Pediatric occipitocervical fixation: radiographic criteria, surgical technique, and clinical outcomes based on experience of a single surgeon

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OBJECTIVE If left untreated, occipitocervical (OC) instability may lead to serious neurological injury or death. Open internal fixation is often necessary to protect the neurovascular elements. This study reviews the etiologies for pediatric OC instability, analyzes the radiographic criteria for surgical intervention, discusses surgical fixation techniques, and evaluates long-term postoperative outcomes based on a single surgeon's experience.

METHODS The charts of all patients < 18 years old who underwent internal OC fixation conducted by the senior author were retrospectively reviewed. Forty consecutive patients were identified for analysis. Patient demographic data, OC junction pathology, radiological diagnostic tools, surgical indications, and outcomes are reported.

RESULTS The study population consisted of 20 boys and 20 girls, with a mean age of 7.3 years. Trauma (45% [n = 18]) was the most common cause of instability, followed by congenital etiologies (37.5% [n = 15]). The condyle-C1 interval had a diagnostic sensitivity of 100% for atlantooccipital dislocation. The median number of fixated segments was 5 (occiput–C4). Structural bone grafts were used in all patients. Postsurgical neurological improvement was seen in 88.2% (17/19) of patients with chronic myelopathy and in 25% (1/4) of patients with acute myelopathy. Preoperatively, 42.5% (17/40) of patients were neurologically intact and remained unchanged at last follow-up, 42.5% (17/40) had neurological improvement, 12.5% (5/40) remained unchanged, and 2.5% (1/40) deteriorated. All patients had successful fusion at 1-year follow-up. The complication rate was 7.5% (3/40), including 1 case of vertebral artery injury.

CONCLUSIONS Occipitocervical fixation is safe in children and provides immediate immobilization, with excellent survival and arthrodesis rates. Of the radiographic tools evaluated, the condyle-C1 interval was the most predictive of atlantooccipital dislocation.

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KEY WORDS fusion; long-term; occipitocervical; outcome; pediatric; spine
Pediatric occipitocervical fusion long-term outcomes

Surgical and clinical studies suggest that fixation, using occipital keel screws, occiput–C1 transarticular screws, and C1–2 transarticular screws (with good screw purchase), provides the best immobilization and fusion rates. The incorporation of structural grafts strengthens the construct and improves arthrodesis rates. However, the most appropriate fixation strategy must be tailored to each patient; surgeons should consider the underlying pathology, size of the spinal osseous structures, any anatomical variations, and extent of pathology. The size of the bone at sites of fixation is most often dictated by the patient’s age and any comorbid congenital syndrome. It is one of the most important considerations in determining a fixation strategy for the craniovertebral junction (CVJ) in children. When the bony fixation points are too small to accommodate screws, wiring techniques are most often used.

Long-term sequelae of OC internal fixation in children have been an area of concern. Adverse effects on spinal alignment, stability, and cervical spine growth have been described. Although several pediatric series on this topic have been previously published, results need to be interpreted cautiously because of relatively small sample sizes. To our knowledge, this study represents one of the largest single-surgeon case series to date describing OC fixation in the pediatric population.

Methods

The institutional review board at St. Joseph’s Hospital and Medical Center (Phoenix, AZ) approved this study. Forty-three pediatric patients with a diagnosis of OC instability underwent OC fixation performed by the senior author (N.T.) between 2004 and 2013, at Barrow Neurological Institute (Phoenix, AZ). Three patients were excluded because of missing data, which left 40 patients for inclusion in this detailed retrospective analysis.

Trauma and relevant comorbidities were documented. Plain lateral and anteroposterior cervical radiographs, CT scans, 3D CT reconstructions, and MR images were used for pre- and postoperative radiological analysis. Etiologies for OC instability included trauma, congenital abnormalities, neoplasms, vascular malformations, and multiple causes (detailed in the Results section). Patients with preoperative neurological deficits were grouped into 2 cohorts according to the time course of symptom evolution. Patients with trauma formed the acute myelopathy cohort. The remainder of the patients, who had progressive evolution of symptoms, formed the chronic injury cohort. All patients were surgically treated on the basis of clinical presentation, radiographic evidence of OC instability, and the senior author’s (N.T.) clinical judgment.

All patients were preoperatively immobilized in the neutral position as needed. When AOD was suspected, external immobilization was achieved by placing sandbags on each side of the patient’s head and securing the head and sandbags to the bed with tape. Rigid cervical collars, which are potentially harmful with distraction injuries, were avoided. Halo vest orthoses were used in some cases when immediate operative intervention was not possible.

