Occipitocervical stabilization is performed in both adults and children for a variety of potentially life-threatening conditions. It has been described as part of the treatment regimen for congenital instability, degenerative spine disease, Down syndrome, traumatic instability, Chiari malformation Type I (CM-I), tumor involvement of the occipital condyle, instability following lateral skull-based approaches, and instability due to inflammatory or infectious sequelae. 

Signs and symptoms of craniovertebral junction (CVJ) abnormalities are varied and insidious in presentation, with the exception of acute trauma. Symptoms may result from involvement of the cervical spinal cord, brainstem, cerebellum, cervical nerve roots, lower cranial nerves, and/or vascular supply to these structures. Abnormalities of general physical appearance can alert the clinician to involvement of the CVJ in patients with concomitant congenital anomalies. For example, the presence of head tilt, abnormal facial configuration, low hairline, short neck, limited cervical range of motion, and scoliosis may alert the clinician to an underlying prob-
Surgical stabilization of the CVJ in the pediatric population presents a unique challenge. The pediatric spine is hypermobile as compared with that of adults because of ligamentous laxity, shallow and angled facets, underdeveloped spinous processes, flat contouring of the occipital condyles, and physiological anterior wedging of vertebral bodies. This hypermobility contributes to high torque and shear forces acting on the C1–2 region. Other factors that make surgery challenging include incomplete ossification of the odontoid process and C-1, a large head compared with body mass, and weak, underdeveloped cervical musculature.

As early as 1910, Pilcher described surgical fixation at the occipitocervical joint following atlantoaxial dislocation. Since then, multiple techniques ranging from bone grafting to placement of instrumentation including wires, screws, rods, and plates have been described to treat a variety of conditions. Because of anatomical complexity and procedural morbidity, improvement in surgical techniques and options are necessary. Adult cadaveric studies have demonstrated the occipital condyles (C-0) to be a feasible fixation point. Subsequently, successful demonstration of this technique has been reported in adults and in 1 pediatric patient. In this paper, we present a series of 4 pediatric patients using our described technique.

Methods

Study Sample

Between 2009 and 2012, 4 children (ranging in age from 3 to 17 years old) requiring craniocervical stabilization were operated on using a technique previously described in adults. Occipitocervical fixation from C0–C2 (n = 3) and C0–T2 (n = 1) was performed with the goal of decompression and stabilization of this region. All patients had preoperative neurological dysfunction. In deciding on the surgical approach, multiple factors were considered preoperatively, including the method and type of instrumentation, pathological origin, presence of bone or vascular abnormalities, and presence of symptomatic CM-I, including the existence of syringomyelia. If the patient had a reducible structural deformity, preoperative traction was employed.

Preoperative Procedure

The occipital condyle height, length, width, and sagittal angle were measured preoperatively to ensure successful C-0 screw placement. Two patients had preoperative traction reduction of their craniocervical deformity and halo stabilization. Cases 2 and 4 underwent 12 and 15 pounds, of halo traction, respectively, applied preoperatively to reduce spinal deformities. After undergoing general anesthesia, the patients were placed in Mayfield pins and turned prone on the operating table, while those in halos were fixed using the Mayfield halo adaptor. Neuronavigation (BrainLab), using a custom-made drill guide to provide real-time feedback from intraoperative images acquired by the SIREMOBIL Iso-C3D (Siemens), was used to ensure proper trajectory, screw length, and screw width to avoid damaging structures such as the vertebral artery and cranial nerve XII in the hypoglossal canal. After positioning each patient prone, but prior to skin incision, images were acquired to use with the BrainLab software to insert the screws.

