Dyspnea and dysphagia from upper airway obstruction after occipitocervical fusion in the pediatric age group

*Meng Huang, MD, David D. Gonda, MD, Valentina Briceño, RN, Sandi K. Lam, MD, MBA, Thomas G. Luerssen, MD, and Andrew Jea, MD

Neuro-Spine Program, Division of Pediatric Neurosurgery, Texas Children’s Hospital, Department of Neurosurgery, Baylor College of Medicine, Houston, Texas

Upper airway obstruction resulting from overflexion of the craniocervical junction after occipitocervical fusion is a rare but potentially life-threatening complication and is associated with morbidity. The authors retrospectively reviewed the medical records and diagnostic images of 2 pediatric patients who underwent occipitocervical fusion by the Neuro-Spine Program at Texas Children’s Hospital and experienced dyspnea and/or dysphagia from new upper airway obstruction in the postoperative period. Patient demographics, operative data, and preoperative and postoperative occiput-C2 angles were recorded. A review of the literature for similar complications after occipitocervical fusion was performed. A total of 13 cases of prolonged upper airway obstruction after occipitocervical fusion were analyzed. Most of these cases involved adults with rheumatoid arthritis. To the best of the authors’ knowledge, there have been no previous reports of prolonged upper airway obstruction in children after an occipitocervical fusion. Fixation of the neck in increased flexion (−18° to −5°) was a common finding among these adult and pediatric cases. The authors’ cases involved children with micrognathia and comparatively large tongues, which may predispose the oropharynx to obstruction with even the slightest amount of increased flexion. Close attention to a satisfactory fixation angle (occiput-C2 angle) is necessary to avoid airway obstruction after an occipitocervical fusion. Children with micrognathia are particularly sensitive to changes in flexion at the craniocervical junction after occipitocervical fixation.

http://thejns.org/doi/abs/10.3171/2015.1.FOCUS14810

KEY WORDS occipitocervical fusion; pediatric spine; spinal instrumentation; respiratory failure; dyspnea; dysphagia

Case Reports

Case 1

History and Examination

This 15-year-old boy had a medical history of a posterior fossa ependymoma that had been resected via suboccipital craniectomy at 1 year of age, followed by radiation therapy. At the age of 4 years, he developed a cervical syrinx associated with Chiari malformation; he was treated with marsupialization of the syrinx into the spinal subarachnoid space via a C1–4 laminectomy. After being lost to follow-up for almost 10 years, the patient presented with complaints of progressive neck pain and posterior headache that worsened with exertion. Findings from his neurological examination were normal. CT and MRI showed...
recurrence of syrinx and persistence of Chiari malformation, with associated platybasia and basilar invagination from settling of the skull base on the upper cervical spine as a result of his prior multiple surgeries and radiation therapy at the craniocervical junction. He was admitted for elective repeat Chiari decompression and concurrent occipitocervical fusion.

Operation
The patient’s preoperative anesthesia evaluation was significant for Mallampati Class II airway, normal thyromental distance and normal mouth opening, but micrognathia. After induction of anesthesia, the patient was intubated uneventfully in 1 attempt with a 6.5-mm endotracheal tube with fiberoptic endoscopy. His head was then placed in a 3-pin Mayfield headholder, and the patient was positioned prone on chest rolls with his head and neck in flexion for the Chiari decompression. After the usual sterile preparation and draping, the patient underwent syringosubarachnoid stenting and expansile duraplasty as the first part of the procedure. At this point, the patient’s head was repositioned to reduce the amount of flexion and achieve a neutral neck position; this repositioning was performed suboptimally underneath the drapes. Unfortunately, a postpositioning radiograph to ensure a neutral position was not obtained. Routine occipitocervical fusion (occiput to C-5) was then carried out. After routine closure, the patient was positioned supine on the recovery bed, taken out of the Mayfield headholder, and extubated.

Immediately after extubation, the patient displayed an obstructive breathing pattern with desaturations that did not respond to noninvasive positive pressure techniques. A subsequent attempt at laryngeal mask anesthesia placement was unsuccessful, and reintubation attempts with direct and fiberoptic visualization were unsuccessful due to airway and tongue edema preventing visualization. The otorhinolaryngology service was consulted, and an emergency tracheotomy was performed. The patient was then transported to the pediatric intensive care unit for ventilator and respiratory support. Fortunately, he remained at his neurological baseline.

Hospital Course
A comparison of the preoperative and postoperative CT scans showed that the patient’s occiput-C2 angles were +3° and −2°, respectively (Fig. 1). The patient remained neurologically stable, and with the tracheostomy in place, he was stable from the respiratory perspective. He was eventually weaned to room air and was discharged home on postoperative Day 10.

