PAPERS AND ORIGINALS

Selection for Treatment in Spina Bifida Cystica*

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Summary

This paper discusses (1) the mortality in 295 patients with myelomeningocele born 1961-9, (2) the "quality of life" of 88 surviving patients born 1961-5, and (3) their implications for selection for treatment in spina bifida cystica. Evidence is presented that high neurological levels, the presence of severe hydrocephalus, especially if present at birth, meningitis and ventriculitis, and gross renal disease are very adverse factors both in mortality and in quality of life. In selection for treatment we recommend immediate repair of the sac in most low lesions, deferring treatment in high lesions, and re-evaluation of the survivors of the latter group at 1 month or later.

Introduction

Dramatic changes have taken place in the treatment of spina bifida cystica over the past 15 years owing to the introduction of more efficient methods of treating hydrocephalus and the sophistication of treatment for paraplegia and urinary incontinence. Ventriculoatrial shunts have been available since 1956 but were not in regular use in this hospital until the end of the 1950s. Patients born from 1960 onwards have had the modern treatment for spina bifida and many of these children are now of school age and available for more accurate assessment.

After more than a decade of energetic treatment it is possible to assess the efficacy of this massive therapeutic programme, involving as it does many disciplines of hospital and other professional personnel. Whereas previously clinical

impressions of many paediatricians have raised doubts whether treatment is worth while, these impressions have lacked the basis of objective evidence of long-term results. These impressions, often correct in themselves, have frequently been generalizations, sometimes to the detriment of individual children who may have benefited by active treatment. The present analysis, comparable with that of Lorber (1971), begins to define the criteria observable in the neonatal period that relate specifically to a good or bad prognosis and to the quality of survival.

There has been much controversy on selection of patients for active surgical treatment. Selection has ranged from complete rejection to complete acceptance of patients for early surgery. The latter policy is exemplified by workers in Sheffield reported by Lorber (1971). The present study shows an intermediate position. Since 1961 an active policy of selection for early surgical treatment has been carried out, and no early surgical treatment was given to 79 (27%) out of 295 patients born during 1961-9 because of an adverse prognosis. This policy, continued over a decade, has influenced the early mortality rate and especially the quality of life of the survivors—points which are elaborated in more detail below.

Patients Studied

During the period 1943-72, 718 patients with spina bifida cystica were treated at the Royal Children's Hospital, Melbourne, 570 before 1970 (table I). The present study is confined to those with myelomeningocele treated by modern

TABLE I—Outcome in Patients with Spina Bifida Cystica Born during 1943-69

	Year of Birth			Total
	1943-60	1961-5	1966-9	
Meningocele { No. dead .. { No. alive ..	3 26	1 8	11	4 45
Total	29	9	11	49
Myelomeningocele { No. Dead { No. Alive	140 (62%) 86 (38%)	71 (45%) 88 (55%)	53 (39%) 83 (61%)	264 257
Total	226	159	136	521
Grand total	255	168	147	570

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methods, including ventriculoatrial shunts, and with a minimum follow-up for mortality and quality of life of three years and seven years respectively. This comprises for the mortality statistics 295 patients born during 1961-9, and for quality of life 88 survivors out of 159 patients born during 1961-5. Only children with myelomeningocele have been considered; children with meningocele with no neurological signs and no disabilities (making up 6% of the 1961-9 series of spina bifida cystica) have been excluded. Two children who could not be traced have been excluded from the study, but otherwise the series is consecutive.

Assessment of Neurological Level

Basic to the study of mortality and the quality of survival is the classification of patients into "high" and "low" lesions. The neurological levels and numbers of patients at each level are shown in table II. It is convenient to divide the lesions into five levels. It should be noted that these levels refer to the neurological level of the spinal lesions rather than the level of the sac.

TABLE II—Neurological Level of Lesion and Mortality Rates in Patients with Myelomeningocele born during 1961-9

Neurological Level	No. Dead	No. Alive	Total
(I) Cervical and upper thoracic	0	2	2
(II) Lower thoracic	54	26	80
(III) Upper lumbar	35	32	67
(IV) Lower lumbar and upper sacral	31	103	134
(V) Lower sacral	4	8	12
Total	124 (42%)	171 (58%)	295 (100%)

Lesions in the cervical (C 1-7) and upper thoracic areas (T 1-6) (level I) usually consist of meningoceles but are occasionally myelomeningoceles with neurological involvement. Level II (T 6-12) comprises spinal cord lesions in the lower thoracic region, and such lesions produce complete paraplegia. Children in level III (L 1-2—that is, normal cord down to L 1 or L 2) have hip flexion and adduction only, all other leg muscles being paralysed, except sometimes for weak, ineffective movements of the quadriceps muscles. Lesions at levels I-III are referred to as "high" lesions, the dividing point from "low" lesions (level IV, L 3-S 2; level V, S 3-5) being the mid-lumbar region. Clinically the distinguishing feature between high and low lesions is the absence or presence of an active quadriceps.

There is little difficulty in determining the neurological level in children beyond infancy, when accurate muscle charting is possible. We believe that it is also almost always possible to determine the level in infancy, including the newborn period. We have compared our recorded notes in this period with more exact muscle charting performed later in childhood and have found close agreement between the two. In the few children where marginal changes have been observed the changes have generally been for the worse—that is, the degree of muscle involvement was underestimated and the subsequent level was slightly higher than first recorded. In the selection of these patients for operation this meant that these children had early surgery when the subsequent course threw some doubt on the wisdom of this, but, conversely, it ensured that all children with low lesions received early surgery unless there were other adverse factors. Three types of observations have been found useful—the type, size, and position of the sac, the presence or absence of active movements, and the posture of the limbs, the last two being the most helpful.

