Occipitocervical instability is a rare condition that can lead to life-threatening complications. It can result from trauma, surgery for posterior fossa decompression, or chronic pathological processes including genetic disorders with inherent ligamentous laxity, rheumatoid arthritis, infections, tumors, and congenital malformations.23,27,37,40 The central role of internal fixation with instrumentation in occipitocervical fusion has clearly been established.9,13 Several surgical strategies have been implemented to achieve this goal.23,27,37 Early techniques involving simple posterior onlay bone grafts demonstrated a high rate of failure and have largely been replaced by rigid posterior fixation systems using rods, screws, or plates, providing superior biomechanical stability and higher fusion rates.36 Plate and screw constructs have been shown to maximize resistance to flexion, axial compression, and shear stress when compared with wire and rod combinations.13,17

Fixation can be particularly difficult in children given their small size, the paucity of suitable autologous bone, and an inadequate match between available hardware and the size and shape needed for this age group. In addition, there may be situations in which the occipital bone surface is inadequate. A common problem faced by pediatric neurosurgeons is the need to perform fusion after posterior fossa decompression in a patient. Transarticular occipitoatlantal screw fixation has been studied as an alternative in these situations.4,5,10,12 Moreover, direct application of screws through the occipital condyles has been reported concurrently in 2 cadaveric studies and in 1 adult patient with pseudarthrosis of a Type II odontoid fracture.21,35,36 The feasibility of this method has been established in these studies, and its risks have been delineated. In the current case we describe the use of occipital condyle screws in a child undergoing occipitocervical fixation. To the best of our knowledge this is the first report of this technique being used in a pediatric patient.

In cadaveric studies and recently in one adult patient the occipital condyle has been studied as an option to allow bone purchase by fixation devices. In the current case the authors describe the use of occipital condyle screws in a child undergoing occipitocervical fusion. To the best of the authors’ knowledge this is the first reported instance of this technique in a pediatric patient.

This girl had a history of posterior fossa decompression for Chiari malformation Type I when she was 22 months of age. When she was 6 years old she presented with neck pain on flexion and extension of her head. Magnetic resonance imaging in flexion and extension revealed occipitocervical instability. She underwent an occiput to C-2 posterior arthrodesis with bilateral screw placement in the occipital condyles, C-2 lamina, and C-1 lateral masses. Postoperatively, she was neurologically intact. Computed tomography demonstrated a stable construct, and her cervical pain had resolved on follow-up. (DOI: 10.3171/2010.4.PEDS09551)

Key Words • Chiari malformation Type I • occipital condyle screw • atlantooccipital instability
Case Report

History and Examination. A 6-year-old girl, whose history was remarkable for suboccipital craniectomy including C-1 and limited medial C-2 laminectomy for Chiari malformation Type I at 22 months of age (Fig. 1), was noted to have increased neck and head pain on flexion and extension of her neck for about 2 years. Her initial presentation earlier in life had been for motor delays and mild weakness of the left side—the arm more than the leg—without myelopathy. The left-sided weakness manifested as a preferential use of the right arm and hand, decreased left arm swing, and a mildly asymmetric gait. Magnetic resonance imaging at that time showed marked cerebellar tonsillar descent to C-2 with mild retroflexion of the odontoid process. Her initial surgery consisted of a 1.5-cm occipital craniectomy, C-1 and partial C-2 laminectomy, and duraplasty. She fared well postoperatively with age-appropriate motor development and resolution of the left-sided weakness and right-sided arm preference. She underwent cervical flexion and extension radiography at the age of 3 years for routine follow-up because of the surgical involvement of C-1 and C-2, and the results showed no instability. She was followed-up annually and re-presented at 6 years of age with increasing mechanical neck pain radiating to the occipital region with flexion and extension but without exertional pain, increased pain on Valsalva maneuver, or any other neurological symptoms. She was neurologically intact, with no evidence of myelopathy. She was slender in build and had scant subcutaneous fat, with a weight at the 28th percentile for her age, height at the 61st percentile, and body mass index at the 10th percentile. The rest of her clinical history and physical examination was unremarkable.

