STUDIES IN SPINA BIFIDA CYSTICA

II WHEN TO REPAIR THE SPINAL DEFECT

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In a recent survey of the problem of spina bifida cystica (Doran and Guthkelch, 1961) it was suggested that developments in the management of hydrocephalus and of paraplegia had improved the outlook in the grosser forms of spina bifida to the point at which it may not be justifiable to regard any but a very small minority of patients as incapable of survival. If this is the case, then it is of some importance to establish the optimum period of life for spinal repair, and also whether there is any time at which its performance carries any special risk.

Concerning operation in the neonatal period, the generally accepted view is that of Ingraham and Matson (1954), who say: 'There is seldom necessity for [this] . . . [except] when the covering membrane is thin, ulcerated or ruptured but uninfected, the lesion is small enough to make repair feasible, the head is normal in size, and there is no neurological deficit', provisos which would restrict operation to cases of simple meningocele and to myelomeningocele without neurological signs or hydrocephalus. From time to time, however, it has been suggested that early repair might be of benefit in cases of complete rachischisis (myelocoele) with partial or complete paraplegia on the ground that closure of the skin defect and return of the spinal cord rudiment to its natural environment within the theca would prevent infection and consequently further destruction of an already inadequately developed part of the nervous system. When spina bifida is produced experimentally in the rat, the exposed neural plate is actually undermined and cast off during the post-natal period by the ingrowing skin (Warkany, 1960), and such a process, if it occurred in the human subject, would necessarily stop transmission across the affected area of any nervous activity which had been present at birth.

On the other hand, it might be argued that manipulation of the delicate tissues of a myelomeningocele at so early an age could lead to further damage, whereas some delay in operative treatment implies a relative decrease in the size of the lesion to the size of the child and therefore facilitates closing the skin without undue tension

or the necessity to fashion extensive skin flaps. Also, to delay operation until the infant is between 3 and 12 months of age gives 'sufficient time for the development and recognition of neurological disabilities, for detection of hydrocephalus, for establishment of some regularity of bowel and bladder habits, for growth of skin, and for the child to develop into a better operative risk' (Ingraham and Matson, 1954).

These points, however, are not beyond challenge. It would be wrong to allow 'time for the recognition of neurological disability' if during that time the disability were increasing for want of treatment; 'time for the detection of hydrocephalus' is of little importance if this complication can be treated whenever it is detected, and recent contributions to anaesthetic and resuscitative technique make operations on the newborn infant less dangerous than they were.

So, in an attempt to establish whether under present conditions the previously accepted rules of management still apply, a further analysis has been made of some of the data which formed the basis of the previous study (Doran and Guthkelch, 1961). Particular attention has been paid to the influence upon the operative results of the complications of infection of the surface of the sac and of cerebrospinal fluid leaking from it, and to whether the infant's age at the time of repair has in itself any significant effect on the prognosis.

RESULTS

REPAIR OF SPINAL DEFECT IN THE FACE OF LEAKAGE OF CEREBROSPINAL FLUID Actual rupture with complete collapse of the sac was almost invariably due to birth trauma, and proved to be a most dangerous complication. In seven patients in whom, for one reason or another, no repair was attempted death followed within a fortnight. Eight patients were treated by operation immediately upon admission to hospital, and three of them survived. Of the survivors, one is normal (the only example of rupture of a simple meningocele which occurred

in this series) and two have developed hydrocephalus, one of these being mentally normal and able to walk with calipers, the other backward but without any defect of cord function. Of the fatal cases, there was one post-operative death from meningitis with suppurative myelitis of the cord stump: this baby had shown no pre-operative neurological deficit but became completely paraplegic before death on the fourth post-operative day. In two cases hydrocephalus proved fatal, chronic meningitis being contributory in one. There was one death from pyonephrosis complicating a neurogenic bladder and in this case it was noted that anal sphincter tone was good and the anal skin reflexes present before operation but that both were absent thereafter, so that operation apparently increased the neurological deficit. The final death occurred outside hospital when the child was 2½ years old and the cause is uncertain. This infant presented at the age of 1 day with a heavily damaged lumbosacral myelomeningocele with complete loss of function below first sacral level. Operation was performed and the immediate recovery was uneventful, but during the ensuing weeks voluntary movement in both legs became progressively less until by the age of 4 months the infant was completely paraplegic. There was no external sign of sepsis and further investigation was refused. This may, in retrospect, have been a case of missed diastematomyelia, but the possibility of chronic intraspinal infection is not excluded.

