Spinal stenosis surgery in pediatric patients with achondroplasia

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Object. Achondroplasia is a hereditary form of dwarfism caused by a defect in endochondral bone formation, resulting in skeletal abnormalities including short stature, shortened limb bones, macrocephaly, and small vertebral bodies. In the pediatric population, symptomatic spinal stenosis occurs at all spinal levels due to the abnormally narrow bone canal. In this study, clinical outcomes were assessed in children with achondroplasia after spinal canal decompression.

Methods. A retrospective review was conducted involving pediatric patients with heterozygous achondroplasia and symptomatic stenosis after decompressive procedures at the authors’ institution within a 9-year period. Measured outcomes included resolution of symptoms, need for repeated surgery, presence of fusion, development of deformity, and complications.

Forty-four pediatric patients underwent a total of 60 decompressive procedures. The average patient age at surgery was 12.7 years (range 5–21 years). Forty-nine operations were performed for initial treatment of stenosis, and 11 were performed as revision surgeries on previously operated levels. A large proportion of patients (> 60%) required additional cervicomedullary decompressions, most often preceding the symptoms of spinal stenosis. Of the initial procedures, decompression locations included 32 thoracolumbar (65%), 10 lumbar (20%), four cervical (8%), two cervicothoracic (4%), and one thoracic (2%). Forty-three of the decompressive procedures (72%) included spinal fusion procedures. Of the 11 revisions, five were fusion procedures for progressive deformity at levels previously decompressed but not fused (all thoracolumbar), five were for decompressions of symptomatic junctional stenosis with extension of fusion, and one was for repeated decompression at the same level due to recurrence of symptomatic stenosis.

Conclusions. Decompression of the spinal canal in pediatric patients with achondroplasia can be accomplished safely with significant clinical benefit. Patients with a history of cervicomedullary compression may be at an increased risk of developing symptomatic stenosis prior to adolescence. Fusion procedures are recommended in patients with a large decompression overlying a thoracolumbar kyphosis to avoid progressive postoperative deformity.

KEY WORDS • achondroplasia • cervicomedullary compression • spinal stenosis • decompression surgery • spinal fusion • pediatric neurosurgery

Achondroplasia, the most common cause of skeletal dysplasia, is an autosomal-dominant inherited dwarfism syndrome that occurs in one of every 15,000 to 40,000 live births. People with this condition typically are of small stature with shortened proximal limbs, small fingers and toes, a large head with a prominent forehead, spinal kyphosis or lordosis, and bow-leg (varus) or knock-knee (valgus) deformities. In addition to the more obvious abnormalities that lead to deformity of the appendicular and axial bones, patients with achondroplasia also possess more subtle abnormalities of the axial skeleton including a relatively small skull base with a narrowed foramen magnum and small vertebral bodies with shortened pedicles. As a result, achondroplasia is commonly associated with several neurological conditions such as hydrocephalus, cervicomedullary compression, cervical or thoracic cord compression, and lumbar spinal stenosis due to bone compression along the neuraxis.

Although it is well established that patients with achondroplasia possess narrowed spinal canals due to shortened pedicles and decreased interpedicular distance, surgical treatment of the ensuing neurological compression in pediatric patients can be technically difficult for the pediatric neurological or orthopedic surgeon and may also be associated with a high rate of postoperative progressive deformity. Specifically, the dura mater is exceedingly thin and friable in patients with achondroplasia, especially in pediatric patients, increasing the likelihood of unintentional durotomy and subsequent CSF leakage during surgical decompression. In addition, skeletally immature patients are at high risk for developing postlaminectomy thoracolumbar

Abbreviations used in this paper: CMD = cervicomedullary decompression; CSF = cerebrospinal fluid; MR = magnetic resonance.
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kyphoses, often necessitating concurrent or delayed instrumented fusion in such patients. In this retrospective study, we reviewed the charts of 44 pediatric patients with achondroplasia who underwent spinal decompression procedures for symptomatic stenosis at our institution within a 9-year period. Particular focus was placed on presentation and resolution of symptoms, need for further surgery, presence of fusion, postoperative deformity, and complications.

Clinical Material and Methods

We performed a retrospective study of the records of all pediatric patients with heterozygous achondroplasia who underwent spinal decompressive procedures at our institution between 1996 and 2005. Forty-four patients were identified who underwent a total of 60 decompressive procedures. The charts were reviewed for demographic data, presenting symptoms, type and location of surgery, presence of fusion operation, postoperative course, and complications.

