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CASE REPORT

Spinal Cord Injury After Extremity Surgery in Children With Thoracic Kyphosis

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Abstract

Background Spinal cord injury is a rare complication after lower extremity surgery in children with skeletal dysplasia and thoracic kyphosis. We encountered two patients who had this complication, from among 51 (39 from Nemours/Alfred I. duPont Hospital for Children and 12 from Seattle Children's Hospital) who underwent lower extremity surgery during an 8.5-year period (June 2004 to December 2012). Because spinal cord injury is a devastating complication likely not known to most physicians treating patients with skeletal dysplasias, we sought to examine factors that may contribute to this rare complication.

Case Description We performed a retrospective review of two patients with skeletal dysplasia who had paraplegia develop after extremity surgery. Outcome measures included operative time, vital signs, and postsurgery recovery of neurologic deficit. MR images were reviewed. Two patients were found—an 8.5-year-old boy with spondyloepiphyseal dysplasia congenita with a 76°-thoracic kyphosis apex at T4 and a 6.5-year-old boy with mucopolysaccharidosis type 1-H with an 80°-thoracic kyphosis apex at T2. Bilateral proximal femoral osteotomies or bilateral innominate and proximal femoral osteotomies had been performed. The spinal cord injuries occurred at the apex of the kyphosis as determined by clinical examination and MRI assessment. In both patients, the mean arterial blood pressure decreased below 50 mm Hg and might be a factor in the etiology of the paralysis. The first patient recovered motor function in 5 months; the second had no recovery.

Literature Review Paraplegia is extremely rare after nonspine operations. Many factors contribute to the risk for a spinal cord event: low mean arterial pressure, duration of the surgery, position on the operating table, the kyphotic spine deformity, or unappreciated vascular disease. Motor-evoked potentials and somatosensory-evoked potentials together potentially provide high sensitivity and specificity for predicting a postoperative neurologic deficit.

Clinical Relevance Based on our two patients with skeletal dysplasia and a literature review of patients with hyperkyphosis undergoing extremity surgery, the surgeon must be aware of the risk of spinal cord injury. Careful preoperative assessment possibly including MRI of the spine is recommended. Mean arterial pressure should be maintained at a safe level; neuromonitoring should be considered.

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Introduction

Sagittal and coronal deformities of the spine are common in skeletal dysplasia, including spondyloepiphyseal dysplasia and mucopolysaccharidosis. These dysplasias are a nonhomogeneous group of growth disorders characterized by shortening of the trunk and extremities [13]. Patients with spondyloepiphyseal dysplasia are characterized not only by disproportionate short posture (dwarfism), but also by progressive involvement of the spine and epiphyses of the long bones, resulting in kyphoscoliosis and limb malalignment with progressive hip deformities [8, 17, 22]. In patients with mucopolysaccharidosis, glycosaminoglycan accumulation affects skeletal structure, connective tissues, and other organs, which may lead to joint contractures and limb and spine deformities. Atlantoaxial instability, spinal stenosis, and progressive kyphoscoliosis are of particular importance and may be associated with spinal cord compression or myelopathy in both types of dysplasias [5, 8, 17, 25, 26]. We identified two patients with skeletal dysplasia who experienced a neurologic complication and present their cases, with a review of the relevant literature.

Case Report

No institutional review board approval was needed for a case report of two patients. We reviewed the cases of two patients: one with mucopolysaccharidosis and one with spondyloepiphyseal dysplasia, who had neurologic injury after lower extremity surgery. Our outcome measures included operative time, anesthesia time, intraoperative complications, estimated blood loss, O_2 saturation, mean arterial pressure (MAP), urine output, patient positioning, time for manifestation of deficit, and neurologic recovery. Postoperative acuity of care and neurologic history also were considered. Pre- and postdeficit neuraxis MR images were compared and radiographic images correlated to deficits were determined. The followup was a minimum of 2 years after surgery.

Two patients with skeletal dysplasia who had a new neurologic deficit with confirmatory neuroimaging after extremity surgery were identified. There were no other complications noted. Blood work including a complete blood count and coagulation test results were normal, and neither patient had a family history of thrombotic disease (Table 1).

