Atlantoaxial Fixation for Basilar Invagination without Obvious Atlantoaxial Instability (Group B Basilar Invagination): Outcome Analysis of 63 Surgically Treated Cases

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**BACKGROUND:** We discuss the rationale of surgical treatment of group B basilar invagination by atlantoaxial facet joint stabilization and segmental arthrodesis.

**METHODS:** From January 2010 to April 2016, 63 patients with group B basilar invagination were surgically treated. All patients had varying degree of myelopathy-related functional disability. Fifty-two patients had both Chiari malformation and syringomyelia. All patients were treated by atlantoaxial plate and screw fixation with the techniques described by us in 1994 and 2004. Foramen magnum decompression or syrinx manipulation was not carried out in any patient. Occipital bone and subaxial spinal elements were not included in the fixation construct.

**RESULTS:** Three patients died in the immediate postoperative phase. In the remaining patients, there was clinical improvement and no patient’s neurologic function worsened after surgery. In 12 of 38 patients in whom postoperative magnetic resonance imaging was possible, at a follow-up of at least 3 months, there was reduction in the size of the syrinx.

**CONCLUSIONS:** The pathogenesis of basilar invagination in group B is related to atlantoaxial instability. The clinical outcome suggests that the surgical treatment in these cases should be directed toward atlantoaxial stabilization and aimed at segmental arthrodesis. Inclusion of the occipital bone in the fixation construct is not necessary. Foramen magnum decompression and procedures involving manipulation of Chiari malformation and syringomyelia are not necessary.

**INTRODUCTION**

In 2004,¹ we discussed a classification that divided basilar invagination into group A and group B depending on the presence or absence respectively of demonstrable instability of the atlantoaxial region that was manifested by distancing of the odontoid process from the anterior arch of the atlas. Group B basilar invagination is a relatively rare clinical entity.¹ There are only a limited number of studies in the literature that discuss the treatment strategy in such patients.⁴ It was earlier observed that in group B basilar invagination, the atlantoaxial joint was either fixed or fused and the atlantodental interval was within its normal range. On the basis of observations, it was identified that posterior foramen magnum decompression was an effective method of surgical treatment for this group of patients.⁴ Several other investigators have treated patients with group B basilar invagination on similar lines.⁷ In general, most investigators agree that it is necessary to perform foramen magnum decompression to increase the volume of the posterior cranial fossa.¹⁵,⁶ Others find craniovertebral stabilization along with foramen magnum decompression a viable treatment option.⁵,⁶ The question of including occipital bone and subaxial vertebrae in the fixation construct is also a debated issue.⁷,⁸

As the experience in the subject increased over the years, it was observed that atlantoaxial instability forms the basis of pathogenesis of both groups of basilar invagination. Even in group B basilar invagination, in which there is no radiologically demonstrable abnormal atlantoaxial mobility of the region on dynamic imaging, the principal pathogenetic issue was atlantoaxial instability. The structural musculoskeletal malformations in this group of patients were related to special functional needs in the face of long-standing atlantoaxial instability rather than a result of embryologic dysgenesis.⁹ In this article, we discuss our experience with 63 patients with group B basilar invagination who were treated by atlantoaxial fixation. The rationale of the discussed treatment and the outcome are analyzed.
METHODS

From January 2010 to April 2016, 407 patients with basilar invagination were treated by atlantoaxial fixation. Of these patients, 63 had group B basilar invagination. These patients are analyzed. The principle guideline that was adopted to classify patients as having group B basilar invagination was that the atlantodental or clivodental interval was within the normal range (maximum, 3 mm) in dynamic images. When measured by the classic parameter of the Chamberlain line, 50 patients had severe basilar invagination, meaning that the odontoid process tip was at least 10 mm above this line.10