Clinical Evaluation

The clinical evaluation of all patients was performed by a multidisciplinary team that included pediatric neurosurgeons, pediatric orthopedic surgeons, pediatric neuroradiologists, and pediatricians. All patients underwent a thorough preoperative neurological examination, with particular attention to signs and symptoms of myelopathy and radiculopathy. Patients were questioned regarding characteristics of back and/or extremity pain, denoting quality, duration, location, and aggravating and relieving factors. Presence and duration of motor weakness, sensory abnormalities, and bowel and bladder symptoms were also recorded. All patients underwent plain radiography and an MR imaging study of the spinal region in question to assess the bone anatomy of the spine and the degree of compression on the neural elements, respectively.

Diagnostic Criteria

The diagnosis of symptomatic spinal stenosis was based on the correlation of clinical presentation with imaging criteria. The criteria used to make the diagnosis included the following: 1) the presence of signs or symptoms of chronic spinal cord compression (weakness, bowel or bladder dysfunction, hyperreflexia or hypertonia, spastic gait, or clonus) or chronic nerve root compression (neurogenic claudication, weakness or sensory disturbance in a radicular pattern, or hyporeflexia); and 2) neuroimaging evidence of spinal stenosis.

Surgical Procedure

All patients underwent a spinal decompressive procedure at the site of radiographic evidence of spinal canal narrowing that corresponded to the clinical symptomatology. If patients were undergoing cervical spine decompressions, they were placed in Mayfield cranial frames. After induction of general anesthesia, patients were placed prone. In patients in whom a Mayfield frame had been placed, it was rigidly fastened to the bed. In patients in whom no such frame was used, the head was carefully supported on a well-padded pediatric horseshoe. Somatosensory evoked potentials were routinely used throughout the positioning and operative stages of the procedure to assess posterior column integrity. In addition, in patients in whom internal fixation was performed, transcranial electrical stimulation-induced motor evoked potentials were used to monitor anterior cord function, and free run and intermittent electromyography were also conducted to intermittently assess nerve root function. Electroencephalography was often used at selected intervals to determine a burst suppression pattern as a surrogate marker of excessive anesthetic suppression of motor evoked potential signals.

In all cases, a midline incision was made over the spinal segments of concern, followed by subperiosteal dissection performed sufficiently laterally to visualize the transverse processes. Care was taken to avoid manipulation of bone elements over stenotic levels. Laminectomies were then performed using rongeurs and high-speed drilling. Care was also taken to avoid entering the dura at this stage of the operation. All resected bone was saved as potential autografts for later fusion procedures. The ligamentum flavum was also removed, and partial or total facet joint removal was completed in cases in which encroachment on the canal or neural foramina was significant. The patient anatomy was invariably notable for thickened ligamentum flavum and thin dura, such that the neural elements could easily be visualized inside the thecal sac. Following removal of these posterior elements, the dura was frequently noted to expand significantly from its previous position.

In those cases in which internal fixation and fusion were performed, lateral mass screws were placed in cervical vertebrae and pedicle screws were placed in thoracic and lumbar vertebrae using external landmarks and fluoroscopy. At every level, pedicles were drilled, measured, and tapped prior to screw insertion. Electromyographic recordings were monitored for indications of bone breaching and potential nerve root damage during hardware placement. Corresponding rods were contoured before construct tightening. The fusion bed was prepared in a standard fashion by decorticating the local bone. Local resected bone from the laminectomy was combined with posterior iliac crest autograft and both were placed around the construct. Drains were placed if deemed necessary, and the wounds were then closed in multiple layers. After the patient awakened from anesthesia, a focused neurological examination was conducted to delineate movement of all four extremities. Patients were subsequently taken to the pediatric intensive care unit.

Postoperative Evaluation

Patients were observed carefully for improvement or resolution of their preoperative neurological symptoms. Serial neurological examinations were conducted in patients prior to discharge. Patients were discharged when their surgical wounds appeared to be healing well, their drains were removed, and they could tolerate pain with the assistance of oral medication. Follow-up data regarding neurological status, spinal alignment, and successful fusion were obtained postoperatively at 1, 3, 6, 12, and 24 months when feasible.

Results

Patient Population

Of the 44 children with achondroplasia undergoing spinal
decompressive procedures, 19 were girls (43%) and 25 were boys (57%). The mean age of the patients at the time of operation was 12.7 years (range 5–21 years), and the mean duration of symptoms was 9.2 months (range 1–54 months) (Table 1).

