The Endoscopic Endonasal Approach for Craniovertebral Junction Pathology

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ABSTRACT

The craniovertebral junction (CVJ) can be affected by a variety of disease processes, which at times may cause irreducible ventral compression that is best addressed by an anterior approach. Over the past decade, the endoscopic endonasal approach (EEA) has emerged as a viable alternative to the established microscopic transoral approach. Published experience using the EEA for CVJ pathology has grown steadily, and a number of anatomic studies have helped to further refine the technique and define its indications and limitations. In this article we provide a description of the surgical technique and present an illustrative case. The indications, complications, and neurologic outcomes of the EEA are discussed, and a comparison is made between the EEA and alternative anterior approaches. Potential complications with the EEA include airway problems, velopharyngeal insufficiency, dysphagia, cerebrospinal fluid leak, and infection.

KEY WORDS: Craniovertebral junction, craniovertebral junction pathology, endoscopic endonasal approach

INTRODUCTION

Pathology of the ventral craniovertebral junction (CVJ) poses significant difficulty with respect to surgical access. Anatomic abnormalities, atlanto-axial instability, basilar impression, or space occupying lesions of the CVJ can cause symptoms through mechanical instability or neural compression. Common causative etiologies include rheumatoid arthritis, fractures, tumors, infection, Chiari malformation, achondroplasia, os odontoideum, osteogenesis imperfecta, Down syndrome, and other congenital anomalies.

A variety of surgical approaches have been used to access the ventral CVJ. Posterior and posterolateral approaches to this region include the midline suboccipital, far lateral, and direct lateral (1) approaches. However, the ventral corridor has afforded the most direct approach to this challenging region. The first transoral approach to the CVJ was reported by Kanaval in 1917 (32) but was used infrequently until the 1970's and 80's when the technique was further refined. The first large case series involving 17 cases was published by Menezes in 1980 (45). Other reports soon followed, and the transoral approach became a well-established and widely used approach to the ventral CVJ, with one published case series including 533 patients (9). Although the transoral approach has provided an effective route for ventral decompression, it has several drawbacks, including limited superior extent of exposure, contamination from oral flora, and delayed resumption of oral feeding, risk of velopharyngeal insufficiency (VPI), prolonged postoperative intubation and occasional need for tracheostomy.

Endoscopy is a relatively new technique in cranial base surgery. Jankowski first reported an endoscopic transsphenoidal approach for treatment of pituitary tumors in 1992 (30). Jho and Carrau adopted the endoscopic approach in 1993, and in 1997 they published the first large case series of endoscopic endonasal pituitary surgery (31). Since then, there has been increased adoption of endoscopic approaches for pituitary tumors. Further modifications have resulted in "extended" EEAs that provide access to the ventral midline skull base along a wide rostral-caudal axis, extending from the frontal sinus to the C2 vertebral body.

The success of other endoscopic endonasal procedures eventually led to the development of an EEA to the CVJ (34). In 2002, Alfieri published a cadaveric study that demonstrated the feasibility of the approach and the extent of rostral-caudal exposure it could provide (3). Kassam provided the first clinical case report of an EEA to the CVJ in 2005 (33). The publication of numerous case reports and small case series soon followed (5–7,10,11,16,19–23,25–27,29,33,35,37,38,40–44,47–50,53,55–57,61,62,64–66,69,70,75,78,80–83), as did numerous anatomical studies that used cadavers or imaging to further refine the approach. Two systematic reviews were recently published which identified over 90 cases reported across 32 studies (17,18).

ENDOSCOPIC ENDONASAL APPROACH to the CRANIOVERTEBRAL JUNCTION

Patient Selection and Preoperative Evaluation

Appropriate preoperative planning is key to ensuring success with an anterior approach to the CVJ. Preoperative imaging studies should ideally include MRI (to assess the degree of neural compression) and CT (to assess bony anatomy). Cervical flexion/extension x-rays may be helpful to evaluate the stability of the CVJ. Attention should be given to the location of the pathology relative to the hard palate, as this dictates whether adequate decompression can be achieved by the planned approach (Figure 1). Volumetric MRI and CT are helpful for the purpose of intraoperative navigation.

