



Endoscopic endonasal approach to the craniovertebral junction

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Abstract

The surgical approach to lesions of the ventral craniovertebral junction (CVJ) has evolved significantly in the last several years with the advent of endoscopic skull base surgery. Differing pathologies of the CVJ can result in irreducible compression of the cervicomedullary region. The endoscopic endonasal approach lends itself well to this region due to the ventral location, and while there is a steep learning curve, is a safe and effective way to perform decompression of the cervicomedullary region. Herein, we discuss the anatomy of the CVJ, preoperative evaluation and surgical considerations, our surgical approach, complications, and outcomes.

KEYWORDS

basilar invagination, craniovertebral junction, endonasal surgery, endoscopic skull base surgery, odontoidectomy

INTRODUCTION

The surgical approach to lesions of the ventral craniovertebral junction (CVJ) has evolved significantly in the last 16 years. The traditional microscopic transoral transpharyngeal (TO) approach was and remains subject to several limitations inherent to the physical and optical characteristics of the microscope when working on a deep target through a narrow corridor. With the advent of endoscopic endonasal approaches (EEA), the possibility of a more direct approach allowing a brighter, more panoramic, and multiangled view with a greater range of motion became a reality. The first cadaveric study to prove the feasibility of an EEA approach to the odontoid/CVJ was published by Alfieri et al. in 2002.¹ This was shortly followed by the first clinical report of an endoscopic endonasal odontoidectomy by Kassam et al.² in 2005. Since those initial reports, the application of the endoscopic approach to the CVJ has been expanded to a greater number of indications and pathologies as the technical aspects have been more comprehensively described and the safety and efficacy of

the approach has been well established. EEA to the CVJ (EEACVJ) remains a challenging procedure with a steep learning curve and the potential for devastating neurovascular complications. As such, it is ideally suited for highly experienced endoscopic skull base teams consisting of a rhinologist and an endoscopic skull base neurosurgeon. Herein we discuss our institutional approach to the CVJ with a discussion of the anatomy, indications, preoperative evaluation/surgical considerations, an overview of the technical aspects of the surgical approach, alternative approaches, complications, and outcomes.

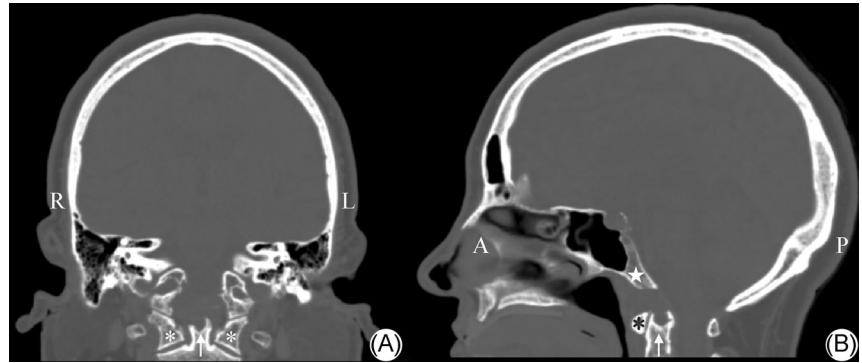
ANATOMY

The CVJ protects the brainstem, upper spinal cord, cranial nerve XII, spinal nerves, and vertebral arteries.³ It is comprised of the occipital bone as well as the first and second cervical vertebrae (C1 and C2, respectively). Superiorly is the foramen magnum where the medulla

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FIGURE 1 Coronal (A) and sagittal (B) computed tomography images of the craniovertebral junction. Note that C1 and the odontoid have been eroded from a lower clival chordoma (* denote C1, arrow denotes C2 and odontoid, ★ denotes clivus)



passes into the spinal canal and laterally, the occipital condyles (OC) connect C1 with the skull. The hypoglossal canals course through the OCs. The occipital bone, C1, and C2 are connected by multiple ligaments. To access the bony structures of the CVJ, both the anterior longitudinal ligament and the underlying atlantooccipital membrane must be opened. The anterior arch of C1 is typically encountered first, and as the posterior surface of the C1 arch joins with the odontoid process of C2, is a useful anatomic landmark. The relationship between C1 and C2 is shown in Figure 1. Finally, a complex system of ligaments comprised of the alar, apical, and cruciform ligaments surrounds the odontoid process, or dens, and must be sectioned to release the odontoid process and allow for removal.

