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Management of Myelomeningocoele - Selection Policy and Indication for Surgery

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V Balasubramaniam, B Chidambaram, - *Ranga Nursing Home, 36, Moubray's Road, Madras - 600 018*

Abstract

This paper discusses the merits and demerits of various surgical procedures in the management of myelomeningocoele. The timing of the operation and the results of the procedures are evaluated with special reference to the quality of life. The direction of future research is indicated.

Key words -
Myelomeningocoele,
Surgical procedure,
Timing and results,
Future trends

In this paper the term myelomeningocoele is taken to refer to the wide variety of dysraphic anomalies from a meningocoele. The same group has been referred to by some as spina bifida aperta or spinal bifida cystica. As in many other problems there are two methods of approaches. The first is to state certain facts of policy and justify them This is the centrifugal method. The other is to collect all the data, discuss the hypotheses and theories and then try to frame out guideline. This is the centripetal method. This centripetal method is followed here.

In myelomeningocoele we encounter a group of disorders where the aetiology is not known, pathogenesis uncertain and treatment controversial. Medical science is still unable to prevent this miserable disorder; nor is surgery able to offer a consistently useful programme for management.

There has not been a uniform selection policy with all the surgeons; nor has there been even an uniform policy in any Institute or even with a single surgeon. Opinions vary between the two extremes - at one and every case being operated upon while at the other end no case is touched till the patient is a few years old perhaps depending upon death to eliminate a certain percentage. It is not a mere coincidence that at these two extremes are poised the junior most surgeon bursting with enthusiasm and at the other end the experienced but disillusioned older group

Before we discuss the selection policy and indication let us first see what the objects of surgery are. In myelomeningocoele there is a varying defect of the neural tube (to wit the adult spinal cord) and the neural elements. The neurological deficit (motor, sensory, autonomic and viscero-motor) depends on the type of defect and the level of lesion. In addition in many of the children hydrocephalus has been a complicating factor. It may exist together or follow surgery in a percentage. Patients who are not operated also develop hydrocephalus. Whether hydrocephalus is due to the same cause that produces the dysraphic state or is the result of the dysraphic state is not decided. Some feel that hydrocephalus follows unoperated cases due to infection. Hydrocephalus following surgery may also be due to infection or bleeding. In other words there may be many causes for the hydrocephalus.

The aims of surgery are

(a) to close the defect in order to prevent infection and to prevent deterioration of the neurological deficit - the latter of course is questionable. Whether repair of the meningomyelocoele will correct the existing neurologic deficit is a highly debatable point. Sufficient studies have not been made to answer the questions either way.
(b) The second aim of surgey is to treat the hydrocephalus either by anticipation or by steps taken after it may have set in.

It is an undisputed fact that exposure of the cord or its part to the atmosphere carries a high risk of meningitis, etc. So one should close the defect. And if this is to be done well it should be done as soon as possible. Surgery immediately after birth is possible but whether this is worthwhile in the new born infant is debatable. The child may have more serious anomalies which may require greater attention or may even prove fatal ; secondly there is no guarantee that surgery will not worsen the condition. This latter statement may raise some discussion. Surgical procedure if confined to mere closure will be safe; but too enthusiastic a dissection might result in a great deficit detectable only at a later age. Secondly it is not known whether hydrocephalus is the result of surgical procedure. There is of course a group where hydrocephalus co-exists with the myelomeningocoele. But in a certain patients the head which was definitely normal in size does increase in circumference following surgical repair of the deformity. Dandy [1] held that this was because the sac which was doing a 'compensatory function' of CSF absorption has been removed. But the problem is not so simple as all that. It is likely that surgery contributes to the hydrocephalus by introducing infection or by bleeding; it is equally likely that the surgical procedure happened to be have been done in cases who in later days to come would have any way developed hydrocephalus. In one series reported by Heimburger [2] children operated on between 24 and 48 hours after birth, while having a better chance of survival also had a greater chance of developing hydrocephalus and spinal cord dysfunction

While the question of hydrocephalus is interesting from the point of its relation to the surgery it is not so difficult to manage; but the occurrence of neurological deficit and its relation to surgery are still undecided.

Does surgery correct the existing deficit, prevent further deficit, or does it make it worse?

To take the last point first, properly done surgery at any age should not make the patient worse. This means a thorough assessment of the neurological state must be made lest an overlooked defect be attributed to the surgery. It is necessary to stress this because adequate neurological assessment is obligatory. If this is done then one can certainly state that properly done surgery cannot make the deficit worse.

One of the most contested claim is the question of arrest of further neurological deficit and even variable degree of recovery from the existing deficit.

In an unoperated meningomyelocoele it is very likely the deficit may become worse in course of time, due to infection and further loss of nervous tissue. In addition when hydrocephalus occurs it adds to the deficit. All can then agree that these can be prevented by early surgery.

But whether early surgery can actually help these children to recover from the existing deficit is debatable.The neurological deficit is probably due to rupture of the neural tube (or failure of formation of the dorsal part of the neural plate) and at surgery the surgeon only detaches the cord from the skin and edges or the cord can come together and grow into a normal cord is not known. Under such circumstances it is difficult to be dogmatic in ascribing to surgery more benefit than it deliver. At the best surgery can prevent further damage.

The next point to be decided is whether surgery can prevent further neurological deficit.

