Craniocervical decompression for cervicomedullary compression in pediatric patients with achondroplasia

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The congenital osseous abnormalities associated with achondroplasia include stenosis of the foramen magnum and the upper cervical spinal canal. In the pediatric achondroplastic patient, such stenosis may lead to cervicomedullary compression with serious sequelae, including paresis, hypertonia, delayed motor milestones, and respiratory compromise. Using a standardized protocol the authors have treated 15 young achondroplastic patients with documented cervicomedullary compression by craniocervical decompression and duroplasty. Following this procedure, significant improvement in presenting neurological or respiratory complaints was noted in all patients. The mortality rate in this series was zero. The major cause of morbidity associated with this procedure was perioperative cerebrospinal fluid (CSF) leakage from the surgical wound, presumably related to coexisting abnormalities of CSF dynamics. This problem was successfully managed by temporary or, when necessary, permanent CSF diversion procedures. It is concluded that craniocervical decompression is an effective and safe treatment for young achondroplastic patients with cervicomedullary compression.

KEY WORDS □9 achondroplasia □9 spinal compression □9 craniocervical decompression □9 hydrocephalus □9 children

ACHONDROPLASIA is an autosomal-dominant condition characterized by dwarfism, macrocephaly, rhizomelic shortening of the extremities, and other skeletal abnormalities, all resulting from a defect in endochondral bone formation.16 Neurological dysfunction is frequent in achondroplasia, resulting from compression of the neuraxis at several distinct levels.4,5,12,18 In adult achondroplastic patients, spinal stenosis (due primarily to spondylosis in the setting of a congenitally narrowed spinal canal) has become well recognized and its treatment has been described by others.4,5,12,19,20 In the pediatric achondroplastic population, cervicomedullary compression secondary to osseous and ligamentous abnormalities of the foramen magnum and upper cervical spine has been increasingly recognized as a cause of morbidity and mortality.1,6,8,13,17,18,25

The clinical manifestations of cervicomedullary compression in these patients may be subtle and are often overlooked or mistakenly attributed to coexistent congenital abnormalities. Furthermore, these coexistent malformations often complicate the surgical and anesthetic management of these patients. For this reason, the diagnosis and treatment of clinically significant cervicomedullary compression in this population are not straightforward. Over the period from 1984 to 1989, 15 pediatric patients with achondroplasia and cervicomedullary compression were treated surgically at our institution. Our experience with the management of this difficult group of patients is presented.

Clinical Material and Methods

The records of all pediatric patients with heterozygous achondroplasia who were admitted to the Johns Hopkins Hospital for clinical evaluation between 1984 and 1989 were reviewed. Of this group of 58 patients, 15 had undergone craniocervical decompression for cervicomedullary compression. Comprehensive chart reviews were performed for these 15 patients.

Clinical Evaluation

The clinical evaluation protocol used was standardized for all patients and has been described previously.18 In brief, patients were evaluated by a multidisciplinary team including neurosurgeons, pediatric neurologists,
pulmonary and sleep specialists, geneticists, anesthesiologists, neuroradiologists, and otolaryngologists. Evaluation in all patients included a detailed neurological and general physical examination as well as assessment of the adequacy of the foramen magnum and upper cervical spine by intrathecaly enhanced computerized tomography (CT) or magnetic resonance (MR) imaging techniques. Previous authors have defined criteria for the radiographic diagnosis of craniocervical stenosis in young patients with achondroplasia. Patients who underwent decompressive surgery fitting those criteria. In some patients, axial CT scans or MR images of the head were available and were evaluated for the presence of bifornal ventriculomegaly based upon measurement of the bifrontal ventricular size index. Other studies, including cineangiography, pulmonary function tests, polysomnography, electromyography, nerve conduction tests, somatosensory evoked potential monitoring, and electromyography, were performed as indicated to clarify the diagnosis in selected patients.

Diagnostic Criteria

Uniform criteria were applied for the diagnosis of cervicomedullary compression. It should be emphasized that the diagnosis of craniocervical stenosis is purely radiographic and may not imply clinically relevant cervicomedullary compression; however, the diagnosis of cervicomedullary compression relies on both radiographic and clinical factors. The criteria used to establish the diagnosis of cervicomedullary compression included: 1) the presence of signs or symptoms suggestive of ongoing brain-stem compression (such as apnea, lower cranial nerve palsies, hyperreflexia or hypertonia, paresis, or sustained clonus); and 2) radiographic evidence of foramen magnum or upper cervical canal stenosis with or without compression of the neuraxis. In addition, whenever possible the tests described above were used to exclude other possible etiologies of respiratory or neurological dysfunction (such as obstructive respiratory difficulty secondary to oropharyngeal hypoplasia). Because hypotonia and delayed motor milestones are present in nearly all young achondroplastic patients, these features were not useful in distinguishing those achondroplastic patients with clinically significant cervicomedullary compression.