INDICATIONS

Irreducible ventral cervicomedullary compression can be caused by several etiologies, including trauma, tumor, and inflammatory diseases such as rheumatoid arthritis (RA).⁴ Congenital and developmental processes, including atlanto-occipital hypoplasia, Chiari malformation, clival hypoplasia, os odontoideum, and platybasia can result in irreducible ventral compression as well.^{3,5} Many of these pathologies result in displacement of the odontoid process into the foramen magnum, resulting in compression of the cervicomedullary region, otherwise known as basilar invagination (BI).⁴ Presenting symptoms can include weakness, numbness, paresthesia's, neck pain, gait difficulty, spasticity, dysphagia, and dysarthria.^{6,7}

Forms of BI that are reducible can be treated with realignment via cervical traction followed by posterior arthrodesis/occipitocervical fusion.^{3,4} For irreducible BI, odontoidectomy is considered the gold standard treatment.^{3,4}

In the setting of degenerative spinal diseases such as RA, instability of this portion of the spine leads to the development of an inflammatory pannus around the odontoid, which can compress the brainstem.⁸ Typically, fixation through posterior arthrodesis is curative and results in regression of the pannus.⁹ When this is unsuccessful in regressing the pannus and compression persists, such as the case shown in Figure 2, the EEA can be utilized to resect the pannus allowing for decompression.

Finally, tumors such as chordoma can arise in this location. Chordomas can involve the skull base as well as the mobile spine.

Up to 23% of clival chordomas arise from the lower clivus, which has a very close relationship to C1 and the odontoid process of C2.¹⁰ Lower clival tumors can, therefore, involve these structures or can arise from C1 or C2 as demonstrated in Figure 3. Atlanto-occipital instability can occur as a result of bony destruction from the tumor or from surgical removal of the chordoma and around 3% of patients require arthrodesis after chordoma resection.¹¹

PREOPERATIVE EVALUATION/SURGICAL APPROACH CONSIDERATIONS

First, each patient being considered for endoscopic endonasal odontoidectomy (EEO) must have confirmation that the abnormality of concern is irreducible. This can be achieved through radiographic studies, including thin cut computed tomography (CT) scans and thin cut magnetic resonance imaging (MRI) of the skull base and CVJ. Second, the stability of the CVJ should be evaluated clinically and with flexion-extension films.³ If the CVJ is found to be unstable, before undertaking EEACVJ, it is recommended that either initial posterior fusion be performed or in the case of previous posterior fusion, that this be revised.³ It is critical in these situations to stabilize the CVJ before performing an endoscopic approach. Third, the patient's anatomy needs to be thoroughly reviewed and the surgical plan based on the origin of the brainstem compression. This is best done in a multidisciplinary setting with neuroradiologists, spinal surgeons, and the endoscopic skull base surgical team. In our experience, MRI with and without contrast provides adequate detail of the vascular structures and CT-angiography is not routinely obtained. The optimal surgical approach is dictated by each patient's anatomy. In certain congenital malformations, the position of the odontoid process is quite high and thus lends itself well to an EEA.¹²

The inferior-most limit of the EEA is considered to be the inferior aspect of the body of C2. Access to structures or lesions inferior to this is limited by the hard palate and may need to be addressed through a traditional transoral approach. Various anatomic "lines" have been defined that represent methods of measuring the inferior limit of the endoscopic approach on preoperative imaging. Perhaps most well-known is the nasopalatine line defined as a line extending from the nasal bones anteriorly to the hard plate posteriorly.¹³

