ELSEVIER
SAUNDERS

Neurosurg Clin N Am 18 (2007) 357–364

NEUROSURGERY
CLINICS
OF NORTH AMERICA

Kyphectomy for Myelodysplasia

Lawrence I. Karlin, MD[a,b,*]

[a]*Harvard Medical School, 180 Longwood Avenue, Boston, MA 02115, USA*
[b]*Children's Hospital Boston, 300 Longwood Avenue, Boston, MA 02115, USA*

Kyphotic deformity of variable severity at the lumbar or thoracolumbar area occurs in up to 15% of children with a myelomeningocele [1–4]. Those involved usually have thoracic level paraplegia. In these children, the combination of complex pathologic spinal anatomy, associated deformities, and neurologic abnormalities creates unique and challenging clinical problems. This article will discuss considerations and techniques in the management of patients with kyphotic deformity.

The pathologic anatomy of the spine in myelodysplasia was described by Hoppenfield [3]. The posterior elements are splayed open, the pedicles are oriented in the coronal plane, and the dysplastic laminae are everted with their anterior surfaces directed posterolaterally (Fig. 1). Kyphotic deformity occurs at the thoracolumbar and lumbar spine as a result of the imbalance of flexor and extensor muscle activity across the spine. The paraspinal muscles are anterior to the apex of the deformity and act as flexors instead of extensors of the spine. The psoas and diaphragmatic crura are unopposed [5]. The aorta is bowstrung across the deformity [6]. A compensatory thoracic lordosis develops above the kyphotic deformity.

There are problematic consequences of this altered anatomy [7–9]. The bony kyphotic deformity is prominent and covered with thin scarred skin; pressure sores are frequent and often refractory to treatment. Progressive difficulty with breathing may occur as the abdominal contents, crowded by the shortened lumbar spine, elevate the diaphragm. The lordotic proximal portion of the deformity further compromises the size of the chest cavity (Fig. 2). The crowded abdominal contents also make eating difficult, so nutritional status may suffer. Without functional spinal extensors, the child cannot maintain a sitting posture without the support of the upper extremities. Using the upper extremities for postural support compromises the ability of the patient to use his or her arms and hands for activities of daily living and wheelchair mobility. There are technical problems with urinary drainage procedures, because kyphotic deformity compromises space for ilial conduits and catheterization procedures. Kyphotic deformity results in significant compromise of function and affects multiple systems in the patient.

In essentially all cases of patients with myelodysplasia and kyphotic deformity, the deformity progresses 8° to 12° per year [8,10]. Orthotic support may help to maintain the upright posture, but the braces are cumbersome, often cause skin breakdown, and do not prevent progression. The only treatment that prevents worsening of the deformity is surgical stabilization. Surgical correction of spinal deformity in myelodysplasia is difficult because of the deficient posterior elements that provide inadequate mass for bony fusion and the absence of anchorage for spinal instrumentation.

The surgical indications were defined by Eckstein and Vora [11] in 1972. Indications include the need to obtain primary skin closure over the protuberant bone, recurrent ulceration over the gibbus, difficulty with placement in a stable upright sitting or standing posture, reduction of the anterior abdominal surface area precluding

* Children's Hospital Boston, 300 Longwood Avenue, Boston, MA 02115.
E-mail address: lawrence.karlin@childrens.harvard.edu

1042-3680/07/$ - see front matter
doi:10.1016/j.nec.2007.02.005

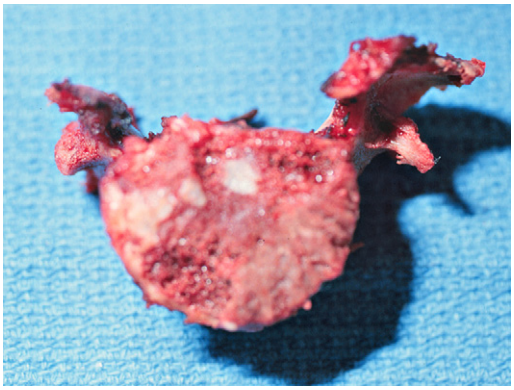

Fig. 1. Lumbar vertebral body resected during a kyphectomy procedure. Note the dysplastic posterior elements. A pedicle screw placed in this vertebral body would have an extremely oblique orientation.

