OSTEOTOMY-EXCISION OF THE SPINE FOR LUMBAR KYPHOSIS
IN OLDER CHILDREN WITH MYELOMENINGOCELE

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In 1968 a technique of osteotomy-excision of the lumbar spine for congenital kyphosis in myelomeningocele in the newborn was described and the initial results were reviewed (Sharrard 1968). Since then the technique has been extended for the management of increasing kyphosis in older children in whom the operation had not been done immediately after birth. This paper describes the progressive development of deformity in these children and the methods found to be satisfactory for the management of the problems resulting from it.

ETIOLOGY

Sharrard (1968) noted that the quadratus lumborum and erector spinae muscles were displaced postero-laterally to become perverted flexors of the lumbar spine and that this was important in the development of kyphosis in myelomeningocele. A necropsy study of kyphosis in myelomeningocele (Drennan 1970) has indicated the importance of the psoas major and minor muscles and the crura of the diaphragm in causing further increase in kyphosis in the absence of any functioning extensors of the lumbar spine. The crura of the diaphragm cross the roof of the kyphos anteriorly to insert into the apical vertebrae (Fig. 1), and the remaining attachments to the diaphragm produce the crowning effect in the lower ribs in the antero-posterior radiographs described by Hoppenfeld (1967). As the kyphotic deformity increases, the effectiveness of the psoas muscles to produce further deformity increases, and the psoas attachment to the lumbar transverse processes (Fig. 2) causes them to rotate into the concavity of the curve, bringing the attachment of the thoraco-lumbar fascia with its enclosed erector spinae to act as a perverted flexor of the trunk.

Kyphosis in myelomeningocele is often associated with good innervation down to the third or fourth lumbar neural segment and with paralysis distal to this level. The hypertrophy of the psoas muscles associated with this level of neural innervation further aggravates the abnormal musculo-skeletal situation.

ANATOMY

Necropsy studies of kyphotic spines have confirmed and amplified the observations made by Hoppenfeld (1967). The abnormal pedicles and laminae in the area of the kyphos are splayed out and their tips are displaced anteriorly with the attached posterior layer of the lumbo-dorsal fascia. Normal zygo-apophysial joints are not developed: they are represented by cartilaginous joints between the laminae which become converted to bone in older children. The effect of this is to create intervertebral foramina similar to those seen in the sacrum, through which the nerves pass forwards into the substance of the psoas. The vertebral bodies do not show any increase in width in the kyphotic area. The apex, which is usually the third—or occasionally the fourth—lumbar vertebra, is wedged anteriorly, but this is thought to be secondary to pressure rather than a primary bone deformity.

Usually the kyphosis begins in the lower thoracic spine and extends to the sacrum. The dura mater covers the posterior aspect of the vertebral bodies and extends laterally where it is adherent to the splayed neural arches. The lumbar nerves pass almost directly forwards (Fig. 2) to the corresponding intervertebral foramina where they are accompanied by spinal
branches of the lumbar vessels. The aorta and inferior vena cava follow the anterior surface of the vertebral bodies and are not adherent to them or to the thickened anterior longitudinal ligament.

![Diagram showing the insertion of the crura of the diaphragm against which the psoas acts to aggravate and increase the kyphosis.](image1)

**Fig. 1**

Diagram to show the insertion of the crura of the diaphragm against which the psoas acts to aggravate and increase the kyphosis.

![Transverse section of the lumbar spine and its attached muscles in the region of the kyphosis.](image2)

**Fig. 2**

Transverse section of the lumbar spine and its attached muscles in the region of the kyphosis. The pedicles and laminae of the vertebrae are splayed out laterally. The erector spinae muscles enclosed in the thoraco-lumbar fascia lie lateral to the vertebral bodies and act as perverted flexors. The psoas muscle pulls the lumbar transverse processes forwards.

**PROGRESS IN UNTREATED CASES**

The radiographs of thirty-five spines with lumbar kyphosis at birth were reviewed. In almost all, the curves measured more than 80 degrees and extended from the first to the fifth lumbar vertebrae. The early stage at which the kyphosis develops is illustrated by the finding of kyphosis in a six weeks old foetus (Emery 1971).