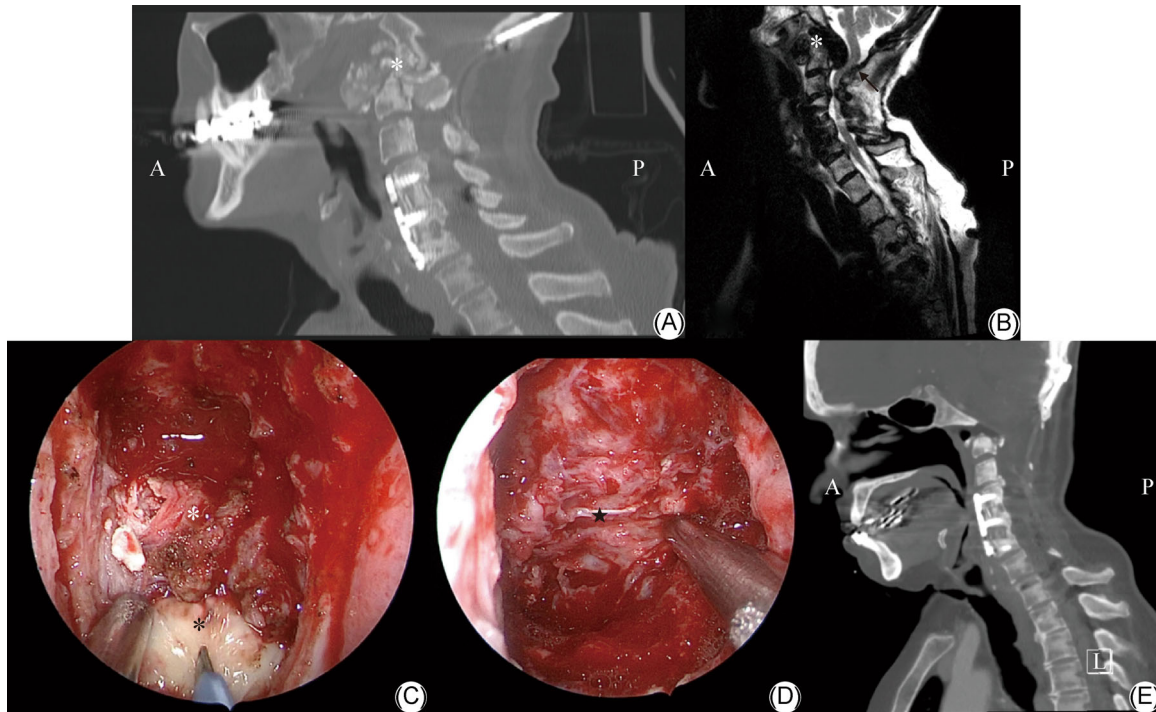


FIGURE 2 (A) Sagittal computed tomography (CT) showing an inflammatory pannus (*); (B) Sagittal T2 magnetic resonance imaging again demonstrating the inflammatory pannus (*) and the degree of cord compression (arrow); (C) Intraoperative picture of the inflammatory pannus (*); (D) Intraoperative picture after removal of the pannus and decompression (★ denotes dura); (E) Postoperative sagittal CT showing the resulting decompression

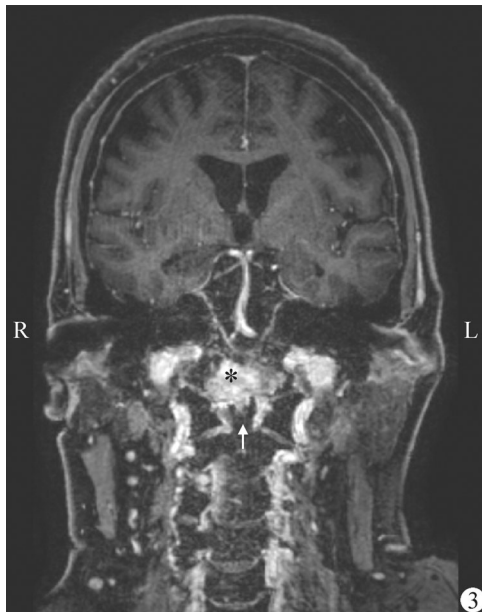


FIGURE 3 Coronal postcontrast T1 magnetic resonance imaging showing a chordoma involving the inferior clivus and C1 (*) with involvement of the odontoid process (arrow)

It is important to note that both head position and BI can affect the nasopalatine line.¹³ Specifically, maximal cervical flexion provides greater caudal exposure than cervical extension.¹³ Other lines defining the lower limit of the endoscopic approach include the

nasoaxial line and the rhinopalatine line.^{14,15} Each of these inferior limit lines can be measured out on preoperative imaging as part of the surgical approach planning phase. If it appears that some dissection caudal to the nasopalatine, nasoaxial, or rhinopalatine line will be necessary, then the surgical team should be prepared for the use of angled instruments or the possible need of converting to a transoral approach or an endoscopic-assisted transoral approach. If a substantial amount of decompression or tumor resection needs to be carried out caudal to the nasopalatine, nasoaxial, or rhinopalatine line, the EEA should not be considered.

SURGICAL APPROACH TO THE CVJ

EEO and EEACVJ are a few of the rare instances at our institution where we will pin the patient with the Mayfield retractor (Integra NeuroSciences Implants) as it allows for stabilization of the skull base and CVJ. Neuronavigation with both CT and MRI is utilized in every case while neuromonitoring is performed on a case-by-case basis.

The surgical procedure starts with outfracturing the bilateral inferior turbinates and lateralizing the middle turbinates. In patients with narrow nasal cavities, the posterior 1/3 of the inferior turbinates can be resected to provide more working space. It is important that thorough cauterization be done after the resection as the inferior turbinate branch of the sphenopalatine artery enters the posterior aspect of the inferior turbinate and can result in significant postoperative epistaxis if not addressed. Next, the posterior septectomy

is performed. Before resecting the septum, the inferior part of the nasoseptal flap (NSF) pedicle and the lower septal arm of the NSF can be delineated with a bovie. The size and height of the septectomy depend upon the size of the nasal cavities and the position of the odontoid process, respectively. In cases of platybasia, the clivus is oriented in a more horizontal configuration and the odontoid is displaced more superiorly than normal. In these cases, access may require drilling through the sphenoid floor and skeletonization of the clivus. Drilling the nasal spine down until it is flush with the nasal floor can improve access inferiorly.

