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# Single-Stage Posterior Decompression and Occipitocervical Fusion Using a Screw-Rod-Plate System for Basilar Invagination with Anterior Spinal Cord Compression and Craniocervical Instability

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# Abstract

Purpose: To review our experience with surgical management of basilar invagination causing foramen magnum compression, focusing on selection of the surgical approach.

Methods: Twelve consecutive patients underwent posterior foramen magnum decompression with occipitocervical fixation and fusion for treatment of basilar invagination causing brain stem compression and instability. Gentle traction and reduction during positioning of the patients also were performed. Ventral decompression (odontoid resection) was performed in none of the patients. Pre- and postoperative neurologic status was graded according to JOA and Nurick scales

Results: All patients had anterior spinal cord compression due to cranial settling of the cervical spinal column as well as instability at the craniocervical junction. The average follow-up period was 31 months (range, 24-42 months). All patients' JOA and Nurick scores improved after surgery, but postoperative neurologic improvement and odontoid reduction were better in patients with atlas assimilation compared with patients with other pathologies.

Conclusion: Odontoid reduction using an occipitocervical fixation system and decompression of the foramen magnum through a single-stage posterior approach is an effective treatment for basilar invagination, particularly in patients with atlas assimilation. Since odontoid reduction and foramen magnum decompression can be achieved through a single-stage posterior approach in most patients, odontoid resection should remain as a secondary procedure when these decompression efforts are insufficient.

**Keywords:** Occipitocervical fixation; Instability; Basilar invagination; Craniocervical junction anomaly; Odontoid resection; Transoral approach

## Introduction

The customary treatment method for patients with basilar invagination and anterior odontoid compression at the foramen magnum level is odontoid resection through a transoral technique (or some variation of endoscopic techniques) followed by occipitocervical fixation and fusion in a second operation [1-7]. On the other hand, recent publications have shown that decompression of the brain stem in these patients can be achieved by odontoid reduction through a posterior approach using a cervical anchor or through preoperative cervical traction using Gardner-Wells tongs [8-11]. The concrete criteria for an "irreducible" odontoid and which degree of reduction is significant for the outcomes of these patients are not clear in the literature. There is also confusion regarding the order of these surgical procedures in this difficult clinical entity and unique anatomical area.

In this study, the outcomes of 12 patients with basilar invagination who underwent odontoid reduction, posterior foramen magnum decompression, and occipitocervical fixation through a single-stage posterior approach are presented. Since we performed odontoid resection in none of the patients in this study, the preoperative selection criteria for the best candidates for this single-stage posterior approach also are discussed.

## Material and Methods

## Patient population and clinical evaluation

Twelve patients diagnosed with basilar invagination and craniovertebral junction instability causing anterior odontoid compression at the foramen magnum level and progressive neurologic deficit between January 2006 and January 2013 were included in this study. All patients underwent odontoid reduction, posterior

craniocervical decompression, and occipitocervical fixation. Patients diagnosed with basilar invagination without neurologic deficit or with nonprogressive neurologic deficit and with no radiographic evidence of instability were excluded from this study since they did not undergo surgical treatment. When instability was uncertain, patients were evaluated with dynamic magnetic resonance imaging (MRI) to disclose brain stem compression by flexion. Data were collected and analyzed retrospectively. The patient population comprised 4 men and 8 women aged 7 to 55 years (mean, 27.58 years). The clinical presentations are summarized in Table 1. Symptom onset occurred at 3 to 25 years (mean, 10.5 years) of age. Progressive weakness in the extremities was the main symptom, and was present in all patients. Pre- and postoperative neurologic status was graded according to the Japanese Orthopaedic Association (JOA) [12] and Nurick [13] scales for determining maximum efficiency of the surgical procedure. All patients were followed for 24 to 42 months (mean, 31 months). Postoperative JOA and Nurick scores were recorded at 1 month and 1 year after surgery, and the scores at 1-year postoperative were used here as the final neurologic condition.