No fixed compensating thoracic lordosis is present at birth. All the kyphoses in which spinal osteotomy was not performed at birth showed a steady increase which became more rapid after the end of the first year when the child started to sit (Figs. 3 and 4). By the end of the third year, all but one of the curves exceeded 120 degrees, and in three patients it was
more than 150 degrees. Three children, aged more than eight years, had curves exceeding 170 degrees, so that the anterior aspect of the first and second lumbar vertebrae came to lie against or to one side of the bodies of the fourth and fifth lumbar vertebrae.

At the junction of the thoracic and lumbar spines secondary lordosis develops which is at first mobile but later becomes fixed, though this is not invariable. The lower part of the rib cage is splayed and eventually the child sits with the rib cage astride the femoral shafts and may even develop an extension deformity of the cervical spine.

In association with increasing kyphosis, any function that might have been present in the lumbar nerves in the early months of life usually disappears, though innervation of the psoas muscle from the first and second nerves may be retained and aggravate the grossly abnormal posture by producing fixed flexion deformity of the hips. The skin covering the defect becomes thin and atrophic (Fig. 5), especially if the open myelomeningocele lesion had originally been allowed to heal spontaneously. It is almost impossible to prevent recurrent skin ulceration over the lateral processes at the apex of the kyphos. The child cannot lie on his back, and even when he sits in a chair the kyphos protrudes as a hump, rubs against the back of the chair however well padded, and completes skin break-down if this has not already occurred spontaneously. At first the ulceration may heal if the child is nursed prone, but in six patients the ulcer remained permanently unhealed and correction had to be performed in the presence of an open ulcer. If the ulceration extends to involve the dura there is a danger of recurrent meningitis.

The abdominal muscles and skin fail to grow and add to the fixity of the kyphosis. The combination of posterior displacement of the kidneys and vertical compression of the abdominal wall may preclude the performance of an ileal conduit operation in the management of urinary incontinence and reflux. In spite of the severity of the abnormality and the extent of paralysis in the lower limbs and bladder, many of those who survive beyond the age of three years continue to thrive. A high proportion of patients born with an extensive myelomeningocele with kyphosis have hydrocephalus of lesser or greater degree, and this was

FIG. 3
Severe lumbar kyphosis at the age of 3, exceeding 150 degrees. A secondary thoraco-lumbar lordosis is developing. The pelvis shows fixed flexion relative to the trunk.
true of all the patients in these series, whether treated at birth or later. All but two patients, whether treated in the newborn period or later, had had ventriculo-cardiac shunts. Their general level of intelligence was always below normal, but in older children the intelligence quotient was not less than 70.

![Image](image_url)

**FIG. 5**
Skin ulceration over a kyphotic lumbar spine in a poorly healed myelomeningocele.

**INDICATIONS FOR OPERATIVE CORRECTION**

The inevitability of progressive increase in the kyphosis and the difficulty in closure of an open myelomeningocele associated with kyphosis at birth make it advisable that osteotomy-resection of the spine should be done in the newborn at the time of surgical closure of the spinal defect. The technique of the latter has been described in an earlier paper (Sharrard 1968).

There are five indications for operative correction of kyphosis in older children. The commonest is chronic or recurrent skin ulceration, which may be associated with episodes of meningitis. Fixed flexion of the pelvis and the prominent kyphos may prevent the fitting of calipers for walking, or a child who has previously been able to walk for a limited distance in calipers may have to discard them because of increasing deformity. All children with kyphosis have bladder paralysis and urinary incontinence for which an ileal conduit diversion may be indicated for hydronephrosis, ureteric reflux, chronic urinary infection, chronic perineal excoriation, or for social reasons in girls. Such procedures may be difficult or impossible to perform if the kyphos becomes severe (Figs. 6 and 7) or, if successfully performed, the management of the ileostomy bag is made extremely difficult. A few children in whom primary closure of the spinal lesion has been possible in spite of the presence of the kyphosis at birth may have some active muscle function in the lower limbs from lumbar innervated muscles; progressive deterioration in such function as exists may occasionally be an indication for correction. Grotesque deformity and abnormal posture may in itself be an additional indication for correction. Occasionally, pain in the kyphotic spine or embarrassment of respiration may be additional indications. In most older children with severe kyphosis two or more of these indications have been present.