Patient 1

An 8.5-year-old boy with spondyloepiphyseal dysplasia congenita and severe coxa vara bilaterally underwent

Table 1. Patient characteristics

Patient	Age; sex	Diagnosis/apex of kyphosis	Unrelated surgery	EBL (cc)	Operative time/anesthesia (minutes)	Time to deficit	Postoperative MRI findings
1	8 years, 6 months; male	SED, hyperkyphosis, coxa vara bilateral Kyphosis 76° (T1–T7, T4 apex)	Bilateral femoral valgization osteotomies	200	293/398	Immediate after surgery	Spinal cord signal abnormality on T2 at apex of kyphoscoliosis
2	6 years, 6 months; male	MPS, hyperkyphosis, bilateral hip dysplasia Kyphosis 80° (C7–T4, T2 apex)	Bilateral hip open reduction and innominate osteotomy	400	252/473	Seen at 4 hours postoperative	Increased T2/STIR signal

Level of paralysis	Treatment	Lowest MAP at surgery	Postoperative care (PICU vs floor)	Time to recovery	Last MRI
T4-T5	Solu-Medrol® 30 mg/kg IV bolus, 5.4 mg/kg for 23 hours	40 mm Hg	PICU	5 months, patient returned to near baseline strength	Increased signal at T4-T5. The cord appears slightly narrowed, 6months postoperative
T2	Methyl-prednisolone IV	48 mm Hg	PICU	None at 2 years, 3 months followup	Atrophy of cord at 2-years, 3-months postoperative

EBL = estimated blood loss; MAP = mean arterial pressure; PICU = pediatric intensive care unit; SED = spondyloepiphyseal dysplasia; IV = intravenous; MPS = mucopolysaccharidosis; STIR = Short tau inversion recovery; Solu-Medrol® (Pfizer Inc, Peapack, NJ, USA).
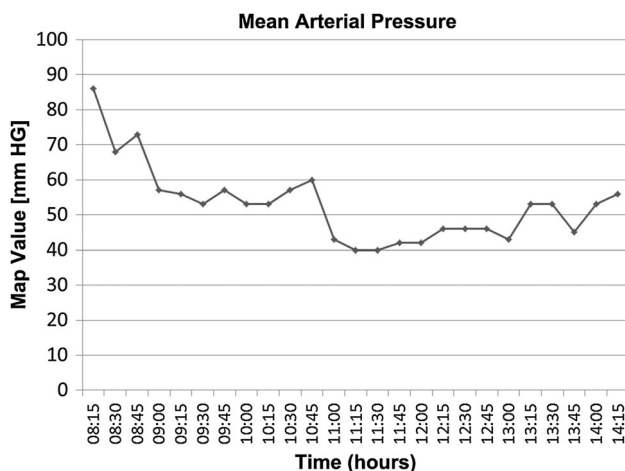
bilateral proximal femoral realignment osteotomies at one surgery. Preoperative neurologic examination was normal. Scoliosis and midthoracic kyphosis were 115° and 76°, respectively. The thoracic kyphosis apex was T4–T5 (Table 1).

General anesthesia was administered with the patient in the supine position. Total anesthesia and surgery time were 398 and 293 minutes, respectively. Neuromonitoring was not used. Estimated blood loss was 200 cc with maintenance of O$_2$ saturation at 95%. Low MAP values (40 mm Hg) occurred twice for 15 minutes (Fig. 1). Urine output was within normal limits (1.2–1.7 mL/kg/hour).
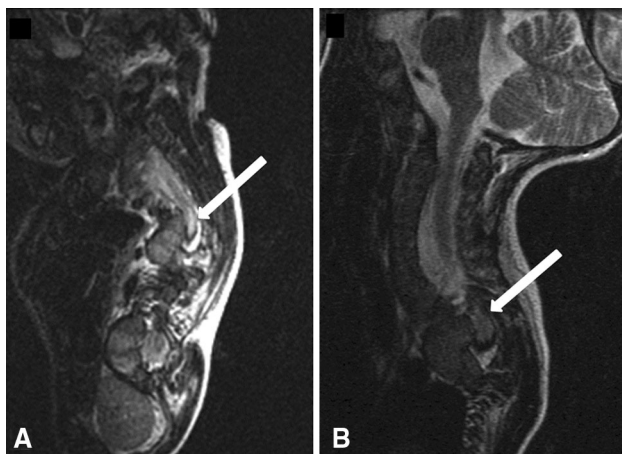
Dense paraplegia of both lower extremities was observed postoperatively but both showed nondermatomal paresthesia and no discernible sensory level loss. Five-hour

Fig. 1 The intraoperative mean arterial pressure (mm Hg) levels of Patient 1 in relation to time of surgery are shown.