The size and position of the sac gives a rough guide to the level of the lesion but is not accurate enough for diagnosis; however, complete rachischisis is associated with high lesions.

In the neonatal period four large muscle groups are tested—flexors of the hips, extensors of the knees, and dorsiflexors and plantarflexors of the feet. Apart from the occasional level I child with arm and upper trunk involvement absence of active movements in the muscles of all four groups places the child in level II, the presence of the hip flexion only in level III, the presence of hip flexion, and knee extension, with or without foot dorsiflexion, in level IV, and the presence of plantar flexion (and so probably no orthopaedic disability) in level V. Some confusion can exist because of the presence of upper motor neurone lesion reflex movements in the legs, generally affecting the knee flexors and the muscles below the knees. These movements are almost always associated with high lesions, and the absence of active hip flexion and knee extension is usually obvious. In many infants more detailed muscle charting is possible but a study of the activity of these four major groups of muscles is sufficient at this early stage. There are sometimes slight differences (and rarely marked differences) in muscle power between the legs; in such cases the level is that corresponding to the more severely affected leg. We have not found sensory loss useful in determining the neurological level in the newborn period.

The posture of the limbs can also be of help. The flaccid, extended, abducted, and externally rotated hips—all by gravity—indicate a complete paralysis. A flexion contracture of the hips indicates the presence of an active psoas. When the lesion is below L 1 the hips lie flexed, abducted, and externally rotated; when below L 2, and the psoas and adductors are strongly active, the hips lie in flexion and adduction. Level IV is indicated when the knees are held in extension or hyperextension (from the quadriceps) and (if the lesion is below L 4) the foot is in dorsiflexion and varus (from active tibialis anterior and posterior and paralysed calf muscles).

Mortality

The drop in mortality after 1960 is shown in table I. (Ventriculoatrial shunts have been used in the hospital consistently from 1960 onwards.) The mortality rate since 1961 is similar to that in many reported series, and about 40% can be considered to be the average mortality rate of spina bifida cases taken as a whole.

As stated above, 79 (62 (78%) of whom had high lesions) of the 295 patients received no active treatment, and of these 77 died, so that death in this group is almost inevitable. Of the 216 patients selected for active treatment 47 subsequently died, a mortality rate of 21.8%. Because most of the children who received no active treatment had high lesions, however, those treated showed a preponderance of children with low lesions—129 (60%) with low lesions compared with 87 (40%) with high lesions. Altogether 18 (14%) of these children with low lesions died compared with 29 children (33%) with high lesions. These mortality rates represent the risk of death of those children treated and are helpful in discussing the likelihood of survival with the parents.

Cause of Death

The two most important causes of death in the total series were hydrocephalus and meningitis, each making up one-third of the total deaths (table III). Among the other causes of death, pneumonia was the most predominant. Six children died immediately postoperatively, five after repair of the

sac and one after an ileal conduit operation. One child dying of peritonitis had a Curling-type perforation on the curvature of the stomach near the pylorus with subsequent peritonitis.

TABLE III—Causes of Death in Patients with Myelomeningocele born during 1961-9

	No. in Total Series	No. of Patients Actively Treated
Hydrocephalus	43 (35%)	16 (34%)
Meningitis	43 (35%)	16 (34%)
Other*	38 (31%)	15 (32%)
Total	124	47

*Pneumonia 16 (4 patients actively treated), multiple anomalies 4, gastroenteritis 2 (1), renal failure 2 (1), septicaemia 2, prematurity 1, congenital heart disease 1, peritonitis 1 (1), atelectasis 1, postoperative 6 (6), not known 2 (2).

The causes of death in the 47 children born during 1961-9 who received active treatment are shown in table III. The causes of death in these patients show no significant differences from those for the total series.

Age at Death

The ages at death of children born during 1961-5 (providing a 7-11-year follow-up) showed a heavy concentration in infancy. Altogether 26 (37%) of the children were dead by the age of 1 month, 59 (83%) by 1 year, and 62 (87%) by 2 years. Seven out of nine deaths in children over 2 years were from hydrocephalus. No child dying of meningitis died after the age of 6 months (except in the 1961-9 series, where one child died of meningitis at 21 months).

Apart from the deaths (48) in children born during 1961-5 who were not offered active treatment 23 children died. The ages at death of these 23 children compared with the ages at death of the 48 children born in the same years and not offered active treatment (numbers in parentheses) were as follows: by the age of 1 month 6 (20), by the age of 1 year 15 (44), and by the age of 2 years 17 (45); 6 (3) children died over the age of 2 years. The later ages at death of the treated children can be seen; 15 out of 23 treated children (65%) dying before the age of 1 year compared with 44 out of 48 untreated children (92%).

FACTORS INFLUENCING MORTALITY

Neurological Level

One of the decisive factors in the mortality of spina bifida children is the neurological level of the lesion. The neurological levels and the outcome are shown in table II. From a prognostic point of view the distinction between high and low lesions is critical. The mortality of high lesions (60%) compares unfavourably with that of low lesions (24%).

Hydrocephalus

The neurological level also influences the incidence of hydrocephalus. In table IV the incidence is shown to fall with the lowering of the level of the lesion. In lower thoracic lesions the incidence is 86%. The mortality relative to the onset of hydrocephalus is shown in table V. The high mortality (78%) when hydrocephalus is clinically present at birth is very marked, and the mortality rapidly drops as the time of onset after birth is delayed. Table V also indicates when hydrocephalus can be expected clinically. In 26% (55/211) it was present at birth, mostly in children with high lesions; in 68% (144/211) it was present by the age of 2 weeks; and in 77% (163/211) it was present by the age of 1 month. In 96% (202/211) the hydrocephalus was present before 6 months of age.