Next, the mucosal incision can be planned. This can either be a straight cut down the midline, or an inverted u-shaped incision (Figure 4). The anatomic landmarks for the inverted u-shaped incision include the clivus superiorly, the eustachian tubes/fossa of Rosenmueller (FOR) laterally, and the C2 arch inferiorly. As the pharyngeal segment of the internal carotid artery is posterolateral to the eustachian tubes, and the FOR can be relatively deep and with redundant mucosa, it is the authors' preference to make the lateral incisions for the mucosal flap just medial to the FOR. Additionally, in an attempt to spare the underlying musculature as much as possible when raising a mucosa-only flap, it is the authors' preference to make the mucosal incision with a two surgeon, four-handed approach with the bovie in the right hand and suction in the left to provide traction/countertraction and to suction smoke. The depth of this incision can be difficult to reach with standard endoscopic instruments. We have found great success with the 6.5 cm protected tip bovie. After the mucosal flap has been elevated, a 2-0 or 3-0 silk suture can be placed endoscopically (Figure 4). The needle is cut and handed off and a

hemostat is placed through the oral cavity and into the nasopharynx. The suture is then grasped under endoscopic visualization by the hemostats (Figure 4) and then pulled through the oral cavity and secured to the drapes. This removes the mucosal flap from the field for the duration of the surgery. If a midline mucosal incision is felt appropriate, this is done in the same two surgeon, four handed technique.

Another option for the mucosal incision is the rhinopharyngeal flap (RPF).¹⁶ Similar to the NSF, the RPF is vascularized with its vascular pedicle based inferiorly and toward the midline.¹⁶ An inverted u-incision is made in the mucosa, similar to described above, and dissection is carried down to the level of the bone of the clivus.¹⁶ The flap is elevated within that plane as close to the bone as possible. This results in elevation of the basopharyngeal fascia and capitis muscle as part of the flap.¹⁶ Similar to described above for the inverted u-incision, the RPF is displaced inferiorly during the case. For closure, the RPF can be reflected back into its original location and affixed with sutures, fibrin glue, or packing.¹⁶

Following the mucosal cut, dissection is carried down through the underlying musculature. Of note, at the midline is the pharyngeal raphe, which represents a plane through which dissection can be carried cleanly and rapidly down between the longus coli and capitis muscles to the anterior longitudinal ligament and the bony spine. The use of the four-handed technique allows for greater ease in identifying this plane and more accurate dissection. The depth of the dissection is carried down to the bone of the anterior arch of C1. This represents a crucial bony landmark that can be reliably identified and confirmed with surgical navigation. Next, the muscle attachments

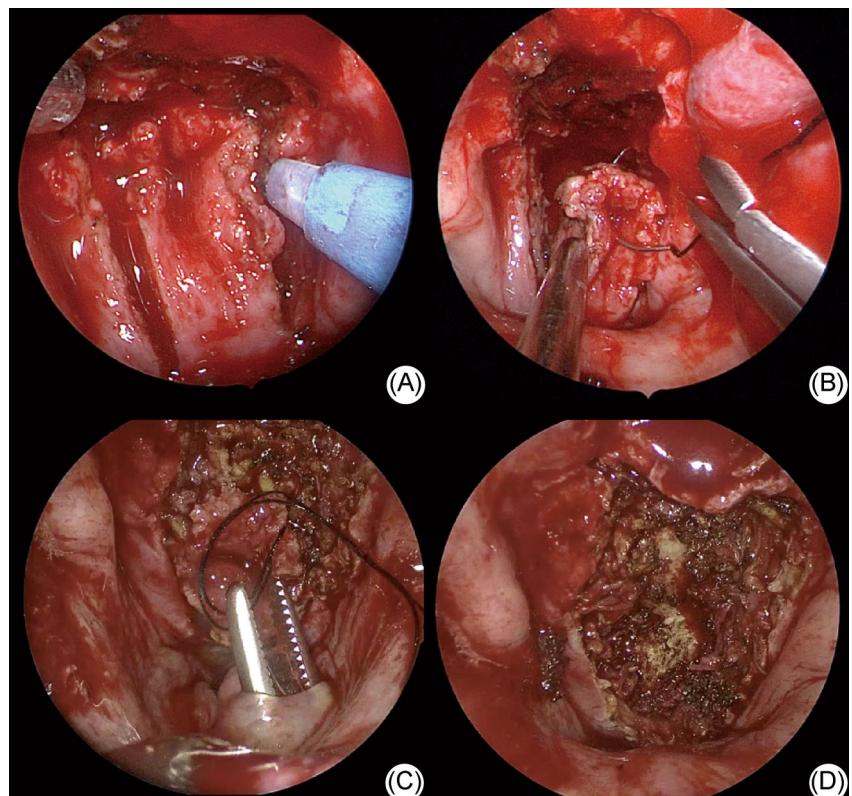


FIGURE 4 Creation and management of the mucosal flap (A): u-shaped mucosal incision; (B): endoscopically placing a silk suture; (C): grasping the suture with hemostats placed through the oral cavity; (D): after the flap has been displaced into the oropharynx

inserting on the clivus can be dissected starting at the midline and moving more laterally. These attachments are notably adherent and are best dissected with monopolar cautery. With the dissection of these attachments, the surgical corridor widens.

