SPINAL OSTEOTOMY FOR CONGENITAL KYPHOSIS IN MYELOMENINGOCELE

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Hoppenfeld (1967), in a review of spinal deformities associated with myelomeningocele in patients admitted to Sheffield Children's Hospital between 1956 and 1960, found that kyphosis of the lumbar spine was present in one in eight patients. He reviewed the anatomy of the deformity and the problems in management to which it gives rise.

This paper describes techniques that have been devised to try to improve the management of children affected by this serious congenital abnormality.

THE DEFORMITY AT BIRTH

If myelomeningocele is associated with lumbar kyphosis the open spinal cord is stretched over the kyphotic lumbar vertebrae (Fig. 1). As Hoppenfeld (1967) observed, the pedicles are very widely spread in the area of the kyphos and the skin defect is extremely large—often more than two-thirds of the width of the back. Figure 2 shows the radiographic extent of the deformity. The kyphos usually involves all the lumbar vertebrae and the angle of the deformity is 90 degrees or more. The pelvis is rotated forwards and the plane of the lower limbs is at right angles to that of the trunk, so that the hips appear to have a severe fixed flexion deformity.

The lower limbs almost always show a variety of paralytic deformities characteristic of myelomeningocele, such as dislocation of the hip, recurvatum of the knee and calcaneo-varus

Fig. 1

Figure 1—Child aged 6 hours with myelomeningocele and lumbar kyphos.

Fig. 2

Figure 2—Lateral radiograph of a severe lumbar kyphos taken on the day of birth.
or equinovarus of the foot. Before operation there is likely to be complete paralysis of the lower limbs or spontaneous activity only in the hip flexor and adductor muscles. Faradic stimulation of nerve trunks and muscles indicates that there is potential for recovery in additional muscles supplied by the lumbar neurological segments, but there is usually congenital paralysis of the muscles supplied by the sacral segments.

**PROBLEMS ASSOCIATED WITH OPERATIVE CLOSURE**

Before 1962 attempts were seldom made to close an extensive defect of this kind. Conservative management left 25 per cent of survivors all with complete paraplegia below the tenth or eleventh thoracic level, and with severe kyphotic deformity of the spine covered by thin irregular skin that was extremely liable to break down. Since 1962 an attempt to close the spinal defect surgically has been made, however extensive and difficult the procedure might appear to be, because it was considered that the prognosis for life and for power in the lower limbs could hardly be worse. Although there have been more survivors, breakdown of the skin suture has been common. The combination of the posteriorly projecting bony hump and the extreme width of the bony elements of the spine has made satisfactory skin closure without tension impossible even with extensive skin flaps or relaxing incisions. Secondary infection of the stretched neural tissues has resulted in complete flaccid paraplegia in nine out of ten patients.

**OSTEOTOMY RESECTION OF THE SPINE IN THE NEWBORN**

Necropsy studies suggested that only osteotomy of the spine with resection of one or two lumbar vertebrae could succeed in reducing the kyphos. This operation was first performed in July 1966 and has been done for five patients since then. The object of the operation is to remove one or more vertebral bodies at the apex of the kyphos, the bone being taken out laterally between the nerve roots to avoid damage to the nervous system or to the large vessels running on the posterior abdominal wall. The patients were operated upon three, five, five and a half, eight and a half, nine and a half and thirty-six hours after birth. **Technique**—The membranous tissue connecting the neural plaque to the skin is excised and the dura mater lining the posterior surfaces of the vertebral bodies and pedicles is dissected medially in the same way as in the closure of other myelomeningocele. The dural layer is often very thin as it lies over the prominent pedicles. The three or four nerve roots associated with the apical vertebrae are identified as they leave the dura to pass through foramina in the bone (Fig. 3). It must be appreciated that in the area of the kyphotic deformity the laterally directed processes are fused together on their outer side to form bony foramina like those normally present in the sacrum. The lumbar fascia is incised at its attachment to the tips of the processes to expose their anterior surface and the nerve roots as they emerge from the foramina to enter the psoas muscle. The osteocartilaginous processes are removed, the nerves being protected by a blunt dissector. The nerve roots are then displayed in their full course from the spinal cord to the point at which they enter the psoas muscle anteriorly. The lateral aspects of the vertebral bodies can now be identified (Fig. 4).