**Location of Symptomatic Stenosis**

All 44 patients underwent at least one decompressive surgery, and five of these patients underwent additional spinal decompressive procedures at another area of the spine because of the development of new clinical and radiographic stenosis. Including these five additional procedures, symptomatic spinal stenosis was diagnosed overwhelmingly in the thoracolumbar segment of the spine (32 [65.3%] of 49 patients) and lumbar spine (10 [20.4%] of 49 patients). The remaining cases of stenosis occurred in the cervical spine in four patients (8%), cervicothoracic spine in two (4%), and thoracic spine in one patient (2%) (Table 2). Of note, 27 (61.4%) of the 44 patients had experienced symptomatic cervicomedullary stenosis in addition to spinal stenosis, and the majority of these patients (25 [93%] of 27) were treated with CMD procedures before the onset of symptomatic spinal stenosis (Table 1). The average age of these 25 patients at the time of CMD was 3.5 years (range 3 months–17.5 years), the average age at the time of subsequent spinal decompressive surgery was 11.5 years (range 5–21 years), and the average interval between the two procedures was 8 years (range 6 months–6.5 years). Interestingly, 27 of the 43 pediatric patients with achondroplasia in our institutional database who underwent CMD later required spinal stenosis decompressive surgery during follow up, prior to 21 years of age. The details of these surgeries were not included in this analysis.

**Clinical Presentation**

Pain was the most common presenting symptom of spinal stenosis in our patient population. Consistent with the anatomical distribution of stenosis in our patients, neurogenic claudication (bilateral leg pain and motor and sensory abnormalities worsened by extended posture) with or without low-back pain or radicular pain was present in 40 (91%) of 44 patients (Table 1). Including one patient who presented with only radicular pain and one patient presenting with only low-back pain, 42 (95.4%) of 44 patients had some type of pain syndrome upon presentation. Urinary and/or bowel incontinence occurring in the context of neurogenic claudication was noted in 11 patients (25%), delineating a cauda equina presentation. Clear myelopathic symptoms and signs were only present in two patients (4.5%). Age distribution at the time of surgery is presented in Fig. 1. Regarding spinal alignment, preoperative kyphosis and scoliosis were present in 22 patients (50%) and in 14 patients (32%), respectively. Nine patients (20%) previously had concomitant hydrocephalus requiring the insertion of CSF shunts (Table 1).

**Surgical Results and Complications**

The forty-four patients underwent a total of 60 spinal decompression procedures between 1996 and 2005. Forty-three operations (72%) involved fusion procedures with internal fixation. When revision surgery was necessary, the most frequent reasons were for progressive deformity in a nonfused spine (5), decompression of junctional stenosis (5), and repeat decompression at the same level (1). Complications occurred in 7 (11.6%) of the 60 procedures: durotomy in 4, wound breakdown/infection in 2, and instrumentation revision in 1.

### TABLE 1

*Baseline characteristics of pediatric patients with achondroplasia before decompressive surgery for symptomatic spinal stenosis*

<table>
<thead>
<tr>
<th>Variable</th>
<th>No. of Cases/Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>no. of patients</td>
<td>44</td>
</tr>
<tr>
<td>sex (%)</td>
<td></td>
</tr>
<tr>
<td>boys</td>
<td>25 (57)</td>
</tr>
<tr>
<td>girls</td>
<td>19 (43)</td>
</tr>
<tr>
<td>mean age at op (yrs)</td>
<td>12.7 (range 5–21)</td>
</tr>
<tr>
<td>mean duration of preop symptoms (mos)</td>
<td>9.2 (range 1–54)</td>
</tr>
<tr>
<td>history of CMD (%)</td>
<td>27 (61)</td>
</tr>
<tr>
<td>preop kyphosis (%)</td>
<td>22 (50)</td>
</tr>
<tr>
<td>preop scoliosis (%)</td>
<td>14 (32)</td>
</tr>
<tr>
<td>hydrocephalus (%)</td>
<td>9 (20)</td>
</tr>
<tr>
<td>presenting symptoms (%)</td>
<td></td>
</tr>
<tr>
<td>pain</td>
<td>42 (95.4)</td>
</tr>
<tr>
<td>neurogenic claudication/cauda equina*</td>
<td>40 (91)</td>
</tr>
<tr>
<td>radicular pain only</td>
<td>1 (2.3)</td>
</tr>
<tr>
<td>low-back pain only</td>
<td>1 (2.3)</td>
</tr>
<tr>
<td>myelopathy</td>
<td>2 (4.5)</td>
</tr>
</tbody>
</table>

* Presence of urinary and/or bowel incontinence occurred in the context of neurogenic claudication in 11 (25%) of 44 patients.

