The development of the vertebral column and spinal cord is a dynamic synchronized event during embryology that depends on the interaction of multiple substrates. Stages of skeletogenesis are based on the initial formation of a mesenchyme matrix and the ossification of cartilaginous scaffolding. The formation of the spinal cord occurs with the separation of mesenchymal tissue from the ectoderm and endoderm and adequate formation of the neural tube. It is believed that the levels of these 2 structures are at a similar height initially but that the vertebral column ascends more rapidly than the spinal cord. The conus medullaris may lie between L-2 and L-4 in fetuses at 30–39 weeks of gestation. The normal level of the conus after 40 weeks of gestation lies between L-1 and L-2. The lowest normal level is considered to be between L-2 and L-3.

Skeletal dysplasias are a large group of disorders, some of which can affect the cartilaginous and ossification process of bone, resulting in reduced vertebral body growth and a short stature. If the ascension of the conus were simply based on the difference in growth between neural tissue and bony elements, would the level of the conus be different? The purpose of this retrospective chart review was to determine the level of the conus in a population of pediatric patients with skeletal dysplasia.

Methods

A retrospective internal review board–approved analysis was undertaken at Nemours Alfred I. duPont Hospital for Children, Nemours Children’s Clinic, Wilmington, Delaware for the period from September 2000 to

Level of the conus in pediatric patients with skeletal dysplasia

Clinical article

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Object. Skeletal dysplasias are a heterogeneous group of disorders that affect bone development and can result in reduced vertebral body growth and short stature. The level of the conus medullaris is unknown in this population. The purpose of this review was to determine the level of the conus in a population of pediatric patients with skeletal dysplasia.

Methods. A retrospective chart review of a 7-year period was undertaken at a tertiary care pediatric hospital to identify patients in whom skeletal dysplasia had been diagnosed. Radiographs and MR imaging studies were evaluated to discern the level of the conus with respect to the bony vertebral column.

Results. Four hundred sixty-seven patients with skeletal dysplasia were identified. Radiographs and MR imaging studies were evaluated to discern the level of the conus with respect to the bony vertebral column. The mean conus level was found at the L-1 vertebral body. No difference was noted with respect to the age of the patients or the type of skeletal dysplasia. Two patients (1.7%) had a conus level lower than L-2.

Conclusions. Skeletal dysplasia leads to abnormal bone formation and can result in short stature. The location of the conus with respect to the vertebral bodies appears to be stable at the L-1 level regardless of patient age or the type of skeletal dysplasia involved. However, the appearance of a low-level conus and associated tethered cord syndrome may be slightly increased in this population. (DOI: 10.3171/2009.12.PEDS09364)

KEY WORDS • skeletal dysplasia • conus • tethered cord

This article contains some figures that are displayed in color online but in black and white in the print edition.
through May 2007. Initially, all electronic and paper charts were examined for patients that were identified by current procedural terminology (CPT) codes 259.4, 277.5, 733.29, 756.4, 756.50, 756.55, 756.56, 756.59, 756.89, and 756.9. Only patients in whom a clear diagnosis of a skeletal dysplasia had been made remained in the study. Demographics, including the specific diagnosis, patient sex, and patient age at the time of MR imaging, were extracted from the electronic chart. Hard-copy radiographic or electronic images obtained in these patients were viewed as well. Only the MR images that adequately demonstrated the level of the conus were used. Letters were sent to the families of patients whose radiographs were not available at our institution. The senior author (A.M.R.) viewed the outside MR images or reports or had a discussion with the referring hospital’s radiologist or neurosurgeon to discern the level of the conus. The tip of the conus was located on MR images by counting downward from the C-2 level or by counting upward on limited lumbar MR images after reviewing plain films for the appropriate numbering of lumbar vertebral bodies. The tip of the conus was based on its position closest to the intervertebral body disc space or midpoint of the vertebral body, and its position was documented as T-12, T12–L1, L-1, L1–2, L-2, L2–3, L-3, L3–4, L-4, L4–5, L-5, L5–S1, or sacral level.