Patients who were diagnosed as having a minor injury to the CVJ that did not require OCF were not included in our analysis. Thus, the incidence of preoperative collar use was not assessed.

Surgical techniques and types of hardware were documented, along with the type of graft used. Use of external orthoses, if required, was documented before and after surgery. In patients with traumatic injuries and suspected AOD, the condyle-C1 interval (CCI) was used to confirm the diagnosis; for those with suspected AAD, the atlantoaxial interval (ADI) was used. Sagittal and coronal reconstructions of preoperative CT scans were used to determine the CCIs (Fig. 1). Four equidistant measurements were made for each occiput–C1 joint (labeled CC1–CC4) at the point of longest contact to generate a mean CCI. A mean CCI ≥ 4 mm on either side was considered abnormal and indicative of AOD. The ADI was determined by measuring the distance from the posterior aspect of the anterior arch of C-1 to the anterior border of the dens, using either sagittal CT reconstructions or lateral radiographs. Flexion-extension radiographs were also obtained if static ADI was < 4 mm but positional instability was still suspected. Atlantoaxial instability was defined as an ADI > 4 mm, or positional change in ADI > 4 mm.

Benzel et al. modified Japanese Orthopaedic Association score (mJOAS) and Hirabayoshi et al. recovery rate were used to assess clinical outcome results in patients with chronic injury preoperatively, postoperatively, and at last follow-up. The Hirabayashi recovery rate (RR) was calculated and used as a prognostic tool and an indicator of clinical outcome, using the formula: RR = (postoperative mJOAS – preoperative mJOAS)/preoperative mJOAS) × 100.

Spinal cord injury in patients with acute myelopathy was assessed with the American Spinal Injury Association (ASIA) impairment scale (AIS) preoperatively, postoperatively, and at last follow-up. Complete injury (AIS Grade A) is defined as no sensory or motor function preserved in S4–5 below the neurological level of injury (NLI). Sensory incomplete injury (AIS Grade B) is defined as sensory function but not motor function preserved below the NLI, including S4–5 and deep anal pressure. Motor incomplete (AIS Grade C) is defined as motor function preserved below the NLI, and more than half of key muscle functions below the NLI have strength scores ≤ 2 out of 5. Motor incomplete (AIS Grade D) is defined as motor function preserved below the NLI, and at least half of key muscle functions have strength scores ≥ 3 out of 5. Normal (AIS Grade E) is defined as neurologically intact (http://www.asia-spinalinjury.org/elearning/ASIA_ISCOS_high.pdf).

Neurological monitoring during surgery with somatosensory evoked potentials and motor evoked potentials was performed in all patients when technically feasible. Baseline tracings were obtained before positioning. Structural autografts were used to provide additional structural support and increase the area of arthrodesis. Most autografts were harvested from a posterior rib; most allografts were from a cadaveric fibula. The curvature of the posterior rib graft usually mirrors the curvature of the posterior OC junction; therefore, it was preferred when suitable. Fluoroscopy was used in all cases for assistance with positioning and intraoperative confirmation of hard-
ware placement, and in some cases the O-arm Surgical Imaging System (Medtronic, Inc.) was used. Intraoperative image-guided navigation was used for surgical planning and hardware placement in most cases.

Postoperatively, all patients were placed in rigid cervical collars for a minimum of 8 weeks. External orthosis was discontinued on the basis of the quality of arthrodesis and clinical progress. Fusion was defined as osseous trabeculation throughout the instrumented OC levels evident on anteroposterior and lateral cervical spine radiographs, and on CT scans when indicated. All radiographs were reviewed by the senior author (N.T.) and by an independent neuroradiologist. Follow-up was ongoing with clinical and radiographic control. Complications were documented intraoperatively and during the early and delayed postoperative periods.

Confidence values were calculated for reported data (significant at $p < 0.05$) with t-tests (2-tailed distribution, 2-sample equal variance). Means and standard deviations are provided when appropriate.

Results

Patient Characteristics

Demographic Data

Twenty (50%) patients were boys, and 20 (50%) were girls, with a mean age of $7.3 \pm 4.4$ years (range 7 months to 16 years). The mean age of the 18 patients with acute traumatic injuries was $6.2 \pm 4.1$ years (range 2–16 years), and the mean age of the other 22 patients, all of whom had chronic pathologies, was $7.7 \pm 4.7$ years (range 7 months to 16 years) ($p = 0.3$).

