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Kyphectomy in Children With Severe Myelomeningocele-Related Kyphosis

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Abstract

Study Design: Retrospective review of prospectively maintained database.
Objectives: To review myelomeningocele patients with severe kyphosis undergoing kyphectomy surgery in terms of complications and clinical and radiographic outcomes.
Summary of Background Data: Because of posterior element abnormality in myelomeningocele, the extensor muscles act as perverted flexors, driving progressive kyphosis that resulted in sitting, respiratory, and skin breakdown problems.
Methods: Clinical case notes and x-rays of seven myelomeningocele patients undergoing kyphectomy surgery were reviewed with a minimum follow-up of 24 months. They consisted of four males and three females with an average age of 9.5 years at surgery. Surgery was performed in three despite open pressure ulcers that failed to heal. These wounds were all closed primarily at initial operation, and no flaps were required. Pedicle screw and sublaminar wire constructs were utilized with iliac screws for distal control.
Results: The median surgical time was 245 minutes (165–285), with an estimated blood loss of 700 mL (500–2,550). The preoperative kyphosis of 142 degrees (90–180) was corrected to 15 degrees (5–45) representing a 92% correction. All experienced improved sitting. There were no early complications but 2 patients with preoperative pressure ulcers returned at 13 months with recurrent sepsis and wound breakdown. Their osteotomy had fused, and the infection settled after instrumentation removal and antibiotic administration.
Conclusion: Although an infrequent presentation today, severe kyphosis in myelomeningocele patients causes not only a major functional impairment but threat to their life with apical pressure sores. Kyphectomy and posterior instrumented spinal fusion can be performed safely, even in the face of an open sore with excellent kyphotic correction and resultant improved functionality and ability to sit. These open sores can be closed primarily without the requirement of plastic surgery as a result of the shortening and extension of the spine.

Keywords: Kyphectomy; Myelomeningocele; Spina bifida; Kyphosis

Introduction

Myelomeningocele is the most common congenital birth defect in the Western Cape, South Africa [1], with a reported incidence of 1 to 2.5/1,000 patients. This defect in neural tube formation occurs within the first 3 to 4 weeks of human embryogenesis and results in failure of closure of the neural structures forming the spine and subsequent paraplegia. With growth, this leads to spinal deformities such as scoliosis and kyphosis [2-4].

The incidence of kyphosis in myelomeningocele patients ranges from 8% to 20%. The congenital rigid deformity occurs within the thoracolumbar region, with kyphotic curves often more than 80 degrees at birth, which progress at 6 to 12 degrees per year [5-12]. With the deficient posterior spinal elements, the erector spinae muscles migrate anteriorly and act as perverted flexors, driving progression [1,2,6].

This kyphosis impairs activities of daily life as a result of the inability to sit upright, forcing the use of their arms to support their trunks. The pelvis is flexed forward, resulting in a sacral sit with pressure ulcer risk. The prominent

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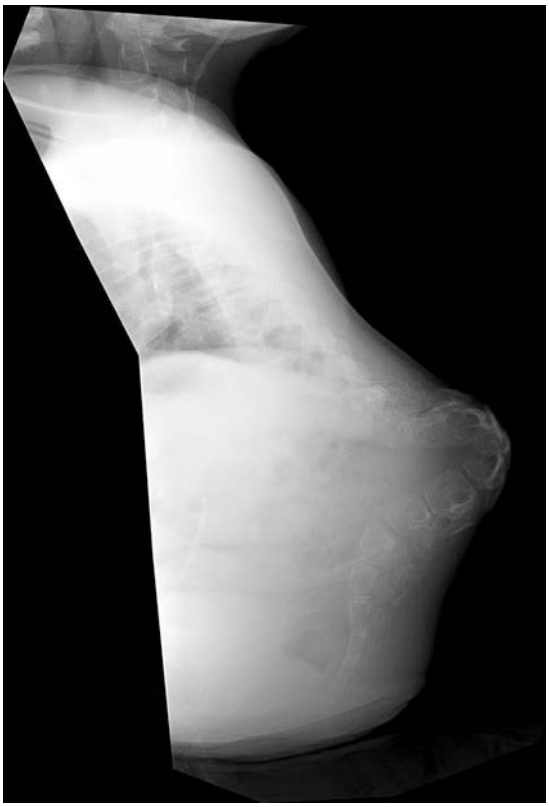

Fig. 1. Preoperative x-ray demonstrating the severe kyphosis.

