A retrospective study of congenital osseous anomalies at the craniocervical junction treated by occipitocervical plate-rod systems

Xianjun Ding · Kuniyoshi Abumi · Manabu Ito ·
Hideki Sudo · Masahiko Takahata ·
Ken Nagahama · Akira Iwata

Received: 22 August 2011 / Revised: 22 February 2012 / Accepted: 14 April 2012 / Published online: 1 May 2012
© Springer-Verlag 2012

Abstract

Purpose To evaluate the effectiveness of posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and plate-rod systems for patients with congenital osseous anomalies at the craniocervical junction.

Methods Twenty patients with congenital osseous lesions who underwent posterior occipitocervical fusion using the anchors of cervical pedicle screws and plate-rod systems for reduction and fixation from 1996 to 2009 were reviewed. The lesions included os odontoideum, occipitalization of the atlas, congenital C2–3 fusion, congenital atlantoaxial subluxation, congenital basilar invagination and combined anomalies. The clinical assessment and the measurements of the images were performed preoperatively, postoperatively and at most recent follow-up.

Results The combined deformity of flexion of the occipitoatlantoaxial complex and invagination of the odontoid process associated with congenital osseous lesions at the craniocervical junction was corrected by application of combined forces of extension and distraction between the occiput and the cervical pedicle screws. Preoperative

myelopathy improved in 94.7 % patients. The mean Ranawat value, Redlund-Johnnell value, atlantodental distance, occiput (O)–C2 angle, and C2–C7 lordosis angle improved postoperatively and was sustained at most recent follow-up. The mean cervicomedullary angle improved from 129.3° preoperatively to 153.3° postoperatively. The mean range of motion at the lower adjacent motion segment remained unchanged at most recent follow-up. The fusion rate was 95 %.

Conclusions The results of the present study indicate that posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and plate-rod systems is an effective technique for treatment of deformities and/or instability caused by congenital osseous anomalies at the craniocervical junction.

Keywords Craniocervical junction · Congenital osseous anomaly · Atlantoaxial instability · Occipitocervical fusion · Cervical pedicle screw fixation

Introduction

Malalignment and/or instability at the craniocervical junction may cause compression of the spinal cord or medulla oblongata, triggering myelopathy, intractable neck pain or progressive disability [1–3]. For this condition, posterior atlantoaxial or occipitocervical reconstructive surgery is one potential option.

Since the first description of occipitocervical fusion by Pilcher in 1910, various surgical procedures for posterior occipitocervical fusion have evolved with the improved maintenance of alignment, increased fusion rate, and decreased incidence of complications [2, 4–6]. Recently, posterior occipitocervical fixation using screw rod/plate

X. Ding · M. Takahata · K. Nagahama · A. Iwata
Department of Orthopedic Surgery,
Graduate School of Medicine, Hokkaido University,
Sapporo, Japan

K. Abumi (✉)
Department of Spinal Reconstruction,
Graduate School of Medicine, Hokkaido University,
Kita-15 Nishi-7, Kita-ku, Sapporo 060-8638, Japan
e-mail: abumi@med.hokudai.ac.jp

M. Ito · H. Sudo
Department of Advanced Medicine for Spine and Spinal Cord
Disorders, Graduate School of Medicine, Hokkaido University,
Sapporo, Japan

systems has gained popularity. Grob et al. [2, 3] recommended posterior occipitocervical reconstruction using C1–C2 transarticular screw fixation. However, it may be unsatisfactory for the reduction of malalignment at the occipitoatlantoaxial region [2]. Direct anterior decompression by odontoidectomy is required for this procedure when the compression of the spinal cord or medulla oblongata by the odontoid process cannot be sufficiently reduced by the posterior approach. Abumi et al. [6] started posterior occipitocervical reconstruction using the anchors of cervical pedicle screws in 1993. By using these systems, the flexion deformity of the occipitoatlantoaxial complex and the upward migration of the odontoid process were corrected through the application of the combined forces of extension and distraction between the occiput and the cervical pedicle screws. Thus, sufficient indirect decompression of the spinal cord or medulla oblongata was achieved by reduction through a posterior approach. Direct anterior decompression was not required for the twenty-six consecutive patients with various lesions at the craniocervical junction who underwent posterior occipitocervical reconstruction in this series [6]. The goal of the present study is to evaluate the results of posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and plate-rod systems for patients with congenital osseous anomalies at the craniocervical junction.