Etiology of OC Instability

The etiologies of OC instability and the indications for surgical treatment are presented in Table 1. The most common cause of instability overall was trauma, which occurred in 18 (45%) patients; occipitoatlantoaxial dislocation was the most often diagnosed instability. None of the patients with trauma had any additional comorbidities (e.g., Down syndrome) at the time of injury. Congenital etiologies were the indication for surgery in 15 (37.5%) patients; Chiari Type I malformation was the most common in this subgroup. One patient with Chiari malformation also had Ehlers-Danlos syndrome (EDS). Hardware failure was the third most common indication for surgery (3 cases [7.5%]) and was associated with multiple primary diagnoses among patients referred from outside hospitals.

Other etiology categories included neoplasm, inflammatory arthropathy, vascular pathology, and multiple causes; each had 1 representative case. The patient with vascular pathology had a rare upper cervical spine meta-

![Fig. 1. A: Parasagittal CT image of the CVJ of a patient with AOD (CCI 0.59 cm). B: Midsagittal CT image of the CVJ of a patient with OC instability (ADI 0.43 cm). Coronal (C) and parasagittal (D) CT images of a patient with a normal CCI (< 4 mm).](image)
meric arteriovenous malformation (AVM) and presented for decompression, biopsy, and OC fixation. Two patients with OC instability were initially treated nonoperatively for decompression, biopsy, and OC fixation. Two patients aged surgically. No patients were treated with halo vest but subsequently required surgical immobilization after surgery.

Of the 18 patients with traumatic injuries, only 2 presented with polytrauma, which in these 2 cases did not require emergency surgery by other services before the neurosurgical procedure. After admission to the emergency department, all patients with traumatic injuries were immediately transferred to the ICU. Mean arterial pressure was maintained at > 85 mm Hg, and patients were optimized hemodynamically and metabolically and cleared for surgery within 24 hours.

Preoperative Neurological Function

Seventeen (42.5%) of the 40 patients presented as neurologically intact preoperatively—12 (70.6%) from the acute injury cohort and 5 (29.4%) from the chronic injury cohort. All 17 patients remained unchanged through the last follow-up visit; therefore, they were excluded from the analysis of neurological improvement. Patients presented preoperatively with neck pain (26 [65%]), weakness (22 [55%]), bladder and/or bowel incontinence (17 [42.5%]), numbness (12 [30%]), gait ataxia (12 [30%]), and quadriplegia (1 [2.5%]) (Table 2). Five cases with unique clinical presentations, comorbidities, and evolution are summarized in Table 3.

Two of the 18 trauma patients presented with severe traumatic brain injury (Glasgow Coma Scale score of 3), were unfit to undergo motor function testing on admission, and had no additional neurological deficit after surgery; both were AIS D at last follow-up. Preoperatively, 75% (12/16) of our patients with trauma were neurologically intact, and none had vascular or cranial nerve injuries. Although only 25% (n = 4) of patients with trauma presented with a neurological deficit, the neurological injuries that did occur were often severe; 75% (n = 3) of those with a deficit were categorized as having AIS Grade A, B, or C impairment. Unlike patients with trauma, those with chronic injury were neurologically intact preoperatively in only 22.7% of cases (5/22), probably because longer periods of myelopathy resulted in irreversible damage to the spinal cord.

Diagnostic Radiological Measurements

It was feasible to obtain ADI and CCI measurements retrospectively from 28 patients (mean age 7.2 years; range 8 months to 16 years). Atlantoaxial and occipitocervical dislocation correlated to an ADI > 4 mm in 5 of 15 (33.3%) patients with suspected C1–2 instability and a CCI > 4 mm in all 16 patients with suspected occiput–C1 instability. The mean ADI was 3.2 mm (range 1.4–6.4 mm), the mean right CCI was 3.7 mm (range 1.1–6.8 mm), and the mean left CCI was 4.0 mm (range 1.3–6.1 mm). The ADI had a sensitivity of 17.6% for atlantoaxial instability, and CCI measurements had a sensitivity of 100% for AOD.

**Occipitocervical Fusion**
Types of Instrumentation

Threaded Steinmann pins and sublaminar wiring (Atlas Cable System, Medtronic Sofamor Danek USA, Inc.)