By careful blunt dissection the dura mater is mobilised from the posterior longitudinal ligament, and the aorta and inferior vena cava are separated from the anterior surfaces of the vertebral bodies. Points are selected above and below the apex of the kyphos that will allow the removal of about one and a half vertebral bodies. The bone is divided at the two levels with a small French osteotome at right angles to the plane of the vertebral bodies (Fig. 5). The bony fragment is removed by extracting it from between the roots above and below the level of the osteotomies. The gap produced by this resection is easily closed by extending the hips to rotate the pelvis into its normal position. The tension on the neural tissues is immediately relaxed and the prominence of the kyphos considerably reduced. The
vertebral bodies are held together by four sutures. In five patients silk sutures were used, and in one steel wire. Some difficulty has been encountered in obtaining a satisfactory hold with the sutures, and the steel wire cut out of the bone. Satisfactory stability was achieved in four patients; in the other two there has been some displacement at the osteotomy after operation, and the ultimate correction has not been quite so satisfactory as that obtained at operation.

Figure 3—The dissected kyphos viewed from the side. The dural layer, lifted up by forceps, has been dissected from the lateral processes. Three nerve roots are shown entering foramina in the bone. Figure 4—Kyphos viewed from the side after removal of the lateral processes. Three lumbar roots are shown passing from the spinal cord forwards into the psoas muscle.

The dura is closed longitudinally by interrupted sutures. It is not possible to obtain any fascial closure: the lumbar fascia is displaced so far laterally that any medial mobilisation of it is impossible. The skin defect is closed by mobilisation of the skin from the flank, and a satisfactory vertical closure has been obtained without undue skin tension. In three out of the six patients lateral V-shaped relieving incisions have been needed on one or both sides.

The measured external blood loss has varied between 118 and 253 millilitres. One and a half times the external blood loss has been replaced through an umbilical intravenous catheter inserted before the commencement of the spinal operation.

**Post-operative management**—After operation the baby is suspended in an incubator by a sling of adhesive strapping to relax tension on the wound for the first three or four days. In two children with rather strong and active abdominal musculature it has been necessary to suspend the baby by tapes from the lower limbs to the top corner of the incubator to maintain extension of the spine until the osteotomy has united after three or four weeks. No additional splintage or plaster has been used because restriction to respiration is not tolerated by a newborn child.

**RESULTS OF NEONATAL OPERATION**

In spite of the extent and severity of the operation on a newborn child, all have survived. Primary skin healing occurred in three by the end of the tenth day and in two more by the end of the third week. In one patient there was necrosis of the skin edges with superficial infection, but healing was complete at the end of the fourth week and there was no meningeal infection.
Two patients developed generalised sclerema which responded to a combination of antibiotic and steroid therapy.

A typical clinical and radiological result is shown in Figures 6 and 7. The spine is covered by sound skin, the kyphos is improved and the position of the pelvis has been restored to normal so that the thighs lie in their correct relationship to the trunk. The spine in every case has united to give continuity of bone centrally at the vertebral bodies and laterally through the medium of the fused lateral processes.

![Fig. 6](image)

**FIG. 6**

**FIGS. 6 AND 7**

Figure 6—Clinical result six months after neonatal osteotomy-resection of spine. Figure 7—Lateral radiographs of lumbar spine fourteen months after neonatal osteotomy-resection.

The paralytic state in the lower limbs at follow-up has been improved compared with the pre-operative assessment in two patients, is the same in three and is worse in one. Neurological recovery has, in general, not been so good as has come to be expected of immediate closure of the spinal lesion in myelomeningocele without kyphosis, but is distinctly better than that after conservative treatment or attempted closure of the lesion without spinal osteotomy.

