An operative technique for correction of the severe kyphosis that may develop in cases of myelomeningocele is described. It includes excision of the apical vertebrae, correction of the full length of the kyphotic segment and of rigid compensatory curves, and extensive fusion aided by internal fixation with longitudinal threaded wires. The technique evolved during the management of eleven children. Of these, in the first two the internal fixation employed was inadequate; two others died; and in the remainder good correction was maintained up to the time of review one to six years later.

Children suffering from myelomeningocele with kyphosis who survive beyond the age of two years frequently present problems which can only be managed operatively. This paper describes a technique that aims to correct not only the primary kyphotic deformity but also the fixed lordotic curves above and below it, and to obtain sound fusion over the whole length of the section of the spine involved.

Hoppenfeld (1967) has reported an incidence of kyphosis at birth of one in eight cases of myelomeningocele. The pathological features of the condition have been described in detail by Hoppenfeld (1967) and again by Sharrard and Drennan (1972).

With regard to surgical treatment, Kilfoyle, Foley and Norton (1965) reported good results in two of three interbody fusions for kyphosis with paraplegia from varying causes. Sharrard (1968) described osteotomy-resection of the spine in the newborn, and in older children transverse osteotomy with overlapping of the fragments and fixation by four screws. For less severe kyphosis in which there was a liability to ulceration of the skin on either side of the midline, he described excision of the laterally directed pedicels and laminae. Sharrard and Drennan (1972) later reported the results of osteotomy-excision of the lumbar spine in eighteen children aged three to twelve years. Fusion was limited to the area of bone excision, where Blount staples and crossed oblique pins were used for internal fixation; compensatory curves were not corrected.

Sriram, Bobechko and Hall (1972) described a case effectively treated by excision of the vertebral body and fusion using Harrington compression apparatus inserted into the intervertebral foramina. Eckstein and Vora (1972) reported the results of spinal osteotomy in sixteen children under the age of eight years; they preferred not to use internal fixation. Five of the sixteen children died after operation.

An anterior approach to the deformity was described by Duncan, Lovell, Bailey and Ransom (1976), where the kyphosis was pulled forwards onto a rigid plate using special screws to transfix the vertebral bodies. McKay (1976) described a method of correction that did not involve excision of bone. The fact that the child regained normal height after this procedure could prove a disadvantage as a shorter child has a lower centre of gravity and therefore greater stability.

PRINCIPLES OF THE PROCEDURE

These were firstly, to remove the convexity of the kyphosis by the excision of vertebrae from the apex; secondly, to correct the residual deformity as completely as possible by extending the dissection proximally and distally and excising or incising the adjacent intervertebral discs; and thirdly, to provide adequate internal fixation by intramedullary wires and thus to promote fusion to the entire involved area of the spine in the corrected position.

In order to prevent recurrence of the deformity it is important that complete obliteration of the kyphos is obtained without tension in the soft tissues. The removal of bone facilitates this, although it is necessary to incise or excise adjacent intervertebral discs to correct any residual kyphosis, and similarly to correct rigid compensatory lordosis above the kyphosis and less commonly below it. As in scoliosis, the whole length of the spine involved in the deformity must be fused. The excised vertebral bodies, supplemented by bank bone, are used as bone grafts anteriorly, laterally, posteriorly and between the remaining vertebral bodies.
Indications for operation
The main indication is chronic or recurrent skin ulceration. However, correction gives subsidiary benefits. Without the kyphos a brace is more easily fitted and pressure over the unstable skin avoided; the child is able to sit in greater comfort; and there is improved access for urinary diversion (Figs. 1 and 2).

The mortality rate of the operation is too high for the last three factors to be regarded as indications unless there is also chronic or recurrent ulceration. Unlike Sharrard and Drennan, we have not operated to prevent neurological deterioration, painful kyphosis or gross deformity alone. In general, children with gross kyphosis are either paraplegic or have so little muscle power in the legs that there need be no concern regarding further loss of neurological activity. As for age, in our opinion the procedure is best performed between the third and sixth year.

Pre-operative management
Vigorous endeavours are made to heal ulceration over the kyphosis. This may involve not wearing any brace for some weeks, the use of protective pads and lying prone; and it may be necessary to admit the child to hospital for several weeks before operation.