Surgical Technique

The screw insertion technique used in our patients was developed during an adult cadaveric study. After exposing the CVJ, the occipital condyle is located rostral to the superior articular facet of the atlas (Fig. 1A–C). The condyle is approached using subperiosteal dissection beginning at the lateral border of the foramen magnum and extending laterally and inferiorly until the C0–C1 joint capsule is removed. By maintaining constant contact with bone surfaces, carefully analyzing preoperative films, and using neuronavigation, the surgeon can prevent injury to the horizontal segment of the vertebral artery coursing along the groove in the superior aspect of the posterior arch of the atlas. The entry point of the occipital condyle is usually lateral to the condylar canal (containing the occipital emissary vein) at the lateral edge of the condylar fossa just below the skull base. The entry point is superior and lateral to the usual path the vertebral artery takes while traveling around the superior articular facet of the atlas. The lateral and medial borders of the occipital condyle are palpated, and a pilot hole is drilled at the center of the condyle using a high-speed drill. The hole is then hand drilled with the custom drill guide merged with the neuronavigation software, and subsequently tapped (Fig. 1C and D).

Posterior cervical polyaxial screws and rods were used from the Ascent LE Posterior Occipital Cervico-Thoracic (POCT) system (Orthofix). Smooth-shank 3.5-mm screws ranging from 26 mm to 32 mm in length were used in the occipital condyle. To prevent screw entry into the hypoglossal canal (Fig. 1B), the caudal-cranial angulation is determined by placing the hand drill in the pilot hole with its shaft tangential to and abutting the skull base (Fig. 1D). At each step a ball-tipped probe is used to check for cortical breach. Depending on condylar, skull base, and hypoglossal canal anatomy, the overall trajectory of occipital condyle screws is usually between 10° cranial and 10° caudal to the horizontal plane. The 2 children with halos had them removed prior to awakening from anesthesia and were subsequently placed in hard cervical collars. See Table 1 for characteristics of each case.

Results

Case 1

An 8-year-old boy presented with emesis, bradycardia, exertional headaches, and decline in academic performance. Magnetic resonance imaging showed a CM-I with medullary compression and basilar impression (Fig. 2A). Surgical intervention was planned to include posterior fossa decompression, patch grafting, and C0–C2 stabilization and fusion using polyaxial screws, and local
Pediatric occipital condyle to cervical spine fixation

**Fig. 1.** Cadaveric photographs and an illustration showing placement of occipital condyle screws. The vertebral artery (a.) is highlighted in black (A) and in red (B). A ball-tipped probe is shown in the pilot hole (C), which is then hand drilled (D). A smooth-shank polyaxial screw is then placed at an angle of 20°–30° degrees from the sagittal plane (E).

**TABLE 1: Summary of cases**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>age (yrs)</td>
<td>8</td>
<td>15</td>
<td>3</td>
<td>17</td>
</tr>
<tr>
<td>preop reduction</td>
<td>no</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>estimated blood loss (ml)</td>
<td>600</td>
<td>230</td>
<td>100</td>
<td>200</td>
</tr>
<tr>
<td>screw lengths (mm)</td>
<td>C-0 (smooth-shank) 3.5 × 30</td>
<td>3.5 × 28</td>
<td>3.5 × 26</td>
<td>3.5 × 32</td>
</tr>
<tr>
<td></td>
<td>C-1 (smooth-shank) 3.5 × 26</td>
<td>3.5 × 28</td>
<td>3.5 × 20</td>
<td>3.5 × 26</td>
</tr>
<tr>
<td></td>
<td>C-2 3.5 × 10 (rt), 3.5 × 16 (lt)</td>
<td>3.5 × 24</td>
<td>3.5 × 26</td>
<td>3.5 × 26</td>
</tr>
<tr>
<td>neurological symptoms</td>
<td>preop</td>
<td>emesis, Bradycardia, Exertional headaches</td>
<td>progressive torticollis &amp; neck pain</td>
<td>Clumsiness, difficulty walking, spasticity</td>
</tr>
<tr>
<td></td>
<td>postop</td>
<td>resolved</td>
<td>neutral neck position, resolution of pain</td>
<td>significant improvement in coordination &amp; decrease in spasticity</td>
</tr>
<tr>
<td>follow-up period (mos)</td>
<td>14</td>
<td>11</td>
<td>6</td>
<td>11</td>
</tr>
</tbody>
</table>
bone graft supplemented with a small collagen sponge containing recombinant human bone morphogenetic protein (rhBMP; Medtronic Sofamor Danek) extending posterolaterally from the decorticated occipital surface to the lateral mass of C-2. Postoperative radiographs showed that the screws were well-placed (Fig. 2B and C). Computed tomography scans completed 1 year postoperatively demonstrated a solid bone fusion (Fig. 2D and E). Clinically, the patient’s symptoms have resolved.