TABLE IV—Neurological Level of Lesion and Incidence of Hydrocephalus in Patients with Myelomeningocele born during 1961-9

Neurological Level	No. with Hydrocephalus	Total
(I) Cervical and upper thoracic	1	2
(II) Lower thoracic	69 (86%)	80
(III) Upper lumbar	53 (79%)	67
(IV) Lower lumbar and upper sacral ..	81 (60%)	134
(V) Lower sacral	7	12
Total	211 (72%)	295

TABLE V—Onset of Hydrocephalus and Mortality Rates in Patients with Myelomeningocele born during 1961-9

Onset	No. Dead	Total
Present at birth	43 (78%)	55
Under 2 weeks	33 (37%)	89
2 Weeks and under 1 month	6	19
1 Month	7	24
2-5 Months	2	15
6 Months and over	0	9
Total	91 (43%)	211

Table VI combines neurological level and the onset of hydrocephalus and shows their influence on the mortality. The mortality rate for children with hydrocephalus and high lesions is 56%, compared with 25% for those with hydrocephalus and low lesions. The earliness of onset of the hydrocephalus compounds this effect, and if a child has a high lesion and hydrocephalus at birth the mortality is 84%.

TABLE VI—Neurological Level, Onset of Hydrocephalus, and Mortality Rates in Patients with Myelomeningocele born during 1961-9

Onset	High Lesion		Low Lesion	
	No. Dead	Total	No. Dead	Total
Present at birth	37 (84%)	44	6	11
Under 1 month	25 (46%)	54	14 (26%)	54
1 Month and over	7 (28%)	25	2 (9%)	23
Total	69 (56%)	123	22 (25%)	88

Of the 159 children born during 1961-5 112 (70%) developed hydrocephalus, and of these 72 (64%) required shunts. Most (34) of the remainder died early (all under the age of 1 year); treatment of these patients was not considered justifiable, and only a few (6), who had minimal and non-progressive hydrocephalus, did not have shunts and survived. Of the 72 patients 16 (22%) died after shunt operations; this refers to death after shunt operations from any cause and does not indicate the immediate operative mortality. It is interesting to note that 11 of the 16 patients died ultimately of hydrocephalus despite the shunts, the remaining five dying of volvulus, gastroenteritis, renal failure, pneumonia, or respiratory failure.

Sac Excision

The mortality related to excision or non-excision of the sac is shown in table VII. There was no significant difference in mortality whether the sac was excised under or over 48 hours after birth. In 81 cases the sac was not excised but in two of these excision was not necessary. Both children had low lesions with flat, healed sacs, were of normal intelligence, and attended normal schools. There was a very high mortality (98%) in the remaining 79 unoperated patients. This last group is a highly selected one, and indicates the predictable mortality when operation is withheld on grounds

of adverse criteria. These adverse criteria governing non-intervention in this series were the presence of very high lesions, early onset of hydrocephalus, infected sacs, ventriculitis, or the presence of another life-threatening disease. The two children surviving in this group had high lesions and were retarded, one with mild and the other with profound retardation.

TABLE VII—*Sac Excision and Mortality Rates in Patients with Myelomeningocele born during 1961-9*

	No. Dead	Total
Excised under 48 hours	30 (24%)	127
Excised over 48 hours	17 (20%)	87
Not excised—deferred	77 (98%)	79
Not excised—not necessary	0	2

Meningitis and Ventriculitis

Out of all the 295 children 52 (18%) developed meningitis and ventriculitis, but the mortality when contracted was very high (43/52; 83%). Some of the children developing meningitis, however, were not treated, so that it is likely that vigorous treatment, had this been indicated, would have reduced this mortality rate. Deaths from meningitis are closely related to sac excision. A much higher proportion of children developed meningitis when the sac was not excised—27/81 (33%) compared with 16/214 (7%) when the sac was excised. Of the 43 deaths from meningitis half occurred in children under the age of 1 month, and all patients except one were under the age of 6 months, the one referred to dying at 21 months.

SUMMARY OF FACTORS IN MORTALITY

Most children with spina bifida cystica have myelomeningoceles (94%). The mortality rate is higher in children with high lesions, and thus though the incidence of high and low lesions is almost equal at birth two-thirds of the survivors have low lesions, the lesions which give the best functional results. The two major causes of death are hydrocephalus and meningitis, about one-third each, and most deaths (87%) occur before the age of 2 years. Owing to the severity of the condition in this series, in more than one-quarter of the cases the sac was not excised and active treatment was not given. When such a selection has been made there is a very high mortality (98%). Three-quarters of the children develop hydrocephalus, and the incidence of hydrocephalus is marginally higher the higher the lesion. Three-quarters of the children develop hydrocephalus under the age of 1 month, and the earlier the onset of hydrocephalus the higher the mortality, being 78% for those with hydrocephalus present at birth. High lesions and the early onset of hydrocephalus compound the mortality—84% with a high lesion and hydrocephalus at birth. There is a 22% mortality after shunt operations, over half of the deaths being still due to hydrocephalus. Though the incidence of meningitis and ventriculitis is low (18%) it carries a very high mortality (83%), and almost all such children die under the age of 6 months.

Hence this series shows that there is objective evidence that high neurological level, the presence of hydrocephalus at birth, and meningitis and ventriculitis are very adverse factors in mortality. Other adverse factors are the presence of other severe anomalies, other severe diseases, and gross renal pathology.

