Spina Bifida Cystica

Results of Treatment of 270 Consecutive Cases with Criteria for Selection for the Future

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Lorber, J. (1972). *Archives of Disease in Childhood*, 47, 854. **Spina bifida cystica: results of treatment of 270 consecutive cases with criteria for selection for the future.** The disappointing results of treatment in 270 consecutive unselected cases of spina bifida admitted during a 27-month period are detailed. Massive effort has led to much avoidable suffering at an exorbitant cost in manpower and money. This study confirms the validity of those adverse prognostic criteria defined in an earlier study and which form a basis for selection. These are (1) thoracolumbar lesions, (2) severe paraplegia, (3) gross enlargement of head, (4) kyphosis, and (5) other severe congenital defects, or birth injuries. It is shown again that selection is possible on the first day of life on purely objective criteria; that it is essential for the benefit of all those affected—whether they are for treatment or no treatment; and that it is in the interest of their families and the community.

The long-term results of treatment of infants born with open myelomeningocele are disappointing in spite of an immense effort to minimize their handicaps. Improved techniques did not lead to fewer, but to more, handicapped children, because with the increased survival rate many more severely affected infants survive (Lorber, 1971a). The policy of offering active treatment to all infants, irrespective of the type and degree of their physical condition and their social background, resulted in great suffering for the patients and in grave family, social, educational, financial, and community problems (Hare *et al.*, 1966; Freeston, 1971; Lightowler, 1971; Walker, Thomas, and Russell, 1971). For these reasons an attempt was made in an earlier study to define certain criteria which can be readily recognized soon after birth, without the use of elaborate investigations, and which would serve as a basis for selection. Patients would be divided into two groups: those who are likely to benefit from and should be offered total care, and those who are unlikely to benefit, either because they are likely to die soon or, if they survive, will be so severely handicapped that active treatment of any kind could be considered to be against their interest (Lorber, 1971a).

In a prospective study of 201 consecutive infants born in 1967 and 1968 and treated from the first day of life onwards, 4 main adverse criteria, present

on the day of birth, were found to be associated with a consistently grave prognosis. These four criteria are:

(1) *Severe paraplegia.* (Grade 5 paraplegia: at most hip flexors acting; Grade 4 paraplegia: at most hip adductors and the quadriceps acting, in addition to hip flexors.) Almost all such infants also have paralysed bladder and rectal sphincters.

(2) *Gross enlargement of the head,* the head circumference being at least 2 cm above the 90th centile, corrected for birthweight (Fig. 1).

FIG. 1.—Head circumference centiles at birth related to birthweight (modified from O'Neill, 1961).

Received 7 April 1972.

FIG. 2.—X-ray of the spine at birth, showing kyphosis.

(3) *Kyphosis* (Fig. 2).

(4) *Associated gross congenital anomalies or major birth injuries* (Fig. 3).

Those in the last category had the highest mortality and the survivors in all 4 groups have very severe multisystem physical handicaps. Only 20% of those admitted were considered to have normal intelligence. 50% were dead by 2 years of age.

It was also shown that infants with thoracolumbar lesions had a particularly bad prognosis, as well as those with severe degree of hydrocephalus, even if their heads were not extremely big. Finally, ventriculitis in the neonatal period in those already suffering from hydrocephalus or paraplegia so much worsened the prognosis that treatment is now considered to be contraindicated, except in infants with little or no handicap.

The age of the survivors in the previous study (Lorber, 1971a) was 2½ to 4 years and they neces-

sarily did not have such full assessment as is possible at a later age. For this reason the present study was undertaken to test the validity of these suggested criteria for selection. An earlier cohort of an equally large group of well-studied patients was chosen for this purpose.

POLICY

Before the first patient was admitted for the present study on 1 June 1962, the unit had considerable experience in the treatment of myelomeningocele, and well over 500 cases had already been treated (Sharrard, Zachary, and Lorber, 1967).

The policy of closure of the back lesion as soon as possible after birth on the first day of life was based on the results of a controlled therapeutic trial (Sharrard *et al.*, 1963). The conclusions of this trial were widely accepted, though the differences in the results between 'early' and 'late' closures were not statistically significant. Any apparent 'improvement' in muscle function was found to be temporary (Duckworth and Brown, 1970), and in several respects the conclusions then reached have not been substantiated by later experience. On the contrary, in my present opinion (but not in the opinion of all my colleagues) the urgency of treatment for all patients demanded in the conclusion probably resulted in more harm than good.

Apart from the emergency closure of the back, the policy of the unit was to offer total care to all infants, irrespective of the degree of their handicap, with all available surgical and medical means (Zachary, 1968). Infections, including ventriculitis, were energetically treated (Lorber and Segall, 1962); and collapsed infants were resuscitated on the operating table. On the surgical wards cardiac massage was practised in cases of cardiac arrest.

The associated hydrocephalus was investigated on the medical wards and only those cases of hydrocephalus were not surgically treated, who died in the neonatal period or were sufficiently mild not to require operation. Unfortunately, routine air-ventriculography was not carried out in all cases. The policy was to operate on all infants whose CSF pressure was over 300 mm (Lorber, 1961b). A fair indication of ventricular size was, however, obtained in all other infants by a ventricular tap carried out to measure the pressure and to ascertain that the CSF was normal before operation. High pressure as a sole indication for operation was not supported by later studies (Lorber, 1969, 1971b) and, based on this indication, many infants

were probably unnecessarily operated on. In patients with a CSF pressure below 300 mm air-ventriculography was carried out and the degree of hydrocephalus was classified as follows: Grade 1 hydrocephalus with a residual cerebral mantle of 35 mm or more, Grade 2 with a mantle of 16 to 35 mm; and Grade 3 with a mantle of 15 mm or less (Lorber, 1961a). During the period under consideration a controlled trial was in progress establishing the need—or lack of need—for surgical treatment of those with moderate (Grade 2) hydrocephalus (Lorber, 1971b). As many 'controls' did not need shunt therapy, it is apparent that several of those who had shunt treatment did not need it either.

Complications of shunt therapy were all operated on, even if the child had severe intellectual or physical defects.

The urinary tract also received constant active attention clinically, radiologically, surgically, and medically. Large numbers of operations were carried out and an extensive system was developed for the prevention (if possible) and the treatment of urinary tract infections (Cook, Lister, and Zachary, 1968; Cudmore and Zachary, 1970; Lister, Cook, and Zachary, 1968; Lorber, 1967b; Lorber and Formby, 1968; Lorber, Menneer, and Allott, 1968; Pekarovic *et al.*, 1968; Pekarovic, Robinson, and Zachary, 1969).

Orthopaedic problems received similar active consideration and an aggressive surgical approach was adopted (Sharrard, 1964, 1968, 1969; Sharrard and Drennan, 1972; Sharrard and Grosfield, 1968).

Such all-out attempts at total care were only limited by existing knowledge and by the facilities of time and space in a unit which, by the time the survivors of the present series were of school age, had dealt with over 1000 cases.

Definitions

In this paper, as in the previous study (Lorber, 1971a), children will be regarded as having *moderate physical handicaps* if they are incontinent, or have an ileal loop diversion, but do not have chronic urinary tract infection or hydronephrosis; if they are able to walk without the help of calipers, though they may limp, waddle, or need surgical boots; if they have analgesia of their legs or perineum but without chronic ulceration or abscess; if they have hydrocephalus which either required no shunt therapy or hydrocephalus well controlled with a shunt. If a child has any one or any combination of these defects, he is considered to have *moderate handicaps*.

Children are regarded as having *severe physical*

handicaps if any handicap is more severe than those just defined; incontinence with pyelonephritis or hydronephrosis, renal stones, or hypertension; paraplegia requiring calipers, crutches, or wheelchair for locomotion; kyphosis or scoliosis; repeated fractures or unreduced dislocated joints; hydrocephalus under precarious control; blindness, fits, or other less common defects.

