Occipitocervicothoracic stabilization in pediatric patients

Clinical article

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Object. Although rarely encountered, pediatric patients with severe cervical spine deformities and instability may occasionally require occipitocervicothoracic instrumentation and fusion. This case series reports the experience of 4 pediatric centers in managing this condition. Occipitocervical fixation is the treatment of choice for cranio cervical instability that is symptomatic or threatens neurological function. In children, the most common distal fixation level with modern techniques is C-2. Treated patients maintain a significant amount of neck motion due to the flexibility of the subaxial cervical spine. Distal fixation to the thoracic spine has been reported in adult case series. This procedure is to be avoided due to the morbidity of complete loss of head and neck motion. Unfortunately, in rare cases, the pathological condition or highly aberrant anatomy may require occipitocervical constructs to include the thoracic spine.

Methods. The authors identified 13 patients who underwent occipitocervicothoracic fixation. Demographic, radiological, and clinical data were gathered through retrospective review of patient records from 4 institutions.

Results. Patients ranged from 1 to 14 years of age. There were 7 girls and 6 boys. Diagnoses included Klippel-Feil, Larsen, Morquio, and VATER syndromes as well as postlaminectomy kyphosis and severe skeletal dysplasia. Four patients were neurologically intact and 9 had myelopathy. Five children were treated with preoperative traction prior to instrumentation; 5 underwent both anterior and posterior spinal reconstruction. Two patients underwent instrumentation beyond the thoracic spine. Allograft was used anteriorly, and autologous rib grafts were used in the majority for posterior arthrodesis. Follow-up ranged from 0 to 43 months. Computed tomography confirmed fusion in 9 patients; the remaining patients were lost to follow-up or had not undergone repeat imaging at the time of writing. Patients with myelopathy either improved or stabilized. One child had mild postoperative unilateral upper-extremity weakness, and a second child died due to a tracheostomy infection. All patients had severe movement restriction as expected.

Conclusions. Occipitocervicothoracic stabilization may be employed to stabilize and reconstruct complex pediatric spinal deformities. Neurological function can be maintained or improved. The long-term morbidity of loss of cervical motion remains to be elucidated. (DOI: 10.3171/2011.4.PEDS10450)

Key Words • occipitocervicothoracic stabilization • Larsen syndrome • Klippel-Feil syndrome • pediatric surgery • spinal instrumentation • spine surgery

Abbreviations used in this paper: OCT = occipitocervicothoracic; VATER = acronym for congenital syndrome defined by vertebral anomalies, anal atresia, tracheoesophageal fistula, esophageal atresia, and renal anomalies.

Occipitocervical stabilization is performed in both adults and children for disorders of the atlantooccipital or atlantoaxial spine as well as those with unstable high cervical traumatic injuries. Occipitocervical fixation is the treatment of choice for cranio cervical instability that is symptomatic or threatens neurological function. In children, the most common distal fixation level with modern techniques is C-2. These patients maintain a significant amount of neck motion due to the flexibility of the subaxial cervical spine. Distal fixation to the upper thoracic spine (occipitocervicothoracic [OCT] stabilization) has been reported in adult series; however, reports of distal fixation in the pediatric literature are sparse. This procedure is avoided in children due to the morbidity of complete loss of head and neck motion. Unfortunately, in rare cases, the pathological condition or highly aberrant anatomy may require occipitocervicothoracic stabilization.
Methods

Records of all pediatric patients undergoing posterior spinal stabilization procedures performed by senior authors at the University of Florida, Columbia University, University of Utah, and New York University between 2000 and August 2010 were reviewed. Patients who underwent OCT fixation were identified. After obtaining institutional review board approval, a retrospective chart review was performed to gather demographic, radiological, and clinical data. Illustrative cases are presented.

