ORIGINAL ARTICLE

A Dual Approach for the Management of Complex Craniovertebral Junction Abnormalities: Endoscopic Endonasal Odontoidectomy and Posterior Decompression with Fusion

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■ BACKGROUND: **Ventral brainstem compression secondary to complex craniovertebral junction abnormality is an infrequent cause of neurologic deterioration in pediatric patients. However, in cases of symptomatic, irreducible ventral compression, 360° decompression of the brainstem supported by posterior stabilization may provide the best opportunity for improvement in symptoms. More recently, the endoscopic endonasal corridor has been proposed as an alternative method of odontoidectomy associated with less morbidity. We report the largest single case series of pediatric patients using this dual-intervention surgical technique. The purpose of this study was to evaluate the surgical outcomes of pediatric patients who underwent posterior occipitocervical decompression and instrumentation followed by endoscopic endonasal odontoidectomy performed to relieve neurologic impingement involving the ventral brainstem and craniocervical junction.**

■ METHODS: **Between January 2011 and February 2017, 7 patients underwent posterior instrumented fusion followed by endonasal endoscopic odontoidectomy at our unit. Standardized clinical and radiological parameters were assessed before and after surgery. A univariate analysis was performed to assess clinical and radiologic improvement after surgery.**

■ RESULTS: **A total of 14 operations were performed on 7 pediatric patients. One patient had Ehlers-Danlos syndrome, 1 patient had a Chiari 1 malformation, and the remaining 5 patients had Chiari 1.5 malformations. Average extubation day was postoperative day 0.9. Average day of initiation of postoperative feeds was postoperative day 1.0.**

■ CONCLUSIONS: **The combined endoscopic endonasal odontoidectomy and posterior decompression and fusion for complex craniovertebral compression is a safe and effective procedure that appears to be well tolerated in the pediatric population.**

INTRODUCTION

Ventral brainstem compression (VBSC) secondary to craniovertebral junction abnormality is associated with a number of underlying etiologies in the pediatric population. Predisposing developmental conditions include Down syndrome, Ehlers-Danlos syndrome, Klippel-Feil syndrome, os odontoideum, atlantoaxial rotatory subluxation, juvenile rheumatoid arthritis (Still's disease), cleidocranial dysostosis, achondroplasia, and a host of mucopolysaccharide storage disorders.[1-7] Pediatric patients with VBSC are commonly found to have an associated Chiari 1

Key words
- Axis
- Basilar invagination
- Chiari
- Endonasal
- Endoscopic
- Odontoidectomy
- Pediatric

Abbreviations and Acronyms

CT: Computed tomography
CVJ: Craniovertebral junction
CXA: Clivoaxial angle
EEA: Endoscopic endonasal approach
MRI: Magnetic resonance imaging
POD: Postoperative day
VBSC: Ventral brainstem compression
WCMC: Weill Cornell Medical College

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malformation.[8] However, the incidence of VBSC in patients with Chiari 1 malformations is much less frequent—estimated at 4% in one study.[9] In cases of symptomatic, irreducible VBSC secondary to craniovertebral junction abnormality, 360° decompression with posterior stabilization and fusion has been advocated.[10] Historically, transoral routes have been used to perform the odontoidectomy.[10-12] However, the endoscopic endonasal corridor has been proposed as an alternative route that may decrease the incidence of postoperative velopalatal insufficiency and dysphagia and allow for earlier extubation and initiation of oral feeds.[13,14] Although numerous publications have demonstrated the safety and efficacy of endoscopic endonasal odontoidectomy in the adult population, there is a paucity of data regarding the use of this approach in pediatric patients.[15] Very few select pediatric cases have been reported in the literature, with majority of them published as part of adult case series (**Table 1**). This is the largest single case series of pediatric patients with complex craniovertebral junction abnormalities who have had endonasal endoscopic odontoidectomy and posterior instrumented fusion.

The purpose of this study was to evaluate the clinical and radiologic outcomes of a consecutive series of pediatric patients who underwent posterior occipitocervical decompression and instrumentation followed by endoscopic endonasal odontoidectomy to evaluate the safety and efficacy of this approach.