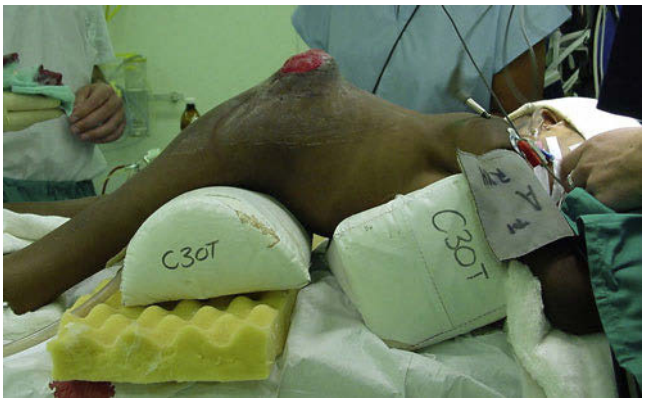

Fig. 3. Preoperative theatre positioning indicating severe kyphosis, flattened buttocks, and open pressure wound that failed to heal despite avoidance of pressure and application of dressings.

gibbus abrades against the chair-back, resulting in skin breakdown (Figs. 1–3). Respiratory impairment and early satiety result from the abdominal contents restricting diaphragmatic excursion [1,6,10].

Once severe, seating and bracing are ineffectual, and surgical correction should be considered [6,10,11,13].

We present our experience in managing this challenging group of patients with respect to technique, complications, and outcomes.

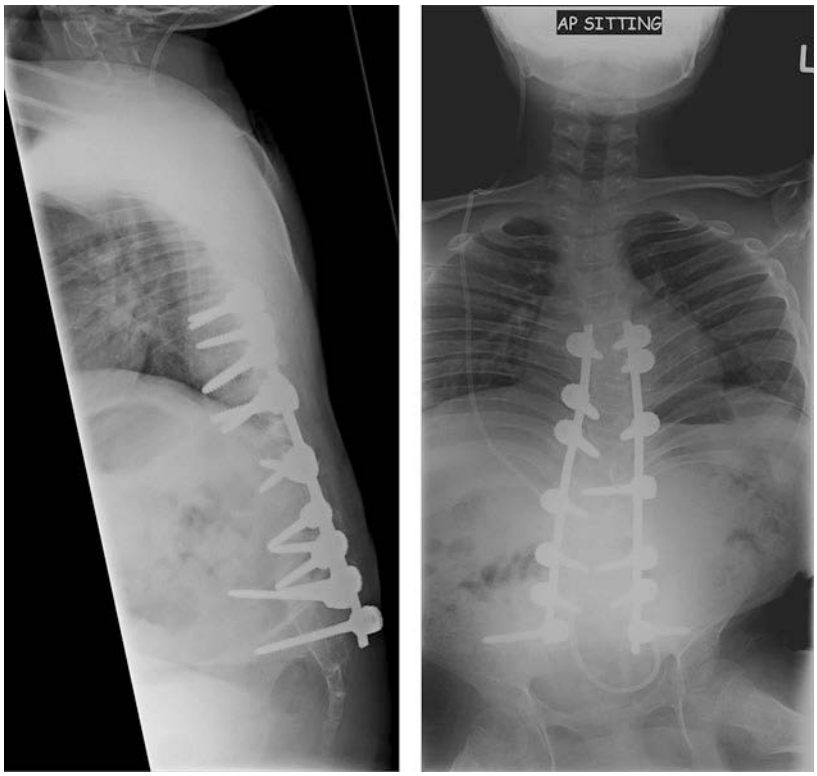

Fig. 2. Postoperative x-ray confirming pedicle screw–based construct correction.

Materials and Methods

Following institutional ethical approval, a retrospective review of clinical notes and radiology was performed on myelomeningocele patients undergoing kyphectomy surgery. They were identified from the senior author's prospectively maintained surgical database.

All surgery and follow-up was performed at a dedicated paediatric hospital by the senior author.

During a 10-year period from September 2004 to 2014, a total of seven patients underwent kyphectomy surgery. There were four boys and three girls, with an average age of 9.5 (8–13) years.

Three of the children had open pressure ulcers over the kyphotic deformity at the time of surgery. These failed to heal despite pressure care and dressings. The surgery was performed with the open sore present.

All patients were wheel chair bound, with no neurologic function distal to their gibbus and a thoracic sensory level. They had a history of relentless progressive kyphotic deformity.