**Case 2**

A 15-year-old girl with a past medical history of medulloblastoma and spinal metastasis was treated with posterior fossa craniotomy and cervical laminectomy for tumor resection, followed by chemotherapy and radiation therapy, and ventriculoperitoneal shunt placement, at the age of 7. She experienced several years of progressive torticollis with her neck flexed to the right and increasing neck pain. Imaging revealed cervical kyphosis (apex of C3–4), stenosis, and head tilt toward the right. Due to her prior surgical treatment, there were extensive defects in her bone anatomy extending from the occiput through the entire cervical spine (Fig. 3A–C). There was not enough bone remaining on the occiput to use for fixation. Surgical intervention was planned to include correction of deformity and stabilization of the spine in a more anatomically correct position. The patient was placed in preoperative halo traction by gradually adding weight over several days at 2-pound increments until a total of 12 pounds was achieved, at which point she demonstrated good radiographic reduction (Fig. 3D and E), while remaining neurologically intact. Halo bars were locked down, and she was taken to surgery. A C0–T2 fusion was performed using autograft supplemented with a small collagen sponge containing rhBMP. Screws were placed in the subaxial cervical spine wherever safely possible, skipping levels of grossly abnormal bone anatomy. Postoperative imaging demonstrated good reduction of her kyphotic and rotatory deformities (Fig. 3F and G). She remained neurologically intact postoperatively.

**Case 3**

A 3-year-old girl with a past medical history of epilepsy diagnosed at age 10 months presented with clumsiness, difficulty walking, spasticity, and decline in her physical and social development. Computed tomography and dynamic MRI in flexion and extension revealed platybasia, a CM-I, and instability at the CVJ (Fig. 4A–C). Surgical intervention was planned to include stabilization of the craniovertebral junction in slight extension, allowing for anatomical decompression of the cervicomedullary junction without the need for posterior fossa decompression and C-1 laminectomy. A C0–C2 fusion was performed using autograft supplemented with a small collagen sponge containing rhBMP (Fig. 4D–I). Clinically, the patient’s walking and development improved and spasticity diminished.

**Case 4**

A 17-year-old boy with Down syndrome presented with a 6-week history of progressive neck pain and 3/5 left hemiparesis. Computed tomography and MRI demonstrated compression of the cervicomedullary junction with severe spinal cord and brainstem compression, basilar invagination, and rotatory subluxation of C-1 on C-2 (Fig. 5A–D). The patient was placed in preoperative halo traction by gradually adding weight over several days until a maximum of 15 pounds was achieved, after which he regained full strength on his left side correlating with good radiographic reduction (Fig. 5E and F). The halo...
ring was locked down and he was taken to surgery. A CO-C2 fusion was performed using autograft supplemented with a small collagen sponge containing rhBMP (Fig. 5G and H). The patient remained at full strength and experienced resolution of neck pain.

**Discussion**

In this paper we describe 4 cases in which occipital condyle to cervical (and thoracic in 1 case) fusion successfully treated instability at the CVJ in the pediatric population. These are challenging cases due to the anatomical characteristics of the involved area, congenital and acquired regional abnormalities, and inherent characteristics of the pediatric spine. Individuals with instability in this area may not have normal anatomy; therefore, neuronavigation is essential to ensure safe placement of screws in the occipital condyle and cervical spine. Table 1 reflects a summary of the cases including the types of screws used in each case. There were no injuries to cranial nerve XII in the hypoglossal canal, vertebral arteries, jugular bulb, or carotid arteries. Both children placed in preoperative halo reduction/immobilization had it removed postoperatively. All patients had excellent postoperative results, including resolution/stabilization of their neurological deficits and solid bone fusion. These patients continue to be followed clinically, and none have experienced hardware failure to date.

