

## THE DECISION TO TREAT MYELOMENINGOCELE ON THE FIRST DAY OF LIFE\*

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### SUMMARY

*An analysis of the factors concerned in deciding to treat myelomeningocele on the first day of life is presented. The results of active treatment in all cases are presented, including the results obtained at the Transvaal Memorial Hospital for Children. These are compared with the results of withholding treatment in all cases. The merits of these forms of management and selection of cases for treatment are discussed.*

Myelomeningocele (spina bifida aperta) is the commonest surgical congenital abnormality encountered on the first day of life. The incidence of the abnormality varies in different countries. An incidence of 4:1 000 live births is reported in Wales and parts of England<sup>1</sup> and probably about 2 500 children with the condition are born annually in Great Britain.<sup>2</sup> Most other countries have a slightly lower incidence, i.e. 2-3:1 000. The incidence in the Republic of South Africa is not known. Since a genetic factor as yet undetermined appears to play a role in the causation of at least a proportion of cases of spina bifida, it would appear that the condition is likely to increase in incidence in the future.

Myelomeningocele is a catastrophic abnormality and exists in a minor form in only a very small proportion of cases which suffer little disability. The child born with the usual major form may die from sepsis or central nervous system infection due to the lesion being uncovered by skin whether closure is undertaken expeditiously or not. Should the child survive, it is likely to develop hydrocephalus in a high proportion of cases (80-90%)<sup>3</sup>—probably unrelated to whether closure of the initial lesion was performed or not.<sup>4</sup> The hydrocephalus will be aggravated by associated central nervous system infection. Mental deficiency may occur as a result of the hydrocephalus.

All children with myelomeningocele will be paraplegic to a greater or lesser extent depending on the level of the lesion. Unfortunately low lesions, e.g. sacral, while giving rise to orthopaedic problems which are relatively minor and involving only the feet, frequently produce the worst bladder lesions due to cauda equina paralysis.

The child who is expeditiously treated from the first day of life and who survives will not have a tumour on his back and, with adequate treatment, should not have a large head. Surgical treatment for the hydrocephalus will be required in 25% - 50% of the survivors. Severe mental retardation is not common but the average intelligence of children with treated hydrocephalus is likely to be lower than that of the average normal child. Though the child is paraplegic, with modern orthopaedic treatment deformities can be corrected and he should be able to walk with the aid of calipers and crutches and not be condemned to a wheel-chair existence, provided he is intelligent.

Adequate urological care should ensure that renal function will be preserved. However, a small proportion of patients will succumb to renal failure though seldom before the age of 15-20 years. The child will be incontinent of urine which is easier to contend with in the male with appropriate appliances than in the female. About 33% - 50% of the children will require urinary diversion, e.g. ileal conduit, for prevention of renal failure or maintenance of socially acceptable continence, the latter especially in females. Rectal continence is seldom a severe problem, being managed by daily evacuation of the bowel by appropriate use of suppositories, bowel wash-outs or laxatives.

At best the child born with myelomeningocele constitutes a severe burden to his family and the community. The mother not only will be forced to spend a large proportion of her time in the care of the child but also frequently lavishes excessive love on it with frequent neglect of its siblings. Unstable marriages may break up under the burden of a handicapped child.

These children nearly always require special schooling in centres where rehabilitation facilities are provided. It is likely that a large proportion of them will require sheltered employment and institutions where they can live once their families are no longer able to care for them. The males are impotent and neither they nor the females are likely to marry. Hospital facilities are strained in the care of these children since they frequently require multiple admissions for their neurosurgical, orthopaedic and urological problems.

It is therefore mandatory that considerable care be given to the decision to treat these unfortunate children on the first day of life and it is the purpose of this article to analyse the factors in this decision.

### Methods of Management

There are 3 methods of management available to the surgeon confronted by infants born with myelomeningocele:

1. Immediate closure of the spinal defect in all cases with continued aggressive treatment of the hydrocephalus (if progressive) and maximal rehabilitation of the orthopaedic and urological disabilities.

2. All infants with myelomeningocele are left untreated in the hope that they will soon die.

3. An attempt is made to select the infant deemed to have a good prognosis regarding both disability and survival and he is treated aggressively. Infants considered to have a poor outlook are not treated. It might also be decided to observe the infant's progress for a period of time, i.e. weeks or months, before making a final decision regarding treatment.

