Revision Surgery After Inappropriate Posterior Fossa Decompression For Craniocervical Junction Malformation

Li-feng CHEN¹, Yang YANG², Xin-guang YU¹, Bo BU¹, Bai-nan XU¹, Ding-biao ZHOU¹
¹The Chinese PLA General Hospital, Beyin ve Sinir Cerrahi Anabilim Dalı, Pekin, Çin ²The Chinese PLA General Hospital, Geriatrik Nöroloji Anabilim Dalı, Pekin, Çin

Summary

Background: A variety of treatment options have been described for craniocervical junction malformation. The authors described a revision surgical technique with intraoperative CT navigation system for treatment of craniocervical junction malformation associated with inappropriate excessive posterior fossa decompression.

Methods: The 12 patients with Chiari I malformation, basilar invagination, and syringomyelia, which were performed extensive posterior fossa decompression in other institutions received the revision surgery from April 2009 to March 2012. Clinical presentation, diagnose, treatment, therapeutic outcome and follow-up results of the patients were retrospectively studied.

Results: 8 patients were performed the posterior fixation and fusion in a reduced position. 3 patients with irreducible craniovertebral junction were only performed the posterior occipitocervical fixation, whereas 1 patient was performed the posterior occipitocervical fixation, and a transoral odontoidectomy was performed as the second procedure. The postoperative stability of the region and decompression of the brainstem was confirmed radiologically, and the 11 patients were relieved of their symptoms. At 9-24 months of follow-up, respectively, solid bone fusion was observed between the occipital bone and axis in all patients.

Conclusions: An excessive large posterior fossa and upper cervical canal decompression should be avoided for basilar invagination associated with Chiari I malformation. The revision occipitocervical fixation and fusion with the intraoperative CT and neuronavigation for treatment of craniocervical junction malformation with inappropriate excessive posterior fossa decompression can be successfully performed. Ventral decompression via the transoral route should be considered in the patients whose clinical symptoms had not been improved after the posterior decompression and fixation.

Key words: Revision surgery; Basilar invagination; intraoperative computed tomography; neuronavigation system; Occipitocervical fixation
Kranioservikal Bileşke Malformasyonlarında Uygunsuz Posterior Fossa Dekompresyonu Sonrası Revizyon

Özet

Giriş: Kranioservikal bileşke malformasyonu için birçok tedavi seçeneği tanımlanmıştır. Yazarlar uygunsuz olarak aşırı posterior fossa dekompresyonu uygulanan Kranioservikal bileşke malformasyonunda intraoperatif BT-navigasyon sistemi ile uygulan bir revizyon cerrahi tekniği tanımlamaktadırlar.


Yargı: Chiari Tip I malformasyonu ile birlikte olan baziler invaginasyon olgularında aşırı geniş posterior fossa ve üst servikal kanal dekompresyonunun kaçınılmak gerekir. İntraoperatif BT ve nöronavigasyon kullanarak oksipitoservikal fiksasyon ve füzyon uygulanması böyle uygunsuz aşırı posterior fossa dekompresyonlarını başarılı şekilde tedavi eder. Posterior dekompresyon ve fiksasyon sonrası klinik semptomları düzelmeyen hastalarda transoral yolla ventral dekompresyon mutlaka düşünülmelidir.

Anahtar Kelimeler: Revizyon cerrahisi; Baziler invaginasyon; intraoperatif bilgisayarlı tomografi; nöronavigasyon sistemi; Oksipitoservikal fiksasyon

INTRODUCTION

Chiari I malformation, basilar invagination, and syringomyelia have been under discussion and evaluation for over a century. A variety of treatment options have been described(2,3,7,9,15,18,21,22). Chiari I malformation generally is treated only with posterior fossa decompression(17). There are several surgical options to treat Chiari I malformation and basilar invagination. Simple posterior fossa decompression may aggravate postoperative instability of the region for Chiari I malformation accompanied with instability of the occipitocervical junction, and clinical symptoms got more serious(3,7,11,15). Because of multiple changes in geometry and the abnormal development of bony structures and vertebral artery anomalies in craniovertebral junction malformation, posterior fixation procedures are obviously complex(24,28). Surgical techniques of occipitocervical fixation and fusion for the patients with the extensive posterior fossa decompression are more technically challenging.