Materials and methods

Demographics

The clinical records and photographs of all patients with congenital osseous anomalies who underwent posterior occipitocervical fusion using the anchors of cervical pedicle screws and rod-plate systems for reduction and fixation from 1996 to 2009 were reviewed. Of these patients, eleven were males and nine were females. Their mean age at the time of surgery was 42.5 years (range 13–75 years). The follow-up term was 56.8 months on average (range 12–151 months). The congenital osseous disorders included os odontoideum in twelve patients, os odontoideum associated with occipitalization of the atlas in one, occipitalization of the atlas associated with congenital C2–3 fusion in three, occipitalization of the atlas in one, congenital atlantoaxial subluxation in two and congenital basilar invagination in one. Among the patients with os odontoideum, three had Down's syndrome and one had spondyloepiphyseal dysplasia congenita (Table 1). The most common presenting symptoms were myelopathy and neck pain. Subjective myelopathy symptoms were reported by 95.0 % patients before surgery. Surgical

indications were atlantoaxial or occipitoaxial instability and/or subluxation, neurological deficit, or intractable pain.

Surgical procedures

Patients were positioned prone in a Mayfield tongs. A midline incision was made with a subperiosteal exposure of the occiput and cervical spine. Posterior occipitocervical fixation was achieved using occipitocervical plate-rod systems. Cotrel-Dubousset occipitocervical rods (CD rod) were used in one patient and occipitocervical plate-rod systems (Robert Reid Inc., Tokyo, Japan) was used in the remaining nineteen patients. Cervical pedicle screws (CPS, AcroMed Inc., Cleveland, OH) were used for cervical fixation in all patients. The combined deformity of flexion of the occipitoatlantoaxial complex and invagination of the odontoid process was corrected through the application of the combined forces of extension and distraction between the occiput and the inserted cervical pedicle screws. Lateral fluoroscopy image was used to control the pedicle screw insertion and to confirm the correction of deformity. Corticocancellous graft was harvested from the iliac crest in all patients. The most caudal instrumented vertebra was C2 in thirteen patients, C3 in three, C4 in two and C5 in two. Additional posterior decompression by C1 or C2 posterior arch resection was performed in four patients; C1 posterior arch resection and enlargement of the foramen magnum was performed in three; and combined laminectomy of C1–2 and T1–2 with laminoplasty from C3 to C7 was performed in one (Table 1). Postoperative immobilization varied mainly with the number of the spinal segments fixed and the associated diseases. Fourteen patients wore a soft cervical collar postoperatively for 2–3 months; Three wore a Philadelphia collar for 2–3 months; and three were immobilized by a halo vest for 3 months.

Clinical and radiographic evaluation

The Ranawat classification for neurological deficits and the Ranawat classification for pain [7] were used to evaluate myelopathy and pain, respectively, before surgery, after surgery, and at most recent follow-up (Table 2).

The radiographic parameters included Ranawat value, Redlund-Johnell value, atlantodental distance (ADD), occiput (O)–C2 angle, cervicomedullary angle (CMA), C2–C7 lordosis angle, and range of motion (ROM) at the lower adjacent motion segment. They were calculated before and after surgery, and at most recent follow-up, using plain roentgenograms, computed tomography (CT) with reconstruction and magnetic resonance imaging (MRI). The Ranawat and Redlund-Johnell values were measured to evaluate the occipitoaxial alignments in the sagittal plane. The ADD and O–C2 angle were measured

