CLINICAL AND RADIOLOGICAL FEATURES OF ATLANTOAXIAL DISLOCATIONS ASSOCIATED WITH CONGENITAL MALFORMATIONS OF THE CRANIOVERTEBRAL JUNCTION

O.M. Pavlova, S.O. Ryabykh, A.V. Burtsev, A.V. Gubin
Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russia

Objective. To analyze clinical and radiological features of congenital atlantoaxial dislocations (AAD) in congenital craniovertebral junction malformations.

Material and Methods. The data of 26 patients with AAD associated with congenital pathology of the craniovertebral junction, who applied to the Ilizarov Center in 2012—2017, were analyzed.

Results. Patients were divided into three groups: with non-syndromic AAD — 6 (23.1 %) patients, with AAD associated with Klippel — Feil syndrome — 11 (42.3 %) and with syndromic AAD — 9 (34.6 %). Odontoid anomalies were observed in 15 (57.7 %) patients, the magnitude of dislocation was determined from the C1 facet displacement relative to that of C2 in different planes. Patients with non-syndromic AAD had local pain syndrome (VAS score 4.20 ± 2.64) accompanied by torticollis and restriction of head movements, and myelopathy. In patients with AAD associated with Klippel — Feil syndrome, the local symptoms prevailed: restriction of neck movements, torticollis, neck pain (VAS score 2.40 ± 2.01), and myelopathy. Myelopathy and unpronounced pain syndrome (VAS score 2.30 ± 1.94) were leading symptoms in patients with syndromic AAD.

Conclusion. Patients with syndromic AAD more often have myelopathy, whereas AAD with the Klippel-Feil syndrome and non-syndromic AAD are often manifested by local symptoms.

Key Words: congenital atlantoaxial dislocations, unstable C1—C2 anomalies, craniovertebral junction.

Please cite this paper as: *Pavlova OM, Ryabykh SO, Burtsev AV, Gubin AV. Clinical and radiological features of atlantoaxial dislocations associated with congenital malformations of the craniovertebral junction. Hir. Pozvonoc. 2018;15(1):32—41. In Russian.*
DOI: http://dx.doi.org/10.14531/ss2018.1.32-41.

Atlantoaxial dislocations (AADs) with underlying congenital malformations of the craniovertebral junction have been most extensively studied in India and China [17, 27, 36–38]. It is known that mortality rate associated with traumatic AAD is 60—80% due to sharp compression of the spinal cord and brain stem [14, 35, 39]. There are no data on mortality in patients with AAD with underlying congenital malformations of the craniovertebral junction, but the reported incidence of myelopathy is 40 to 100% [17, 25, 28, 29, 36]. Traumatic injuries and excessive flexion in the cervical region are the risk factors for neurological instability in patients with AAD. The main factors predisposing to the development of AAD in children include odontoid hypoplasia or aplasia, odontoid bone (os odontoideum), occipitalization of the atlas in combination with C2–C3 block, dysplasia of the articular processes of C1 and C2 vertebrae accompanied by symmetric or asymmetric enhancement of the angles of the lower facets of C1 and superior facets of C2, bone dysplasia, Down syndrome, connective tissue dysplasia accompanied by ligamentous laxity at the craniovertebral junction.

The study was aimed at determining clinical and radiological features of AAD associated with the congenital pathology of the craniovertebral transition.

Study design. Retrospective and prospective analysis of a series of clinical cases, level of evidence IV.

Material and Methods

A retrospective and prospective monocentric cohort of 26 patients with AAD with underlying congenital pathology of the craniovertebral junction, who applied to the Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics (Kurgan) in 2012–2017.

Inclusion criteria were as follows: AAD with underlying malformations of the craniovertebral junction, adequate X-ray archive.

The following criteria were assessed: sex, age at the time of admission, the nature of the onset of symptoms, the presence of a trigger, the duration of the disease, AAD type, including classification according to Fielding and Hawkins [9],

Samartzis et al. [33]), myelopathy (JOA score as modified by Benzel), local symptoms, including neck pain (VAS score), concomitant pathology, the data from introscopic methods. The following radiological parameters were assessed based on X-ray images, MRI, and CT: the type of craniovertebral junction malformation, the angle of the inferior facets of C1 in the sagittal and frontal planes according to Salunke et al. [31], displacement of the inferior facet of C1 in the sagittal and frontal planes on the median sagittal and frontal sections through the facet, sagittal diameter of the canal at the level of C1 and C2, sagittal diameter of C1 and the foramen magnum, vertical atlantoaxial index (VAAI according to the Kulkarni and Goel), anterior atlantodental interval (ADI), atlantooccipital dissociation (BAI and BDI indices), displacement of the os odontoideum bone (if any).