urinary diversion procedures, compression of the abdominal cavity resulting in elevation of the diaphragm and respiratory compromise, and pain attributable to costal margin impingement on the iliac crests. An additional indication is documented or predicted neurologic deterioration in those with less angular deformities and significant or progressive deformity [8]. Because virtually all kyphotic deformities increase, this last indication assumes that all involved children have eventual problems. It is important to recognize that there is significant variability in clinical presentations of kyphotic deformity [12].

Preoperative evaluation

Kyphosis in patients with a myelomeningoceles is a multisystem problem, and the preoperative evaluation is important to the success of surgical intervention and may require extensive workup involving multiple disciplines. A complete neurologic evaluation must be performed. Some children may have neural function below the level of the deformity, and the consequence of spinal cord or nerve root resection should be determined. Lesions, such has syringomyelia, above the area of surgery can be detected by MRI. Shunt function must be established as cord transaction, or manipulation may alter the cerebrospinal fluid dynamics, causing catastrophic complications [13]. Urologic evaluation should include urodynamics to determine the status of bladder function and urine cultures to minimize urosepsis [9]. The skin over the defect needs assessment to determine potential difficulties in primary wound closure; tissue expanders may be needed. Latex precautions are necessary. The orthopedic assessment should include evaluation of hip contractures. The patient must have sufficient flexion so that the extension of the pelvis produced when the sacrum is brought into a more physiologic position does not compromise the ability to sit. The aberrant dysraphic bony elements pose problems for in strumentation anchorage. A preoperative CT scan helps to plan the appropriate fixation and orientation or bony anchors. Nutritional status

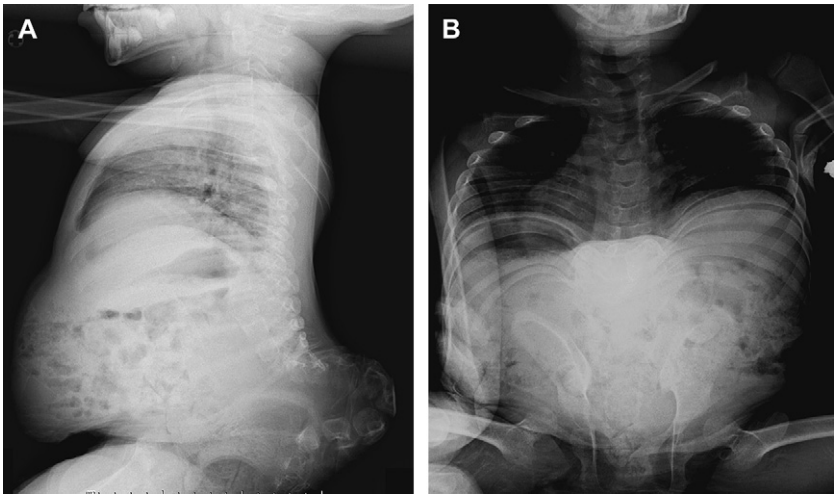

Fig. 2. Typical "S"-shaped deformity. On the lateral view, the lordotic nature of the superior limb of the kyphotic deformity is noted. The trunk is shortened, and the abdominal contents extend into the chest. The lung volume is additionally diminished by the compensatory thoracic lordosis.

is assessed to minimize chances of postoperative infection or delayed wound healing. The child and family must be counseled on the consequences of the procedure and possible complications. A surgically straightened spine is a stiffer one, and a more rigid spine may make self-catheterization, transfers, and ambulation more difficult. These considerations are especially important in patients with marginal independence at baseline.