Table 1 Clinical characteristics and details of surgery

Case no.	Sex	Age (years)	Main osseous lesion	Associated osseous lesion	Associated systemic disorder	Lowermost fixed vertebra	Applied additional surgery	Follow-up term (m)
1	F	53	Os odontoideum			C2	C1 posterior arch resection	151
2	M	13	Os odontoideum			C2		137
3	M	15	Os odontoideum		Down's syndrome	C2	C1 posterior arch resection; enlargement of the foramen magnum	103
4	F	66	Os odontoideum			C2	C1 posterior arch resection	12
5	M	26	Os odontoideum			C2		72
6	M	45	Os odontoideum	Occipitalization of the atlas		C2		26
7	M	45	Os odontoideum			C2		54
8	F	16	Os odontoideum		Down's syndrome	C2		45
9	F	60	Os odontoideum			C2		21
10	F	75	Os odontoideum			C2		42
11	M	13	Os odontoideum		Spondyloepiphyseal dysplasia congenita	C2		14
12	F	69	Os odontoideum			C3		19
13	M	27	Os odontoideum		Down's syndrome	C4		110
14	M	43	Occipitalization of the atlas	Congenital C2–3 fusion		C5		46
15	M	71	Occipitalization of the atlas	Congenital C2–3 fusion		C4	C1 posterior arch resection; enlargement of the foramen magnum	37
16	M	67	Occipitalization of the atlas	Congenital C2–3 fusion		C2	C2 posterior arch resection	28
17	F	45	Occipitalization of the atlas			C3	C1 posterior arch resection; enlargement of the foramen magnum	64
18	F	26	Congenital atlantoaxial subluxation		Down's syndrome	C3	C1–2, T1–2 laminectomy; C3–7 laminoplasty	76
19	F	59	Congenital atlantoaxial subluxation		Achondroplasia	C2	C1 posterior arch resection	60
20	M	15	Congenital basilar invagination		Aarskog syndrome	C5		18

to evaluate the atlantoaxial instability. These parameters were measured on three occasions, and the average value was used for comparisons.

The Ranawat value indicates the distance between the line connecting the center of the anterior and posterior arch of the atlas and the center of the C2 pedicles (Fig. 1a, b).

The Redlund-Johnell value indicates the vertical distance between the midpoint of the C2 endplate and McGregor's line drawn from the posterosuperior aspect of

the hard palate to the most caudal point on the midline occipital curve (Fig. 1e, f).

The ADD indicates the distance between the dens and the anterior ring of C1 (Fig. 1h).

The O–C2 angle indicates the angle between McGregor's line [8] and the line of the endplate of the axis (Fig. 1e, g).

The CMA [9] indicates the angle between the lines on the ventral side of the cervical spinal cord and the medulla oblongata calculated on MRI image (Fig. 1k, l). In the

Table 2 Criteria for pain and neurological evaluation [7]

Pain evaluation	
Grade 0	None
Grade I	Mild; intermittent, requiring only aspirin for analgesia
Grade II	Moderate; a cervical collar was needed
Grade III	Severe; pain could not be relieved by either aspirin or collar
Neurological evaluation	
Class I	No neurological deficit
Class II	Subjective weakness with hyperreflexia and dysesthesia
Class IIIA	Objective findings of paresis and pyramidal tract signs, but ambulation possible
Class IIIB	Quadriplegia with resultant inability to walk or feed oneself

present study, it was used to evaluate the correction of the malalignment of the spinal cord and the medulla oblongata. As MRI was conducted in 17 patients preoperatively and 12 postoperatively, average CMA in 12 patients who underwent MRI examination preoperatively and postoperatively was used for comparison.

The C2–C7 lordosis angle indicates the angle between the lines drawn at the posterior margin of C2 and C7 vertebral bodies (Fig. 1i, j).

The ROM at the lower adjacent motion segment [10] was defined as follows: to evaluate the postoperative instability at the lower adjacent motion segment, the angle formed by the two lines between the inferior margin of the upper vertebral body and the superior margin of the lower

Fig. 1 Radiologic evaluation using plain roentgenograms, CT or MRI. Ranawat value: the distance between the line connecting the center of the anterior and posterior arch of the atlas and the center of the C2 pedicles (a, b). Range of motion (ROM) at the lower adjacent motion segment: the difference of the angles between flexion and extension formed by the two lines between the inferior margin of the upper vertebral body and the superior margin of the lower vertebral body at the motion segment below the most caudal instrumented vertebra (c, d). Redlund-Johnell value: the distance between

McGregor line and the midpoint of the C2 endplate (e, f). O–C2 angle: the angle between McGregor line and the line of the endplate of the axis (e, g). Atlantodental distance (ADD): the distance between the dens and the anterior ring of C1 (h). C2–C7 lordosis angle: the angle between the lines drawn at the posterior margin of C2 and C7 vertebral bodies (i, j). Cervicomedullary angle (CMA): the angle between the lines on the ventral side of the medulla oblongata and the cervical spinal cord (k, l)

vertebral body at the segment below the most caudal instrumented vertebra was measured using plain radiographs (Fig. 1c, d). The ROM at the lower adjacent motion segment was defined as the difference of the angle between flexion and extension.