Results

Study Population

Four hundred sixty-seven patients with a skeletal dysplasia over the 7-year time period were identified. Forty-seven different skeletal dysplasias were observed, and 23 types were demonstrated on corresponding images (Table 1). No radiographs were available from patients with cleidocranial dysplasia, thanatophoric dysplasia Type 1, Maroteaux-Lamy syndrome, acrodyplasia, metaphyseal chondrodysplasia, diaphyseal dysplasia, primordial dysplasia, frontometaphyseal dysplasia, Kabuki syndrome, Kniest dysplasia, melorheostosis, Russell-Silver syndrome, trichorhinophalangeal dysplasia Type 1, dysplasia epiphyseal hemimelica, Camurati-Engelmann disease, Leri-Weill syndrome, Beals syndrome, Goldenhar syndrome, Schwartz-Jampel syndrome, hypophosphatemia, osteopetrosis, and chondrodystrophy.

Level of the Conus

One hundred eleven patients had quality MR images allowing for the determination of the level of the conus. The male/female ratio was 1.3:1. The mean level of the conus was at L-1. Figure 1 features a graph of the level of the conus in the entire study population. The most common skeletal dysplasia encountered in this group was achondroplasia. Several closely related skeletal dysplasias were then combined to form the second most common type in our cohort: multiple epiphyseal dysplasia, spondyloepiphyseal dysplasia, spondyloepimetaphyseal dysplasia, spondylocostal dysplasia, and Stickler syndrome. The third most commonly observed type of skeletal dysplasia included the mucopolysaccharidoses. Lastly, metatropic dysplasia was observed with relative frequency. In all of these groups, the conus level was observed to be between L-1 and L-2 (Fig. 2).

The age range of patients at the time of MR imaging was 1 month to 21 years. There was no statistical difference in the conus level of 8 children younger than 1 year (T12–L1, 2 patients; L1, 4 patients; L1–2, 1 patient; and L-2, 1 patient) compared with that in the 103 children older than 1 year.

Patients With Low-Lying Coni

Two patients in our cohort exhibited low-lying coni. Their respective clinical scenarios were evaluated in more detail. One patient had Conradi-Hünermann syndrome. (Four of 5 patients with Conradi-Hünermann syndrome had available MR images.) This patient had a progressive kyphotic deformity to 100°. There were no symptoms of tethered cord prior to imaging. The MR imaging studies showed the conus to be at the level of the sacrum with an associated fatty filum. The patient underwent spinal cord detethering and fusion without difficulty. There were no further neurosurgical issues.

The second patient with a tethered cord had diastro-

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### TABLE 1: Summary of skeletal dysplasias demonstrated on MR imaging

<table>
<thead>
<tr>
<th>Skeletal Dysplasia</th>
<th>Total No. of Patients</th>
<th>No. w/ MRIs</th>
<th>Location of Dysplasia</th>
</tr>
</thead>
<tbody>
<tr>
<td>achondroplasia</td>
<td>204</td>
<td>36</td>
<td>T-12 15 7 2 0 0</td>
</tr>
<tr>
<td>camptomelia</td>
<td>8</td>
<td>5</td>
<td>T12–L1 1 0 0 0 0</td>
</tr>
<tr>
<td>chondrodysplasia punctata</td>
<td>8</td>
<td>2</td>
<td>L-1 1 0 0 0 0 0</td>
</tr>
<tr>
<td>Conradi-Hünermann</td>
<td>5</td>
<td>4</td>
<td>L1–2 1 0 2 0 1</td>
</tr>
<tr>
<td>diastrophic dysplasia</td>
<td>16</td>
<td>5</td>
<td>L-2 1 0 2 0 0</td>
</tr>
<tr>
<td>hemihypertrophy</td>
<td>2</td>
<td>2</td>
<td>L-3 1 0 2 0 0</td>
</tr>
<tr>
<td>Hunter syndrome</td>
<td>3</td>
<td>2</td>
<td>L-4 1 0 2 0 0</td>
</tr>
<tr>
<td>metatrophic dysplasia</td>
<td>14</td>
<td>7</td>
<td>L-5 1 0 2 0 0</td>
</tr>
<tr>
<td>mucopolysaccharidosis</td>
<td>27</td>
<td>10</td>
<td>L5–S1 1 0 2 0 0</td>
</tr>
<tr>
<td>pseudochondrodysplasia</td>
<td>21</td>
<td>6</td>
<td>S-1 1 0 2 0 0</td>
</tr>
<tr>
<td>spondyloepiphyseal dysplasia</td>
<td>44</td>
<td>17</td>
<td></td>
</tr>
<tr>
<td>unspecified</td>
<td>12</td>
<td>4</td>
<td></td>
</tr>
</tbody>
</table>