Intelligence has been classified on the basis of repeated Terman-Merrill tests. Those with an IQ of 80 or over are considered normal, but those with normal intelligence will be divided into those whose IQ is over 100 and those with IQ of 80 to 99; those with IQ of 60 to 79 are retarded, and those with less than 60 are severely retarded.

Muscle power was assessed and a muscle chart was prepared, just before operation in the operating theatre, with the infant fully warmed up, by the orthopaedic team under Mr. W. J. W. Sharrard, who himself carried out the large majority of tests. These tests form the basis for classifying muscle paralysis into 5 grades: Grade 1 apparently normal, Grade 5 most severely paralysed—intermediate grades as defined earlier (Lorber, 1971a).

CASE MATERIAL

(1) **Main series of 200 infants with open myelomeningocele.** It was decided to study 200 consecutive cases of open myelomeningocele admitted on the first day of life from 1 June 1962. The 200th patient was admitted on 5 September 1964. Closure of the spinal lesion was carried out in 195 cases by Mr. R. B. Zachary and Mr. J. Lister, and their surgical team, within 36 hours of birth (usually much sooner). In the remaining 5, technical difficulties or extreme illness precluded surgery, but these 5 were given all possible treatment, including antibiotic treatment of ventriculitis.

There were 116 girls (58%) and 84 boys (42%).

135 infants (67.5%) had one or more of the 4 major adverse criteria on admission and 65 (32.5%) did not. However, 16 of the latter had thoracolumbar or thoracolumbosacral lesions. Previous results (Lorber, 1971a), together with the additional evidence presented in this paper, indicate that infants with such lesions could be added to those with the 4 other major criteria.

Thus 49 (24.5%) infants are left who had no major adverse criteria on admission and 151 (75.5%) who did. This high proportion with adverse criteria should not be taken as typical of all cases of myelomeningocele. During the years these infants were born, the unit accepted large numbers of difficult cases from regions outside Sheffield, whereas the less severe cases were being treated in their own region. The series to be analysed is completely unselected from the receiving end, but selection of various types undoubtedly existed among those who referred cases to the unit.

Finally, ventriculitis in those with adverse criteria and in those with hydrocephalus at the time of diagnosis is

a contraindication to treatment, but without such criteria ventriculitis can and should be successfully treated. If the selection at birth is made correctly, very few need be excluded later because of ventriculitis or other reasons.

(2) **31 later admissions of infants with open myelomeningocele.** During the same period 31 other infants, aged 2 days to 10 months, were admitted for primary treatment of their open myelomeningocele. Closure of the myelomeningocele was carried out in 18 of these. In 13 closure was no longer necessary as the lesion was epithelialized, or was impossible either because of infection or for technical reasons. Nevertheless all other treatment was given to these infants, including antibiotic treatment of ventriculitis. 20 (64%) were girls; 28 (90%) had adverse criteria on admission.

(3) **36 infants with meningocele.** There were also 36 cases of membrane or skin-covered spina bifida without central nervous tissue in the lesion, and with or without associated hydrocephalus or diastematomyelia. 35 were surgically treated; the remaining normal child had a skin-covered lesion. 15 (42%) were girls. None had adverse criteria.

(4) **3 infants with skin-covered myelocele.** 2 were surgically treated, but the third died with extensive multiple congenital defects early in infancy. 2 were girls.

Thus a total of 270 cases of spina bifida were admitted to the unit for primary treatment during a period of just over 27 months. 153 (56%) were girls, whose prognosis was much poorer. 179 (65%) had adverse criteria on admission.

The results in the main series of 200 cases will be given in more detail than in the other 3 smaller groups.

RESULTS

Main Series of 200 Infants

103 children (51.5%) were living in December 1971, and were between 7 and 9 years of age. 65% of the boys and 41% of the girls survived. The difference is due to the much higher incidence of ventriculitis and, to a lesser degree, to the higher mortality from renal complications in girls (Tables I and II). 84% of those 49 without the 5 adverse criteria or 77% of those without the original 4 adverse criteria survived, as compared with the 58% sur-

TABLE I

Case Material and Survival Rate All Admissions between 1 June 1962 and 5 September 1964

	Admitted					Alive (December 1971)					
	Boys		Girls		Total	Boys		Girls		Total	
	No.	%	No.	%		No.	%	No.	%	No.	%
Open myelomeningocele	84	42	116	58	200	55	65	48	41	103	51.5
First day admissions											
Open myelomeningocele	11	36	20	64	31	5	50	10	50	15	50
Later admissions	21	58	15	42	36	19	90	14	93	33	92
Meningocele	1		2		3	—		2		2	
Closed myelocele											
Total	117	44	153	56	270	79	67.5	74	48.4	153	56.6

TABLE II

200 Cases of Open Myelomeningocele, First Day Admissions Life Table

Alive at	(1) No Adverse Criteria n = 49		With Adverse Criteria on Admission						All with Adverse Criteria n = 151	
			(2) Paraplegia Only n = 65		(3) Other Major Criteria n = 70		(4) T-L Lesion Other Than (2) and (3) n = 16			
		%		%		%		%		%
28 dy	45	92	56	86	40	57	15	94	111	73
1 yr	42	86	45	69	26	37	12	75	83	55
5 yr	41	84	39	60	17	23	10	63	66	43
7-9 yr	41	84	38	58	15	21	9	55	62	41

Note: The difference in mortality between those with no adverse criteria and those with them is highly significant ($\chi^2 = 11.6$; $P < 0.001$).

vival of those with severe paraplegia with or without thoracolumbar lesions, and 21% of those 70 who had at least one of the other major adverse criteria with or without major paralysis or a thoracolumbar site (Table III). Table IV shows the same results, in more detail according to the type of initial major criteria.

The difference between the survival rate of those with and without adverse criteria is highly significant ($\chi^2 = 11.6$; $P < 0.001$).

TABLE III

Results in 151 Infants with Adverse Criteria on Admission

	Total	Alive	IQ 100 +
Gross paralysis and/or thoracolumbar lesions without other criteria	81	47 (58%)	5 (6%)
Kyphosis, huge head, or congenital anomalies with or without gross paralysis and T-L site of lesion	70	15 (21%)	—

Note: All survivors are physically severely handicapped.

Children who died. 19 infants (9.5%) died within a week and 44 (22%) died by 28 days of age (Table II). 40 of these had one or more of the 5 major criteria, especially cerebral birth injury and major congenital defects, which accounted for 18 deaths. Ventriculitis and/or septicaemia with 20 cases was the commonest cause of death during the first month. Full details of the age and causes of death are given in Tables V and VI.

After the first 28 days, birth injuries and congenital defects no longer caused death. During the 2nd and 3rd months ventriculitis remained the principal cause of death with 12 out of 20 cases.

In 2 instances infants who developed ventriculitis survived with pyocephalus much longer. Altogether, primary ventriculitis was responsible for 34 of the 97 deaths (35%) and as 4 of these occurred in infants without initial adverse criteria, it is apparent that improved techniques in prevention and in treatment (Lorber, Kalhan, and Mahgreffe, 1970) could have reduced mortality among the less affected infants. In addition, ventriculitis associated with colonized shunts was responsible for 7 more deaths.