CVJ stability must be considered during the preoperative planning, because significant instability may be present preoperatively as a result of an underlying destructive pathology, or it may result from the surgical approach itself. When faced with a patient presenting with symptomatic compression of the brainstem or upper cervical spinal cord secondary to CVJ pathology, an initial attempt should be made at non-operative reduction if applicable to the pathology. For potentially reducible pathologies, traction can be applied using Gardner-Wells tongs over the course of several days. If adequate reduction is achieved, then a posterior instrumented fusion may be all that is indicated. If there is non-reducible deformity or continued ventral compression, then it is appropriate to proceed with anterior decompression. We typically favor posterior instrumented fusion first to avoid any period of instability that is created by anterior decompression. A full discussion of the indications and techniques for reduction, fixation, and fusion in the setting of CVJ instability is beyond the scope of this paper.

Surgical Technique

The patient is placed under general anesthesia. Neuromonitoring with motor and somatosensory evoked potentials is utilized. The navigation system is registered. A portion of the abdomen is prepared for possible fat graft harvest. Intravenous antibiotics are given for infection prophylaxis, and the nasal mucosa is infiltrated with lidocaine and epinephrine. We perform all of our endoscopic endonasal surgeries in conjunction with rhinology colleagues, using a two-surgeon, four-handed technique. After bilateral inferior turbinate outfractures, endoscopic endonasal access to the nasopharynx is obtained via both nares. Additional maneuvers such as middle turbinate resection and posterior septectomy may be considered for cases with significant superior and lateral extension but are not commonly needed to approach the CVJ. The inferior nasal meatus leads directly to the nasopharynx overlying the CVJ (Figure 2A). The Eustachian tubes serve as important landmarks as they lie at approximately the level of the occiput-C1 junction and correspond to the lateral limit of exposure (Figure 2B).

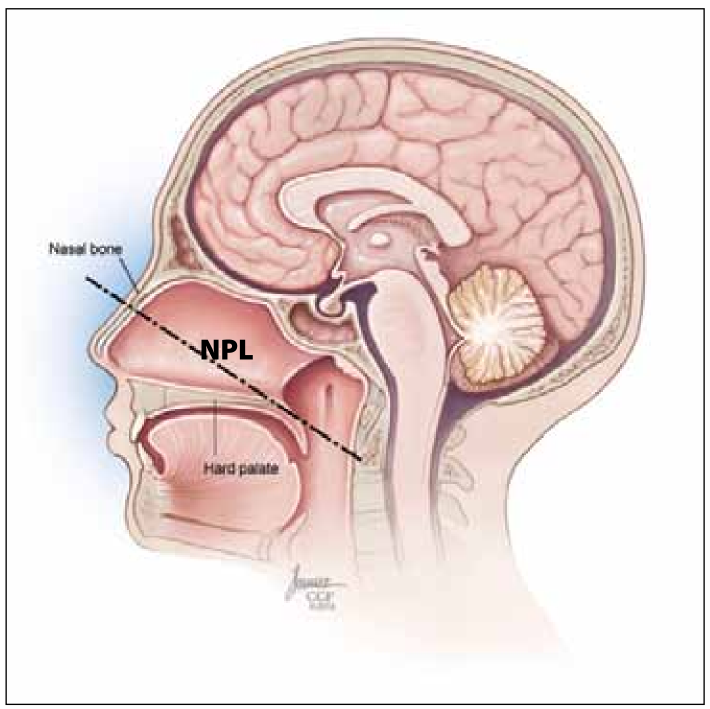

Figure 1: Illustration demonstrating the endoscopic endonasal approach to the craniovertebral junction. The dashed line indicates the nasopalatine line (NPL), which is limited by the nasal bone anteriorly and the hard palate posteriorly. The NPL approximates the inferior limit of exposure with this approach, and usually corresponds to the mid-C2 level, below the base of the dens.

The anterior tubercle of C1 can usually be palpated through the mucosa, and the navigation system is used to confirm the desired location. While some surgeons have described an inferiorly based U-shaped flap (33), we prefer to make a vertical linear incision in the midline using monopolar cautery (Figure 3A). The myomucosal layer is dissected off the ventral surface of the spine in subperiosteal fashion to expose the C1 anterior arch, dens, and lower clivus (Figure 3B).