Demographic data, surgical parameters, intra- and postoperative complications, and radiographic deformity correction was collated.

All patients had urine cultures a week preoperatively and treated prophylactically with oral antibiotics.

Surgical Technique

Following prone positioning and prophylactic first-generation cephalosporin antibiotic administration, the spine was exposed through a posterior midline incision. In the three patients with persistently open pressure sores over the gibbus, an elliptical incision was made to excise the sore back to clean, bleeding edges.

The spine was exposed taking care to avoid durotomy where the theca or meningocele was exposed by the deficient posterior elements. Generally, there was no thecal sac present at the apex of the gibbus. This may well be due to the underlying pathology, that is, the meningocele that was closed at birth, leaving predominately scar tissue. Alternatively, especially in the patients with ulceration over the gibbus, the chronic abrasion of the seat back may well have caused this scarring.

During surgical exposure, the distal end of the theca was identified by cephalad exploration from the apex of the gibbus. If it was identified distal to the planned proximal osteotomy site, it was mobilised in a cephalad direction to expose the vertebral bodies' posterior wall as required. Generally, this entailed a 1- to 2-cm mobilisation. If there was any concern regarding the integrity of the distal thecal sac as regards cerebrospinal fluid (CSF) leakage, the end was oversewed with a 4/0 Prolene suture.

Any epidural bleeding was controlled with bipolar coagulation and application of bone wax into the bony sinuses.

The soft tissue of the kyphotic segment was subperiosteally dissected off with cautery and bluntly with a cottonoid "peanut" to circumferentially expose the spine and push the aorta and abdominal contents anteriorly (Figs. 4 and 5).

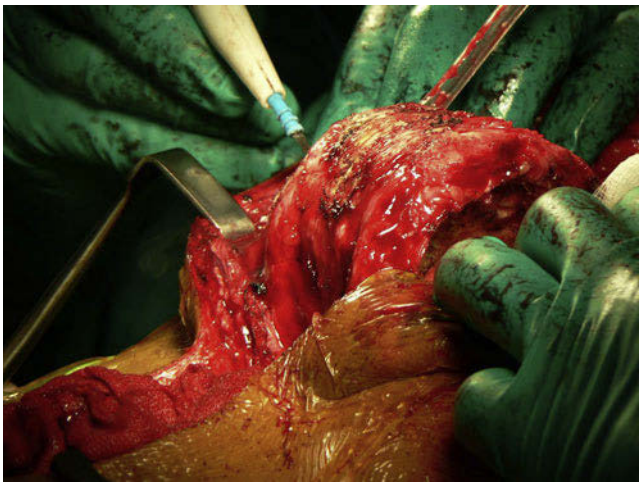

Fig. 4. Intraoperative exposure of the gibbus demonstrating absence of posterior elements and deficient thecal sac.

The posterior iliac crest was exposed to facilitate pelvic fixation. The lateral iliac blade was exposed subperiosteally to visualize the greater sciatic notch, as associated pelvic rotation made screw direction difficult to predict without this anatomic reference. Pedicle screws were placed from the posterior most inferior part of the iliac blade parallel to the sciatic notch after preparation with an awl and sounding with a pedicle probe to confirm circumferential bone.

Pedicle screws were placed in the distal spine. The number of fixation points was determined by residual distal bone. Typically, in addition to the pelvic screw, an S1 pedicle was placed and a more proximal body screw as there was unlikely to be a pedicle higher up. Thus, six fixation points were used distally. The coronally angled body screws facilitated subsequent deformity correction and pelvic rotation in the sagittal plane.

Proximally, the number of fixation points was determined subjectively by the perceived screw bone interface strength. Typically, the surgeon would place pedicle screws and

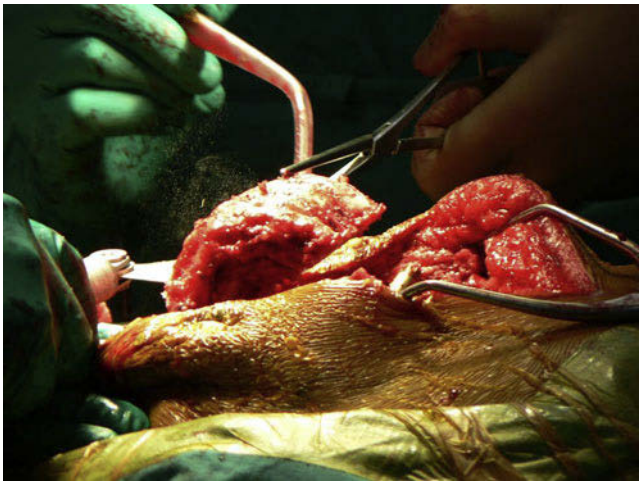

Fig. 5. Intraoperative osteotomy of the gibbus angling cuts to allow contact on correction of deformity.