TABLE IV

Babies with Adverse Criteria on Admission

Adverse Criteria	Total	Alive	IQ 100 +
Grade 4 paralysis alone	18	9 (50%)	2
Grade 5 paralysis alone	47	29 (60%)	3
Kyphosis or congenital anomaly or grossly enlarged head (without gross paralysis)	12	3	—
Grade 4 paralysis + at least one other	12	3	—
Grade 5 paralysis + at least one other	46	9	—
Thoracolumbar lesions (without any above)	16	9 (56%)	—
All with adverse criteria	151	62 (41%)	5 (3.3%)

TABLE V

Principal Causes of Death Related to Age at Death and to Presence of Adverse Criteria on Admission

Age at Death	Congenital Defects Including Hydrocephalus	Birth Injury	Primary Ventriculitis and Septicaemia		Other Sepsis	Blocked and Infected Shunt		Renal	Other and Unknown		Total		
	Cr.*	Cr.	No Cr.	Cr.	Cr.	No Cr.	Cr.	Cr.	No Cr.	Cr.	No Cr.	Cr.	All
Under 28 days	10	8	3	17	2	—	1	1	1	1	4	40	44
1-3 mth	—	—	1	11	1	1	2	3	—	1	2	18	20
3-12 mth	—	—	—	1	1	1	4	2	—	2	1	10	11
1-9 yr	—	—	—	1	1†	1	10	7	—	2	1	21	22
All ages up to 9 years	10	8	4	30	5	3	17‡	13	1	6	8	89	97

*Cr. = 1 or more of 5 major adverse criteria on admission.

†Fibrocystic disease of the pancreas.

‡7 had ventriculitis in association with colonized shunts.

TABLE VI
Principal Causes of Death in Main Series

Cause	No Adverse Criteria (n = 49)		With Adverse Criteria (n = 151)		Total (n = 200)	
	No.	%	No.	%	No.	%
Congenital defects	—		10	7	10	5
Birth injury	—		8	5	8	4
Primary ventriculitis and septicaemia	4	8	30	20	34	17
Other sepsis	—		5	3	5	2.5
Blocked or infected shunt	3	6	17	11	20	10
Renal	—		13	9	13	6.5
Other	1	2	6	4	7	3.5
All	8	16	89	59	97	48.5

Complication of shunt therapy was directly responsible for 20 deaths (blocked, overworking, and infected shunts). 3 children who died of these complications had no adverse criteria. Some shunt-treated children did not require a shunt, as judged by retrospective evidence. This is another area in which prevention and better treatment could result in a lower mortality especially among the less severely handicapped.

Altogether 101 of the 200 patients had shunt therapy. A 20% mortality from a therapeutic procedure in an average of 8 years' observation should make one think very seriously before inserting a shunt—just as one would very seriously think about using a drug which has a 20% fatal complication rate. In addition there was severe morbidity attached to shunt therapy. The 33 shunt-treated infants who died had 75 operations (average 2.3) to deal with their hydrocephalus (Table VII).

TABLE VII
97 Dead Children

	Without Adverse Criteria	With Adverse Criteria	Total
Died under 1 year	7	68	75
Died over 1 year	1	21	22
Total number of operations	12	191	203
Duration of hospital stay (wk)	42	856	898

Renal failure and acute or chronic pyelonephritis was the principal cause of death in 13 children (13%). Gross hydronephrosis caused the death of one infant at 10 days of age. Renal causes of death were more common at later ages and were nearly as important causes of mortality as shunt complications. None of the renal deaths to date occurred in children without adverse criteria at

birth, but 7 of the 41 survivors in this category already had hydronephrosis or pyelonephritis—another challenge for improved techniques of prevention.

To illustrate the intensity of the effort to treat all infants against all odds, here are a few examples: all cases of primary ventriculitis were treated with the most suitable antibiotic drug by the intra-ventricular and systemic route, often till death. 8 other very severely affected infants were given cardiac massage or assisted ventilation, including one who collapsed before primary closure, could not breathe spontaneously, was intubated, and then the operation was carried out. Her heart then stopped, but she was resuscitated again with cardiac massage. She died at 5 days of age. Another infant's heart stopped before closure and was revived by cardiac massage.

One severely affected baby had imperforate anus (which was operated on), a thoracolumbar lesion and Grade 3 hydrocephalus; another, with a family history of cystic fibrosis, had meconium ileus. There were 2 mongols with severe paraplegia. One infant had ectopia vesicae which was operated on; 2 had 'Ondine's syndrome' of intermittent respiratory arrest (Mellins *et al.*, 1970); one had such a huge head that she could only be delivered after obstetric craniotomy. Many had gross skeletal defects (Fig. 3) in addition to their spina bifida.

Though the large majority of the deaths occurred in the first year of life, 22 (23%) took place later (Table V) and more will come. Practically all the late deaths occurred in children with adverse criteria. They and their families have been through an extremely difficult life. They had up to 14 operations, and these 22 had 105 (average 5) operations in all.

8 infants died who had no adverse criteria on

FIG. 3.—Infant with gross skeletal defects, after closure of the myelomeningocele. This child also has multiple internal congenital defects.

conditions, once developed, were treatable with a good chance of success. These facts indicate a possible improvement in prognosis of those infants who initially had no serious handicap.

Altogether the 97 dead children underwent 203 operations. 191 of these were performed on those with adverse criteria. They spent 898 weeks in hospital in Sheffield (average 9 weeks), not to mention admission to other local hospitals. Some who died at a later age spent their entire life in hospitals (Table VII). During the same time the surgical waiting list of the Children's Hospital went up by about the same number.

Survivors

Assessment of survivors. By December 1971, 103 children (51.5%) were alive. No child has been untraced. Information about 4 who had moved too far to be followed up personally has been obtained from colleagues within the past months. Several others who were also out of the area made a special visit back to Sheffield for a full assessment. All those who remained in the area underwent full serial physical, radiological, and psychometric assessment, and school reports have been obtained for nearly all children. All intelligence tests were carried out by an educational psychologist with special experience with these children (Dr. J. G. Parsons). The Terman-Merrill modification of the Stanford-Binet test (form L-M) has been used as a routine for the sake of uniformity.

admission. The cause of all these deaths (ventriculitis 4, shunt complications 3, and gastroenteritis 1) was potentially preventable, and the

Condition of survivors (Tables VIII and IX). There is no survivor without some physical defect. 10 children have *moderate* physical defects, as

TABLE VIII
Intelligence Quotient Related to Physical Handicap

	Boys n = 84		Girls n = 116		Total n = 200	
	No.	%	No.	%	No.	%
IQ 100+	11	13	2	2	13	6.5
Moderate sequelae	3		—		3	
Severe sequelae	8		2		10	
IQ 80-99	19	23	26	22	45	22.5
Moderate sequelae	—		4		4	
Severe sequelae	19		22		41	
IQ 60-79	21	25	14	12	35	17.5
Moderate sequelae	2		1		3	
Severe sequelae	19		13		32	
IQ Below 60						
All severe sequelae	4	5	6	5	10	5
All alive	55	65	48	41	103	51.5

Note: The difference between survival rate of boys and girls is significant at the 0.01 level ($\chi^2 = 6.8$ p = <0.01).

TABLE IX

Results of Treatment Related to Adverse Criteria on Admission (Including all Thoracolumbar Lesions), Main Series 1962-1964

	No Adverse Criteria n = 49 (%)	With Adverse Criteria n = 151 (%)
Alive	84	41
Moderate physical sequelae	20	—
IQ 100 +	6	—
80-99	8	—
60-79	6	—
Severe physical sequelae	64	41
IQ 100 +	10	3
80-99	27	18
60-79	23	14
Below 60	4	6

versely, no child with adverse criteria survived without severe handicaps (Table VII).

The remaining 93 survivors all have severe, usually multisystem handicaps. 10 have an IQ of 100 or more and 8 of these are boys. Only 2 of the 116 girls survive with an IQ of over 100 and both have very severe physical handicaps.

41 children with severe sequelae have an IQ of 80-99, 32 have an IQ of 60-79, and 10 have an IQ of less than 60.

Table IX shows the considerable difference in favour of those without adverse criteria, as far as survival rate (double), the overall physical defects, and intelligence are concerned. One-half of those admitted are alive with normal intelligence as compared with only one-fifth of those with adverse criteria.

defined earlier. 3 of them, all boys, have an IQ of over 100. The boy with the highest IQ of 117 has no hydrocephalus. There are 4 children with IQs of 80-99 and 3 with 69-79. The boy with the lowest IQ of 69 has a ventriculocardiac shunt which required 1 revision, but he is the only child in the entire series who walks normally and is continent.