Urine culture and tests for the antibiotic sensitivity of any organisms present are performed. A course of the appropriate antibiotic is commenced twenty-four hours before operation and continued until the wound has healed. If the urine is sterile, prophylactic antibiotics are used, cephalosporin being the one currently preferred. The volume of blood cross-matched before operation should exceed the patient's blood volume as the loss may range from 500 to 1500 millilitres.

OPERATIVE TECHNIQUE
The patient lies prone with pillows beneath pelvis and chest and the abdomen free. Blood loss is estimated throughout the procedure. Dilute adrenaline, one part in a quarter of a million, is injected along the line of the proposed incision. This is longitudinal and extends well beyond the limits of the kyphos; it may have to be modified to follow old suture lines of primary repair of the myelomeningocele. Although there is a redundancy of skin after excision of the kyphos, it is wise to preserve all of it until the end of the operation, and then to excise skin only if it is particularly redundant, thin and unstable.

The incision is deepened down to bone over the apex of the kyphos. The profuse bleeding is controlled by diathermy. The periosteum is raised at the apex of the kyphos and then laterally, giving flaps of maximal thickness. Nerve roots and spinal theca cannot be identified at this level. The subperiosteal dissection is carried proximally and distally. Proximally the dura will at last be opened, usually inadvertently as it is difficult to identify at its lowermost point. Having been identified, the dura is dissected off the back of the vertebral bodies and turned proximally a short distance; any opening made in it is carefully closed. Care is taken to avoid damage to nerve roots.

At this stage it is noted that the vertebral bodies are exceedingly wide and that the stumps of the pedicles lie so far laterally that the skin cannot easily be retracted to expose them. The lateral exposure is improved if bone is nibbled away from the apex of the kyphos in order to release tension on the flaps. At least one vertebra is removed from the apex of the kyphos and then the discs above and below are removed. The exposed bone of the vertebrae above and below is either rawed or more bone is excised according to the fit of the bones as the deformity is corrected. When bone has been excised from the midline of the apical vertebra, it becomes possible to nibble away bone further laterally both at this level and above and below it, so that the bulk of bone in the back is greatly reduced even if only one vertebral body has been excised.

Once the anterior longitudinal ligament has been exposed at the site of the excised vertebral body it is possible to strip the soft tissues off the front of the proximal and distal vertebrae. The intervertebral discs are then excised from between any vertebrae which form part of the residual kyphosis. If there is a fixed lordosis above or below it, then the intervertebral discs are excised at these levels also. This may be difficult or impossible in the upper portion of the wound without jeopardising intact nerve roots. It may only be possible to incise one or two discs at this level and then to correct the deformity by digital pressure.

The remaining vertebral bodies, thus converted into something like a string of beads, have been narrowed by excision of bone laterally and are held together by the anterior longitudinal ligament, which has been stripped only a short distance above and below the level of the kyphosis. The upper and lower segments of the spine are then brought to end to end and a decision made as to whether it is necessary to excise more bone. If there is an overlapping of bone another vertebral body is excised.
The resected vertebral bodies are nibbled into small fragments which are laid anterior to the remaining vertebral bodies, packed between them at the site of disc excision, and pressed into any lateral crevices. A small gutter may be made and packed with cancellous bone if this seems appropriate. Bank bone may be used to supplement the autogenous bone.

Three or four heavy threaded Kirschner wires (gauge 7/64 inch or 2.75 millimetres) have then to be passed along the length of spine to be fused (Fig. 3). It is generally easiest to pass these wires from above downwards, deferring an endeavour to correct the deformity at the site of bone excision until the wires emerge at this level. Then the upper and lower segments are brought together with the spine held in a position of lordosis, by digital pressure and by lifting the ankles off the table so that the pelvis is tilted. The wires are then passed distally into the caudal segment of spine. These wires are inserted high and laterally and cross the midline at the site of bone excision. If possible they are driven into the iliac crests. In some circumstances it may be easier to pass the wires in a cephalic direction; a puncture wound is made over the iliac crest and the wires passed proximally to emerge near the midline at the level of bone excision. The spine is then held in a position of lordosis whilst the wires are driven upwards. If there is doubt, the position of the wires is checked by radiography.

Once satisfactory internal fixation has been achieved more bone grafts are packed along the length of the spine to be fused, two suction drains are inserted, one deep and the other superficial to the vertebral bodies, and the wound is closed in layers.

Variations in technique. The first time this operation was performed a single long screw was used and proved inadequate. Unthreaded wires, Steinmann pins and Knowles pins have been employed, but the former two may migrate, and Knowles pins are more bulky and less flexible than the heavy threaded Kirschner wires. Staples were used to supplement longitudinal wires in two instances but are no longer employed.