The presence of a homogeneous fusion mass on the lateral cervical radiographs or sagittal CT reconstructions was considered a direct evidence of fusion. Patients without a homogeneous fusion mass who nonetheless maintained the integrity of their hardware and had less than 2° of segmental motion on flexion–extension radiographs without a clear zone around the pedicle screws and the occipital screws were considered to have an indirect evidence of fusion.

Results

Clinical results

On average, operative time was 134 min (range 90–220 min), and intraoperative blood loss was 140 ml (range 50–250 ml).

According to Ranawat classification for pain, the pain was assessed as grade 0 in one patient, grade I in four, grade II in twelve and grade III in three preoperatively. Nine patients had grade I pain, two had grade II pain and the remaining nine experienced no pain postoperatively. At most recent follow-up, three patients had grade I pain, and the remaining eighteen had no pain (Table 3).

Among the nineteen patients with myelopathy preoperatively, Ranawat classification of neurologic deficits was: class II in nine patients, class IIIA in six and class IIIB in four. Seventeen improved by 1 class and one gained improvement of 2 classes postoperatively. Another patient remained at the same class, but his spinal cord symptoms improved to some extent. No patient suffered neurological deterioration as a result of the procedure. At most recent follow-up, fourteen patients were graded as I, four as II and two as IIIA (Table 3).

Radiographic evaluation

The mean preoperative Ranawat value of 11.3 mm (range 1–22 mm) improved to 15.4 mm (range 5–25 mm) postoperatively and 15.5 mm (range 6–25 mm) at most recent follow-up. The mean preoperative Redlund-Johnell value of 24.6 mm (range 5–36 mm) improved to 30.8 mm (range 11–48 mm) postoperatively and 30.1 mm (range 12–47 mm) at most recent follow-up.

The mean preoperative ADD of 5.2 mm (range 0–10 mm) was reduced to 2.1 mm (range 0–8 mm) postoperatively and 1.8 mm (range 0–8 mm) at most recent follow-up. The mean preoperative O–C2 angle of 5.9°

Table 3 Clinical evaluation [7]

Case no.	Pain grade			Neurological class		
	Preop	Postop	F-up	Preop	Postop	F-up
1	I	0	0	II	I	I
2	II	I	0	II	II	I
3	II	0	0	IIIA	II	I
4	I	0	0	II	I	I
5	II	0	0	IIIA	I	I
6	II	I	0	II	I	I
7	II	I	0	IIIA	II	II
8	0	0	0	I	I	I
9	II	0	0	II	I	I
10	II	I	0	II	I	I
11	I	0	0	II	I	I
12	III	II	I	IIIB	IIIA	IIIA
13	II	I	0	IIIA	II	I
14	III	I	I	IIIA	II	II
15	III	II	0	IIIB	IIIA	II
16	II	0	0	IIIA	II	I
17	I	0	0	II	I	I
18	II	I	0	IIIB	IIIA	IIIA
19	II	I	0	II	I	I
20	II	I	I	IIIB	IIIA	II

Preop preoperative, *Postop* postoperative, *F-up* follow-up

(range -37° to 30°) improved to 13.5° (range -37° to 36°) postoperatively and 14.2° (range -35° to 36°) at most recent follow-up.

The mean preoperative CMA of 129.3° (range 115° – 146°) in 12 patients improved to 153.3° (range 140° – 160°) postoperatively. The mean preoperative C2–C7 angle of 30.1° (range 5° – 63°) improved to 19.6° (range 6° – 47°) postoperatively and 19.9° (range 0° – 48°) at most recent follow-up. The mean ROM at the lower adjacent motion segment was 5.2° (range 0° – 11°) preoperatively and was sustained at 5.3° (range 0° – 11°) at most recent follow-up (Table 4).

