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Variation of craniocervical junction volume as an effective parameter for basilar invagination treatment

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Abstract. – OBJECTIVE: The major pathological change in basilar invagination (BI) is represented in the decrease of craniocervical junction (CVJ) volume resulting from abnormal bone protrusion around the foramen magnum. The diagnosis and clinical evaluation of BI is mainly based on the clinical manifestations and radiographic measurements by means of calculation of the scan lines of CVJ in X-ray, CT and MRI. With the transoral decompression atlantoaxial reduction plate (TARP III) system, the decompression, reduction and fixation can be achieved to decompress and stabilize medulla spinalis change the position of the dens in CVJ, thus expand the CVJ relative volume, relieve the compression on medulla spinalis and the nerve injury. However, the correlation between the dens position change and the variation of CVJ has not been established previously. This study focused on the clinical significance of the variation of craniocervical junction (CVJ) volume caused by the dens position change for the treatment of BI.

PATIENTS AND METHODS: We've performed an analysis of data from 62 BI patients admitted from January 2008 to May 2013, who were treated by TARP III system. The data include preoperative, postoperative JOA scores (Japanese Orthopaedic Association scores, 17 points method), preoperative and postoperative X-ray, thin-slice CT scan with three-dimensional reconstruction and MRI scan to measure the cervicomedullary angle (CMA). We have analyzed the preoperative and postoperative three-dimensional CT data by means of MIMICS 10.01 software system according to the Box volume (BV) method to determine the changes of CVJ volume resulting from preoperative and postoperative dens position change, assessed the correlation between the CVJ volume changes and the JOA scores with correlation between CMA change and the JOA scores. All data were analyzed by paired *t*-test and Pearson correlation analysis.

RESULTS: In all 62 patients, JOA scores were recovered from preoperative 9.26 ± 1.66 to postoperative 13.02 ± 1.44, CMA change rate was 21%, and CVJ volume change rate was 36%. The CMA change rate and the JOA score recovery rat exhibited relevance, as Pearson's correlation coefficient was 0.46 ($p < 0.005$). The Pearson's correlation coefficient between CVJ volume change rate and JOA score recovery rate was 0.63 ($p < 0.005$), and the CVJ volume change rate was significantly different while compared with the correlation between CMA change rate and JOA score ($p < 0.005$).

CONCLUSIONS: the CVJ volume change rate is a sensitive and reliable parameter for the evaluation of neurological function improvement in patients with BI. It can be used as a predictor to evaluate the postoperative neurological recovery.

Key Words:
Basilar invagination, Dens, Volume, Nerve function.

Introduction

The basilar invagination (BI) is a rare congenital or acquired craniocervical junction (CVJ) progressive abnormality. The diagnosis of BI primarily depends on the measurement of radiographic scan lines in CVJ and relevant clinical manifestations[1-5]. Up to now, most treatments are focusing on the direct decompression via posterior fossa and relieving the compression of *medulla spinalis* ventral from dens[6-10]. Posterior screw-rod indirect distraction, fixation and fusion (recoverable BI) and the anterior release and decompression together with posterior screw-rod

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fixation and fusion (irrecoverable BI) can indirectly alter the position of the dens, thus relieve the compression of *medulla spinalis*, achieve the goal of expanding CVJ space volume[2,11-13]. Transoral direct surgical resection of dens together with subsequent posterior fixation and fusion can also relieve the compression. However, both risks and costs will be significantly increased due to two prerequisite operations[2,12,14].

In this study, we have performed a retrospective analysis of the radiographic data from 62 patients that were admitted from January 2008 to May 2013 due to BI. The patients completed the first phase transoral anterior release and transoral atlantoaxial reduction plate (TARP) fixation and fusion. We have analyzed the correlation between preoperative, postoperative dens position change, postoperative nerve function recovery (JOA scores, 17 points method) and cervicomedullary angle (CMA) to investigate the potential correlations among CVJ volume change, CMA and nerve function recovery after decompression.