Chronic leakage of cerebrospinal fluid without frank rupture of the sac is a common event in cases of unrepaired myelomeningocele. The origin of the leak can sometimes be difficult to determine once the neonatal period is over and the exposed cord rudiment has become covered by dense, puckered scar tissue. Often the cerebrospinal fluid appears to be coming from the central canal of the cord, an occurrence which is made possible by the fact that in the grosser forms of spina bifida there is an associated hydromyelia which often extends from the level of the lesion to the fourth ventricle. In other cases fissures develop between the epithelium clothing the membranes of the sac and that which covers the area medullo vasculosa, while sometimes local pressure by clothes or dressings causes ulceration and breakdown.

Such a chronic leak is less immediately fatal than frank rupture of the sac. Of 18 cases in which (under the standards prevailing at the time when they were first seen) operation was not advised, 10 survived for at least a year, though all except two eventually died of meningitis or hydrocephalus. The remaining eight failed to survive beyond the age of 3 months. Of the 23 patients with cerebro-

spinal fluid fistula on whom operation was performed, nine showed clear evidence of hydrocephalus before operation, and another nine developed signs of increased intracranial pressure within a month. Thirteen of the hydrocephalic patients have died, 10 of hydrocephalus itself, two of meningitis, and one of an undetermined illness: only five are still living.

All the five patients who avoided hydrocephalus survived. In one such hydrocephalic case, operated upon at the age of 6 weeks, there has been some slight deterioration in cord function since the operation, which was performed while the sac was ulcerated and septic. Ventricular drainage was required for several weeks and the wound was inflamed for a similar period though a generalized meningitis was avoided; it seems reasonable to attribute the increased paralysis to infection of the replaced tissues.

ELECTIVE REPAIR IN FIRST WEEK OF LIFE A group of seven cases of complete rachischisis was operated upon electively, that is to say in the absence of cerebrospinal fluid fistula though before the central area had become fully epithelialized, within seven days of birth. Three of these, who all survived, have shown no evidence of hydrocephalus at any time and in a fourth case the hydrocephalus, though present at birth, was mild and became arrested at the age of 1 year, the patient being now mentally normal. The other three patients developed hydrocephalus within the first post-operative month and this proved fatal in two, meningitis being contributory in one; the third hydrocephalic child died of urinary infection.

Among the four survivors there was one case of definite post-operative deterioration in cord function, recovery being complicated by the development of wound sepsis with a cerebrospinal fluid fistula which only healed after several weeks. This was a child with thoracolumbar myelomeningocele, leg movements being thought to be adequate at all times up to the age of 6 months. Between then and 1 year the child progressively lost function in both legs, one becoming completely paralysed. This case, like the one of ruptured sac with slowly progressive paraplegia mentioned in the previous section, may possibly be one of intraspinal infection but in the absence of evidence of spinal block, re-exploration was not performed.

There was no instance of substantially improved neurological function after spinal repair in any of the cases which were deliberately operated upon early.

LATER ELECTIVE REPAIR Ninety-seven cases which were not complicated by cerebrospinal fluid fistula

were operated upon after the first week of life but seven of the patients were first seen at the age of more than 2 years and have been excluded from consideration here. Table I presents an analysis of the remaining 90 to which have been added for comparison the seven cases of early elective operation of open myelocoele mentioned in the previous section. It shows, as might be expected, that the proportion of infants showing evidence of hydrocephalus before operation rises with increasing age during the first six months of life. The slight fall thereafter is probably only due to the fact that after six months many hydrocephalic infants were either dead or too crippled for surgery to be considered, so that those operated upon in later infancy were relatively well-preserved. The total incidence of hydrocephalus, pre- and post-operative, remains remarkably constant at between 45 and 50%, and though this complication was more frequently fatal in patients operated upon in the first month of life, the death rate thereafter shows no significant variation.