None of the 10 children with moderate physical defects had adverse criteria on admission. Con-

Hydrocephalus (Tables X and XI). 7 of the 41 (17%) survivors without adverse criteria but only 3 of the 62 (5%) with adverse criteria do not have hydrocephalus. 93 survivors do have hydrocephalus, but whereas in those without adverse criteria it was usually mild or moderate (Grade 1 and 2), in the majority of those with adverse criteria it was severe (Grade 3). Consequently, 13 of the 41 (32%) without, but only 12 of 62 (19%) with criteria, had no surgical treatment. Altogether

TABLE X

Hydrocephalus in the 103 Survivors, Related to Adverse Criteria on Admission

	No.	No Shunt			With Shunt			
		No Hydrocephalus	Grade 1 and 2 Hydrocephalus	Average IQ	No.	Average IQ	Revisions	
							Patients	Operations
No adverse criteria	41	7	13		21		12	33
Adverse criteria	62	3	12		47		40	115
Total	103	10	25	92	68	79	52	148

TABLE XI

Main Series, Intelligence in 103 Survivors

	No Shunt		Shunt				Total
	No.	%	No.	%	Operations	Average Operations	
IQ 100 +	7	20	6	9	10	1.7	13
80-99	20	57	25	37	89	3.4	45
60-79	7	20	28	41	98	3.5	35
Below 60	1	3	9	13	19	2.1	10
All	35		68		216	3.0	103

Note: Average IQ, without shunt 92; with shunt 79.

half the survivors without but only one-quarter of those with adverse criteria had no shunt therapy. The average IQ of the 35 children without shunt is 92, and of the 68 with shunt it is 79. 20% of those without shunt as compared with 9% of those with shunt have an IQ of 100 or more, and 77% compared with 46% have an IQ of 80 or more. In contrast, 54% of those with shunt are retarded including 13% who are grossly retarded (Table XI). Out of the original 101 infants who had shunt therapy only 6 survive with an IQ of 100 or more. The highest is a boy whose IQ is 109; he is incontinent and wears full length calipers.

A smaller proportion of children required shunt revisions among those without adverse criteria (12 of 21 against 40 of 47) and the 41 had 54 operations to deal with hydrocephalus (average 1.3) whereas the 62 with adverse criteria had 162 operations (average 2.6).

Several children with shunt-treated Grade 2 hydrocephalus probably did not need the shunt (Lorber, 1971b) and would have been better off without it. Those who died due to shunt therapy have already been mentioned. Among the survivors is a girl now 8 years of age who had a stormy progress. She had 8 revisions and her IQ is only 68. At one stage her ventriculoperitoneal shunt perforated the colon, resulting in ascending ventriculitis due to enteric organisms. Her shunt had to be exteriorized. During this episode she lost her sight, but on appropriate drug therapy she recovered, the shunt was re-established, and she fully regained her vision. This was only one of several examples in which ventriculitis or blocked shunt led to blindness, and which was followed by full recovery of vision after appropriate therapy (Lorber, 1967a).

Another child had 3 revisions and became profoundly retarded; a third had colonization of the shunt with ventriculitis. This was only cured after removal of the shunt (which did not need replacement). A fourth developed craniosynostosis which had to be relieved by operation. A fifth child had 17 operations on his hydrocephalus

and now his ventricles are of almost normal size. Almost certainly all these 5—and some others—would have been better off without the original shunt operation.

No child in this series with Grade 3 hydrocephalus had an IQ of 100.

Locomotion (Tables XII–XIV). Only 1 child

TABLE XII

Main Series, Locomotion in 103 Survivors

Walks normally	1	<i>Orthopaedic operations</i>	
Walks nearly normally	8	No. of patients	83
Limps, waddles	6	No. of operations	436
Walks with below-knee calipers	25	Range	up to 14
Walks with below-knee calipers, has wheelchair	3	Average per operated patient	5
Full-length calipers, chest band, fully or almost chairbound	60		

walks normally and 8 walk almost normally; 6 limp or waddle. The rest rely on calipers with sticks, elbow crutches, or rollators, etc. The large majority, 60 of 103, are entirely or largely chairbound. Most of these have thigh-length calipers with abdominal or chest bands, and with these, plus crutches, etc. many can take steps. They are only able to raise their feet off the ground by tilting their pelvis. They move far faster and are more comfortable in their chairs which they must use out of doors, though 'walking' for some gives a great moral uplift and a sense of achievement. None can put on their own calipers. They are unable to negotiate steps. They would be helpless if left to their own devices in any emergency.

83 children underwent 436 orthopaedic operations—an average of 5, with a maximum of 14. The child with 14 operations is completely chairbound. 9 children had 10 or more orthopaedic operations: 3 of them did not have complete paraplegia at birth and can walk with below-knee calipers; 6 are completely chairbound: 5 had complete paraplegia at birth, but 1 had normal legs.

TABLE XIII

Gross Paraplegia and Scoliosis or Kyphosis Related to Muscle Assessment at Birth, Survivors (n = 103)

On Admission	Alive	Chairbound	Gross Kyphosis or Scoliosis	Orthopaedic Operations	
				Number	Average
No paralysis	16	3	6	28	2
Grade 2–3 paralysis	37	13	9	147	4
Grades 4 and 5 paralysis	50	44	21	261	5
All survivors	103	60	36	436	4.4

TABLE XIV
103 Survivors, Main Series, Locomotion and Orthopaedic Aspects

Legs on Admission	Total	Locomotion						Scoliosis or Kyphosis	Orthopaedic Operations		
		Normal	Near Normal	Limp or Waddle	Below-knee Calipers	Below-knee Calipers and Chair	F/L Calipers Chair ± ±		Patients	Total	No. per Operated Patient
<i>No adverse criteria</i>											
Grade 1 No paralysis	15	1	5	—	7	—	2	5	8	28	3
Grade 2	8	—	3	1	3	—	1	—	5	14	3
Grade 3	18	—	—	4	10	—	4	3	13	92	7
Total	41	1	8	5	20	—	7	8	26	134	5
<i>With adverse criteria</i>											
Thoracolumbar lesion											
Grade 1 to Grade 3	9	—	—	1	—	—	8	5	8	41	5
Other adverse criteria											
Grade 2 and Grade 3	3	—	—	—	—	2	1	2	3	6	2
Grade 4 paralysis alone	9	—	—	—	5	—	4	2	9	63	7
Grade 4 paralysis with other adverse criteria	3	—	—	—	—	—	3	2	3	14	5
Grade 5 paralysis alone	29	—	—	—	—	1	28	10	28	156	6
Grade 5 paralysis with other adverse criteria	9	—	—	—	—	—	9	7	6	22	4
Total	62	—	—	1	5	3	53	28	57	302	5
All survivors	103	1	8	6	25	3	60	36	83	436	5

She is now completely paraplegic with a dislocated hip and with scoliosis.

Among those 53 survivors who had none to moderate paralysis on admission (Grade 1 to 3)* at least 26 suffered *severe* loss of muscle function in spite of early closure. Similar loss of muscle function also occurred in those who died at a few weeks of age and who were not grossly paralysed at birth. Out of 16 surviving children who were considered to have normal legs at birth* only 1 walks normally; 3 are completely paralysed and chairbound; and 6 have gross scoliosis (Table XIII). Among the 37 survivors who had Grade 2 or 3 paralysis on admission, 13 are chairbound and 9 have severe scoliosis or kyphosis. Finally, out of the 50 survivors who had Grade 4 or 5 paralysis, 44 are chairbound and 21 have gross scoliosis or kyphosis (Fig. 4 and 5).

Taking all those with initial adverse criteria, 53 of the 62 survivors are mostly or entirely chairbound and 28 have severe scoliosis or kyphosis. Only 1 child waddles without needing calipers. In contrast, the proportion of children requiring wheelchairs who had no adverse criteria is 7 of 41, and 9 walk normally or nearly normally. No child improved on his initial muscle assessment.

