Congenital kyphosis in myelomeningocele
THE EFFECT OF CORDOTOMY ON BLADDER FUNCTION
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To determine the effect of cordotomy on the function of the bladder during surgical correction of congenital kyphosis in myelomeningocele, we reviewed 13 patients who had this procedure between 1981 and 1996.

The mean age of the patients at operation was 8.9 years (3.7 to 16) and the mean follow-up was 4.8 years (1.3 to 10.8). Bladder function before and after operation was assessed clinically and quantitatively by urodynamics.

The mean preoperative kyphosis was 117° (52 to 175) and decreased to 49° (1 to 89) immediately after surgery. At the latest follow-up, a mean correction of 52% had been achieved.

Only one patient showed deterioration in bladder function after operation. Eight out of the nine patients who had urodynamic assessment had improvement in bladder capacity and compliance, and five showed an increase in urethral pressure. One patient developed a spastic bladder and required subsequent surgical intervention.

Cordotomy, at or below the level of the kyphosis, allows excellent correction of the structural deformity.

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Congenital kyphosis is the most severe spinal deformity associated with myelomeningocele and is seen in 12% to 20% of patients. It is caused by anomalous vertebral development in addition to the spina bifida. The deformity is present at birth with an associated visible gibbus in about 10% of cases and is usually located in the upper lumbar spine. The kyphotic curve progresses markedly in the first years of life and a compensatory lordosis rapidly develops above the kyphosis.

The erector spinae muscles, quadratus lumborum and the thoracolumbar fascia are located anterior to the pedicles where they act as flexors of the spine leading to a fixed deformity.

Treatment using spinal braces is difficult. Children with kyphosis have trouble sitting in a wheelchair and develop ulceration over the prominent spinal elements. Progression of the curve occurs, despite the use of braces, and may occasionally cause both feeding and breathing difficulties secondary to increased intra-abdominal pressure. Most patients are incontinent with flaccid bladders, and the increased flexion of the trunk further interferes with the drainage of urine. The spinal deformity may lead to technical problems with placement of a urethrostomy or vesicostomy.

Surgical correction of congenital kyphosis is a formidable procedure and a number of methods have been described. Typically, it involves a two-staged anterior and posterior approach without disturbance of the spinal cord, but can be achieved using a single-stage circumferential exposure through a posterior incision. This is the procedure which we used and includes transection of the spinal cord as well as a spinal osteotomy and vertebral resection to shorten and straighten the spine in preparation for instrumentation. Transection of the spinal cord (cordotomy) greatly enhances the ability to obtain correction of the structural deformity. Although it may have the added orthopaedic benefit of reducing spasticity in the limbs, it also has the potential to alter bladder function.

Our aim was to review the results of the surgical correction of congenital kyphosis in myelomeningocele with cordotomy to determine the effect on bladder function.

Patients and Methods

Between 1981 and 1996, 13 patients with myelomeningocele and a severe kyphotic deformity had surgical correction of the kyphosis with cordotomy at the Children’s Hospital of Eastern Ontario.

Information regarding the reason for kyphectomy, the age at operation, the surgical technique and complications were obtained from the hospital records.

Anteroposterior and lateral radiographs were taken with the patient sitting whenever possible, and measurements were obtained at the initial presentation, before operation,
after operation and at the latest follow-up. The severity of the kyphosis and the lordosis cephalad to it were measured on the lateral radiograph. 13 The height of the kyphosis was determined by the perpendicular distance from the dorsal edge of the apex to a line drawn from the posterior aspect of the first lordotic vertebra of the thoracic spine and the posterior aspect of the first sacral vertebra. 13 Any accompanying scoliosis was measured on the anteroposterior radiograph using the Cobb technique.

Patients were seen on a regular basis in the urology clinic and urodynamics laboratory by one of two urologists and a clinical nurse. Urine samples were taken from all patients for culture and sensitivity before surgery. Urodynamic evaluation included monitoring of urethral pressures and cystometry which analyses bladder capacity and compliance. Studies on bladder filling and voiding depicting accepted age- and gender-adjusted values for bladder capacity, compliance and a urethral pressure profile were used to interpret the results of urodynamic testing. 14-16

Operative technique. The number of vertebrae to be resected was determined before operation; the extent of the resection was sufficient to remove the lordotic segment cephalad to the kyphosis. The operation was carried out under general anaesthesia. Patients were placed prone and operated on using a posterior approach. All patients received prophylactic antibiotics perioperatively.

Through a longitudinal incision, the paraspinal muscles were elevated subperiosteally from the posterior spinal elements. The dura and remnants of the spinal cord were identified. The cord was elevated subperiosteally from the back of the vertebral bodies, both cephalad and caudal. Nerve roots below the neurological level were clamped and cauterised. The spinal cord was transected at the apex of the kyphosis. The cord was occasionally resected depending on the preference of the surgeon. The ends of the dural sac were then oversewn with silk.