One child had activity in the psoas and quadriceps muscles; the appropriate nerve roots were carefully identified and protected throughout the operation.

Postoperative management. The child is nursed on a Bradford frame for three months (Figs. 4 to 7), most of the time prone, but supine for a few hours every day. With three exceptions these children have been nursed by the parents at home after the first three weeks.

RESULTS

Eleven children were treated by the technique described in the period 1970 to 1975, all the operations being performed by one of the authors (MBM). The average age at operation was three and a half years, the youngest child being two and a half and the oldest five and a half (Table 1).

The time of follow-up on the nine survivors averaged three years, with a range of one to six years. The early results in three of these patients have already been published (Menelaus 1971). All but two have now been reviewed personally by one of the authors (GL). Case 1 was reviewed to some extent by correspondence with the child's parents and Case 8 with the aid of a colleague in another State. The clinical result was considered satisfactory if problems of recurrent skin

Fig. 3
Case 9. Radiographs taken after operation showing the heavy threaded Kirschner wires that are now preferred for internal fixation.

Fig. 4
Case 4. Figure 4—The appearance before operation. Figure 5—The same child immediately after operation being nursed prone on a Bradford frame. The bulk of the kyphosis has been greatly reduced.
Fig. 6

Case 4. Figure 6—A lateral radiograph of the kyphosis before operation. Figure 7—A lateral radiograph taken immediately after operation with fixation by Knowles pins. This child died two and a half months later (see text).

Table I. Clinical details of the eleven children

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Age at operation (years)</th>
<th>Indications for operation</th>
<th>Condition of local skin at operation</th>
<th>Internal fixation</th>
<th>Follow-up (years)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>4½</td>
<td>Ulceration. Marked kyphosis. Difficulty with sitting</td>
<td>Scarred</td>
<td>1 screw</td>
<td>6</td>
<td>Loss of fixation. Relapse. Failure</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>5½</td>
<td>Ulceration</td>
<td>Old skin graft</td>
<td>3 Steinmann pins</td>
<td>6</td>
<td>Extrusion of pins. Relapse of kyphosis. Harrington fusion later. Unsatisfactory result</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>3</td>
<td>Ulceration</td>
<td>Scarred</td>
<td>3 Knowles pins</td>
<td>6</td>
<td>No further ulceration. Correction maintained</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>2½</td>
<td>Ulceration. Progressive kyphosis</td>
<td>Scarred</td>
<td>4 Knowles pins</td>
<td>—</td>
<td>Good correction. Died two and a half months after operation from urinary infection. Back explored at necropsy—fusion sound</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>3</td>
<td>Ulceration</td>
<td>Scarred</td>
<td>2 Knowles pins+staple</td>
<td>4</td>
<td>No further ulceration. Correction maintained</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>3</td>
<td>Ulceration</td>
<td>Scarred</td>
<td>3 Knowles pins+staple</td>
<td>4</td>
<td>Ulceration relieved. Good correction. Minimal relapse</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>3</td>
<td>Ulceration. Progressive kyphosis</td>
<td>Scarred</td>
<td>3 Knowles pins</td>
<td>—</td>
<td>Wound breakdown and infection. Septicaemia. Died one month after operation</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>2½</td>
<td>Ulceration</td>
<td>Scarred</td>
<td>4 unthreaded Kirschner wires</td>
<td>3</td>
<td>Good correction though slight relapse. Ulceration relieved</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>3</td>
<td>Ulceration</td>
<td>Ulcerated</td>
<td>3 threaded Kirschner wires</td>
<td>2</td>
<td>Ulceration relieved. Good correction</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>4</td>
<td>Ulceration. Poor sitting posture</td>
<td>Old skin graft</td>
<td>3 threaded Kirschner wires</td>
<td>1</td>
<td>Ulceration relieved. Only fair correction. Non-union and progressive relapse</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>4</td>
<td>Ulceration</td>
<td>Old skin graft Ulcerated</td>
<td>2 threaded Kirschner wires +2 Knowles pins</td>
<td>1</td>
<td>Good correction. Recurrent ulceration over pedicles causing minor trouble</td>
</tr>
</tbody>
</table>
ulceration had been relieved and the gross degree of kyphotic deformity reduced.