In 1959 a programme was started in Sheffield in which all the children were operated within the first few hours of life [3]. Hydrocephalus was also tackled. It was a multidisciplinary approach. Though initially much enthusiasm was generated by the plan, a long term appraisal found that a considerable number of patients with severe handicap had been saved. In addition to the mental agony for the patient, and the strain for the people around there was considerable expense for the state as the National Health Service had to provide for them.

In the series reported by Heimburger [2] out of 71 patients operated 37 survived. Of these seven (i.e. 19%) were quite

normal. Of these 28 (i.e. 75%) were attending school. Only 9 (i.e. 24%) were disabled. The conclusion arrived was that "Myelomeningocoele repair within the first 24 hours after birth provides the best quality of survival. Children operated on between 24-48 hours after birth have a better chance of surviving, but also a greater chance of developing hydrocephalus and spinal cord dysfunction.The high incidence of hydrocephalus, spinal cord dysfunction and both indicate that myelomeningocoele repair should not be done between 1 week and 1 month after birth. If surgery is not done during the first week of life it is safer from all point of view to postpone it until the child is 1 or more month old". The conclusion of the report is that "the chances of survival are better if the child is operated within 24 hours".

The policy of wait and see

In many series a policy of waiting for a few weeks or a few months is followed. The main reason is that many children when they are brought have already established infection. (This can certainly come on in a few hours). Surgery in these cases carries the risk of meningitis and other complications. Another reason for waiting is that more serious defect either in the nervous system or elsewhere may come to the forefront and surgery on the dysraphic anomaly may have to be modified or even given up. This policy has certainly considerable measure of persuasion. But what must be remembered is that in this process quite a number of cases can be eliminated either by sepsis or other complications and a bias is introduced in selection.

There are certain condition listed definitive contraindications for surgery.

Hydrocephalus if already present will have to be treated first and this may postpone surgery on the dysraphic anomoly.

Mental retardation either primarily (or due to hydrocephalus) will certainly contraindicate surgery. And this can be determined only long after the first few days.

The presence of cranio-lacunia as shown in the x-rays has been taken to indicate mental retardation and has been listed as one of the factors against surgery. But this is not agreed to by all.

Most neurosurgeons, who have operated meningomyelocoele when they followed their cases over a period have had some disquieting moments. The doubt they all have experienced is whether surgery only helped to prolong a miserable existence. Many of their children who are incontinent hydrocephalic and retarded, are painful evidence of enthusiasm superseding scientific judgement. Every surgeon at one time or other must have felt whether he was guilty of misjudgement in operating on a particular patient. When the parents and relatives are confronted by a new born with reddish cord lying exposed they would first want it to be closed and any amount of talking may not at that time indicate to them the agony and misery that is awaiting 50 per cent of the children operated. At a later date they too would feel that it would have been better if the child had been left alone.

This raises in its wake a scientific cum ethical question about not operating. Many purists have exclaimed that in these cases, the involved party (namely the child) is not consulted. More serious than this is the charge that the surgeon cannot take the decision not to operate and thus allow the child to die. Both these objections can be considered as frivolous by many. While much discussion can centre around this, it cannot be denied that this situation is certainly not unique to dysphoric state only. When the surgeon takes a decision in many situations for eg. a cancer stomach or a glioblastoma of the brain such objection too can be raised. But surely no surgeon will operate when he knows that he cannot help the patient. If he did he will be guilty of hypocrysy.

In all such situations a well known escape route is to state each must be judged on its individual

merits. This is only a euphemistic way of saying "we do not know".

Let us face it. As our knowledge stands no definite guidelines can be given. And it is likely to be so for a long time.

Every surgeon must act on according to his own judgement, the facilities available including the family background and help from State.

One has only got to see once, a child operated for meningo-myelocoele with double incontinence, paralysed and with trophic ulcers to realise the drain on the family much to the detriment of other healthy childrens.

Ultimately the answer will lie only in prevention if possible or correction in utero. Estimation of alpha fetoprotein and ultrasonography may be of great value here. However till that millienieum arrives one has to do something.

Our Experience

Our case material for the first twenty years of our Department between 1950 and 1970 is shown:

Table

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For a long time our Department was the only one of few neurosurgery centres in India. Cases came from all parts of India and they came at varying intervals and we were not in a position to lay down rigid criteria for surgery. But we refused as general rule to operate if the deficit was severe and there were complicating factors like infection. Quite a number of cases operated within a few hours of birth, did well, and quite a number did not. It looks what we are dealing with certain other factors as such unknown. This is not a scientific assessment but it is a frank statement of our experience. Since last few years many neurosurgical centres have came up and many others are also taking up cases of dysraphic anomaly. Perhaps the only solution is to have a collaborative study each centre doing one line of treatment or all centres treating them in a particular order. After five or ten years the cases can be assessed and conclusions drawn. Until then let us be cautious in our approach to the problem of myelomeningocoele. The situation is even worse than that of hydrocephalus.

Dandy's statement regarding treatment of hydrocephalus must be remembered here too. "For infants, therefore assurance can never be given that a cure is attainable; it is just a chance, less than even, but the only chance and it may be worth taking" [4]. Failure to adopt a proper non-emotional scientific attitude will result in haphazard surgical attempts with children who cannot be helped and who cry out "Oh Death where is thy sting".

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