Contra-indications to operation may be severe uncontrolled hydrocephalus or recurrent ventriculitis, cardio-respiratory failure or incipient renal failure. In some children poor renal function with hydronephrosis or mental retardation seemed to contra-indicate operation, but because it was found that such children might survive to the age of ten years or more, this view has been modified and correction has been successfully obtained in four such children.
The aim of operation is not to obtain complete radiological correction of the deformity—which is never possible—but to eliminate recurrent skin ulceration, to improve posture and appearance and to provide access to the abdomen for the ileal conduit operation. A child previously confined to bed can at least be expected to be able to use a wheel-chair, and those who have previously been able to walk for a limited distance with calipers should be able to do so again.

**INVESTIGATIONS BEFORE OPERATION**

The renal function is assessed by examination of blood urea nitrogen and by intravenous pyelography. Any defects in electrolyte balance must be corrected. Urinary infection, which is often present, should be controlled by antibiotic drugs. The adequacy of any shunts to control hydrocephalus should be assured. The haemoglobin level of the blood is assessed and blood is cross-matched. The blood loss is likely to be about 50 per cent of the blood volume, though it may on occasions exceed this. Any deformity of the lower limbs that will make nursing of the spine in the prone or supine position difficult should have been corrected.

A lateral radiograph should be taken with the patient prone and sitting to assess the mobility of the spine above and below the level of the kyphosis. The vertebrae in the region of the kyphos almost always shows fixed deformity with ankylosis of the lateral processes. In older children there is usually a fixed thoraco-lumbar lordosis of between 45 and 90 degrees but this is not invariable; the lumbo-sacral angle may be normal or increased. An assessment should be made of the degree of fixed flexion of the pelvis relative to the thorax.

Provisional assessment can be made of the level of vertebral osteotomy and of the number of vertebrae to be removed (Figs. 8 and 9). The main aim of the operation is to decrease
the vertical height of the projecting kyphos and to correct the alignment of the pelvis relative to the trunk. Where there is fixed thoraco-lumbar lordosis, the aim should be to remove part of the apical vertebra and one or two vertebrae proximal to it, in order to correct both the kyphos and the lordosis. If the degree of thoraco-lumbar lordosis is not marked, correction may be obtained in the same way as in osteotomy-excision in the newborn, by removal of the apical vertebra and half of each vertebra above and below the apex.

THE OPERATION

Anaesthesia—After induction with Pentothal this is maintained with nitrous oxide, oxygen and Halothane with intubation. Intravenous infusion is begun. The patient is laid prone with pillows beneath the chest and pelvis, leaving the abdomen free to assist pulmonary ventilation and to diminish bleeding. In children below the ages of five to ten years, individually placed pillows are preferred to the use of a standard frame, the size of which cannot usually be accommodated satisfactorily to the kyphotic deformity. Blood loss is best estimated by a haemoglobin dilution-extraction technique and a positive balance of blood replacement is maintained throughout the operation in preparation for possible sudden blood loss, though this rarely occurs. Continuous electrocardiograph monitoring is advisable.

Skin incision—The skin at the operation site is always scarred, often extensively. If the original myelomeningocele lesion has healed spontaneously, the skin covering the apex of the kyphos is likely to be thin and closely adherent to underlying dura. If surgical closure has been made, it may have been achieved by means of flaps or by vertical or transverse closure. There may be a chronic ulcer over the lateral bony processes at the apex of the lesion.