We have accumulated surgical experience using the revision rigid occipitocervical fixation technique with the intraoperative CT navigation for the patients with the extensive posterior fossa decompression. In this article, we will review our experience with 12 patients, describe the technique, the long-term follow-up results, and discuss the advantage of the technique.

MATERIAL AND METHODS

Patients

During a 3-year period between April 2009 and March 2012, we have surgically treated 12 patients with Chiari I malformation, basilar invagination, and
syringomyelia, which were performed extensive posterior fossa decompression in other institutions. The 12 patients included 5 females and 7 males. The mean age was 37.4 years and ranged from 16 to 51 years. The amount of time between the initial decompressive surgery and evaluation at the Department of Neurosurgery in our hospital ranged from 6 months to 2 years (average 8.7 months). 8 patients (66.7%) were performed extensive posterior fossa decompression, 4 patients (33.3%) were performed extensive posterior fossa and upper cervical canal decompression, and 2 patients (16.7%) were performed transoral odontoidectomy (Table 1). The neurological function of the patients was graded according to the scale described by McCormick, et al (Table 2)(14). KPS scores were also determined for each patient based on clinical evaluations. The clinical information was obtained by hospital charts, clinic notes, and operative reports.

<p>| Table 1 Characteristics in 12 patients with Chiari I malformation and basilar invagination |
|---------------------------------|-----------------|----------------|----------------|----------------|----------------|------------------|------------------|</p>
<table>
<thead>
<tr>
<th>case No.</th>
<th>Age(y)</th>
<th>Sex</th>
<th>Main diagnosis</th>
<th>Concomitant</th>
<th>Previous surgery</th>
<th>Fixation area</th>
<th>Preop Mc grade</th>
<th>Complication</th>
<th>F/U (mos)</th>
<th>F/U Mc grade</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>16,M</td>
<td></td>
<td>CM,AA,BI</td>
<td>-</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>II</td>
<td>NA</td>
<td>24</td>
<td>I</td>
</tr>
<tr>
<td>2</td>
<td>35,F</td>
<td></td>
<td>CM, AA, AAD</td>
<td>Syringomyelia, cerebellar ptosis</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>II</td>
<td>NA</td>
<td>24</td>
<td>I</td>
</tr>
<tr>
<td>3</td>
<td>51,F</td>
<td></td>
<td>CM,AA,AAD</td>
<td>-</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>II</td>
<td>NA</td>
<td>24</td>
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</tr>
<tr>
<td>4</td>
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<td>PFDD</td>
<td>C0-C2-3</td>
<td>IV</td>
<td>NA</td>
<td>18</td>
<td>I</td>
</tr>
<tr>
<td>5</td>
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<td></td>
<td>CM,AA,BI,AAD</td>
<td>C2-3 fusion</td>
<td>PFDD</td>
<td>C0-C2-4</td>
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</tr>
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<td>syringomyelia</td>
<td>PFDD</td>
<td>C0-2</td>
<td>II</td>
<td>NA</td>
<td>18</td>
<td>I</td>
</tr>
<tr>
<td>7</td>
<td>39,M</td>
<td></td>
<td>CM, AA, BI</td>
<td>-</td>
<td>TO+PFDD</td>
<td>C0-2</td>
<td>III</td>
<td>NA</td>
<td>18</td>
<td>II</td>
</tr>
<tr>
<td>8</td>
<td>45,F</td>
<td></td>
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<td>PFDD</td>
<td>C0-C2-3</td>
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<td>NA</td>
<td>12</td>
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</tr>
<tr>
<td>9</td>
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<td>syringomyelia</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>II</td>
<td>NA</td>
<td>12</td>
<td>I</td>
</tr>
<tr>
<td>10</td>
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<td>-</td>
<td>TO+PFDD</td>
<td>C0-C2-3</td>
<td>III</td>
<td>NA</td>
<td>12</td>
<td>I</td>
</tr>
<tr>
<td>11</td>
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<td></td>
<td>CM,AA,AAD</td>
<td>Syringomyelia, cerebellar ptosis</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>III</td>
<td>NA</td>
<td>12</td>
<td>I</td>
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<tr>
<td>12</td>
<td>46,M</td>
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<td>syringomyelia</td>
<td>PFDD</td>
<td>C0-C2-3</td>
<td>II</td>
<td>NA</td>
<td>9</td>
<td>I</td>
</tr>
</tbody>
</table>