Once the exposure is felt to be adequate, the neurosurgeon takes over. For odontoidectomy, the C1 arch is resected using either a high-speed drill or the ultrasonic aspirator (Sonopet, Stryker). The amount of the C1 arch removed depends upon the position of the odontoid process relative to C1. In cases where sufficient exposure can be obtained without removal of any part of C1, it is our preference to preserve C1. It is important to not extend the resection laterally to include the OC as preservation of the OCs decreases postoperative instability, and also protects CN XII. Figure 5 shows the removal of the anterior C1 arch. Typically, between C1 and the odontoid process of C2 is a variable amount of inflammatory tissue or pannus. Dissection is carried down through this until the bone of the odontoid process is identified. Soft tissue attachments, including multiple ligamentous attachments, are dissected off the odontoid process and the odontoid is cored out centrally down to the inner cortex. This can be achieved with either a high-speed drill or an ultrasonic aspirator. Over time, our team has begun to prefer the ultrasonic aspirator as it has triple functionality (bone removal, suction, and irrigation) and creates less heat and energy transfer to nearby structures.

The remaining cortex is removed with Kerrisons leaving the cap of the odontoid. The residual cap is then carefully dissected from its ligamentous attachments and soft tissue with various dissecting instruments as shown in Figure 6. Once the cap is delivered, decompression is confirmed with visualization of dural pulsations. In different inflammatory disease processes, ligaments and surrounding soft tissue can become thickened and contribute to the compression. This pathologic tissue must be cut to provide optimal decompression.

When the indication for surgery of the CVJ is a tumor, the approach described above is utilized and altered accordingly to provide adequate access for tumor resection. At our institution, the most common tumor involving the lower clivus and CVJ we manage is chordoma. As noted earlier, chordomas of the lower clivus can often also involve C1 and chordoma can develop anywhere along the mobile spine. With chordoma surgery, the goals differ from odontoidectomy. With odontoidectomy,

the preservation of functional capacity and improvement of this is the main goal. While functional outcomes are just as important in chordoma surgery, completeness of tumor resection is also essential. It has been well established that gross total resection of chordomas significantly decreases recurrence rates and has a significant impact on overall survival.^{11,12} Therefore, in chordoma cases like the case demonstrated in Figure 3, the lateral extent of the tumor necessitated the removal of the bone up to the OC. Postoperatively, the patient in Figure 3 was immobilized in a cervical spine collar until they could be cleared via flexion/extension films. Fortunately, no instability was identified, and the patient did not require posterior arthrodesis.

Following resection, the dura is closely examined for any defects or evidence of cerebrospinal fluid (CSF) leakage. If a low flow CSF leak is noted, a dural substitute can be placed over the leak and secured in place with dissolvable packing and/or fibrin glue. When a linear mucosal incision had been made, this can be reapproximated with 4-0 vicryl sutures placed endoscopically. The u-shaped mucosal flap or RPF can be repositioned over the defect and either secured in place with endoscopically placed 4-0 vicryl sutures as shown in Figure 7, or with fibrin glue or packing.

High flow CSF leaks of this region present a reconstructive challenge. Similar to other regions of the skull base, a multilayered closure is preferred. Dural onlay grafts or fat can be used as the initial layer of the reconstruction depending on the size of the defect cavity. A NSF can then be harvested and reflected over the defect and secured in place with packing. Postoperative lumbar drainage can be utilized depending on the surgeon's preference.

In the absence of a CSF leak, the reconstruction can vary from coverage with dissolvable packing +/- fibrin glue to the closure of the mucosal flap. Our preference is to reapproximate the mucosal cuts with 4-0 vicryl sutures whenever possible.

POSTOPERATIVE CARE

At our institution, patients are extubated before leaving the operating room unless there are specific circumstances that dictate otherwise. Each patient receives a CT scan of the head and cervical spine postoperative

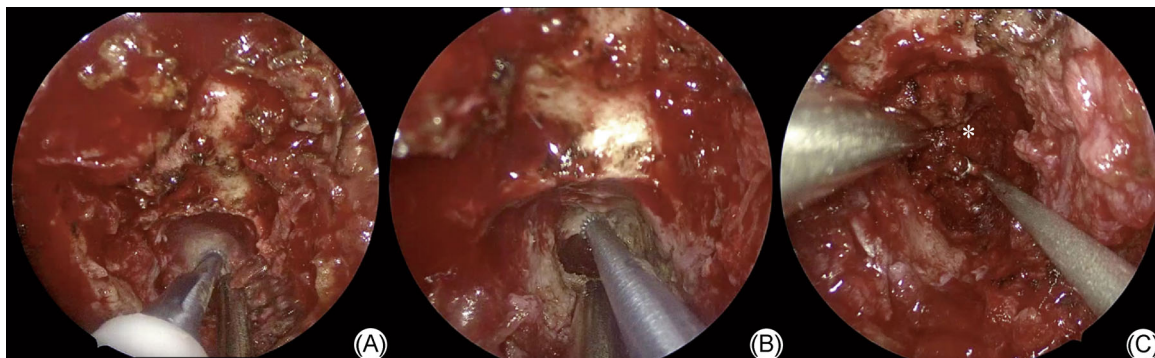


FIGURE 5 Progressive removal of the C1 arch in (A) and (B) down to inflammatory tissue (* in C) found between C1 and the odontoid process