Attempts to fashion an incision that follows previous incisions has been found to be unsatisfactory and the best results have been obtained by using a transverse incision crossing the apex of the kyphos, and enclosing and excising any chronic ulcer. After the prominence of the kyphos has been removed, there is likely to be more than sufficient skin available to allow inadequate skin to be excised at the end of the operation. Full thickness flaps are developed proximally and distally. At the apex of the lesion the skin may be very thin indeed and difficult to dissect from the dura. Any incision through the dura should be closed promptly to prevent excessive loss of cerebrospinal fluid that might precipitate cerebellar herniation in the presence of a Cleland-Arnold-Chiari deformity which is almost always present. The skin should be dissected sufficiently to expose the whole of the bony arch.
**Dural dissection**—Vertical incisions are made on each side of the midline following the oval margins of the bony defect. Incisions can be carried boldly down to bone without fear of incising nerve roots or vessels, all of which lie medially. Dissection is continued beneath the dura close to bone using a combination of blunt and sharp dissection. As the dissection proceeds medially there is usually no difficulty in establishing a loose plane between the posterior aspect of the dura and the vertebral bodies and pedicles (Fig. 2), but the dural layer is more adherent to the posterior longitudinal ligament and it may be safer to separate this ligament from the bone than from the dura if a dural leak is to be avoided.

A careful watch is kept for nerve roots passing downwards, forwards and slightly laterally to intervertebral foramina. The dura is mobilised carefully from the apical vertebra and from two vertebrae above and one below it. The corresponding nerve roots are identified. Sharp dissection close to the nerve roots should be avoided because substantial vessels are sometimes in their vicinity.

If there has been clinical evidence of complete lower limb paraplegia and if nerve root stimulation confirms the absence of any response in the lower limb muscles, the nerve roots and their accompanying vessels can be ligated and divided, the dura divided transversely just below the level of the apex of the kyphos and the meningeal layers sutured with a continuous catgut suture to prevent leakage of cerebrospinal fluid. The dural layer can then be mobilised proximally until a complete exposure of the kyphotic vertebrae is obtained.

If clinical assessment before operation has shown evidence of active muscle innervation in the lower limbs, individual roots may be stimulated with a nerve stimulator to establish their identity and to show whether they are capable of active conduction. If so, the dissection of the vertebrae needs to be made without transverse division of the dura, working between the neural tissues posteriorly and the vessels anteriorly. This dissection, though more tedious, is compatible with adequate resection of bone, though the bony fragments have to be removed from the side between the nerve roots.

The intervertebral foramina at the apex of the kyphos and above and below it are defined by passing a blunt dissector through each foramen. This instrument serves to protect the nerves and vessels during subsequent dissection. The thoraco-lumbar fascia, which is attached to the tips of the lateral processes, is incised and mobilised forwards. Beneath it, the erector spinae muscle passing on the lateral side of the kyphos is exposed and preserved. The anterior surfaces of the lateral processes are defined by blunt dissection and are removed with bone nibblers over the whole length of the spine where it is intended to perform osteotomy-excision. In doing so, the outer walls of the intervertebral foramina are removed, exposing the nerve roots passing from the dural layer forwards along the side of the vertebral bodies into the iliopectoas muscle. At least four nerve roots are normally exposed on each side.

**Exposure of the vertebral bodies**—Each nerve root is mobilised carefully from the side of the vertebral body to the point where it passes into the psoas muscle, which can now be seen. Using a blunt dissector, the anterior surface of the body of the apical vertebra is defined between the nerve roots proximal and distal to the apex. Dissection is made close to the vertebral body, pushing forward the contents of the apex of the kyphos which include the great vessels, though neither the aorta or inferior vena cava is firmly attached to the anterior surface of the vertebral bodies and, in severe kyphosis, they do not occupy the apical part of the bony arch. Surprisingly little bleeding is encountered during this part of the dissection and it is seldom necessary to ligate or coagulate any lumbar vessels. At the upper end of the incision the diaphragmatic crura can be identified and are mobilised forward as are the iliopectoas muscles, the dissection being extra periosteal. When this part of the dissection is complete it is usually possible to see right through the bony arch from side to side.