Surgical principles

Although surgical treatment is the most direct and effective method to address the problems of myelomeningocele kyphosis, there remains debate about the proper timing for surgery, the method of kyphosis reduction, the extent of instrumentation, and the role of instrumentation with limited arthrodesis. In general, earlier surgery on less severe deformities is arguably safer and may allow inclusion of fewer motion segments and permit some spinal growth. A high recurrence rate has been reported when a limited spinal fusion is performed [4,7,11,12,14–16]. Definitive surgery in older children does not have an adverse affect on spinal growth; when performed with modern spinal instrumentation, it can yield dramatic and stable corrections. In the older age group, however, larger and more rigid deformities require more extensive and, consequently, more dangerous procedures [17–24]. A reasonable compromise may be early surgery with contemporary segmental instrumentation performed using a technique that preserves growth potential [25,26]. In any scenario, surgery is fraught with high complication rates that must be weighed against the anticipated benefits.

It is helpful to consider first the method of bone resection performed at the site of the deformity and then the forms of spinal instrumentation used to obtain and maintain that correction. There are basically two methods for addressing the site of the deformity: vertebrectomy and vertebral body decancellation. Vertebrectomy was the original surgical treatment. It was first described in 1968 by Sharrard [27], who advocated correction in the newborn by resection of 1.5 vertebrae; he and others noted a high complication rate with loss of correction over time [4,7,11,12,14–16]. Since then, major advances have been made in our understanding and surgical treatment of the deformity. Lindseth and colleagues [28] refined the surgical technique and defined the indications for its use. They classified the kyphosis into a collapsing "C"-shaped deformity and a more rigid "S"-shaped deformity (Fig. 3). In the "S"-shaped deformity, vertebral resection is usually required. In the less rigid "C"-shaped type, Lindseth [29] has suggested treatment by decancellation, the removal of the cancellous vertebral bone, of the bodies above and below the apex vertebra.

"C" type deformity

Although vertebral resection techniques can be used for all forms of myelomeningocele kyphosis, the less severe "C" type of the deformity would seem ideal for the decancellation method, as advocated by Lindseth [29]. Compared with excisional kyphectomy, the neural elements are much less at risk with this technique, making it preferential in the rare patient with intact neurologic function distal to the deformity. Additionally, the anterior column is not lengthened, so vascular structures are not put into jeopardy, and the technique is less extensive and may be associated with less blood loss. Theoretically, this method spares the growth centers of the vertebral bodies, allowing continued growth.

Age-Specific considerations for surgical correction of "C" type deformities

Newborn

Surgical correction is indicted in the newborn if the magnitude of the deformity precludes skin closure or if the philosophy of the treating surgeon is for earliest correction. The procedure is done at the time of surgical sac closure. The vertebra above the apical vertebra is exposed, and the posterior elements are removed to the level of the posterior vertebral cortex. Working from both sides by way of the pedicle base, the vertebral body cancellous bone is removed. Portions of the posterior and lateral cortices are then removed while preserving the anterior cortex. Care is used to avoid injury to the cartilaginous end plates. The same procedure is performed on the vertebra below the apex. Heavy nonabsorbable sutures or wires are placed around the pedicles of the apical vertebra and those of the bodies adjacent to the resected vertebra, the kyphosis is manually corrected compressing the posterior vertebral bodies, and the sutures are tightened (Fig. 4). An important component of the closure is repair of the paraspinal musculature behind the dural sac. Recurrence of deformity is a given with this early

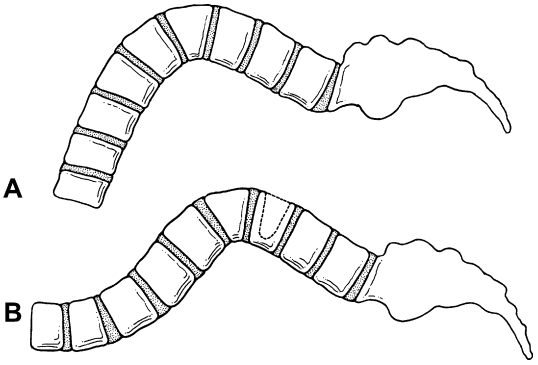

Fig. 3. The "C" deformity (*A*) and the "S" deformity (*B*). (*From* Lindseth RE. Myelomeningocele spine. In: Weinstein SL, editor. The pediatric spine: principles and practice. New York: Raven; 1994. p. 1058; with permission.)

surgery, but proponents believe that it is well tolerated and that further surgery, if required, is less difficult [15].

