Pathology of the craniocervical junction represents one of the more challenging spinal abnormalities in terms of surgical management. Numerous pathologies can lead to abnormal degeneration of the craniocervical junction, including osteoarthritis, rheumatoid arthritis (RA), Down’s syndrome, neoplasia, trauma, and Chiari malformation. Lesions in this location have been traditionally accessed through an anterior approach to reduce mass effect on the brainstem and high cervical spinal cord. Often a large pannus forms in this location as a result of instability among the osseous and ligamentous elements as well as the joint complexes, leading to an abnormal fibrous complex. Therefore instability can result in an upward translation of the upper cervical elements into the cranial vault and compress the neural elements at the cervicomedullary junction. This anterior pathology can lead to numerous symptoms including cranial neuropathies, bulbar pathology, intracranial hypertension, cervical myelopathy, respiratory suppression, pain, and even hydrocephalus.

Several terms are used to describe degenerative pathology at the craniocervical junction. Benke et al. describe the unique pathologies of “basilar invagination,” “basilar impression,” and “cranial settling.” Basilar invagination is a superior protrusion of the dens and loss of skull height due to congenital abnormalities. Basilar impression is attributed to skull base softening, usually caused by an acquired condition such as Paget’s disease or osteomalacia. Cranial settling occurs when there is vertical subluxation of the dens caused by the loss of ligamentous support structures commonly seen in rheumatoid or psoriatic arthritis.

Multiple radiographic measurements have been developed to quantify the degree of pathology at the craniocervical junction (Table 1). They all seek to address malalignment of the upper cervical spine with regard to the skull base (Fig. 1).

Just as there are multiple etiologies of craniocervical pathologies, there are multiple surgical approaches to treat disorders of this region when patients are symptomatic and unstable.

**Key Words**
- craniocervical instability
- basilar invagination
- cranial settling
- cervical fusion

This article contains some figures that are displayed in color online but in black-and-white in the print edition.

Abbreviations used in this paper: OC = occipitocervical; RA = rheumatoid arthritis; SSEP = somatosensory evoked potential.
warrant surgery. Traditionally a transoral approach to this pathology has been used.\textsuperscript{10,14,29,32,42} Recent improvements in spinal instrumentation technology\textsuperscript{43} have given spine surgeons other methods for treating craniocervical pathologies. In this paper we present cases illustrating that a posterior-only approach with intraoperative reduction can achieve the desired fusion construct in cranial settling.

**Methods**

**Case 1**

*History and Examination.* A 51-year-old woman with known RA treated with adalimumab presented with chronic neck pain, headache, and left-sided facial pain and numbness. Her physical examination was unremarkable. Magnetic resonance imaging of the brain demonstrated cranial settling with subsequent cervicomedullary kinking (Fig. 2). Further workup with flexion and extension radiographs of the cervical spine revealed instability (Fig. 3). Her radiographic parameters are listed in Table 2, showing measurements consistent with cranial settling.

**Surgical Technique.** Preoperatively the patient was admitted and placed in cervical traction with Gardner-Wells tongs with 15 lb of weight applied in neutral distraction for 2 days, while progress was monitored via daily cross-table lateral radiographs. With signs of only mild reduction, the patient was brought to the operating room and underwent fiberoptic endotracheal intubation. Baseline somatosensory evoked potentials (SSEPs) were obtained prior to patient positioning. The patient lay prone on a radiolucent table with her head resting on a foam headrest to allow for reduction maneuvers. The weights were then reattached to the tongs to provide neutral traction.

Following standard exposure, an occipital keel plate was attached, with bicortical screw purchase at multiple fixation points. Bilateral C-2 laminar screws and C3–5 lateral mass screws were placed under fluoroscopic guidance. Rods were contoured to fit the angle between the occipital keel plate and the cervical screw heads without tension. The rods were initially left long at both ends to allow for reduction maneuvers. Screw caps were then provisionally tightened.
Posterior fusion for craniocervical instability

At this point, several reduction maneuvers were used to facilitate reduction in essentially two vectors: 1) anterior reduction of C-2 relative to the foramen magnum, and 2) inferior reduction of C-2 relative to the foramen magnum. To facilitate anterior reduction, a rod extender was temporarily placed on the rod posterior to the keel plate (Fig. 4). After loosening the screw caps on the keel plate, compression between the keel plate and the rod extender facilitated anterior reduction of C-2 relative to the foramen magnum (Fig. 5B).