Follow-Up
By the most recent follow-up, more than 3 years after the occipitocervical fusion, a sleep study for snoring and apneas showed persistent severe obstructive sleep apnea (apnea-hypopnea index 32.6); this was not present prior to the fusion.

The patient’s tracheostomy has been maintained given his complex airway and the possibility for future surgery. The patient is currently undergoing evaluation for mandibular advancement secondary to his severe obstructive sleep apnea, attributable to the iatrogenic flexion-induced oropharyngeal constraint.

Case 2
History and Examination
This 15-year-old girl had a history of central sleep apnea, Chiari malformation, and cervicothoracic syrinx that developed after suboccipital craniectomy, syringosubarachnoid marsupialization, and expansile duraplasty at an outside institution at the age of 11 years. She presented to our service with new symptoms of progressive neck and shoulder pain. Neurological examination findings were normal. Chronological review of her CT scans and MR images before and after surgery demonstrated persistence and progression of her holocord syrinx associated with platybasia and basilar invagination. She was scheduled for placement of a syringosubarachnoid shunt as well as occipitocervical fusion.

Operation
The patient’s preoperative anesthesia evaluation was significant for Mallampati Class I airway, normal thyromental distance and normal mouth opening, but micrognathia. The patient was intubated after induction of anesthesia without complication. Her head was placed in a 3-pin Mayfield headholder and she was positioned supine. After the usual preparation and draping, spinal instrumentation was placed from the occiput to C-4 in a standard manner. A C-4 laminectomy was then performed, followed by durotomy and myelotomy, and a syringosubarachnoid shunt was placed. After the usual closure, the patient was extubated without complication.

Hospital Course
A comparison of the preoperative and postoperative CT scans showed that the patient’s occiput-C2 angles were −8° and −14°, respectively (Fig. 2). The patient seemed to tolerate the procedure well and was discharged on the 5th day after surgery.

Of note, during her hospital course, the patient did complain of new difficulty with swallowing food with a tough consistency such as meat, and the need to sleep in an upright 90° position to prevent a perception of shortness of breath. Regrettably, no workup was initiated for these apparent minor complaints, which were thought to be related to the intubation technique and endotracheal tube itself.

Follow-Up
Over the course of 2 years after the occipitocervical fusion, the patient did develop more obvious postoperative dyspnea and dysphagia. Swallow evaluation showed no gross aspiration but did reveal atypical oropharyngeal anatomy with markedly reduced oropharyngeal space with the posterior tongue close to the posterior pharyngeal wall. A sleep study was performed and showed mild obstructive sleep apnea (apnea-hypopnea index 13.7) also attributable to a large tongue and small posterior pharyngeal space.

Like our other patient, this patient is currently under evaluation for mandibular advancement given her persis-
tent symptomatology due to her iatrogenic flexion-induced oropharyngeal constraint in the setting of atypical anatomy.

**Discussion**

Upper airway obstruction after extubation is a life-threatening complication of anterior or posterior cervical spine surgeries. Moreover, even mild dyspnea or dysphagia after cervical spine surgery carries significant morbidity for patients, necessitating notable alterations of lifestyle such as sleeping in an upright position or changing to a soft mechanical diet. In the present cases, the upper airway obstruction was thought to be related to a reduction in the pharyngeal space as evaluated by our otolaryngology colleagues. Our patients had micrognathia but with no signs of airway obstruction or mouth opening restriction prior to surgery. We suspected that the occipitocervical fusion in a flexed position may have caused the upper airway obstruction in these patients.

Although it is known that a posterior occipito-cervico-thoracic fusion in a flexed position may cause dysphagia or, rarely, dyspnea, we were unsure that this was applicable to relatively shorter occipitocervical fusions because the subaxial cervical range of motion is not completely restricted. Thus, we regrettably did not perform a revision of the occipitocervical fusion to realign the craniocervical junction. If a similar situation presented to us in the near future, however, we would give strong consideration to immediate revision.

**FIG. 1.** Case 1. Preoperative (left) and postoperative (right) sagittal reconstructions of the cervical CT scans demonstrating occiput-C2 angles (subtended by McGregor's line and a line through the inferior endplate of C-2) measuring +3° and −2°, respectively. The angle is positive if it opens anteriorly; the angle is negative if it opens posteriorly. The difference between the postoperative and preoperative occiput-C2 angles is −2° − (+3°) = −5°.