CM=Chiari I malformation; AA=atlas assimilation; BI=basilar invagination; AAD=atlantoaxial dislocation; NA=not applicable; preop Mc grade=preoperative McCormick clinical grade; F/U Mc grade= follow-up McCormick clinical grade; F/U= follow-up.

| Table 2 McCormick clinical grading scale for neurological function * |
|-----------------|-----------------|
| Grade | Definition |
| I | Neurologically normal, mild focal deficit not significantly affecting function of involved limb, mild spasticity or reflex abnormality, normal gait |
| II | Presence of sensorimotor deficit affecting function of involved limb, mild to moderate gait difficulty, severe pain of dysesthetic syndrome impairing patient’s quality of life, still functions and ambulates independently |
| III | More severe neurological deficit, requires cane/brace for ambulation or significant bilateral upper extremity impairment, may or may not function independently |
| IV | Severe deficit, requires wheelchair or cane/brace with bilateral upper extremity impairment, usually not independent |

Surgical technique and Workflow

We worked with the intraoperative CT unit during surgical procedure. The setup of the operating room (Fig.1) and CT workstation were introduced in our previous article (28). After general anesthesia, the patient was placed on the operating table in the prone position with the head moderately flexed using a radiolucent head clamp. The original midline incision was performed to expose the region from the external occipital protuberance to the spinous process of the 4th cervical vertebra. Care must be taken to decrease the risk of dural laceration and cerebrospinal fluid leakage during the operation because of the expansive posterior fossa decompression and occipital bone defect.

An initial CT scan was performed to localize the desired surgical level. Multiplanar reconstructions were performed. The intraoperative CT imaging data were transferred to the neuronavigation system (Vector Vision; BrainLab, Germany) and registered. Axial and sagittal CT scan was conducted in combination with the neuronavigation system to determine screw trajectory and to ascertain whether the screws had penetrated a pedicle wall (Fig.2). The pedicle screws were inserted into the pedicle on C1, C2, C3 or C4 bilaterally. On the cranial side, the range of occipital bone defect and the thickness of the residual occipital bone were estimated with the intraoperative neuronavigation system. The goal was to find an adequate site of residual occipital bone that could accept the screw without causing injury of transverse sinus and dura. Adjusted the radiolucent head clamp with the head cephalic traction and more extended. Intraoperative CT scan was performed to confirm reducing the atlanto-dens interval and reduction of occipitocervical junction. One rod was contoured to span from the occipital screws to the cervical instrumentation. The occipital screws and the cervical instrumentation were connected with the rod. For calibration references, we used small titanium screws that were inserted into the spinous process before the intraoperative scans were performed, and they were removed afterwards (28). We used screws of 3.5 mm in diameter and 6-34 mm in length (Vertex, Sofamor Danek).

Autologous bone flap was secured between the rostral decorticated edge of residual occipital bone and decorticated lamina of axis with titanium wires or coarse wires. Post-operatively the neck of the patient was immobilized with a cervical collar for at least 3 months.