Surgical Procedure

All 15 patients diagnosed as having clinically significant cervicomedullary compression underwent foramen magnum decompression, duroplasty, and upper cervical laminectomy. The surgical procedure has been described previously. Because of several instances of cerebrospinal fluid (CSF) leakage from the surgical wound (see Discussion), the original procedure was modified after the first six patients to include a ventriculostomy prior to the foramen magnum decompression. The ventriculostomy served to divert CSF flow and protect the duroplasty until adequate wound healing could occur. Preoperatively, an external ventricular drain (EVD) was inserted via a right frontal approach into the anterior horn of the right lateral ventricle. The EVD was left in place (closed) throughout the subsequent surgical procedure, and was opened immediately postoperatively to continuous drainage at an intracranial pressure (ICP) of greater than 10 cm H2O. The EVD was removed on the 2nd or 3rd postoperative day in all cases.

For the craniocervical decompression itself, patients were positioned prone on the operating table with the head and neck carefully supported in slight flexion using a padded pediatric horseshoe headrest. Upper-extremity somatosensory evoked potentials were assessed routinely during positioning as well as during the decompression procedure itself. A midline suboccipital incision was made and subperiosteal dissection performed to expose the occiput and the spinous processes of C-1 and C-2. The arch of C-1 was then removed using a high-speed drill and small curettes. In several patients, compression of the cervical cord necessitated the removal of the arch of C-2 as well. Subsequently, the posterior rim of the foramen magnum was removed with a high-speed drill and small, straight and angled curettes. Invariably, the bone rim of the posterior foramen magnum was greatly thickened and oriented more horizontally than usual, severely indenting the underlying theca. Once the bone decompression was complete, an abnormally thick fibrous band or pannus was often observed at the level of the foramen magnum. Duroplasty was performed by incising the pannus and opening the dura in the midline along the area of constriction. Adequate cord pulsations and CSF flow were confirmed, and a dural patch graft was performed utilizing paraspinal fascia or commercially available human cadaveric dura. The wound was then closed in several layers and, once movement was confirmed in all four extremities, the patients were sent to the pediatric intensive care unit. Extubation was often performed immediately postoperatively; however, in some cases, facial and laryngeal edema delayed extubation for 12 to 24 hours.

Postoperative Evaluation

Postoperatively, patients were observed carefully and evaluated for improvement or resolution of presenting signs and symptoms. Serial neurological examinations were performed prior to discharge, and follow-up studies were scheduled as needed. Patients were discharged when stable medically and when all wounds appeared healed.

Pediatric follow-up data were obtained for all patients, either by return clinic visits or by communication with the referring physician. Resolution or persistence of presenting signs and symptoms was determined, and neurological status assessed.