The C1 anterior arch is removed using a high speed electric drill (Figure 3C). The odontoid process is then hollowed out using the drill (Figure 3D), and its remaining posterior cortical shell and ligamentous attachments are removed with a Kerrison rongeur or microdissectors (Figure 3E). The inflammatory pannus is removed, and visualization of the dura verifies adequate decompression (Figure 3F). If a cerebrospinal fluid (CSF) leak is encountered, it is repaired with a dural substitute or fascia lata, and fibrin glue. A fat graft can be utilized to fill significant dead space, and a foley balloon can be inflated in the nasopharynx to bolster the repair. Cerebrospinal fluid diversion via a lumbar drain is an option if a significant leak is encountered.

Postoperative Care

Patients are extubated immediately if possible. A postoperative CT is helpful to judge the adequacy of bony decompression. Prophylactic antibiotics are used for 24 hours postoperatively or for the duration of time period that nonabsorbable packing is in place. In the event of a CSF leak, lumbar drainage of CSF is performed for 3-5 days to minimize the risk of a postoperative leak. After assessment of swallowing function, a clear liquid diet is started on the first postoperative day and advanced as tolerated. Aggressive, early mobilization is highly encouraged. Following discharge, post-operative visits are used to assess for adequate wound healing, CVJ stability, and posterior bony fusion. A detailed review of postoperative care following endoscopic skull base surgery has been provided by Tien et at (68).

Illustrative Case: Endoscopic Endonasal Odontoidectomy

A 91-year-old man presented with bilateral arm paresthesia and weakness, unsteady gait, and impaired fine motor control in his hands. His symptoms had been present for several years and he had previously declined surgery but reconsidered after significant progressive neurologic deterioration. An MRI of the cervical spine showed a pannus dorsal to the odontoid process causing severe compression of the spinal cord and lower brainstem (Figure 4A). The degree of compression had progressed compared to an MRI performed several years previously. Decompression and stabilization were recommended.

Posterior instrumented fusion and combined anterior and posterior decompression of the craniocervical junction was done in two stages. Stage I consisted a midline suboccipital craniectomy, C1 laminectomy for posterior decompression, and occiput-C5 instrumented fusion using an occipital plate and screw/rod construct. Two days later he underwent Stage II, which consisted of an EEA and odontoidectomy as described above. The C1 anterior arch and odontoid process were removed, and the inflammatory pannus was resected. An incidental CSF leak was encountered through a pinhole dural opening. This was repaired using a collagen matrix dural substitute (Integra Lifesciences, Plainsboro, NJ), a small piece of which was placed into the dural opening as a "bath plug," with a larger piece placed in onlay fashion. This

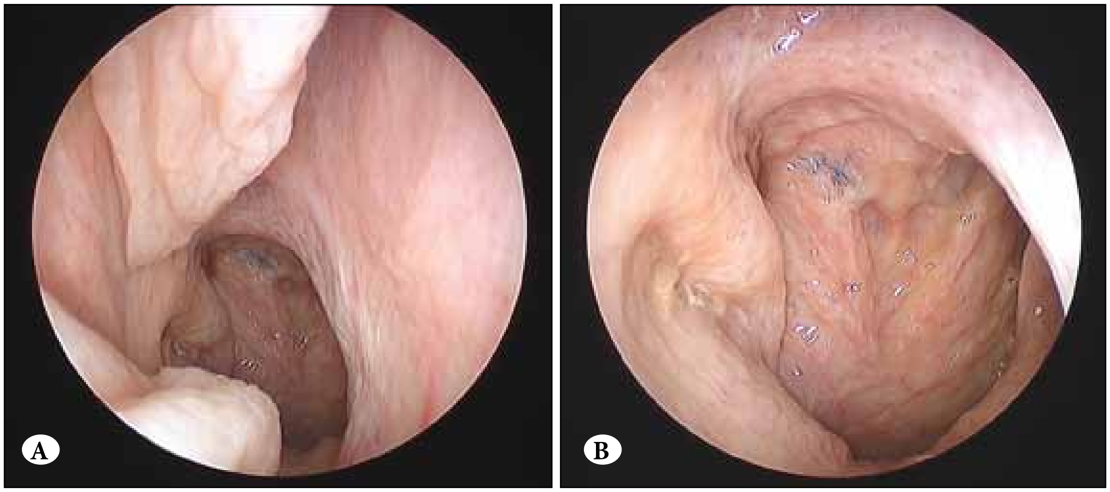

Figure 2: *Cadaveric images of key landmarks in approach the CVJ via an EEA.* ***A)*** *As the endoscope is introduced and the nasal floor followed posteriorly, the first important landmark is the choana (Ch), which represents the transition between the nasal cavity and the nasopharynx.* ***B)*** *After entering the nasopharynx, the midline corridor is identified between the Eustachian tubes (ET).*