Fusion rate

The fusion rate was 95 %. These patients showed no radiographic evidence of non-union: implant failure, screw loosening, or translucent zone formation around the pedicle screws and occipital screws. In this series, one patient had dislodged occipital screws during the operation. The screws were reinserted, and a Halo-vest was used for 2 months postoperatively. Another patient had delayed union and correction loss of craniocervical deformity, and became myelopathic again at 2-year follow-up. This patient underwent salvage surgery by fusion mass

Table 4 Radiological evaluation

Case no.	Ranawat value		Redlund-Johnell value		ADD		O-C2 angle		CMA		C2–C7 angle		ROM at lower adjacent segment	
	Preop	Postop	F-up	Preop	Postop	F-up	Preop	Postop	F-up	Preop	Postop	F-up	Preop	F-up
1	10	16	15	24	33	29	4	4	4	13	14	13	124	153
2	9	12	16	29	34	34	6	5	4	4	24	10	35	10
3	12	14	15	23	29	32	6	4	3	23	29	20	20	20
4	13	18	18	36	48	47	8	0	0	0	−31	20	115	158
5	5	10	12	17	23	24	10	0	0	20	9	7	34	21
6	11	15	16	32	37	37	5	3	2	30	33	34	146	151
7	10	19	17	16	20	19	6	3	2	4	15	18	145	12
8	14	17	16	25	32	29	9	0	0	2	25	27	139	155
9	13	14	13	30	32	34	5	0	0	3	25	25	120	140
10	16	22	22	31	35	35	0	0	0	23	13	13	140	153
11	12	15	14	18	22	21	5	1	1	−15	−6	−5	133	38
12	12	19	18	30	38	35	4	0	0	20	12	12	127	160
13	9	22	17	35	46	39	8	5	5	5	14	29	126	159
14	1	5	6	5	11	12	3	0	0	18	12	10	130	154
15	20	20	19	35	36	36	0	0	0	30	36	36	131	153
16	20	25	25	31	36	35	6	1	1	−13	10	12	130	157
17	6	8	9	18	27	25	4	2	2	−14	7	8	123	146
18	7	8	10	15	20	19	8	3	2	16	10	11	103	20
19	22	18	20	34	37	39	3	2	2	16	18	20	165	5
20	4	11	11	7	20	20	4	8	8	−37	−37	−35	104	35
Ave.	11.3	15.4	15.5	24.6	30.8	30.1	5.2	2.1	1.8	5.9	13.5	14.2	129.3 ^a	153.3 ^a

ADD atlantodental distance, O-C2 angle occiput-C2 angle, CMA cervicomedullary angle, ROM range of motion, Ave. average

^a Average values in 12 patients who underwent MRI examination preoperatively and postoperatively

Fig. 2 The patient was a 66-year-old female with AAS and VS in os odontoideum. Radiographic examination showed rigid atlantoaxial subluxation, flexion deformity at the occipitocervical junction and cranially migrated odontoid process (**a–c, e**). A round ossicle was observed above the anterior arch of the atlas (**e**). MRI showed impingement of the spinal cord or medulla oblongata by the tip of the

odontoid process (**e**). Posterior occipitocervical reconstruction using cervical pedicle screws and plate-rod systems was performed (**d**). The malalignment at the occipitocervical junction was satisfactorily improved (**d, f**). MRIs demonstrated the improvement of CMA from 115° preoperatively to 158° postoperatively and the decrease of the anterior compression of neural tissue (**e, f**)

osteotomy and extension of fixation caudal to C4. Sufficient correction of deformity and bone union was achieved at most recent follow-up.

Complications

Complications occurred in five patients. The reoperation rate was 10 %. Early complications occurred in three patients, including intraoperative dislodging of occipital screws in one and postoperative transient cerebrospinal fluid (CSF) leakage in two. The CSF leakage was managed successfully through lumbar subarachnoid drainage for 14 days without sequelae. Late complications included loss of correction by delayed union in one patient and cutaneous irritation of the plate in the other. The cutaneous irritation of the plate was managed through the removal of the

implant after bone union. Satisfactory results were achieved after the second operation.