Patients and Methods

From January 2008 to May 2013, 62 patients in our hospital were definitively diagnosed as BI and received surgeries. The exclusion criterion was the previous surgical treatment (either anterior or posterior) with unsatisfied therapeutic efficacy. The patients included 29 males and 33 females, aged from 19 to 71 years old (average age of 42.1). The average preoperative JOA scores were 9.26 ± 1.66 and the average preoperative cervicomedullary angle (CMA) was 110-142° (average 126 ± 12°). The postoperative results are shown in Table I, involving a 59-year-old female patient whose preoperative JOA score was 6 and the postoperative result was 0. The CTA scan showed that the bilateral vertebral artery embolization resulted in the cerebral edema and she died after operation one month later.

CT-scan

By means of Siemens Dual Source CT scan, we analyzed the bone tissue window fault from the inion to the 7th cervical vertebrae. The voltage was 120 KV with the pixel of 0.43 mm and slice of thickness of 0.625 mm. The data were saved in DICOM format and analyzed by means of the Mimics 10.01 software (Materialized interactive medical image control system, Belgium) to reconstruct a three-dimensional model of CVJ and calculate the volume change.

MRI

By cervical coil of the Intra-Operative Magnetic Resonance Imaging System (IMRIS, GE Sigma HDX 3.0T and Siemens Snata 1.5), the sagittal images of cervical vertebrae T2W1 (TR 2600-3000 ms, TE 90-100 ms, thickness of 3 mm, space of layers of 1 mm, FOV of 240 mm × 240 mm), T1W1 (TR500-550 ms, TE10-20 ms, thickness of 3 mm, space of layers of 1 mm, FOV 240 × 240 mm and transverse images of T2WI I (TR 3000-3500 ms, TE 90-100 ms, thickness of 3 mm, space of layers of 0.5 mm, FOV of 200 mm × 200 mm) were collected.

CMA Measurement

On the MRI sagittal image of CVJ, the angle of medulla ventral surface and the parallel lines of cervical intramedullary ventral surface were measured according to the method (Figure 1) by Bundschuh et al[15]. The changing rate of CMA = postoperative CMA-preoperative CMA/preoperative CMA (results is showed in Table II).

CVJ Volume Measurement

The establishment of Box volume (BV): the data of CT-scans from BI patients were analyzed by means of the Mimics 10.01 software to reconstruct a three-dimensional model of CVJ. The redundant data were excluded by threshold method. With the function of cavity fill and crop mask, the object region was constructed.

Table I. Radiographic results of preoperative and postoperative BI patients (mean ± SD, n = 62).

	(CMA) (°)	CVJ Box volume (mm^3)	JOA score
Pre- (mean)	126 ±12	19007.9 ± 1200.2	9.3 ± 1.7
Post- (mean)	150 ± 9	25898.9 ± 1326.1	13.0 ± 1.4
Change rate (%)	21	36	3.4 ± 1.7

CVJ change rate = (post-Box volume - pre-Box volume)/pre-Box volume × 100%.