Results

Mean age of these patients at the time of evaluation at our institution was 3.5 years (range 8 months to 18
TABLE 1
Summary of clinical characteristics*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Presenting Sx</th>
<th>Preop Duration of Sx (yrs)</th>
<th>Ventriculomegaly</th>
<th>Surgery</th>
<th>Complications</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5, F</td>
<td>sleep apnea, rt hemiparesis</td>
<td>3</td>
<td>no</td>
<td>CCD</td>
<td>none</td>
<td>hemiparesis improved, apnea gone post-tracheotomy</td>
</tr>
<tr>
<td>2</td>
<td>1.2, M</td>
<td>apnea &amp; cyanotic episodes</td>
<td>0.66</td>
<td>yes</td>
<td>CCD</td>
<td>none</td>
<td>apnea gone then returned → 2nd surgery</td>
</tr>
<tr>
<td>3</td>
<td>3, M</td>
<td>apnea, hyperreflexia, quadriparesis, shunted hydrocephalus</td>
<td>1</td>
<td>no (previous shunt)</td>
<td>CCD</td>
<td>none</td>
<td>↑ strength, apnea gone post-T + A</td>
</tr>
<tr>
<td>4</td>
<td>2, F</td>
<td>quadriparesis, hyperreflexia</td>
<td>0.66</td>
<td>no</td>
<td>CCD</td>
<td>none</td>
<td>↑ LE strength</td>
</tr>
<tr>
<td>5</td>
<td>2, F</td>
<td>apnea, respiratory arrest × 2, lower-extremity paresis</td>
<td>0.5</td>
<td>yes</td>
<td>CCD</td>
<td>CSF leak → shunt</td>
<td>apnea gone, normal LE strength</td>
</tr>
<tr>
<td>6</td>
<td>2.5, M</td>
<td>quadriparesis, hyperreflexia</td>
<td>1</td>
<td>no</td>
<td>CCD</td>
<td>CSF leak → meningitis</td>
<td>no change</td>
</tr>
<tr>
<td>7</td>
<td>2, M</td>
<td>apnea unrelied by tracheotomy, delayed motor milestones</td>
<td>0.5</td>
<td>no</td>
<td>CCD with EVD</td>
<td>meningitis</td>
<td>apnea gone, ↑ LE strength</td>
</tr>
<tr>
<td>8</td>
<td>4, F</td>
<td>quadriparesis, gait disturbance</td>
<td>1</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>subdural hygroma → shunt</td>
<td>↑ gait &amp; LE strength</td>
</tr>
<tr>
<td>9</td>
<td>1, M</td>
<td>apnea, respiratory arrest, hypertonia</td>
<td>0.25</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>none</td>
<td>apnea gone, ↑ tone</td>
</tr>
<tr>
<td>10</td>
<td>9, M</td>
<td>paraparesis, spasticity</td>
<td>4</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>none</td>
<td>↑ gait &amp; LE strength</td>
</tr>
<tr>
<td>11</td>
<td>18, F</td>
<td>spasticity, hyperreflexia, severe gait disturbance</td>
<td>6</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>none</td>
<td>↑ strength in UE &amp; LE, ↑ gait</td>
</tr>
<tr>
<td>12</td>
<td>1.25, M</td>
<td>spasticity, delayed motor milestones, hyperreflexia</td>
<td>1</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>CSF leak → shunt</td>
<td>attaining milestones, hypertonia</td>
</tr>
<tr>
<td>13</td>
<td>1.25, M</td>
<td>hypertonia, delayed motor milestones</td>
<td>0.25</td>
<td>no</td>
<td>CCD with EVD</td>
<td>none</td>
<td>attaining milestones, walking</td>
</tr>
<tr>
<td>14</td>
<td>0.66, M</td>
<td>apnea, cyanotic episodes</td>
<td>0.33</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>CSF leak → shunt</td>
<td>apnea gone</td>
</tr>
<tr>
<td>15</td>
<td>9, F</td>
<td>apnea, paraparesis, hyperreflexia</td>
<td>6</td>
<td>yes</td>
<td>CCD with EVD</td>
<td>none</td>
<td>apnea gone, ↑ strength &amp; reflexes</td>
</tr>
</tbody>
</table>

* Sx = symptoms; CCD = craniocervical decompression; EVD = external ventricular drain; CSF = cerebrospinal fluid; T + A = tonsillectomy and adenoidectomy; LE = lower extremity; UE = upper extremity. --- = not measured; → = leading to; ↑ = improved.

years). However, the majority of patients had been symptomatic for a significant period before initial evaluation. The mean duration of symptoms prior to evaluation was 1.6 years (range 3 months to 6 years). Of the 15 patients, six were female and nine male.

Clinical Characteristics

The clinical characteristics of this patient group are presented in Table 1. The most frequent presenting clinical features were upper- or lower-extremity paresis (10 patients), apnea or cyanosis (eight patients), hyperreflexia or hypertonia (eight patients), and general delay in motor milestones even beyond that normally expected with achondroplasia (three patients). Many patients had more than one presenting problem: the constellation of mono- and paraparesis with hyperreflexia and episodic cyanosis or apnea was particularly common. Such patients presented a striking contrast to the usual floppy, hypotonic achondroplastic infant. In all eight patients with a history of cyanosis or apneic episodes, polysomnography was performed. Three patients were found to have primarily obstructive apnea, three had elements of both central and obstructive apnea, and in two patients no abnormalities were detected.