Quality of Life

In this section we analyse 88 surviving children born during

1961-5 and who at the time of writing were aged 7 to 11 years. All but two uneducable children were attending school and the whole group was available for assessment.

The neurological level of the lesion, which is so important a factor in the mortality rate, is also significant in the quality of life of the survivors. The much higher mortality of children with high lesions compared with the mortality of children with low lesions has the important corollary that patients with low lesions make up the majority of survivors—64 out of 88 (73%) in the 1961-5 series. This clearly influences the quality of life and the survivors taken as a whole. The type of selection in spina bifida also has a marked influence on the neurological levels of the survivors. Before 1960, when very few children were actively treated, more children with high lesions died, and therefore among the survivors there was a higher proportion of children with low lesions. By contrast, when all cases are treated more children with high lesions survive, as in Lorber's (1971) series.

The level of neurological deficit is particularly important in relation to ambulation but also influences the mental state and subsequent schooling, primarily due to the fact that higher lesions are associated with a higher incidence of hydrocephalus than low lesions.

Quality of survival is discussed under the following headings.

AMBULATION

The extent of the splintage required for independent mobility is shown in table VIII. All children were ambulant except one severely retarded child. All children with high lesions (except one with a cervical lesion) wore calipers and many had pelvic and thoracic extensions to their calipers. Owing to the extent of their equipment all had limited mobility. Though all could walk to some degree, in this high group many were so slow or walking so laboured that about half were either using wheelchairs in the primary school or were expected to do so permanently as adolescents or young adults. By contrast 56% of the children with low lesions used splints, but these consisted only of short irons, and 41% could walk without splints. All the children with low lesions were actively mobile and none were likely to require a wheelchair unless there was some other unusual complication—for example, two children had severe mental retardation. As shown by table VIII, the level of the lesion is decisive on the quality of ambulation.

TABLE VIII—*Use of Splints by Patients with Myelomeningocele born during 1961-5*

	No. with High Lesions	No. with Low Lesions	Total
Calipers	23	1+	24 (27%)
Short irons		36 (56%)	36 (41%)
No splints	1*	26 (41%)	27 (31%)
Not ambulant		1+	1 (1%)
Total	24	64	88

*Cervical lesion.

†Severe mental retardation.

MENTAL STATE

The children are tested in three ways. (1) They are referred to the psychiatrist and clinical psychologist attached to the spina bifida clinic in the second year of life and are usually tested in the hospital two or three times in the preschool period. (2) In the year preceding school the children are tested by the educational psychologist of the State Education Department. (3) In addition to formal testing the children are observed in the clinic, and assessments have also been sought

from the teachers in charge of the preschool centres, and especially from the teachers in the special schools. This last information is helpful in checking the accuracy of formal testing, especially in checking academic achievement.

The children are classified into broad groups according to the International Classification of Disease—namely, normal, I.Q. 86 and above; borderline mental retardation, I.Q. 68 to 85; mild mental retardation, I.Q. 52 to 67; moderate, severe, and profound mental retardation, I.Q. below 51. The normal range is, however, divided into two groups classified as “high normal” and “low normal” in order to identify children who, though they can manage the normal school curricula, are unlikely to proceed beyond the primary grades or the first few forms of secondary school. These low normal children, who will not reach the upper grades of secondary school or tertiary education, have difficulties in achieving skilled occupations which may be desirable because of their physical handicaps.

The numbers of children at the various mental levels are indicated in table IX. Three-quarters of the children were within the normal range of intelligence, and half of these were likely to be able to manage the higher levels of secondary education; a few should achieve tertiary education. One-fifth of the children were mentally retarded but educable in special schools. Only two children (2%) had moderate, severe, or profound mental retardation.

TABLE IX—*Mental State of Patients with Myelomeningocele born during 1961-5*

Mental Level	No. with Shunt	No. with No Shunt	Total
High normal and above ..	17 } (66%)	21 } (97%)	38 } (77%)
Low normal ..	20 }	10 }	30 }
Borderline mental retardation ..	14 } (30%)	1 }	15 } (21%)
Mild mental retardation ..	3 }	3 }	6 }
Severe mental retardation ..	2 } (4%)		2 } (2%)
Total	56	32	88

Table IX also shows the distribution of intelligence in those children with shunts. Of the 88 children 56 (64%) had progressive hydrocephalus requiring ventriculoatrial or other shunts. Table IX indicates that almost all children who do not develop hydrocephalus or whose hydrocephalus is clinically insignificant and do not require a shunt are of normal intelligence. In those with shunts two-thirds were in the normal range and almost one-third were retarded but educable. There is therefore a significant difference in the intellectual outlook of children who require ventriculoatrial shunts.

Many children had multiple operations for shunts. Only nine of the 56 children did not require revision of the shunt apparatus, and the average number of revisions in the remaining 47 was two; 19 children had one revision, 15 two revisions, 6 three revisions, 4 four revisions, 2 five revisions, and 1 six revisions. The figures are too small for detailed analysis of the mental state in relation to the number of shunt revisions, but the records show that even after many revisions the children can still remain within the normal intellectual range.

It is difficult to find comparable series of unoperated cases of hydrocephalus in spina bifida in order to compare the results with those of modern treatment. The mortality rates have altered and so has the distribution of severe cases, so that the figures are not comparable. Two series have been reported by Laurence and Coates (1962) and Hagburg and Sjorgen (1966). The former dealt with 81 survivors of 182 unoperated cases of hydrocephalus of mixed aetiology; of the survivors 38% had normal intelligence, 35% borderline or mild retardation, and 27% moderate, severe, or profound mental retardation. Hagburg and Sjorgen, reporting on the chronic brain syndrome in hydrocephalus, studied 63 spontaneously arrested cases, also of mixed aetiology. Their results were

similar to those of Laurence and Coates, being normal 37%, borderline and mild mental retardation 38%, and severe mental retardation 25%. Even allowing for the difficulties in comparing these series with modern treated series it can be seen that the introduction of the ventriculoatrial shunts has greatly improved the mental status of the children.