manually check the integrity with a manual ''pull out test.'' By lifting the patient partially up off the bed with the pedicle screw driver, the value of the screw purchase was determined. Generally, a minimum of three levels of bilateral screw insertion would be considered adequate proximal fixation. The longest possible screws were utilized. The USS 1 titanium (Synthes, West Chester, PA) was used for most cases. As the shortest screw was 30 mm, this often required trimming with a bolt cutter. With the USS 2, 25-mm screws are now available, obviating this step. Again, adjacent to the gibbus, the pedicles were frequently absent and the use of coronally directed body screws employed.

In the earlier part of the series, instrumentation use was restricted by cost and sublaminar wires were employed in the thoracic spine. This necessitated a longer construct.

Because of the absent posterior elements near the apex of the deformity, body screws were used. This involved placing the screws posterolaterally, or even laterally, directly into the bodies proximal and distal to the planned osteotomy (Fig. 6).

The apex was then resected, either through the discs or through the bodies. If through the discs, the residual disc material was removed to expose endplates to promote fusion. The number of vertebral bodies resected depended on the degree of deformity and mobility of the extremes of the curve. It was largely subjectively determined by judging how the ends would engage on reduction without taking too much to inadvertently unnecessarily shorten the torso but excising enough to remove the prominence. If unsure, cuts would be made conservatively and the pelvis anteverted with downward pressure on the proximal spine, to reduce the cut ends and simulate correction before rod application. If it was deemed too long, that is, the force of reduction placed the instrumentation at risk of pull or cut out, a further osteotomy was performed to shorten one end or both until the anterior aspects of the vertebral column could be engaged and the pelvis rotated to close the gap posteriorly and allow rod application safely.

The osteotomies were angled in a manner to facilitate subsequent bony contact when corrected. This was either

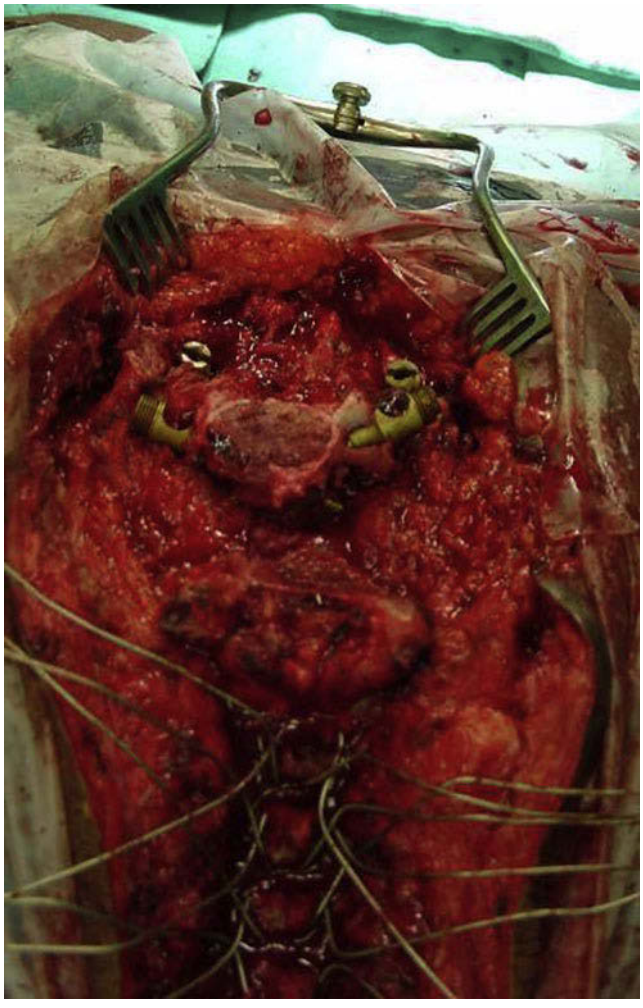

Fig. 6. Intraoperative view of defect following osteotomy with vertebral body screws and sublaminar wires placed ready for correction.