**Bone resection**—Division of the lumbar vertebrae is most easily made with an oscillating saw passed between adjacent nerve roots at the apex of the kyphos. Two cuts are made to allow removal of a wedged piece of the apical vertebra. Once this has been done, the distal fragment
sinks forward and the dura and other tissues become much more slack, allowing them to be retracted so that further vertebral bodies can be defined and resected. Removal of one and a half vertebrae above the level of the apical vertebra and half a vertebra immediately distal to the apical vertebra can then be achieved without difficulty, the bony cuts being made through the cancellous bone of the vertebra and not through the intervertebral disc. The lines of the osteotomies are so arranged that the two cut surfaces can be brought together with correction of the projection and restoration of the normal alignment of the pelvis. Approximation of the cut vertebrae is aided by hyperextending the lower limbs.

**Internal fixation**—A combination of three, four or five Blount staples on the dorsal and lateral aspects of the vertebrae with crossed oblique pins has proved to be the best method of internal fixation of the osteotomy. There has seldom been any difficulty in obtaining firm coaptation of the fragments. Cancellous bone obtained from the removal vertebrae is packed around the osteotomy site anteriorly. Any remaining prominent bone in the lateral processes above or below the level of the osteotomy is removed before the soft tissues are closed (Figs. 10 and 11).

**Closure**—The erector spinae enclosed in its layer of lumbo-dorsal fascia is mobilised posteriorly as far as possible. It has never been possible to suture the two erector spinae muscles across the midline but it should be possible to suture them to the thickened dura in such a way as to retain them as posteriorly placed as possible. As far as possible the subcutaneous layer is sutured separately from the skin layer. If a transverse incision has been used, there is seldom any difficulty in obtaining skin closure without tension and redundant skin can be excised. Two suction drainage tubes are inserted between the subcutaneous and deep tissue layers. Temporary external fixation is provided by a carefully moulded plaster shell extending from the scapulae to the buttocks and held in place by an elastic bandage.

**Fig. 10**
Radiographs of the lumbar spine before and after vertebral osteotomy-excision. The fixed flexion of the pelvis has been corrected and the osteotomy has been held by staples. Bone from the removed vertebrae has been packed anteriorly into the apex of the residual kyphos.
MANAGEMENT AFTER OPERATION

Blood loss during the operation has usually been replaced by the end of the operation but sometimes a further infusion of up to 50 millilitres of blood may need to be continued to allow for any further internal loss. Intravenous parenteral fluid is continued for up to forty-eight hours until normal fluid intake and output has been established and electrolyte balance has recovered. The child is nursed prone or obliquely on one side or the other on pillows with regular turning to avoid pressure ulceration. A watch is kept for possible respiratory complications or paralytic ileus, though neither have given cause for serious concern. Antibiotic drugs are only needed to control urinary or respiratory infection.

When skin healing is sound, posterior and anterior plaster bed shells can be made with care to position the lower limbs to avoid pressure or secondary limb deformity. Union of the osteotomy is usually sound by the end of three months and the child can be allowed to sit up. The lower limbs are mobilised with the aid of hydrotherapy and braces are fitted to allow walking.

COMPLICATIONS

Primary skin healing has occurred in all but one patient, a twelve-year-old boy who developed an indolent ulcer over the level of the osteotomy which slowly increased. At the tenth week the staples were removed and the ulcer healed after a further six weeks. Two patients developed secondary ulceration after uncomplicated primary skin healing but these healed after removal of the staples. One of these two patients had been allowed to return home one month after the operation in the plaster jacket and ulceration had occurred from pressure. In three patients one or two of the staples extruded during the period of immobilisation and one of the crossed pins extruded in two. Fixation remained sufficient to allow sound union without significant loss of correction.

RESULTS IN OLDER CHILDREN

Osteotomy-excision of the lumbar spine has been done in eighteen children between 1966 and 1970. The mean age was seven years, the youngest being three years and the oldest twelve years. There were twelve girls and six boys. In twelve patients the primary indication for operation was recurrent ulceration of the skin over the kyphos; in six prolonged conservative treatment had failed to heal the ulcer. Three children who had previously been able to use calipers had become unable to do so and two other children had become unable even to sit in a wheel-chair. One of these two also needed an ileal conduit operation for hydronephrosis. Another child had also the need for an ileal conduit as a primary indication. The eighteenth child, who had had the kyphos for many years, developed pain at the junction of the kyphos and a secondary thoraco-lumbar lordosis.