Hagburg and Sjorgen (1966) described the clinical pattern of hydrocephalus, and our findings were similar—spastic paraparesis (above the level of the spina bifida lesion), ataxia, squint, mental retardation, and some behaviour deviations. The behavioural features found in some patients with hydrocephalus (25% of the patients in Hagburg and Sjorgen's series) are striking. The children are sociable and “pseudo-bright,” with an outgoing personality. They show constant chatter, described vividly by Ingram and Naughton (1962) as the “chatter-box syndrome” and by Hagburg and Sjorgen (1966) as the “cocktail party syndrome.” There is, however, poor understanding of the stream of words and the educational abilities of the children are below the levels of their verbal scores. We have also noticed with these children impairment of body image and spatial disorientation, such as confusion between right and left—for example, in dressing and in positioning skills—and the conception of the body in space—for example, in standing and walking. Teachers have also reported the increased tendency to reversal of letters and words and confusion of lines of print or diagrams.

These features which are seen in some children with hydrocephalus, especially those with borderline or mild mental retardation, have several pitfalls. (a) The child with the sociable, alert personality and with good speech misleads the parents, who are often difficult to convince that the child is not especially bright and may be retarded. (b) The high verbal scoring in testing and functioning in the preschool and early school years gives an overestimate of the child's ultimate educational ability as he moves to more abstract and formal thinking. On the positive side, the sociable personality is an advantage in social contacts at school, in employment, and later in integration into the community.

SCHOOLING

The distribution of children between normal schools and special schools for the physically handicapped is in table X. Altogether 39% of the children attended normal school, 59% attended special schools for the physically handicapped, and 2% were not educable. Many factors determine the placement of a child in a special school, the three main ones being the mental state, the presence of extensive splints, and bowel incontinence. If the urinary incontinence is adequately controlled by means of an ileal conduit diversion operation this is not a serious factor. Only three out of 23 children with high lesions and wearing calipers were being educated in a normal school compared with 31 out of 62 wearing short irons or not requiring splints. All the children in normal schools had normal intelligence except two with borderline mental retardation, who were being educated in country schools; by contrast 16 of the 52 children attending the special schools were retarded. There was also a different distribution between children with “high normal” and “low normal” intelligence; in

TABLE X—*Mental State and Schooling in 88 Patients with Myelomeningocele born during 1961-5*

Mental State	No. at Normal School	No. at Special School	No. Not Educable
High normal and above ..	25 } 32	13 } 36	
Low normal ..	7 }	23 }	
Borderline mental retardation ..	2 }	13 }	
Mild mental retardation ..		3 }	
Severe mental retardation ..			2
Total	34 (39%)	52 (59%)	2 (2%)

normal schools 25 of the 32 were in the "high normal" range compared with 13 out of 36 in the same range in the special schools.

RENAL TRACT

Urinary Incontinence

Only six children (7%), all with low lesions, were continent. These children did not have completely normal control but could hold urine over two hours, had awareness of the desire to micturate, and wore normal underclothes at school and at home. They occasionally had accidents, especially at night. It was considered, however, that their urinary control was socially acceptable. The remainder of the children (82; 93%) had some form of urinary collecting apparatus and all except five boys, who wore penile bags, had had an ileal conduit diversion operation. The use of an ileal conduit diversion bag is quite compatible with attendance at normal school and sporting activities, including swimming, and, in fact of the 34 children attending normal school 28 wore ileal conduit appliances, 2 wore penile bags, and 4 were continent. During the preschool period the children were taught to empty their bags, and all were able to do this by the time they entered school. The changing of the flange of the ileal conduit apparatus is a more difficult procedure and we have found that children can do this themselves at 10 to 12 years of age. It is our aim to have all children independent of the help of the parent with their urinary apparatus before they enter adolescence.

Smith (1972) reviewed 150 patients with ileal conduits and studied their prognosis in more detail. The most persistent problems affecting quality of life after ileal conduit diversions were peristomal ulceration, stomal stenosis, leakage at the apparatus, and discharge from the disconnected bladder. The worst results in terms of quality of care were in socially deprived families, indicating the necessity for careful and regular hygiene and supervision.

Renal State

During the first admission in the neonatal period intravenous pyelography and expression cystourethrography are performed, and these are repeated in the second year. Pyelography is repeated at about two-year intervals in the preschool period and at three-year intervals thereafter. In addition urinary culture is performed three-monthly before the ileal conduit operation and subsequently at least six-monthly. In the light of these investigations an assessment was made of the renal tract over a period of years and the presence or otherwise of any deterioration was noted. The children's renal state has been divided into three grades.

Grade 1 children were those who before treatment had a normal renal tract or insignificant abnormalities and who remained normal over the period of observation. This group made up 78% of the survivors and we expect that these would have a normal renal prognosis. *Grade 2* children were those who were normal before the conduit operation or had moderately severe abnormalities and showed slight deterioration. This group made up 14% of the children and we consider that these would have some risk of renal failure in adult life. *Grade 3* children were those who had all degrees of involvement of the renal tract from normal to severe abnormalities before the conduit operation and showed gross deterioration. This group made up 7% of the total and there was in these a severe risk of renal failure; we would expect this group to die of uraemia in adolescence or early adult life. There was inadequate information on one patient (1%). We would therefore expect that almost four-fifths of the survivors should have a normal renal prognosis (Smith, 1972). In the

present series there was no relation between the neurological level of the lesion and the ultimate prognosis for the renal tract.

