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 improvements 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 craniocervical 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
Surgical technique

Under general anesthesia, in the prone position gentle traction was applied, and the odontoid was applied by the surgeon under fluoroscopic control for reduction (combined forces of distraction and extension) of the odontoid. Computed tomography (CT) scans also were obtained when necessary. 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.

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 thin-slice 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 (McRaes line) were impossible to draw on postoperative images, and were not recorded.

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.

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

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

| Patient No. | CMA, Deg | ADI, mm | WL, mm | CL, mm | ML, mm |
|-------------|----------|---------|--------|--------|--------|
|             | 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      | 3      | 0      | 4     | -      | -2    | -      |
| 11          | 80       | 80      | 3      | 3      | 3      | 0      | 28    | -      | -2    | -      |
| 12          | 105      | 105     | 3      | 3      | 3      | 0      | 12    | -      | -2    | -      |

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
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° ± 16.12°, while the mean decrease in the WL after surgery was 2.58 ± 1.68 mm. These differences were statistically significant (p = 0.007 and 0.006, respectively). These radiographic findings as well as reappearance of the subarachnoid 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 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 (arrow). The patient underwent a reoperation for dural decompression and graft application (asterisk = metallic artefact of the fixation system).

![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.](image1)

![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 due to dural thickening, despite bony decompression (arrow). The patient underwent a reoperation for dural decompression and graft application (asterisk = metallic artefact of the fixation system).](image2)
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 resection 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 craniovertebral 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 the odontoid through a single-stage posterior approach [8-10,17-19].

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.

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