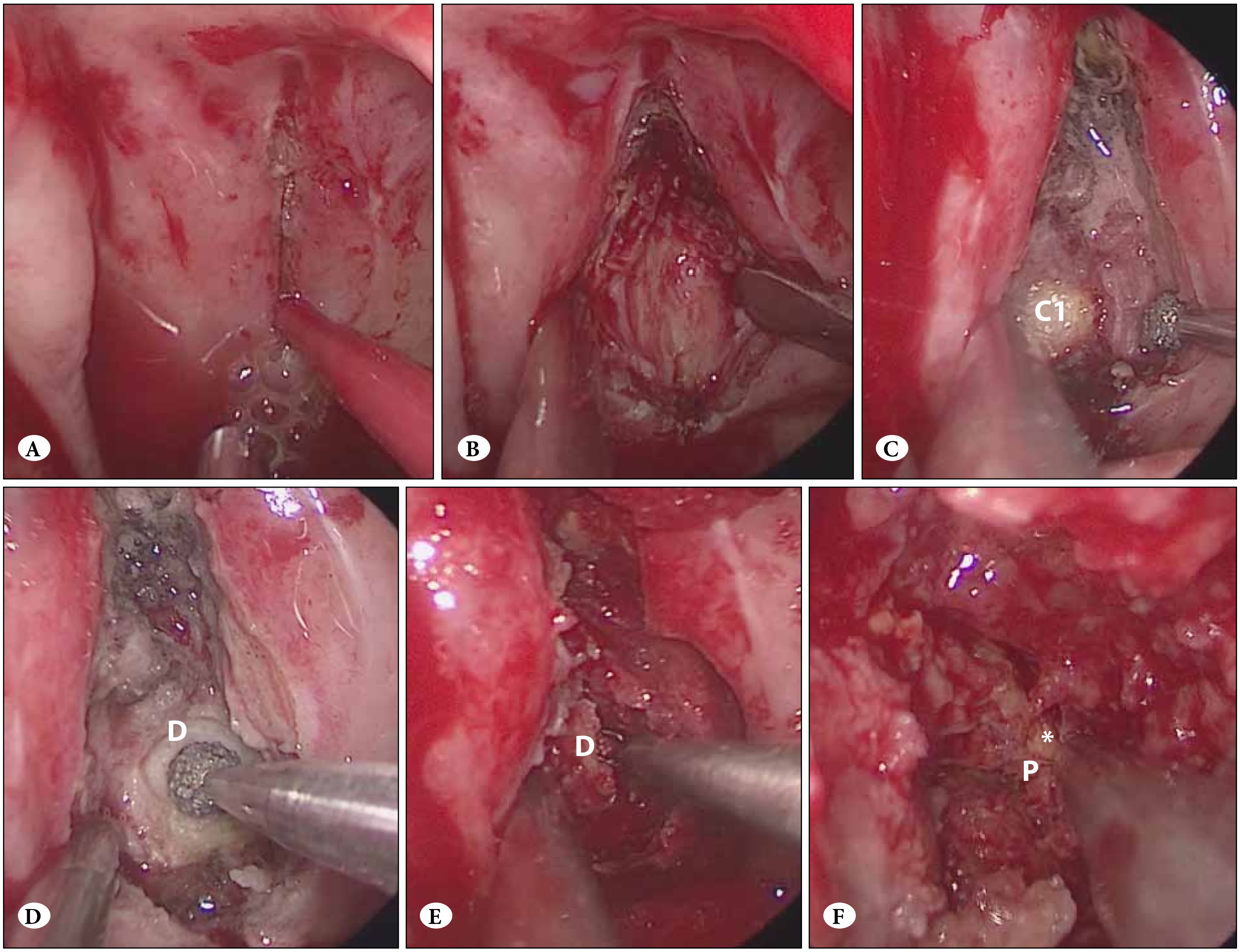

Figure 3: A) *A midline linear incision is made through the mucosal, muscular, and ligamentous layers using monopolar cautery.* ***B)*** *The anterior C1 and C2 vertebrae are exposed by subperiosteal elevation of the prevertebral muscles using a Cottle elevator. C) The C1 anterior arch (C1) is removed using an electric drill with diamond-tipped burr.* ***D)*** *After removal of the C1 anterior arch, the dens (D) is cored out using the drill, leaving a shell of cortical bone dorsally.* ***E)*** *The remaining cortical bone of the dens (D) is removed using a Kerrison rongeur.* ***F)*** *After removal of the dens, the inflammatory pannus (P) is removed using a microdebrider. Visualization of the dura (*) verifies adequate decompression.*