BOWEL INCONTINENCE

Since the treatment of urinary incontinence has become more satisfactory bowel incontinence has become a more noticeable disability for these children. The problem has varied from frequent and extensive bowel soiling in some children to those who have only an occasional bowel accident, and this range of soiling is difficult to classify.

Children who have urinary continence all have bowel continence, and many children with bowel incontinence can achieve satisfactory toilet routines. In all about three-quarters of the children did not have any appreciable soiling that would interfere greatly with normal daily living. About one-quarter of the children did have some soiling, in a few cases severe. There were some children who had low lesions, normal intelligence, and good urinary care who were attending the special school solely because of bowel soiling. Fortunately all these children improve with age, and the problems of soiling are greatest in the preschool and early school periods, and in socially deprived families.

Discussion

The quality of survival in children of school age with high and low lesions is summarized in table XI. In quality of survival there was a definite difference between children with high and low lesions. Because of a higher incidence of hydrocephalus in patients with high lesions the proportion of children with normal intelligence was lower. Only 17% of children with high lesions were attending normal school, compared with 47% of those with low lesions. All the children with high lesions wore calipers and many had pelvic or thoracic extensions; about half either were using wheelchairs in primary school or were expected to use wheelchairs permanently as adolescents or young adults. By contrast only 56% of the children with low lesions used splints, and these consisted only of short irons; 41% could walk without splints. All were actively mobile and none were likely to require a wheelchair unless some other, unusual complication occurred. Almost all the children with lesions at all levels were incontinent and required urinary apparatus, only a few of those with low lesions (7%) being continent. Altogether 78% had normal renal function after conduits, and with this group we would expect a normal renal prognosis. Up to a quarter of the children had difficulties with urinary and bowel care, most marked in those with high lesions.

TABLE XI—Quality of Survivors with Myelomeningocele born during 1961-5

	High Lesion	Low Lesion
Normal mental state	63%	81%
Normal school	17%	47%
Splints	All	56%
Actively mobile	None	All
Wheelchair	About half	None
Urinary appliance	All	93%
Good renal prognosis	75%	81%
Urinary and bowel care	>25% poor	<25% poor
Expected normal employment ..	33%	66%

These children were not old enough to assess for employment but we would expect on the basis of our adolescent patients that not more than one-third of the patients with high lesions would be normally employable, compared with two-thirds of these with low lesions. The remaining two-

thirds and one-third respectively could be employed in sheltered workshops, with a small number being unemployable.

Lorber (1971) attempted to grade the degree of handicap into five categories. With minor modifications these are as follows. (1) *No handicap*: this comprises meningoceles (these are not further considered). (2) *Moderate handicap*: these children have normal intelligence, are actively mobile (no splints or only short irons), and have urinary continence or control with apparatus, good renal prognosis, and no other major handicaps; this is the most favourable group among survivors. (3) *Severe handicap*: children with normal intelligence but severe physical handicaps. (4) *Very severe handicap*: children with borderline or mild mental retardation and physical handicaps. (5) *Extreme handicap*: children with moderate, severe, or profound mental retardation and physical handicaps.

Table XII grades the surviving children born during 1961-5 into these categories; for comparison is the series reported by Lorber (1971) from Sheffield. In Lorber's series every child was treated actively. By contrast it has always been our policy at the Royal Children's Hospital to select certain patients for treatment and to defer others. Table XII shows the striking difference in the quality of survival resulting from this policy of selection. Particularly noticeable is the greater percentage in our series in category 2 and the greater percentage in Lorber's series in category 5. The policy of treating every child without selection appears to result in a large number of children with severe physical and intellectual handicaps surviving who will be a burden to themselves, their families, and the community for very many years.

TABLE XII—Classification of Patients with Myelomeningocele by Degree of Handicap

Category	Lorber (1971) 1959-63	Present Series 1961-5
2	20 (15%)	42 (48%)
3	66 (51%)	26 (30%)
4	28 (22%)	18 (20%)
5	16 (12%)	2 (2%)
Total	130	88

Selection for Treatment

"Selection" of patients for treatment is not a static or single decision made for all time. In our clinic most patients present as newborn infants, and the first decisions on selection are made on criteria presenting at that time. Once a decision not to treat at birth has been made, further criteria apply if the infant is continuing to survive, so that re-evaluation then becomes necessary.

In the light of the factors of mortality and the quality of survival the following are our recommendations for selection.

TREATMENT AT BIRTH

Low Lesions

We recommend an immediate repair of the sac on all low lesions, except in infants with hydrocephalus clinically present at birth, gross anomalies, or life-threatening disease or who present a few days after birth with an already infected sac. Low lesions without these complications augur almost certain survival and a good prognosis, and maximum treatment is offered.

High Lesions

We would defer treatment of all high lesions on this criterion

alone with or without other associations. About one-quarter of these children will die under the age of 1 month.

At birth spina bifida admissions are almost equally divided between high and low lesions (table II). About one-tenth of the children with low lesions have contraindications to early surgery, so that about 90% of children admitted with low lesions and 45% of all children admitted have early surgery.

TREATMENT AT 1 MONTH

During the first month about one-third of those whose sac has not been excised will already have died. In the survivors re-evaluation will be necessary and is made along the following lines.

Low Lesions

During this month some patients whose back lesions have been repaired will develop hydrocephalus; a decision will be made to perform a shunt operation or to keep under observation depending on neurosurgical principles of the rapidity of progress of the hydrocephalus and its type.