### TABLE 2. Preoperative findings among acute and chronic injury cohorts

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Acute Injury (n = 18)</th>
<th>Chronic Injury (n = 22)</th>
<th>All (N = 40)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>Neck pain</td>
<td>11</td>
<td>61.1</td>
<td>15</td>
</tr>
<tr>
<td>Incontinence</td>
<td>1</td>
<td>5.6</td>
<td>16</td>
</tr>
<tr>
<td>Weakness (≤2 of 5)*</td>
<td>4</td>
<td>22.2</td>
<td>11</td>
</tr>
<tr>
<td>Altered reflexes</td>
<td>2</td>
<td>11.1</td>
<td>12</td>
</tr>
<tr>
<td>Numbness</td>
<td>1</td>
<td>5.6</td>
<td>11</td>
</tr>
<tr>
<td>Gait ataxia</td>
<td>1</td>
<td>5.6</td>
<td>11</td>
</tr>
<tr>
<td>Weakness (≥3 of 5)*</td>
<td>1</td>
<td>5.6</td>
<td>6</td>
</tr>
<tr>
<td>Flaccidity</td>
<td>1</td>
<td>5.6</td>
<td>0</td>
</tr>
<tr>
<td>Decreased rectal tone</td>
<td>1</td>
<td>5.6</td>
<td>0</td>
</tr>
</tbody>
</table>

* ASIA scale for strength (0 = no strength to 5 = full strength).

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**TABLE 1. Diagnosis of occipitocervical fusion**

<table>
<thead>
<tr>
<th>Detailed Diagnosis</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma</td>
<td>18 (45.0)</td>
</tr>
<tr>
<td>Occipitoatlantoaxial dislocation</td>
<td>13 (32.5)</td>
</tr>
<tr>
<td>Occipitocervical dislocation</td>
<td>3 (7.5)</td>
</tr>
<tr>
<td>Atlantoaxial dislocation</td>
<td>2 (5.0)</td>
</tr>
<tr>
<td>Congenital</td>
<td>15 (37.5)</td>
</tr>
<tr>
<td>Chiari Type I malformation</td>
<td>5 (12.5)</td>
</tr>
<tr>
<td>Multiple congenital causes</td>
<td>3 (7.5)</td>
</tr>
<tr>
<td>Morquio syndrome</td>
<td>2 (5.0)</td>
</tr>
<tr>
<td>Spondyloepiphyseal dysplasia</td>
<td>2 (5.0)</td>
</tr>
<tr>
<td>Wolf-Hirschhorn syndrome</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>VACTERL association</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Hardware failure</td>
<td>3 (7.5)</td>
</tr>
<tr>
<td>Failed arthrodesis (outside hospitals)</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Redo of Chiari decompression</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>After resection of neurofibroma (intra- &amp; extradural)</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>After resection of yolk sac tumor</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Neoplasms</td>
<td></td>
</tr>
<tr>
<td>C-2 osteoid osteoma</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Inflammatory arthropathies</td>
<td></td>
</tr>
<tr>
<td>Juvenile rheumatoid arthritis</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Vascular pathology</td>
<td></td>
</tr>
<tr>
<td>Metameric AVM in upper cervical spine</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Multiple causes</td>
<td></td>
</tr>
<tr>
<td>Chiari Type I malformation, Klippel-Feil, basilar invagination, &amp; juvenile rheumatoid arthritis</td>
<td>1 (2.5)</td>
</tr>
</tbody>
</table>

VACTERL = vertebral, anal atresia, cardiac, tracheal, esophageal, renal, and limb defects.
were used in 20 (50%) patients, screw-based constructs were used in 19 (47.5%) patients, and a sutured structural autograft was used in 1 (2.5%) 7-month-old patient. Neuro-navigation was used in all patients undergoing placement of screw-based constructs. Multiple fixation systems, such as the Synthes Synapse system (Synthes, Inc.), Synthes Axon (Synthes, Inc.), and Medtronic Vertex (Medtronic, Inc.), were used. All patients were able to undergo intraoperative neuromonitoring, and no changes were noted in any case.

Most patients included in this series presented with severe and sometimes devastating injuries to the occipital and cervical spine requiring extensive instrumentation to correct basal deformities and to improve stability. The median number of instrumented levels was 5 (from occiput to C-4 in 17 [42.5%] cases), followed by 4 levels (from occiput to C-3 in 10 [25%] cases). There were 2 occipit–C2 (5%), 1 occipit–C5 (2.5%), and 1 occipit–C6 (2.5%) constructs. Among constructs extending to the thoracic spine, there were 6 (15%) at occiput–T2 and 1 (2.5%) each at occiput–T1, occiput–T3, and occiput–T4.