METHODS

Patients

After institutional review board approval from Weill Cornell Medical College (WCMC), we compiled de-identified data of pediatric patients who underwent occipitocervical posterior instrumented fusion followed by endoscopic endonasal odontoidectomy over a period of 6 years from January 2011 to February 2017. Per the recommendation of the lead pediatric neurosurgeon (JPG), the patients underwent a posterior instrumented occipitocervical fusion paired with a tailored posterior fossa decompression before staged (same day or 48 hours later) endonasal odontoidectomy. Of note, some patients had undergone suboccipital decompression at an earlier date before the decision to proceed with odontoidectomy and occipitocervical fusion. All patients were 18 years or younger at the time of surgery and had clinical and radiological evidence of brainstem compression. The electronic medical records and operative notes were examined for relevant preoperative data (eg, age, gender, associated diagnoses, presenting signs/symptoms). Preoperative radiological studies (computed tomography [CT] and magnetic resonance imaging [MRI]) were used to assess the clivoaxial angles (CXA) and Grabb-Oakes measurement to gauge the severity of ventral compression before surgery.[25,26] Relevant intra- and postoperative data (eg, estimated blood loss, complications, time to extubation and initiation of oral feeds, evaluation for abnormality in speech or swallowing, length of hospital stay, relevant nasal outcomes, and postoperative symptomatic improvement) were subsequently assessed. The modified Rankin scale was used to assess the clinical outcome after surgery.[27] Routine postoperative plain films were obtained per protocol to evaluate for pseudarthrosis and/or instrumentation failure. In addition, we routinely assessed for improvement in the thickness of the ventral subarachnoid space after odontoidectomy by comparing sagittal pre- and postoperative T2-weighted MRI. This metric has previously been described in great detail and has correlated well with clinical improvement in past studies.[15] Pre- and postoperative values for both modified Rankin score and radiologic thickness of ventral subarachnoid space were evaluated using a paired *t*-test for statistical significance.

Furthermore, a systematic review of PubMed Central, Google Scholar, Scopus Database, Cochrane database, and Science Research, using the key words “odontoid endoscopic,” “pediatric,” “craniovertebral endoscopic,” “cervicomedullary endoscopic,” “endonasal endoscopic,” “children,” and relevant case series and case reports were identified. Articles reporting only

Table 1. List of Published Pediatric Cases (Endonasal Endoscopic Odontoidectomy and Posterior Instrumented Fusion)

Reference	Year	Study Type	Age(s)	Underlying Pathology(ies)
Magrini et al[16]	2008	Case report	11	DS + OO
Hankinson et al[13]	2010	Case series (only pediatric patients)	15, 11	CM + BI
Lee et al[17]	2010	Case series (adult and pediatric patients)	6	TO
Tomazic et al[18]	2011	Case report	11	CM + BI
Beech et al[19]	2012	Case report	10	BI + OO
Patel et al[20]	2012	Case report	10	AAS + BI
Sinha et al[21]	2012	Case report	13	OI + BI
Nagpal et al[22]	2013	Case report	12	BI
Hickman et al[23]	2013	Case series (only pediatric patients)	12, 11	CM + OI, DS + BI
Tan et al[24]	2014	Case series (only pediatric patients)	3, 12, 13	CM + BI, BI + AAS, RM
Goldschlager et al[15]	2015	Case series (adult and pediatric patients)	7, 14	BI + CM, BI + CM

AAS, atlantoaxial subluxation; BI, basilar invagination; CM, Chiari malformation; DS, Down syndrome; OI, osteogenesis imperfecta; OO, os odontoideum; RM, rhabdomyosarcoma; TO, telangiectatic osteosarcoma.