Illustrative Cases

Case 2

This 12-month-old Hispanic male was initially seen in consultation for severe cervical kyphosis and myelopathy in the presence of multiple congenital abnormalities consistent with Larsen syndrome. His cervical spinal canal was severely stenotic, with a 2-mm diameter (Fig. 1A and B). The kyphotic deformity measured 135°, with the apex at C4–5. The posterior elements of C-2 were absent and the atlantodental interval was increased. Remarkably, the patient moved all extremities, but he was hypotonic and had diminished spontaneous movement. Correction of the deformity was postponed until the patient was older, given that stable neurological findings were maintained throughout the interval and there were limited options for reconstruction in such a young child. At 26 months of age, he underwent a tracheostomy and a staged procedure consisting of anterior cervical corpectomies followed by occiput–T1 stabilization with autologous rib grafting. He tolerated the procedure without any complication. He was placed in a halo vest postoperatively. Imaging at 4 months demonstrated good alignment (Fig. 1C), and the halo vest was replaced with a rigid cervical collar that was removed 6 weeks later. He remained ventilator dependent for 6 months after correction. A CT study performed 6 months postoperatively demonstrated bony fusion. The patient is currently almost 6 years old; he remains nonambulatory, has significant learning disabilities, and is dependent on a gastrostomy tube.

Case 4

This 2-year-old Hispanic female born in the Dominican Republic presented with poor weight gain, a weak cry, bony anomalies of the hands, and shortness of breath with head flexion. In addition, the parents reported her neck had progressively shortened. A chin-on-chest deformity was present, but the results of her neurological examination were normal. Imaging demonstrated complete absence of anterior elements and hypoplastic posterior elements of the upper cervical spine (Fig. 2A and B). Buckling of the soft tissues anterior to the spinal cord was present (Fig. 2C). Flexion-extension films revealed hyperextension with passive extension. A diagnosis of unspecified spondylocostal dysostosis was made after evaluation by a pediatric geneticist. The patient underwent occiput–T3 fixation with use of occipital screws, multilevel cable and rod fixation, and autologous rib grafting. She was placed in a halo vest postoperatively (Fig. 2D). Unfortunately, she developed a superficial infection at one of the halo pin sites necessitating readmission and intravenous antibiotic therapy. The halo vest was removed after completion of the antibiotic course with evidence of satisfactory alignment (Fig. 2E). The child remained neurologically intact throughout her postoperative course, but was lost to follow-up 6 months after surgery.

Case 5

This 18-month-old white male child underwent evaluation for labored breathing and was found to have a cervical spine anomaly with severe kyphosis at C-5 (Fig. 3A and B). The patient had not yet begun to walk but otherwise

Fig. 1. Case 2. A and B: Preoperative MR image (A) and CT scan (B) demonstrating severe cervical stenosis and kyphosis. C: Postoperative lateral radiograph demonstrating excellent deformity correction.
had met most developmental milestones and was neurologically intact on examination. Imaging demonstrated an incompletely ossified clivus with anterior displacement of the dens below the skull base. In addition, fusion of multiple contiguous facet joints was noted, consistent with Klippel-Feil syndrome. Surgical intervention was postponed until a later date given the child’s young age and intact neurological status. At 30 months of age, he was admitted to the hospital for preoperative traction under sedation, which resulted in some correction of the deformity (Fig. 3C). Surgery was then performed. He was initially placed supine and underwent anterior C-4 and C-5 partial corpectomy, C6–7 anterior cervical discectomy, and anterior plating from C-4 to C-7. He was subsequently placed prone and underwent occiput–T1 stabilization with rib allograft (Fig. 3D). He returned to the operating suite 2 days later for revision of the anterior bone graft. Postoperatively, the patient was weaned slowly from sedation and the ventilator after development of ventilator-associated pneumonia. After extubation, it was noted that he was moving the left arm less than the right. Imaging did not demonstrate any cause for his motor asymmetry. At discharge to a rehabilitation hospital, the patient had evidence of mild left upper-extremity weakness. A CT study performed 6 months postoperatively demonstrated bony fusion (Fig. 3E). Ten months after surgery, the patient is doing well with good alignment. He has persistent left upper-extremity weakness (approximately 4/5 strength), but otherwise remains neurologically intact.