*Including 12 who had adverse criteria, other than severe paralysis, i.e. the two first groups with adverse criteria in Table XIV.

FIG. 4.—Gross scoliosis and dwarfing. Case of hemimeningomyelocele.

FIG. 5.—Gross kyphosis and secondary chest deformity, with dwarfing.

83 of the 103 survivors had a total of 436 orthopaedic operations (average 5). The 16 children who had no paralysis on admission had 28 operations (average 2), the 37 with moderate paralysis had 147 (average 4), and the 50 with severe paralysis had 261 (average 5).

Table XIV shows in more detail the orthopaedic aspects of the survivors, indicating the very severe sequelae in almost all the children with initial adverse criteria.

Table XIV does not show that many children who could walk with the aid of calipers and other appliances are often in wheelchairs because their legs are in plaster after operations. They are often chairbound for prolonged periods, either because of problems and delays in fitting calipers, broken, ill-fitting, or outgrown calipers, or because the caliper rubbed sore places on their legs which usually take a long time to heal, or because they sustained fractures. These fractures are numerous and sometimes only a big callus draws attention to them, or they are accidentally detected on a routine x-ray, e.g. during intravenous pyelography. These fractures are usually due to severe osteoporosis, particularly after prolonged immobilization and most commonly affect the femora (Fig. 6 and 7).

Many children have or had dislocated hip joints. This may be of no great practical consequence for those who are chairbound, though a great deal of active surgery has been carried out to replace the

FIG. 6.—Severe osteoporosis with healed fractures.

hip in joint (Sharrard, 1964). Some hips, however, tend to redislocate even after posterior ileopsoas transplants and adductor tenotomies.

4 children had spinal osteotomy carried out at a few years of age, with improvement in the degree of their kyphosis (Sharrard and Drennan, 1972). Some children's scoliosis is of extreme degree.

Children with kyphosis tend to have chronic or recurrent ulceration of the skin over the kyphos requiring special attention.

FIG. 7.—Multiple recent fractures of both femora. Note excess callus formation.

Almost all children with severe paraplegia are dwarfed—their height being well below the 3rd centile.

Urinary tract (Table XV). Absence of sphincter control and its sequelae are among the most serious medical, and are probably the most serious social, problems in persons with myelomeningocele. The urinary tract, therefore, requires intensive investigative monitoring, especially because infection and damage to the upper renal tract are usually silent until the trouble is far advanced. This means frequent urine cultures on samples taken with the utmost care; prolonged treatment of any infection which may be found; repeated intravenous pyelography (soon after birth, at 1 year of age, and thereafter approximately at 2-yearly intervals as well as at special times, e.g. immediately before surgery on the renal tract); micturating cystourethrograms; isotope renograms; bladder pressure studies; biochemical studies of renal function; and measurement of the blood pressure (for references see p. 856).

Only 10 children are continent and 3 are almost continent. None of these had severe paralysis on admission though 3 had thoracolumbar lesions. All the remaining 90 were incontinent, but 28 of these had their urinary stream diverted and now have cutaneous, ileocutaneous, or sigmoid loop ureterostomies. Almost all these operations were carried out after recurrent infections, after the development of hydronephrosis, and often after operative procedures on the bladder neck had failed. Occasionally the operation was carried out for social reasons. There are many children with trabeculated bladder and diverticula who do not have a bypass. These diverticula predispose to chronic infections. 6 children have or had renal stones and 1 had a nephrectomy.

Altogether 73 operations were carried out on the renal tract—almost certainly not enough.

25 children have hydronephrosis which in 13 is so severe that they are unlikely to survive long. One of these has a single hydronephrotic kidney

with stones. Another had an episode of hypertensive encephalopathy (BP 220/160 mm Hg). 4 have sufficient arterial hypertension to require hypotensive therapy (Lorber and Lyons, 1970).

The major differences between the groups without and with adverse criteria on admission are that one-quarter of the former but only one-twentieth of the latter are continent or almost continent, and that hydronephrosis is nearly twice as common in the latter (17% and 29%). Unfortunately, gross kidney damage was not avoided in many better children with sacral lesions and whose main problems are urinary. Two of these who can walk unaided have extreme degree of hydronephrosis.

It is apparent that even closer watch on the renal tract, better methods of prevention of infection, and earlier surgical intervention to prevent hydronephrosis (Smith, 1966, 1972) could substantially improve the outlook, especially for those with less severe handicaps.

Infections in incontinent children are very frequent, but their detection, including the correct interpretation of urinary culture results, is difficult. It was impossible to culture the urine often enough. The distances involved and the numbers attending the clinic made this impossible. Major recent improvements in this field have been the introduction of routine urine cultures in all the residential schools (with reports coming to the centre and recommendations going out for treatment, without disturbing the child's education) and the introduction of dip-slide culture methods at schools and at some homes.

Another major role of the special schools is bladder training. This is only possible when the child has good intelligence and strong motivation to stay dry. Bladder expression is often abandoned by parents and children for fear of injury and lack of success. It is not recommended in the presence of ureteric reflux.

Rectal incontinence. This is almost invariable among children with urinary incontinence and occasionally it is the major social handicap. It

TABLE XV
Urinary Tract in Survivors Related to Adverse Criteria on Admission

	Total	Continent or Almost Continent	Incontinent Infections	Diversion	No. of Operations	Hydronephrosis Pyelonephritis	Raised BP
No adverse criteria	41	10	20	12	27	7	1
Adverse criteria	62	3*	42	16	46	18	3
All	103	13	62	28	73	25	4

*None of these had severe paralysis.

TABLE XVI
Schooling

Adverse Criteria	Total	Ordinary	Physically Handicapped	ESN*	Blind	Training School	None or ?
None	41	16 (40%)	21	2	—	1	1
Yes	62	8 (13%)	51	—	1	1	1
All patients	103	24 (23%)	72	2	1	2	2

*Educationally subnormal.

TABLE XVII
Infants with Thoracolumbar Lesions, First Day Admission, Alive

Leg Grade on Admission	No. Admitted	Total Alive No.	Shunt Treated Hydrocephalus No.	Chairbound No.	Loop and/or Hydronephrosis No.
1	3*	2	} 8 (80%)	1	—
2	8*	8 } 53%		4 } 80%	2 } 40%
3	8*	4		3	2
4	8	2	} 29 (90% ^o)	2	1
No other criteria	4	3 } 30%		3	1
With other criteria	9	3		3	1
5	32	18 56%	} 29 (90% ^o)	18 } 32 (100%)	4 } 9 (28% ^o)
No other criteria	5	9 20%		9	3
With other criteria	45	9		9	3
All	113	42 37%	37 (90%)	40 (95%)	13 (30%)

Note: Percentages relate to survivors.

*1 in each group had other 'adverse criteria' on admission.

does not threaten life, except by predisposing to urinary tract infection in severely constipated children.

Schooling (Table XVI). Nearly a quarter of the children (24) attend an ordinary school in spite of severe physical handicaps, and some are even in a wheelchair. 72 attend day or residential schools for the physically handicapped and the rest go to schools for the blind, the educationally subnormal, or to training schools. Only 13% of those with adverse criteria are at ordinary school, compared with 40% of those without.

The headmasters' school reports about scholastic achievement almost always confirm what might be expected from the children's IQ assessment. In several instances, however, school marks fall well below this standard through a variety of reasons, especially lack of concentrating ability and because of frequent absences.

Those who are badly physically handicapped will not be able to stay at an ordinary school once they reach the secondary school age.

One of the major problems of handicapped children attending ordinary schools is incontinence: faecal as well as urinary. Accidents inevitably occur and this exposes them to the ridicule of other

children. Such events necessitated the removal of children from ordinary to special schools.