Neuroimaging Studies. She underwent serial MR imaging of the cervicomedullary junction because of concerns arising from the initial slight posterior angulation of the odontoid and decompression involving C-2. On presentation with the new symptoms, repeat imaging with flexion and extension views demonstrated instability of the cranio cervical junction with significant deformity of the lower brainstem and upper cervical cord (Fig. 2). The instability was worse on flexion (Fig. 2 left). The distance from the basion-C2 line to the ventral dura mater was seen to increase over the serial images: from 3.5 mm before the Chiari malformation decompression at 22 months old, to 7.6 mm in the neutral position at 4 years old, to 7.7 mm on extension and 12.9 mm on flexion at 6 years old. She also underwent CT studies of her head and cervical spine, which confirmed the misalignment and delineated the bone anatomy of her occipitocervical region (Fig. 3). Both scans aided in evaluating the course of the VA and delineating the venous anatomy of the condylar fossa.

Treatment. Given the instability of her occipitocervical region, persistent headaches, and the potential for future brainstem or spinal cord compression, the decision was made to proceed with occipitocervical fixation. Because of the previous posterior fossa decompression, the occiput did not provide adequate bone mass for placement of an occipital plate. Furthermore, the remaining bone appeared too thin for direct fixation, and thinness of the skin and soft tissue made low-profile hardware desirable. For these reasons, it was decided to proceed with occipital condyle screw placement.

Preoperative CT scans of the occipital skull and upper cervical spine with fiducials were obtained using pediatric protocols to minimize radiation exposure. The pediatric CT protocol involves a lower kilovolt peak (kVp) and milliampere (mA) setting to reduce the radiation dose; we use 100 kVp and approximately 125 mA. The slice thickness remains at 1.25 mm. The patient was anesthetized and placed in pins in a neutral prone position while under continuous neurophysiological monitoring using somatosensory and motor evoked potentials and brainstem evoked potentials. The upper cervical and suboccipital regions were exposed, and the occipital condyles were subsequently revealed, mobilizing the horizontal segment of the VA inferiorly. With the assistance of intraoperative image guidance and limited fluoroscopy, pilot holes were drilled in the condyles bilaterally to a depth of approximately 14 mm. The pilot hole was drilled with 15° of medial angulation and 5° of cranial angulation in the sagittal plane, with the tip pointing toward the basion. This optimal trajectory has been validated in cadaveric studies. A ball-tip probe was used in both instances to verify a bony channel. There appeared to be no cortical breaches along the walls of the channel. A smooth-shafted cervical screw measuring 26 mm in total length with a total thread length of 15 mm was then placed, with several threads remaining exposed. Overall, screw penetration within the condyle was estimated at approximately 12–13 mm based on preoperative measurements.

Screws were also placed into the C-1 lateral mass-
Placement of occipital condyle screws for occipitocervical fixation

During exposure of the condyles, the occipitocervical joint was revealed such that the synovial surfaces were clearly visualized above the VA. The dorsal aspect of the joint surfaces was then decorticated using a high-speed drill and curettes to allow bone placement for fusion. Bone that had been locally harvested from the thicker portions of the occipital bone and from drill hole placement was then packed into both of these joints (Fig. 4). A similar procedure was performed below the C-2 nerve root in the C1–2 joint space.

**Postoperative Outcome.** Postoperatively, the patient was neurologically intact and headache free, and was discharged on postoperative Day 3. Her incision healed well without problems of hardware protrusion. She underwent CT studies of the head and cervical spine, which demonstrated a distance between the basion-C2 line and the ventral dura of approximately 8.2 mm as well as appropriate positioning of the hardware (Figs. 5 and 6). At the 6-month follow-up she had good functional range of motion in her neck with complete resolution of the prior cervical pain and headache, and her neurological examination remained within normal parameters. Follow-up flexion and extension films are planned for her 8-month follow-up visit.