Older child (3+ years old)

This author agrees with others [30] that correction is best left to children older than 3 years of age. At the level of the meningocele, the exposure and vertebral decancellation are performed as in the neonate. In this age group, the reduction is obtained by instrumentation that must be anchored proximally and distally. After manual cantilever correction, the sublaminar wires placed proximally are tightened sequentially. Three or four anchors are needed for each rod. When performed in an older child, more rigid constructs may be used, along with fusion proximally and distally; in the younger child, fusion is not performed.

"S" type deformity

These deformities are rigid and severe, and they consist of the angular kyphosis and a rigid lordotic cephalad limb. Successful reduction of the deformity requires vertebral resection. The technique was refined by Lindseth and Stelzer [4], who demonstrated improved results when the lordotic cephalad portion, along with a portion of the apex vertebrae, was resected [15].

Age-specific considerations for surgical correction of "S" type deformities

Newborn

In general, this extensive procedure is not recommended in the newborn, but it may be necessary when the severity of the deformity precludes skin closure. It is accomplished at the time of neurosurgical sac closure. Although

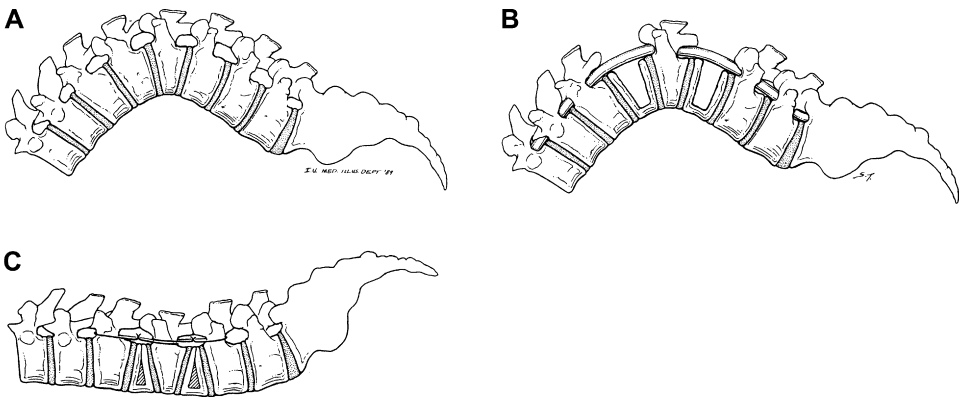

Fig. 4. Decancellation of one or more levels at the apex of the deformity permits significant correction in a "C"-shaped deformity. (*From* Lindseth RE. Spine deformity in myelomeningocele. Instr Course Lect 1991;40:276; with permission.)

significant complications have been reported in older series, Crawford and colleagues [31], using modern techniques, have reported excellent results with limited complications. Recurrent deformity does occur but seems to be well tolerated and can be addressed by less demanding procedures than those required in the older child.

Older child

The spine is approached through a posterior midline incision, and the kyphotic deformity is exposed circumferentially (Fig. 5). The everted posterior elements of the apical and two proximal vertebrae are removed. Lindseth [28] recommends freeing the dural sac from the bony elements, ligating the segmental nerves and vessels, dividing the neural tube one segment distal to the apical vertebra, and oversowing the distal cord. When the neural tube is divided, the repair of the proximal limb must be meticulous, because hydrocephalus has been described when the proximal limb closure occludes the central canal [13]. This author has found it possible to preserve the dura as additional soft tissue coverage. The vertebra above the apical vertebra is now resected with the disc on either side. It is then usually necessary to resect the next cephalad vertebra and disc to correct the deformity (Fig. 6). The end plate is removed from the proximal remaining vertebral body, and a trough is made in the body. With its end plate removed, the apical vertebra is now shaped into a wedge and, after manual deformity correction, placed in the groove of the proximal vertebra. The correction can be held by wires placed around the pedicles of the proximal and apical vertebrae. Segmental instrumentation provides excellent fixation and minimized recurrence. Closure should involve repair of the paraspinal muscles over the sac, which may require relaxing incisions. Additionally, when the procedure is performed in the older child, a definitive fusion may be performed at this time (see Fig. 4A, B). This is an extensive procedure with significant blood loss. Nolden and colleagues [26] have reported on the early results with a decancellaton technique in rigid deformity. Correction of the deformity required decancellation of multiple vertebral bodies, and the reduction was maintained by contoured rods. The procedure is less extensive than a vertebrectomy and theoretically allows continued growth.