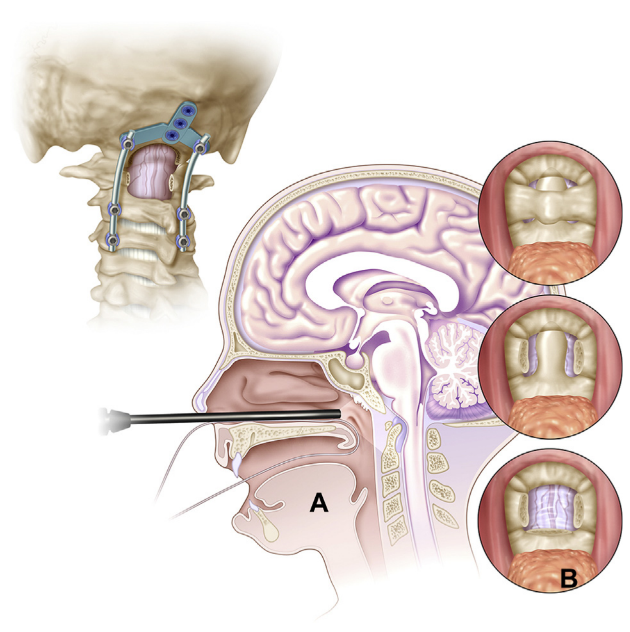

Figure 1. (**A**) Illustration showing the endoscopic endonasal approach to the odontoid along the hard palate. Note the absence of trauma to the oropharynx, which facilitates early extubation and feeding. *Inset*: View of the odontoid, after removal of the anterior ring of C-1. (**B**) Staged views of operation. *Upper left:* The nasopharyngeal fascia between the Eustachian tubes overlies the bottom of the clivus and the top of the odontoid. The sphenoid sinus is opened here for orientation purposes but does not need to be opened in this operation. *Upper right:* Anterior arch of C-1 after removal of nasopharyngeal fascia and longus colli muscles. *Lower left:* Odontoid process after removal of the anterior arch of C-1. *Lower right:* Ventral dura in front of CMJ after odontoid removal. CMJ, cervicomedullary junction.

pediatric cases were identified. In addition, pediatric cases were also selected from the articles with large series of adult and pediatric patients.

Operative Technique

At WCMC, the surgical team consists of a pediatric neurosurgeon (JPG), endoscopic skull base neurosurgeon (THS), and an otorhinolaryngologist (VKA or AK). The otorhinolaryngological surgeon performs the endonasal endoscopic approach phase of the procedure up till the floor of the sellar fossa is adequately exposed, and then the neurosurgeon continues with the surgical procedure. The posterior suboccipital decompression and occipitocervical fusion is performed by a team of spinal neurosurgeons before the odontoidectomy (RH, AB, JPG), either on a previous day or earlier on the same day. The fusion is performed in neutral anatomic alignment using routine neurologic monitoring (ie, motor and somatosensory evoked potentials) before the anterior decompression to minimize any potential risk of cord injury relating to underlying spinal impingement and/or instability. Great care is taken in optimizing anatomic alignment, as postoperative pharyngeal edema in the context of excessive flexion or posterior translation of the head in the fixed position can contribute to difficulty with ventilation and/or swallowing. Fusion is done before the endoscopic endonasal odontoidectomy to prevent any cord compromise that could result from inadvertent movement during an anterior procedure with an unstable cervical or craniocervical spine region. All endonasal endoscopic procedures in this institution are performed with the head secured in a Mayfield (Integra, Ohio, USA) clamp, to prevent unnecessary neck or head movement. In the majority of cases presented, the fusion construct involved an occipital plate, C2 pars or pedicle screws, and lateral mass screws in the subaxial cervical spine based on desired length of construct. Sublaminar wiring was used in lieu of screw instrumentation in select cases.

The operative technique of endonasal endoscopic odontoidectomy has been previously described in great detail.[15,28-30] In brief, the patient is positioned supine with the head immobilized in Mayfield clamp 3-point fixation. Navigation using CT and CT angiography to identify bony and vascular landmarks is vital for intraoperative localization and to help optimize decompression.[31] The nasopharynx is approached using a binostril approach (**Figure 1**). A posterior septectomy is performed. The adenoids can be resected to allow for improved visualization and access, particularly in younger children with smaller anatomy.[13] This is a major feature unique to the pediatric cases. In adult cases, a septoplasty or turbinectomy (superior or middle) could be performed to improve surgical access. Surgical manipulation of the endoscope within the narrow nasal apertures in pediatric cases could also be a bit more technically challenging than when done in adults.

