diagnosis vary considerably from those where a meningocele is diagnosed only after extensive clinical and pathological study, to a diagnosis of exclusion and only cases that have no hydrocephalus or no neurological deficit falling into that category (Ingraham and Swan, 1943). It is the author's experience that the more fully a 'meningocele' is investigated, the more likely is it found to be in fact a myelocele. Though some of these differences are undoubtedly due to selection of cases, one might wonder whether in some of those series that include a very high proportion of meningoceles all the cases were really always fully investigated.

The sex ratio for the meningoceles, like that in most other investigations, is about 1 to 1, but the 368 myeloceles have a preponderance of females, with a ratio of about 1 to 1.25 males to females. This compares with 1 to 1.15 for Doran and Guthkelch (1961) and 1 to 1.17 for Record and McKeown (1949), differences that are hardly significant. It has been suggested that the female embryo is more liable to develop this malformation (Doran and Guthkelch, 1961), and as evidence for this the high sex ratio of 1 to 1.51 males to females amongst stillbirths is quoted (Record and McKeown, 1949). It has, however, been found that in man a large proportion of conceptuses becomes addled (Hertig and Rock, 1949), and, experimentally, a high percentage of embryos of rats treated with trypan blue during the teratogenic period become absorbed (Beck and Lloyd, 1963). Differential mortality of the malformed embryos early in gestation, rather than different liability of the sexes to develop this malformation, may thus account for the disturbed sex ratio. Doran and Guthkelch further contend that spina bifida cystica is a more serious condition in the female. This is hardly supported by the present figures; in fact, neither the sex ratio of the children dying at the various ages nor that of the survivors of the various ages up to 12 years showed any statistically significant variation, nor even any recognizable consistent trend.

As any paralyses associated with spina bifida cystica must be regarded as an integral part of the condition, hydrocephalus is, therefore, the commonest and the most serious complication. It is probably present at birth in the majority of cases, though obviously without undue skull enlargement (Lorber, 1961) if there is no aqueduct malformation

and if the basal cistern compression is not too marked. Superadded ascending infection from the sac greatly increases the incidence and the severity of the hydrocephalus. Every effort must therefore be directed at reducing the incidence of this infection (Laurence, 1957), for, once present, it is not easy to control (Lorber and Bruce, 1963). The survival of cases with hydrocephalus here recorded is similar to the 34 cases with spina bifida included in an investigation into the natural history of hydrocephalus (Laurence and Coates, 1962); however, this survival rate is rather low in both groups and was largely dependent on whether spontaneous arrest occurred, as the surgical operations employed for the relief of hydrocephalus were not generally effective. In fact, nearly all the deaths in the present series were amongst those with hydrocephalus. Hydrocephalus thus appears to be a grave prognostic sign in spina bifida cystica. However, this survival rate could be used as a baseline on which results of the newer and, generally, more satisfactory operative procedures, such as that employing the Holter valve, might be judged. Early and resolute surgery should also be undertaken with a view to preventing some of the damage consequent upon ventricular dilatation. Only if the hydrocephalus is mild, or if spontaneous arrest is obviously taking place, is it justifiable to temporize provided of course there are no other contra-indications to surgery such as an infected cerebrospinal fluid, overwhelming neurological deficit or other associated gross malformations. Severe hydrocephalus itself should not be a contra-indication for it has been shown that even the presence of severe hydrocephalus need not necessarily mean that the infant will grow up to be retarded (Laurence and Coates, 1962). This is even underlined by the relatively few intelligence assessments available on cases of this series. Indeed, cases of hydrocephalus with spina bifida cystica tend to show fewer electroencephalographic abnormalities than other types of hydrocephalus (Pampiglione and Laurence, 1962), presumably because the brain is often damaged by ventricular dilatation only, implying that parenchymal damage is less frequent at least in the survivors.

A considerable proportion of infants are born with relatively normal reflex activity and a good deal of spontaneous movements in their legs. Sharrard and his co-workers (1963) have found that even muscles that appear to be paralysed generally give a positive response to short-duration stimulation or to nerve conduction stimulation. Of 40 unselected newborn infants with lumbar myeloceles so tested, only one showed mainly reflex activity in the