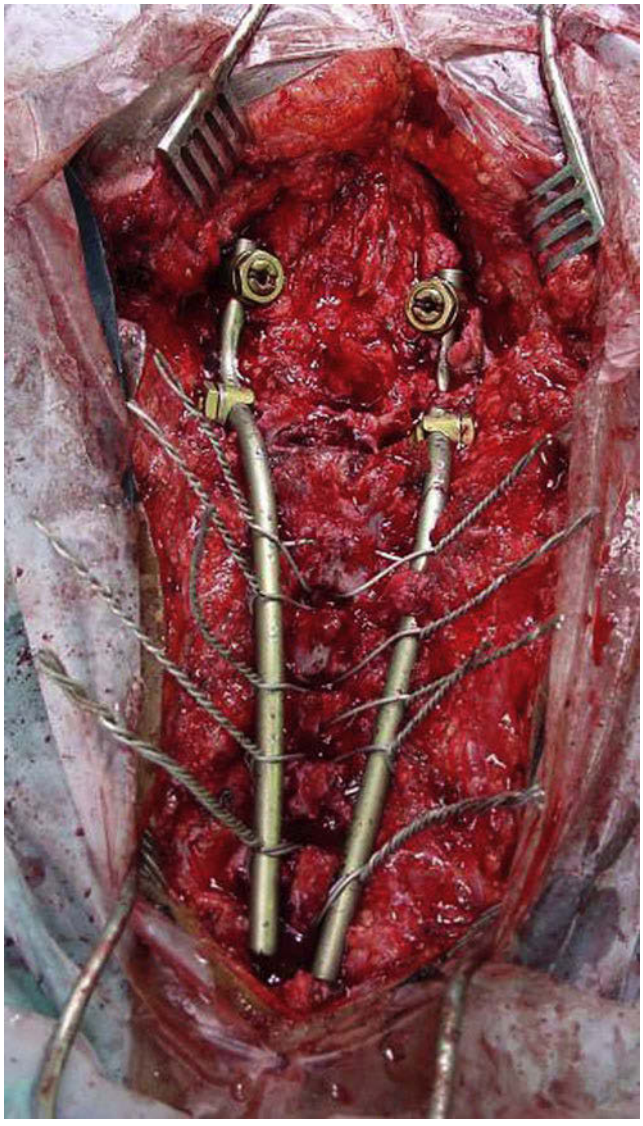

Fig. 7. Intraoperative view of reduced deformity.

perpendicular to the axis of the vertebral body selected or even sloped to facilitate slight lordosis on reduction. To achieve this, the distal cut would be in an anterior cephalad to posterior caudal slope and the proximal cut anterior caudal to posterior cephalad direction.

The rods were applied and a cantilever correction force applied to correct the kyphotic deformity and rotate the pelvis back to a more normal position to allow subsequent ischial seating. Bone grafting was performed utilizing morselized bone from the resected apex (Figs. 6 and 7).

Soft tissue closure was performed in layers. Despite the three elliptical incisions to excise the pressure ulcers, soft tissue closure was possible as a result of the spine shortening, flattening of gibbus, and anteversion of the pelvis. There was often difficulty closing the deep muscular layer as this was frequently deficient from the underlying meningocele. This was tacked as best as possible. The skin and subcutaneous tissue was abundant because of the realignment of the spine and relative shorting of the posterior length, as there was no longer the gibbus to be draped over. A ¼-inch drain was placed deep to the muscle layer. Redundant skin from the previous gibbus site was trimmed to facilitate closure. Plastic flaps were never needed.

Initially the children were nursed supine for 6 hours, as is the author's routine, to tamponade bleeding. They were then regularly turned with pressure care because of their insensate lower limbs.

After initial High Care Unit admission, they were mobilized to their wheel chairs in the ward without a brace.

The children were then followed up at 6 weeks and 3, 6, and 12 months postoperatively and then annually where the wound was inspected and x-rays checked for fusion.

Results

The median operative time (skin to skin) was 245 (165–285 ± 40.8) minutes. The median estimated blood loss was 700 (500–2,550 ± 722.5) mL (Table 1). A cellsaver was used and blood transfused in those with the larger losses. The larger losses were in the patients with the longer fusions, that is, proximal extent reaching T2/3.

The median preoperative kyphotic angle was 142 degrees (90–180 degrees ± 30.9). The postoperative median angle was 15 degrees (5–45 degrees ± 14.6). This represents a 92% (66% to 94% ± 10.2%) median correction (Table 2).