Occipital condyle fixation has been reported in 1 pediatric patient in the literature. The authors used image guidance and placed the screws with 15° of medial angulation and 5° of cranial angulation in the sagittal plane, with the screw tip pointing toward the basion. This procedure was based on prior publications validating the technique in cadavers. Our technique, used in the pediatric cases described here, was adapted from previous work published by Frankel et al. analyzing the occipital condyles in 40 patients using CT scans, and confirming screw length, trajectory, and feasibility in 4 adult cadavers. From this analysis the authors concluded that, on average, a 3.5-mm-diameter screw, 20- to 30-mm in length, can be safely placed at an angle of 20°–33° from the sagittal plane (Fig. 1E). This was based on the overall measurements of condylar heights (mean ± SD) 10.8 ± 1.5 mm, range 8.1–15.0 mm), widths (mean 11.1 ± 1.4 mm, range 8.5–14.2 mm), lengths (20.3 ± 2.1 mm, range 15.4–24.6 mm), and angles (mean 32.8° ± 5.2°, range 20.2°–45.8°).

This analysis was subsequently followed by a case report of a 70-year-old woman with rheumatoid arthritis, basilar invagination, and atlantoaxial instability who was successfully treated with this novel technique. Furthermore, it is our opinion based on this study that caudocranial angulation should be determined by keeping the hand drill shaft tangential to and abutting the skull base. This is a key element of the technique because it prevents screw entry into the hypoglossal canal. Angulation, even as slight as 5° as suggested by others, may cause entry of the screw into the hypoglossal canal at some point in its trajectory. The use of neuronavigation is an important adjunct to the safe placement of C-O screws, but does not serve as a replacement for the careful understanding of this complex anatomy.

There are several intraoperative and postoperative complications of occipitocervical fusions, regardless of
technique. Intraoperative complications include excessive venous hemorrhage, vertebral artery injury, and dural lacerations with CSF leakage. Postoperative complications include limitations of future growth potential, hardware failure, wound infections, neurological deficit, and failure of fusion. Complication rates in the literature are varied. One study reported a 10.4% complication rate in a total patient population of 67 patients. The complications experienced were infection, vertebral artery injury, and hardware failure. Another study reported a complication rate of 15% consisting of transient dysphagia, pseudarthrosis, wound infection, worsening quadriplegia, CSF leakage, and transverse sinus injury in patients undergoing occipitocervical fusion. The authors contended that pursuit of bicortical purchase of occipital screws increases the chance of CSF leakage or dural sinus injuries.

Fig. 4. Case 3. A: Sagittal CT scan exhibiting platybasia. B and C: Flexion and extension sagittal MR images show severe kinking of the medulla when the child was in flexion. D–F: Postoperative axial CT scans showing screw placement in the occipital condyle (D), lateral mass of C-1 (E), and C-2 pedicle (F). G and H: Sagittal CT reconstructions exhibiting screw placement. I: Intraoperative neuronavigation screenshots of screw placement.
patient population is limited to 4 cases, but we did not experience any of the mentioned complications. Placing a fixation point in the occipital condyle eliminates the risk of CSF leakage, dural sinus injuries, and cerebellar hemorrhage potentially encountered when placing occipital bone screws, an important point of distinction.

Pediatric spinal surgeries have a high rate of successful fusion. In 1 series of 67 patients undergoing C1–2 transarticular screws, the authors reported a 100% fusion rate. Menezes reviewed 850 craniovertebral fusions, and reported 98% success with bone fusion alone, and 100% success using rigid instrumentation. In a recent analysis of outcomes of instrumented fusions in the pediatric cervical spine, occipitocervical fusions had a 99% fusion rate in 285 patients. In our 4 cases, we have a 100% fusion rate to date. To aid in fusion, there are multiple options available to the surgeon including autograft with iliac crest or rib graft, and rhBMP. Iliac crest and rib grafts are associated with a 25.3% and 3.7% complication rate, respectively. The complications include donor site pain, wound dehiscence, pneumonia, meralgia paresthetica, hematoma, and iliac spine fracture with iliocrest graft, and pneumonia, persistent atelectasis, and wound dehiscence with rib graft. In our patients, we used a mixture of the patient’s bone harvested during decompression and rhBMP to avoid such complications. The rhBMP was especially useful in the case in which no decompression was completed (Case 3) and in the patient with a history of radiation to the cervical spine (Case 2).