### TABLE 2

*Characteristics of surgical procedures in 44 patients with achondroplasia*

<table>
<thead>
<tr>
<th>Variable</th>
<th>No. of Cases/Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>total no. of procedures</td>
<td>60</td>
</tr>
<tr>
<td>initial procedures</td>
<td>49</td>
</tr>
<tr>
<td>revision procedures</td>
<td>11</td>
</tr>
<tr>
<td>location of initial procedure (%)</td>
<td></td>
</tr>
<tr>
<td>thoracolumbar</td>
<td>32 (65.3)</td>
</tr>
<tr>
<td>lumbar</td>
<td>10 (20.4)</td>
</tr>
<tr>
<td>cervical</td>
<td>4 (8)</td>
</tr>
<tr>
<td>cervicothoracic</td>
<td>2 (4)</td>
</tr>
<tr>
<td>thoracic</td>
<td>1 (2)</td>
</tr>
<tr>
<td>fusion procedures w/ internal fixation</td>
<td>43</td>
</tr>
<tr>
<td>nonfusion procedures</td>
<td>17</td>
</tr>
<tr>
<td>mean follow up (mos)*</td>
<td>34 (range 8–93)</td>
</tr>
<tr>
<td>reasons for revision surgery</td>
<td></td>
</tr>
<tr>
<td>progressive deformity in nonfused spine</td>
<td>5</td>
</tr>
<tr>
<td>decompression of junctional stenosis</td>
<td>5</td>
</tr>
<tr>
<td>repeat decompression at same levels</td>
<td>1</td>
</tr>
<tr>
<td>complications (%)</td>
<td>7 (11.6)</td>
</tr>
<tr>
<td>durotomy</td>
<td>4</td>
</tr>
<tr>
<td>wound breakdown/infection</td>
<td>2</td>
</tr>
<tr>
<td>instrumentation revision</td>
<td>1</td>
</tr>
</tbody>
</table>

* Nine of 44 patients had limited follow up because of moving out of state/country; not included in mean follow up.

Fig. 1. Bar graph showing the age distribution (in years) of 44 patients with pediatric achondroplasia at the time of surgery.
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instrumented fixation and autografts. Forty-nine decompression operations were performed on stenotic segments of the spine that had not undergone previous operations, and 11 operations were revision surgeries for previously operated levels (Table 2). Of these 11 revision procedures, five were fusion procedures for progressive deformity and back pain at levels previously decompressed but not fused, five were for decompressions of symptomatic junctional stenosis with extension of fusion, and one was for repeated decompression due to an acute disc herniation at the L2–3 interspace within a previous L1–5 laminectomy (Table 2). Although this latter patient had previously undergone an L1–5 in situ fusion without instrumentation, during the reoperation she underwent L2–3 pedicle screw fusion and placement of an interbody graft at that level to ensure solid fusion. No patient experienced progressive kyphosis at the segments previously fused, and no patient experienced instrumentation failure.

Nine of the patients had limited follow up (less than 6 months). These patients largely lived out of state or out of the country and chose to continue follow up with neurological and/or orthopedic surgeons closer to their places of residence. The remaining patients were followed up for an average of 34 months (range 8–93 months). All patients experienced significant improvement or resolution of symptoms of spinal stenosis. Thirty-two patients experienced durable resolution throughout the entirety of their follow up, and 11 patients had recurrence or new symptoms during their follow up necessitating repeated surgery as described previously. One patient had clinical persistence of myelopathic signs and symptoms despite adequate decompression of the cervical spinal canal and no obvious stenosis elsewhere in the neuraxis as corroborated by MR imaging and computed tomography myelogram results. New lower-extremity radicular pain occurred in three patients immediately following surgery. The pain was managed conservatively in two patients with good improvement. The third patient returned to the operating room during hospitalization for repositioning of a pedicle screw postulated to be responsible for the new radicular pain. After screw repositioning in this patient, radicular pain drastically improved, and the patient experienced no lasting pain syndrome after the revision.