**Results**

A total of 13 patients who underwent OCT stabilization were identified (Table 1). Distal fixation terminated in the thoracic spine in 11 patients and extended distally to the lumbosacral spine in 2 patients who had additional caudal spinal anomalies. The patients’ ages at the time of surgery ranged from 1 to 14 years (mean 4.6 years). Seven patients were female. The diagnoses included Klippel-Feil syndrome, Morquio syndrome, neurofibromatosis Type I, Larsen syndrome, postlaminectomy kyphosis, VATER syndrome, and spondylocostal dysostosis. Four patients were neurologically intact at presentation, and 9 had evidence of myelopathy. Five children were treated with preoperative traction. Eight children underwent posterior stabilization alone; the remaining 5 underwent combined anterior and posterior procedures. Two patients had extension of the posterior constructs to include the lumbosacral spine due to caudal pathology. Allograft was used anteriorly and autologous rib grafts were used for posterior arthrodesis. Duration of follow-up ranged from 0 to 43 months. Fusion was confirmed
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Diagnosis</th>
<th>Age (yrs),†</th>
<th>Sex</th>
<th>Surgical Intervention</th>
<th>FU</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>CM-II, postlaminectomy (C1–5) progr kyphosis w/ myelopathy</td>
<td>9, F</td>
<td></td>
<td>1) preop traction; 2) C4–5 &amp; C5–6 ACDF; 3) occiput–T1 pst instr &amp; fusion</td>
<td>12 mos</td>
<td>improvement in Sx, solid fusion</td>
</tr>
<tr>
<td>2</td>
<td>Larsen syndrome w/ severe skeletal dysplasia, dev delay &amp; myelopathy</td>
<td>2, M</td>
<td></td>
<td>1) C4–5 laminectomy &amp; traction; 2) C-3, C-4, &amp; C-5 corpectomies w/ ant fusion from C-2 through C-7; 3) C5–6 laminectomy &amp; occiput–T1 pst instr &amp; fusion; 4) halo vest</td>
<td>43 mos</td>
<td>nonambulatory but remains neurologically stable, solid fusion</td>
</tr>
<tr>
<td>3</td>
<td>Klippel-Feil syndrome w/ atlantooccipital instability &amp; agenesia of pst elements of C-2, C-3, C-4 &amp; partial agenesia of C-5, C-6, &amp; C-7 w/ chin-on-chest deformity; neural intact</td>
<td>8, M</td>
<td></td>
<td>1) C4–5 &amp; C5–6 ACDF; 2) occiput–T1 pst instr &amp; fusion</td>
<td>6 mos</td>
<td>neurointact, markedly improved posture, solid fusion</td>
</tr>
<tr>
<td>4</td>
<td>spondylocostal dysostosis w/ complete agenesia of ant elements of upper cervical spine &amp; partial agenesia of pst elements</td>
<td>2, F</td>
<td></td>
<td>1) occiput–T3 multilevel cable &amp; rod instr; 2) halo vest</td>
<td>6 mos</td>
<td>neurointact, LTFU</td>
</tr>
<tr>
<td>5</td>
<td>Klippel-Feil syndrome w/ atlantooccipital instability &amp; multi cervical abnormalities w/ severe kyphosis at C-5, myelopathy w/ motor delay</td>
<td>2, M</td>
<td></td>