All patients were corrected to a functional sitting position as observed by the surgeon and reported by the caregiver.

There were no inpatient complications and all wounds healed primarily.

Subsequently, at outpatient follow-up, two of the patients with preoperative pressure sores over the apex presented with recurrent peri-implant sepsis. They were not systemically ill but had percutaneous pus drainage. One had skin breakdown over the iliac crest instrumentation with the screw head visible. In both cases, this occurred at 13 months postoperatively. The osteotomy was confirmed to be fused radiographically and the spinal instrumentation removed. Antibiotics were administered for 6 weeks with resolution of the infections.

There was no loss of spine correction in the follow-up period.

Because of resource restriction, sublaminar wiring was employed in the thoracic area to reduce cost. The latter cases were screw-only constructs. There were no implant failures in this cohort. There was no x-ray evidence of nonunion.

Table 1
Intraoperative data involving the kyphotic deformity correction.

Patient	Time (min)	Blood loss (mL)	Cell saver infusion (mL)	Levels fused	Cordotomy	Instrumentation	Preoperative pressure ulcer
1	255	1,000	440	T2–pelvis	Yes	Pedicle screws	
2	245	800	None	T7–pelvis	Yes	Screws and sublaminar wires	
3	165	700	None	T6–pelvis	No	Pedicle screws	
4	285	2,550	1,100	T3–pelvis	No	Screws and sublaminar wires	Yes
5	240	550	None	T4–pelvis	Yes	Screws and sublaminar wires	Yes
6	270	600	None	T5–Pelvis	No	Screws and sublaminar wires	Yes
7	205	500	None	T6–pelvis	No	Screws and sublaminar wires	

Table 2
Demographics and kyphotic deformity measurements.

Patient	Age	Sex	Preoperative kyphotic angle	Postoperative kyphotic angle	Correction	Correction percentage
1	13	Male	132	45	87	65.9
2	8	Female	90	5	85	94.4
3	10	Male	142	10	132	92.9
4	10	Male	145	8	137	93.1
5	10	Male	180	15	165	91.7
6	9	Female	180	32	148	82.2
7	9	Female	135	15	121	88.9

Discussion

Myelomeningocele patients are prone to spinal deformity. Although typical neuromuscular deformities may occur, they are at risk of a congenital kyphotic deformity that is present and progressive since birth. This focal kyphosis may progress at 6–12 degrees per year because of the anatomical nature of the pathology [2,6,9,10,14-16].

With the absent posterior spinal elements, the anteriorly and laterally displaced erector spinae and quadratus lumborum muscles act as perverted flexors, driving flexion instead of extension. In addition, the psoas muscle and diaphragmatic crura act as unopposed flexors. The ventral pull by the psoas muscles on the transverse processes leads to ventrolateral migration of the thoracolumbar fascia.

As the kyphosis progresses, the pelvis anteverts, resulting in a sacral sit. There is approximation of the costal margin and pubis, with reduced abdominal space when seated and reduced diaphragmatic excursion. With the insensate lower body, there is increased risk of sacral ulcers and abrasion of the attenuated skin over the gibbus itself. The patient is often forced to employ their upper limbs to support their sitting position. They may experience early satiety as a result of the compressed abdominal compartment.

These drivers render conservative treatment of the kyphotic deformity in myelomeningocele a poor choice. Braces are poorly tolerated, ineffective, and may cause decubitus ulcers [6,7,10,11,14-20].

Surgery is thus indicated in this group of severe kyphotic deformity; however, it is reported to have a high morbidity [6,10].

Sharrard described the first vertebral body resection procedure in 1968 [21]. Since then, numerous variations have evolved, including anterior fusions, posterior fusions, combination fusions, the use of Luque and Harrington rods, screws, plates, sublaminar hooks, cables, and pedicle screws. There is no current consensus in literature on which method is the most superior and depends on the surgeon's preference [10,15,19,22-25].

In our series, two cases were instrumented with pedicle screws throughout whereas the rest had a combination of pedicle screws and sublaminar wires to reduce cost. Sublaminar wires are at risk of failure because of the cartilaginous nature of the young patient's lamina and required increased length of instrumentation. From a technical perspective, we prefer the use of pedicle screws to facilitate correction.