Fig. 5. In a vertebral resection kyphectomy, the deformity is exposed circumferentially through a posterior approach. (*From* Lindseth RE. Myelomingocele. In: Lovell, Winter, editors. Pediatric Orthopaedics. Philadelphia: Lippincott Williams & Wilkins; 2001. p. 617; with permission.)

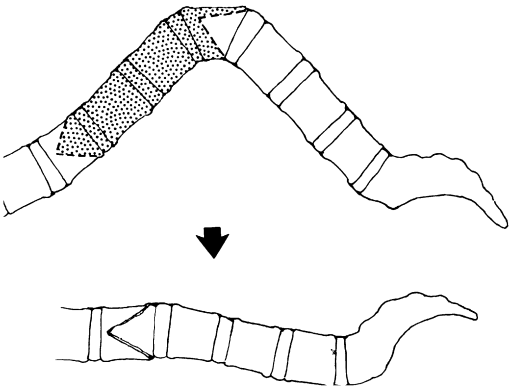

Fig. 6. An apical resection of two or more segments may be required for more severe deformity. The apical resection permits significant shortening of the spinal column and is useful in patients with skin compromise.

Spinal instrumentation

As initially described, the correction obtained at the time of vertebrectomy was stabilized by wires or sutures placed around the dysplastic posterior elements. Although this fixation may be all that can be placed in the extremely small child, it produces minimal corrective force and does not control the adjacent spinal segments, so that the deformity recurs at the junction of the fused and mobile spine. Procedures in children with myelomeningocele spinal deformities using longer constructs, such as Harrington rods,

attempted to control the entire length of the deformity but were fraught with difficulties, including the inability to control the spine segmentally [32]. Current spinal instrumentation techniques have addressed the problem of recurrent deformity that plagued earlier surgical techniques. After deformity reduction, dramatic stresses are placed on instrumentation anchorage points osteoporotic and dysplastic bone. Those forces are reduced when distributed over multiple sites by segmental spinal instrumentation (SSI), a technique that uses physiologically contoured vertically oriented rods connected to the spine at multiple sites by various forms of instrumentation. Heydemann and Gillespie [17], using Luque instrumentation and pelvic fixation, first reported the use of this technique for myelomeningocele kyphosis. They and others noted a dramatic im provement in the amount of correction and maintenance of correction with this method [17,21,24]. Increased fixation strength is now available with a wide selection of hooks and screws for the intact portion of the spine and pedicle screws for the dysraphic portion (Fig. 7).

The best distal bony anchor is the pelvis and sacrum, but fixation to these structures poses unique problems in the kyphotic myelodysplastic spine. The Galveston technique is commonly used for pelvic fixation, and the low profile makes it ideal for patients with poor soft issue coverage. Unfortunately, the stress placed on this form of fixation by the kyphosis may produce dorsal displacement of the implants. The Dunn-McCarthy technique addresses this problem by placing the end of the rod anterior to the sacrum. A stronger construct, devised by Warner and Fackler [24], places the rod anterior to the sacrum by way of the S1 foramina (Fig. 8). The rods are cross-linked to produce rotational stability (Fig. 9).