was covered with a layer of fibrin glue (Baxter, Deerfield, IL). An abdominal fat graft was harvested and packed into the odontoid defect, and foley balloon was inflated in the nasopharynx to bolster the repair. A lumbar drainage catheter was inserted at the end of the case.

The patient was extubated at the end of the case. A postoperative CT of the cervical spine showed adequate bony decompression (Figure 4B). He was ambulating with assistance the day after surgery. A clear liquid diet was started on the first postoperative day, and he was tolerating a regular diet by the end of the first week. The nasal foley and continuous lumbar drainage were continued for three days. There was no sign of CSF leak postoperatively, and he was discharged to a skilled nursing facility after nine days. Histopathology from the resected C2 pannus showed fibrocartilage with degenerative changes and deposition of calcium pyrophosphate crystals, consistent with a diagnosis of pseudogout. He experienced significant improvement in his gait and right hand motor function within six weeks of surgery, and an MRI six months later showed resolution of the ventral cervicomedullary compression (Figure 4C).

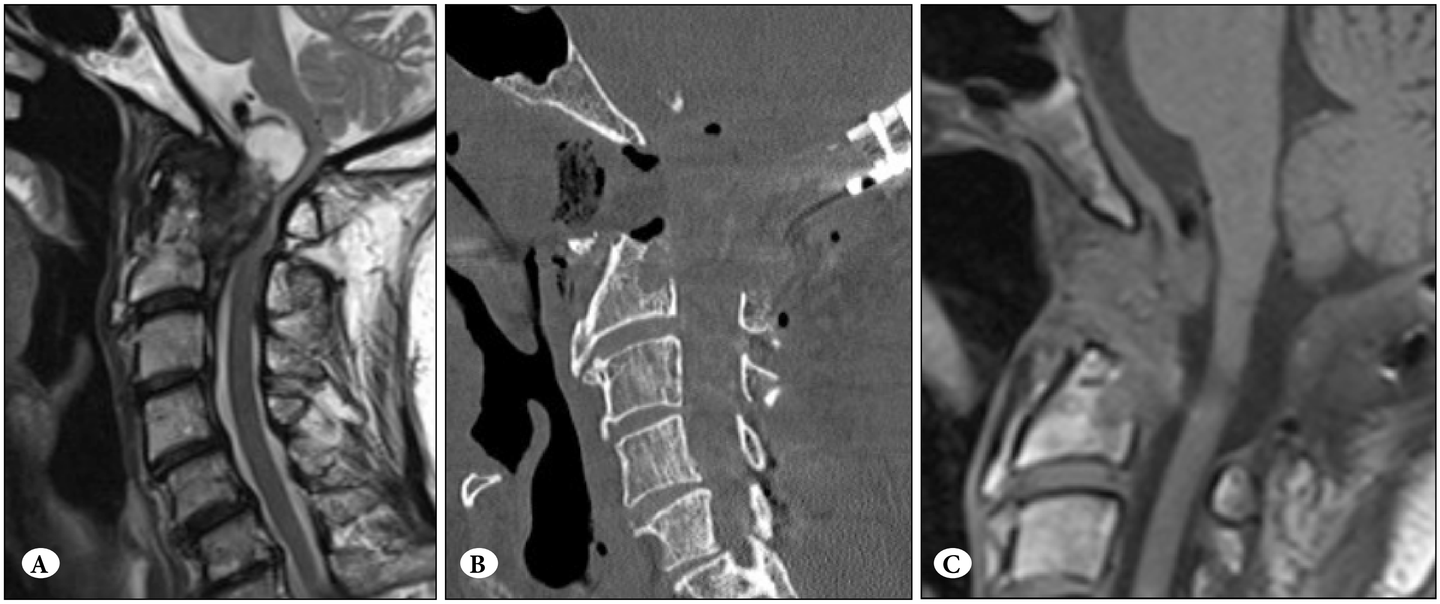

Figure 4: ***A)*** *Preoperative MRI (sagittal T2-weighted image), showing an inflammatory pannus dorsal to the odontoid process and compressing the upper cervical spine and lower brainstem. The patient had signs and symptoms of myelopathy.* ***B)*** *Postoperative image of the same patient following staged dorsal decompression and instrumented occipito-cervical fusion, followed by endoscopic endonasal odontoidectomy. This sagittally reconstructed CT of the cervical spine demonstrates adequate anterior bony decompression by the endoscopic endonasal approach, with interval removal of the dens and the C1 anterior arch.* ***C)*** *Six-month post-operative MRI (sagittal T1-weighted image) showing resolution of the ventral compression.*

DISCUSSION

Endoscopic Endonasal Compared to Alternative Approaches

A variety of anterior and posterior approaches are available to reach pathology of the CVJ. Anterior options include microscopic transoral, endoscopic transoral, endoscopic endonasal, and transcervical retropharyngeal approaches. Comparison of EEA to the other anterior approaches focuses on two main aspects: surgical exposure and complications.

We will focus on the comparison of the EEA to the microscopic transoral approach as the "gold standard," but two other techniques that deserve consideration are the endoscopic transoral and the endoscopic transcervical approaches. Welch and colleagues reported using an endoscope-assisted approach in 2001 (73), and the first clinical report of a fully endoscopic transoral approach was made by Hussain in 2006 (28). Three clinical case series have been reported using a purely endoscopic transoral approach to the CVJ, with a combined 50 patients (28,54,77). The endoscopic transcervical approach has been described by Wolinsky and colleuages (74), who used a tubular retractor to perform an odontoidectomy in 15 patients with basilar invagination, resulting in significant neurologic improvement (14). Wu et al. also used an endoscopic transcervial approach to successfully perform ventral release and instrumentation in 10 patients with irreducible atlanto-axial dislocations (76).