Radiographic Findings

Radiographic investigations typically revealed a small, misshapen foramen magnum. Frequently, measurements of the coronal and mid-sagittal foramen magnum diameter on axial CT scans or MR images demonstrated dimensions more than 3 standard deviations smaller than those of age-matched normal controls. Effacement of subarachnoid spaces surrounding the brain stem and upper cervical spinal cord and abnormal protuberance of the odontoid and posterior rim of the foramen magnum were also observed. In several instances, indentation or compression of the neuraxis was visualized (Fig. 1 upper pair).

Measurements of the bifrontal ventricular size index on CT scans and MR images revealed ventriculomegaly in nine of 12 patients for whom these studies were
available. However, none of these patients had preoperative clinical evidence of elevated ICP. Four patients did develop evidence of increased ICP postoperatively (see below) and eventually required permanent CSF diversion. One patient had undergone shunting for symptomatic hydrocephalus several years before developing signs of cervicomedullary compression. A number of patients had radiographic evidence of ventriculomegaly but did not develop symptomatic hydrocephalus postoperatively; thus, in this series there was no obvious correlation between ventriculomegaly and the eventual need for a shunt.

Surgical Results and Complications

The mortality rate in this series was zero. The major morbidity of this procedure included a high incidence of CSF leakage from surgical wounds (four patients). In one patient, this leakage was controlled by local measures (oversewing the wound and compressive/occlusive dressings). In the remaining three patients, ventriculoperitoneal (VP) shunting was eventually required to control the hydrocephalus that was responsible for continued leakage.

After the first six patients in the series had been treated and the problem of CSF leakage from surgical wounds was identified (two of these patients developed leakage from the suboccipital wound and one required a shunt), the procedure was modified to include controlled perioperative CSF diversion via an EVD. The subsequently treated nine patients underwent both foramen magnum decompression and ventricular drainage via an EVD. The EVD protected the suboccipital suture line well: no patient treated with perioperative CSF diversion developed CSF leakage from the suboccipital wound. Interestingly, however, two patients in this later group developed CSF leakage from the site of the EVD itself. Both of these patients eventually required CSF shunting procedures to control this problem. Intracranial pressure was monitored while the EVD was in place. Periodic resting ICP spikes to 30 to
Cranio cervical decompression in achondroplasia

50 mm Hg were noted in several patients without apparent Valsalva maneuvers or stimulation; however, plateau elevations of ICP were uncommon. While the small sample size did not permit correlation of ICP readings with the eventual need for ventricular shunting, relatively high ICP values and more frequent ICP spikes were observed in one patient who eventually went on to require a shunt.

Two patients developed postoperative Gram-negative meningitis (one with and one without demonstrated CSF leakage from the wound); this condition was successfully controlled with antibiotics in both cases. A single patient developed symptomatic bilateral subdural hygromas subsequent to VP shunt placement; these resolved after switching to a higher-pressure shunt system and shunting the subdural space.

Clinical Follow-Up Review

The mean follow-up period was 19.1 months (range 2 months to 5 years). One patient was lost to follow-up review after the first postoperative visit and is excluded from analysis. At the last follow-up contact, no patient had suffered clinical deterioration following surgery. Improvement or resolution of presenting signs or symptoms was noted in all 14 patients with follow-up data. A breakdown of surgical results by presenting symptom reveals that nine of 10 patients who presented with paresis showed significant improvement in strength postoperatively, some returning to normal. Three children who presented with delayed motor milestones but no other neurological deficits began to attain milestones rapidly following surgery. Six of the eight patients with hyperreflexia or hypertonia demonstrated resolution of these signs postoperatively. Finally, of the eight patients who presented with cyanosis or apnea, six had total resolution of these respiratory problems by the time of last follow-up, whereas two continued to have episodes of cyanosis or apnea even after foramen magnum decompression; these two children were discovered to have an additional component of obstructive apnea which in both cases resolved following surgery directed toward the cause of the obstruction. One patient did well initially, only to develop recurrence of apnea some 3 years following surgery. Subsequent evaluation revealed incomplete decompression of the foramen magnum; repeat cranio cervical decompression resulted in total resolution of symptoms.

Radiographic Follow-Up Findings

In several patients, follow-up radiological studies were performed to evaluate the adequacy of decompression. These studies typically showed a wide decompression with no residual cranio cervical stenosis or brain-stem compression (Fig. 1 lower pair).