Generally, spinal instrumentation is intended to obtain deformity correction and maintain that correction while fusion occurs throughout the instrumented area. The dilemma facing the surgeon who must deal with a young patient who has a rigid severe deformity is that correction requires rigid instrumentation but fusion would cause stunting of the spinal growth. In this case, instrumentation without fusion can be helpful.

The spine is approached, whenever possible, in an extraperiosteal fashion. Fusion is performed at the level of the osteotomy only, and adjacent segments are left unfused, preserving potential for growth. Instrumentation anchors are appropriately chosen, and the physiologically contoured rods are applied. McCall [25] reported a series using cross-linked contoured spinal rods placed anterior to the sacrum and fixed to the thoracic spine with sublaminar wires. Fusion was performed at the resection site only. An average 111° of kyphosis was reduced to 15° and was

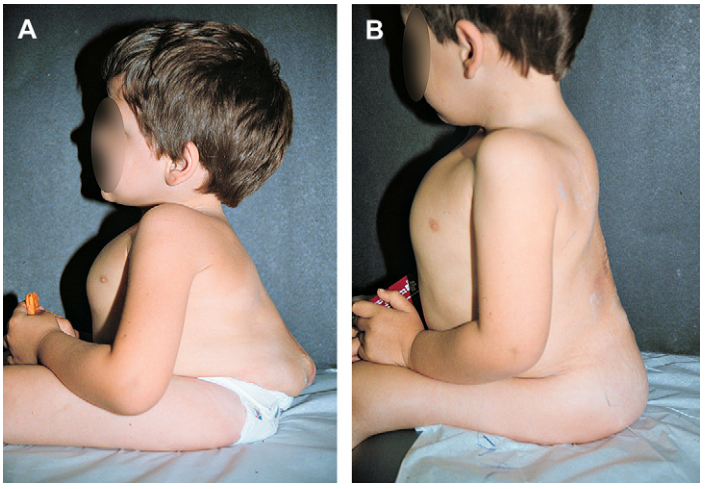

Fig. 7. (*A*) This child must use his upper extremity to maintain an upright sitting posture and has had recurrent skin breakdown over the gibbus. (*B*) After kyphectomy with segmental instrumentation, the gibbus is corrected and the trunk height is increased. (*From* Warner WC Jr, Fackler CD. Comparison of two instrumentation techniques in treatment of lumbar kyphosis in myelodysplasia. J Pediatr Orthop 1993;13:704–8; with permission.)

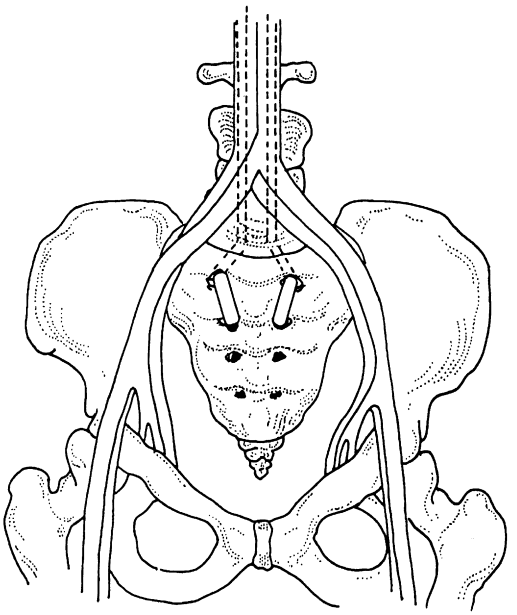

Fig. 8. The sacral rod fixation technique of Warner and Fackler [24] requires a transverse connector for rotational stability and spinal anchorage for vertical stability. Tremendous corrective forces may be applied with this method.