Surgical Exposure

Based on studies that have directly compared exposure provided by the EEA to that of the transoral route (4,39,58,72), it can be concluded that the EEA can reach higher along the clivus but may be limited in its inferior reach compared to the transoral route. The standard microscopic transoral approach can reach as high as the lower third of the clivus, and this superior exposure is limited by the posterior palate and the mandible. The inferior limit of this approach, the lower margin of C2, is imposed by the posterior tongue (12,46). Modifications to the transoral approach include palatotomy or maxillotomy, which increase the superior limit of exposure; and mandibuloglossotomy, which increases both the superior and inferior limits (79).

The endonasal corridor, on the other hand, provides essentially unlimited superior exposure but is limited in its inferior exposure. This inferior limit of exposure via the EEA is dictated by the posterior hard palate and the nasal bone and cartilage. De Almeida and colleagues defined the nasopalatine line (Figure 1) as a method for predicting the inferior-most exposure based on preoperative imaging (15). A subsequent cadaveric study by Aldana et al. suggested that the nasopalatine line may overestimate the inferior exposure and as an alternative proposed the naso-axial line, which connects the posterior edge of the hard palate to a point midway between the rhinion and the anterior nasal spine (2). Based on clinical experience, La Corte and colleagues proposed the rhinopalatine line, whose anterior point lies one third of the way from the anterior nasal spine to the rhinion (36). It should be noted that these studies utilized 0-degree endoscopes, and the use of angled-lens endoscopes provides lower exposure (60). Development of angled drills and instrumentation enhanced our ability to work with the visualization provided by angled endoscopes.

Applying the endoscope to the transoral approach could neutralize some of the benefits of the transnasal over the transoral route, perhaps by providing increased visualization or by reducing the need for soft tissue retraction. Pillai et al conducted a cadaveric study utilizing a 30-degree endoscope in a transoral approach.They found that a larger working area and higher clival exposure are obtained compared to the microscope, without needing to split the palate (52). Inferiorly, the endoscopic transoral route provides greater exposure than can be obtained transnasally. In addition, Visocchi and co-workers demonstrated that the endoscopic transoral route provides wider working angles in the sagittal and axial planes compared to the EEA in a cadaveric study (72). Given that they provide overlapping but unique areas of exposure, the transnasal and transoral routes should be viewed as complementary approaches, which can even by used simultaneously with the endoscope during the same surgery (65,69).