FIGURE 6 (A) Drilling the odontoid to the inner cortex; (B) Removal of the inner cortex (*); (C-E) Dissection and delivery of the odontoid cap (arrow); and the decompressed dura (★ in F)

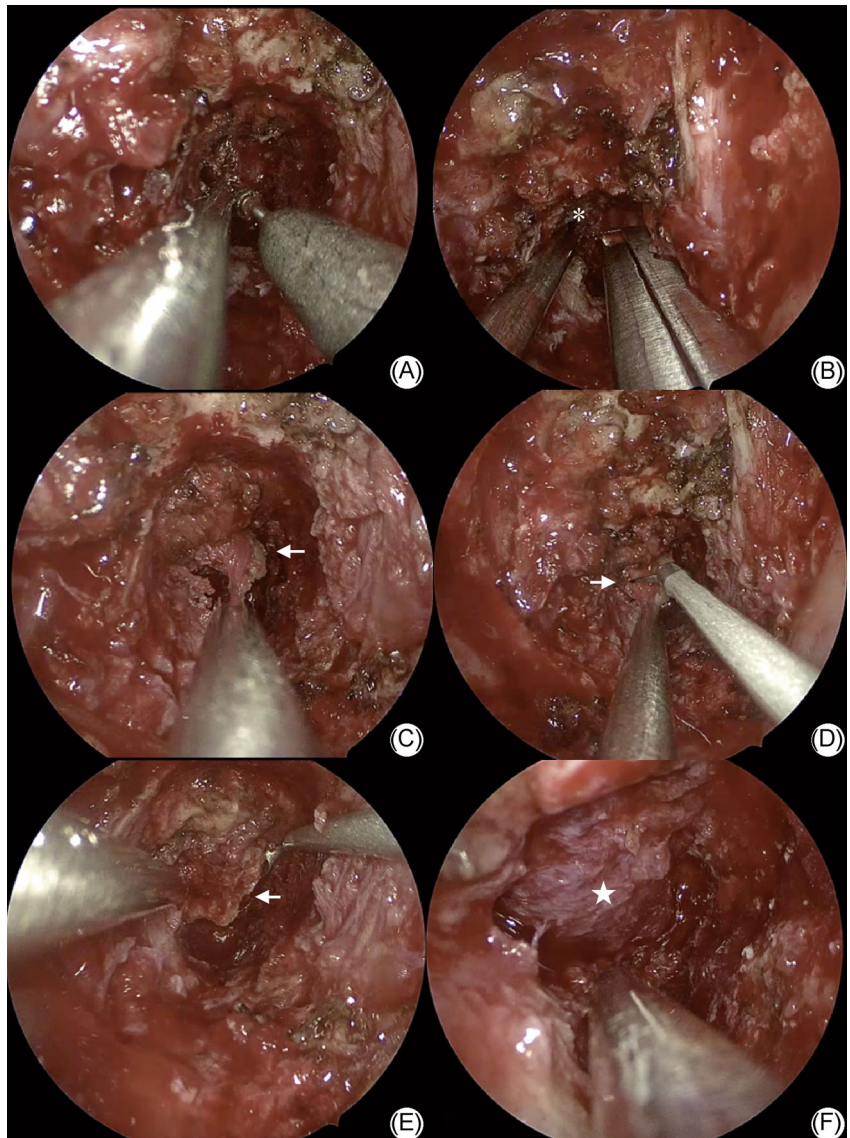
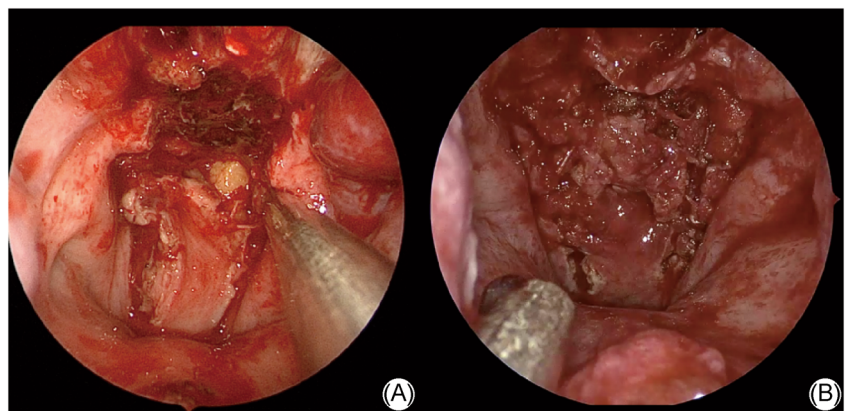


FIGURE 7 (A, B) Reapproximation of the mucosal flap in two different cases



day (POD) 0 to confirm adequate resection of the pathologic process and adequate decompression. In patients who have not had previous posterior fusion, the neck is immobilized with a cervical collar until clearance with flexion/extension films. If flexion/extension films suggest instability

of the CVJ, patients will undergo a posterior fusion during the same hospitalization. Patients at our institution begin taking po POD 1. Any patients with pre-existing dysphagia undergo a bedside swallow evaluation on POD 1 before taking po unsupervised.