The exclusion of cases of cerebrospinal fluid fistula means that the group is composed of infants in whom the sac was soundly epithelialized at the time of operation and the risk of meningitis would therefore be expected to be small. In fact, no fatality occurred from this cause. However, it would have been unsafe to conclude immediately that because no case of open myelocoele died of meningitis if operated upon after epithelialization of the sac had become complete, therefore it is best to postpone operation in all such uncomplicated cases, for there might have been a large number of deaths from meningitis in those who awaited operation. To test this point, the group of unoperated cases reported in the previous communication (Doran and Guthkelch, 1961) was re-examined, excluding those in whom a cerebrospinal fluid fistula was present when the patient was first seen. There were in fact, only five instances of death from meningitis between birth and 3 months of age. Two of these

were examples of thoracolumbosacral myelomeningocoele with complete paraplegia from mid-thoracic level, and were originally rejected for surgery on the basis that their degree of neurological deficit was considered to be incompatible with survival, so that no effort was made to prevent possible complications. Had the sacs been adequately protected in these cases, death from meningitis might not have resulted. One newborn infant had a gravely septic, almost gangrenous sac, with established meningitis, and died within an hour of admission to hospital. This leaves only two patients out of a total of 93 unoperated (but without cerebrospinal fluid fistula) in whom postponement of operation to the age of 3 months, had they been thought suitable for it on other grounds, could have led to death from meningitis.

The number of instances of post-operative deterioration in function of the central nervous system in the group of patients in whom repair was performed after the first week was small (four out of 90 or 4.5%). In one of these the cause was surgical trauma, the other three were associated with prolonged wound sepsis. Some improvement in neurological status was noted in the post-operative phase in two cases, but it was only trivial, amounting to increased movement in the toes in one case and the appearance of previously absent tendon reflexes in the other.

DISCUSSION

The general rules by which some cases of spina bifida cystica should be accepted and others rejected for treatment have been discussed elsewhere (Doran and Guthkelch, 1961). Here, the question to which attention is directed is this: Given a case of spina bifida cystica, at what age is the repair of the spinal defect normally most safely performed, what complications occur in the presence of which a more general rule must be waived, and in what way do these complications modify the results obtained?

TABLE I
AGE AT REPAIR OF DEFECT, RISK OF HYDROCEPHALUS, AND EFFECT ON CORD FUNCTION¹

Time of Operation (mth.)	Total Cases	Incidence of Hydrocephalus			Post-operative Neurological Deterioration
		Pre-operative	Post-operative	Fatalities	
0-1	20	3 (15%)	6 (30%)	5 (25%)	1
1-3	22	5 (23%)	6 (27%)	3 (14%)	1
3-6	21	7 (33%)	3 (14%)	3 (14%)	2
6-24	34	9 (26%)	7 (21%)	5 (15%)	1
Totals	97	24	22	16	5

¹Cases with rupture of the sac or a cerebrospinal fluid fistula are excluded.

Whatever the answer, it will not be the ideal one. In the early stages of rachischisis, the open neural plate contains abundant developing neural tissue but during later foetal life this suffers a progressive degeneration, the common result being myelomeningocele with paraplegia (Warkany, 1960). Clearly, therefore, short of preventing its occurrence, the ideal treatment of rachischisis would be to close the neural plate and repair its coverings long before birth, but this, in the present state of surgical knowledge, is impossible and we can only consider how best to time the operation once the infant is born.