There were 40 males and 23 females in the series and their ages ranged from 14 to 58 years (average, 28 years). The presenting symptoms are outlined in Table 1. The clinical assessment was based on a 5-point scale11 as shown in Table 2. In addition, Japanese Orthopaedic Association scores12 (Table 3) were used to assess the patients both before and after surgery. All patients underwent preoperative dynamic plain radiography, computed tomography (CT), and magnetic resonance imaging (MRI). Appropriate imaging of the vertebral artery included CT and/or magnetic resonance angiography and was carried out in all cases. In 3 patients, three-dimensional models were made to evaluate the pathology, rehearse the operation before surgery, and provide anatomic assistance during the operation.12 Sixty-one patients had Chiari malformation. Fifty-two patients had both Chiari malformation and syringomyelia. Twenty-eight patients with Chiari malformation were included in our earlier study on the subject.11 Six patients had been treated earlier by foramen magnum decompression. Based on type of facetal alignment and atlantoaxial instability in a neutral head position, the patients were divided into 3 groups.14,15 Type A patients (n = 4) had anterior atlantoaxial instability, in which the facet of the atlas was dislocated anterior to the facet of the axis. Type B patients (n = 28) had posterior atlantoaxial instability, in which the facet of the atlas was dislocated posterior to the facet of the axis. Type C patients (n = 31) had no demonstrable facetal malalignment on dynamic imaging and the patients were labeled as having central or axial atlantoaxial instability or dislocation. In 3 patients, dynamic images showed vertical, mobile, and at least partially reducible atlantoaxial dislocation.16 All patients underwent surgery with the aim of segmental atlantoaxial arthrodesis using the techniques discussed in 1994 and 2004.1,17-19 The patients were placed in a prone position. Midline craniocervical exposure was performed. The exposure of the atlantoaxial joint was considerably difficult because of the presence of severe basilar invagination and the atlantoaxial joint being remarkably rostrally located. The degree of difficulty in exposure of the atlantoaxial joint was related to the presence of occipitalization or assimilation of the atlas and decrease of the length of the clivus. The C2 ganglion had to be sectioned in all cases to achieve the exposure. Subperiosteal exposure along the pedicle of the C2 vertebra assisted in exposing the joint. The articular cartilage was widely denuded. Bone chips harvested from the iliac crest were packed into the joint cavity. Monoaxial self-tapping screws and plates were used in all cases for atlantoaxial facetial fixation. The implants were of stainless steel in 25 patients treated in the earlier part of the series. Titanium implants were later used in 38 patients. All muscle attachments to the C2 spinous process were sharply sectioned. In addition, bone graft was placed in the midline over the appropriately prepared host bone of the posterior arch of the atlas and lamina of C2.

Problems During Surgery

In 8 patients, the vertebral artery had an abnormal course and travelled over the posterior surface of the facet of the atlas. In 2 cases, the vertebral artery was injured during dissection in this region. Although the presence of extensive venous bleeding in the region made suturing of the lacerated vessel difficult, successful suturing of the vertebral artery was possible in 1 patient. In 3 patients, the artery was injured during the insertion of a screw guide into the C2 facet. Bleeding was successfully controlled in all patients by hurriedly completing the screw insertion; the procedure essentially sacrificed the vertebral artery. Difficulty in dissection and adequate exposure of the joint did not permit completion of metal fixation as planned on 1 of the 2 sides in 8 patients. Metal implantation involving screw insertion in the facets of the atlas and axis and plate/rod construct was performed only on 1 side in 2 patients. In 3 patients, a transarticular fixation was performed on the contralateral side. In 2 patients, a spacer was impacted into the joint on the contralateral side joint17 and no additional instrumentation was used. In 1 patient, occipitocervical fixation was performed on the contralateral side. Three patients died in the postoperative phase. Death appeared to be related to vertebral artery injury in 2 patients.

Postoperatively, the patients were placed in a 4-post hard cervical collar for 8 weeks. They were mobilized as soon as they were able to sit or stand on their own. Postoperative investigations included plain radiography and CT. MRI was possible in patients in whom titanium implants were used. The radiologic investigations were performed within 48 hours of surgery and at 3 months and 1 year follow-up (Figures 1 and 2).

There was no implant failure or infection in any case. The follow-up ranged from 3 to 72 months (average, 32 months). Radiologic follow-up of more than 6 months was available for analysis in 55 patients. All patients’ symptoms improved after surgery. The degree of improvement varied and is outlined in Tables 2 and 3. No patient had delayed worsening of neurologic symptoms. Postoperative MRI at a follow-up of at least 3 months showed reduction in the size of the syrinx in 12 patients (of 38 patients in whom MRI was possible). In 2 patients, a reduction in size of the syrinx was observed in the immediate postoperative phase. Rostral or reverse migration of the herniated tonsil was appreciated in 8 patients.