**Discussion**

Occipitocervical fusion was first described in 1927. It consisted of simple onlay bone graft techniques, which were supplemented in the 1970s by wire fixation. These constructs evolved with the use of rods that were fixed with wires or cables and attached to the skull via bur holes at one end and to the posterior part of the cervical spine at the other. Although the latter constructs were more stable than the ones used initially, they remained largely nonrigid and did not resist cranial settling or rotation, requiring prolonged external immobilization. Constructs using fully rigid fixation have gradually replaced previous fixation techniques. Occipitocervical plate fixation became popular, offering rigid skeletal fixation and immediate stability, and thus increasing the fusion rates and

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**Fig. 2.** Preoperative sagittal T2-weighted MR images of the head and cervical spine, flexion (left) and extension (right) views, demonstrating basilar invagination and instability of the craniocervical junction with significant deformity of the lower brainstem and upper cervical cord. The findings worsen with flexion. White dot indicates the tip of the basion; dotted line shows the plane of the hard palate. The increased laxity allows the basion to slip forward and inferior with flexion and, coupled with the retroflexion of the dens, results in compression of the medulla.

**Fig. 3.** Preoperative reconstructed sagittal (left) and 3D (right) CT of the cervical spine, demonstrating basilar invagination (left) and changes due to suboccipital craniectomy and C1–2 laminectomies (right).
decreasing the need for external immobilization. However, the plate design of these constructs forced screw placement into a suboptimal location—laterally directed on the occiput, where bone is thinner. As a result, plate systems have been gradually replaced by systems of independent occipital plates and cervical rods. These implants are rigidly secured to the midline suboccipital bone, allowing improved intraoperative contouring of the construct to the craniocervical junction.

The occipital condyle has been studied as a viable structure for fixation in cadaveric studies. Note that the location and shape of this structure can differ. It is bordered rostrally by the hypoglossal canal, caudally by the occipitocervical joint, medially by the foramen magnum, and laterally by the jugular bulb and a concave area called the “condylar fossa,” located posterior to the bulb. The condylar canal opens at the bottom of the condylar fossa. The posterior condylar emissary vein runs through the condylar canal and courses between the posterior part of the jugular tubercle, whose external surface is formed by the condylar fossa and occipital condyle. Preservation of this structure during exposure of the condyle is extremely important since it can serve as major drainage for the sinus in cases of jugular occlusion or congenital anomalies. Venography, either MR venography or CT venography, should have a central role in preoperatively identifying patients with such congenital anomalies. The condylar emissary vein can be seen more easily as it exits the posterior surface of the condylar fossa. The VA passes medially around the posterior margin of the atlantooccipital joint. The carotid artery is located 5–8 mm lateral to the condyle.

This complex anatomy and the surrounding vital structures have prevented surgeons from attempting occipital condyle screw placement. With the evolution of neuronavigation, however, transarticular C-1 to occipital condyle screw placement has been attempted in cadaveric studies. The biomechanics of this construct have shown that the condyle provides enough solid bone purchase to permit insertion of a screw vertically up into the condyle through a caudal-rostral trajectory. Authors of more recent cadaveric studies have investigated the feasibility and

Fig. 4. Postoperative axial CTs of the cervical spine demonstrating bone graft in the occiput to C-1 space (left) and the C-1 to C-2 space (right).

Fig. 5. Postoperative axial CT at the level of the occipital condyles demonstrating the position of the occipital condyle screws.

Fig. 6. Postoperative reconstructed sagittal CT of the cervical spine showing the position of the left (A) and right (B) construct from the occiput to C-2.
Placement of occipital condyle screws for occipitocervical fixation

The use of occipital condyle screws in occipitocervical fixation appears to be a feasible and safe method for the treatment of occipitocervical instability. While more traditional stabilization and fusion methods can be used in many cases, this procedure offers an additional option for surgeons. It is particularly useful in patients with prior posterior fossa decompression and therefore an inadequate amount of occipital bone for appropriate purchase and/or fusion. Despite the overall favorable profile of occipital condyle screws, further biomechanical studies are needed to compare their use with the current techniques of occipitocervical fixation.

Conclusions

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

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