A vertical incision with electrocautery is made in the posterior pharyngeal musculature. Gentle dissection is used to expose the anterior arch of C1. The incision in the posterior pharynx can be further extended cephalad to expose the inferior portion of the clivus. In cases of severe platybasia, the inferior portion of the clivus sometimes must also be removed to achieve adequate ventral decompression. The anterior arch of C1 is resected to allow for exposure of the odontoid. The odontoid is hollowed out with a high-speed drill. The apical and alar ligaments are sectioned, whereas the base of the dens remains intact. Leaving a thin shell of bone before final decompression can delay early visualization of dura within the operative field. Full ventral decompression is completed using curettes and small Kerrison rongeurs (Codman/Johnson and Johnson, Raynham, Massachusetts, USA) to remove the posterior shell of cortical bone from the tip of the dens, taking care of dissecting the bone from dura from a margin that is often very adherent. In the cases where the posterior shell of cortical bone is adherent to the cruciate ligament, they can be removed together to expose the dura. The posterior pharyngeal tissue defect is then reapproximated using absorbable stitches. In case of cerebrospinal fluid leak, a small fat graft is placed over the defect and held in place with the reapproximated pharyngeal tissue. In this situation, a lumbar drain is also placed. An online video has been provided as a reference of the surgical technique (**Video 1**).

Video

Video available at www.sciencedirect.com

If the occipitocervical fusion and odontoidectomy are performed on the same day, the patient is extubated the following morning. If the odontoidectomy alone is performed, patients are extubated immediately after the procedure. Diet is advanced shortly thereafter.

Table 2. Preoperative Clinical Characteristics of Patients Who Underwent Endoscopic Endonasal Odontoidectomy for Basilar Invagination

Patient	Age/Sex	Preoperative Signs/Symptoms	Associated Diagnoses	Clivoaxial Angle (Degrees)	Grabb-Oakes Measurement (mm)
1	10/M	SOP, MY, SD, GI, DZ	C1.5M, EDS	115	18.2
2	10/M	SOP, OSA, DZ	C1.5M	116	14.4
3	11/M	SOP, SD	C1M	118	9.1
4	18/M	SOP, MY	C1.5M	106	17.2
5	16/F	SOP	C1.5M, SC	93	9.4
6	14/M	MY, OSA, SD, GI	C1.5M, AOA, BI	109	14.8
7	7/F	SOP, MY, SD, VE, GI	KF, C1-2AF, BI	111	11.6

AOA, atlanto-occipital assimilation; BI, basilar invagination; C1-2 AF, C1-2 autofusion; C1.5, Chiari 1 malformation; C1M, Chiari 1 malformation; DZ, dizziness; EDS, Ehlers-Danlos syndrome; GI, gait instability; KF, Klippel Feil; MY, myelopathy; OSA, obstructive sleep apnea; SC, scoliosis; SD, swallowing difficulty; SOP, suboccipital pain; VE, vertigo.

RESULTS

A total of 14 operations were performed on 7 pediatric patients for the treatment of VBSC associated with craniovertebral junction abnormalities (preoperative variables can be found in Table 2). Five of the patients were male, and 2 were female. One patient had Ehlers-Danlos syndrome, 1 patient had a Chiari 1 malformation, and the remaining 5 patients had Chiari 1.5 malformations. Three of the patients had undergone prior suboccipital decompression at outside institutions before presentation; 1 additional patient had undergone prior suboccipital decompression at WCMC. Common presenting signs and symptoms included suboccipital pain ($n = 6$), myelopathy ($n = 4$), swallowing dysfunction ($n = 4$), gait instability ($n = 3$), obstructive sleep apnea ($n = 2$), dizziness ($n = 2$), and vertigo ($n = 1$). The CXA ranged from 93° to 118° (mean of 110°). Grabb-Oakes measurement ranged from 9.1 to 18.2 mm (mean of 13.5 mm).