Table 1. The Presenting Symptoms

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck pain</td>
<td>52</td>
</tr>
<tr>
<td>Arm pain</td>
<td>14</td>
</tr>
<tr>
<td>Paresthesias</td>
<td>38</td>
</tr>
<tr>
<td>Weakness/stiffness</td>
<td>56</td>
</tr>
<tr>
<td>Hoarseness/nasal regurgitation</td>
<td>8</td>
</tr>
<tr>
<td>Bowel/bladder weakness</td>
<td>4</td>
</tr>
</tbody>
</table>
The subject of basilar invagination has been under discussion for more than a century. Several landmark contributions worldwide have helped to understand the subtleties of the pathogenesis of the malformation and evolution of the treatment strategy.\(^{1,6,19,21-27}\)

From being a radiologic curiosity in the middle of the twentieth century,\(^{28}\) the clinical entity is now viewed as eminently treatable. The rapidity in the evolution of understanding of various issues of craniovertebral junction epitomizes the scientific and medical revolution that has taken place in the last few decades hallmarking by the growth of computer technology. Most importantly, patients with problems secondary to myelopathy related to compression of critical neural structures at the craniovertebral junction can now hope to live with pride and independence.

In 1998, a classification of basilar invagination divided it into 2 groups based on the absence (group 1) or presence (group 2) of Chiari malformation.\(^{29}\) The pathogenesis in group 1 patients seemed to be herniation of the spine into the brain via the foramen magnum (as summarized by von Torklus and Gehle\(^{27}\) in 1972). The odontoid process seemed to be the compressive factor over the cervicomedullary cord and accordingly, transoral surgical decompression of the odontoid process formed the bottom line of the treatment strategy. In group 2 patients, reduction of the posterior cranial fossa volume seemed to be the primary pathogenetic factor. Chiari malformation was speculated to be a result of reduction of the space available for the cerebellum in the smaller posterior cranial fossa. According to the understanding of mechanical issues, posterior fossa volume expansion by foramen magnum decompression was considered a rational surgical strategy.

The surgical strategy for basilar invagination as mentioned earlier and as was current during that time was based on the understanding that the craniovertebral junction and atlantoaxial joint were stable or fixed in such cases.\(^{29}\) Surgical decompression, either by an anterior transoral surgical route or by posterior foramen decompression, formed the primary modalities of treatment. Surgical stabilization was essentially a nonissue or was highly debated.

In 2004, an alternative classification of basilar invagination divided these patients into 2 groups on the basis of presence (group A) or absence (group B) of manifest instability of the atlantoaxial junction shown by an abnormal increase in the atlanto-dental or clivodental interval.\(^{2}^{1}\) In group A basilar invagination, the atlantoaxial dislocation was not fused or fixed as was described in the existing literature; it was not only mobile but was excessively and abnormally so. More importantly, there was a possibility of reduction of the dislocation and realignment of the craniovertebral junction by facetal distraction using bone graft with or without titanium spacer-implant impaction. This concept had a great impact on the treatment design of the entity of basilar invagination and several subsequent studies have discussed the need for stabilization of the craniovertebral junction and the possibility of reduction of group A basilar invagination.\(^{21,29,9}\) Posterior alone treatment is considered a standard form of treatment by many groups, and transoral surgical decompression is slipping into the historical domain.

Although surgical treatment for group A basilar invagination is standardized, the treatment strategy for group B basilar invagination is still under discussion. A literature survey does not show any major clinical series that delineates or describes the surgical principles of this group of patients. The anomaly of group B basilar invagination was considered to be stable or fixed and there was no manifest instability that could be identified by using standard radiologic parameters. Foramen magnum decompression formed the primary mode of surgical treatment. In the

### Table 2. Distribution as per Clinical Grading System

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Number of Patients (Preoperative)</th>
<th>Number of Patients (Postoperative)</th>
<th>Lower Cranial Nerve Deficits</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Independent and normally functioning</td>
<td>6</td>
<td>23</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>Walks on own but needs support/help to carry out routine household activities</td>
<td>17</td>
<td>18</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Walks with minimal support and requires help to carry out household activities</td>
<td>20</td>
<td>11</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>Walks with heavy support and unable to carry out household activities</td>
<td>9</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>5</td>
<td>Unable to walk and dependent for all activities</td>
<td>11</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>

### Table 3. The Preoperative and Postoperative Clinical Assessment as per Japanese Orthopaedic Association Scoring System

<table>
<thead>
<tr>
<th>Japanese Orthopaedic Association Score</th>
<th>Preoperative (Number of Patients)</th>
<th>Postoperative (Number of Patients)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;7</td>
<td>15</td>
<td>3</td>
</tr>
<tr>
<td>8–12</td>
<td>26</td>
<td>11</td>
</tr>
<tr>
<td>13–15</td>
<td>20</td>
<td>26</td>
</tr>
<tr>
<td>16–17</td>
<td>2</td>
<td>20</td>
</tr>
</tbody>
</table>
literature, the validity for stabilization of the craniovertebral junction has not been clearly evaluated for this group of patients.