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**Phosphor dystrophy.** He was 1 of 15 patients with this syndrome and 1 of 4 who had MR images suitable for study. Magnetic resonance images showed the conus to be at L2–3. This patient was observed from 2001 to 2003, showing progressive kyphosis from 40 to 60°. Neurosurgical evaluation prior to orthopedic fusion revealed a 1.5-year history of back pain and an instance of left anterior thigh paresthesia with decreased endurance. No bowel or bladder symptoms were documented. The neurosurgical examination findings at that time were normal. Pulmonary function tests showed mild restrictive lung disease. Chest radiography showed mild enlargement of the heart. Initially, an anterior transthoracic approach from T4–9 was undertaken. When the patient was turned prone to complete the posterior approach, the motor evoked potentials decreased significantly. The patient was turned to the right lateral position with a return of the potentials to 80% of baseline. The procedure was aborted. The patient exhibited transient lower-extremity weakness. He returned to the operative arena 5 days later for a posterior decompression from T-3 through T-5 with fusion in the lateral position. Thirty minutes into the procedure, there was a bilateral loss of motor evoked potentials, which responded to blood pressure manipulation. The procedure was completed. The patient was unable to move his lower extremities on emerging from anesthesia.

**Discussion**

**Embryological Development**

The development of the vertebral column and spinal cord is a well-orchestrated, synchronized event. Their origins stem from the same primitive blastocyst that gives rise to 2 separate cell layers: epiblast and hypoblast. The epiblast then gives rise to the ectoderm, mesoderm, and endoderm. The mesoderm then becomes segmented into somites, which are precursors to the bony vertebral column and associated musculoskeletal tissues. The neural groove, which develops in the ectoderm cell layer, folds around itself to form the neural tube. Neural tube closure progresses rostrally to caudally and is thought to correlate with the cranial to caudal development of the somites.

**Developmental Ascension of the Conus**

Multiple studies in the embryology literature suggest that the conus ascends as gestational development progresses. This ascension is thought to occur early in development, with numerous authors estimating it to take place prior to 30 weeks’ gestation.
An ultrasonography evaluation during the prenatal period to the time of birth revealed that the conus ascends from approximately the L-4 level at the 13th week of gestation to rostral to L-2 at 40 weeks’ gestation. A second study further isolated the timing of the ascent to be between the 30th and 40th weeks of gestation, with 77% of babies between the 30th and 39th weeks having a conus medullaris between L-2 and L-4, compared with 84% of babies at 40 weeks’ gestation. A postmortem study focused on the location of the conus of the fetal spine showed the level of the conus to be between L1–2 and L2–3 in fetuses between the 30th and 49th weeks having a conus between the 30th and 40th weeks of gestation, with 77% of study further isolated the timing of the ascent to be between the second and third trimesters. The youngest patient in our study was 1 month old. We did not observe any significant difference between the levels of the conus with respect to age.