A circumferential exposure of all three columns of the spine was carried out at the apex of the kyphosis. The amount of shortening required was determined based on preoperative radiographs and on the intra-operative flexibility of the spine. Correction of the kyphosis was obtained either by excision of the vertebral body or by removal of the disc and end plates. Step-cut osteotomies were carried out on the vertebral bodies immediately above and below the kyphectomy to allow them to interdigitate. On several occasions, they were held together at the site of the osteotomy using 18-gauge wire passed through the neural foramina and around the pedicles.

Stabilisation after correction of the kyphosis was achieved by one of four techniques; Harrington compression rods, Luque rods with the Galveston fixation to the pelvis, interforaminal wiring alone 11,17,18 or by the method described by Torode and Godette. 19 In the Torode technique, two holes are drilled in the most posterior portion of the most cephalad lumbar vertebral body and passed caudally into the anterior portion of the body of the fifth lumbar or first sacral vertebra. Rods, 6.2 mm in diameter, are passed into these holes and their position checked on posteroanterior and lateral radiographs of the lumbosacral spine. The thoracic spine was prepared as for a Luque fusion with sublaminar wires and/or Wisconsin button wires, depending on the integrity of the posterior elements. 19

The excised vertebral bodies provided adequate bone graft for fusion of the spine over the length of the instrumentation. Bank bone was occasionally used in addition to autogenous bone.

The dural sac was returned to its preoperative position. The kyphectomy allowed the thoracolumbar spine to translate anteriorly, restoring a more normal sagittal alignment and making it possible to approximate the paraspinal muscles over the spine posteriorly. If necessary, redundant skin was excised and the remaining skin closed with sutures.

After operation the patients remained in bed, prone or in the lateral decubitus position, until the wound was dry. After seven to ten days, they were fitted with a custom polypropylene spinal brace and allowed to sit upright. For some patients, a thigh extension was added to the brace if the fixation was felt to be tenuous. The brace was used for nine months to one year after the operation, except when the patient was recumbent. Care was taken to monitor the pressure areas under the brace.

There were nine girls and four boys. The mean age of the patients at the time of the operation was 8.9 years (3.7 to 16). One patient died four months after the operation and another was lost to follow-up after four months. The mean follow-up for the remaining 11 patients was 4.8 years (1.3 to 10.8).

Results

All patients had undergone closure of the myelomeningocele in the neonatal period and insertion of a ventriculoperitoneal shunt. One patient had had a previous operation for the kyphotic deformity with an anterior release and cortical strut graft which was revised through a posterior approach for progression of the kyphosis.

The neurological levels in the 13 patients were T8 in one, T9 in one, T10 in six, T12 in two, L1 in one and L3 in two. The range of movement of the hips before operation was sufficient to allow sitting after. The gibbus was at T12 in one patient, L2 in three, L2 to L3 in two, L3 in six and L4 in one. Eight of the 13 patients had pressure sores over the apex of their kyphosis before surgery. Three required insertion of tissue expanders for skin coverage.

The indication for operation was increasing spinal deformity in five patients, repeated skin breakdown in four and poor sitting balance in four. None had symptoms of abdominal compression or pulmonary compromise.

The mean length of the operation was 6.8 hours (5.00 to 8.6). The mean estimated blood loss was 1805 ml (350 to 4000) and the mean amount of blood transfused was 5 units (3 to 8).
The number of vertebrae excised depended on the severity of the kyphosis. It ranged from zero, with only the discs and endplates excised, to six.

Stabilisation, after correction of the kyphosis, was achieved with Harrington rods in one patient, Luque rods with the Galveston technique in three and by interforaminal wiring alone in two. The remaining seven had spinal stabilisation using the technique described by Torode and Godette.\(^\text{19}\)

The mean kyphosis at presentation was 82° (30 to 147); immediately before surgery, it was 117° (52 to 175). The mean deformity in the immediate postoperative period was 49° (1 to 89) while at the latest follow-up it was 55° (1 to 88). This represents a mean correction of 52%.

The mean height of the kyphosis was 3.0 cm (0.6 to 6.1) at presentation and 5.2 cm (2.0 to 7.9) immediately before surgery. Immediately after operation, it was 2.2 cm (0.2 to 4.0) and at the latest follow-up, 3.5 cm (0.4 to 5.2).

The mean thoracic lordosis above the kyphosis was 44° (9 to 100) at presentation and 57° (15 to 100) immediately before surgery. Immediately after surgery, it was 29° (10 to 51), and at the latest follow-up, 27° (6 to 38).

