Spina bifida cystica

An orthopaedic criterion for the early closure of myelomeningocele

C. C. MICHAEL JAMES

From the W. J. Sanderson Orthopaedic Hospital, Newcastle upon Tyne

SUMMARY There is an urgent need for criteria upon which to base the decision to close a myelomeningocele soon after birth. Orthopaedic experience advises that early closure of the spinal lesion should not be done unless there is active flexion power at both hips. This criterion is offered with the knowledge that there may occasionally be other exceptional factors to contradict it, but it is based on experience of 96 personal cases aged between 18 months and 7 years. These cases are analysed and discussed. A further possible criterion for not closing the spinal lesion early is the presence of a gross lumbosacral kyphosis.

Sufficient time has elapsed for preliminary conclusions to be drawn about the consequence of early closure of spina bifida cystica, as advocated by Sharrard, Zachary, Lorber, and Bruce in 1963. This operation, together with control of hydrocephalus, has saved life, but very often has not preserved the muscle function in the lower limbs that had been hoped, so that there are numerous survivors whose severe disability is not compatible with an independent and happy existence. It has been clear for a long time that there must be selection of cases for early closure of the spinal lesion in the first few hours after birth, but it has been difficult to know on what criteria the decision should be based.

Because of the paralysis in their lower limbs, large numbers of these children require orthopaedic treatment, and with the passage of time their patterns of development can be watched and classified. The cases divide themselves into two principal groups, those with power of hip flexion and those without. Reviewing the progress made by the children in these two groups and anticipating their difficulties in the long-term, it becomes clear that one consideration to be taken into account when deciding whether a newborn infant should have early closure of the spinal defect or not is the presence or absence of power of hip flexion. Orthopaedic experience suggests therefore that *early closure of the spinal lesion should not be done unless there is active flexion power at both hips*.

This criterion is offered with the knowledge that it is only one of several factors which have to be taken into consideration. It is based upon experience with 96 cases referred for orthopaedic treatment at

about 12 months old from several different sources. The children were born between February 1964 and August 1969 and were all submitted to early closure of the spinal lesion. Seventy-four of the 90 survivors are now between 3 and 7 years old.

FLEXION POWER OF THE HIP

Sixty-three children have bilateral hip flexion power, including seven with paraplegia, and at least 10 with reflex activity of the muscles of one hip. Depending on the extent of muscle power to control the knees and feet, these patients can all be expected to be encumbered finally by at most a pair of calipers—apparatus which they can put on and take off themselves without assistance. Many will not require splints. Consequently the majority can be expected to live an independent existence and be able to move around in the world outside their own homes. Many of these children compare with the best survivors, now adults, of the years before early closure of the spinal lesion was attempted and when hydrocephalus was untreatable; even so, these adults commonly suffer from trophic lesions and their complications. Sharrard's operation of posterior transplantation of the iliopsoas muscle (1964) has enabled those cases with paralysed gluteal muscles to walk with minimal supporting splints and has prevented the hip flexion contracture which developed in those previously abandoned to a chair existence and the trophic ulceration so commonly associated with it.

By contrast, it is clear that the other 33 cases in this series who have no power of flexion of both hips

are permanently condemned to much more elaborate splintage to enable them to walk, and which they will not be able to put on and take off themselves without assistance. They also have total sensory loss from T12 dermatome downwards. It is possible that a very few individuals will learn to deal with this splintage themselves, but none of this group of severely paralysed patients will be able to live an independent existence. Twenty-nine children have complete paralysis of both hips; two cases have one flail hip and reflex muscle activity in the other, and two have good power of flexion in one hip only, the other being flail. In the two latter cases, each with one flail hip and one with control, the prognosis is better, although it will probably be necessary to amputate the useless flail limb to facilitate ambulation on the good limb using crutches, presuming that the foot has good sensation. If there is sensory loss in the foot, trophic ulceration will occur and this leg will also have to be amputated but, at least, the proximal part of the limb will be a valuable member in stabilizing sitting and in movement between chair and bed.