Fig. 2A–B (**A**) An MR image taken 30 minutes after manifestation of clinical signs of paralysis in Patient 1 shows the T2 signal changes in the spinal cord at the kyphosis level (arrow). (**B**) This MR image shows the same zone (arrow) with the heterogeneous signal 6 months later.

postoperative MR images showed a T2-signal change in the dorsal spinal cord at T4–T5 (apex of kyphosis), without evidence of compression at any level (Fig. 2).

Postoperatively, the patient was started on methylprednisolone (30 mg/kg bolus, then 5.4 mg/kg/hour × 23 hours infusion). The patient had spontaneous movement of his left great toe on postoperative Day 3. He had voluntary urine voiding on the sixth day after surgery after initial neurogenic bladder with urinary retention. Motor strength was 4 of 5 and there was no clinical evidence of myelopathy at his 5-month postoperative visit. MR images of the spine indicated a decrease in T2 signal abnormality with slight loss of cord volume at the corresponding region. The patient is able to complete activities of daily living to his satisfaction 5 years postoperatively.

Patient 2

A 6-year-old boy with mucopolysaccharidosis 1-H underwent bilateral innominate osteotomies and proximal femoral varus osteotomies during one surgical event. Cervicothoracic kyphosis was 80°. The apex of the thoracic kyphosis was T2–T3 (Table 1). His preoperative neurologic examination was normal. General anesthesia was administered with the patient in the supine position. Total anesthesia and surgical time were 473 and 252 minutes, respectively. Three additional procedures were performed: dental restorations, MRI, and bilateral medial tibial hemiepiphysiodesis. For pediatric hip reconstruction, the surgeons did not deem any dental procedure as a contraindication. No neuromonitoring was done during surgery. Estimated blood loss was 400 cc and O$_2$ saturation level was greater than 95% during surgery. Urine output was within normal limits (1.3–1.7 mL/kg/hour). The patient's MAP decreased to 48 mm Hg 30 minutes before the end of the procedure and persisted for 2 hours.

Flaccid paralysis was discovered 4 hours postoperatively as the patient started to recover from the epidural anesthesia. The entire case was done with the patient receiving epidural anesthesia and sedation. Deep tendon reflexes and movement were absent. Skin sensation was present. MR images obtained the following morning showed ischemia of the spinal cord at the T2 level. High-dose, intravenous corticosteroid therapy and pharmacologic treatment for hypertension were started. Discharge occurred on Day 52 postoperatively with the patient having continued complete flaccid paraplegia and bladder incontinence. At the last followup 27 months after surgery, paralysis of both lower extremities with complete sensory and motor loss and bladder incontinence were still present. MR images obtained 3 months and 27 months postoperatively showed atrophy of the spinal cord.

Discussion

Patients with skeletal dysplasia may require operative procedures for long bone or hip reconstruction [4, 24]. Children with skeletal dysplasia also may have associated atlantoaxial instability, spinal stenosis, or kyphoscoliosis.

There were several limitations to this study. First, we had only two patients and therefore a statistical analysis for potential causes of paraplegia were not possible; however, as this complication is not well appreciated by treating physicians, it is worth noting that it can occur. We explored many plausible explanations for the cause of the neurologic injury. We believe that the decrease in MAP during the procedure might be a cause but we cannot document that maintaining MAP at a consistent level would have prevented the injuries. The actual cause or causes of the neurologic injury, other than the spinal cord ischemia observed on MR images, remain unknown in these two patients. We also cannot comment on the frequency of this complication because we could not accurately assess every similar patient with skeletal dysplasia and spine deformity who underwent lower extremity surgery at our institutions, but we think there were 52 during an 8½-year period.

To our knowledge, our study is the first report of spinal cord injury associated with extremity surgery in patients with skeletal dysplasia. Neurologic sequelae related to the spinal cord are rare after nonspinal operations, with the overall incidence estimated to be 0.08% [2, 19].

Many factors contribute to a patient's risk for a spinal cord event: low MAP, low O_2 saturation, duration of the operation, positioning on the operating table, and the presence of a spine deformity, or unappreciated small vessel vascular disease unique to patients with skeletal dysplasias. We know of no evidence that a synergistic action occurs between these many factors; however, MAP may be the main factor. Both patients in our study had kyphosis (76° and 80°) at the time of the procedure but neither showed any spinal or peripheral nerve dysfunction preoperatively. These kyphosis measurements are not exceptional for patients with skeletal dysplasia.