Two reduction maneuvers were available for inferior reduction of C-2 relative to the foramen magnum. Analogous to the above maneuver, a rod extender can be fashioned on the rod caudal to the C-5 lateral mass screw. The screw caps can be loosened cephalad to the rod extender, and compression can be applied between the rod extender and the lateral mass screws (Fig. 5C). Alternatively, a rod extender can be attached above the most cephalad spinal fixation point, and distraction between this point and the lateral mass screws can facilitate the same maneuver (Fig. 5D).

**TABLE 2: Preoperative and postoperative radiological results in 3 cases of craniocervical instability**

<table>
<thead>
<tr>
<th>Line</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
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<tbody>
<tr>
<td></td>
<td>Preop</td>
<td>Postop</td>
<td>Preop</td>
</tr>
<tr>
<td>Ranawat's (mm)</td>
<td>5.1*</td>
<td>125*</td>
<td>10.8*</td>
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<tr>
<td>McRae's</td>
<td>above</td>
<td>below</td>
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<tr>
<td>McGregor's (mm)</td>
<td>10.6</td>
<td>2.7</td>
<td>7.4</td>
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<tr>
<td>Chamberlain's (mm)</td>
<td>8.4</td>
<td>1.5</td>
<td>4.4</td>
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* Indicates measurements for females.

Fig. 3. Case 1. Sagittal cervical spine CT (A) demonstrating the extent of bony elements not previously seen well on MRI findings (Fig. 2). Extension (B) and flexion (C) radiographs showing instability at the C1–2 junction with an increase in distance between the atlas and odontoid process.

Fig. 4. Case 1. Intraoperative fluoroscopic images showing anterior translation (upper), moving the odontoid process away from the spinal cord, and vertical translation (lower), which in turn pushes the skull/occipital plate away from the cervical spine.
These maneuvers were performed under fluoroscopic guidance while monitoring for any changes in intraoperative SSEPs. Following reduction, final tightening of all hardware, the removal of temporary rod extenders, and trimming of any excess rod previously used for reduction were performed. Bone decortication was performed along with allograft and autograft placement to facilitate fusion. After surgery, the patient was maintained in a rigid cervical collar for immobilization (Fig. 5E).

**Postsurgical Course.** The patient tolerated the procedure well without any complications, and her symptoms resolved after surgery. Follow-up imaging 8 months after surgery (Fig. 6) showed no hardware complications.

**Case 2**

A 34-year-old woman with a history of RA and currently taking prednisone and certolizumab pegol, in whom RA had been diagnosed at 20 years of age, presented in a halo brace for a second opinion. Four months earlier at an outside hospital, she had undergone a C1–2 fusion via a sublaminar wiring technique for craniocervical instability. Imaging demonstrated pseudarthrosis, and she became symptomatic with electric shocks and body weakness when putting her head on a pillow, thus prompting treatment with halo bracing. Radiological imaging showed occipitocervical (OC) instability with minimal correction after halo reduction. Closed reduction was not performed in this case since the sublaminar wires were bowed into the spinal canal and there was atlantoaxial instability.

**Surgical Technique.** Similar to the prior case, the patient was brought to the operating room, and awake fiberoptic endotracheal intubation was performed. Prior to taking off the halo brace, we put a hard cervical collar in place and obtained baseline SSEP measurements. A Mayfield skull clamp (Integra) was used to attempt closed reduction with the patient under anesthesia. Because there was no need for vertical translation of the C-2 vertebra, the patient underwent surgery in the Mayfield frame. Intraoperative closed reduction with neuromonitoring was attempted to correct the subluxation under fluoroscopy, but it was unsuccessful. The surgery was performed without using the monopolar electrocautery since the sublaminar cables abutted the spinal cord. A laminectomy was performed en bloc to remove the previous hardware without injuring the spinal cord. Somatosensory evoked potentials showed improvement after the decompression.

Manual reduction of C-1 was attempted, but the vertebra did not move. An occipital plate and C-2 pars screws with C-3 lateral mass screws were placed bilaterally, skipping C-1 given an abundance of scar tissue and the absence of a tissue plane to safely identify normal point...
anatomy. A reduction maneuver was performed after contouring a rod into the screw heads without tension, with the screw caps provisionally tightened (Fig. 7). At this point, a temporary heavy rod holder was placed caudal to the occipital plate and a distractor instrument was placed in-between (Fig. 8). Under fluoroscopy, we saw the action of the distractor instrument allow for anterior translation of the axis in relation to the atlas (Video 1).

**Video 1.** Saw-bone model demonstrating the anterior reduction maneuver using a distractor between the occipital plate and rod holder. Copyright Joseph O'Brien. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

**Postsurgical Course.** The patient tolerated the procedure well without any complications, and her symptoms resolved after surgery. Follow-up imaging at 1 year after surgery (Fig. 9) showed no hardware complications.