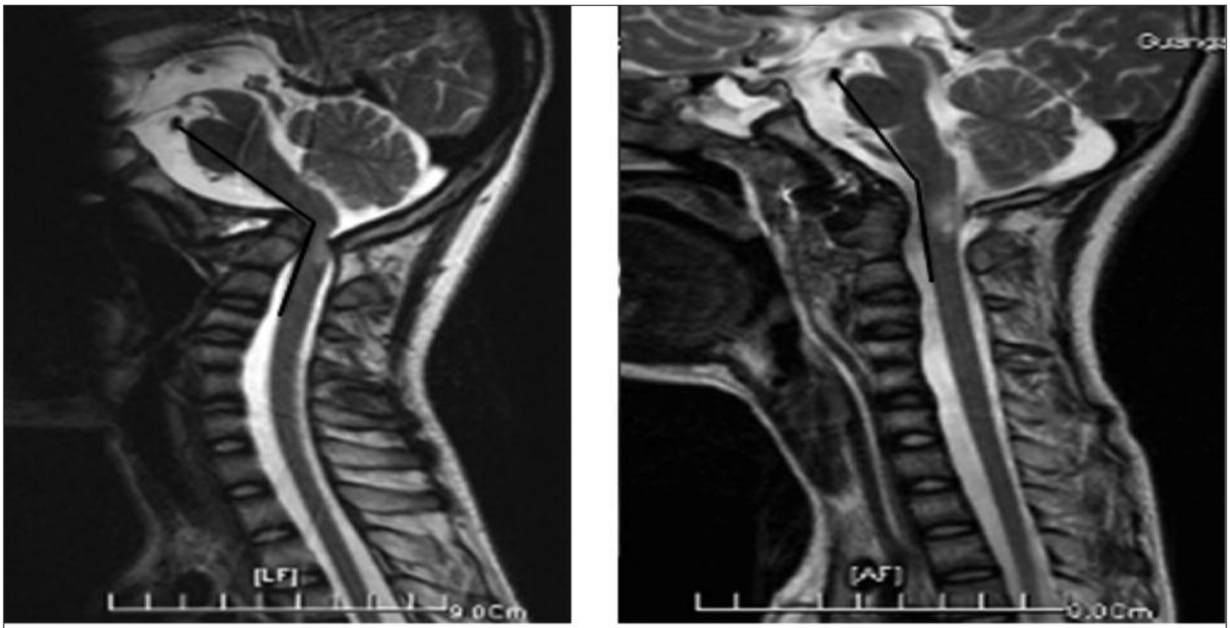

Figure 1. The measurements of preoperative, postoperative CMA by TARP III treatment.

At this step, the measurement area was set (anterior and posterior boundary was the anterior hyaloid and posterior hyaloid of the inferior margin of sagittal *foramen magnum*; upper boundary was the plane of dens tip; inferior boundary was the inferior of axis). A 3D model of Box volume in the object region was established in CVJ. The axis part in the BV was excluded from Boolean operation and the rest volume in the object region was calculated (Figure 2, ***1-2***). With the same method, a 3D model of postoperative CVJ data was established by 3D calculation. The same BV volume (upper, inferior, right and left boundary of BV was the same as the postoperative BV) was established, so the BV position from preoperative and postoperative remained the same. By the Boolean operation, we removed the position-changed axis and the rest BV is the postoperative BV in the object region (Figure 2, ***3-4***). The change in BV postoperative and preoperative was equal to the postoperative BV minus preoperative BV (Figure 2). The change rate of CVJ = postoperative BV – preoperative BV/preoperative BV.

Mechanism of Transoral Atlantoaxial Reduction plate

TARP III procedures can be referred from Figure 3.

Statistical Analysis

All data were shown as mean ± SD and analyzed by SPSS 16.0 (SPSS Inc., Chicago, IL, USA). Statistical comparisons were completed by paired *t*-test and correlations of BV with JOA score and CMA with JOA score were described by Pearson's coefficient. $p < 0.05$ is considered statistically significant.

Table II. Correlation between change rate of radiography results and JOA score in BI patients.

	Persons correlation coefficient	*p* value
CMA change rate	0.46	0.05
CVJ volume change rate	0.63	0.05