There has not, in this series, been evidence that any important further loss of functional neural elements occurs after birth, for although occasionally leg movements were reported immediately after delivery of a baby born with a myelomeningocele, who was subsequently found to be paraplegic, such as were personally observed seemed to be in the nature of reflex spasms rather than voluntary movements. Equally, although the possibility of regeneration of the malformed cord, once it is restored to its normal environment, should not be completely ruled out, there has been no evidence in this series of substantial increase in neurological function following spinal repair, whether the operation was performed early or late. Consequently, there does not at present seem to be any *a priori* reason to operate upon infants with myelomeningocele within the first few days of life, though it should be stressed that as the hospital at which this work was done does not lie close to any maternity unit, the writer has no experience of operation performed immediately after delivery. The results of such a study would be interesting, but the present enquiry must confine itself to the situation as it presents itself when the routine of delivery (at home or in hospital) and transfer of the baby to a neurosurgical unit has been accomplished, and the baby is already several hours old.

Further, since it was possible to avoid any

mortality from shock and haemorrhage throughout the series and since surgical trauma to the cord should be rare, the discussion is in effect limited to consideration of the likelihood and consequences of infection and to the effect of operating on the spine on the potential for developing hydrocephalus which exists in all, or almost all, infants suffering from myelomeningocele.

It has not been thought necessary to extend the analysis of operative results to cases of simple meningocele since in these the only danger which needs consideration is that of meningitis and in minimizing this the same principles apply as hold for the more serious types of spina bifida. A general comparison of the results of treatment in these latter (Table II) suggests the three following conclusions:

1 Operation upon a myelocele within the first week of life is liable to be followed by impaired function of the cord in a substantial proportion of cases. This deterioration was sometimes associated with manifest infection of the cord, and its relatively greater frequency in cases with ruptured sac suggests the same cause even though definite signs of meningitis were not always present. When operation was postponed until the sac was epithelialized this post-operative neurological deterioration occurred less frequently and when it did the post-operative period usually proved to have been complicated by such evidence of sepsis as an inflamed wound and a temporary cerebrospinal fluid fistula.

Consequently, it is suggested that in a case of complete rachischisis, the surface of which is not as yet epithelialized, operation within the first week of life is justified only when the sac has been ruptured by trauma or is on the point of rupture, for it is only in these conditions that the danger of loss of life from loss of fluid and meningitis is greater than the risk of impairing the function of the cord through local infection in the post-operative phase. It had originally been hoped that if the spinal defect was repaired sufficiently soon after birth, infection could be avoided, but in practice, since the baby was

TABLE II
AGE AT REPAIR OF DEFECT, RISK OF HYDROCEPHALUS, AND EFFECT ON CORD FUNCTION
IN ALL CASES

Time of Operation		No. of Cases	Incidence of Hydrocephalus		Post-operative Neurological Deterioration
			Affected	Fatalities	
First week	Sac intact	7	4 (57%)	2 (29%)	1 (14%)
	Sac ruptured	8	4 (50%)	2 (25%)	3 (37%)
After first week	Without C.S.F. fistula	90	42 (47%)	14 (16%)	4 (4%)
	With C.S.F. fistula	23	18 (78%)	10 (43%)	1 (4%)
Totals		128	68	28	9

never received in the neurosurgical unit until several hours after delivery, there was always bacterial contamination of the surface of the sac, and the presence of exposed nervous tissue limited the choice of skin-cleansing agents and the vigour with which they could be used. It is possible that a series of cases operated upon within an hour of birth might show different results.

Most of the cases of myelocoele without cerebrospinal fluid fistula became soundly epithelialized within a few weeks of birth. In a few instances, in which the swelling was large and tense, the fundus was not yet covered by healthy skin, and in addition there was heavy infection of the bare area, the baby was admitted to hospital and epithelialization allowed to continue with the aid of protective pads or cages and the local and systemic administration of antibiotics. Operation was not performed until this process was complete.

The group of babies operated upon electively between the ages of 1 week and 3 months was mainly composed of those in whom the sac was already epithelialized (sometimes from birth) but in whom other considerations such as its size or the likelihood of rupture, the desire to avoid a long period in hospital, or a request that the spinal repair should be completed so as to facilitate orthopaedic treatment of paralytic limb deformities, made relatively early operation desirable. The results obtained in such cases were, apart perhaps from a slight extra danger that if hydrocephalus developed in cases operated upon in the first month, it would prove fatal, neither better nor worse than those secured from waiting longer.