Other features. Among the 103 survivors, 13 are subject to fits (unassociated with blockage of the shunt system). Only one of 31 recovered from ventriculitis in the newborn period, but 4 of 11 did survive ventriculitis occurring later in life which was associated with colonization of the shunt system; one has an IQ of 94 but the other 3 are retarded (IQ 61–65). 1 child is blind and 1 has paralysed arms presumably due to syringomyelia. The spine was re-explored in 4 patients to remove diastematomyelic spurs or dermoids. These children usually had upper motor neurone paralysis of their legs which was converted to lower motor neurone paralysis after re-exploration.

Several children had trophic ulcers of the buttocks which had to be excised and 1 had his foot amputated.

Family and nervous breakdown occurred in 10 families; in 6 of these the patients are retarded, and 3 are 'in care'.

Infants born with thoracolumbar lesions (Tables XVII and XVIII). Having dealt with all

TABLE XVIII

113 Infants with Thoracolumbar Lesions, First Day Admissions (Alive : 42 (37%))

Chairbound	40	No sphincter control	39
Gross kyphosis or scoliosis	20	Hydronephrosis, ileal loop diversion	13
Shunt treated hydrocephalus	37	IQ 100 + Retarded	None 22
All operations on survivors	391	Average	9

Note: All survivors have severe physical handicap.

the children first, it is now necessary to consider briefly the 113 children (56.5% of the series) born with thoracolumbar (or thoracolumbosacral) lesions, because it is suggested that infants with such lesions should be, with rare exceptions, considered to have such a poor prognosis that treatment should not be given.

As Table XVII shows, the large majority (77) had Grade 5 paralysis, but there were 3 who had no detectable paralysis on admission. 57 also had one or more adverse criteria, other than paralysis on admission.

Only 42 (37%) survive and *all* 42 have severe, multisystem physical defects. 40 (95%) are chairbound. They are all dwarfed; 20 have gross kyphosis and/or scoliosis; 4 had spinal osteotomy. The gross deterioration in muscular power and function in those few who were not severely paralysed on admission is noteworthy. All have severe sensory loss with its risks and complications. 40 have no sphincter control and 12 of these have hydronephrosis, including 10 with diversions of the urinary stream. 1 had a diversion but is not hydronephrotic.

Half the survivors are of normal intelligence but there are none with an IQ of over 100; 14% of the survivors are profoundly retarded. Several have fits, 1 is blind, 1 has quadriplegia, and 1 is permanently in hospital, having been abandoned by her parents.

Only 5 have no shunt. The 42 survivors have had 391 operations (average 9) so far (Table XVIII).

Possible breadwinners. The children in this survey are only between 7 and 9 years of age. Nevertheless, it is possible to get an estimate of the number who could, subject to survival to adult life, earn their own living in competitive employment and be self-supporting with a secure, independent place in society. It is assumed that the patients who have a chance to achieve this are either those whose physical handicaps are only

'moderate' (as defined) along with an IQ of at least 85, or who combine a severe, multisystem handicap with an IQ over 100 and with an absence of chronic or frequent illness. This is an optimistic assumption which takes no account of the fact that these severely handicapped persons are unlikely to be able to perform tasks requiring mobility and physical strength, that they may often be absent from work for hospital attendances, etc., and that they will have to compete for jobs against the physically fit. Under these circumstances an IQ of well over 100 may be required to achieve success. A severely handicapped person will have to have an unusual degree of perseverance and determination to overcome grave odds against him. He will need great strength of will to overcome many inevitable disappointments and what must appear to him as injustices and discrimination against handicapped persons.

At this stage there are 13 children (6.5% of all first day admissions and 13% of all survivors) who with good fortune might reach adult life and a chance of independent existence; but not all will do so. (None of the 31 'later admissions' fall into this group.) Their IQ range is between 99 and 117 (average 107). 4 had a major adverse criterion on admission. In 2 cases this was Grade 4 and in 2 others Grade 5 leg paralysis—none had other adverse criteria. No child had a thoracolumbar lesion; 3 had trivial or no hydrocephalus; all the others had moderate (Grade 2) hydrocephalus, 5 of whom did not need operative therapy; 5 had shunt therapy, but only 2 required revisions. Only one child walks normally (after two foot operations) and this girl has an ileal loop. Both children with initial Grade 4 paralysis can walk with below-knee calipers, both remain incontinent, and one is much troubled with faecal soiling. Both children with initial Grade 5 paralysis remain completely paraplegic, one is chairbound, the other walks with full-length calipers. One is incontinent, and the other has an ileal loop diversion, chronic pyelonephritis, and gross hydronephrosis, with hypertension (controlled with guanethidine). This girl is unlikely to reach adult life in a fit condition to work.

Only 3 children are fully or almost continent. 2 have ileal loop diversions and the rest are incontinent.

Of these 13, the 7 who are most likely to reach adult life and earn their living are all 4 with moderate handicaps and 3 with severe handicaps.

From these observations it is probably fair to conclude that, even with much optimism and with maximal goodwill of many persons concerned, at

most 5% of all first day admissions (and no later admissions) will earn their own living.

Hospital admissions of survivors (Table XIX). It is only possible to calculate the time the children spent in our own hospitals in Sheffield. Many were admitted to their local hospitals, often for prolonged care (including permanent hospital life for 3 abandoned children). In assessing the significance of their hospital stay it must be realized that they were only in hospital for acute conditions; and because of the great pressure on the beds, they spent much less time in hospital than would have been desirable. Because also, early discharge is a policy in the interest of children, who should spend as little time away from home or school as possible, they were often sent home a few days after shunt revision or after other operations. Orthopaedic admissions were particularly kept short, often only a day or two for an operation and immediately sent home in plaster. For these reasons the total time spent in hospitals is remarkably little, bearing in mind the number of admissions and procedures.

The 41 survivors with no adverse criteria spent altogether 793 weeks (average 19 weeks per patient) in our hospitals (range 3–55 weeks). One spent over a year in hospital.

The 62 survivors with adverse criteria spent 2003 weeks in hospital (average 32, range 3–107 weeks). 13 spent over 1 year in hospital. Altogether, all the survivors spent some 3,000 weeks in hospital, so far.

Many children had over 20 admissions in 8 years.

Cost. It is impossible to calculate the total cost with any precision. The inpatient cost of one of these children, especially while in the neonatal surgical unit must be several times the average of some £80 a week per child in this hospital. They need the help of an exceptionally large and senior medical and nursing staff, highly expensive equipment, drugs and dressings, cubicle accom-

modation, and extra heating. The cost of the operations and theatre staff as well as the complexity and number of radiological and laboratory investigations is considerably in excess of the usual patient need in hospital. At a conservative estimate £250 per week on average may be a fair figure. Thus the total inpatient cost would be of the order of £750,000 or £7,500 per patient.

To this must be added the cost of outpatient visits, often from long distances. Patients travel up to 160 miles, usually by ambulance, a journey which costs £50 for one child. The cost of outpatient investigations, including radiography, drugs, various orthopaedic appliances such as calipers and wheelchairs, renal appliances, i.e. belts, plasters, and disposable napkins, and other items must be added. The correspondence about these children to parents, family doctors, referring consultants, school medical officers, and medical officers of health, runs into hundreds of letters in many cases with a commensurate cost of secretarial staff, equipment, and postage. Most of the children after reaching 4 to 5 years of age go to special schools. The cost per child in the residential spina bifida school (65 pupils) in Sheffield is £1,600 per annum. Most parents with a seriously handicapped child now (since December, 1971) receive a more than badly needed and well-deserved £240 per annum as tax-free 'attendance allowance'.

The average cost per child per annum cannot be much less than £3,000. In the first years the bulk of this will be hospital treatment—later it will be schooling. This would mean about £300,000 per year for the 103 survivors. Over an average of 8 years this amounts to £2,400,000. By adding the cost of those who died, the total cost could approach £3,000,000, but the margin of error in this calculation is appreciable. There is no doubt, however, that Lightowler's (1971) calculation is a long way below a realistic figure.