In deciding which of these three courses to take, the results of each should be known.

### RESULTS OF AGGRESSIVE TREATMENT IN ALL CASES

For the past 8 years (June 1962 - June 1970) all infants with major myelomeningocele admitted soon after birth to

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the Transvaal Memorial Hospital for Children, Johannesburg, received active surgical treatment, with the exception of those with severe associated abnormalities incompatible with survival—e.g. severe cardiac anomalies, rectal agenesis, vesico-intestinal fissure (5 cases)—who received no treatment. All these latter patients died. Of the 54 patients treated, 49 underwent closure of the spinal lesion on the first day of life. Five patients admitted from the 2nd to the 5th day of life had the spinal lesion closed on the day of admission but are included in this series. Twenty-nine patients were males and 25 females. There were 43 lumbar lesions, 9 sacral and 2 thoracic. Seven patients were admitted with the sac already ruptured and 5 infants weighed under 2.5 kg.

Closure of the spinal lesion was performed by 'decapting' the sac followed by dural suture. Where the defect was large, skin closure was achieved in most cases by extensive mobilization to the flanks with or without release incisions. A 2-layer closure was attempted in all cases except in sacral lesions where lumbar fascia is not available. Prophylactic antibiotics were used as a routine initially but were discontinued later in the series with no change in the sepsis rate.

The decision to undertake drainage of the hydrocephalus was taken on head size and ventriculography was not performed as a routine. If head-growth was deemed ab-

normally rapid (usually during the first 4 weeks of life) ventriculo-atrial shunt was performed using a Pudenz-Heyer valve provided the back lesion was healed and the CSF not infected. If the CSF protein was markedly raised or obvious infection was present, drainage was instituted by ventriculostomy catheter only and appropriate antibiotics were given until the CSF cleared and shunting was permissible. During the past 2 years ventriculoperitoneal shunts have been used when the CSF has been mildly infected or the protein raised. Ventriculo-atrial and ventriculoperitoneal shunts have required frequent revision for complications.

The orthopaedic and urological management of these patients will not be commented on here since it has no material bearing on the survival and intelligence of these patients and will be subject of a further communication.

Thirty of the 54 children in the series have died (mortality rate 55.5%). The age at death is given in Fig. 1. It will be noted that all patients who died, did so before the age of 2 years. This compares well with other series and indicates that the child who survives 2 years is unlikely to die from the effects of myelomeningocele unless renal failure supervenes, usually at a much later date, i.e. 10-15 years.

The deaths may be divided into 3 groups (Fig. 1). Those within the first 7 days of life were due to various post-

operative complications following spinal closure. Those from 2 weeks to 3 months were due to sepsis and central nervous system infection and those from 5 to 18 months were due to complications associated with ventriculo-atrial shunting, either sepsis or blockage of the drainage system. Six of these last deaths were in patients admitted *in extremis* from outlying areas. They constitute failures of adequate supervision and should have been avoidable. Since insistence on regular follow-up since 1967 no similar deaths have occurred.

These 6 deaths have caused a loading of the mortality rate which, had they not occurred, would have been 44.4%. This is in keeping with the mortality rates in most centres which have treated myelomeningocele energetically. Some centres claim an even lower mortality rate, e.g. Liverpool (39%)<sup>5</sup> and Sheffield (44%).<sup>6</sup> From a survey of the literature it would seem that an average mortality rate of 40-45% can be expected in most institutions.

The intellectual quality of the survivors in this series is summarized in Table I. The age at follow-up varies from 8 months to 8 years with only 2 children under 1 year.