**FIG. 2.** Case 2. Preoperative (left) and postoperative (right) sagittal reconstructions of cervical CT scans showing occiput-C2 angles (subtended by McGregor's line and a line through the inferior endplate of C-2) measuring −8° and −14°, respectively. The angle is positive if it opens anteriorly; the angle is negative if it opens posteriorly. The difference between the postoperative and preoperative occiput-C2 angles is −14° − (−8°) = −6°.
The oropharynx is the area from the soft palate to the upper border of the epiglottis. It forms an upper aerodigestive tract with many functions, including deglutition, respiration, and phonation. The tongue root provides the primary force for the movement of food from the oropharynx, around the epiglottis, and into the laryngopharynx. The soft palate is elevated to seal the nasopharynx, and the suprahypoid muscles pull the larynx up and forward. The epiglottis moves downward to cover the airway while striated pharyngeal muscles contract to move the food bolus.

As shown in Table 1, most of the adult patients (7 of 11) carried a diagnosis of rheumatoid arthritis. Because it is difficult to intubate many patients with rheumatoid arthritis, the intubation maneuver may cause pharyngeal injury and edema. Likewise, children with micrognathia may be challenging to intubate, and in severe cases mandibular distraction or elective tracheostomy prior to other major surgery may be required. Taking these points into consideration, micrognathia seems to be a major risk factor for postoperative upper airway obstruction. Furthermore, operative times for occipitocervical fusion may be fairly long (range 2.75–8 hours). Operations in the prone position over a long period of time could increase the risk of pharyngeal edema.

In a few of the tabulated cases (2 of 13), the craniocervical junction was fixed in a neutral or extended position, rather than a flexed position. Moreover, in some cases, postoperative dyspnea and dysphagia were managed without revision of the spinal instrumentation and spinal alignment. In most cases (9 of 13), dysphagia persisted even with the resolution of dyspnea. These factors suggest that the causes of upper airway obstruction after occipitocervical fusion are multifactorial.

Nonetheless, of the factors described above, the occiput-C2 angle should be a key factor in preventing postoperative dyspnea and dysphagia after occipitocervical fusion. It may be measured intraoperatively with the aid of fluoroscopy or lateral radiography after the patient is positioned.

### TABLE 1. Cases of upper airway obstruction after occipitocervical fusion

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Indications for Fusion</th>
<th>Procedure</th>
<th>Fixed Position (difference)*</th>
<th>Length of Op (hrs)</th>
<th>Revision</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kainuma &amp; Yama-da, 1985</td>
<td>1</td>
<td>53, F</td>
<td>RA</td>
<td>Oc–C4 fusion</td>
<td>Flexed</td>
<td>8</td>
<td>No</td>
<td>Prolonged intubation</td>
</tr>
<tr>
<td>Miyata et al., 2009</td>
<td>2</td>
<td>70, M</td>
<td>RA</td>
<td>NA</td>
<td>Extended</td>
<td>4.75</td>
<td>Yes</td>
<td>Dysphagia</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>61, F</td>
<td>RA</td>
<td>Oc–C3 fusion</td>
<td>Neutral</td>
<td>2.75</td>
<td>No</td>
<td>Prolonged intubation</td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>63, F</td>
<td>Epidural tumor</td>
<td>Oc–C4 fusion</td>
<td>Flexed</td>
<td>6.7</td>
<td>Yes</td>
<td>Dysphagia persisted</td>
</tr>
<tr>
<td></td>
<td>5</td>
<td>69, M</td>
<td>RA</td>
<td>NA</td>
<td>NA</td>
<td>4.6</td>
<td>No</td>
<td>Tracheostomy, dysphagia persisted</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>77, F</td>
<td>RA, atlantoaxial subluxation</td>
<td>Oc–C2 fusion</td>
<td>Flexed (−18°)</td>
<td>4.9</td>
<td>Yes</td>
<td>Dyspnea resolved after revision, dysphagia persisted</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>78, F</td>
<td>Atlantoaxial osteoarthritis, occipitalization of C-1</td>
<td>Oc–C3 fusion</td>
<td>Flexed (−14°)</td>
<td>NA</td>
<td>NA</td>
<td>Dysphagia resolved after 7 mos</td>
</tr>
<tr>
<td></td>
<td>8</td>
<td>41, M</td>
<td>Atlantoaxial subluxation, occipitalization of atlas</td>
<td>Oc–C2 fusion</td>
<td>Flexed (−14°)</td>
<td>NA</td>
<td>NA</td>
<td>Dysphagia persisted</td>
</tr>
<tr>
<td></td>
<td>9</td>
<td>63, F</td>
<td>RA, atlantoaxial subluxation</td>
<td>Oc–C3 fusion</td>
<td>Flexed (−5°)</td>
<td>NA</td>
<td>NA</td>
<td>Dysphagia persisted</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>56, F</td>
<td>RA, atlantoaxial subluxation</td>
<td>Oc–C2 fusion</td>
<td>Flexed (−16°)</td>
<td>NA</td>
<td>No</td>
<td>Dysphagia persisted</td>
</tr>
<tr>
<td>Tagawa et al., 2011</td>
<td>11</td>
<td>63, F</td>
<td>RA</td>
<td>Oc–C4 fusion</td>
<td>Flexed</td>
<td>5.5</td>
<td>Yes</td>
<td>Dyspnea resolved</td>
</tr>
<tr>
<td>Present cases</td>
<td>12</td>
<td>15, M</td>
<td>Basilar invagination, platybasia, cervical cord syrinx</td>
<td>Oc–C5 fusion</td>
<td>Flexed (−5°)</td>
<td>5</td>
<td>No</td>
<td>Tracheostomy, severe obstructive sleep apnea</td>
</tr>
<tr>
<td></td>
<td>13</td>
<td>15, F</td>
<td>Basilar invagination, platybasia, holocord syrinx</td>
<td>Oc–C4 fusion</td>
<td>Flexed (−6°)</td>
<td>4.5</td>
<td>No</td>
<td>Mild obstructive sleep apnea, dysphagia persisted</td>
</tr>
</tbody>
</table>