## **Radiographic evaluation**

Diagnosis was determined mainly from measurements of cranial

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Patient No.	Age,y/Sex	Duration of symptoms	Pathology	Surgery Fusion level	JOA		Nurick		Comp.	
					Preop	Postop	Preop	Postop	•	Follow-up mo
1	21 M	4 years	Assimilation of atlas, MM	Dec + Oc-C2	6	11	4	2	-	42
2	41F	18 years	Congenital, MM	Dec + Oc-C2	7	11	3	2	+	38
3	42 M	20 years	Ankilosing spondylit, MM	Dec + Oc-C5	11	14	3	2	-	24
4	15 F	6 years	Assimilation of atlas, MM	Dec + Oc-C3	6	11	4	2	+	36
5	31 F	14 years	Congenital, SM	Dec + Oc-C4 syr.sh.	7	11	3	2	-	24
6	16 F	6 years	Assimilation of atlas, Chiari, MM	Dec + Oc-C3	6	10	3	1	-	24
7	7 F	3 years	Down Syndrome, MM	Dec + Oc-C4	6	12	4	1	-	36
8	34 M	8 years	Down Syndrome	Dec + Oc-C4	9	11	2	1	+	24
9	13 F	4 years	Assimilation of atlas, MM	Dec + Oc-C3	10	14	2	1	+	36
10	20 F	3 years	Congenital, MM	Dec + Oc-C2	12	14	2	2	-	36
11	36 F	16 years	Congenital + Severe Chiari	Dec + Oc-C2	12	14	2	1	-	28
12	55 M	25 years	Congenital, MM	Dec + Oc-C2	14	15	1	1	-	24

 Table 1: Summary of clinical presentations. MM: Myelomalasia, SM: Syringomyelia, JOA: Japanese Orthopedic Association Score, Dec: Decompression, Oc-C2,C3,C4,C5:

 Occipito-Level of Cervical Lateral Mass Fixation.

Batiant No.	CMA, Deg		ADI, mm		WL, mm		CL, mm		ML, mm	
Fallent NO.	Preop	Postop	Preop	Postop	Preop	Postop	Preop	Postop	Preop	Postop
1	105	140	12	3	5	2	4	-	4	-
2	125	140	4	2	6	3	5	-	5	-
3	120	160	7	0	4	1	6	-	5	-
4	125	160	10	7	5	1	4	-	5	-
5	110	145	5	2	4	1	4	-	4	-
6	115	147	6	3	7	2	5	-	6	-
7	130	150	7	2	3	0	3	-	6	-
8	128	140	6	2	3	0	3	-	4	-
9	115	155	8	4	4	0	4	-	3	-
10	130	130	3	3	0	0	4	-	2	-
11	80	80	3	3	0	0	26	-	2	-
12	105	105	3	3	0	0	12	-	2	-

 Table 2: Documentation of spinal cord and/or brain stem compression by radiographic measurements. ADI: Atlantodens Interval; CL: Chamberlain's Line; CMA: Cervicomedullary Angle; ML: Mcrae's Line; WL: Wackenheim's Line.

settling on upper cervical MRIs and dynamic roentgenograms. Computed tomography (CT) scans also were obtained when necessary. Concrete documentation of spinal cord and/or brain stem compression was made by radiographic measurements (Table 2). Associated Chiari malformation, syringomyelia, or other intramedullary signal changes also were evaluated.

After surgery, all patients underwent plain radiography and thinslice CT with reconstructed views to define the positions of the screws and the extent of reduction. MRI was performed approximately 6 months later to assess the extent of decompression of the spinal cord and medulla oblongata; change in syringomyelia also was evaluated. All relative radiographic measurements were repeated after surgery. However, owing to intraoperative removal of a significant part of the posterior margin of the foramen magnum, the CL (Chamberlain's line) and ML (McRae's line) were impossible to draw on postoperative images, and were not recorded.

# Surgical technique

Under general anesthesia, in the prone position gentle traction

for reduction (combined forces of distraction and extension) of the odontoid was applied by the surgeon under fluoroscopic control before fixing the head by Mayfield headrest. A small part of the posterior margin of the foramen magnum (approximately 10–15 cm<sup>2</sup>), C1 posterior arch, and C2 lamina (when necessary) were removed completely by a Kerrison Rongeur. This decompression procedure was performed in all 12 patients, but dural opening with graft application was performed only in 1 patient (see Complications section). After satisfactory decompression, the occipitocervical fixation procedure was accomplished (Cervical Posterior System 1; Tasarimmed). Finally, the wound was closed in layers after bone fusion. Figure 1 includes an illustrative case of a basilar invagination in children with Down's syndrome.

## Statistical analysis

The results of this study were evaluated by statistical analyses. Comparisons between groups in which the parameters did not show normal dispersion were evaluated with the Mann-Whitney U test. On the other hand, comparisons within groups in which the parameters

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did not show normal dispersion were evaluated with the Wilcoxon signed-rank test. Since the number of patients in this study was 12, interactions within the parameters were evaluated with Spearman's Rho nonparametric test.