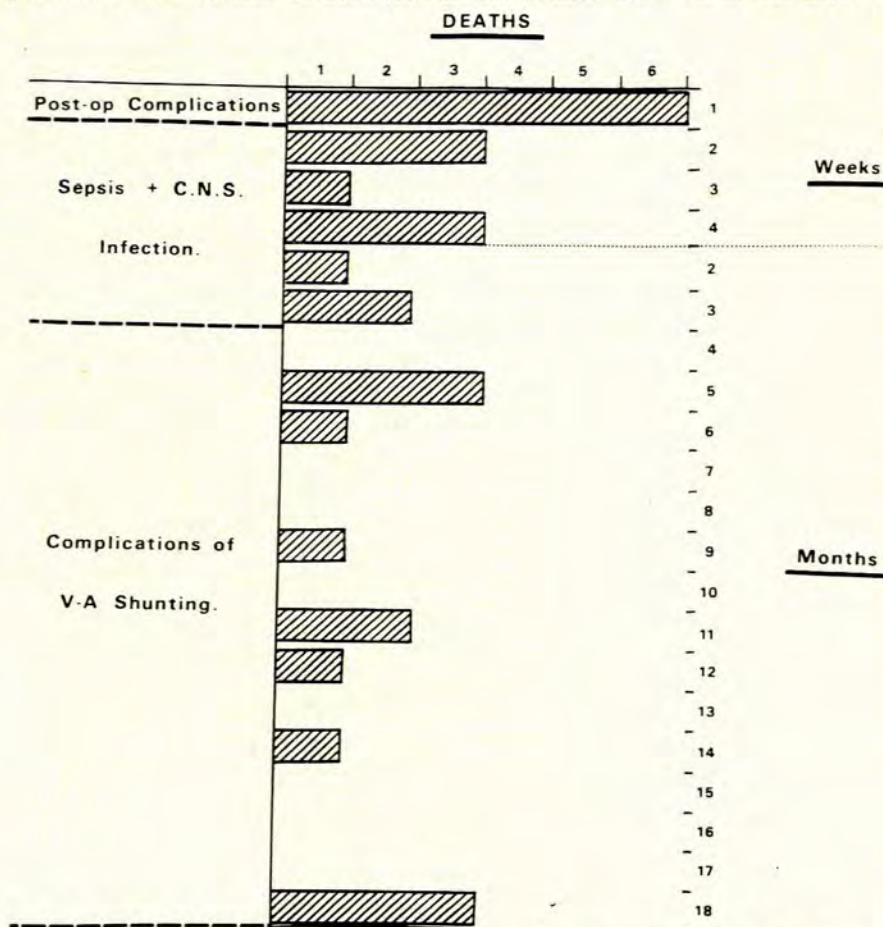


Fig. 1. Age at death of infants treated for myelomeningocele on first day of life.



TABLE I. INTELLECTUAL QUALITY OF SURVIVORS

	Intelligent	Educationally subnormal	Vegetative
V-A shunt .. ..	6	4	2
No shunt .. ..	9	1	2
Total .. ..	15	5	4

These last 2 children appear particularly intelligent even at this age and it is probably fair to include them as 'good' results intellectually. Detailed intelligence testing has not been performed in all cases and the results regarding intelligence have been assessed empirically by the members of the follow-up clinic and the staff of the special school which they attend. They are considered reasonable assessments as far as educability is concerned.

It will be noted that the children not requiring ventriculo-atrial shunting fared better intellectually than those requiring it, and this is common experience. It is accepted that children with significant hydrocephalus, whether treated or not, will have an average lower intelligence than those where hydrocephalus is mild or absent.

Though all the survivors have urological and orthopaedic handicaps of varying severity, it is accepted that they will all be incontinent to a certain extent (even if urinary diversion is performed) and all will walk with the aid of appliances if sufficiently intelligent. The yardstick by which they should be judged is their intelligence and educability, since this will determine the extent to which they can cope with their urological and orthopaedic disabilities and whether they can learn skills to enable them to lead useful lives in the future.

It will be noted that 4 children (16.6%) are vegetative and will require permanent care in mental institutions. In 2 of these cases severe sepsis prevented adequate ventricular drainage at an early stage and their condition can be considered potentially avoidable. Five children (20.8%) are considered educationally subnormal though attempts are being made to educate them and perhaps they will learn some useful skill. The remainder (62.5%) are considered of good intelligence.

The levels of intelligence in this series are not as good as those achieved in some other centres, e.g. Liverpool.<sup>5</sup> Possibly their poorer quality is due to a high sepsis rate and a rather conservative attitude to ventriculo-atrial shunting early in the series. This will be discussed in a further communication. In a Liverpool series<sup>5</sup> 7% of the children were vegetative and 10% educationally subnormal and 83% of the children were of good intelligence. These results are considered exceptional and in average hands the results can be expected to be slightly inferior to these.