The patients were carefully reviewed by a multidisciplinary team of neurosurgeons, spinal surgeons, neuroanesthetists, and so on, and the dual procedure of endonasal endoscopic odontoidectomy and posterior instrumented fusion was advocated in specific cases based on the presenting neurology, radiological findings/measurements, and where the posterior approach was not deemed optimal enough to address the VBSC.

All patients underwent a suboccipital decompression and tailored instrumented occipitocervical fusion followed by endonasal endoscopic odontoidectomy (intra- and postoperative variables can be found in Table 3). Estimated blood loss associated with endonasal odontoidectomy was 50 mL or less in all cases. Average extubation day was postoperative day (POD) 0.9. Average day of initiation of postoperative feeds was POD 1.0. The subsequent length of hospital stay was 5 to 9 days (mean hospital stay, 6.7 days), which included the time between the 2 surgeries. One patient aspirated nasal packing and fibrin sealant after extubation on POD 0 and required reintubation. This patient was subsequently extubated without incident on POD 2. Another patient experienced transient postoperative dysphagia that resolved with conservative measures. The mean follow-up duration was 33.7 months (range, 13–76 months). All 7 patients showed improvement in their postoperative modified Rankin scores. The mean preoperative score was 2.3 ± 0.8. The mean postoperative score was 1.0 ± 0.8. The difference between the 2 scores, as evaluated by a paired *t*-test, was statistically significant ($P < 0.0001$). The mean width of the T2 signal ventral to the brainstem significantly increased from 0.99 ± 0.47 mm before surgery to 3.37 ± 0.53 mm postoperatively (paired *t*-test; $P \leq 0.0001$).

Illustrative Case

A 12-year-old male patient initially presented to our clinic with complaints of headaches that were mainly frontal and not exertional. He did not complain of swallowing difficulties. His parents reported that he had problems with snoring. He had no significant past medical history and his neurologic examination was nonfocal. MRI of the brain demonstrated a Chiari 1.5 malformation with tonsillar herniation of 24 mm and VBSC (Figure 2). In light of the nonspecific nature of his presenting symptoms, a trial of medical management and close clinical follow-up was recommended. At his 2-year follow-up visit, he endorsed progressive dizziness, blurry vision, persistent headaches, and daytime somnolence. Polysomnography was notable for 60 recorded episodes of central sleep apnea during the duration of the test. Neuro-ophthalmology evaluation did not reveal papilledema, esotropia, or nystagmus. Recommendation at the time was for posterior fossa decompression with duraplasty, occipitocervical instrumentation, and endoscopic, endonasal odontoidectomy. The multistage procedure was performed successfully and the patient was discharged uneventfully. At his 1-year follow-up, the patient reported complete resolution of headaches and blurry vision with improvement in sleep quality and decreased daytime somnolence leading to improved school performance.

DISCUSSION

The clinical presentation in pediatric patients with VBSC is highly variable and can include head and/or neck pain, paresthesias, quadriparesis, ataxia, vertigo, dysphagia, dysphonia, sleep apnea, urinary dysfunction, and/or progressive scoliosis.[9,32] Fang et al[33] demonstrated myelopathy as the commonest presenting feature (75% of the patients) in their systematic review, as opposed to

Table 3. Intra- and Postoperative Clinical Characteristics of Patients Who Underwent Endoscopic Endonasal Odontoidectomy

Patient	Levels Fused	Complications	Extubated (POD)	Oral Feed (POD)	Follow-Up (Months)	Preoperative MRS	Postoperative MRS
1	O-C3	Transient dysphagia	1	1	17	3	2
2	O-C3	None	1	1	13	1	0
3	O-C3	None	1	1	23	3	1
4	O-C3	Reintubation	0/2*	2	24	2	1
5	O-C4	None	1	1	32	2	0
6	O-C5	None	0	1	76	2	1
7	O-C5	None	0	0	51	3	2

MRS, Modified Rankin Score; POD, postoperative day.
*Patient coughed postextubation and aspirated nasal packing and sealant requiring reintubation; subsequently extubated without incident on POD2.

suboccipital pain in this series. However, most of their patients were adults, and rheumatoid arthritis was the commonest underlying pathology noted (36.5%). Consequently, the more unfavorable rate of complications in adults could be consequent to the associated comorbidities.