Most surgeons agree that surgery can be performed from 5 to 12 years of age [12,19,24]. The optimal age is 8 years and above as long fusions can restrict growth and chest wall development [26]. Our cohort resembled this, with a mean age at surgery of 9.5 (8–13) years. We accept that the older the child, the better in terms of blood volume and bony architecture to facilitate instrumentation. In addition, fusion reduces torsal growth; however, we accept that 70% of torsal height is achieved by 5 years of age. In reality, one's hand is forced because of the severe progressive kyphosis and threatened or actual breakdown of soft tissue over the gibbus.

The extent of fixation is debatable, but most surgeons currently include the sacrum in the fixation. Inclusion of the pelvis allows improvement of the pelvic inclination, thus shifting the centre of gravity, ensuring greater stability, leading to better alignment and correction of the deformity [15,22,27] (Fig. 8). Frequently, their bone is soft and the iliac screws are required to ensure adequate distal purchase.

Many iliac fixation methods have been described but we employed a modified Galveston technique with pedicle screws placed in the iliac just above the greater sciatica notch. By placing them as distal as possible, rod connection from the S1 screw to the iliac screws is possible [6,26].

Reported postoperative complications include death, excessive bleeding, wound dehiscence, infection, CSF leak, pseudoarthrosis, implant failure, recurrent decubitus ulcers, and loss of deformity correction [2,3,6,9,11,16,17,19,23,26]. The shortening of the column allows good soft tissue coverage even when an ulcer has been excised. We were able to obtain muscle cover and not only skin.

We experienced no acute wound problems. This is surprising, as three had open ulcers at the time of surgery. This may well be explained by the protocol of preoperative urine culture and administration of a week's preoperative oral antibiotics. It is the author's experience that this patient group all have bacteriuria, if not low-grade urinary infection, as a result of their paraplegia and lack of bladder sensation and control. Typically, the culture would yield a mixed growth and oral amoxicillin–clavulanic acid would be prescribed for the week preoperation.

In speculating on possible reasons for the relatively low complication rate, it may be the environment we find ourselves in. These patients are largely from rural impoverished areas with poor access to health care. This paradoxically may positively impact the outcomes by naturally selecting better hosts. In addition, they avoid the frequent admissions often seen in the sophisticated world with the danger of MRSA colonisation and the like.

Fig. 8. Clinical view of postcorrection alignment where not only is the gibbus area not flat but the pelvis has been rotated to restore the buttocks to improve seating.

Delayed wound infection occurred in two of the three cases operated on with the preoperative open wounds. The instrumentation was removed without loss of correction with subsequent control of the infection. There is always the concern of loss of correction following implant removal. In these two cases, the osteotomy sites were confirmed to be fused preremoval on x-ray and subsequently intraoperatively. The rest of the instrumented area likewise had fused and we did not observe loss of correction on follow-up.

Intraoperatively, we made an effort to identify the residual thecal sac and oversew the end to ensure there was no CSF leak. We did not see any CSF-related complication.

All our patients maintained their deformity correction, even the two who underwent implant removal. Despite a 10% to 15% reported incidence, we did not identify any pseudo-arthrosis at the osteotomy site [3,9,17].

Conclusion

Although an infrequent problem today, severe kyphosis in myelomeningocele patients causes not only a major functional impairment but threat to their life with apical pressure sores.

Kyphectomy and posterior instrumented spinal fusion can be performed safely, even in the face of an open sore with excellent kyphotic correction and resultant improved functionality and ability to sit.

These open sores can be closed primarily without the requirement of plastic surgery because of the shortening and extension of the spine.