Case presentation

Case 4

A 66-year-old female with os odontoideum had mild myelopathy by AAS and VS. Rigid atlantoaxial subluxation was observed on the flexion–extension radiographs (Fig. 2a–c). A round ossicle was observed above the anterior arch of the atlas (Fig. 2e). MRI showed impingement of the medulla oblongata or spinal cord by the tip of the odontoid process (Fig. 2e). Posterior occipitoaxial reconstruction using cervical pedicle screws and plate-rod systems was performed (Fig. 2d). Screws were inserted

into the bilateral C2 pedicles and the combined forces of extension and distraction were applied to reduce the flexion deformity and the cranial migration of the odontoid process. The malalignment at the occipitocervical junction was satisfactorily improved. MRIs demonstrated the improvement of CMA from 115° preoperatively to 158° postoperatively, and the decrease of the anterior compression of the neural tissue (Fig. 2e–f). Her neurologic deficits showed improvement from class II preoperatively to class I postoperatively based on the Ranawat classification.

Discussion

Various congenital osseous anomalies at the craniocervical junction have been reported [11–21]. In the current study, the congenital lesions include os odontoideum, occipitalization of the atlas, congenital C2–3 fusion, congenital atlantoaxial subluxation, congenital basilar invagination and combined anomalies. Os odontoideum is defined as an independent ossicle of variable size with smooth circumferential cortical margins separated from the odontoid process. It has been classified into two anatomical types according to the location: orthotopic and dystopic [12, 13]. Orthotopic os odontoideum consists of an ossicle located in the normal position of the odontoid process, which usually moves with the anterior arch of the atlas, while dystopic os odontoideum consists of an ossicle near the basion, which can be fused with the clivus. In the present study, orthotopic os odontoideum was found in five patients, while dystopic os odontoideum was found in eight. In the absence of a united odontoid process, the atlantoaxial movements appear to be supported only by ligaments, resulting in atlantoaxial instability. The presence of dystopic os odontoideum can also affect the stability of the occipitoatlantal articulation. Occipitalization of the atlas is caused by the failure of segmentation between the basal occipital sclerotome and the first spinal sclerotome. It is frequently associated with atlantoaxial subluxation and basilar invagination and often occurs in conjunction with other anomalies [14, 15]. In the present study, occipitalization of the atlas was associated with congenital C2–3 fusion in three patients and os odontoideum in one. Occipitalization of the atlas, in association with congenital C2–3 fusion, exerts more strain on the atlantoaxial articulation during flexion and extension [15, 16]. Repeated excessive strain at this joint more likely predisposes a patient to atlantoaxial instability. In a study of 30 patients with occipitalization of the atlas, Gholve et al. [15] reported that eight of the seventeen patients with atlantoaxial instability had an associated C2–3 fusion.

In the present study, these congenital osseous anomalies at the craniocervical junction were associated with such

deformities as: anterior translation of the atlas on the axis, vertical subluxation of the odontoid process or flexion deformity caused by anterior subluxation of the occipitoatlantal complex on the axis, resulting in the compression of the spinal cord or medulla oblongata by the odontoid process. Since neural tissues are compressed anteriorly, direct anterior decompression, using the transoral or mandible splitting approach, has been employed for this condition. However, the above mentioned procedures have several disadvantages: (1) a high risk of infection and the need for complex perioperative management; (2) the difficulty of obtaining solid fusion; and (3) the required additional posterior fixation [17]. Alternatively, in the present study we described posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and occipitocervical rod systems. For the effective correction of malalignment in the occipitoatlantoaxial region, a rigid fixation anchor for the occiput and the cervical spine is typically required. By evaluating the biomechanical stability provided by five types of occipitoatlantoaxial reconstruction techniques in a human cadaveric spine model, Oda et al. [22] showed that the combination of occipital screws and C2 pedicle screws provided the highest stability under axial rotation, flexion/extension, lateral bending, and anterior–posterior translation loading modes. In the current study, indirect anterior decompression was sufficiently achieved through the correction of the flexion deformity at the occipitocervical junction and the reduction of upward migration of the odontoid process by applying the combined forces of extension and distraction in most patients, including in cases with rigid atlantoaxial subluxation (Fig. 2). The mean preoperative CMA of 129.3° , which was calculated in 12 patients, improved to a mean postoperative value of 153.3° . Postoperative clinical and radiographic improvements were maintained until mean follow-up of 56.8 months. The fusion rate was 95 %, which is consistent with the findings of previous reports [23]. At most recent follow-up, no evidence of instability was observed at the lower adjacent motion segment. The results of the present study demonstrate that posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and plate-rod systems is effective in treating deformities and/or instabilities caused by congenital osseous anomalies at the craniocervical junction.