<td>1) preop traction; 2) C-4 &amp; C-5 partial corpectomy, C6–7 ant cervical discectomy &amp; fusion from C-4 through C-7; 3) occiput–T1 pst instr &amp; fusion; 4) revision of ant bone graft 2 days later</td>
<td>10 mos</td>
<td>persistent II UE weakness (4/5) after op, solid fusion</td>
</tr>
<tr>
<td>6</td>
<td>Morquio syndrome w/ severe C2–3 instability after C1–2 fusion performed at age 5 &amp; multi skeletal abnormalities of cervical, thoracic, &amp; lumbar spine, myelopathy</td>
<td>12, M</td>
<td></td>
<td>1) occiput–L4 pst instr &amp; fusion; 2) halo vest</td>
<td>3 mos</td>
<td>neuro stable</td>
</tr>
<tr>
<td>7</td>
<td>NF-1 w/ swan-neck deformity after multi cervical laminectomies &amp; neurofibroma resections, paraparesis</td>
<td>3, F</td>
<td></td>
<td>1) occiput–T6 pst instr &amp; fusion</td>
<td>27 mos</td>
<td>improvement in neurological function, solid fusion</td>
</tr>
<tr>
<td>8</td>
<td>multi congenital spine anomalies w/ absence of C-8 vertebra &amp; w/ C5–6 spondyloptosis</td>
<td>1, M</td>
<td></td>
<td>1) C5–6 &amp; C6–7 ACDF; 2) occiput–T4 pst instr &amp; fusion</td>
<td>16 mos</td>
<td>neurologically intact, solid fusion</td>
</tr>
<tr>
<td>9</td>
<td>unclassified skeletal anomaly similar to osteogenesis imperfecta w/ cervical instability, kyphotic deformity, stenosis, &amp; quadriparalysis</td>
<td>2, F</td>
<td></td>
<td>1) C1–3 decompressive laminectomies; 2) occiput–T2 pst instr &amp; fusion; 3) revision of construct due to occipital screw pullout w/ placement in halo vest 2 days later</td>
<td>19 mos</td>
<td>neurologically stable, solid fusion</td>
</tr>
<tr>
<td>10</td>
<td>Morquio syndrome w/ multilevel vertebra plana &amp; cervical stenosis, myelopathy w/ quadriparesis &amp; airway compromise w/ neck flexion</td>
<td>14, F</td>
<td></td>
<td>1) occiput–sacrum pst instr &amp; fusion w/ elective tracheostomy prior to procedure</td>
<td>3 wks</td>
<td>death due to respiratory complications from tracheal wound infection</td>
</tr>
<tr>
<td>11</td>
<td>multi congenital spine anomalies &amp; swan-neck deformity, paraparesis</td>
<td>2, F</td>
<td></td>
<td>1) occiput–T4 pst instr &amp; fusion</td>
<td>1 wk</td>
<td>neuro stable</td>
</tr>
<tr>
<td>12</td>
<td>skeletal dysplasia of unknown type w/ progr cervical swan-neck deformity extending to C-6, dev delay but neural intact</td>
<td>2, F</td>
<td></td>
<td>1) preop traction; 2) occiput–T3 pst instr &amp; fusion; 3) postop hard collar</td>
<td>25 mos</td>
<td>neurointact, solid fusion</td>
</tr>
<tr>
<td>13</td>
<td>VATER syndrome w/ congenital C5–6 dislocation &amp; severe stenosis w/ spinal cord impingement, dev delay w/ signs of myelopathy</td>
<td>1, M</td>
<td></td>
<td>1) preop traction; 2) cervical laminectomy &amp; resection of pst elements followed by traction; 3) occiput–T3 pst instr &amp; fusion 5 days later; 4) placement in halo ring &amp; body cast for 10 weeks</td>
<td>4 mos</td>
<td>neurointact, solid fusion</td>
</tr>
</tbody>
</table>