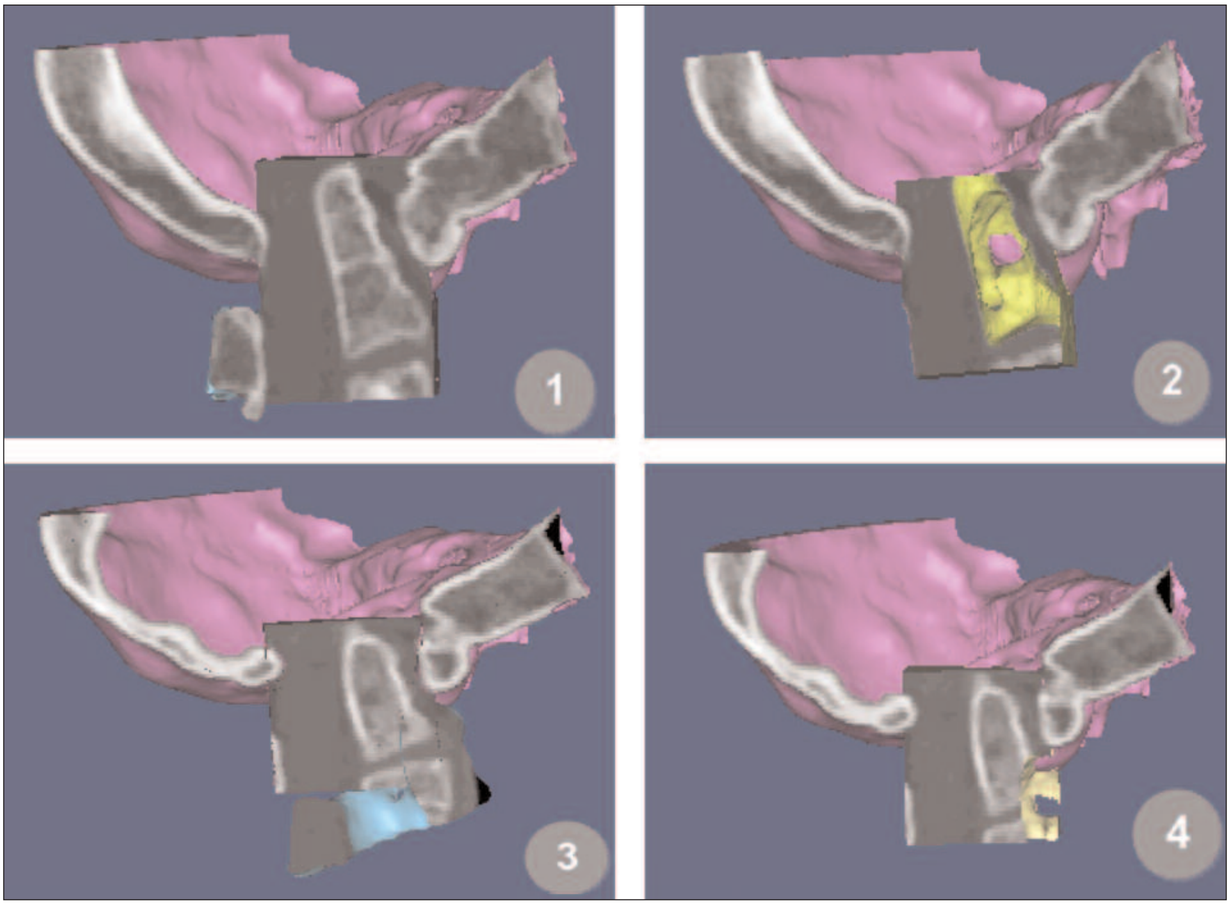

Figure 2. *(1)* The schematic diagram of CVJ measurement by Box volume method. *(1)* Preoperative box position in CVJ; *(2)* Structure and volume of box after resection of epistropheus; *(3)* Postoperative box position with epistropheus move down or move forward; *(4)* Structure and volume of box in CVJ after resection of epistropheus.

Results

CVJ change rate and CMA are correlative to the improvement of JOA score. The correlation of CVJ volume change rate with JOA was significantly different from that of CMA with JOA. The change of effective volume in CVJ region resulting from the dens position change is able to predict the prognosis of patients.