NA = not available; Oc = occiput; RA = rheumatoid arthritis.

* “Difference” indicates the difference between the postoperative and preoperative occiput-C2 angles.

Our review of the published literature revealed 11 Japanese-language cases of prolonged upper airway obstruction after occipitocervical fusion (Table 1). There were no other reports outside of Japan. It is interesting to note that all of the previous cases reported (11 of 11) involved adult patients. Our patients represent the youngest patients in whom the condition has been reported. Ten of the 13 patients included in Table 1 were fused in varying degrees of flexion, with a decrease in the postoperative compared with the preoperative occiput-C2 angles (−5° to −18°). The difference between postoperative and preoperative occiput-C2 angles has been shown to correlate linearly with the cross-sectional area of the oropharynx and has an effect on dyspnea and dysphagia after occipitocervical fusion.

As shown in Table 1, most of the adult patients (7 of 11) carried a diagnosis of rheumatoid arthritis. Because it is difficult to intubate many patients with rheumatoid arthritis, the intubation maneuver may cause pharyngeal injury and edema. Nonetheless, of the factors described above, the occiput-C2 angle should be a key factor in preventing postoperative dyspnea and dysphagia after occipitocervical fusion. It may be measured intraoperatively with the aid of fluoroscopy or lateral radiography after the patient is positioned.
prone. The margin for error for the occiput-C2 angle is small. Miyata et al.\textsuperscript{9} showed that even with small differences in postoperative and preoperative occiput-C2 angles (−10° to −5°), postoperative upper airway obstruction can be seen, as confirmed in our patients with differences of −5° and −6°. Conversely, the same authors demonstrated no postoperative dyspnea or dysphagia in patients with a positive difference between postoperative and preoperative occiput-C2 angles.

Based on our experience with these complications, we have implemented an operating room checklist to minimize the risk of occipitocervical fusion in overflexion or overextension (Fig. 3). Other authors may find that it is safer and more prudent to use the halo vest before surgery to ascertain an adequate craniocervical junction alignment and to avoid unexpected dyspnea and dysphagia.\textsuperscript{7} However, the halo vest may be poorly tolerated in the pediatric age group, and still may be unable to control the intricate and subtle craniocervical alignment precisely.

**Conclusions**

The occiput-C2 angle, measured between McGregor’s line and the inferior endplate of C-2, has considerable im-

---

**FIG. 3.** Proposed algorithm for minimizing inadvertent occipitocervical fusion in overflexion or overextension. Oc = occiput.
pact on dyspnea and/or dysphagia after occipitocervical fusion. It may be used as a practical and reliable indicator during surgery in the hope of avoiding the devastating sequelae of upper airway obstruction. It is important to maintain an occiput-C2 angle not less than the preoperative angle to avoid inadvertent postoperative dyspnea or dysphagia, especially in children with a smaller oropharynx and a history of micrognathia. These patients may benefit from preoperative consultation with an oromaxillofacial surgeon, otolaryngologist, and anesthesiologist regarding risk of surgery and necessity for pre-fusion mandibular distraction or tracheostomy in extreme cases. Intraoperative fluoroscopy or lateral radiography to check the occiput-C2 angle is a recommended part of the process of positioning the patient prone and confirming that the head and neck are in neutral position.

References

Author Contributions
Conception and design: Jea. Acquisition of data: Jea, Gonda, Briceño. Analysis and interpretation of data: Jea, Lam. Preparation of manuscript: Jea, Huang, Gonda, Briceño. Study supervision: Jea.

Correspondence
Andrew Jea, Department of Neurosurgery, Texas Children’s Hospital, 6621 Fannin St., CCC 1230.01, 12th Fl., Houston, TX 77030. email: ahjea@texaschildrens.org.