**Case 3**

**History and Examination.** A 73-year-old woman with RA presented with progressive worsening of neck pain and frequent falls, which were attributed to balance issues. On physical examination she had limited motion in her cervical spine, and when she looked down she had shooting pain down her neck and back. She had limited use of her hands, was hyperreflexive, and required a walker. Preoperative closed reduction was not performed.

**Surgical Technique.** The setup and approach for this case were similar to those in Case 2. A longer construct extending from the occiput to C-6 was used given concerns regarding osteoporosis. Lateral mass screws were placed from C-3 to C-6, skipping C-2 since C2–3 was autofused.

Alignment was achieved with a single reduction maneuver. The offset connector/rod extender was placed slightly caudal to the occipital plate, serving two purposes. First, it allowed the distractor instrumentation to have a solid backing and the distractor to use the rod as a “track.” Second, it allowed maximal torque to be applied since the vector parallels the force applied by the distractor anteriorly (Fig. 10).

Again, cervical reduction was performed under fluoroscopy, and continuous SSEP monitoring demonstrated no adverse change in signals. Following the instrumentation, a cervical laminectomy was performed, as was decortication and placement of allograph and autograph to aid with fusion.

**Postsurgical Course.** The patient tolerated the procedures well without any complications. She continues to use her walker for assistance; however, she has reported improvement in the use of her hands. All postoperative radiographic evaluation demonstrated good reduction, with craniocervical measurement parameters all showing reduction into normal ranges (Table 2).

**Discussion**

The number of surgical cases caused by RA is on the decline as a result of awareness, early treatment, and improved medications. However, spine surgeons still encounter cranial settling due to a rheumatoid process that can be difficult to correct. Progression of this disease in the cervical spine oftentimes results in a large pannus that compresses the cervicomedullary junction, causing multiple symptoms as mentioned above. Matsunaga et al. showed that cervical decompression and fusion in symp-

**Fig. 6.** Case 1. Preoperative (upper) and postoperative (lower) images obtained at 8 months, showing reduction of the dens with a stable construct.
tomatic patients is necessary. At the 3-year follow-up, their entire nonsurgical arm had become bedridden, and by 8 years these same patients had all died, with a mean survival of 4.2 years. On the other hand, the surgical arm not only showed improvement, but also lived for up to 18 years, with a mean survival of 9.7 years. Without a doubt, symptomatic patients with cranial settling require surgical intervention.

The anterior surgical approach has been traditionally applied in treating this pathology and has a very high success rate. Hadley and colleagues described a series of 53 patients with basilar invagination and brainstem compression due to RA, and surgical morbidity and mortality in these patients was 6% and 0%, respectively. The morbidities associated with this approach include infection, breathing difficulties requiring a tracheostomy, swallowing difficulties, and CSF fistulas. Furthermore, the instability caused by ligamentous disruption from the anterior approach does require supplemental stabilization from a posterior approach. Other anterior surgical techniques have been explored, such as transoral robotic surgery, endoscope-assisted transnasal surgery, and transcervical surgery. Ultimately, the goal of
these new methods is to decrease the morbidities associated with the transoral approach.

To avoid the morbidities associated with anterior surgery, solely posterior approaches to treat cranial settling have been explored over the years.\textsuperscript{1,11,17,38} The difficulty lies with indirect decompression of the anterior pathology since the spine has vertically translated into the cranial vault. Reduction techniques via cervical traction, with Gardner-Wells tongs or a halo ring, are safe and have been commonly used to help reduce and realign the spine.\textsuperscript{11,25,38} If closed reduction is successful, patients then undergo posterior fusion to maintain alignment. However, sometimes posterior reduction with cervical traction is unsuccessful, thus requiring a more focused and persuasive means of achieving normal alignment. Regardless of the methodology, reduction methods rely on relative mobility and the lack of fusion—congenital, autofusion, or iatrogenic. Preoperative CT scanning is crucial when planning intraoperative reduction.

The goals in treating cranial settling from the posterior approach are 1) to decompress the anterior pathology and 2) to create a stable spinal construct to prevent further insult to the spinal cord with neck movements. Cervical traction with the use of Gardner-Wells tongs or halo ring placement has been safe and very effective in achieving the first goal of decompressing the anterior pathology.\textsuperscript{5,30} In some cases, cervical traction has been used presurgically for up to 30 days before posterior fixation.\textsuperscript{34,40} Often, patient symptoms resolve with traction alone prior to surgery;\textsuperscript{12,13,20,25} in fact, traction alone without surgery has been successful.\textsuperscript{38} In our experience, preoperative traction is most useful in cases with severe kyphosis, to facilitate surgical approaches. In particular, we have used it successfully in cases of severe chin-on-chest deformity.