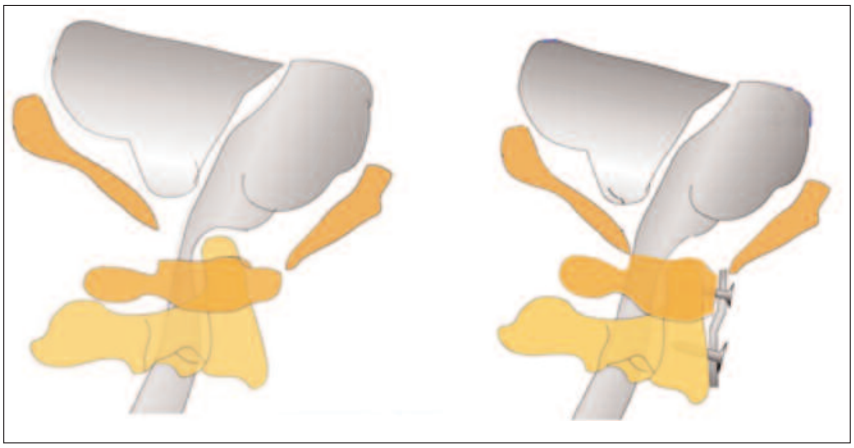

Figure 3. The schematic diagram of treating BI patient by TARP

Discussion

The main pathological change in BI is the abnormal protrusion of the bone around the *foramen magnum* caused by the decrease of the CVJ volume. The move-up of the apex of dens results in the compression of the brain stem and the medulla, thus, producing a series of neurological abnormalities. BI is often accompanied by other deformities, such as clivus dysplasia, atlanto-occipital assimilation and atlanto-axial assimilation[16,17]. The radiographic diagnosis relies on the measurement of the scan lines by X-ray, CT and MRI, which mainly consist of Chamberlain's line, clivus baseline, basal angle and CMA, etc.

The CT tomography and the three-dimensional reconstruction can clearly visualize abnormal bone structures; therefore, the two-dimensional CT measurement of anatomical marks is better than the X-ray measurements. With the three-dimensional reconstruction, the anatomical position changes of superior cervical bone structure are even clearer, but the correlation of CVJ with brain and spinal cord and the correlation between CVJ and clinical manifestations have not been previously investigated. With the digital imaging technology, we have analyzed the CT scan data using mimics software, built the Box volume and measured the preoperative and postoperative CVJ volumes. The results have shown that the move down or forward of dens position after surgery increased the CVJ effective volume (locational space of the *medulla spinalis*) around the dens, directly relieved the compression on *medulla spinalis* and improved nerve function. The instant JOA score also significantly improved, CVJ volume change rate and JOA score showed a clear correlation (Pearson's coefficient = 0.63, $p < 0.05$). These results implied that the change of CVJ volume rate can be used as one of important indicators to evaluate the effectiveness of surgical decompression.

From the MRI images of CVJ region, we can clearly determine the pathological changes of the brain stem, the medulla, the spinal cord, ligaments and other soft tissue structures surrounding of the CVJ. However, the imaging of bone structures is inferior to CT and the scan lines mentioned above can indirectly reflect the pathologic conditions of the brain, spinal cord after being pinched. In addition, only CMA measured by MRI can directly reflect the degree of compression on the nerve structure in the CVJ region without being affected by radiographic magnification. Previous studies have shown inconsistent results on the correlation between CMA and the improvement of clinical manifestations[4,17,18]. Bunschuh et al[15] suggested that patients with CMA < 135° exhibited cervical myelopathy, brain-stem compression or C2 radiculalgia. With important clinical significance, CMA measurement can be used to evaluate the severity of the BI. CMA less than 135° has obvious relevance to myeleterosis or symptoms of brain-stem compression. The increased mean CMA after decompression and symptom improvement were also significantly correlated. Reiter et al[19] found that the patients with CMA less than 135° were most suitable for posterior atlanto-axial joint arthrodesis. CMA can be used to evaluate the effects of CVJ surgery on relieving the compression of *medulla spinalis*. The postoperative CMA closer to the normal range indicates better effects of decompression surgeries. Reijnierse et al[20] previously reported that CMA < 135° was not correlated with the occurrence of the neurological symptoms. Wang et al[18] have also suggested that CMA should not be significantly correlated with age, sex, and etc., but it is a good indicator to evaluate the *medulla spinalis* compression caused by the skull base lesions. Besides, it can also be used as a significantly crucial guidance in terms of the treatment choice and parameter for the evaluation of treatment efficacy.