Two patients have been operated on too recently for full assessment of the result. Two have died from causes unrelated to the operation, five months and eighteen months after operation. In the remaining fourteen children recurrent ulceration has ceased in all except two, in whom there are small intermittent ulcers that arise if excessive pressure is applied to the lumbar area but which heal when pressure is removed. Nine children are now able to walk (Fig. 12) with the help of calipers with hip hinges and trunk support and using either elbow crutches or a walking aid. The two patients who were unable to use even a wheel-chair are now able to do so without the skin ulcerating and the two children who needed an ileal conduit have had this done successfully. The child in whom pain was a significant feature has had none since operation.

All the osteotomies have united. An attempt has been made to measure the radiographic improvement in the curve. This shows an average improvement of 33 degrees. This measurement is more an index of the correction of the alignment of the pelvis relative to the trunk than it is of correction of the bony prominence, which was always substantially improved.
(Fig. 13). In long-term follow-up there is a tendency to recurrence of deformity but at a much reduced rate compared with before operation, the average loss two years after osteotomy being 11 degrees.

RESULTS OF OSTEOTOMY-EXCISION IN THE NEWBORN

In 1968 the initial results of osteotomy-resection of the spine in the newborn were described in six babies; a further seven operations have been done since then. Three of these children have died, one at two days due to hypercapnia and the others at six weeks and fifteen months, from causes not associated with the spinal osteotomy. In two children the osteotomy failed to unite and was repeated, with subsequent union. All the survivors have satisfactory skin cover. In some the correction has been maintained, but overall there has been a tendency to a slow recurrence of deformity, with an average loss of 15 degrees during the first two years of life. This compares with a mean increase of over 45 degrees in the first two years in those who did not have an osteotomy-excision at birth. It is probable that some of the survivors of neonatal osteotomy-excision will need a further localised correction in later childhood.

DISCUSSION

All the older children in this series were already severely handicapped by extensive paraplegia. Most of them, because of the severity and extent of their spinal lesion at birth, had been expected to die during the early months of life but did not do so. In later childhood they had often been expected to die of renal complications, but, in spite of the fact that urinary diversion operations were often precluded by the deformity, they continued to survive, some of them almost into adolescence. The level of intelligence, though not normal, and the normal
function of the upper limbs were additional indications to strive for some improvement in their quality of life.

The primary aims of the operation—to relieve ulceration, to allow the possibility of limited walking with calipers or to sit in a wheel-chair rather than to be left lying prone permanently—have been achieved for every child. Because it has proved possible only partly to overcome the main factor in the development of the deformity—the absence of active extensors of the spine—the deformity is likely to recur, but much more slowly compared with the rate at which it was progressing before operation.

It was hoped that it might be possible to make some general comparison between the result of osteotomy-excision in the spine in the newborn compared with a similar operation done in older children, but a true comparison has not proved possible, because of the many factors involved and because the main indication for the procedure in the newborn is to assist in closure of the primary spinal lesion. It is possible that a compromise approach to the problem in some infants may be to obtain a primary closure of the spinal defect by excision of the prominent lateral processes and to await subsequent events. If the child proves to have a tenacious hold on life and survives beyond the second year, plans can be laid for osteotomy-excision with correction of the kyphosis before deformity becomes excessive and before the skin develops recurrent ulceration.

SUMMARY

1. The etiology and natural progress of lumbar kyphosis in children from three to twelve years of age with myelomeningocele are reviewed.
2. The indications for operation have included intractable or recurrent skin ulceration, inability to wear calipers for walking, inability to sit in a wheel-chair and inability to perform ileal conduit operations.
3. The technique of osteotomy-excision of lumbar vertebrae used in eighteen cases is described.
4. The results in fourteen children are described. The primary aims of operation have been achieved in all patients.
5. A comparison is made with the results of neonatal osteotomy-excision of the spine in the newborn. Recurrence of deformity, but at a much reduced rate, must be anticipated after either operation.

REFERENCES