## Results

Weakness, particularly in the upper extremities, and neck pain were the primary symptoms, and were present in all patients. Other symptoms included posterior column dysfunction, bowel and bladder dysfunction, ataxia, and paresthesia. The preoperative causing pathologies were ankylosing spondylitis in 1 patient, Down syndrome in 2, atlas assimilation in 4, and congenital basilar invagination in 5. The clinical follow-up ranged from 24 to 56 months (mean, 36.5 months).

Decompression and some degree of reduction of the craniocervical junction were achieved in all patients. Radiography and CT were performed immediately postoperatively and at 3 months to 1 year after surgery. These imaging studies were performed until bone fusion was confirmed. Correction of sagittal plane dislocation was evaluated by comparing pre- and postoperative CMA (cervicomedullary angle) and WL (Wackenheim's line) values. The mean increase in the CMA after surgery was  $22.00^{\circ} \pm 16.12^{\circ}$ , while the mean decrease in the WL after surgery was  $2.58 \pm 1.68$  mm. These differences were statistically significant (p = 0.007 and 0.006, respectively). These radiographic findings as well as reappearance of the subarachnoidal space around the foramen magnum on postoperative MRI confirmed good decompression of the spinal cord and medulla oblongata in all 12 patients.

According to our results there is a remarkable difference at



Figure 1: Images of a 7-year-old female patient with Down syndrome admitted with sudden syncope. She had a history of previous occiput-C2 posterior wire fixation at another institution when she was 5 years old. (a) Direct lateral cervical roentgenogram, showing spontaneous rupture of the wire. (b) Lateral computed tomographic reconstruction, revealing occipitocervical instability, basilar invagination, and severe compression of the foramen magnum. (c, d) Occipitocervical fixation was performed by inserting pedicle screws from the C2 to C4.



**Figure 2:** Images of a 13-year-old female patient admitted with quadriparesis and difficulty standing. Preoperative magnetic resonance image (MRI) (a) and computed tomography (CT) scan (b), showing basilar invagination with severe spinal cord compression. (c) Postoperative sagittal CT reconstruction, showing bony decompression at the foramen magnum level. (d) T2-weighted sagittal MRI taken 2 months after surgery, showing remaining posterior spinal cord compression durat thickening, despite bony decompression (arrow). The patient underwent a reoperation for dural decompression and graft application (asterisk = metallic artefact of the fixation system).

postoperative changes in the CMA and WL values in patients with atlas assimilation. Correspondingly, postoperative changes in JOA and Nurick scores were also remarkably higher in patients with atlas assimilation. On the other hand, these differences were not statistically significant, probably due to restricted number of cases.

#### Complications

One patient with congenital instability underwent a reoperation due to a broken rod. Another patient with Down syndrome, who also had hyperactivity, underwent 3 reoperations due to loosening or breaking of the fixation system. In the last operation, fixation was achieved by transcondylar screws instead of an occipital plate. A third patient underwent a reoperation 36 months after the first operation due to intractable pain. Her pain was alleviated after the fixation system was completely removed. The overall nonfusion rate was 16.6% (2 of 12 patients).

A 13-year-old female patient underwent surgery for treatment of atlas assimilation and basilar invagination causing severe brain stem compression and instability (Figures 2a and 2b). She received standard posterior reduction, decompression, occipitocervical fixation, and fusion. The patient was readmitted approximately 2 months later because her symptoms did not recover. Her new MRIs revealed remaining spinal cord compression due to dural thickening, despite bony decompression (Figures 2c and 2d). A second operation comprised excision of the tight dura at the foramen magnum and fascia graft application for widening of the neural channel. Therefore, the overall complication rate associated with the fixation system and/or decompression procedure was 33.3% (4 of 12 patients). Neural tissue injury, infection, or wound complication did not occur in this series.

## Discussion

Odontoid resection carries several serious risks because of its difficult application, including infection, unmanageable instability, swelling of the tongue, and also death [14,15]. Besides these risks, concomitant occipitocervical fixation will be compulsory in a second operation. On the other hand, posterior C0–C1 decompression and odontoid reduction with the help of an occipitocervical rigid instrumentation system is a single-stage and safer procedure since it eliminates all probable risks associated with an anterior approach.