The pathology that causes cranial settling is important to differentiate from basilar invagination. Goel et al.\textsuperscript{13} noted that basilar invagination due to congenital phenomena was much harder to reduce with preoperative traction than cranial settling from acquired conditions such as RA. Many patients with basilar invagination in that series did not have reduction of the malalignment in preoperative traction. The rheumatoid disease process causing cranial settling results from degeneration of the ligaments and joints and eventually causes instability of the cranio-cervical junction. Generally, rheumatoid patients have a quasi-stable condition once cranial settling has occurred. The relative stability is mostly attributed to docking of the skull base on the spine, but vertical vectors will generally undock the skull from its position of immobility.

The second goal—having a stable posterior spinal construct—is relatively easy to achieve with our current technology in spinal hardware. Compared with prior wiring techniques, placement of current screw and rod constructs has proven to be the superior technique in spinal fixation.\textsuperscript{21,37} In the treatment of anterior cervical pathology, there are a limited number of cases involving the posterior-only approach. Distraction between the occipital plate and C-2 pedicle screws has been described.\textsuperscript{19} Additionally, others have described distraction between C-1 lateral mass screws and C-2 pedicle or pars screws, with placement of a spacer between the C1–2 articular space after distracting C-2 caudally away from C-1.\textsuperscript{1,2,12,21} Hsu et al. described a technique similar to ours; however, their two cases involved the treatment of cranio-cervical instability caused by a retropharyngeal abscess and bony destruction from a breast metastasis.\textsuperscript{37}

Rigid OC fusion constructs are not without risk. Screw pullout and subdural hematomas in the cerebellum from drilling the occipital bone are possible complications.\textsuperscript{40} To avoid screw pullout, long C-2 pars screws or C1–2 transarticular screws in addition to longer fusion constructs are options. Understanding the anatomy is crucial in placing the occipital plate in order to decrease the risks of subdural

\begin{figure}
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\caption{Case 2. Sagittal CT cervical spine (upper) showing preoperative atlantoaxial instability with sublaminar wiring. Lateral radiograph (lower) showing stable reduction and instrumentation at the 1-year follow-up.}
\end{figure}
hematoma caused by screw insertions. The occipital plate screw is optimally placed at the keel and at the occipital protuberance where screw lengths up to 16 mm can be placed. It is important to pay special attention when drilling lateral to the keel since the bone is only 3–6 mm thick.

Other complications associated with OC fusion include rod breakage, fusion failure due to disease progression from underlying pathology, poor bone quality due to age and medications (for example, chronic steroid use), pseudarthrosis, or vertebral artery injury from cervical screw placement. We assume that this patient population would have poor bone quality; thus it is beneficial to extend the spinal construct to use several fixation points to distribute the forces of movement and stabilization across multiple levels. Additionally, vertical reduction maneuvers do not provide pullout forces on the screws, and neither does anterior translation on C-2 provide pullout forces, but instead provides compressive loads. Ultimately, the surgeon would decide intraoperatively which maneuver to use on a case-by-case basis.

Careful patient selection is needed when dealing with cranial settling from RA. A bone density scan may be helpful in determining if a patient is severely osteoporotic, which could lead to an increased screw-pullout risk and instrumentation failure, thus warranting a longer OC construct. The postoperative use of external bracing and bone-growth stimulator may help with the fusion, as could nutritional supplementation and/or counseling.

Conclusions

Occipitocervical junction pathology can be difficult to manage, and there are many surgical options. We demonstrated that an intraoperative reduction technique was immediate and did not require prolonged bed rest while in external cervical traction. With the posterior-only approach, we accomplished indirect decompression of the spinal cord, provided a stable rigid fusion construct, and circumvented the morbidities associated with an anterior approach for nonfused cases of cranial settling.

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Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Joseph O’Brien is a paid consultant for Relivant, Globus Medical, NuVasive Inc., and Stryker; has received royalties from Globus Medical; has ownership of Spinicity; and has received clinical or research support from Globus Medical for the study described.

Author contributions to the study and manuscript preparation include the following. Conception and design: Young. Acquisition of data: Young, Sherman, O’Brien. Analysis and interpretation of data: Young, Wind, O’Brien. Drafting the article: Young, Wind. Critically revising the article: Young, Wind, O’Brien. Reviewed submitted version of manuscript: Young, Wind. Approved the final version of the manuscript on behalf of all authors: Young. Administrative/technical/material support: O’Brien. Study supervision: Litvack, O’Brien.

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