Our study showed that the instant postoperative JOA scores improved significantly in patients with significantly increased CMA and the improvement of CMA is definitely correlated with the JOA score (Pearson's coefficient = 0.46, $p < 0.05$); however, no correlation was observed with significantly increased CMA. This may be related to insufficient recovery of *medulla spinalis* function resulting from surplus or insufficient anterior distraction in the process of reduction.

Compared with CMA, the correlation between CVJ volume change rate and JOA score was significantly different and the results demonstrated that CVJ volume change rate was more sensitive in evaluating neurological function recovery, implying that the change in the space around dens may reflect the neurological function recovery of medulla in CVJ.

Theoretically, the BI surgeries on either the anterior or posterior CJV can relieve compression on the nerve to improve symptoms. The decompression by fenestration on posterior of *foramen magnum* can significantly improve the cord compression from the dorsal side, but the oppression from the ventral side cannot be relieved effectively. Currently, the screw-rod indirect reduction and decompression, or by ventral resection of dens and releasing on the ventral side, then indirect reduction posteriorly is the classic surgical treatment to release the dorsal compression by BI[6,8,21]. The risk is significantly increased due to two required operations[10]. We used TARP III to treat BI. During the first phase, direct devices distraction and reduction release posterior tissue, and TARP III direct fixation, the purpose of spinal cord decompression and fixation can be achieved, this surgical process can directly extend the space of decompression on CVJ medulla region. The success rate is higher because of the exact effect[22-26]. Although this study showed that the volume change of CVJ was clearly relevant to clinical JOA score. However, in some cases, for example, the dead patient in our group, it is clinically ineffective and some patients were even worsened in the clinical practice or the literature, which may attribute to the lack of position change in dens or the effective expansion in CVJ volume. Further studies are needed to achieve the desired volume to obtain the best recovery of neurological functions by preoperative design of the dens position change.

Conclusions

The change rates of CVJ volume and CMA are correlated to the improvement of neurological functions. CVJ volume change rate is more sensitive while compared to CMA. The CVJ volume change can be regarded as an important curative criterion for BI operation.

Acknowledgements

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Conflict of Interest

The Authors declare that there are no conflicts of interest.

References

1) Menezes AH. Honored guest presentation: craniocervical developmental anatomy and its implications. Clin Neurosurg 2005; 52: 53-64.
2) Menezes AH. Craniocervical developmental anatomy and its implications. Childs Nerv Syst 2008; 24: 1109-1122.
3) Smoker WR, Khanna G. Imaging the craniocervical junction. Childs Nerv Syst 2008; 24: 1123-1145.
4) Wang XW, Jian FZ, Chen Z, Wu H, Bao YH. An analysis of surgical outcome and influencing factors in patients of congenital basilar invagination with atlantal-axial dislocation: report of 120 cases. Zhonghua Wai Ke Za Zhi 2013; 51: 207-210.
5) Riew, K.D., Hilibrand AS, Palumbo MA, Sethi N, Bohlman HH. Diagnosing basilar invagination in the rheumatoid patient. The reliability of radiographic criteria. J Bone Joint Surg Am 2001; 83: 194-200.
6) Goel A, Shah A. Atlantoaxial joint distraction as a treatment for basilar invagination: a report of an experience with 11 cases. Neurol India 2008; 56: 144-150.
7) Goel A, Bhatjiwale M, Desai K. Basilar invagination: a study based on 190 surgically treated patients. J Neurosurg 1998; 88: 962-968.
8) Botelho, R.V., Neto EB, Patriota GC, Daniel JW, Dumont PA, Rotta JM. Basilar invagination: craniocervical instability treated with cervical traction and occipitocervical fixation. Case report. J Neurosurg Spine 2007; 7: 444-449.
9) Collignon FP, Cohen-Gadol AA, Krauss WE. Circumferential decompression of the foramen magnum for the treatment of syringomyelia associated with basilar invagination. Neurosurg Rev 2004; 27: 168-172.
10) Chandra PS, Kumar A, Chauhan A, Ansari A, Mishra NK, Sharma BS. Distraction, compression, and extension reduction of basilar invagination and atlantoaxial dislocation: a novel pilot technique. Neurosurgery 2013; 72: 1040-1053; discussion 1053.
11) Zileli M, Cagli S. Combined anterior and posterior approach for managing basilar invagination associated with type I Chiari malformation. J Spinal Disord Tech 2002; 15: 284-289.
12) Yu Y, Hu F, Zhang XB, Ge JQ, Sun CJ. Endoscopic transnasal odontoidectomy combined with posterior reduction to treat basilar invagination: technical note. J Neurosurg Spine 2013; 19: 637-643.
13) Wang S, Wang C, Yan Ming, Zhou HT, Jiang L. Syringomyelia with irreducible atlantoaxial dislocation, basilar invagination and Chiari I malformation. Eur Spine J 2010; 19: 361-369.
14) Goel A, Sharma P. Craniovertebral junction realignment for the treatment of basilar invagination with syringomyelia: preliminary report of 12 cases. Neurol Med Chir (Tokyo) 2005; 45: 512-517.