Os odontoideum was the largest subgroup in the present study. Currently, conservative management is normally used in the treatment of an asymptomatic or minimally symptomatic, or radiologically stable os odontoideum; meanwhile surgical intervention is preserved for patients with persistent neck pain, occipitocervical instability, and neurological symptoms [12, 18, 19]. However, delayed neurological deterioration may occur in patients with asymptomatic or radiologically stable os odontoideum at

initial presentation. Clements et al. [20] reported a patient who had a documented os odontoideum without instability and developed symptomatic C1–C2 instability, requiring surgical stabilization and fusion after a 5-year follow-up. Kim et al. [12] found that minor trauma can provoke neurological deterioration in patients with os odontoideum. Thus, a lack of C1–C2 instability at initial presentation does not guarantee that instability will not develop over time. Close clinical and radiographic follow-up is recommended for such patients. Some authors described os odontoideum as an unstable craniocervical junction, and advocate surgical intervention regardless of the clinical presentation. Therefore, further investigations of the role of surgery in patients with asymptomatic or radiologically stable os odontoideum are necessary.

First described by Goel and Laheri [24], posterior atlantoaxial fixation using independent C1 and C2 screws, is a generally adopted and accepted surgical method for os odontoideum. However, this method still has some limitations in the treatment of os odontoideum. First, the method cannot be used to treat the dystopic os odontoid, or os odontoideum associated with other abnormalities affecting the occipitoatlantal articulation because of the possible associated occipitoatlantal instability. Second, the conjunction of occipitalization of the atlas increases the risks for the insertion of lateral mass screws into the atlas due to high incidence of anomalous vertebral artery at the craniocervical junction [21]. Third, the method is contraindicated in os odontoideum associated with a massive defect of the lateral masses of the atlas [25]. Moreover, the method is not applicable for the treatment of os odontoideum with irreducible anterior compression, which usually needs to be completed by anterior approach for direct decompression [17]. Nevertheless, posterior occipitocervical reconstruction using the anchors of cervical pedicle screws and plate-rod systems is suitable for these conditions.

In conclusion, the combined deformity of flexion of the occipitoatlantoaxial complex and invagination of the odontoid process associated with congenital osseous lesions at the craniocervical junction was corrected through posterior occipitocervical reconstruction, using the anchors of cervical pedicle screws and plate-rod systems. Postoperative clinical and radiographical improvements were maintained until the most recent follow-up. No evidence of instability was observed at the lower adjacent motion segment. The fusion rate was 95 %. Therefore, the procedure described here is effective in treating deformities and/or instabilities caused by congenital osseous lesions at the craniocervical junction.

Conflict of interest None.