**Level of the Conus and Low-Lying Conus**

Cadaveric studies dating back to 1894 document the level of the conus between L-1 and L-2. Sonographic and myelographic studies verified this older data. Magnetic resonance imaging has become the modality of choice to assess spinal pathology in both the pediatric and adult populations. A retrospective MR imaging analysis of 639 patients with back pain without congenital anomalies of the spine documented the level of the conus between T-12 and L-1, with the lowest level at the upper portion of L-3 in 2 male patients (0.31% of population). A study of 504 adults who had undergone lumbar MR imaging for the evaluation of low-back pain showed the mean conus position to be at the lower third of L-1 with a range between T-12 and L-3. A low conus was seen at L-3 in 1 patient (0.20% of population). Note, however, that patient selection in these excellent studies was slightly flawed. It is possible that the patients with low-lying spinal cords on MR imaging were also clinically symptomatic. Wilson and Prince won the John Caffey Award for their work aimed at determining the level of the conus in a normal pediatric population by using MR imaging. They performed a retrospective analysis of MR images from 184 children ranging in age from newborn to 20 years. The average level of the conus among all age groups was between L-1 and L-2. The range extended from T-12 to L-3. Patients with proven tethered cords had an average level of termination from L-4 to L-5 and ranged from L-3 to S-4. A well-formulated study by Kesler and associates revealed that the conus terminated between T-12 and the middle of L-2 in 100 normal children without spinal symptomatology or congenital malformation. Not one child’s conus extended below L-2, in contrast to findings in older studies noted above. Kesler et al. firmly asserted that any conus terminating below the midportion of L-2 should be considered tethered.

A low-lying conus has historically been associated with tethered cord syndrome, which encompasses a clinical scenario characterized by back pain, lower-extremity paresthesias, motor weakness, and bowel or bladder difficulty with associated radiographic abnormalities such as the presence of an intradural lipoma, myelomeningocele, meningocele, syringomyelia, neuroparalytic tract, diastematomyelia, and a low-lying conus. However, a low-lying conus is not mandatory for the development of tethered cord syndrome, as was illustrated in Warder and Oakes’ study in 73 patients, 13 of whom exhibited characteristic symptomatology with a normal-lying conus.

Therefore, the radiographic evaluation and clinical presentation of a patient must be considered when discerning the diagnosis of low-lying conus and/or tethered cord. Data in our study suggest that the level of the conus remains within the same defined range in persons with skeletal dysplasia as in the normal adult and pediatric populations. No significant difference was observed between the different types of skeletal dysplasia. However, 2 (1.7%) of 111 of the children with skeletal dysplasia had either a low-lying conus or clinical symptoms that led us to consider a tethered cord. Conradi-Hünermann syndrome was diagnosed in 1 patient and diastrophic dystrophy in the other. The incidence of tethered cord in either of these 2 types of skeletal dysplasia has not been reported in the literature. The 1.7% incidence of tethered cord is higher in this population of children when compared with normal population studies.

**Conclusions**

Skeletal dysplasia accounts for a variety of disorders manifested predominantly by short stature. The level of the conus has not been previously elucidated in this unique population with skeletal dysplasia. The results of this study verified that the mean level of the conus was at L-1, which correlates well with published literature on the level of the conus in normal adult and pediatric populations. Spinal cord tethering was observed in 2 patients—based on clinical symptoms in 1 patient and radiographic imaging in the other. This finding may reflect a slight increase in low-lying coni and tethered cords in this population. Any physician taking care of a child with skeletal dysplasia should be diligent in evaluating the radiographs and subtle findings of tethered cord.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: JW Campbell, G Bajelidze, MC Assis, WG Mackenzie, AM Ritter. Acquisition of data: JW Campbell, G Bajelidze, MC Assis, WG Mackenzie, AM Ritter. Analysis and interpretation of data: DM Sasaki-Adams, AM Ritter. Drafting the article: DM Sasaki-Adams. Critical revision of the article: DM Sasaki-Adams. Reviewed final version of the manuscript and approved it for submission: DM Sasaki-Adams, JW Campbell, G Bajelidze, MC Assis, WG Mackenzie, AM Ritter.

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