Radiological assessment was attempted by the comparison of lateral radiographs of the spine. Measurements were made by the Cobb method, the angle of kyphosis being that made by the intersection of lines drawn through the end plates of the end vertebrae of the kyphosis. Distortion of the anatomy by the of the operation. Both had had good initial correction (Figs. 4 and 5).

Non-union occurred at the lower end of the fused area in one child (Case 10) and a slow relapse of the deformity followed, but skin ulceration did not recur.

In the other six children the clinical results were satisfactory, with no further ulceration over the kyphosis and only slight loss of correction in two cases.

![Fig. 8](image1)

Case 6. Figure 8—A lateral radiograph of the kyphosis before operation. Three Knowles pins have been removed. The relapse of deformity has been quite minimal.

![Fig. 9](image2)

Fig. 9

excision of vertebral bodies made the postoperative measurements quite inaccurate, however, and more emphasis was therefore placed on the clinical assessment.

Pre-operative condition. Skin healing was obtained in all but two children in whom it was necessary to proceed despite the ulceration. Otherwise the skin was either of poor quality due to previous scarring or unstable from previous skin grafting procedures.

Clinical results
Two operations have failed. In Case 1 a single screw was used, but the fixation soon failed and the deformity recurred. Nevertheless the skin did not ulcerate again, presumably because of a reduction in the bulk of the lump. In Case 2 three Steinmann pins were used for fixation and good immediate correction was obtained. Within three months, however, all the pins had extruded spontaneously and the kyphosis relapsed. The development of a scoliosis later required a posterior spinal fusion and Harrington instrumentation to a higher level.

Two children (Cases 4 and 7) died less than three months after the operation, one as a direct consequence Two children had minor breakdown of the skin over the pedicles well lateral to the midline, and one will require excision of the prominent pedicles. After this limited procedure a child can resume walking in his usual appliances within a week.

Radiological results
Radiographs showed all but one of the fusions to be solid (Figs. 8 and 9). Kyphosis measurements showed the average degree of deformity to be 109 degrees before operation, and immediately after about 65 degrees. Of the six children with a satisfactory clinical result the correction was fully maintained in four.

Complications
Deaths. Two children have died since operation. One (Case 4) died two and a half months later from a severe urinary tract infection. At necropsy the site of fusion was exposed by the referring orthopaedic surgeon, who reported solid fusion with no evidence of infection. The second child (Case 7) died a month after operation from septicemia secondary to necrosis of a large area of skin at the site of operation.
Problems with fixation devices. Unthreaded Steinmann pins and Kirschner wires tended to migrate. Steinmann pins ulcerated through the skin and needed early removal. In Case 8 unthreaded Kirschner wires caused an abscess in the buttock which resolved after removal of the wires. In five other children actual or imminent ulceration over the end of a single wire necessitated its removal three to four months after operation. Fracture of the pins occurred twice; portions have been left in situ and have caused no problems up to the time of review.

DISCUSSION

The treatment of spinal deformity in myelomeningocele is made difficult by densely adherent and scarred soft tissues and by the degree of rigidity of both primary and compensatory curves (Sriram, Bobechko and Hall 1972). In cases of kyphosis, thin, anaesthetic, unstable, previously grafted or ulcerated skin over the apex presents an additional difficulty. Failure of skin healing and recurrence of deformity may be expected unless tension is relieved by reducing the bony bulk. This requires the removal of several vertebral bodies at the apex of the kyphosis, a principle established by Sharrard and Drennan (1972).

The operation described here creates a spinal fusion over a long extent, thus overcoming the tendency to recurrence after local fusion (Sharrard and Drennan 1972); the rigid lordosis proximally is corrected by disc incision or excision and incorporation in the fusion. The extent of the fusion raises the problem of internal fixation, for which multiple heavy threaded Kirschner wires have to date proved the most efficient.

The operation described is an extensive procedure for children who often have other medical problems. Blood loss is heavy and the risk of a fatal outcome considerable. For this reason we suggest that the operation be considered only when conservative treatment fails to prevent recurrent ulceration over the kyphosis. Unfortunately, ulceration is generally initiated by pressure from bracing, and healing requires a prolonged period in which the child is unable to walk. Should ulceration recur when walking is resumed, operation is usually indicated.

No long-term results of treatment of kyphosis in myelomeningocele have been reported. In this small series, children followed for up to six years have maintained the clinical improvement and the tendency to ulceration has been almost completely relieved. Up to date any slight relapse in deformity has not raised a further problem.

REFERENCES