The 10 patients requiring instrumentation from occiput to C-3 had injuries compromising bony or ligamentous structures extending to C-2. In 17 cases involving instrumentation placed from the occiput to C-4, 7 patients (41.2%) had injuries compromising structures extending at least to C-3. Two of the 17 (11.8%) required tumor excision, hardware removal, and repeated occipitocervical fusion (OCF), and 8 (47.1%) had severe congenital cervical abnormalities. The patient who underwent placement of occipit–C5 instrumentation had hardware removal and repeat OCF requiring extension of the instrumentation. The patient who underwent placement of occipit–C6 hardware had severe congenital cervical abnormalities.

All 9 of the patients with instrumentation extending to the thoracic spine and 2 patients with instrumentation extending to the lower cervical spine had occipitoatlantal, atlantoaxial, or occipitoatlantoaxial instability with additional abnormalities. These conditions included occipit–T2 instability with Morquio syndrome (2 patients), C5–6 and C6–7 (1 patient each) traumatic spondylolisthesis, C6–7 complete 3-column injury (1 patient), metamerism AVM obliteration and instrumentation extending from the occiput to T-2 (1 patient), multilevel intra- and extraxial tumor excision (1 patient), severe congenital cervical abnormalities (3 patients), and severe kyphosis (1 patient).

**Patients With Arthrodesis**

Structural bone grafts were used in all patients, with allografts taken from a posterior rib in 39 of these patients and a fibular allograft used in 1 patient. Morcellized autograft and/or allograft bone was placed posterolaterally over the decorticated CVJ in all patients.

**Decompression Procedures**

Decompression of the neural elements was performed in 14 (35%) patients. Procedures included laminectomies, suboccipital craniectomies, corpectomies, hardware removal, and tumor/mass resections.

**Outcome Measures**

**Clinical and Radiographic Follow-Up**

The mean clinical and radiographic follow-up was 31.9 months (range 16–119 months). Preoperatively, 42.5% (17/40) of patients were neurologically intact and remained unchanged at last follow-up, 42.5% (17/40) had neurological improvement, 12.5% (5/40) remained unchanged, and 2.5% (1/40) deteriorated. Follow-up is ongoing.

**Chronic Injury**

Neurological outcome was assessed in all patients with
chronic myelopathy by use of the mJOAS and the recovery rate. A total of 17 patients were included in the analysis (Table 4). The first clinical follow-up for all patients occurred between 2 and 8 weeks postoperatively, and the last follow-up for patients with chronic injuries occurred at a mean 28.3 ± 12 months postoperatively. Six (35.3%) patients had regained full neurological function at last follow-up. The mean mJOAS improved from 10.9 ± 4.2 (range 4–17 points) preoperatively to 14.0 ± 4.3 (range 3–18 points) postoperatively (p < 0.001), and to 13.9 ± 4.5 (range 3–18 points) at last follow-up. The recovery rate was 20.8% postoperatively and 20.6% at last follow-up (p = 0.3).

Incidence of Complications

Complications occurred in 3 patients (7.5%) (Table 5). There were 2 intraoperative complications and 2 postoperative complications.

One patient had a left vertebral artery injury during placement of a C-1 lateral mass screw, which caused a dissecting aneurysm of the vertebrobasilar junction. During this event, the patient suffered bradycardia and hypertension, which were managed promptly by the anesthesiologist. The occipitot–C4 screw-rod system was secured, precluding the placement of the contralateral C-1 screw, and the surgery was terminated on an emergency basis.

In the second intraoperative complication, the patient had a traumatic pneumothorax during rib harvest. Two months postoperatively, this patient also developed wound dehiscence, which was managed conservatively.

The second postoperative complication occurred in a third patient. This patient had a delayed complication of a dehiscent and infected wound, which required open débridement.

Arthrodesis Results

Successful fusion was confirmed in all patients in the series by use of lateral flexion-extension and anteroposterior radiographs and CT scans between 4 months and 1 year after surgery.

Acute Injury

Four patients with traumatic spinal cord injury were included in the outcome analysis of acute myelopathy. Three patients presented with incomplete spinal cord injury (1 each with AIS Grade B, C, and D), and 1 patient presented with complete spinal cord injury (AIS Grade A). The patient with AIS Grade B impairment improved 1 categorical grade postoperatively. The status of each of the remaining 3 patients was unchanged at last follow-up. Two patients with acute myelopathy had a severe traumatic brain injury, which limited complete assessment of spinal cord injury with the AIS; those patients were excluded from the analysis.

Etiology of OC Instability

Occipitocervical instability results from the disruption of normal osseous and ligamentous structures. This damage can be caused by trauma or by anatomical variations due to several underlying abnormal conditions. Trauma represented the most common cause of OC instability in this series (45% of cases). Although traumatic OC destabilization occurs in all age groups, young children (<8 years) are especially vulnerable due to hypermobility of the CVJ and increased head-to-torso size and weight ratios.