Complications

Various complications have been associated with anterior approaches to the CVJ, and a thorough consideration of these complications is necessary when considering the indications for surgery and the choice among multiple surgical approaches. The desire to minimize morbidity was a major motivation behind the adaptation of the EEA to the CVJ. There have been two systematic reviews which have examined complications rates associated with this approach (17,18), and a recently published meta-analysis compared complication rates between EEA and transoral approaches (59).

Respiratory complications are a concern with any anterior craniocervical approach. A systematic review by Fujii et al. found that the mean time of post-operative intubation for transnasal cases was 0.54 days, that tracheostomy occurred in five of 57 (8.8%) cases, and two of 57 (3.5%) patients required re-intubation (18). A similar systematic review by Fang and colleagues found a 7% rate of respiratory failure among 85 cases, with 62% of patients extubated on the day of surgery and only 16% requiring intubation longer than one day (17). A meta-anlaysis by Shriver et al. evaluated differences between transnasal and transoral surgeries. Need for postoperative tracheostomy was the only complication with a statistically significant difference (10.8% of transoral cases vs 3.4% of transnasal cases) between the two approaches (59). This difference may be due to less trauma to the airway from the EEA compared to the transoral approach, which may require significant retraction of the tongue and soft palate.

Velopalatal insufficiency (VPI) is the failure of the appropriate closure of the velopharynx. VPI can result in hypernasal and poorly intelligible speech, or regurgitation of oral contents during swallowing. It has been proposed that VPI may occur more often with the transoral approach given that it requires more aggressive retraction, or even splitting, of the soft palate. This theory was not supported by the meta-analysis, in which the incidence of VPI was 3.3% for transoral cases and 6.4% of transnasal cases, a difference which was not statistically significant. Nonetheless, earlier resumption of oral feeding has been reported with the transnasal approach, with over half of patients commencing oral feeding within one day of surgery, and all patients doing so within five days (17,18,22). In comparison, oral feeding is routinely postponed for several days following a transoral approach (46).

Postoperative dysphagia is another concern when dealing with anterior approaches to the upper cervical spine or lower skull base. It has been hypothesized that the transnasal route should allow for a lower rate of dysphagia given that the mucosal incision lies mainly above the palate. This theory is supported by a cadaveric study that demonstrated a lower density of pharyngeal nerve fibers above the hard palate as compared to below it (71). A meta-analysis of published studies, however, did not find a difference in dysphagia rates between the transoral (3.8%) and transnasal (3.6%) routes (59).

As with any skull base approach, CSF leak is a significant concern and a strategy to avoid or to robustly repair a CSF leak is a prerequisite when planning an EEA. Intraoperative CSF leak was reported in 30.0% of transnasal approaches and 0.3% of transoral approaches; while postoperative leak was reported in 5.2% and 0.8% of cases, respectively. Although this represents a large absolute difference, it was not statistically significant due to small study sizes (59). Dural repair through the narrow transnasal route is challenging given that dural suturing is generally not practical. Repair techniques for incidental durotomies includes abdominal fat grafts, fibrin glue, CSF diversion with a lumbar drain (63,67), and nasal packing (43,44,75,78,80). Suturing the mucosal layer has been reported but is technically challenging (43,44). The problematic nature of CSF leaks is the reason why the EEA to CVJ pathology has been limited mainly to extradural pathologies. One exception is a study by Crockard that described a series of seven patients who underwent microscopic transoral approaches for treatment of intradural lesions. His patients routinely underwent shunt procedures and there were three postoperative CSF leaks and one case of meningitis (13). It is clear that improved techniques for dural closure are needed if the EEA can be routinely expanded to intradural lesions. Although the pedicled nasoseptal flap has proven to be a valuable tool for repairing many ventral skull base defects (24), its geometry may be less favorable for defects as low as the C1-2 region. An anatomical study found that even using an extended nasal septal flap, the inferior edge of the flap only reached as far as the foramen magnum (51).

The transnasal approach may have a theoretically lower infection rate given that the mucosal incision lies above the hard palate, lowering the chance of contamination with oral flora. However, the infection rate for both procedures remains low among the published studies. Shriver and colleagues found a wound infection rate of 1.9% for EEA (compared to 3.3% for transoral), while meningitis occurred in 4% (versus 1.0% of transoral cases), with neither difference being statistically significant (59). Of note, Yen and colleagues reported a death ten days postoperatively from meningitis in the setting of a postoperative CSF leak (78). This reinforces the importance of avoiding or adequately closing CSF leaks.