Discussion

The neurological sequelae of compression of the neuraxis in achondroplastic dwarfs have been well described. As early as 1952, Spillane reported three cases of achondroplastic dwarfism with lower-extremity paralysis secondary to spinal cord stenosis. Many subsequent reports have served to define both the condition and its surgical treatment by decompressive laminectomy.

Cervicomedullary Compression

A more dangerous and, until recently, relatively unrecognized form of compression of the neuraxis in achondroplastic patients is compression of the brain stem and upper cervical cord at the level of the foramen magnum. This condition has gained increasing attention as a cause of respiratory and neurological impairment in the pediatric achondroplastic population. Several reports have served to define both the condition and its surgical treatment by decompressive laminectomy. The natural history of this condition has not been systematically studied; however, anecdotal clinical and pathological evidence suggests that the condition may be progressive and frequently lethal. Several studies have shown an increased incidence of sudden death in achondroplastic dwarfs under 4 years of age. In these studies, the increased mortality in this age group has been attributed directly to acute compression of the brain stem and upper cervical cord. Hecht, Reid, et al., calculated a 7.5% risk of sudden death in achondroplastic patients during the 1st year of life, a risk calculated to be at least three times greater than that of the general population.

Cranio cervical Decompression

Given this relatively bleak picture, several centers have performed cranio cervical decompression on achondroplastic infants with cervicomedullary compression, generally obtaining good results. Frequent screening of young patients with achondroplasia for respiratory and neurological impairment and performing early decompression in cases of proven cervicomedullary compression have therefore been recommended. Unfortunately, the small numbers of surgical patients in these studies and the lack of follow-up data over a longer period have not allowed a more definitive assessment of the efficacy of this treatment in individuals with homozygous achondroplasia. Evidence supporting surgical intervention in patients with homozygous achondroplasia is even scantier. The present study, although necessarily limited by its retrospective nature, provides data by which to evaluate the efficacy of cranio cervical decompression in young achondroplastic patients with cervicomedullary compression.

As our results indicate, decompression of the cervicomedullary junction in these patients will often bring about a dramatic and sustained improvement in neurological and respiratory function. No patient in our
Respiratory Compromise

A previous study has reported a 35% incidence of respiratory difficulties in pediatric achondroplastic patients. In some of these patients, respiratory abnormalities were not suspected by referring physicians or parents, and were only detected after workup for unrelated problems. Our results confirm that respiratory dysfunction is a common problem in pediatric achondroplastic patients, and one which may indicate ongoing brainstem compression.

In achondroplastic patients, respiratory compromise may be caused by any of several different mechanisms. Purely obstructive etiologies, including upper airway obstruction secondary to micrognathia, facial and laryngeal hypoplasia, and hypotonia of laryngeal musculature, are common. Restrictive pulmonary disease caused by thoracic cage and chest wall deformities may also contribute to respiratory compromise. Compression of brain-stem respiratory centers by a small foramen magnum can interfere with central respiratory drive, causing central apnea. In addition, compression of the brain stem and upper cervical cord may directly compress the lower motor neuron systems controlling respiratory muscles, resulting in weak, ineffective respirations. Because primary and neurogenic causes of respiratory dysfunction may coexist in the same patient, differentiation between these forms of respiratory compromise is important but often difficult. Patients with primarily obstructive or restrictive pulmonary disease would not be expected to benefit from craniofacial decompression. Conversely, apnea of central origin would not be improved by efforts directed toward a coexistent but minor obstructive component. One of our patients had undergone tracheostomy prior to craniofacial decompression without relief of respiratory difficulties. Two of our patients with cervicomedullary compression required tracheostomy or tonsillectomy and adenoidectomy subsequent to craniofacial decompression because of concomitant obstructive respiratory compromise. Thus, treatment of both obstructive and central apnea may be necessary in this group of patients. Our experience suggests that the coordinated efforts of experienced pediatricians, neurologists, neurosurgeons, radiologists, otolaryngologists, and pulmonologists are vital in determining which achondroplastic children may benefit from foramen magnum decompression.

Motor Deficits

Although the most dangerous consequence of cervicomedullary compression is respiratory arrest via mechanisms described above, other neurological deficits may result from compression of descending and ascending white matter tracts. Hypertonia, upper- and lower-extremity weakness, and spasticity have all been described. Long-standing compression may result in irreversible spinal cord changes and a fixed deficit. For this reason, early decompression is recommended in all proven cases of cervicomedullary compression.