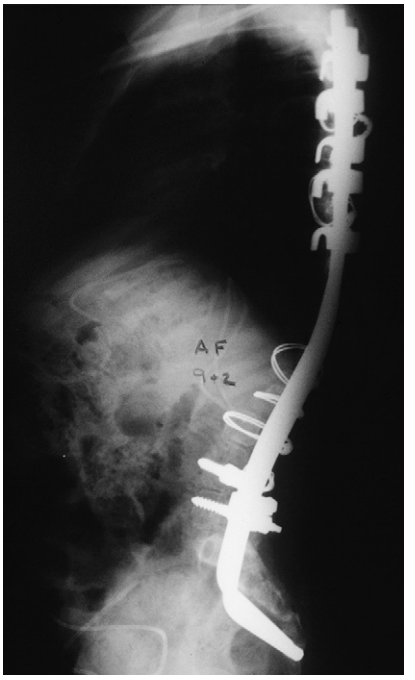

Fig. 9. Postoperative radiograph of the child in Fig. 2. The paired rods are contoured to what is believed to be the maximum correction. They pass through the S1 neuroforamen and then angle laterally so that they abut the anterior aspect of the sacrum away from the midline structures. Such positioning allows one to apply significant corrective forces. Further stability is provided by lumbar pedicle screws and a cross-connector. Fixation cephalad to the bifid spine is obtained by sublaminar wires and laminar hooks. The youngster was reasonably mature at the time of the surgery, and a growth-preserving technique was not used.

20° at last follow-up. The lumbar spine growth after the initial correction averaged 3.9 cm. The thoracic spinal growth was not measured but was noted by Luque wire migration.

It is not surprising that the complication rates for the surgical treatment of kyphosis in children with myelomeningocele are extremely high. These children have associated neurologic abnormalities, poor soft tissue coverage, urosepsis, osteopenia, and reactions (including anaphylaxis) associated with latex allergy. The operations involve rigid deformities in osteopenic bones with deficient dysplastic elements and require partial or complete vertebral resections and placement of instrumentation in close proximity to unprotected neural elements. Complications include high rates of infection, excessive hemorrhage [4,12,15], wound healing difficulties [4,17], pseudarthrosis [4,12,21], instrumentation failure or exposure [14,17], long bone fractures [12], recurrent deformities [4,7,11,12,14–16], neural injury, acute hydrocephalus [13], vascular injury, and death [11]. With proper preoperative evaluation, antibiotic coverage, and modern surgical techniques, some of these problems can be limited, but intraoperative and perioperative complications remain an important consideration for patients and physicians considering spinal reconstructive surgery for kyphosis in myelodysplasia.

Summary

Major advances in the surgical treatment of the deformity of myelomeningocele kyphosis have been made. Further experience should help to determine the best timing for the surgery. At present, excision kyphectomy, vertebral decancellation, segmental instrumentation, and instrumentation without fusion are all part of the menu of treatments allowing individualization of care. SSI has certainly demonstrated its superiority as a fixation method. The role of decancellation

procedures and instrumentation without fusion remains to be determined by further prospective investigation.

References

[1] Banta JV, Hamada JS. Natural history of the kyphotic deformity in myelomeningocele. J Bone Joint Surg Am 1976;58:279.

[2] Carstens C, Koch H, Brocai DRC, et al. Development of pathological lumbar kyphosis in myelomeningocele. J Bone Joint Surg Br 1996;78(6):945–50.

[3] Hoppenfield S. Congenital kyphosis in myelomeningocele. J Bone Joint Surg Br 1967;49:276–80.

[4] Lindseth RE, Stelzer L. Vertebral excision for kyphosis in children with myelomeningocele. J Bone Joint Surg Am 1979;61:699–704.

[5] Drennan JC. The role of muscles in the development of human lumbar kyphosis. Dev Med Child Neurol 1970;12(Suppl 22):33–8.

[6] Loder RT, Shapiro P, Towbin R, et al. Aortic anatomy in children with myelomeningocele and congenital lumbar kyphosis. J Pediatr Orthop 1991;11:31–5.

[7] Sharrard WJW, Drennan JC. Osteotomy-excision of the spine for lumbar kyphosis in older children with myelomeningocele. J Bone Joint Surg Br 1972;54: 50–60.

[8] Mintz H, Sarwark JF, Dias L, et al. The natural history of congenital kyphosis in myelomeningocele. A review of 51 children. Spine 1991;16(Suppl): 348–50.