Vascular injury can be a devastating complication of skull base surgery. However, the incidence of carotid injury during endoscopic endonasal surgery is very low. The risk of carotid injury during odontoidectomy and similar CVJ procedures should be very low given that this inferior approach corresponds to the level of the cervical segment of the internal carotid artery, which remains far from midline with only rare exceptions. There were only 50 reported cases of ICA injury from EEA identified by a recent systematic review (8), with none involving the cervical segment. In addition, ICA injury during a CVJ approach has not been reported.

Neurologic injury is a potential complication in any cranial or spinal procedure. In the case of anterior craniocervical decompression, injury could occur as a result of manipulation of neural structures, vascular insult, inadequate decompression, improper reduction of deformity, or inadequate fixation. We found no reports of devastating neurologic injuries in the perioperative period, and the meta-analysis showed that none of the patients undergoing EEA worsened (59). Both EEA and microscopic transoral approaches can provide good neurologic outcomes. Possible strategies to minimize neurologic complications include establishing judicious surgical goals (i.e. debulking versus gross total resection in a tumor case), use of intraoperative neuromonitoring (i.e. SSEP's, and MEP's), adequate perfusion pressure, and close postoperative follow-up to detect mechanical instability that could result in delayed neurologic deterioration.

While the EEA is most often compared to the transoral approach, the endoscopic transoral technique deserves separate attention. Shriver's meta-analysis grouped all transoral complications together, but the endoscopic group was a very small proportion of all the transoral surgeries (39 out of 1238 cases). Husain et al. performed endoscopic transoral decompression on 11 patients and reported two patients each with wound infection and wound dehiscence, and one patient with postoperative neurologic deterioration despite adequate decompression and postoperative instrumentation (28). Quihang reported no complications in five patients undergoing this procedure (54). Yadav reported only a single case of CSF leak resolving with lumbar drainage in a series of 34 endoscopic transoral cases (77). There were no reports of tracheostomy or feeding tube placement among these studies.

Assessment of complications with the endoscopic transcervical route is based on 25 reported cases. Wolinsky's series of 15 patients included complications of airway swelling (two patients), dysphagia (two patients, with one requiring placement of a percutaneous endoscopic gastrostomy tube), and intraoperative CSF leaks (3 patients, of which two had no sequelae and one had an asymptomatic pseudomeningocele) (14). In a series of ten patients undergoing an endoscopic transcervical approach for ventral release of irreducible atlanto-axial dislocations, Wu and colleagues reported no complications (76).

In summary, the most common complications from ventral CVJ approaches are CSF leak, respiratory problems, dysphagia, and VPI. A meta-analysis comparing complication rates between transnasal and transoral (of which most used the microscope) approaches found a statistically significant lower rate of tracheostomy with the transnasal route. However, a comprehensive comparison of complication rates is hindered by the relatively low number of reported cases of EEA, endoscopic transoral, and endoscopic transcervical approaches.

Clinical Outcomes

Patients with symptomatic craniocervical compression and instability most often present with myelopathy, although they can also have neck pain, occipital radicular pain, and dysfunction of the lower brainstem and cranial nerves leading to dysphagia and respiratory failure. Stabilization or recovery of neurologic deficits is perhaps the most important concern for patients undergoing EEA. Published studies have generally reported excellent neurologic outcomes, with approximately 90% of patients improving and the rest showing stability without progression of their symptoms (17,18,59).

CONCLUSIONS

While ventral pathology of the CVJ remains challenging to treat surgically, the EEA approach has given us a valuable tool to provide optimal access to this region while minimizing morbidity. The EEA provides a direct route to ventral midline pathologies from the lower clivus to C2. It often needs to be combined with an instrumented fusion due to pre-exiting or iatrogenic instability. Its main limitations are the inferior extent of exposure that can be obtained and the width of the operative corridor, for which transoral techniques provide a complementary anterior approach. Reported complications from the EEA include CSF leak, airway problems, dysphagia, VPI, and infection. The EEA is likely to be less morbid than the transoral approach, as evidenced by the lower rate of tracheostomy (59). High rates of neurologic improvement have been reported following decompression using the EEA. Our experience as well as the volume of published cases suggests that the EEA approach is both effective and safe.

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