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Basilar Invagination: Case Report and Literature Review

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Running Title: Anterior vs. Posterior Decompression for Basilar Invagination

Key Words: Anterior odontoid decompression, posterior odontoid decompression, basilar invagination, cervicomedullary junction compression, endoscopic endonasal odontoidectomy
Abstract

**Background:** Basilar invagination (BI) is a rare clinical condition characterized by upward protrusion of the odontoid process into the intracranial space, leading to bulbomedullary compression. It is often encountered in adults with rheumatoid arthritis (RA). Transoral microscopic or endonasal endoscopic decompression may be pursued, with or without posterior fixation. We present a case of basilar invagination with C1-C2 autofusion and discuss an algorithm for choice of anterior versus posterior approaches.

**Case:** A 47-year-old woman with RA presented with severe occipital and cervical pain, dysphagia, hoarseness, and arm paresthesias. MRI revealed moderate cranial settling with the odontoid indenting the ventral medulla, with no posterior compression. CT demonstrated bony fusion at C1-C2 without lateral sag. Given autofusion of C1-C2 in proper occipito-cervical alignment and the absence of posterior compression, the patient underwent endoscopic endonasal odontoidectomy without further posterior fusion, with satisfactory resolution of symptoms.

**Conclusion:** Endoscopic endonasal odontoidectomy offers a safe and effective method for anterior decompression of basilar invagination. Pre-operative assessment for existing posterior fusion, absence of posterior compression, and preservation of the anterior C1 ring during operative decompression help stratify the need for lone anterior approach versus the need for combined anterior and posterior treatment.
Introduction

Symptomatic anomalies of the craniovertebral junction (CVJ) present a unique challenge for neurosurgical management with regard to surgical approach selection and planning and the accompanying technical difficulty. Of the CVJ anomalies, basilar invagination (BI) confers a particular therapeutic challenge. BI often coexists with or is a result of other medical conditions (e.g., Chiari malformation, Down syndrome, Klippel Feil, osteogenesis imperfecta, and rheumatoid arthritis (RA)), which complicate achieving atlanto-axial stability and increases perioperative complications (23). In patients with BI, the ventral aspect of the brainstem or the craniovertebral junction is often compressed leading to neurological symptoms necessitating decompression. Because of the mechanical and anatomical complexities, surgical treatment continues to center on varying combinations of anterior or posterior decompression with or without traction or fusion. Common routes for decompression include endonasal and transoral access ventrally or an occipital-cervical posterior decompression and fixation (15). We present a RA case with odontoid basilar compression and autofusion of C1-C2, in whom an endonasal decompression approach was pursued, without posterior fusion. We further discuss a systematic algorithm for selection of operative approaches to treatment of basilar invagination based on our experience and literature published.

Case

A 47-year-old woman with long-standing RA presented with 6 months of increasing neck and occipital pain, occasional dysphagia, dysphonia, and difficulty breathing. Her MRI and CT revealed basilar invagination and moderate cranial settling (Figure 1). Review of previous cervical spine films revealed mild subluxation of C1 and C2 since 1998. Operative intervention was indicated given her symptomatic and radiographic progression of brainstem compression. Observation of autofusion at the C1-C2 joint in proper occipito-cervical alignment prompted an endoscopic endonasal approach for odontoidectomy and decompression, deferring an accompanying posterior fusion. The odontoid was exposed following a small inferior posterior septectomy and lateralization of the buccopharyngeal fascia from the level of the Eustachian tubes to C2. The inferior aspect of the clivus was drilled to expose foramen magnum and the odontoid peg was resected in the setting of pannus and hypertrophied ligamentous tissue. Adequate decompression was achieved with preservation of the arch of C1. The patient tolerated the procedure well and reported near-resolution of occipital and cervical pain, with only intermittent mild dysphagia, on follow-up.

Discussion

Currently, no defined guidelines exist for management of patients with basilar invagination. Each patient is different and complex due to frequently accompanying concomitant diseases. Decisions are based on imaging findings, often complicated by abnormalities such as cranial settling, autofusion of cervical vertebrae, platybasia, and syringomyelia. Management should account for both compression of the brainstem and cervical cord as well as for craniospinal instability. There is no clear consensus as to treatment approaches for this condition, with options ranging from cervical traction, lone anterior decompression posterior sagittal realignment and fusion with or without bony decompression, as well as combined anterior/posterior approaches (9,13-15,22,26).