Although in the present study the mean age of patients with acute traumatic injuries (6.2 ± 4.1 years; range 2–16 years) differed from that of those with all other chronic pathologies (7.7 ± 4.7 years; range 7 months to 16 years), this difference was not statistically significant (p = 0.3). Both traumatic and congenital etiologies are frequently reported as the most common causes of OC instability in children.

Bollo et al. reported an association between posterior fossa decompression for Chiari malformations and OCF, with 6% of patients requiring OCF after decompression due to medullary kinking, odontoid process retroflexion, and basilar invagination. Some authors have proposed a relationship between EDS and OC instability, due to inherent joint hyperlaxity in patients with EDS. However, a clear association between either AOD or AAD and EDS has not yet been established. We had 1 patient with a his-
tery of Chiari malformation and EDS who developed progressive neurological deterioration and signs of myelopathy, leading to suspicion of occipitoatlantoaxial dislocation and incomplete spinal cord injury. However, the relative contributions of the Chiari malformation and EDS to the patient’s instability are unknown.

Diagnostic Radiological Measurements

Detailed imaging of the OC junction is essential in cases of suspected OC instability. The ADI measurement is a radiographic tool that is used to evaluate the integrity of the atlantodental, alar, and transverse ligaments, and, in turn, OC stability. A static ADI ≥ 4 mm is consistent with AAD and OC instability. However, even in an unstable CVJ, neutral radiographs may show a normal ADI (< 4 mm); in this setting, dynamic imaging with flexion-extension views is required to establish the diagnosis. An ADI ≥ 4 mm in any position is considered consistent with atlantoaxial instability.\(^\text{50}\)

The CCI measurement is considered an excellent tool for radiological diagnosis of AOD.\(^\text{3,41,45}\) Other diagnostic tools for AOD\(^\text{31,30,46,53,56}\) have been shown to have inferior power compared with the CCI in the pediatric population.\(^\text{31,42}\) Our results support previously published data showing that the CCI has excellent diagnostic sensitivity and negative predictive value for AOD in pediatric patients.\(^\text{41}\)

Preoperative Neurological Function

Neurological deficits in children who survive high-energy traumas to the CVJ are usually partial or absent, and may be associated with cranial nerve (most commonly abducens and hypoglossal nerves)\(^\text{1, 5, 29,37,43}\) and vertebral artery injuries.\(^\text{39,42}\) Survival rates after AOD, occipitoatlantoaxial dislocation, and AAD range from 39% to 63%.\(^\text{5,23,28}\)

Surgical Planning

Detailed imaging of the CVJ is essential for the surgical planning of OC fixation. Imaging allows for a complete understanding of the patient’s anatomy and the consideration of any anatomical variations. An understanding of the size and interrelationship of the osseous structures dictates the instrumentation strategy. For example, smaller and less ossified vertebral bodies are often unable to accommodate screws, and wiring techniques should be used. Up to 25% of patients have unfavorable vertebral artery anatomy for screw placement. In some cases, 3D models of the spine have been used to assess the appropriateness of screw placement.\(^\text{15,57}\) Intraoperative image guidance with neuronavigation is becoming more common and is especially useful in pediatric patients with diminutive anatomy. Safe placement has been reported in up to 93% of pediatric patients, even in those with anatomy that might otherwise appear to preclude screw instrumentation.\(^\text{40,47}\) We found neuronavigation especially helpful in patients with severe congenital cervical abnormalities or a small osseous anatomy, as well as for orientation and assessment of adequate screw placement. Neuronavigation can also be very helpful for determining the midline and optimal placement of keel screws. Neurophysiological monitoring also serves as an important adjunct and is recommended in all cases.\(^\text{40,47}\)

Fixation Techniques

An almost equal proportion of wire- and screw-based constructs was used in this case series (50% vs 47.5%) (Figs. 2–4). The decision to use either one was carefully determined on the basis of each individual’s presentation, age, and bony anatomy, as well as on the basis of its technical feasibility and the surgeon’s experience. The 7-month-old patient required special consideration and underwent fixation using a posterior rib structural autograft sutured to the CVJ due to the inability to place screws and sublaminar wiring.