Fig 1: Photographs of setup of the intraoperative CT navigation system. (A) multislice computed tomographic scanner with a sliding gantry. (B) The radiolucent head clamp. (C) The radiolucent operating table. (D) Navigation reference frame. (E) Touch LCD. (F) Infrared locator. (G) Anesthesia machine.
RESULTS

Preoperative Characteristics

The instability of the occipitocervical junction and clinical symptoms got more serious followed the initial decompressive surgery. The most common symptoms were motor deficit 12(100%), sensitive deficit 7 (58%), and gait/coordination abnormalities 6 (50%). Of the patients, three(25%) had hoarseness and difficulty in breathing. Three(25%)patients got cerebellar ptosis. Syringomyelia occurred in 8 patients. The range of preoperative McCormick grade was II- IV. The range of preoperative KPS score was 50–80. The preoperative diagnosis and other characteristics for the combined series of 12 cases were shown in the Table 1.

Surgical characteristics and results

8 patients with reducible craniovertebral junction were performed the posterior fixation and fusion in a reduced position. 3 patients with irreducible craniovertebral junction were only performed the posterior occipitocervical fixation, whereas 1 patient with irreducible craniovertebral junction was performed the posterior occipitocervical fixation, and a transoral odontoidectomy was performed as the second procedure. Dural tear was detected in 1 patient. The patient had fever after surgery. She was cured after treatment with antibiotics and lumbar drainage in one week. No other patient had dural, neural, or vertebral artery injury, infections, CSF leakage, or pneumonia. 38 occipital screws and 52 lateral mass or pedicle screws including 4 C1 lateral mass screws, 24 C2, 20 C3 and 4 C4 pedicle screws were inserted into the 12 patients. Intraoperative CT neuronavigation was found to correlate well with the intraoperative findings and the recalibration was uneventful in all cases. 1 (1.9%) patient was found C2 pedicle perforation, and no any neural or vessel injury.

Postoperative Course

Patients were reexamined during scheduled visits at 3 months, 6 months, and 12 months postoperatively and intermittently as needed. Symptomatic improvement was achieved in 11 patients. The 11 patients (92%) were improved by at least 1 McCormick grade, whereas the grade did not change in 1 patient (8%). Transoral odontoidectomy as the second procedure was performed in the patient. Her clinical symptoms improved in the follow-up. The range of 6 months postoperative McCormick grade was I- III and the range of 6 months postoperative KPS score was
Postoperative CT and MRI showed that the goal of decompression of the neuraxis and fixation was achieved in 11 patients. Syringomyelia in 7 (87.5%) patients improved. Cerebellar ptosis of the 3 patients resolved. No patient developed progressive symptoms or dislocation during follow-up period ranged from 9 to 24 months (mean 17 months).

Illustration of cases

A 26-year-old man underwent expanded posterior fossa and upper cervical canal decompression in another hospital because of basilar invagination, partially reducible atlantoaxial dislocation, occipitalization of the atlas. Rapid deterioration ensued in the patient at 12 months after surgery. He got weakness and numbness of limbs. Physical examination showed right extremities weaknesses were grade 2/5 and left extremities weakness were grade 3/5. Babinski sign was positive in bilateral lower extremity. Sagittal MRI scan showed severe ventral compression of the cervicomedullary junction, serious cerebellar ptosis after a newly created expanded posterior fossa decompression, and syringomyelia that extended from C2–T2 (Fig. 2–3).

Preoperative CT showed dysplasia of the C1 lateral mass (Fig. 2). Intraoperative simulation of pedicle screws insertion of C2, C3 and C4 by use of the computerized navigation system demonstrated that screw insertion would be safe on both sides. The residual occipital bone was suitable for screw implantation. The original midline incision was performed. The region from the external occipital protuberance to the spinous process of the 4th cervical vertebra was exposed. Intraoperative CT scan and multiplanar reconstructions were performed. 6 pedicle screws and 5 occipital screws were inserted with intraoperative CT navigation system (Fig. 2). Adjusted the radiolucent head clamp with the head cephalic traction and more extended. Intraoperative CT scan was performed to confirm reducing the atlanto–dens interval and reduction of occipitocervical junction. One rod was contoured to span from the occipital screws to the cervical instrumentation. The occipital screws and the cervical instrumentation were connected with the rod.