Preoperative considerations
Preoperative imaging of the connective tissue, musculature, and osseous alignment should be closely evaluated. In cases where there is evidence of ventral tissue contraction, posterior reduction may be limited, thus necessitating an anterior approach. Identification of the relationship of the vertebral arteries to the C1-C2 lateral masses, posterior arch, and entry at the occiput is essential. The course of the vertebral arteries is also known to course irregularly along the paraesophageal tract and ventral body of C2, affecting the operative approach (20,21).

**Considerations for posterior approaches**

In cases of BI without cranial nerve palsies, suggesting low-level ventral compression, posterior realignment and fusion has shown to be successful when a mobile and reducible C1-C2 complex is present (1). This realignment can be achieved with preoperative traction, a Halo-vest, distraction of C1-C2 by intra-articular spacers, or by intraoperative traction, followed by subsequent posterior instrumentation and fusion (1,9,28).

**Considerations for anterior approaches**

In cases of moderate to severe ventral compression, initial criteria to consider are presence of cranial nerve palsies, in which case, an anterior approach is often necessary independent of posterior complex stability (17,26). When C1-C2 is fused, unsafe to distract, or if distraction fails to alleviate the ventral compression, anterior approaches have been shown to be successful and necessary (8,11,26,32). Autofuson of nearby bodies such as C2-C3 is insufficient if the C1 integrity is not preserved (12). In select cases, anterior decompression can be safely pursued without causing significant instability, thereby avoiding a posterior fusion (3,8,11,27,32) (**Table 1**). Deteriorating neurological function any time following a basilar decompression without posterior fusion would need to be further investigated, as a posterior fusion may eventually be required.

If there is evidence of auto-fusion of the C1-C2 lateral masses, either transoral or endonasal odontoidectomy is effective. The approach is best decided in accordance to the patient’s anatomy and surgeon’s intuition regarding the exposure needed to successfully treat the patient. The endonasal approach is favored when the lesion is located 2 cm above the palate (6). The nasopalatine line can assist with determining the most inferior point of dissection possible (2,4).

Morbidities related to the endonasal approach involve disruption of the nasal anatomy (i.e. middle turbinates, ethmoids, and posterior septum). Disruption of the nasal mucosa can results in tissue atrophy and ongoing nasal complications, some of which can be avoided if an additional corridor is used through the oral cavity to avert damage (4,6).

The majority of complications related to the trans-oral approach involve the oral cavity. There is risk of damage to the tongue, teeth, phonation, and speech due to velopharyngeal incompetence (12). Risk of infection is also high with pharyngeal incisions due to constant contamination from oral flora (16). As such, these patients often require prolonged extubation times and nasogastric tube feedings (12,18,32).

**Consideration for a combined anterior and posterior approach**

A caveat of the anterior approach is that odontoidectomy may cause cervical instability, especially if the C1-C2 lateral masses have ligamentous laxity. In cases where there is clear evidence of C1-C2 instability, such as demonstrated lateral expulsion of the C1 lateral mass, due to transection of the C1 anterior arch, posterior instrumentation and fusion is necessary (8,24,25). The location and orientation of the odontoid is important. If the odontoid is 7-8 mm above the
Chamberlain line, has a posteriorly oriented tip, or there is severe medullary compression anterior approach and maximal decompression will be necessary. Of note, in elderly and frail patients without evidence of C1-C2 instability that may not be able to tolerate the posterior approach, anterior decompression through an odontoidectomy with maintenance of C1 anterior arch may be preferred (5,17,31).

The combined procedure may be staged or done in one setting. Reasons for staging include minimizing operating room time, anesthesia related risks, and blood loss; however, there is potential for complications from instability as great as subluxation, which has known to occur even in the OR during positioning (21). Although there are many reports of anterior decompressions followed by posterior fusion in patients with a high concern of spinal instability, posterior cervical fusion may be performed first in order to better stabilize the cervical spine with the skull base (2,10). The age of the patient may also influence the choice for a one- versus two-stage procedure.