Blood Pressure and MAP

Both patients had their MAP measured on a continuous basis during surgery. Low MAP (< 60 mm Hg) is an important risk factor for spinal cord injury intra- and postoperatively [6]. Data regarding spinal ischemia injury in patients in the supine position are limited [19]. Blood pressure changes and heart rate instability may not be sensitive enough to caution the surgical team regarding low cardiac output [25]. In a young, healthy adult patient, a MAP of 50 mm Hg to 60 mm Hg is well tolerated, but higher pressures may be required in patients with cardiovascular disease. According to Haque and Zaritsky [11], the MAP for patients at age 6 years should be 70 ± 20 mm Hg.

Spinal Cord Ischemia

The diagnosis of spinal cord ischemic injury was made with MRI in both children. Most often, the clinical presentation of spinal cord ischemia is anterior spinal syndrome, with paraplegia and loss of pain, temperature, and touch sensation (but with relative sparing of pressure, vibration, and proprioception sensation). Urinary and fecal incontinence, impotence, and spinal dysautonomia also may occur [12]. Specific risk factors, such as spinal stenosis, vascular disease, intraoperative hypotension, or the use of epinephrine in the local anesthetic solution may cause spinal cord ischemia [1, 18]. In a case report of spinal cord ischemia after esophagectomy, the patient had a thoracic epidural catheter placed for postoperative pain control [21]. The spinal catheter was used in our second patient but at the lower lumbar level. Tong et al. [25] showed the upper thoracic level (T2–T4) to be a region at particular risk for ischemic injury, which corresponds to the watershed region of the spinal cord and therefore is most susceptible to changes in blood flow. The anterior spinal artery arises from the intercostal arteries and is one of three arteries associated discontinuously along the spinal cord. The anterior spinal artery supplies the anterior ⅔ of the spinal cord [16]. The posterior spinal arteries are fed by segmental arteries that arise from the aorta. There are more segmental arteries supplying the posterior spinal arteries than the anterior arteries, which accounts for anterior spinal artery syndrome being more frequently observed [19]. The diameter of spinal vessels may influence the blood flow through the spinal cord [15, 19]. In a dynamic flexion cadaveric study, Farley et al. [9] found that, in thoracic kyphosis exceeding 63°, spinal cord intramedullary pressure increased significantly compared with less substantial curves in the same specimen. However, a change of intramedullary pressure with progressive increases in the kyphosis angle on specimens did not correlate with spinal cord stenosis in the thoracic spine [9].

Positioning on the Operating Table

There are numerous reports of reduction of evoked potentials during patient positioning for surgery [6, 14, 21, 23, 25]. All but one of the patients in these studies sustained a spinal cord injury while they were in the prone position, whereas our patients were in the supine position [1, 3, 19–21]. Paraplegia attributable to a thoracic spinal

cord infarction was described in a 16-year-old patient with Morquio syndrome (mucopolysaccharidosis IV) [25]. The child had surgery on the foramen magnum and atlantal decompression led to paraplegia because of a thoracic spinal cord infarction while the patient was in the prone position [25].

Intraoperative Monitoring

Neither of our study patients had neuromonitoring during surgery. It might be an important consideration when performing lower extremity procedures on patients with skeletal dysplasia, but neuromonitoring presents some challenges in maintaining a sterile field. We now use sterile neuromonitoring leads with adhesive tape, and pass over the end of the table after prepping the patient. The sterile field was maintained and allowed for manipulation of the lower extremity as needed.

Motor evoked potentials and somatosensory evoked potentials usually are performed to monitor spinal cord function of patients during spine surgeries. The somatosensory evoked potentials may identify 90% of clinically relevant neurologic events [25]. The motor evoked and somatosensory evoked potentials together obtain almost 100% sensitivity and specificity [7, 10, 25].

Some patients with severe thoracic kyphoscoliosis are at risk for ischemic cord injury during lower extremity surgery. In our series, the ischemic injury was located near the apex of the thoracic kyphotic deformity. The avoidance of low MAP intraoperatively and during postoperative care may be important for children with substantial thoracic kyphosis in the watershed area. Surgeons should consider neuromonitoring whenever possible in these patients, although this presents some logistic challenges in maintaining a sterile field with the placement of monitoring electrodes.

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