Iliac bone graft was secured between the rostral decorticated edge of residual occipital bone and decorticated lamina of axis with coarse wires. The neurological deficits of patients gradually recovered after surgery. Postoperative MRI revealed significant reduction of cervicomedullary compression, tonsillar herniation, cerebellar ptosis, and syringomyelia (Fig. 3). CT scan obtained at 6 months after the operation demonstrated bony fusion between occipital bone and axis (Fig. 2). The patient got remarkable recovery from the presenting symptoms.

Fig 3: (A) MRI showing ventral compression of the cervicomedullary junction before the extensive posterior fossa decompression. (B) MRI showing basilar invagination, and subcutaneous fluid at 6 months after the first surgery. (C) MRI showing basilar invagination, cerebellar ptosis, downward displacement of tonsil, and syringomyelia at 12 months after the first surgery. (D) MRI showing significant reduction of cervicomedullary compression, tonsillar herniation, cerebellar ptosis, and syringomyelia at 6 months after occipitocervical fixation and fusion.
DISCUSSION

It seems that craniovertebral anomalies are more frequently found in China than anywhere else in the world. Perhaps, it has relationship with the ethnic and economic condition. The presence of concomitant bone abnormalities with Chiari I malformation and syringomyelia has been well described (3,7,8,22). A variety of treatment options have also been described about the patients with basilar invagination associated with Chiari I malformation and syringomyelia (3,12,20). The need for stabilization with instrumentation in the patients remains under discussion. Some authors reported a fixation procedure could be avoided and recommended anterior surgery (12,25,27). Other authors suggested that there was potential or manifest instability in this complex anomaly, and a fixation procedure was necessary (1,4,9,11,16,20). Nishikawa et al (19) described the importance of simultaneous performance of both posterior fossa decompression and occipitocervical fixation in patients with Chiari I malformation associated with craniovertebral junction instability. However, the surgical indications for a given approach together with the timing of the surgical stages are still controversial.

In our series, the patients had undergone a previous primary operation consisting of excessive posterior fossa and upper cervical canal decompression in other institutions. Rapid deterioration ensued in these patients or an initial improvement was followed by progressive neurological deterioration. MRI in each of these instances showed an increase in the ventral cervicomedullary junction compression and the angulation secondary to the peg-like basilar invagination. Another complicating feature was cerebellar ptosis after a newly created expanded posterior fossa decompression, and this further worsened the situation as well as caused further impaction at foramen magnum. The stabilization was paramount to maintain the neural decompression for the special patients. Revision occipitocervical reconstruction for this kind of patients presented many serious challenges and issues because of the extensive bone defects, cerebellar ptosis and the surgical scars.

Since Foerster (6) first attempted occipitocervical arthrodesis using a fibular graft in a case of progressive atlantoaxial dislocation in 1927, various instrumentation systems or procedures for internal fixation have been developed in the past century. Most of these consist of rigid and nonrigid fixation constructs, including cables, hooks, screws, rods, and plates (6,13). Each fixation construct has its particular advantages and disadvantages (6). Nishikawa et al (19) demonstrated posterior fossa decompression and simultaneous occipitocervical fusion with an intradiploic screw fixation technique in the patients with Chiari I malformation and basilar invagination. The occipital diploic layer of the selected patients should be robust enough to accept a screw and excessively long screws would be prone to break out because the occiput was roughly spherical in shape. Some authors (3,7-9,18,22) showed a foramen magnum craniotomy followed by occipitocervical instrumentation with lateral fixation and a plate and screw on each side. Because the C1–C2 screw fixation had biomechanically been demonstrated to be superior in providing immediate rigidity and eliminating the need for external immobilization when compared with other methods, leading to higher rates of fusion, the method was the “gold standard” for occipital fixation (13).