In using occipital condyle fusion techniques it is important that this type of construct should be as strong as the traditional occipital plate to cervical spine fusion. Prior cadaveric biomechanical studies in adults have determined that an occipital condyle to cervical spine fusion is as strong as an occipital bone plate in an occipitocervical fusion. Uribe et al. compared occipital condyle screw fixation to occipital plate fixation, analyzing range of motion and stiffness in a cadaveric model, and demonstrated no statistically significant difference between the two techniques. These authors hypothesized that the longer screws in the occipital condyle improved the pull-out strength of the construct when compared with shorter screws used in the occipital plate. It is unknown, however, if the same model can predict equivalence between these two techniques in the pediatric spine. Performing this type of analysis would be exceedingly difficult in terms of specimen procurement.

The advantages of using the occipital condyle screw fixation compared with occipital bone screw fixation are multiple. For example, in patients with CM-I (Case 1) or those requiring extensive foramen magnum decompression for other reasons (Case 2), fixation and fusion can still be performed safely. Alternatively, as in Case 3, the spine can be stabilized in slight extension, thus allowing for anatomical decompression of the cervicomedullary junction without the need for posterior fossa decompression and C-1 laminectomy. Further advantages include the lack of a requirement for placing a bulky plate on the occipital bone; therefore, the operative exposure and blood loss are reduced, which may have healing implications in the thin pediatric tissue found in young patients (Case 3). Condyle screws eliminate the need to bend a rod into abnormal configurations to articulate with the obtuse angles required to engage the occipital plate (as in Case 4). This results in a lower-profile construct with less chance of hardware-related wound breakdown and potentially shorter operative times.

The majority of the scientific work pertaining to occipital condyle fusion has been in the adult population, with pediatric experience limited to 1 case report. Pediatric anatomical studies must be completed to better characterize the anatomy at the CVJ in the immature spine. Ideally, cadaveric studies could be conducted to confirm those studies, but cadaveric pediatric specimens are not practical to obtain. Radiological reviews are the next best option to study the pediatric bone anatomy in this region. Although prior studies report that there is no increased risk of developing a spinal deformity after undergoing occipitocervical or C1–2 fusions, it is imperative that we follow these patients into the future to determine how the fusions affect their growth and development. We will

Fig. 5. Case 4. A–D: Preoperative sagittal (A) and coronal (B) CT scans, and sagittal MR images (C and D). E and F: Sagittal CT scans obtained before (E) and after (F) traction reduction of 15 pounds. G and H: Postoperative anteroposterior and lateral radiographs.
continue to use this technique for stabilization of the CVJ in the pediatric population when the appropriate patient need arises.

Conclusions

In this study we have shown that the use of an occipital condyle screw is a useful option to have in the surgical repertoire for stabilization at the CVJ in the pediatric population. It allows simultaneous foramen magnum decompression (fusion of the occiput to the cervical spine at the same time as decompression of the medulla and cervical spinal cord), and it allows surgical fixation to occur in patients who have previously undergone posterior fossa surgery. The use of neuronavigation allows for accurate placement of screws in the small targets present in the pediatric spine. Whereas the highest degree of accuracy of neuronavigation would be relative to the occipital condyle because the reference array is affixed to the head frame, we have been satisfied with the guidance provided for placement of screws in the atlas and axis as well. We have seen positive fusion results in the patients thus far, and they continue to be followed clinically as time elapses from the original surgery dates. Using the condyles as a point of fixation avoids a large plate on the occiput, thus allowing a lower profile construct, less rod bending, and lower likelihood of CSF leak, dural sinus laceration, and intracranial hemorrhage.

Disclosure

Dr. Frankel is a patent holder for Hitachi Aloka Ultrasound, Zimmer Spine, and Orthofix; has received support of non–study related clinical or research effort from NIH/RO1 grants; and serves as a consultant to Orthofix.

Author contributions to the study and manuscript preparation include the following. Concept and design: Glazier, Frankel. Acquisition of data: Kosnik-Infinger. Drafting the article: Kosnik-Infinger. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Glazier. Administrative/technical/material support: Kosnik-Infinger. Study supervision: Glazier, Frankel.

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