[9] Lalonde F, Jarvis J. Congenital kyphosis in myelomeningocele: the effect of cordotomy on bladder function. J Bone Joint Surg Br 1999;81:245–9.

[10] Doers T, Walker JL, van der 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.

[11] Eckstein HG, Vora RM. Spinal osteotomy for severe kyphosis in children with myelomeningocele. J Bone Joint Surg Br 1972;54:328–33.

[12] Martin J, Kimar SJ, Guille JT, et al. Congenital kyphosis in myelomeningocele: results following operative and nonoperative treatment. J Pediatr Orthop 1994;14:323–8.

[13] Winston K, Hall J, Johnson D, et al. Acute elevation of intracranial pressure following transaction of non-functional spinal cord. Clin Orthop 1977;128: 41–4.

[14] Christofersen MR, Brooks AL. Excision and wire fixation of rigid myelomeningocele kyphosis. J Pediatr Orthop 1985;5:691–6.

[15] Lintner SA, Lindseth RE. Kyphotic deformity in patients who have a myelomeningocele. J Bone Joint Surg Am 1994;76:1301–7.

[16] Lowe GP, Menelaus MB. The surgical management of kyphosis in older children with myelomeningocele. J Bone Joint Surg Br 1978;60:40–5.

[17] Heydemann JS, Gillespie R. Management of myelomeningocele kyphosis in the older child by kyphectomy and segmental spinal instrumentation. Spine 1987;12:37–41.

[18] McCarthy RE, Dunn H, McCullough FL. Luque fixation to the sacral ala using the Dunn-McCarthy modification. Spine 1989;14:281–3.

[19] Niall DM, Dowling FE, Fogarty EE, et al. Kyphectomy in children with myelomeningocele: a long-term outcome study. J Pediatr Orthop 2004;24: 37–44.

[20] Odent T, Arlet V, Ouellet J, et al. Kyphectomy in myelomeningocele with modified Dunn-McCarthy technique followed by an anterior inlayed strut graft. Eur Spine J 2004;13:206–12.

[21] Rodgers BB, Williams MS, Schwend R, et al. Spinal deformity in myelomeningocele: correction with posterior pedicle screw instrumentation. Spine 1997;22:2235–43.

[22] Thomsen M, Lang RD, Carstens C. Results of kyphectomy with technique of Warner and Fackler in children with myelodysplasia. J Pediatr Orthop B 2000;9:143–7.

[23] Torode I, Godette G. Surgical correction of congenital kyphosis in myelomeningocele. J Pediatr Orthop 1995;15:202–5.

[24] Warner WC Jr, Fackler CD. Comparison of two instrumentation techniques in treatment of lumbar kyphosis in myelodysplasia. J Pediatr Orthop 1993;13: 704–8.

[25] McCall RE. Modified Luque instrumentation after myelomeningocele kyphectomy. Spine 1998; 23:1406–11.

[26] Nolden MT, Sarwark JF, Vora A, et al. A kyphectomy technique with reduced perioperative morbidity for myelomeningocele kyphosis. Spine 2002; 27(16):1807–13.

[27] Sharrard WJW. Spinal osteotomy for congenital kyphosis in myelomeningocele. J Bone Joint Surg Br 1968;50:446–71.

[28] Lindseth RE. Myelomeningocele spine. In: Weinstein SL, editor. The pediatric spine: principles and practice. New York: Raven; 1994. p. 1043–67.

[29] Lindseth RE. Spine deformity in myelomeningocele. Instr Course Lect 1991;40:273–86.

[30] Hall JE, Poitras B. The management of kyphosis in patients with myelomeningocele. Clin Orthop 1977; 128:33–40.

[31] Crawford AH, Strub WM, Lewis R, et al. Neonatal kyphectomy in the patient with myelomeningocele. Spine 2003;18(3):260–6.

[32] Geiger F, Parsch D, Carstens C. Complications of scoliosis surgery in children with myelomeningocele. Eur Spine J 1999;8:22–6.