The main goals of surgical intervention for these patients with complex craniovertebral junction (CVJ) abnormalities are to relieve compression and stabilize the CVJ. In some cases, both goals can be accomplished via a single posterior midline approach. In some cases with basilar invagination and VBSC, preoperative halo traction may be required to relieve the decompression. Endonasal endoscopic odontoidectomy with posterior instrumented fusion is our proposed treatment option for cases where the aforementioned algorithm management steps do not achieve the intended goals.

Surgical decision making is driven mostly by the severity of clinical symptoms and their progression; however, imaging provides important information that may guide treatment strategies. The severity of VBSC can be gauged using lateral radiographs and/or midsagittal CT and MRI.[34] We also routinely add a CT angiography to the CT to provide a road map of the carotid artery through the skull base to integrate into our navigation and preoperative planning in children in whom distances are often small and anatomy distorted by the pathology. Flexion/extension radiographs can be additive to assess for instability at O-C1 or C1-2 that may not be appreciated on static imaging.

A number of treatment algorithms have been proposed for the treatment of pediatric patients felt to be symptomatic from VBSC secondary to basilar invagination.[10] If the ventral compression is

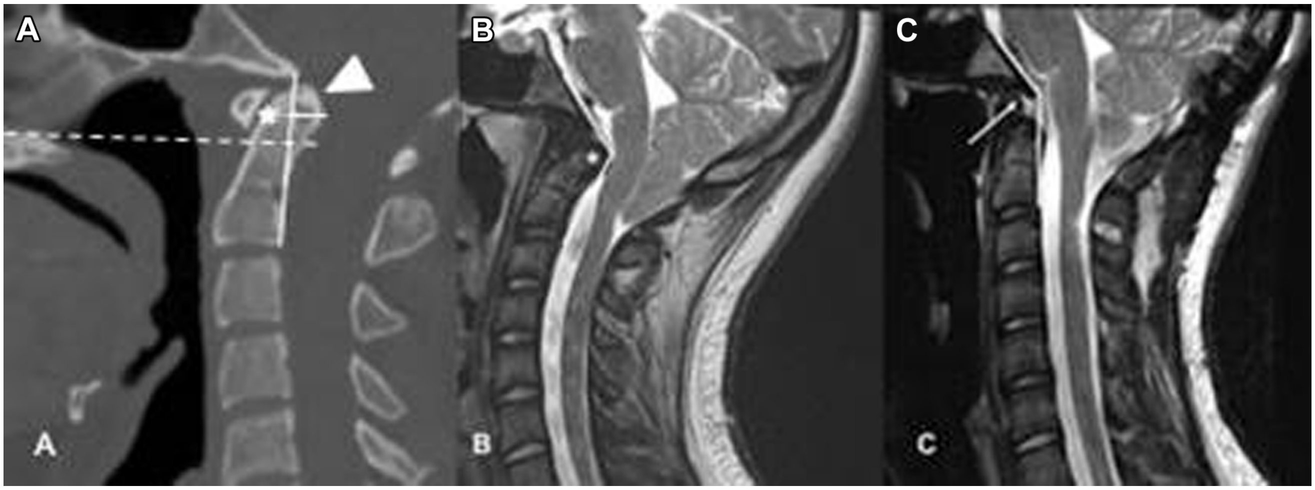

Figure 2. (**A**) Midsagittal preoperative computed tomography scan of the cervical spine demonstrates a retroflexed odontoid (designated with *white arrowhead*) and assimilated arch of C1. (**B**) Midsagittal T2-weighted cervical spine magnetic resonance imaging (MRI) confirms ventral brainstem compression secondary to a retroflexed odontoid. There is medullary kinking and indentation with radiologic evidence of severe obstruction of cerebrospinal fluid (CSF) (cerebrospinal fluid) flow across the craniocervical junction. Clivo-axial angle (CXA) (designated with *asterisk*) was measured at 118°, and the Grabb-Oakes measurement was noted at 9.1 mm. (**C**) Postoperative midsagittal T2-weighted MRI of the cervical spine shows an improved CXA (designated with *arrow*; measured at 140°) with complete resolution of brainstem compression and adequate ventral CSF flow.