It is too early to be sure that the kyphos in children treated by neonatal spinal osteotomy will not increase when the child starts to bear weight. Of the three children who are now able to sit, the correction has been maintained in two. In one, relapse has been treated by further osteotomy at the age of ten months. The presence of sound skin over the residual kyphos makes it possible to apply splintage when the child is older to help to support the spine and to diminish the liability to recurrence.

**MANAGEMENT OF KYPHOSIS IN OLDER CHILDREN**

If no osteotomy has been performed soon after birth the kyphos remains extremely prominent or increases with growth (Figs. 8 and 9). Calipers cannot be fitted, or a child who has previously been able to walk with the help of calipers and trunk support becomes unable to do so because of fixed flexion of the pelvis.

When there is complete paralysis of the lower limbs and bladder, transverse osteotomy of the kyphos can be performed. This procedure is made possible because the paralysed neural structures can be dissected from the lumbar spine and the dura mater closed at the upper level of the kyphos. The bony arch of the kyphos can then be dissected out through a retroperitoneal exposure. Wedge osteotomy cannot be performed because of the shortness of the anterior abdominal structures, but correction can be obtained by overlapping the fragments.
and fixation with four screws (Fig. 10). This method has given satisfactory correction with sound skin closure and with realignment of the lower limbs that makes it possible to fit apparatus once more to allow walking.

![Image](https://via.placeholder.com/150)

**Fig. 8**

**FIGS. 8 AND 9**

Figure 8—Severe lumbar kyphos in a child aged 12 years. Until three years before she had been able to walk with the help of calipers, but increasing deformity had made it impossible for her to continue to use them. Figure 9—Lateral radiograph of the lumbar spine.

![Image](https://via.placeholder.com/150)

**Fig. 9**

In a less severe kyphos in which there is a liability to ulceration of the skin on either side of the midline at the point at which the processes protrude postero-laterally, improvement can be obtained by excision of the laterally directed pedicles and laminae. They are dissected away from the nerve roots in the same way as has been described for the first part of the dissection of the spine in the newborn. In two patients in whom this had been done the tendency to ulceration has been relieved.

**DISCUSSION**

No explanation has yet been offered for the development of kyphosis in association with myelomeningocele. Observation of the differences between the lesion in newborn children with and without kyphosis suggests that in those who have no kyphosis the defect is not so wide and there is still some extensor musculature on either side of the defect capable of producing active extension of the spine. In those with kyphosis the lesion is very wide, the erector spinae muscles are atrophic or absent and the quadratus lumborum is displaced postero-laterally so that it becomes a perverted flexor of the lumbar spine. It is suggested that the unopposed action of the abdominal muscles produces an intra-uterine paralytic lumbar kyphosis.

A good result as regards correction of deformity and improvement in lower limb paralysis has been obtained in two out of six; in the remaining four the result has been less than ideal and it is still too early to know how
lasting the results will be. Even so, the results of conservative management or of closure of the defect without spinal osteotomy have all been bad, and it may be that with further experience of the operation more consistent improvement may be obtained.

SUMMARY
1. The management of severe kyphosis of the lumbar spine in association with myelomeningocele is discussed.
2. Neonatal spinal osteotomy-resection has been performed in six patients with partial correction of the deformity and a greatly improved ease of closure and healing of the skin defect. The severity of lower limb paralysis has been diminished compared with the complete paraplegia that almost always results from conservative management of closure of the defect without osteotomy.
3. In an older child who has not had the benefit of neonatal osteotomy and who has complete lower limb paralysis, transverse spinal osteotomy or excision of the prominent laminae and pedicles on each side of the midline makes possible the fitting of apparatus for walking and diminishes the liability to recurrent ulceration of the skin.

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REFERENCE