Unintentional durotomy occurred during four procedures, two of which occurred in the same patient during a successive decompressive procedure. All durotomies were primarily closed with sutures and supplemented with fibrin glue. Postoperatively, these patients were kept in a flat position for at least 72 hours, and none experienced CSF leaks through the skin. Two patients experienced wound breakdown. One wound was washed out and primarily closed in the operating room without significant sequelae. The second patient had a pressure ulcer that developed over the underlying instrumentation with eventual visualization of the internal hardware. After revision of this wound, plastic surgeons rotated a muscular flap to cover the defect, but the instrumentation and graft were kept in place, and the patient’s spine healed and fused adequately. Each of these two patients was treated for a wound infection with a standard course of antibiotics. No other wound infections or meningitides occurred. No deaths resulted from these procedures.

Discussion

Achondroplasia occurs because of a point mutation on the long arm of chromosome 4, leading to a defect in fibroblast growth factor Type 3 receptors and subsequent abnormal limb growth and spinal segment formation.\textsuperscript{5,7,22,24,26,29,37} Common abnormalities found in pediatric patients with achondroplasia include delays in mental and motor development, feeding and sleep disorders, hypotonia, and macrocranium with or without hydrocephalus, but neurosurgical interest is largely centered on the impaired formation of vertebrae and foreshortening of the basicranium, which lead to spinal stenosis and cervicomedullary compression, respectively.\textsuperscript{3,9,11–13,27,30,33,36,38} In addition, thoracolumbar kyphosis can be a common cause of pain and debility in such patients, especially when it contributes to stenosis of the spinal canal and foramina.\textsuperscript{1,31,32}

Spinal Stenosis

Patients with achondroplasia have both regional and global spinal abnormalities (Fig. 2). Pedicles are shortened in anteroposterior length, and the interpedicular distance is narrowed, leading to a smaller cross-sectional area of the spinal canal for the neural elements.\textsuperscript{10} Jeong and colleagues\textsuperscript{14} used MR imaging to evaluate lumbar stenosis in patients with achondroplasia. These investigators found that, in the normal spinal canal, the interpedicular distance widens from L-1 to L-5, whereas in patients with achondroplasia this distance progressively decreases from L-1 to L-5. In addition, because of this narrower canal, patients with achondroplasia were more likely than others to develop symptoms of spinal stenosis. It is this developmentally narrow canal combined with degenerative changes that seems to be the most important factor for the development of symptomatic stenosis of the lumbar canal in achondroplasia.

When stenosis results in pressure on the spinal cord, cauda equina, or nerve roots, pain syndromes and/or neurological compromise may become progressive. It has been estimated that between 10 and 25% of patients of all ages with achondroplasia and spinal stenosis will require surgical treatment for such progression.\textsuperscript{10} Most commonly, patients experience numbness, weakness, cramping, or general pain in the legs, representing a classic neurogenic claudication appearance. If spinal stenosis is particularly severe, patients may have problems with bowel and bladder function. Such symptomatology was found in the majority of the patients in our cohort; 40 (91%) of 44 patients presented with neurogenic claudication (bilateral leg pain and motor and sensory abnormalities worsened by extended posture), and of those 40, 11 (25%) also complained of progressive urinary and/or bowel incontinence. Although some investigators believe that foraminal stenosis that leads to isolated radicular pain is more clinically relevant than central canal narrowing in patients with achondroplasia,\textsuperscript{11} such pain syndromes were very uncommon in the present series and only one patient presented with this syndrome.

It is generally accepted that spinal stenosis in patients with achondroplasia is a problem that develops primarily in young adults, due to concurrent degenerative spondylosis and ligamentum flavum hypertrophy, which further tightens an already stenotic canal.\textsuperscript{2,15,34} Furthermore, it is likely that such stenosis is exacerbated by the thoracolumbar kyphosis or the compensatory lumbar hyperlordosis below thoracolumbar deformities that often occur in patients with achondroplasia.\textsuperscript{15,38} This cohort showed a broad distribution of ages, however, with over half of our patients presenting pri-
or to 12 years of age, suggesting that symptomatic stenosis may be more common in these children than previously suspected. At least three possible explanations exist for this discrepancy. First, our institution is a large referral center for pediatric patients with achondroplasia and possibly admits patients with more significant pathology who present at earlier ages. Second, a large number of patients in this cohort underwent prior CMDs at a young age. Such patients are routinely observed very closely by a neurosurgeon, perhaps allowing earlier detection of spinal stenosis symptoms. And third, patients with pediatric achondroplasia with a history of cervicomedullary compression may form a subset of patients that experience more severe bone constriction of the entire neuraxis. Such compression may be so severe that symptoms of spinal stenosis occur prior to the ages at which degenerative changes contribute to spinal canal narrowing. Such a theory is not only supported by the data in the present study, in which 27 (61%) of 44 patients underwent additional decompressive procedures for symptomatic cervicomedullary compression, but also by previously published work by Bagley and coworkers. In this cohort of patients with pediatric achondroplasia undergoing CMD at our institution, 27 (63%) of the 43 patients studied by Bagley and colleagues have thus far eventually required spinal decompressive surgery for symptomatic stenosis prior to 21 years of age. Pediatric patients with achondroplasia requiring CMDs may therefore be at greater risk of developing symptomatic spinal stenosis prior to adulthood.