reducible, posterior decompression and occipitocervical fusion without anterior decompression has been recommended.[32] In cases of irreducible VBSC, anterior and posterior decompression in addition to posterior stabilization and fusion is often advocated.[10,13] This case series of our patient group describes the latter. In the past, semirigid wire-and-loop fixation devices have been used for stabilization.[5,35,36] However, more recent studies have indicated superior results with rigid screw fixation.[37-40]

All 7 of the patients evaluated in this series had evidence of complex craniovertebral junction abnormalities with VBSC. Notably, 5 of the seven patients had radiologic evidence of Chiari 1.5 malformation. The designation of Chiari 1.5 involves caudal descent of the brainstem, in addition to cerebellar tonsillar herniation characteristic of the Chiari 1 malformation. Patients in this subgroup appear to be twice as likely to fail posterior fossa decompression alone when compared with Chiari 1 patients.[41] Moreover, progressive retroflexion of the odontoid may correlate with increased caudal displacement of brainstem structures, as evaluated by the position of the obex in relation to the foramen magnum on MRI.[42] Grabb et al[26] created a measurement to evaluate the extent of VBSC in patients with Chiari malformations. All patients with a Grabb-Oakes measurement (pB-C2) <9 mm were successfully treated with posterior fossa decompresssion alone. In select patients with VBSC and a pB-C2 measurement of >9 mm, treatment to directly reduce the degree of VBSC was recommended along with suboccipital decompression to prevent neurologic deterioration.[26] Bollo et al[25] also sought to investigate patients with Chiari 1 and 1.5 malformations at high risk of failing routine posterior decompression. This study found that patients with basilar invagination, the Chiari 1.5 malformation, or a CXA measurement <125° were at an elevated risk of failing standard suboccipital decompression alone. Collectively, these studies support the idea that consideration should be given to directly addressing VBSC in a subgroup of patients with complex craniovertebral junction abnormalities felt to be at high risk of failure with posterior decompression alone. All 7 patients in this study had a pB-C2 measurement of >9 mm and a CXA measurement of <125°.

Historically, the transoral route has been considered the gold standard for anterior decompression of craniovertebral junction abnormalities associated with VBSC.[10,15] Although this approach has been found to be efficacious in numerous series, it is associated with a number of drawbacks as well.[5,10,11] A systematic review of transoral approaches in the literature of adult and pediatric patients by Komotar et al[43] reported that an incision in the soft palate was necessary to improve exposure in 33.6% of cases. Additional factors inherent in the transoral approach contribute to perioperative morbidity. Swelling secondary to intraoral retraction can result in airway edema that may require prolonged intubation and/or tracheostomy after surgery.[10,11] Disruption of oropharyngeal mucosa can adversely affect the swallowing mechanism, which in some cases may already be compromised from lower cranial nerve deficits. In the previously referenced literature review,[10,11] gastrostomy tube placement was necessary in 14.8% of patients who underwent transoral odontoidectomy. Baseline risks of postoperative velopalatal insufficiency, nasal regurgitation, and hypernasal speech increase considerably when transpalatal, transmaxillary, and transmandibular extensions are used.

The endoscopic endonasal approach (EEA) for odontoidectomy was proposed as a way to potentially decrease the risk profile associated with the transoral approach. The advantages allow for an earlier time to extubation and return to oral feeding in patients following EEA for odontoidectomy when compared with the transoral approach.[15,44] In addition, the EEA offers superior access to severe VBSC above the level of the hard palate, an area notoriously difficult to reach via the transoral approach. Access can be obtained inferior to the level of the hard palate as well. Surgeons considering implementing the endonasal odontoidectomy in their practice should be familiar with appropriate methods of preoperative radiographic evaluation. The authors prefer to use the rhinopalatine line to ensure that the area of compression is not beyond the inferior limit of visualization afforded by the EEA.[28] In this method, the starting point is calculated 2/3 of the way from the rhinion to the anterior nasal spine. A line drawn from this point traversing through the posterior nasal spine is extended posteriorly to illustrate the inferior extent of exposure.