* ACDF = anterior cervical discectomy and fusion; ant = anterior; CM-II = Chiari Malformation Type II; dev = developmental; FU = follow-up; instr = instrumentation; LTFU = lost to follow-up; multi = multiple; neurol = neurological or neurologically; NF-1 = neurofibromatosis Type 1; progr = progressive; pst = posterior; UE= upper extremity.
† The age given refers to the child’s age at surgery.
Occipitocervicothoracic stabilization

by CT in 9 patients; the remaining patients were lost to follow-up (1 patient) or had not yet had repeat imaging (3 patients). Most patients with preoperative myelopathy either improved (4 patients) or stabilized (4 patients). One child with congenital kyphosis (Case 5) had mild postoperative arm weakness that persisted at his latest follow-up visit. One quadriparetic patient who underwent occipit-sacrum instrumentation died 3 weeks after surgery as a result of respiratory compromise after developing a tracheostomy-site infection. This patient is included in the 3 who had not had repeat imaging. Two patients required reoperation within 48 hours of the first surgery; one of these patients required revision of the occipital construct due to screw pullout. Preoperatively, most of the children had significant loss of neck motion as a result of their pathological condition. All patients had severe, complete movement restriction postoperatively. The cervical motion restriction was well tolerated in all patients, with no loss of gross motor ability or impediment to ambulation (in those who were previously ambulatory).

Discussion

In pediatric patients with severe cervical deformity and instability, posterior OCT instrumentation and fusion is a viable and effective management strategy. Occasionally, this procedure may be combined with an anterior cervical decompression, fusion, and instrumentation to achieve relief of bony compression causing neurological deficit. Successful OCT arthrodesis was documented in 9 of 13 patients, with 1 patient having been lost to follow-up, 1 having died in the early postoperative period, and 2 other patients having not yet undergone follow-up imaging. In these 13 patients, highly aberrant anatomy with lack of caudal cervical fixation points necessitated inclusion of the thoracic spine in the constructs. The expected cervical motion restriction postoperatively was not a significant motor impediment for any patient.

Occipitocervicothoracic stabilization is rarely employed to reconstruct complex spinal deformities in adults. Published series of OCT stabilization exist in adult patients with progressive and severe kyphosis from rheumatoid arthritis or secondary to cervical neoplasia. These series indicate good midterm results (average follow-up 5 years in one study) in these patient groups with preservation of alignment and neurological function as well as improvement in pain. However, few case reports of OCT fusion in pediatric patients can be found in the literature. The goals of the present study are to report a multicenter pediatric experience with severe craniocervical disease necessitating OCT stabilization and to provide insight into the indications and outcomes of this procedure in children.

While occipitocervical fusion is a relatively common procedure in pediatric patients with craniocervical abnormalities, OCT fusion is rarely performed. In fact, only 2 reported cases of pediatric OCT stabilization could be located in the literature, both of which were in the setting of Larsen syndrome, a rare autosomal dominant disorder of growth hormone receptors resulting in joint hypermobility as well as extremity and spinal deformities. Generally, OCT fusion is reserved for severe deformity or highly aberrant pathological conditions necessitating total craniocervical stabilization to prevent neurological injury. The disadvantage of this procedure is severe restriction of movement of the head and neck postoperatively.

Although the 13 cases presented in this paper have been gained from 4 different institutions and involve a variety of surgical techniques, they still serve to illustrate several important principles regarding the indications and techniques of OCT fusion. First, 9 of the 13 patients in this series were surgically treated at or before the age of 2 years, demonstrating that these cervical anomalies are typically diagnosed early in life. After the diagnosis is made, a cervicothoracic orthosis is typically used until the decision is made to proceed with surgery. For most patients, indications for early surgery are progressive cervical deformity that threatens neurological function, progressive neurological deficit, or the patient’s bony anatomy reaching the stage where internal cervical screw fixation is a viable option. Although there are no established criteria to indicate when a patient’s bone has reached a stage of development adequate for fusion, in our experience delaying surgery until about 24 months of age provides both an adequate bony substrate and a satisfactory bone growth potential for rigid internal fixation. In many cases, multiple posterior cervical fixation points are used to achieve stability and load-sharing as part of the constructs. In other cases, the cervical bony anatomy is so deficient or distorted that cervical fixation is impossible and direct fixation from the occiput to the upper thoracic region is the only viable option. Careful judgment must be exercised when making these decisions. One must keep in mind that although the modern techniques of spine surgery have made these procedures possible, we are still in the very early stages of determining their ultimate effectiveness. Second, a skilled pediatric spine surgeon is required to place the thoracic pedicle screws. These screws can be difficult to insert and are critically important, as they anchor the construct. As a corollary, the breadth of the patient’s occipital bone is also important, as the pullout strength of the occipital screws is directly related to bone quality and thickness. Third, although posterior instrumentation and fusion fixations are frequently stand-alone constructs (8 of the 13 reported procedures), additional anterior procedures are sometimes necessary. Each clinical scenario needs to be evaluated to determine whether such anterior procedures are necessary. Finally, a halo orthosis, or halo-like device, was used postoperatively in only 4 cases. This is a significant departure from historical controls; in the past, postoperative use of a halo orthosis was routine. Such changes are a testimony to the significant advances seen in spine surgery, particularly in pediatric craniocervical surgery.