TABLE XIX
Operations and Hospital Stay

	Operations				Hospital Stay (in Sheffield) (wk)			
	Alive (153)	Dead (117)	All (270)	Average	Alive	Dead	All	Average
Main series of 200								
First day admissions with myelomeningocele	885	203	1088	5	2796	898	3694	18
31 late admissions with myelomeningocele	130	25	155	5	396	153	549	18
36 cases of meningocele	51	5	56	1.6	60	8	68	2
3 cases of closed myelocele	5	—	5		12	2	14	3
	1071	233	1304		3264	1061	4325	16

This is the cost of the 200 patients now analysed and admitted during a 2-year period. With late admissions and transfers from other hospitals, the unit admitted some 1,500 babies with myelomeningocele in 14 years since 1958. The overall cost of all these patients may have been approximately as much as the annual budget of the Medical Research Council.

The cost of the research in our spina bifida unit has not been included in the calculation. It is a minute proportion of the total spent on service needs.

Open Myelomeningocele—Late Admission

31 infants with myelomeningocele were admitted for primary treatment between 2 days and 10 months of age (Table XX). 12 were under 1 week old and all but 1 were under 6 months of age. Closure of the back lesion was carried out in 18, of whom 13 are alive; no closure was necessary or possible in 13, of whom only 2 are alive.

3 children without adverse criteria. The neurological condition of these infants at birth is not known, but by the time they were admitted only 3 had no adverse criteria, as defined for newborn infants. One of these, who had no hydrocephalus and had good sphincter control and normal legs with a straight spine at 5 weeks of age, developed severe paraplegia with kyphoscoliosis and died at home aged 9 months from inhalation of vomitus. The second child has a cervical myelomeningocele with only minor neurological disability (slight weakness and clumsiness of one hand), but has an IQ of only 68. The third girl has a lumbosacral lesion, and no hydrocephalus; she is of normal intelligence but attends a school for the physically handicapped because of incontinence and a limping gait.

28 children with adverse criteria. All the remaining 28 of the 31 cases had adverse criteria on admission, 17 having thoracolumbar lesions; there

are 13 survivors and all have severe multisystem handicaps.

The dead. Of the 31 children of this group 15 died; 11 under 6 months of age, usually within a few weeks of admission, the remaining 4 between 13 months and 2½ years. Ventriculitis was the immediate cause of death in 8, blocked shunt in 4, pyelonephritis, acute respiratory illness, and gross multiple deformities in 1 each.

Survivors with Initial Adverse Criteria

Hydrocephalus, intelligence, and schooling.

Of the 13 survivors 1 has an IQ as high as 100; 7 fall within the range of 80–99; and 3 each between 60–79 and below 60 (Table XX).

Only 1 child has no hydrocephalus, but her IQ is only 73; she is paraplegic, has severe hydronephrosis, and had multiple operations to create an ileal loop ureterostomy. The remaining 12 children have had 43 operations to deal with their hydrocephalus.

Of the 13 survivors, only 1 child manages to attend an ordinary school; 1 each goes to a school for the blind and a 'training centre', 1 has no schooling. The rest are in schools for the physically handicapped.

Locomotion and orthopaedic aspects. 2 children (both with initial Grade 4 paralysis) can walk with below-knee calipers and the help of sticks and need a chair only for longer distances out of doors. The rest have full-length calipers and can take steps indoors or are entirely chairbound. 1 child fractured her femora 12 times, but still walks indoors with full-length calipers and sticks. Many others had fractures and have dislocated hips; 5 have gross kyphosis or scoliosis (Fig. 5). 12 children underwent 54 orthopaedic operations (average 4.5).

Urinary tract. 5 children have hydronephrosis and 4 had their urinary stream diverted; all the rest remain incontinent. The 4 children had 12 operations on their urinary tract.

TABLE XX
31 Late Admissions with Myelomeningocele

No adverse criteria on admission	3		
Alive, moderate sequelae	2		
Adverse criteria on admission	28		
Alive	13	All operations	148 (average 5)
Severe sequelae IQ 80–99	7	Weeks in hospital	546 (average 28)
IQ 60–79	3		
IQ below 60	3		

Ventriculitis. One child survived neonatal ventriculitis with a normal IQ. 2 others who had ventriculitis are severely retarded. 12 others who had ventriculitis are dead, though 4 'recovered' from their ventriculitis with profound intellectual and physical sequelae before they died of other causes.

It is clear from this brief account that the prognosis of these 'late admissions' was even graver than those admitted on the first day, because almost all had at least one, and most several, adverse criteria. Half developed ventriculitis.

Cases of Meningocele

During the period under review 36 infants were admitted with membrane or skin-covered meningocele (Table XXI). Unlike the patients with myelomeningocele, boys were more often affected (21:16). Of those who had a skin-covered lesion, all except one were operated on: 33 (92%) survive. 19 had sacral lesions and the rest cervical, thoracic lumbar, or lumbosacral. 27 had no hydrocephalus and the 26 who survive are physically normal children with an IQ range of 79-129. 9 have an IQ of 100 or more (average 101). One infant was found to have diastematomyelia as well, developed ventriculitis, and died at 2 weeks.

Nine children had hydrocephalus and were treated by a ventriculocardiac shunt, many requiring more than one revision. 7 are alive, all are of normal intelligence (range 80-133, average 104), and 5 are also physically normal. 1 has spastic legs and needs a wheelchair; another with a typical diastematomyelic spur has a weak, flaccid leg.

Two shunt-treated children died, one at 3 months and the other at 6 years of age, both due

hydrocephalus was mild in most of those children with meningocele, but the indication for surgery was an initial rapid rise in head circumference after closure of the spina bifida. Subsequent experience has shown that waiting a little longer would have led to arrest of the abnormal rate of head growth in most.

Infants with Skin-covered Myelocele

The last 3 patients had physical signs identical with those found in open myelomeningocele, but their lesions were fully covered by skin. One died soon after admission with multiple gross congenital defects. The 2 survivors have no hydrocephalus.

The first of these, a girl, had a large thoracic swelling which was explored 3 times to arrest the development of gross spasticity of her legs; her spasticity was only converted to flaccid paraplegia and she is wheelchair-bound. She has an IQ of 96 and is continent.

The second survivor is a girl who had a sacral lesion with normal legs and sphincter function at birth, but after removal of the lump she progressively became paraplegic and incontinent, and later an ileal loop diversion was carried out because of her hydronephrosis. Her IQ is 118.

DISCUSSION

This study confirms the grave prognostic significance of the previously examined 'adverse criteria' as well as that of thoracolumbar lesions and of ventriculitis. Together, between the two series studied, 267 first day admissions with open myelomeningocele had one or more such adverse criteria.

None survived without severe multisystem physical handicaps; only one-fifth of all admitted have an IQ of 80 or over and very few have an IQ of 100 or more. The few brighter children all have very severe physical handicaps.

There was no improvement in the mortality rate of the children with initial adverse criteria between 1962-64 and 1967-68. By 2 years of age half were dead, and of the older group 59% died by 7 to 9 years of age.

None of the survivors with only *moderate* sequelae had been initially assessed as having adverse criteria, so that a decision to withhold treatment from the latter group would not have reduced the numbers surviving with *moderate* handicap.

If one agrees in principle to selection, then difficulties are bound to arise as to where to draw the line. Details have therefore been given separately

TABLE XXI
Cases of Meningocele

	No Hydrocephalus	Shunt-treated Hydrocephalus	Shunt Operations
Normal	26	5	13
Diastematomyelia and/or weak leg	—	2	5
Dead	1	2	3
Total	27	9	21

to shunt complications. A review of the indication for shunt treatment in these children strongly suggests that they did not need a shunt and would be alive and well if they had no operation. The

about the results in different categories with adverse criteria, so that anyone may decide for himself where he wishes to draw the line.