Abumi et al. [16] introduced posterior occipitocervical reconstruction using the anchors of cervical pedicle screws in 1999. With these systems, flexion deformity of the occipitoatlantoaxial complex and upward migration of the odontoid process were corrected through the combined forces of extension and distraction between the occiput and cervical pedicle screws. In the last 10 years, several methods have been reported describing correction of occipitocervical alignment and reduction of the odontoid through a single-stage posterior approach [8-10,17-19]. All these methods use either preoperative traction with Gardner-Wells tongs or an intraoperatively placed occiput plate with lateral mass screw traction and compression of the rods before fixing the system for odontoid reduction and indirect anterior decompression. In our series, we used a similar method, previously described by Kim et al. [20], which provided sufficient correction of malalignment of the craniocervical junction. In our series, the underlying cause of cranial settling of the odontoid was primarily congenital, except for ankylosing spondylitis in 1 patient and Down syndrome in 2 patients. The postoperatively increased CMAs in this study support that reduction of basilar invagination based on congenital reasons (particularly atlas assimilation) can be achieved indirectly through a posterior approach without the need for anterior odontoid resection. This situation should be taken into account while treatment modalities are planned preoperatively.

Atlas occipitalization or assimilation is one of the most common osseous congenital anomalies of the craniovertebral junction, with an incidence of 0.08%–3% in the general population [21]. Basilar invagination and occiput–C1 instability are most commonly secondary phenomena in these patients. In which type of atlas assimilation and which mechanism is responsible for the development of basilar invagination are obscure in these patients. Menezes asserted that as a child grows, this pathology is in reducible form up to approximately age 14 or 15 years [22]. Beyond this age, the lesion becomes irreducible



**Figure 3:** (a) T2-weighted sagittal magnetic resonance image of a patient with congenital basilar invagination, showing periodontoid dural and ligamentous thickening. (b) Cranial computed tomography scans of the same patient, showing the complex congenital C1, odontoid, and clivus configuration.

basilar invagination [22]. There were 4 patients with atlas assimilation in our study, aged 21, 15, 16, and 13 years. The greatest amount of CMA correction was achieved in these patients, as was also mentioned in Menezes' report. This finding supports that after decompression and realignment, posterior occipitocervical fusion involving intraoperative cervical distraction and extension is a reasonable option, particularly in children with atlas assimilation and basilar invagination.

Compression of the brain stem due to dural thickening (or dural band) appeared as a reason for revision surgery in 1 of our patients with atlas assimilation (Figure 2a). Idiopathic dural thickening at the craniocervical junction has been reported in some patients with Chiari type 1 malformation as a probable causative factor for neural structure compression and syringomyelia [23,24]. This pathologic entity should be considered by the surgeon, and preoperative MRIs should be evaluated from the aspect of probable dural thickening by the neuroradiologist. If there is a high suspicion of dural thickening preoperatively, dural widening with graft application should be done in advance during the operation. On T2-weighted MRI, a hypointense signal, consistent with fibrotic thickening, appearing as a dural tail at the foramen magnum area, particularly in patients with atlas assimilation, creates a high suspicion of dural thickening (dural band); this finding was present on the preoperative MRIs of our patient (Figure 2a). On the other hand, there is a group of patients in whom basilar invagination occurs as a result of a congenital anatomical anomaly, and besides an irreducible upward-settled odontoid, the odontoid can be inaccessible due to a bizarre configuration of the clivus, occiput, C1, and C2 complex (Figures 3a and 3b). In these patients, odontoidectomy procedures can result in failure because of this bizarre configuration. Additionally, the above-mentioned posterior dural thickening can be present at the periodontoid area anteriorly (Figure 3a). Even when we assume that successful odontoid resection has been performed, probable dural and tectorial ligament thickening and periodontoid fibrosis will prevent satisfactory neural decompression in such patients. In these cases, posterior foramen magnum decompression with dural graft, C1 arch resection, and odontoid reduction with an occipitocervical rigid fixator can be an alternative solution.

# Conclusion

In conclusion, intraoperative reduction and fixation with posterior decompression through a single-stage posterior approach should be the first choice of treatment for patients with basilar invagination and craniocervical instability causing progressive neurologic deficit. In particular, patients with basilar invagination due to atlas assimilation are the best candidates for this type of treatment. For patients who do not show relief of clinical findings despite these posterior treatment efforts, odontoid resection can be performed as a second procedure when there is persistent anterior compression at follow-up radiographic investigations. Nevertheless, dural thickening and fibrosis at the periodontoid area should be considered before performing odontoid resection through detailed evaluation of preoperative MRIs together with a radiologist.

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