In those whose surgical treatment was deferred at birth, and who have overcome their severe associated diseases, surgical treatment is given in the appropriate order for the sac and (if present) the hydrocephalus. We would still, however, defer any active treatment in those whose hydrocephalus had its onset at birth, in those with severe and rapidly fulminating hydrocephalus, and in the continued presence of meningitis and ventriculitis, gross anomalies, and life-threatening disease. Most of these patients will die within the following few months.

High Lesions

We would still exclude from active treatment those exceptions listed above; the presence of severe kyphosis is an additional contraindication to treatment. With these exceptions, however, survival at this stage often indicates permanent survival, and surgical treatment cannot reasonably be withheld; as most patients will have hydrocephalus this assumes the greater priority, and thus shunting is performed before back repair.

TREATMENT LATER IN INFANCY

Except for those with an obviously hopeless prognosis only a few children will not have begun treatment at this stage. Treatment may be started at any time in those children who are continuing to survive despite predictions to the contrary.

TREATMENT AT LATER AGE

There is one further group, a very small one. There are surviving children who at any age have severe or profound mental retardation with physical handicaps and then develop meningitis and ventriculitis, shunt blockage, or a life-threatening disease. The prognosis for such children is so poor that serious consideration should be given at that stage to withholding active treatment.

We have deliberately excluded the question of a neurogenic bladder and bowel. Since all patients with high lesions and 93% of patients with low lesions will have incontinence the presence of the signs of incontinence in the newborn period cannot be used as a criterion in selection. Gross renal disease is usually not diagnosed until after the newborn period becomes a factor in selection, as one of the adverse prognostic features, only at a later age.

Discussion

Three questions about this recommended programme may be asked.

(1) Does deferring the sac excision worsen the neurological state? Many studies have been made on this subject since the original claim by Sharrard *et al.* (1963) that there is less neurological involvement if the sac is repaired immediately after birth. It is true that excess drying of a sac and the secondary complication of meningitis may worsen the neurological state and that the incidence of these may be diminished by immediate repair. It is for this reason that we advocate urgent repair in low lesions where the prognosis is so favourable. But we have never seen the neurological status of a high lesion change to that of a low lesion by immediate repair; any minor changes in muscle tone and reflexes observed after repair are of such a marginal nature that the basic prognostic distinctions between high and low lesions still persist. We cannot confirm a major change of status as reported by Sharrard *et al.*, and thus we are prepared to accept the risk of worsening complications in unoperated high lesions in the first month of life rather than offer immediate treatment to this group, whose ultimate outlook is so unfavourable even with treatment. Nor has sac excision any significant influence on the development of hydrocephalus. The only danger of deferring sac excision is the increased risk of meningitis, as indicated above.

(2) Does the delay until 1 month of age or later in the treatment of the hydrocephalus have an adverse effect on the ultimate mental state? In principle the sooner the hydrocephalus is relieved the better, but shunt operations before 1 month of age have a high incidence of complications, especially blockage of the proximal tube and the development of shunt dependence; it is the general view of neurosurgeons that where possible shunt operations be deferred until 4 to 6 weeks of age. In table XIII the mental state is analysed in relation to the interval between the onset of the hydrocephalus and the time of the shunt operation. The figures are small but indicate that at all intervals, except, perhaps, when greater than six months, the proportion of retarded children remains constant. It seems, therefore, that a short delay in the introduction of a shunt is not likely to have an adverse effect on the child's ultimate mental state, except in rapidly progressive hydrocephalus; on our criteria such as infant will be excluded from treatment in any case.

TABLE XIII—Mental State of Patients with Myelomeningocele born during 1961-5 correlated with Onset of Hydrocephalus and Time of Shunt Operation

Interval	Mental State		Total
	No. Normal	No. Retarded	
1-2 Weeks	18	8	26
3-4 Weeks	6	3	9
5-6 Weeks	3	1	4
7-8 Weeks	1	1	2
2 Months	6	3	9
3 Months	2	1	3
4 Months			
5 Months	1		1
6 Months and over		2	2
Total	37 (66%)	19 (34%)	56

(3) How long will untreated patients survive? As indicated in table VII, out of 295 children admitted 79 (27%) received no active treatment because of an adverse prognosis. Thirty-seven of these children (47%) were dead by the age of 1 month, 55 (70%) by 3 months, 66 (84%) by 6 months, 71 (90%) by 12 months, and 74 (94%) by the age of 2 years. Three children (4%) died over the age of 2 years (2 year 3 months, 2 years 6 months, and 5 years 1 month, respectively) and two children (2%) survived. The mean age at death was 19

weeks and the median age at death 5 weeks. Hospital accommodation, within either the specialist hospital or the local hospital, is required for these children, if not throughout then certainly in the terminal stage. This must, however, be compared with the far greater number of bed days and the tremendous use of staff time and hospital and community facilities which are involved if children with gross lesions are treated and survive over a period of years, necessitating many readmissions and continued care.

It is difficult to decide whether to introduce a shunt into the severely hydrocephalic child solely to improve the cosmetic appearance and to make the situation less distressing for the parents. The procedure has been done with a few patients and though most of these eventually died the length of life has been greatly extended and the occasional survivor has been left with very severe mental and physical handicaps. In general it is probably more humane and easier for the parents to allow the children to die more quickly without operative treatment for the hydrocephalus.