Although much has been written about the EEA for odontoidectomy in adult patients, there is a relative paucity of data in the pediatric literature. Alfieri et al[45] first reported the EEA to the CVJ and odontoid process in a cadaveric model in 2002. Three years later, Kassam et al[46] performed the procedure on a patient with symptomatic cervicomedullary compression secondary to compressive odontoid and rheumatoid pannus.[37] Zwagerman et al[47] recently published the largest series to date on 34 adult patients who underwent EEA for odontoidectomy. This series demonstrated a high rate of postoperative dysphagia—however, in the majority of cases, this was a transient phenomenon. In the study by Zwagerman et al, the EEA allowed for thorough decompression in all cases and no patients suffered from postoperative velopalatal insufficiency. The experience with endonasal odontoidectomy in the pediatric population is less robust. Magrini et al[16] published the first pediatric report of EEA for odontoidectomy in 2008 in a patient with Down's syndrome; the endonasal approach was chosen in this patient in part because of a small oral cavity. Hankinson et al[13] reported that the EEA provided excellent access for ventral decompression in 2 pediatric patients with VBSC in 2010. However, two patients in this study had difficulty with postoperative ventilation. Specifically, one patient required bedside tracheostomy for respiratory arrest after extubation on POD1 and the other patient required reintubation after extubation on POD2.

From the literature, only 15 pediatric cases have been reported till date from a total of 5 case series and 6 case reports (**Table 1**). As this is still a relatively new surgical treatment modality, the first one was reported in 2008. The average age of the published cases is 11.4 years (range, 6—15 years). Causal pathologies included Chiari malformation with basilar invagination, Down syndrome, os odontoideum, and atlantoaxial subluxation. There were 2 cases of malignancies, that is, rhabdomyosarcoma and telangiectatic osteosarcoma.

As far as we know, this is the largest single pediatric case series regarding this procedure (**Table 1**). One patient aspirated nasal packing after extubation on POD0 and required reintubation, before subsequent extubation without incident on POD2. Aspiration during anesthesia occurs most frequently during induction, but could also occur during the maintenance phase and emergence stage from anaesthesia.[48] To prevent such a complication, patients at risk should be identified early during the preoperative assessment and an appropriate anesthetic technique be instituted. Overall, the average extubation day for this group of patients was POD0.9.

Another patient experienced transient dysphagia that resolved with conservative measures alone. Otherwise, no complications were recorded. Patients in this series resumed oral feeds an average of 1.0 day after surgery. All patients noted significant clinical improvement postoperatively, and radiographic results of the decompressed area matched the preoperative goals in each case. Despite the clinical results that have been achieved, the authors do not recommend universally adopting an endonasal approach for all cases requiring odontoidectomy. Select anatomical features such as very small nostril size, robust presence of adenoid tissue, short intercarotid distance, limited sinus pneumatization, rhinopalatine line, and long dens-nares distance should be considered before the decision to proceed with an endonasal route. Surgeons should also be aware of the risks associated with the endonasal approach, as well as the posterior fusion. Overall, the scores on the modified Rankin scale improved postoperatively, but these were collated retrospectively during their follow-up reviews.

Although this study has inherent limitations, we feel that as the largest series to date in a purely pediatric population, our data demonstrate that this dual technique is beneficial for a specific subset of patients.

CONCLUSIONS

Our goal in this article was to describe our growing experience demonstrating the safety and efficacy of EEA for odontoidectomy in the pediatric population. We report on 7 consecutive patients within this series who underwent staged occipitocervical fusion followed by endonasal odontoidectomy.

This study demonstrates that the endoscopic endonasal approach for odontoidectomy is a safe and effective procedure that is well tolerated in the unique age group, particularly as an option in the setting of irreducible, symptomatic VBSC.

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