#### RESULTS OF WITHHOLDING TREATMENT IN ALL CASES

Laurence,<sup>2</sup> analysing a series of 425 children with spina bifida and encephalocele in South Wales who received no treatment whatsoever from birth, found 36 survivors in the group with myelomeningocele after 2 years (mortality 85%). The survivors were found to have severe handicaps including large heads and persistent tumours on their backs. The IQ was over 85 in only 4% of the survivors.

In contrast to this, Findlay<sup>7</sup> analysed a series of cases from Glasgow and noted that, of a series of 354 children with myelomeningocele, 118 were not considered suitable for treatment and received no therapy at all. After 3 years

30.5% of this group were still alive. No mention is made of their condition but it is likely that they were similar to the group of survivors in Laurence's series.<sup>2</sup>

#### RESULTS OF SELECTION OF CASES FOR ACTIVE TREATMENT

No series in which selection of cases for treatment at birth was practised is available which compares with those cited above. Some authors,<sup>4,8</sup> state various criteria in their decision to treat from birth and some<sup>9</sup> practise a period of observation to determine whether treatment is worth while. However, no author is able to offer a comprehensive follow-up as regards survival and disability on all cases referred from birth which were either selected or rejected for treatment.

#### DISCUSSION

Though it is the doctor's duty to save life, he is entitled to use his discretion in not prolonging severe suffering under special circumstances. It has been mooted that it is not only ethical, but it may be desirable, that children with myelomeningocele should not be treated and should be allowed to die to save themselves, their families and the community the burden of their severe disability.<sup>10</sup> This view might be tenable were it not for the fact that not all children with untreated myelomeningocele die in infancy. Laurence<sup>2</sup> found that 1/6 of his series survived but Findlay's<sup>7</sup> figures suggest that Laurence's experience is not universal and that one-quarter or even one-third might survive with severe physical and mental handicaps.

It must be stressed that it is impossible to predict at birth which of these untreated cases will survive. Should the patient live, then the best chance of salvaging his disabilities will have been missed. Sharrard *et al.*<sup>11</sup> and Brocklehurst *et al.*<sup>12</sup> have shown that failure to close the spinal lesion expeditiously may frequently result in deterioration in the extent of the paraplegia. (Sharrard *et al.*<sup>11</sup> showed that early closure may actually improve the initial state of paraplegia but this is controversial.) Also, the cerebral damage caused by hydrocephalus is probably maximal at an early stage, i.e. within the first few weeks, and the opportunity for remedying this will have been missed by the time that it is assumed that the child will survive. Apart from these major considerations, two ethical problems are apparent: Firstly, with the rapid advances in urology and orthopaedics is it not possible that the residual disabilities may be so considerably reduced in the future that treatment may become much more worth while than it is at present? Secondly, does the doctor not expose himself to a charge of negligence if the parents construe that he has not done the maximum to both save and rehabilitate their child?

Though not applicable to the individual, it might also be argued that considerable advances in knowledge have accrued in the management of hydrocephalus and childhood paraplegia as a result of the continued treatment of myelomeningocele and this has been of benefit not only to children with this condition but to those with related disorders.

It would therefore appear that active treatment should be attempted in all cases unless the child has associated abnormalities which would preclude survival. This approach has the advantage that the doctor and the parents will feel that the maximum has been done to save the



child's life and to rehabilitate it. Naturally, depending on the ultimate disability, this may eventually prove not to have been the best course of action and disappointing results must be accepted. It is impossible to predict the reaction of the parents and siblings to the burden of a handicapped child. However, this approach is likely to be preferable to that in which the handicapped child was expected to die but did not and a lingering doubt exists as to whether alleviation of the disability could not have been achieved with active treatment.

From the practical point of view, active treatment does at least ensure that, even if the child should eventually die, the mother will not have to nurse a child with a large tumour on its back, requiring special care and positioning, and a head which she watches steadily increasing in size.