References

[1] Horn A, Dix-Peek S, Mears S, et al. The orthopaedic management of myelomeningocele. *S Afr Med J* 2014;104:314–7.
[2] Ko AL, Song K, Ellenbogen RG, et al. Retrospective review of multilevel spinal fusion combined with spinal cord transection for treatment of kyphoscoliosis in pediatric myelomeningocele patients. *Spine* 2007;32:2493–501.
[3] Hwang SW, Thomas JG, Blumberg TJ, et al. Kyphectomy in patients with myelomeningocele treated with pedicle screw–only constructs: case reports and review. *J Neurosurg Pediatr* 2011;8:63–70.
[4] Yoshioka K, Watanabe K, Toyama Y, et al. Kyphectomy for severe kyphosis with pyogenic spondylitis associated with myelomeningocele. *Scoliosis* 2011;6:5.
[5] Banta JV, Hamada JS. Natural history of the kyphotic deformity in myelomeningocele. *J Bone Joint Surg Am* 1976;58:279.
[6] Kocaoğlu B, Erol B, Akgülle H, et al. Combination of Luque instrumentation with polyaxial screws in the treatment of myelomeningocele kyphosis. *J Spinal Disord Tech* 2008;21:199–204.
[7] Comstock SA, Cook PC, Leahey JL, et al. Posterior kyphectomy for myelomeningocele with anterior placement of fixation. *Clin Orthop Relat Res* 2011;469:1265–71.
[8] Kadic MAC, Verbout AJ. Treatment of severe kyphosis in myelomeningocele by spinal instrumentation with Luque rods. *Acta Orthop Belg* 1991;57:45–51.
[9] Schroeder JE, Barzilay Y, Hasharoni A, et al. Long-term outcome of surgical correction of congenital kyphosis in patients with myelomeningocele with segmental spino-pelvic fixation. *Evid Based Spine Care J* 2011;2:17–22.
[10] Altiok H, Finlayson C, Hassani S, et al. Kyphectomy in children with myelomeningocele. *Clin Orthop Relat Res* 2011;469:1272–8.
[11] Smith JT, Novais E. Treatment of gibbus deformity associated with myelomeningocele in the young child with use of the vertical expandable prosthetic titanium rib (VEPTR). *J Bone Joint Surg Am* 2010;92:2211–5.
[12] Duddy JC, Caird J, Connolly P. Repair of a large thoracolumbar myelomeningocele with associated lumbar kyphosis. *Acta Neurochir* 2013;155:1965–8.
[13] Fürderer S, Eysel P, Hopf C, et al. Sagittal static imbalance in myelomeningocele patients: improvement in sitting ability by partial and total gibbus resection. *Eur Spine J* 1999;8:451–7.
[14] Carstens C, Koch H, Brocai DR, et al. Development of pathological lumbar kyphosis in myelomeningocele. *J Bone Joint Surg Br* 1996;78:945–50.
[15] Nolden MT, Sarwark JF, Vora A, et al. A kyphectomy technique with reduced perioperative morbidity for myelomeningocele kyphosis. *Spine* 2002;27:1807–13.
[16] Lintner SA, Lindseth RE. Kyphotic deformity in patients who have a myelomeningocele. Operative treatment and long-term follow-up. *J Bone Joint Surg Am* 1994;76:1301–7.
[17] Odent T, Arlet V, Ouellet J, et al. Kyphectomy in myelomeningocele with a modified Dunn-McCarthy technique followed by an anterior inlayed strut graft. *Eur Spine J* 2004;13:206–12.
[18] Lowe GP, Menelaus MB. The surgical management of kyphosis in older children with myelomeningocele. *J Bone Joint Surg Br* 1978;60:40–5.
[19] Garg S, Oetgen M, Rathjen K, et al. Kyphectomy improves sitting and skin problems in patients with myelomeningocele. *Clin Orthop Relat Res* 2011;469:1279–85.
[20] Kose KC, Inanmaz ME, Uslu M, et al. Kyphectomy for congenital kyphosis due to meningomyelocele: a case treated with a modified approach to skin healing. *Int Wound J* 2012;9:311–5.
[21] Sharrard WJ. Spinal osteotomy for congenital kyphosis in myelomeningocele. *J Bone Joint Surg Br* 1968;50:466–71.
[22] Niall DM, Dowling FE, Fogarty E, et al. Kyphectomy in children with myelomeningocele. A long-term outcome study. *J Pediatr Orthop* 2004;24:37–44.
[23] Doers T, Walker JL, van den Brink K, et al. The progression of untreated lumbar kyphosis and the compensatory thoracic lordosis in myelomeningocele. *Dev Med Child Neurol* 1997;39:326–30.
[24] Hoffman EB. The orthopaedic management of myelomeningocele. *S Afr Orthop J* 2011;10:48–55.
[25] de Amoreira Gepp R, Quiroga MRS, Gomes CR, et al. Kyphectomy in meningomyelocele children: surgical technique, risk analysis, and improvement of kyphosis. *Childs Nerv Syst* 2013;29:1137–41.
[26] Samagh SP, Cheng I, Elzik M, et al. Kyphectomy in the treatment of patients with myelomeningocele. *Spine J* 2011;11:e5–11.
[27] Saraste H, Stark A. Vertebral resection and fusion for paralytic kyphosis: 9 patients followed up for 6 (2-14) years. *Acta Orthop Scand* 1991;62:515–6.