These findings are similar to those of Hide *et al.* (1972) in a series of 99 children for whom early surgery was considered inadvisable. By the end of one year 92 and the 99 children were dead. Only four children attained their second birthday and only two survived beyond that age, and in both of these the quality of life was poor. We agree with their conclusion that "a policy of careful selection of the most severely affected infants is unlikely to result in the survival of large numbers of extremely handicapped people."

The recommendations on selection for treatment are similar to those advocated by Lorber (1971, 1972) and accord with the main principles set out by Matson (1969). Matson wrote just before his death: "In this Clinic it is not customary to operate upon newborn infants or those in the first few months of life with thoraco-lumbar or upper lumbar lesions who exhibit total paraplegia and complete sphincter paralysis; this is true whether or not there is significant hydrocephalus at the time. However if there is partial motor function of the lower extremities and it seems apparent that the child may well become ambulatory independently then every effort is made to repair the myelomeningocele as soon as other factors would appear to make it possible." This view was also advocated by Smith (1965).

Two general points concerning treatment may be added. Firstly, assessment and selection should be carried out in the spina bifida units in the major hospitals. We generally advise the local medical officers to refer the children to a spina bifida unit immediately after birth unless the medical practitioner considers that death is likely to occur in the first 24 hours. Selection is difficult and can be done satisfactorily only by the co-ordinated efforts of specialists with extensive experience in this condition. Delay or non-referral based on an inexperienced opinion that "treatment is not worth while" can lead to poor-quality survival when with treatment the prognosis could have been good.

Secondly, parents of spina bifida children always require a great deal of discussion, counselling, and help, and this is particularly so when no active treatment is to be given. The parents have an extremely difficult period during the months when the child is gradually deteriorating and there is an obligation on the staff to give the necessary support and help to these parents until the death of the child occurs. Many children die within the first month of life but some will survive for up to a year, and a few children will survive longer or even permanently. The clinician must discuss the prognosis with the parents, based on predictable medical facts, simply, honestly, sensitively, and without dogmatism, and be flexible enough to change the treatment programme with the changing status of the child. Whether the child remains in hospital or goes home also requires discussion, but if the child goes home terminal care in hospital must always be available if the parents desire it. The quality of a unit depends just as much

on its care of a child with a hopeless prognosis as on its success with the survivors.

The feasibility and desirability of treatment of a grossly handicapped infant, especially in the neonatal period, is one of the most difficult decisions in paediatrics. Many factors are involved and the varying attitudes of individual surgeons and physicians and the pressures from parents accentuate the difficulties. For centuries the mainstream of medical opinion has abhorred the extremes and has trod the middle path. This was expressed in classical form by the nineteenth century English poet Arthur Clough (1819-61):

Thou shalt not kill; but need'st not strive
Officiously to keep alive.

This attitude has been re-enforced by a letter written by Cardinal Jean Villot (1970), Vatican Secretary of State, to the International Federation of Catholic Medical Associations. After stressing the sacred character of life that forbids a doctor to kill and that imposes on him the duty to use all the resources of his profession to fight against death, the Cardinal writes: "But for all that, a medical man does not have to use all the techniques of survival offered by a constantly creative science."

There will always be differences of opinion between individual physicians and surgeons whether in a particular child active treatment is justified or not, but we believe that complete rejection and complete acceptance for treatment in spina bifida are both untenable—the former denies active treatment where the prognosis and the results of treatment are good and gives the survivors less than an optimum future; the latter

keeps alive grossly handicapped children. Reasonably reliable objective criteria are now available for selection for treatment—their application has a marked effect on the quality of survival.

We wish to record our thanks to the following members of the spina bifida team who have worked with us in the clinic since 1961; Mr. R. S. Hooper, neurosurgeon, and his staff; Mr. P. F. Williams and Mr. M. B. Menelaus, orthopaedic surgeons; Dr. E. Wann, psychiatrist; Miss Ruth Magnus, assistant urologist; and a number of medical social workers, appliance sisters, and paramedical staff. All have contributed in various ways and have given freely of their knowledge and experience. The teachers in the preschool centres and special schools for the physically handicapped and, not least, the parents of the children have also provided essential information. To all we are greatly indebted.

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Predictive Factors in Open Myelomeningocele with Special Reference to Sensory Level

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Summary

A total of 113 cases of open myelomeningocele operated on shortly after birth were followed up and the 80 survivors (71%) were assessed one and a quarter to seven and a half years later. Their disability was classified in terms of mobility, intelligence, continence, and major complications; these when combined provided an assessment of overall disability. The overall disability of the survivors was minimal in 6%, moderate in 40%, severe in 39%, and very severe in 15%.

A number of clinical features present at birth were analysed for their predictive value. Of these the sensory level, which frequently differed from both external and radiological levels of the lesion, correlated with the outcome in terms of mobility, intelligence, continence, major complications, and overall disability; and also with deaths caused by renal failure.

A policy of confining operation to those patients with a reasonable chance of achieving independence would involve selecting for treatment a minority of all infants born with open myelomeningocele.

Introduction

In the past few years there has been a change in the climate of opinion, away from routine operative treatment in all cases of myelomeningocele (Lorber 1971, 1972). At the same time it has been confirmed that the most severe cases, if not operated on, rarely survive for long—some 90% dying before the end of the first year (Hide *et al.*, 1972; Smith and Smith, 1973; Stark and Drummond, 1973). A policy of selection of cases for operation, therefore, seems to be indicated and attention needs to be directed towards criteria by which cases likely to benefit by surgery—in the sense of achieving a reasonable degree of independence—can be more accurately identified at the time of birth or soon after.

In this paper some predictive factors are examined. The social consequences of this condition will be discussed in a separate paper.

Subjects and Methods

This survey covers 113 consecutive cases of open myelomeningocele.

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