It is implicit in recommending active treatment that facilities must be provided for maximal rehabilitation. This implies regular follow-up at a multidisciplinary clinic including neurosurgical, urological and orthopaedic facilities. An ophthalmologist must be available for the treatment of squints which are common. A social worker, physiotherapist and occupational therapist will also be necessary. Special schooling must be provided, preferably at an institution where physiotherapy and basic nursing care are available. With increasing numbers of children with myelomeningocele surviving, these facilities will have to be provided on an ever-increasing scale including ultimate opportunities for employment and housing.

It must be emphasized that if facilities for maximal rehabilitation cannot be provided, e.g. in underdeveloped areas and in underprivileged groups, then it is likely that it is best not to treat myelomeningocele at all in the hope that few will survive and the families of these children and the community will bear a minimal burden.

The place for selection of cases in deciding on active treatment is, as yet, uncertain. Several criteria are quoted as precluding active treatment.<sup>4,5,9</sup>

1. *The child born with hydrocephalus.* Two children in the present series born with obvious hydrocephalus are now highly intelligent and it is clear that the presence of hydrocephalus at birth does not imply that cerebral function is beyond salvage.

2. *Rupture of the myelomeningocele at birth.* Seven children with rupture of the sac at birth were treated in the present series. Three died from meningitis but 4 survived. Without treatment, rupture of a myelomeningocele does not necessarily imply certain death from infection and prompt treatment may frequently forestall meningitis.

3. *Paraplegia.* All children with myelomeningocele have some degree of paraplegia but its extent is notoriously difficult to gauge on the first day of life. Sacral defects, though producing fewer orthopaedic problems, invariably produce the worst urological lesions and are therefore not necessarily more favourable than higher defects. Extensive thoracic defects do not produce significantly worse disabilities than high lumbar defects and these children may still walk with supports and crutches. One child in the present series with a thoracic defect is one of the best results as regards walking. Lower limb deformities, including dislocated hips, are correctable with modern orthopaedic care and should not preclude active treatment.

On the above criteria it would appear that it is not

possible to exclude cases for active treatment on the first day of life. However, it has been suggested<sup>5,9</sup> that a limited period of waiting is permissible to assess whether the child will either die, develop hydrocephalus or suffer deterioration in the extent of the paraplegia. This approach does not seem tenable. Firstly, as stated previously, should an untreated child survive, the best opportunity for preserving cerebral and lower limb function has been missed by not providing early treatment. Secondly, if an untreated child should survive and develop hydrocephalus, ventriculo-atrial or peritoneal shunting is likely to fail due to sepsis in the presence of an unhealed spinal lesion. Thirdly, regarding the paraplegia, it is probable that not only would delay cause deterioration in its extent but infection of the open lesion is certain to occur and late closure is very likely to result in wound infection with breakdown and secondary central nervous system infection. Delayed selection of cases for treatment is therefore not permissible.

Despite the difficulties of selection of cases for treatment, it is likely that the ultimate solution to the problem of deciding to treat children with myelomeningocele may depend on the ability to determine on the first day of life who will survive without treatment and to treat only those patients. It is important that large series of treated and untreated cases, fully documented, should be collected from many centres and the results correlated in the hope that such selection will ultimately become possible.

#### CONCLUSIONS

1. Since some children with untreated myelomeningocele do not die, and it cannot be predicted at birth who will survive, it is not permissible to leave all cases of myelomeningocele untreated.

2. It is not only impossible to predict the likelihood of survival at birth but also mental and physical disability, and therefore selection of cases for treatment is not acceptable. Because the best chance of early salvage of the disability might be missed, delayed selection of cases is also not permissible.

3. It is suggested that all cases of myelomeningocele (except those mentioned under 4 and 5 below) should be treated actively from the first day of life. This will, in average hands, produce a survival rate of 55-60% and about 75-85% of the survivors should have adequate intelligence for schooling. All will have orthopaedic and urological disabilities of varying degree but most of them should walk (with calipers and/or crutches if necessary) and their urinary incontinence should be made socially acceptable by appliances.

4. If a child with myelomeningocele has a major associated abnormality incompatible with life, unless treated surgically, it is suggested that treatment of the myelomeningocele and the associated abnormality should be withheld.

5. Should facilities for future rehabilitation not be available, then it is suggested that active treatment should not be undertaken.

6. It is suggested that the ultimate solution in the decision to treat myelomeningocele may lie in the ability to predict survival and disability accurately at birth but this is not possible at the present time.



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