15) Bundschuh C, Modic MT, Kearney F, Morris R, Deal C. Rheumatoid arthritis of the cervical spine: surface-coil MR imaging. AJR Am J Roentgenol 1988; 151: 181-187.

16) Simsek S, Yigitkanli K, Belen D, Bavbek M. Halo traction in basilar invagination: technical case report. Surg Neurol 2006; 66: 311-314.

17) Klimo PJ, Rao G, Brockmeyer D. Congenital anomalies of the cervical spine. Neurosurg Clin N Am 2007; 18: 463-478.

18) Wang S, Wang C, Passias PG, Li G, Yan M, Zhou H. Interobserver and intraobserver reliability of the cervicomedullary angle in a normal adult population. Eur Spine J 2009: 18: 1349-1354.

19) Reiter MF, Boden SD. Inflammatory disorders of the cervical spine. Spine (Phila Pa 1976) 1998; 23: 2755-2766.

20) Reijnierse M, Breedveld FC, Kroon HM, Hansen B, Pope TL, Bloem JL. Are magnetic resonance flexion views useful in evaluating the cervical spine of patients with rheumatoid arthritis? Skeletal Radiol 2000; 29: 85-89.

21) Ogihara N, Takahashi J, Hirabayashi H, Mukaiyama K, Kato H. Surgical treatment of Klippel-Feil syndrome with basilar invagination. Eur Spine J 2013; 22 Suppl 3: S380-387.

22) Yin Q. Ai F, Zhang K, Chang Y, Xia H, Wu Z, Quan R, Mai X, Liu J. Irreducible anterior atlantoaxial dislocation: one-stage treatment with a transoral atlantoaxial reduction plate fixation and fusion. Report of 5 cases and review of the literature. Spine (Phila Pa 1976) 2005; 30: E375-381.

23) Wu Z, Xu J, Wang Z, Xia H, Zhang Q, Ma X, Zhang K. Transoral approach for revision surgery of os odontoideum with atlantoaxial dislocation. Orthopedics 2014; 37: e851-855.

24) Yin QS, Xia H, Wu ZH, Ma XY, Ai FZ, Zhang K, Wang JH, Zhang T, Bai ZH, Wang ZY. Surgical site infections following the transoral approach: a review of 172 consecutive cases. J Spinal Disord Tech 2013; Oct 30. [Epub ahead of print].

25) Ai FZ, Yin QS, Xu DC, Xia H, Wu ZH, Mai XH. Transoral atlantoaxial reduction plate internal fixation with transoral transpedicular or articular mass screw of c2 for the treatment of irreducible atlantoaxial dislocation: two case reports. Spine (Phila 1976) 2011; 36: E556-562.

26) Yin QS, Xia H, Wu ZH, Ai FZ, Ma XY, Zhang K, Wang JH, Mai XH, Wan L, Chen XQ. Diagnosis and treatment for complicated atlantoaxial dislocation. Zhonghua Wai Ke Za Zhi 2010; 48: 1301-1304.