Patients are discharged from the hospital after they are able to take adequate po and their pain is well controlled. In cases with no intraoperative CSF leak who did not undergo posterior fusion during the same hospitalization and do not have any evidence of instability postoperative, discharge occurs on POD 2 or POD 3 typically. Factors that extend the length of stay include intraoperative CSF leak, posterior fusion before EEACVJ during the same hospitalization, or the need for fusion due to new-onset instability post-EEACVJ. At our institution, patients that have known or pre-existing CVJ instability undergo posterior fusion 48–72 h before the EEA. All patients with new post-EEACVJ instability undergo posterior fusion 48–72 h after the endoscopic approach.

ALTERNATIVE APPROACHES

As mentioned previously, if the patient's anatomy renders the endoscopic approach not feasible, an alternative approach will need to be considered. The following includes a brief description of each of these alternatives.

The transoral approach is set up in a similar fashion as the EEACVJ with the patient supine and their head affixed in the Mayfield. A transoral retractor is then placed to retract the tongue and to open the oral cavity widely to provide exposure to the posterior pharyngeal wall. Care must be taken while placing the retractor to avoid injury to the teeth, pressing the tongue against the teeth, and putting too much pressure on the tongue. It is generally recommended that the oral retractor be periodically relaxed to release the compression on the tongue. Palpation is performed along the posterior pharyngeal wall to identify the tubercle of C1. Depending on the relationship of the uvula with C1, the soft palate may require retraction. This can be achieved with the use of flexible catheters passed transnasally and brought out through the oral cavity and secured above the upper lip. A midline mucosal incision is then made with monopolar cautery and the identification of anatomy and dissection is carried out in a similar fashion as described above.

To avoid splitting the palate into cases where the lesion extends above the reach of the TO approach, either an endoscopic TO approach (ETOA) or a transnasal and transoral approach (TNTOA) can be utilized. In the ETOA, the endoscope is passed through the oral cavity and under the retracted soft palate to help visualize and access the area above the soft palate. A TNTOA incorporates both the EEA and TO approaches to allow for decompression both above and below the palate.

Finally, the transpalatal approach (TPA) can be used. In the TPA, the entire hard palate is disarticulated. This involves a sublabial incision to expose the bony hard palate on the nasal side with submucosal dissection of the nasal floor and septum. A mucosal incision is then made on the inferior aspect of the hard palate continuing through the soft palate. The mucosa is elevated subperiosteally to the alveolar margin. A bony cut is then made around the margin of the palate near the alveolar ridge and the hard palate is separated from the nasal septum and lateral nasal wall through the sublabial incision.

The bone is removed from the oral cavity for the duration of the surgery and is reattached during the closure with plates. This is followed by reapproximation of the mucosal cuts along the palate in addition to the closure of the sublabial incision.

COMPLICATIONS AND OUTCOMES

Compared to the traditional transoral or TPAs, endoscopic approaches have been shown to result in shorter time spent on the ventilator/earlier extubation, lower rates of tracheotomy, earlier postoperative po intake, and shorter hospital stays.^{6,7,17} Certainly the EEA negates the need for prolonged tongue retraction and splitting of the soft or hard palates, which can result in upper airway edema and velopalatal insufficiency and dysphagia, respectively.