legs, all others having muscle activity that was independent of stimulation of the limbs. Unfortunately, within a few hours of birth, one after another of the muscle groups that show spontaneous movement become flaccid, and the sphincter becomes paralysed, and this, clinically, reaches its full extent in about 72 hours (Laurence, 1958a). This deterioration in the neurological state is probably due, at least, in part to biochemical alteration in the neural plate because of drying or over-hydration of the neural plate or from the chemicals used to prevent infection, or as a result of stretching of the nerve roots because of fluid accumulating in the thecal space below the plate, or because of infection of the neural plate itself. This infection not only leads to the partial or complete destruction of any functioning neurones in the plate, but it also leads to infection of the subarachnoid space beneath it and often gives rise to further distension and the loculation of the space so frequently seen. It also leads to the ascending meningitis which then may cause hydrocephalus or aggravate any hydrocephalus already present. The spinal lesion should be operated upon as an emergency within as short a time as possible of the delivery of the infant, ideally within one or two hours, with the lesion being kept slightly moist in the meanwhile. Sharrard and his co-workers in a controlled trial, found that such an early operation, as opposed to conservative treatment, led not only to a preservation of motor function but to an improvement upon the apparent neurological status at birth, and also to a reduction in the incidence of meningitis and to a shortening of the time that the infant remained in hospital. Operation in the immediate neonatal period appears to be a much easier and less hazardous procedure as there is at this time no gross infection, oedema or fibrosis, and the infants appear to stand surgical procedures more easily then. In the past, the type of operation often employed was a radical removal of the sac, frequently carried out after the age of 3 months or later for cosmetic or nursing reasons alone. This has led to some tragic cases where the paralysis was made more complete; there is no place for such a procedure except in the large encephaloceles. Especially since 1932 when Penfield and Cone suggested that the sac ought to be preserved in order to assist in the absorption of cerebrospinal fluid and thus reduce the incidence of post-operative hydrocephalus, surgeons have progressively been more reluctant to remove the spina bifida sac completely. Many, however, advocate dissecting the neural element from the sac wall and returning the former to the thecal space; they still advocate waiting for some months before doing so (Macnab,

1958; Guthkelch, 1962), by which time the sac is generally epithelialized; however, tissue planes are generally distorted because of fibrosis. Inevitably much damage may be done to the nervous tissue during such a procedure. On the assumption that one should not bury epithelial elements under skin flaps, many surgeons remove membranous portions of the sac which, however, may include neural and sometimes even central nervous system elements. It has been suggested that the operation of choice in an uninfected case, seen in the earliest neonatal period, might be deflation of the sac by means of a syringe with a wide-bored needle (if this is necessary), and merely covering the sac with skin flaps without removing anything (Laurence, 1957). A similar procedure has been advocated by Kredel (1950) who suggests that epithelial elements buried under skin flaps tend to disappear, or only lead to microscopic cyst formation, causing no trouble. This early but conservative surgical treatment of the sac should result in the maximum preservation of function and, as Sharrard and his co-workers suggest, eventually enable a considerable proportion of these children to walk unaided. It should also reduce the incidence of severe hydrocephalus.

The new attitude towards this group of malformations may mean that this was the last opportunity for examining the natural course of the condition. A review of the cases born during the 10 years from 1956 should yield different and better results, just as the survival in this series was somewhat better than that reported from The Hospital for Sick Children by Norman (1949), largely because of the use of the antibiotics.

Summary

This investigation was based on 407 children with spina bifida cystica born between 1947 and 1956, seen at The Hospital for Sick Children or the Westminster Children's Hospital, London, by the end of 1957. All cases were followed up in the summer of 1958 and the survivors were re-examined. It was considered appropriate to subdivide them into meningoceles and myeloceles only.

The series was found to be fairly representative of the disease for infants surviving beyond the age of 3 months, and 55% of the whole series were still alive. When prognosis is calculated on actuarial principles, using a life table, it is found that only 35.4% of cases attending a large children's hospital can be expected to survive to 12 years. A plea is made for survival figures to be presented in this way.

The meningoceles, which are a difficult diagnostic problem and which formed 9% of the series, all

survived. Only a few suffered from mild paralysis or hydrocephalus.

The 368 myeloceles had an expectation of survival to the age of 12 years of 29%. Their neurological condition usually becomes worse soon after birth, probably because of biochemical change, stretching or infection of the neural plate. Nevertheless, the pattern of paralysis appeared to be largely determined by the position and extent of the lesion, with maximal skeletal paralysis with lumbar lesions, while sphincter paralysis became more frequent and severe the lower the lesion along the neural axis.

Moderate or severe hydrocephalus that is due to malformations or post-inflammatory lesions from ascending meningitis, or one superimposed upon the other, was present in 235 cases. It was clinically present at birth in at least 28% of these and was diagnosed in all but one by 6 months. The incidence was highest in the dorsi-lumbar (88%) and lowest in the sacral myeloceles (37%). Death occurred soon except in the encephalocèles (40%) who seem to have a better prognosis with regard to survival but not intelligence. In 53 cases arrest took place spontaneously, mostly between the ages of 9 months and 1 year and 9 months, and in three following an operation; nine of these died subsequently from other causes.

185 cases died, all but three having hydrocephalus of moderate or severe degrees. Infection was the commonest immediate cause (83 cases) followed by progressive hydrocephalus (59 cases). Eight cases died of renal failure or infection. Although intracranial infection was the commonest cause of death the development of hydrocephalus appears to be a grave prognostic sign, as comparatively few of these cases survived.

It is suggested that results may well be improved if the raw spinal lesion is covered with skin grafts as a surgical emergency within hours of birth without dissecting any neural elements from the sac wall or even removing part of the sac. This should lessen the risk of damage to any functioning cord tissue in the myelocèle and also reduce the incidence of ascending infection and aggravation of any hydrocephalus. The mortality in this condition should also be reduced as a result of the newer surgical methods for the treatment of hydrocephalus.

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