But the specific anatomical constraints caused by congenital abnormalities, such as a high-riding transverse foramen, small or narrow pedicles of C2, dysplasia of the C1 lateral mass, may preclude placement of the screws (26), and there was a risk of
dural, neural, or vertebral artery injury from errant screw placement\textsuperscript{(24,26)}. The recent development of image guidance systems has increased the percentage of successful screw insertions and decreased risk of complications associated with screw malposition\textsuperscript{(10,28,29)}. According to meta-analysis\textsuperscript{(23)}, the rate of penetration of the pedicle cortex by an inserted screw without the use of navigation was almost 10\%, and with the use of spinal navigation, the rate of screw misplacement improved considerably. In our series, the combination of intraoperative CT and neuronavigation provided the choice of suited lateral mass or pedicle for us. It improved the accuracy of screw placement and avoided the risk of dural, neural, or vertebral artery injury. The overall accuracy of screw placement was 98.1\%. No major complications occurred in all the patients. As some pedicles of C2 were narrow, The C3 or C4 screw fixation could provide superior immediate postoperative stability. Residual occipital of the patients was different in shape, and little bone was left for fixation in our series. The flexibility of the rods according to the shape of residual occipital provided by intraoperative CT neuronavigation was the reason that it was used in our instrumentation. No patient was detected the penetration of the occipital cortex by an inserted screw. Another advantage of this technique was rigid stability. Rigid stability increased osseous fusion rate. All the patients in our series got osseous fusion in the follow-up. Moreover, categorizing patients according to McCormick functional scale is useful to assess operative outcomes in perioperative period. The scale provides a practical means of comparing pre- and postoperative neurological condition. 11 patients (92\%) were improved by at least 1 McCormick grade, whereas the grade did not change in 1 patient (8\%) in our series.

It should be emphasized that an excessive large posterior fossa and upper cervical canal decompression is not usually necessary for a Chiari malformation\textsuperscript{(23)}. This can lead to cerebellar ptosis and the reemergence or recurrence of Chiari-related symptoms. Cerebellar ptosis is an infrequent and under recognized potential delayed complication following craniocervical decompression for Chiari I malformation. The condition is principally caused by a suboccipital craniectomy that is too large for the individual patient. Partial suboccipital cranioplasty is successful in treating the headache by supporting the cerebellum and alleviating stretching the dura mater. In our series, 3 patients got cerebellar ptosis, and their clinical symptoms of the patients got more serious. Iliac bone graft was an effective substitution of suboccipital cranioplasty, and cerebellar ptosis of the patients resolved.

Although the indications for simultaneous decompression and fixation in patients with basilar invagination associated with Chiari malformation is controversial\textsuperscript{(23)}, our impression is that it is necessary that simultaneous fixation and fusion after an appropriate posterior decompression in the patients of basilar invagination associated with Chiari malformation. If clinical symptoms of the patients had improved after the posterior decompression and fixation, anterior decompression could be avoided. Ventral decompression via the transoral route should be considered in the patients whose clinical symptoms had not improved after the posterior decompression and fixation. Transoral odontoïdectomy as the second procedure was performed in 1 patient whose clinical symptoms had not improved after the revision posterior fixation in our series.

CONCLUSION

An excessive large posterior fossa and upper cervical canal decompression should be avoided for basilar invagination associated with Chiari I malformation. The revision occipitocervical fixation and fusion with the intraoperative CT and
neuronavigation for treatment of craniocervical junction malformation with inappropriate excessive posterior fossa decompression can be successfully performed. Ventral decompression via the transoral route should be considered in the patients whose clinical symptoms had not been improved after the posterior decompression and fixation.

Correspondence to:
Xin-guang Yu
E-mail: javelins@126.com

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