It is to be expected that ultimately almost all those cases with flail hips will prefer to discard supporting splints and lead a wheelchair life, which will at least enable them to go out of their homes. There are great advantages in teaching them to walk when very young, although this is very burdensome on the parents and on the physiotherapists who instruct them, but these advantages often disappear later on when the child becomes frustrated or discovers the pleasures of a wheelchair at school which gives him so much greater freedom of movement. In any case, he will never be able to live an independent existence without someone to look after him, in spite of all the mechanical aids available to lift him between bed and chair and to make his life generally easier. His future capacity for employment and earning a living will be very limited, and he is likely to be dependent on an institutional home and on National Assistance. He will always run the risk of trophic ulceration over his sacrum, buttocks, and ischial tuberosities, and he will not have the stabilizing effect of thigh muscles to help him sit without support all round. This tale of future tribulation can be further extended to include burns of the legs from sitting too close to sources of heat, and the incidence of fractures from careless movement (James, 1970), to mention only two of the many complications which occur.

The difference between these two groups is very striking and the quality of life to be expected for those with flail hips is obviously very poor, taking into consideration their orthopaedic disabilities alone. Some of those with good hip function will also be permanently dependent on outside assistance, but

for non-orthopaedic reasons—for example, mental retardation which cannot be anticipated at birth.

DISCUSSION

There are many conditions which condemn an individual to a wheelchair life and to dependence on other people; they are conditions which develop after birth. They are not comparable with spina bifida cystica which is present at birth, and cases with flail lower limbs and severe sensory loss who have never known any other physical state. They are consequently deprived of the normal early development and independence which has benefited cases with acquired conditions. Cases of amelus or phocomelus suffer severe disability but they can learn to walk and do not have to contend with incontinence, trophic ulceration, or the consequences of hydrocephalus.

Many difficulties attach to the decision whether or not to operate on the back of a newborn infant and a number of factors have to be taken into consideration. Nevertheless, the presence of active hip flexion is a valid criterion for the clinician who has to assess the infant's total state when deciding whether early closure is advisable or not. It will be found that other factors may preclude early operation when hip flexion is present, but orthopaedic experience leads to the view that, where there is no active power of hip flexion, early closure is not justified unless there are exceptional circumstances.

Examination of the movements of the lower limbs of the newborn infant is not easy and does not always provide immediate information. Sometimes the child will not exhibit the movement of which it is capable and it is often difficult to know whether the movements present are voluntary or reflex. When it is impossible to decide this latter point, the child should be given the benefit of the doubt if other factors are favourable. Voluntary movements carry a good prognosis, although a number of patients are known to lose muscle power later just as some of those with apparently no muscle power at birth develop a limited function later. For children with paraplegia the future is dubious but for those without movement the outlook is bad and, although their expectation of life is short, it is not short enough to avoid the trials they bring upon their families. Many, however, have much happiness in their childhood. It might be thought that those with functionless lower limbs were liable to die earlier than the more fortunately-muscled children, but, in this short series of 96 cases, only two of the six deaths which have so far occurred have been those with flail limbs, both aged 3 years; the other deaths occurred at 2, 3, 4, and 5 years.

A possible further criterion and contraindication

to early spinal closure is the presence of a gross kyphosis. Most cases will have no muscle power to control the hips and would be excluded on that account alone. Those with power will probably have reflex muscle activity, but an occasional case has normal muscle function. The disadvantages of a gross kyphosis are the difficulty of preventing chronic ulceration, the impossibility for the child to lie on its back, the difficulty of splint-fixing, and the ultimate tendency for the angulation to increase with weight-bearing, which leads to stretching of the spinal cord and nerve roots with resultant progressive increase in paralysis. Osteotomy of the spine may be feasible if done early in cases with good limb musculature.

It is only those clinicians who personally follow-up these paralysed cases who can realize the problems which are created and it is necessary that the sur-

geons responsible for primary closure should have their attention drawn to the consequences of their decisions. In time, there will develop a consensus of opinion which may possibly establish that a less active management in the early days after birth is kinder in the long run to the child and to the family into which it is born.

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