References

1. Nockels RP, Shaffrey CI, Kanter AS, Azeem S, York JE (2007) Occipitocervical fusion with rigid internal fixation: long-term follow-up data in 69 patients. *J Neurosurg Spine* 7:117–123
2. Grob D, Dvorak J, Panjabi MM, Antinnes JA (1994) The role of plate and screw fixation in occipitocervical fusion in rheumatoid arthritis. *Spine* 19:2545–2551
3. Grob D, Schutz U, Plotz G (1999) Occipitocervical fusion in patients with rheumatoid arthritis. *Clin Orthop Relat Res* 366:46–53
4. Sandhu FA, Pait TG, Benzel E, Henderson FC (2003) Occipitocervical fusion for rheumatoid arthritis using the inside–outside stabilization technique. *Spine* 28:414–419
5. Vale FL, Oliver M, Cahill DW (1999) Rigid occipitocervical fusion. *J Neurosurg (Spine 2)* 91:144–150
6. Abumi K, Takada T, Shono Y, Kaneda K, Fujiya M (1999) Posterior occipitocervical reconstruction using cervical pedicle screws and plate-rod systems. *Spine* 24:1425–1434
7. Ranawat CS, O’Leary P, Pellicci P, Tsairis P, Marchisello P, Dort L (1979) Cervical spine fusion in rheumatoid arthritis. *J Bone Joint Surg (Am)* 61:1003–1010
8. Logroscino CA, Genitempo M, Casula S (2009) Relevance of the cranioaxial angle in the occipitocervical stabilization using an original construct: a retrospective study on 50 patients. *Eur Spine J* 18(Suppl 1):7–12
9. Reijnierse M, Bloem JL, Dijkman BAC, Kroon HM, Holscher HC, Hansen B, Breedveld FC (1996) The cervical spine in rheumatoid arthritis: relationship between neurologic signs and morphology on MR imaging and radiographs. *Skelet Radiol* 25:113–118
10. Sasai K, Umeda M, Saito T, Ohnari H, Wakabayashi E, Iida H (2006) Microsurgical posterior foraminotomy with laminoplasty for cervical spondylotic radiculomyelopathy including cervical spondylotic amyotrophy. *J Neurosurg Spine* 5:126–132
11. Hosalkar HS, Sankar WN, Wills BP, Goebel J, Dormans JP, Drummond DS (2008) Congenital osseous anomalies of the upper cervical spine. *J Bone Joint Surg Am* 90:337–348
12. Kim IS, Hong JT, Jang WY, Yang SH, Sung JH, Son BC, Lee SW (2011) Surgical treatment of os odontoideum. *J Clin Neurosci* 18:481–484
13. Fielding JW, Hensinger RN, Hawkins RJ (1980) Os odontoideum. *J Bone Joint Surg Am* 62:376–383
14. Goel A, Kulkarni AG (2004) Mobile and reducible atlantoaxial dislocation in presence of occipitalized atlas: report on treatment of eight cases by direct lateral mass plate and screw fixation. *Spine* 29:E520–E523
15. Gholve PA, Hosalkar HS, Ricchetti ET, Pollock AN, Dormans JP, Drummond DS (2007) Occipitalization of the atlas in children: morphologic classification, associations, and clinical relevance. *J Bone Joint Surg Am* 89:571–578
16. Wang S, Wang C, Passias PG, Yan M, Zhou H (2010) Pedicle versus laminar screws: what provides more suitable C2 fixation in congenital C2–3 fusion patients? *Eur Spine J* 19:1306–1311
17. Wang C, Yan M, Zhou HT, Wang SL, Dang GT (2006) Open reduction of irreducible atlantoaxial dislocation by transoral anterior atlantoaxial release and posterior internal fixation. *Spine* 31:E306–E313
18. Klimo P Jr, Kan P, Rao G, Apfelbaum R, Brockmeyer D (2008) Os odontoideum: presentation, diagnosis, and treatment in a series of 78 patients. *J Neurosurg Spine* 9:332–342
19. Spierings EL, Braakman R (1982) The management of os odontoideum. Analysis of 37 cases. *J Bone Joint Surg B* 64:422–428

20. Clements WD, Mezue W, Mathew B (1995) Os odontoideum congenital or acquired? That's not the question. *Injury* 26:640–642
21. Wang S, Wang C, Liu Y, Yan M, Zhou H (2009) Anomalous vertebral artery in craniovertebral junction with occipitalization of the atlas. *Spine* 34:2838–2842
22. Oda I, Abumi K, Sell LC, Haggerty CJ, Cunningham BW, McAfee PC (1999) Biomechanical evaluation of five different occipito-atlanto-axial fixation techniques. *Spine* 24:2377–2382
23. Deutsch H, Haid RW Jr, Rodts GE Jr, Mummaneni PV (2005) Occipitocervical fixation: long-term results. *Spine* 30:530–535
24. Goel A, Laheri V (1994) Plate and screw fixation for atlanto-axial subluxation. *Acta Neurochir (Wien)* 129:47–53
25. Stulik J, Vyskocil T, Sebesta P, Kryl J (2007) Atlantoaxial fixation using the polyaxial screw-rod system. *Eur Spine J* 16:479–484