Algorithm for operative approaches to basilar invagination

In our patient, the particular anatomic orientation of cranial settling and basilar impression allowed for adequate decompression through an isolated endonasal approach. Specifically, the coronal alignment of the occipital condyle with the C1 lateral mass and C2 facet joints suggested C1-C2 stability. Foregoing posterior fusion in patients with these characteristics is a viable option, but requires close post-operative follow-up. Improvement in neurologic symptoms and absence of neck pain calls for continued clinical observation. On the other hand, deteriorating neurological function or progressively worsening neck pain may be a harbinger of cervical instability and posterior stabilization should be considered. Intraoperative CT following a resection may provide valuable utility in determining decompression was adequate (7). We highlight our proposed algorithm in Figure 2.

Limitations

This is a single case report and our algorithm may not be applicable to other surgeons. Based on the literature reviewed and our experience with this case, we believe our algorithm offers a way to stratify surgical approaches to basilar invagination. Surgeon preferences and experience may dictate the most appropriate approach to treatment of BI.

Conclusions

Basilar invagination is a CVJ anomaly that poses considerable management challenges. No well-established studies clearly delineate a treatment algorithm for this condition. From review of the literature and our experience, we propose that an anterior approach should be considered first in patients with high cervicomedullary compression and lack of C1-C2 mobility. During this procedure, effort should be made to preserve the anterior ring of C1 and the lateral masses of C1-C2 to avoid fusion (8). In cases without cranial nerve palsies and presence of C1-C2 mobility, a posterior approach with fusion is preferred. If the posterior decompression fails to relieve the symptoms or there is evidence of a high-level compression, then a combination of anterior and posterior approaches is necessary. Throughout the management triage process, careful preoperative planning and thorough discussion of peri-operative risks should be carried out with the patient.
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References


**Figure Legend**

**Figure 1. Basilar invagination pre-operative and post-operative imaging.** Pre-operative cervical spine CT demonstrating (A) basilar compression of the medulla in mid-sagittal view, and (B) cranial C1-C2 settling on coronal view. (C) Pre-operative sagittal view of the cervicomedullary compression on T2-weighted FLAIR MRI. (D) Intra-operative drilling of odontoid peg. (E) Post-operative CT illustrating successful anterior decompression following endoscopic endonasal odontoidectomy.

**Figure 2.** Management algorithm for anterior and posterior approaches to basilar invagination.
Table Legend

Table 1. Reported cases of successful basilar decompression via an endoscopic endonasal approach without posterior fusion.
Table 1

<table>
<thead>
<tr>
<th>Year</th>
<th>Author</th>
<th>Age/Gender</th>
<th>Disease</th>
<th>Procedure</th>
<th>Posterior Fusion</th>
<th>Neurosurgical Outcome</th>
<th>Follow Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>2014</td>
<td>Chaudhry et al.</td>
<td>47F</td>
<td>RA</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>4 months</td>
</tr>
<tr>
<td>2013</td>
<td>Yu et al. (31)</td>
<td>55M</td>
<td>BI, KF</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>30 months</td>
</tr>
<tr>
<td>2012</td>
<td>Dasenbrock et al.</td>
<td>54F</td>
<td>BI, KF</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>21 months</td>
</tr>
<tr>
<td>2012</td>
<td>Gladi et al. (11)</td>
<td>80M</td>
<td>RA</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>27 months</td>
</tr>
<tr>
<td>2011</td>
<td>Scholtes et al.</td>
<td>76F</td>
<td>BI</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>12 months</td>
</tr>
<tr>
<td>2011</td>
<td>Grammatica et al.</td>
<td>56F</td>
<td>ACM</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>9 months</td>
</tr>
<tr>
<td>2011</td>
<td></td>
<td>20M</td>
<td>ACM</td>
<td>ETO</td>
<td>No</td>
<td>Improved, Spine Stable</td>
<td>1 month</td>
</tr>
</tbody>
</table>

Legend: RA Rheumatoid Arthritis, BI Basilar invagination, KF Klippel-Feil Syndrome, ACM Arnold Chiari Malformation, ETO Endoscopic Transnasal Odontoidectomy
Figure 2: Proposed treatment algorithm for basilar compression
Highlights:

- We here present a case with odontoid basilar compression that was decompressed anteriorly without posterior fusion.
- Evidence of autofusion of C1-C2 should be investigated in pre-operative imaging.
- Basilar compression with cranial nerve palsies often necessitates anterior approach when distraction is not possible.
- We propose a systematic algorithm for selection of operative approaches.
- Intraoperative CT following a resection may provide valuable utility in determining decompression was adequate.
Abbreviations:
Basilar invagination (BI), Rheumatoid Arthritis (RA), Craniovertebral Junction (CVJ),