As experience in the subject grew over the years, it was realized that atlantoaxial instability formed the basis of basilar invagination not only in group A patients but also in group B patients. Neck pain as a constant presenting or associated symptom, presence of facetal malalignment, and identification of instability during direct bone handling were the factors that indicated an unstable atlantoaxial joint. A successful clinical outcome of treatment that was based on fixation without any form of bone or soft tissue decompression validated the observation. The atlantoaxial dislocation in group B is long-standing and subtle. In some cases, the instability could have started in the fetus or in early infancy. The morphologic musculoskeletal changes and neural malformations seem to be natural protective responses that are initiated and progress because of the presence and persistence of instability. In our earlier study, the issue of reversibility of musculoskeletal changes after surgical treatment that stabilizes the atlantoaxial joint was discussed. The fact that short neck, torticollis, and soft tissue neural changes reverse after fixation suggests that these alterations are not related to embryologic dysgenesis but are a result of special neural needs in the face of atlantoaxial instability. It was recently hypothesized that both Chiari malformation and syringomyelia are neural manifestations of

![Figure 1. Images of a 30-year-old man. (A) Sagittal T2-weighted magnetic resonance image (MRI) showing group B basilar invagination, Chiari malformation, and syringomyelia. (B) Sagittal image of showing severe group B basilar invagination. Assimilation of the atlas can be observed. (C) Sagittal cut of computed tomography scan with the cuts passing through the facets showing type B atlantoaxial instability. (D) Delayed (3 months after surgery) postoperative T2-weighted MRI showing reversal of the Chiari malformation and reduction in the syringomyelia. (E) Postoperative MRI scan showing the atlantoaxial fixation construct.](image-url)
atlantoaxial instability and are secondary and probably protective natural responses. The early recognition of instability and its subtle character provide an opportunity for the natural musculoskeletal and neural processes to mold the soft tissues to their advantage and avoid, minimize, and delay neural compromise.

Identification of 3 types of atlantoaxial facet dislocation formed the basis of an alternative classification system for basilar invagination. Type A facet dislocation occurs when the facet of the atlas is dislocated anterior to the facet of the axis. This dislocation was likened to lumbosacral spondylolisthesis. The odontoid process is displaced posterior in such an event and the atlantodental interval increases abnormally. Type A facet dislocation is more frequently encountered in group A basilar invagination. Type B facet instability occurs when the facet of the atlas is dislocated posterior (retrolisthesis) to the facet of the axis. This dislocation was likened to lumbosacral spondylolisthesis. The short clivus can be visualized. Type C basilar invagination occurs when the facet of the atlas is dislocated anterior to the facet of the axis, but the odontoid process is displaced posterior. The atlantodental interval remains normal.
axis. Type C facet instability occurs when there is no radiologic evidence of facet malalignment, but the instability was identified only during surgery. The atlantoaxial interval is not remarkably abnormal in type B and type C facet instability. Such forms of instability are also called axial or central instability. As the odontoid process does not compress on the neural elements, the spinal neural symptoms are subtle and the disease progresses over several years or decades. The average duration of presenting symptoms was 35 months. Morphologic and structural musculoskeletal alterations and neural abnormalities such as Chiari malformation and syringomyelia are more frequent in this group of cases. In none of the cases was there fusion between the facets of the atlas and axis.

Our experience suggests that atlantoaxial instability forms the basis of pathogenesis of group B basilar invagination. Stabilization of the atlantoaxial joint forms the primary form of treatment. Inclusion of the occipital bone and occipitoatlantal joint in the fixation is neither necessary nor does it provide optimum stabilization to the region. Direct facet fixation of the atlantoaxial joint provides a ground for firm and long fixation of the screws and implants and the short segmental stabilization provides significant biomechanical advantage to the construct. However, the conduct of surgery and direct facet fixation was technically challenging in the group of patients presented. Despite 28 years of experience with fixation of the region, we were forced to perform unilateral fixation in 2 patients and had to resort to alternative methods of fixation on the contralateral side in 6 patients. Extensive venous bleeding in the region, frequent presence of an anomalous course of the vertebral artery posterior to the facet of atlas, and rostral location of the atlantoaxial joint make the exposure technically difficult and are wrought with a danger of vertebral artery injury. C2 ganglion resection is almost always necessary to achieve exposure in these cases. Considering the complexity of surgery, elaborate preoperative investigations are essential before surgery. The aim of surgery is to obtain atlantoaxial fixation and segmental arthrodasis. Wide denuding of the articular cartilage, introduction of bone chips within the joint cavity, and direct fixation of the facets form the key steps of surgical treatment. Distraction of facets, particularly using titanium implants as has been advocated for selected cases of group A basilar invagination, is not necessary or mandatory for this group of patients because the aim of the treatment is not craniovertebral realignment but stabilization of the atlantoaxial joint. Because the atlantodental interval is not altered or is not remarkably increased, craniovertebral realignment is not the issue. Foramen magnum decompression, either alone or in association with atlantoaxial or craniovertebral fixation, is neither necessary nor does it provide a viable long-term solution; it may even be countereffective.22