**Thoracolumbar Kyphosis**

Patients with achondroplasia often develop a thoracolumbar kyphosis that is present in approximately 11% by 10 years of age, but may be greater than 30% in patients older than 30 years of age. Such deformity is thought to be due to limited anterior longitudinal growth that usually occurs at the epiphyseal plates between birth and the attainment of adult height at the age of 18 to 20 years. There is often a compensatory lumbar lordosis, pelvic tilt, and subsequent fixed flexion deformity of the hip joints. Although it has been suggested that the incidence of thoracolumbar kyphosis in infants with achondroplasia can be greater than 90%, the overwhelming majority of such patients will show spontaneous resolution of the kyphosis by 3 years of age. Thus, a thoracolumbosacral orthotic brace will be indicated in patients at very young ages for a progressive kyphosis of 30° or greater or for the appearance of anterior vertebral wedging. Using these criteria, Pauli et al. demonstrated control of the thoracolumbar kyphosis when brace therapy was initiated prior to age 3.

If patients experience thoracolumbar kyphosis into adolescence, however, the deformity will often be fixed in nature, necessitating operative reduction and fixation. Moreover, decompressive procedures in this area lead to loss of the posterior ligamentous tension band across the apex of deformity, placing the patient at risk for progressive kyphosis. Tolo noted in adult patients the probable progression of thoracolumbar kyphosis following laminectomy without supplemental fusion in the presence of a preoperative kyphosis greater than 40°. Ain and colleagues also noted the progression of thoracolumbar kyphosis in children following decompressive procedures without fusion. In the present study cohort, five patients developed progressive kyphosis at the thoracolumbar junction following decompressive procedures for symptomatic stenosis.

![Fig. 2. Illustration of the thoracolumbar spine in a pediatric patient with achondroplasia showing the abnormal bone anatomy that leads to neural compression.](image)
Surgical Complications

Regional anatomy of the spine in pediatric patients with achondroplasia can be particularly challenging for the surgeon who is decompressing the canal and/or placing internal fixation. The dura is typically thin and transparent and may be densely adherent to the underlying ligamentum flavum and periosteum. The intervertebral discs and ligamentum flavum are frequently hyperplastic, and the amount of epidural fat is decreased, resulting in less protection for the neural elements than normal. For this reason, it is generally advised that the laminae should first be drilled down to expose the ligamentum flavum before complete bony resection using rongeurs. In the present patient series, four (6.7%) of the 60 procedures resulted in unintentional durotomies, two of which occurred in the same patient. In each case, the dura was closed with sutures supplemented with fibrin glue. All patients were kept in a flat position for at least 72 hours, and none experienced CSF leaks through the skin.

In regard to the placement of pedicle screws, meticulous care must be taken when palpating the pedicle due to its diminutive size and proximity to neural structures. In the present patient series, one patient awoke with new radicular pain corresponding to an aberrantly placed pedicle screw. After return to the operating room for pedicle screw repositioning, the radicular pain improved and no lasting pain syndrome occurred. Although not necessary in all cases, fluoroscopy may provide corroboration of the ideal pedicle screw trajectory, especially in severely dysmorphic segments. Finally, similar to the treatment of compressive cervical myelopathy, adequate spinal cord perfusion must be maintained when operating over the spinal cord in such patients; constant monitoring of blood pressure, volume status, and electrophysiological integrity of the cord are required.

Conclusions

Pediatric patients with achondroplasia complaining of pain or fatigue in their legs, paresthesias in the anterior region of the thigh, and decreased walking distance might be experiencing spinal stenosis, indicating a need for MR imaging studies even without manifestations of other neurological symptoms. Such a possibility should be especially great in patients with previous CMD surgery, who may be predisposed to develop symptomatic spinal stenosis prior to adolescence or adulthood. Decompressive spinal surgery can be accomplished safely if particular attention is given to local anatomy and overall spinal alignment, with fusion recommended in cases of a large decompression overlying a thoracolumbar kyphosis.

References


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