We acknowledge that the limitation of motion after OCT fusion is poorly understood and may, after long-term follow-up, have unintended consequences. We were able to locate only one biomechanical study on OCT constructs, which evaluated the effect of partial versus sequential fixation on the mobility of adult spines. No similar studies could be found in the pediatric literature. Conversely, the effects of occipitocervical fusion on spinal curvature and cervical motion in children have been investigated. Some studies suggest that progressive exaggeration of cervical
lordosis occurs after occipitocervical fusion due to continued growth of the anterior elements. Other studies have not found any kyphotic, lordotic, or adjacent-segment pathology following occipitocervical fusion in children. However, the effects of growth seen after occipitocervical fusion may not be easily translated to a rigid fixed segment from the occiput to the thoracic spine. It is widely accepted that OCT fusion results in near-total absence of cervical motion; however, this consensus has not been confirmed by biomechanical studies. Although no negative sequelae on neck growth were seen during the follow-up period in this series, the long-term impact of such constructs on spinal growth remains to be determined, and delayed deformities will likely take several years to develop. Therefore, it is mandatory that children’s families understand that a likely consequence of this procedure is the permanent inability to move the head or neck on the chest. This may not always be a major issue since patients with these severe anomalies may have significant preoperative movement restriction.

One patient from our series developed a postoperative neurological deficit without evidence of neural compression on postoperative imaging. One patient died of complications associated with preoperative elective tracheostomy. The remaining 11 patients had either improvement or preservation of neurological function, although follow-up was brief (0–43 months). A review of 26 cases of congenital kyphosis indicated an association between perioperative neurological deficit and increased age, preexisting myelopathy, and greater kyphotic deformity prior to cervical stabilization in children. Overall, this case series demonstrates that OCT fusion is well tolerated in the majority of patients. Consideration of OCT stabilization in pediatric patients should only be entertained when all other stabilization options have been exhausted. Candidacy for this procedure should be reviewed only after the patient’s age, underlying diagnosis, functional status, individual anatomy, and family wishes have been reviewed in detail. Given the near-total restriction of head and neck movement resulting from OCT stabilization, we believe this procedure should be avoided unless absolutely necessary.

**Conclusions**

We report 13 cases of severe cervical deformity in pediatric patients necessitating OCT instrumentation and fusion. Occipitocervicothoracic stabilization is rarely employed to reconstruct complex spinal deformities in adults and is even more rare in the pediatric population. Neurological function in children with severe cervical kyphosis and resultant myelopathy can be preserved or improved by this procedure if it is carefully planned and executed. The long-term morbidity of loss of cervical motion remains to be elucidated. For this reason, OCT stabilization should be performed only when other treatment options have been exhausted.

**Disclosure**

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Fargen, Pincus. Acquisition of data: all authors. Analysis and interpretation of data: Fargen, Angevine, Brockmeyer, Pincus. Drafting the article: Fargen, Anderson, Harter, Coon, Brockmeyer, Pincus. Critical revising the article: Fargen, Anderson, Harter, Brockmeyer, Pincus. Study supervision: Pincus.

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