2 So long as sepsis is avoided, the timing of the operation on the spinal defect has little effect on the final neurological state. Although, at first sight, the data presented in Table II appear to constitute a powerful plea for deliberately avoiding operation in the newborn period, yet on analysis what seems to matter is not the age at operation but the state of the surface of the swelling. If this is not covered by skin, then it is contaminated, and to replace this contaminated nervous tissue within the spinal canal is to court the dangers of suppurative meningitis and myelitis.

Conversely, though meningitis is often a fatal complication of unrepaired spina bifida it need not be feared as a complication of operation on the spinal defect, provided always that the latter is covered by healthy skin.

3 The factor which, from the clinical point of view, most increases the risk of progressive hydrocephalus in spina bifida babies is the presence of a cerebrospinal fluid fistula. Not only is hydrocephalus commoner in the presence of a cerebrospinal fluid

fistula (74%) than in its absence (47%) it is also more dangerous, the mortality rates being 41% and 16% respectively. It may seem strange at first sight that within the cerebrospinal fluid fistula group the risk of hydrocephalus and the mortality from this cause are rather higher for cases operated upon *after* the first week of life than for those in which the spinal defect was repaired immediately after birth, but the reason for the difference is probably this, that whereas rupture of the sac at birth is an accident which is rendered more likely by such incidental factors as a difficult labour and a particularly thin sac, a chronic cerebrospinal fluid fistula represents the natural external drainage of what would otherwise be a severe, progressive hydrocephalus. If this drainage ceases, the dynamics of the cerebrospinal fluid circulation are immediately interfered with and even such cases as have shown no sign of increased intracranial pressure before the operation are likely to do so, often to a dangerous degree, immediately after it. This is one type of case in which it would probably be wise to attempt some sort of internal drainage of cerebrospinal fluid as a routine before embarking upon any repair of the spinal defect.

SUMMARY

In the management of spina bifida cystica, the local complications which most strongly militate against successful surgical repair are infections of the surface of the sac and the presence of an acute or chronic cerebrospinal fluid fistula.

When the sac has frankly ruptured, at whatever age, immediate closure of the defect is necessary to save life though the ultimate results are inferior to those obtained after operations of election. Apart from this over-riding consideration, it is unwise to operate on a myelocoele before epithelialization of its surface because of the danger of introducing sepsis within the theca and thereby causing secondary deterioration in spinal cord function due to suppurative myelitis.

Once epithelialization is complete the danger of sepsis should be minimal and there is no reason to wait for the infant to attain any particular age before repairing the spinal defect. Surgical intervention may be performed as soon as is necessary, *e.g.*, as a preliminary to the correction of associated deformities.

Where a cerebrospinal fluid fistula is present (without frank rupture) operation should be delayed only for so long as is necessary to secure the optimum conditions for its performance, the most important being the eradication of surface infection. In all these leaking cases an acute post-operative hydrocephalic episode must be anticipated, and it is

therefore suggested that in these circumstances a trial should be given to the creation of some form of internal diversion of the cerebrospinal fluid circulation before any attempt to repair the spine.

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ADDENDUM

Some mention should be made of Laurence's (1960) suggestion that in infants suffering from myelocoele the area medullovasculosa should be covered by a skin graft within 24 hours of birth. The cases reported above were, of course, treated before Laurence's communication had appeared. But in two recent cases in which closure of the bare area by a free skin graft has been attempted, the graft has not

taken, perhaps because of the difficulty of maintaining firm pressure on the grafted area without raising the intraspinal and intracranial pressures to dangerous heights. Also, in one case, leakage of cerebrospinal fluid may have contributed to the failure. Such leakage can hardly be avoided, since the central canal of the cord opens at the upper end of a myelocoele, and there is often a gross hydro-myelia above it. This difficulty could, however, be avoided by using divided grafts, and if, contrary to the view expressed in this paper, early closure is felt to be of advantage, then the method suggested is worth a further trial.

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