Pathogenesis of Foramen Magnum Stenosis

The pathogenesis of foramen magnum stenosis in achondroplastic patients appears to be related to the generalized defect in endochondral bone formation. This results in an abnormally small posterior fossa with an extremely horizontally oriented posterior rim. Typically, the posterior lip of the foramen magnum itself is thickened and indents the cervicomedullary junction. In addition, a thick band of connective tissue often constricts the theca further at the foramen magnum. Wang, et al., and Hecht, et al., have studied the CT appearance of craniofacial junction abnormalities in pediatric achondroplastic patients. A small, teardrop-shaped foramen magnum was characteristic. In the study by Wang, et al., 96% of patients evaluated had sagittal and coronal foramen magnum dimensions at least 3 standard deviation smaller than those of age-matched control individuals.

Surgical Decompression

Relief of cervicomedullary compression requires removal of both the osseous and soft-tissue constrictive elements. The use of the high-speed drill provides for rapid controlled removal of bone around the cervicomedullary junction, and should be considered an essential part of the procedure. Removal of bone alone is in general not sufficient to relieve the compression, however. In many cases the release of thick dural constricting bands was required to allow free CSF flow and pulsation around the brain stem and upper cervical cord. Since it is difficult to evaluate the degree of dural impingement upon the cervicomedullary junction by simple inspection, we recommend that the dura be opened in all cases and that patch duroplasty be performed when dural constriction exists.

Cerebrospinal Fluid Dynamics

Unfortunately, the dural opening required to achieve adequate decompression was also the cause of the major early morbidity associated with this procedure, namely CSF leakage from the surgical wound. Despite meticulous attention to the dural closure techniques, two patients developed postoperative CSF leakage from the suboccipital incision. Under ordinary circumstances a
properly closed dural suture line would be expected to be watertight; however, it is well recognized that, in achondroplastic patients, CSF dynamics are often abnormal.\textsuperscript{15,21,24} In these patients, ventriculomegaly, abnormally spacious subarachnoid spaces over the cerebral convexities, elevated ICP, and even frank hydrocephalus may be observed.\textsuperscript{15,21,24} The etiology of the abnormal CSF dynamics associated with achondroplasia is not clear; however, recent work points to stenosis of skull outlet foramina and resultant raised jugular and intracranial venous pressures as a prime causative factor.\textsuperscript{21} In the achondroplastic patient with ventriculomegaly but without clear-cut symptomatic hydrocephalus, ventricular shunting is probably best avoided as evidence exists that the condition is not progressive in most patients.\textsuperscript{3,24} Unfortunately, a significant percentage of achondroplastic patients will eventually develop symptomatic hydrocephalus and require a shunting procedure.

At least nine of the 15 patients in the present study had evidence of ventriculomegaly on CT scans or MR images, and four eventually underwent shunt placement. Venticulomegaly alone need not signify active hydrocephalus; however, it probably does indicate a common underlying abnormality of CSF dynamics which, in some patients, may progress to symptomatic hydrocephalus. The high incidence of postoperative CSF leakage from the suboccipital wound in the early cases in this series probably reflected the tendency toward elevated ICP in these patients. Undoubtedly, the inherent friability of pediatric skin and subcutaneous tissues further compounded the problem by providing a weak barrier against elevated CSF pressures. The institution of controlled perioperative CSF diversion via ventriculostomy in the later patients in this series dramatically reduced the incidence of suboccipital wound CSF leakage. In an analogous situation, shunt placement in the newborn with hydrocephalus and a repaired myelomeningocele protects the myelomenigocele wound. Development of CSF leakage from the EVD site itself in two later patients further underscores the existence of abnormal CSF dynamics in patients with achondroplasia, as we have not encountered this problem in individuals with normal CSF dynamics. In addition to CSF diversion, the ability to monitor ICP postoperatively may yield information important in determining which patients might ultimately need shunt placement. Thus, the benefits of perioperative ventriculostomy appear to outweigh the risks associated with the procedure in this group of patients.

**Conclusions**

The treatment of pediatric achondroplastic patients is challenging and requires a coordinated multidisciplinary approach. Although a condition not frequently encountered, the high incidence of compression of the neuraxis and its dangerous sequelae in this patient population mandate familiarity with the available therapies. In documented cases of cervicomedullary compression in pediatric achondroplastic patients, early surgical decompression utilizing the techniques described above appears to improve significantly the natural history of this condition.

**References**


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