As experience has continuously increased, the senior author now uses screw-rod and structural autograft constructs as the preferred method of fixation in appropriate patients because of their proven safety and effectiveness. In a systematic review by Hwang et al.\(^\text{25}\) of 285 patients with OCF, including 20 patients from their experience, a slight predilection was found for screw (55.1%) versus wiring constructs (44.9%). Our youngest patient with safe placement of screw-based constructs was 3 years old. Patients as young as 15–18 months were reported to have screw fixation by Anderson et al.\(^\text{34}\) and Gluf and Brockmeyer.\(^\text{3}\)

The use of short constructs (2–3 levels) has been most frequently reported in the literature.\(^\text{5,15,28}\) Longer constructs are sometimes needed to correct complex deformities and for patients with complicated presentations and

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### TABLE 5. Operative and postoperative complications

<table>
<thead>
<tr>
<th>Case</th>
<th>Complication(s)</th>
<th>Initial Surgery</th>
<th>Etiology</th>
<th>Description, Management, &amp; Sequelae</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>VA injury</td>
<td>Occiput–C4</td>
<td>Congenital</td>
<td>Lt VA injured during C-1 lateral mass screw placement, resulting in vessel occlusion; management, Lt VA embolization; sequelae included brainstem infarction, basilar SAH, quadriplegia, locked-in syndrome, &amp; death 18 mos postop</td>
</tr>
<tr>
<td>B</td>
<td>Traumatic pneumothorax</td>
<td>Occiput–C4</td>
<td>Congenital</td>
<td>Traumatic pneumothorax during rib harvest; management, chest tube placement; sequelae, none</td>
</tr>
<tr>
<td></td>
<td>Wound dehiscence</td>
<td></td>
<td></td>
<td>Dehiscence of posterior OC wound 2 mos postop; management, conservative; sequelae, none</td>
</tr>
<tr>
<td>C</td>
<td>Wound dehiscence</td>
<td>Occiput–T4</td>
<td>Congenital</td>
<td>Superficial breakdown of incision 7 mos postop; management included open debridement &amp; antibiotics; sequelae, none</td>
</tr>
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</table>

SAH = subarachnoid hemorrhage.
multilevel pathologies. Such was the case in the majority of our patients. The most frequent construct length in our series was occiput–C4 (42.5%), and the longest construct required a total of 12 levels. Structural autografts are widely used in children2,11,12,15,17,25,34,48,52 to confer superior biomechanical rigidity across the CVJ (where joint surface arthrodesis is not readily attainable) and to provide structural support across the most mobile junction in the spine. Furthermore, the combined cortical/cancellous autografts possess ideal osteogenic, osteoinductive, and osteoconductive properties for the promotion of arthrodesis.6 Morcellized autograft/allograft was used in all patients in this series to augment fusion. Posterior rib harvest was favored due to much lower harvest-site morbidity, according to the senior author’s experience. The size and shape of the rib are also ideally suited for the anatomy of the CVJ.2,33,51 Augmentation with recombinant human bone morphogenetic protein was not performed in any of the patients in this series to augment fusion. Posterior rib harvest was favored due to much lower harvest-site morbidity, according to the senior author’s experience. The size and shape of the rib are also ideally suited for the anatomy of the CVJ.2,33,51 Augmentation with recombinant human bone morphogenetic protein was not performed in any of the patients in this series. Concern about the risks of recombinant human bone morphogenetic protein (e.g., heterotopic bone formation, osteolysis, neoplasia, and the paucity of published literature regarding its use in the pediatric population) was believed to outweigh the potential benefit of its use for this procedure in a pediatric population.12,25

Clinical Outcomes

The recovery rate for patients with chronic myelopathy was almost 21%, with neurological improvement in 88.2% of 17 patients. Of the patients who had neurological improvement, 35.3% regained complete neurological function and 58.8% improved 3–10 categorical points. One (5.9%) patient remained unchanged. Twelve of the 18 patients with trauma were neurologically intact on admission, and only 1 of the 4 patients with initial spinal cord injury showed any improvement. Patients with complete spinal cord injury tended to remain unchanged despite surgical intervention.

The overall survival rate was 100% up to 1 year after the surgery, with 42.5% (17/40) of patients having significant neurological improvement and the same proportion (42.5%, 17/40) remaining neurologically intact. Overall, 1 (2.5%) patient deteriorated (chronic myelopathy), and 5 (12.5%) remained unchanged. In contrast, Astur et al.5 reported neurological impairment in 50% of their patients on the first follow-up; their result was similar to that of Labbe et al.,28 who reported that one-half of their patients had persistent neurological deficits (mostly hemiparesis or hemiplegia).