There will be occasional exceptions, e.g. the infant with thoracolumbar lesions and normal legs and bladder. Individual consideration can and must be given to every baby. Among these considerations the *social situation* is vitally important. To treat a badly handicapped, abandoned, illegitimate baby is total disaster for the baby if he survives. To treat a similarly handicapped baby of an unmarried girl who is willing to keep her baby is total disaster for the mother as well.

Strong support for selection on very similar, though even stricter, criteria to those suggested in this paper comes from Melbourne, Australia, from one of the largest, long-established spina bifida units in the world. According to the *Medical Journal of Australia* (1971) the indications for non-intervention are thoracolumbar lesions, severe paraplegia, kyphosis, hydrocephalus at birth, other serious anomalies, very large lesions, infected sac, and meningitis. Only those with small lesions and minimal CNS involvement, without obvious hydrocephalus, are for immediate treatment. Cases that fall between these groups are observed, and treatment is given later to those whose early progress is good and if investigations suggest a favourable outcome.

Others may propose different criteria. Maybe not enough weight has been given to absence of sphincter control; maybe early intravenous pyelography could be carried out so that those with hydronephrosis at birth could be identified and excluded. Gross hydrocephalus can be ascertained quickly by echoencephalography in infants without excessive head circumference. These and other investigations need not take long. The need for urgency of operative closure will be discussed later.

The fate of infants born with myelomeningocele is of increasingly grave concern to doctors (Holt, 1970; Laurence, 1972; Miller, 1972; Slater, 1971), the general public, and most of all, to parents. Many, including parents, would prefer that such babies be allowed to die peacefully in early infancy, if their predictable quality of life would be beset with grave, multiple handicaps.

Many paediatricians and surgeons, other than the author, have exercised or advised 'selection' (Forrester, 1965, 1968; Freeman, 1972; Hide, Williams, and Ellis, 1972; Ingram, 1965; James, 1971; Matson, 1968; Smith, 1965; *Medical Journal of Australia*, 1971; Stark, 1971; Wickes, 1968).

The three main concerns of doctors who wish to select patients for treatment are firstly, the fate of

those few badly affected infants who survive in spite of absence of treatment; secondly, the fear that it is impossible to formulate adequate criteria for such selection (Forrest, 1965; Katzen, 1971); and thirdly, the fear of criticism by parents, the public, and by colleagues if operation or other treatment is denied to an infant. The view that operative treatment, to be successful, needs to be carried out within a day of birth, has led to hasty referral of infants to surgical centres, and to hasty, automatic operation, carried out without full consideration of the effect of such action on the infant, his family, and the community.

The first fear has been exaggerated: facts have been presented to show that very few truly untreated, initially severe cases survive for any length of time (Hide *et al.*, 1972; Lorber, 1971a). 'No treatment' in this context does not mean just 'no operation': it means the provision of normal tender nursing care (Matson, 1968) with ordinary feeding but no more; no incubators, no oxygen, no tube feeding, and no antibiotic drugs. To deny an infant the potential benefit of an operation and then prolong life by other means is irrational. One should either offer total care or nothing operative at all (Finlay, 1971). An untreated infant should be protected against all pain and discomfort. An occasional initially severe case will survive; if after some months an infant's general condition is good, then he can be brought back to the group given total care.

The second fear that adequate criteria cannot be formulated soon after birth has been allayed by the data provided in the previous paper, and confirmed by those in this study.

No significance should be attached to the high proportion of cases in our present series with adverse criteria, and that would therefore have been excluded from treatment on a 'selection' basis. Cases were often referred to us that would not have been treated elsewhere, and hence our material was heavily loaded with difficult cases. A different type of selection occurred in Liverpool; in 1960-62, 55 infants born in the city with myelomeningocele were not referred for treatment: all were dead by 6 months (Rickham and Mawsdley, 1966).

It is argued that poor results could be improved by greater skill, better techniques, and by more research, and not by 'abandoning' those with more serious defects. There is no doubt that substantial improvement should be obtained using these means. The fact that many initially almost normal infants develop ventriculitis, that those with normal or near normal legs become severely paralysed, that large numbers with normal upper renal tracts

become hydronephrotic, that some 20% with shunt therapy die as a direct result of that treatment within 8 years, are only a few but outstanding examples of how the results could be improved, even with existing knowledge. The impressive reduction of early mortality by the prevention of infection, especially of ventriculitis, in Liverpool (Mawdsley and Rickham, 1971) since the opening of a neonatal surgical centre in 1968 shows how much more could be done for the better babies. Ventriculitis is a particularly tragic and preventable complication because it occurs at least as often in infants with little or no paralysis (sacral lesions) as in those who are more severely affected (Lorber, 1971a). A major reduction of shunt complications can be achieved by a conservative approach to the milder degrees of hydrocephalus (Lorber, 1971b).

There is, however, no advance in sight which could make more than a marginal difference to the quality of the survivors with adverse criteria. No amount of orthopaedic skill could create muscle power where the spinal cord has failed to develop; no medical or surgical treatment of extreme hydrocephalus could restore lost brain tissue; and no method of treatment could lead to an acceptable quality of life in infants with gross multiple malformations. It would be wiser to concentrate on research projects aimed at discovering the causes of deterioration and failure in the better cases.

Is early closure really so essential, or is it of benefit only to some infants? Considerable doubt was thrown on the necessity of routine early closure by Doran and Guthkelch (1961) who were particularly worried about the substantial increased risk of clinical hydrocephalus in those treated early. They did not find that deterioration in muscle function was any greater in those treated later. Of course, it is true that many infants do lose muscle power even while being prepared for operation, and this is a dramatic event which is bound to colour the views of the surgeon. However, severe and often rapid loss of muscle function also occurs in spite of or because of early closure. Our own earlier controlled trial of early closure (Sharrard *et al.*, 1963) was regrettably interrupted before any conclusion of statistical significance was reached. Brocklehurst, Gleave, and Lewin (1967) did not find significant increase in useful leg function compared with preoperative levels in infants who were operated on within the first 24 hours of life. In a recent controlled trial by Piggot (1971), limited to better children, of 108 cases, half treated by early closure, 32 died (equal proportion in the two groups) before 3 months of age. Of the 40 survivors with early closure, the motor power of

13 deteriorated, and of the 36 with delayed closure 9 deteriorated by 3 months.

The third fear militating against selection is fear of criticism. It is the author's hope that the facts now presented will help to allay this fear. Our current approach to an infant born with spina bifida is for an assessment to be made urgently by myself or my senior assistant on the lines admirably described by Stark (1971). The motor function is also assessed separately by an orthopaedic colleague. A final decision is arrived at after a full discussion with the father, but preferably with both parents, and perhaps grandparents. No decision to treat or not to treat should be made without their full knowledge of all the facts, but the decision must be that of the doctor. It would be most unfair to thrust such responsibility on parents at any time, but especially during the first hours after the news of such a profound tragedy. It is relatively easy to forecast the *minimum* likely disability, but impossible to forecast the maximal disability, if things fail to go according to plan. If the likely minimal disability is such that one cannot recommend treatment, then the parents almost always agree.

CONCLUSIONS

The introduction of the Holter valve 14 years ago, more than anything else, has revolutionized the treatment of spina bifida. Enthusiasm for its remarkable potential resulted in large numbers of babies being treated uncritically. The long-term results now show that we overreached ourselves and that the pendulum has swung too far. We should concentrate our attention on the many unsolved but soluble problems of the less handicapped children. If we did so, our long-term survivors would be hardly fewer, but their quality should be better.

I wish to thank all my colleagues (many named in the text), doctors, nurses, headmasters, and headmistresses, too numerous to mention all by name. The opinions expressed in this paper are mine and in some respects they differ from those of colleagues.

I am particularly grateful to Dr. Ann M. Bruce who was my Research Assistant during the years the children in this series were admitted and whose case records made my task of analysis so much easier. I would also like to thank the parents and their local associations for their trust and for supporting my conclusions.

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