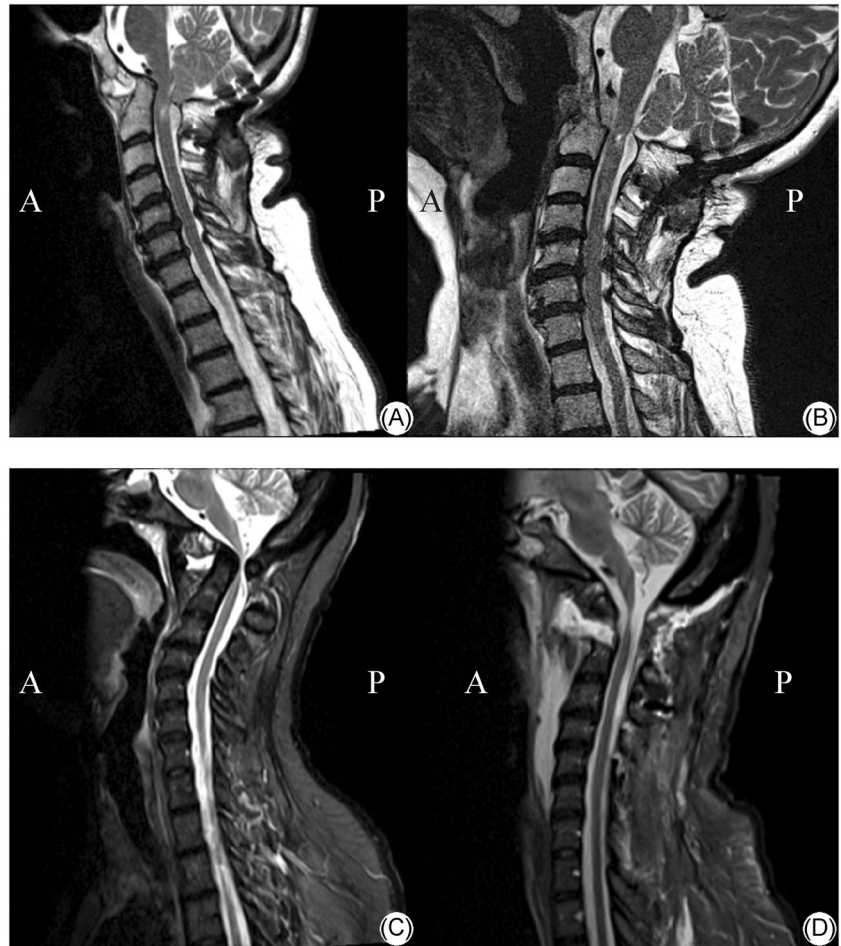
Series published in the literature have shown that the majority of patients undergoing EEA can be successfully extubated by POD 1 and the rate of postoperative tracheostomy is significantly lower after the endoscopic approach compared to TO or TP.^{6,7,18} Regarding dysphagia, Van Abel et al.¹⁹ demonstrated that by keeping the incision of the nasopharyngeal musculature above the palatal line, the EEA decreases the risk of dysphagia by decreasing the risk of damage to the neural plexus within the oropharyngeal walls critical for swallowing. Interestingly, some series in the literature still describe transient postoperative dysphagia in 47%–62% of patients following EEA.^{6,17} It is unclear what the underlying mechanism for the dysphagia was in these series. The rates of intraoperative and postoperative CSF leaks following EEA have been reported as between 24%–30% and up to 5.2%, respectively.^{6,7,20}

The outcomes following EEA for BI are impressive and equal to those seen with traditional approaches. Following EEA for BI, 78%–89% of patients experience neurologic improvement.^{6,20,21} Improvement in quality of life (QOL) following EEA, specifically for odontoidectomy has been demonstrated as well.^{17,21,22} Based on one study, nasal-specific QOL does not appear to be negatively impacted by EEO.²¹ Figure 8 shows the pre- and postoperative results in two patients.

To help reduce sinonasal morbidity following EEACVJ, several steps can be taken starting intraoperatively. Careful evaluation of the sinonasal passages at the end of the case to confirm adequate hemostasis can decrease the risk of postoperative epistaxis requiring nasal packing or a return to the operating room. If the posterior 1/3 of the inferior turbinate is resected, as stated previously, this should be cauterized thoroughly to prevent postoperative epistaxis from the inferior turbinate branch of the sphenopalatine artery. For the reconstruction, the use of absorbable packing over rigid or nonabsorbable packing can reduce the level of postoperative discomfort.

Postoperatively, saline sprays or saline rinses should be initiated relatively early to provide symptom relief and mechanical debridement of sinonasal cavities. An initial debridement is performed at 1 week postoperative followed by additional debridements if necessary. Both saline rinses and postoperative debridements have

FIGURE 8 (A) and (B) demonstrate the preoperative (A) and postoperative (B) T2 sagittal magnetic resonance imaging (MRI) of a 62-year-old patient with basilar invagination (BI). Preoperative, the patient was wheelchair-bound. Four months postoperative, she is walking with the assistance of a walker and continues to work on her recovery. (C) and (D) show the preoperative (C) and postoperative (D) T2 sagittal MRI of a 17-year-old woman with BI who had been experiencing progressive loss of sensation and strength in her upper extremities. Eighteen months after surgery, she has had a complete recovery



been shown to reduce the risks of scarring, synechiae formation, and ostial stenosis after endoscopic sinus surgery.²³

CONCLUSIONS

The endoscopic approach to the CVJ is a safe alternative to traditional approaches with favorable outcomes and low rates of complications. There is a steep learning curve, as is the case with all endoscopic skull base approaches, and therefore it is important that the surgical team be well experienced in endoscopic approaches. Appropriate patient selection is essential as is the evaluation and management of CVJ instability and therefore requires a multidisciplinary team.

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None.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

AUTHOR CONTRIBUTIONS

Ashleigh A. Halderman and Samuel L. Barnett: Writing, review, and editing of the paper.

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