The mean magnitude of the scoliosis immediately before surgery was 22° (2 to 64). Only one patient had a curve severe enough to warrant specific attention. She had congenital scoliosis with multiple rib anomalies and hemi-vertebrae with no pedicles on the left side at T2, T3 and T4. Immediately before operation, she had two curves (58° to the left C7 to T7, 64° to the right T9 to L3). She had a five-level anterior release of the upper thoracic spine one week before the correction of her kyphotic deformity. At the latest follow-up, the curves measured 60° and 56°, respectively.

All patients had functioning ventriculoperitoneal shunts at the time of surgery. After operation, no problems with hydrocephalus occurred as a result of the cordotomy.

**Bladder function.** Pre- and postoperative information on bladder function was available for 12 of the 13 patients. The one patient with incomplete urological follow-up died from intraoperative complications shortly after surgery. All patients had a flaccid bladder at the time of presentation. Most were dry between clean intermittent catheterisations with occasional use of oxybutynin chloride and/or phenylpropanolamine hydrochloride to help with bladder control. There was deterioration in bladder function after kyphectomy and cordotomy in only one patient. Currently, three patients are taking either phenylpropanolamine hydrochloride or oxybutynin chloride. Two of these were already on these drugs before operation.

Pre- and postoperative urodynamic testing was carried out in nine patients. The values for bladder capacity and compliance are outlined in Table I. All but one patient (case 1) showed improvement in bladder capacity and compliance after surgery. Of the nine patients who had evaluation of urethral pressures during voiding, chronic urinary infection rendered interpretation of postoperative data meaningless in cases 2 and 6 (Table I). Another patient (case 5) without urinary infection showed deterioration in urethral pressures after surgery. Although no clear explanation for this could be found, there was a favourable response to phenylpropanolamine hydrochloride. The remaining six patients showed improvement or maintenance of the urethral pressure profile after operation.

One patient (case 1), without evidence of infection of the urinary tract, had postoperative deterioration in bladder capacity and compliance (Table I). This patient, with a T10 neurological level and small bladder capacity, good compliance and a low urethral pressure profile on preoperative urodynamics, developed a spastic bladder and urinary incontinence refractory to oxybutynin chloride after surgery. The spinal cord had been transected at the apex of the kyphosis at L2 to L3. It was dissected proximally to T11 and distally to L5. It was left attached distally. The cord and dura were transected at T11 and oversewn. The patient required multilevel bilateral sacral rhizotomies and transection of the neural placode 28 months after surgical correction of the kyphosis. Marked scarring and epidural adhesions were noted. Only the highest left nerve root was generating bladder activity on intraoperative urodynamics, suggesting that the residual cord segment was tethered and functioning autonomously causing spasticity. Bladder function was satisfactory after the rhizotomies.

**Complications.** During the operation, one patient had hypovolaemia and hypothermia because of interstitial displacement of the central venous line. This resulted in severe

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**Table I.** Preoperative and postoperative bladder capacity (ml), bladder compliance and the urethral pressure profile for the nine patients who had urodynamic assessment

<table>
<thead>
<tr>
<th>Case</th>
<th>Bladder capacity</th>
<th>Bladder compliance</th>
<th>Urethral pressure profile</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Preop</td>
<td>Postop</td>
<td>Preop</td>
</tr>
<tr>
<td>1</td>
<td>190 (315)*</td>
<td>105 (420)</td>
<td>Good</td>
</tr>
<tr>
<td>2</td>
<td>200 (240)</td>
<td>480 (550)</td>
<td>Good</td>
</tr>
<tr>
<td>3</td>
<td>220 (405)</td>
<td>346 (480)</td>
<td>Fair</td>
</tr>
<tr>
<td>4</td>
<td>172 (510)</td>
<td>352 (600)</td>
<td>Poor</td>
</tr>
<tr>
<td>5</td>
<td>Low (165)</td>
<td>Adequate (195)</td>
<td>Good</td>
</tr>
<tr>
<td>6</td>
<td>318 (390)</td>
<td>318 (465)</td>
<td>Excellent</td>
</tr>
<tr>
<td>7</td>
<td>130 (225)</td>
<td>260 (405)</td>
<td>Fair</td>
</tr>
<tr>
<td>8</td>
<td>185 (135)</td>
<td>200 (150)</td>
<td>Good</td>
</tr>
<tr>
<td>9</td>
<td>300 (330)</td>
<td>365 (360)</td>
<td>Good</td>
</tr>
</tbody>
</table>

* age-adjusted normal values in parentheses
† presence of infection of the urinary tract
diffuse cerebral ischaemia. The patient died from respiratory failure a few months later.