Technical difficulties in exposure of the region are more pronounced in patients in whom there was occipitalization of the atlas. The length of the clivus is also a guide regarding the degree of surgical difficulties that can be encountered in exposing the atlantoaxial joint. Although occipital fixation in such cases can provide a similar form of craniovertebral fixation, direct screw fixation of the facets is a significantly superior and stronger technique that provides remarkable stability.33 Placement of the screws in the facet of the atlas even in patients with an assimilated atlas provides a strong fixation point for atlantoaxial stabilization. Facetal stabilization provides fixation at the point of the fulcrum of all movements. The possibility of insertion of long screws in the substance of the thick and cortical bone structure of the facet seems to be stronger and more effective compared with insertion of multiple short length screws in the squama of the occipital bone. Opening of the joint, denuding of articular cartilage, and stuffing of bone graft pieces within the joint provide stability to the construct and an additional space for bone fusion. However, considering the potential surgical difficulties and issues related to venous bleeding and vertebral artery in cases with group B basilar invagination, it does seem that if one is not conversant with exposure of the lateral masses and atlantoaxial joint and facetal screw placement, inclusion of occipital bone in the fusion construct can provide suboptimal but reasonable stability in general and in patients with occipitalization of the atlas in particular. Some investigators have discussed the role of stabilization of the subaxial spinal vertebrae as a fixation point for the cervical spine. However, involvement of multiple joints in the fixation process and use of longer implants have a biomechanical disadvantage and should be avoided, unless other techniques are either not possible or not feasible.

The vertebral artery adopts an unusual course in some patients and can traverse posterior to the facet of the atlas. Insertion of a screw in some patients even involves mobilization of the artery away from the field. The complex bone anomalies and their unusual distortions also make the surgical procedure technically difficult and challenging. Our experience with vertebral artery injuries over several years suggests that sacrifice of the artery is safe in most cases, but in general, the clinical outcome after vertebral artery injury is unpredictable. It is clear that vascular compromise of a major vessel such as the vertebral artery should be avoided and the outcome of this arterial sacrifice can sometimes be clinically hazardous for neurologic outcome and for life. For this reason, elaborate preoperative evaluation of the anatomic location of the vertebral artery is mandatory, particularly as it relates to the facets of the axis and atlas. In this series, death of 2 patients could be directly related to vertebral artery injury during the surgical procedure. Although no correlation could be made, the dominance of the artery and age of the patient seem to be determinants that could affect clinical outcome in patients with vertebral artery injury.

The outcome was evaluated on the basis of a clinical grading system that we described recently.11 Although the scale needs to be validated on the basis of a multi-institutional experience, it does seem to provide a reasonable, simple, and easily reproducible method of clinical evaluation. To avoid deficiency in assessment, we in addition used the more widely used evaluation system of the Japanese Orthopaedic Association. Foramen magnum decompression has been uniformly advocated for patients with Chiari malformation and syringomyelia. The positive clinical outcome in all surviving patients confirms the usefulness of atlantoaxial fixation as the primary surgical procedure. The fact that 6 patients who had earlier failed posterior decompressive surgery and improved after atlantoaxial fixation reinforces the validity of the surgical procedure. Although the size of the syrinx reduced in only 32% of patients (when MRI was possible), the clinical recovery in 100% of patients suggests the prime role of atlantoaxial instability in the pathogenesis of both Chiari
malformation and syringomyelia. Postoperative deaths in 3 patients reinforce the need for technical and surgical improvements in operations and for diligently analyzing the course to save the vertebral artery during surgery.

CONCLUSIONS

Atlantoaxial fixation forms the mainstay of treatment of patients with group B basilar invagination. Foramen magnum decompression is not necessary in these patients.

REFERENCES


Conflict of interest statement: The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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