Successful OCF rates in children have been reported as ranging from 84% to 100%.2,3,5,10,17,20,24,27,33 In a systematic review of 285 patients, Hwang et al.25 found that in
those with screw constructs, the fusion rate was 99% after 3 months, and that in patients with wire constructs, the rate was 95% (94.4% overall average); the difference was not statistically significant. Radiological confirmation of fusion for both screw- and wire-based techniques in our series was 100% within the 1st year after surgery. The mean clinical and radiographic follow-up was 31.9 months (range 16–119 months).

External orthotics were used postoperatively to ensure rigid mobilization and to optimize fusion. Rigid collars are most often used, but halo vests or Minerva jackets can also be applied.5,12,20,26 A radiological analysis of the curvature, alignment, and growth of patients included in this case series (18 of 40 patients; Martinez-del-Campo et al., our unpublished data, 2014) was done immediately after surgery and at the last follow-up. The results show that vertical and longitudinal growth continues after long-term follow-up in the instrumented levels and that there is not a statistically significant change in cervical curvature or alignment.

It is our opinion that adequate patient selection into different fixation modalities contributed to our fusion success rate. No patients required postoperative placement of a halo vest. External orthotics were sufficient to provide and ensure postoperative rigid mobilization and to optimize fusion. All of our patients used a rigid collar for 8 weeks postoperatively, or until fusion was confirmed.

Complication Rates

Complication rates of OC fixation reported in the literature range from 7.8% to 26%.20,25 Complications related to wire techniques are usually more serious and frequent than those related to screw-based techniques (50% vs 14%, respectively). Wiring has the same complications as screw-based techniques, including graft resorption, unintended extension of fusion, hardware failure, quadriparesis, and death.25 Vertebral artery lesions have been described in 1.6%–4% of cases,12,17,20,24,35 hardware failure in up to 31%40 transverse sinus injuries in 10%,24 pseudarthrosis in 10%,24 CSF leak in 7.1%–15%,5,24,26 hydrocephalus in 28.6%,2 neurological deterioration in 50%,3 surgical re-intervention in 10%–13.6%,12,26 rib donor-site morbidity in 3.7%,31 pseudomeningocele in 5%,10 and death in 3.6%–10%.9 These rates were obtained from the reviewed literature, when readily available.

Other complications are wound problems (dehiscence, nonunion, and infection), graft resorption, fusion failure and extension, subfascial fluid collection, airway/respiratory problems, dysphagia/vocal cord paresis, and quadriparesis/quadriplegia.2,3,5,10,12,17,20,24–26,33,36,40,57 The overall complication rate in this series was comparable to what has been previously reported in the literature (7.5%). Our rates were consistent with previous reports for vertebral artery injury (2.5%),2,3,5,10,12,17,20,24,39,57 pneumothorax after rib harvest (2.5%),2,15,33,51 and issues with wound healing (2.5%).2,5,24,26,33,36

Study Limitations

This study contains all of the inherent limitations of a single-surgeon, retrospective case series. Clinical and radiographic follow-up in these patients is ongoing, and the durability of the results will need to be reassessed as follow-up continues.

Conclusions

Pediatric patients with OC instability are at high risk for neurological devastation; these patients benefit from early and definitive treatment. In children, OC instability that requires surgical fixation is most often caused by either trauma or congenital conditions. If there is suspicion of OC instability, a complete diagnostic workup is warranted. The diagnosis can most often be established with a combination of CT, MRI, and/or dynamic radiography. In patients with an unstable CVJ, surgical fixation is indicated. Fixation strategies have evolved as newer technologies have become available. Screw-rod constructs have been demonstrated to be safe and effective, and their use has become increasingly popular.

A screw-rod construct combined with a structural autograft is now the preferred fixation strategy of the senior author in patients for whom it is appropriate. However, for patients with small osseous structures, poor bone quality, and/or unique anatomical relationships, screw-rod constructs may not be appropriate and alternative fixation strategies, such as sublaminar wiring, need to be considered. Long-term follow-up supports favorable clinical and radiographic outcomes. In experienced hands, OC fixation has been demonstrated to be a safe and effective means of treating OC instability and preserving neurological function in pediatric patients.

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Disclosures
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
Conception and design: Theodore, Martinez-del-Campo, Kalb. Acquisition of data: Martinez-del-Campo. Analysis and interpretation of data: Martinez-del-Campo. Drafting the article: Martinez-del-Campo. Critically revising the article: Theodore, Turner, Rangel-Castilla, Soriano-Baron, Kalb. Reviewed submitted version of manuscript: Theodore. Statistical analysis: Martinez-del-Campo. Study supervision: Theodore.

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