In the immediate postoperative period, four patients developed skin breakdown either along the incision or at the site of the previous gibbus. In three, local wound care and removal of the brace resulted in uneventful healing. One patient who had segmental necrosis required excision and closure of the defect by an advancement flap on the 12th postoperative day. Skin breakdown occurred in six patients during the intermediate follow-up period. In four this was felt to be secondary to bracing which was discontinued temporarily while the wounds were managed with dressings. In the other two patients, skin problems were related to prominent instrumentation. One patient had removal of a Wisconsin button wire at one level. Another with complete pull-out of the rods at L5 and local skin breakdown was managed conservatively and the rod was left in situ. There has not been any progression of the kyphosis and at the latest follow-up, 5.8 years after surgery, the patient was doing well with no skin or seating problems.

One patient had partial pull-out of the rods at L5 without skin problems. Another, with a 108° kyphosis, which corrected to 50° after kyphectomy and Harrington compression rods, had a broken rod one year after operation. This was associated with progression of the kyphosis to 60° and a small pressure sore over the lower back. A bivalved cast brace was reapplied once the skin had healed. Revision of the kyphectomy and posterior spinal fusion with Luque instrumentation were carried out at another hospital two years after the initial operation. At 10.8 years after the original procedure, the kyphosis measured 73°, which represents a long-term correction of 32%.

There were no superficial or deep wound infections.

Discussion

Surgical correction of congenital kyphosis can be accomplished by either a two-staged anterior and posterior procedure or a single-stage circumferential operation through a posterior incision, as used in our study. Although technically demanding, with a high potential for complications, surgical correction of kyphosis usually provides considerable benefits including reduction of soft-tissue problems, improved sitting balance, and improved urological function. Based on these criteria, 11 of our 13 patients achieved a good or excellent result. Ten, however, developed skin breakdown either immediately or shortly after kyphectomy underlying the potential for ongoing skin problems in these patients. Two of these patients had considerable difficulty and required aggressive management to resolve the problem. In spite of the need for early intervention, these two patients have achieved a good overall result with no further skin breakdown. Correction of the bony deformity should eliminate recurrent skin problems if long-term use of a brace is avoided.

It has been generally agreed that some loss of correction of the kyphosis after operation is to be expected with time. Progression of the deformity, however, is probably less than that of an untreated kyphosis. There appears to be gradual complete bony union in all patients, although radiological evaluation is unreliable. The broken rod in one patient suggested a pseudarthrosis at the distal aspect of the fusion mass, but radiographs at the final follow-up showed consolidation of the graft over the entire length of the fusion, with no recent progression of the kyphosis. The mean correction of 52% achieved at final follow-up is comparable to that quoted in the literature.

Cordotomy greatly enhances the ability to obtain correction of the structural deformity. Although it may have the added benefit of reducing spasticity in the limbs, it also has the potential to alter bladder function. Clinically, the latter was unchanged in all except one patient. Patients were dry between clean intermittent catheterisations. Urodynamics studies before and after surgery confirmed maintenance or improvement in bladder capacity and compliance after kyphectomy with cordotomy in eight out of nine patients. Although this can be partially explained by the natural increase which accompanies an increase in age, correction of the kyphotic deformity is important for reducing persistent compression of the bladder and allowing an overall gain in bladder capacity.

The urethral pressure profile seems to be less affected by the physical constraints resulting from lumbar kyphosis and subsequent abdominal crowding, but appears more sensitive to internal factors, such as urinary infection. Two patients with chronic persistent urinary infections during postoperative urodynamic testing had results for measurement of urethral pressure which could not be interpreted satisfactorily. All except one of the remaining patients, showed improvement or maintenance of their profiles for urethral pressure after surgery. The reason for the mild deterioration in pressures in one patient was not understood.

In all patients cordotomy was carried out at the apex of the kyphosis, which was located below their neurological level. In the patient who developed a spastic bladder and urinary incontinence refractory to oxybutynin chloride after operation, the residual segment of the cord after kyphectomy and cordotomy was tethered, resulting in an autonomic bladder which required subsequent surgical treatment. It is difficult to determine clinically before operation which patients with high-level myelomeningocele have a tethered cord, since they have minimal or no neurological function in the lower legs and a flaccid bladder. A high index of suspicion should be maintained in these patients, particularly if the management of bladder maintenance becomes problematical. Preoperative MRI of the spinal cord will demonstrate any tethering, which can be dealt with at the time of cordotomy.

Cordotomy, coupled with surgical excision, allows good correction of congenital kyphotic deformity in myelomeningocele. Careful perioperative follow-up has shown...
that deterioration of bladder function was rare in our study. Kyphectomy with cordotomy improves bladder function, provided that the cordotomy is carried out at the apex of the kyphosis below the neurological level and there is no tethering of the cord.

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References