Literature sources were searched in the PubMed (NCBI), e-Library, and Google scholar databases. Search keywords: congenital atlantoaxial dislocation, congenital atlantoaxial displacement, congenital atlantoaxial subluxation, basilar invagination, congenital atlantoaxial instability, odontoid bone (os odontoideum), dens hypoplasia.

Results

There were 14 (54 %) male and 12 (46 %) female patients. The average age at the time of admission was 9.0 ± 5.4 years (2 to 24 years).

All patients had C1 vertebra dislocation in various planes and directions: anterior— 22 (84.6 %), posterior— 2 (7.7 %), central — 17 (65.4%), rotational — 19 (73.1 %). The dislocation was biplanar in 12 (46.2 %) patients, triplanar in 11 (42.3 %), and monoplanar in 3 (11.5 %).

Developmental anomalies of C2 odontoid process were found in 15 patients, odontoid hypoplasia in 4 (15.4 %), os odontoideum in 11 (42.3 %).

We classified the patients into three groups based on the degree of dislocation of the inferior facets of C1 with respect to C2 in the sagittal plane, depending on displacement magnitude: up to 3 mm – Grade I, 3–5 mm – Grade II, more than 5 mm – Grade III (Fig. 1). When an equal displacement of the facets in opposite directions was observed (anterior and posterior), this dislocation was considered as rotational. Grade I dislocation was detected in 2 (8.3 %) of 24 patients with C1 dislocation in the sagittal plane, Grade II in 12 (50.0 %), Grade III in 10 (41.7 %); in the latter two cases, posterior dislocation was observed.

Identification of the rotational AAD type according to Fielding and Hawkins [9] was not possible in 10 patients with odontoid anomalies, since ADI could not be measured. In these patients, the severity can be determined based on C1 rotation value. The following types of rotational dislocations (according to Fielding and Hawkins) were observed in 9 patients with AAD without odontoid malformations: type I in 5 patients, type II in 1 patient, type III in 2 patients, type IV in 1 patient.

The severity of the rotational dislocation was determined based the displacement of C1 facets in the axial plane, namely based on the difference in displacement of the right and left inferior facets of C1 in opposite directions with respect to the facets of C2: Grade I with the difference in facet displacement of 3 mm, Grade II – 3–5 mm, Grade III – more than 5 mm on both sides (Fig. 1). The following types of rotational dislocations were observed in 19 patients: Grade I in 6 (31.6%) patients, Grade II in 9 (47.4 %) patients, Grade III in 4 (21.0 %) patients. Traditionally, this classification matched the classification of Fielding and Hawkins in only one of nine patients. This can be attributed to the fact that the classification of Fielding and Hawkins takes into account dislocation in the axial and sagittal planes, while our staging is based only on dislocation in the axial plane. We evaluated dislocation in the sagittal plane based on the aforementioned individual criteria.

The subtypes of rotational AADs according to Samartzis et al. [33] were determined in all patients: subtype A accompanied by abnormal development of C1–C2 and sub-axial cervical spine was observed in 13 (68.4 %) patients, subtype B (developmental anomalies of C1–C2 without developmental anomalies of the subaxial cervical spine – 6 patients).

Identification of the grade of central dislocation according to Kulkarni and Goel [15] was not possible in 13 patients with odontoid malformation, 2 (11.8 %) patients had a mild Grade I dislocation (11.8 %), 2 patients – Grade II dislocation. Additionally, identification of the lower boundary of C2 could be inaccurate in some patients with the Klippel – Feil syndrome in the case of C2 and C3 block. In 12 (75 %) patients with developmental abnormalities of the odontoid process, central dislocation was identified based on the presence of lateral displacement of C1 facets.

Patients with a central dislocation were divided into three groups depending on the lateral displacement magnitude of the facets of C1: less than 3 mm — Grade I, 3—5 mm — Grade II, more than 5 mm — Grade III (Fig. 2). In the case of equal displacement of the facets of C1 in one direction (to the right or to the left), the dislocation was interpreted as a rotational one. According to this classification, 2 (11.8 %) patients had central dislocation — Grade I, 11 (64.7 %) — Grade II, 4 (23.5 %) — Grade III. These types matched those of Kulkarni and Goel in 4 patients without odontoid abnormalities.

The patients were divided into three subgroups depending on the etiology of the process:

1) 6 (23.1 %) patients with non-syndromic AAD associated with isolated craniovertebral defect;

2) 11 (42.3 %) patients with AAD associated with Klippel-Feil syndrome;

3) 9 (34.6 %) patients with syndromic AAD associated with various genetic and metabolic syndromes: Down syndrome (n = 1), mucopolysaccharidosis (n = 1), spondyloepiphysic dysplasia (n = 3), spondylometaphysial dysplasia (n = 1), and undifferentiated bone dysplasia

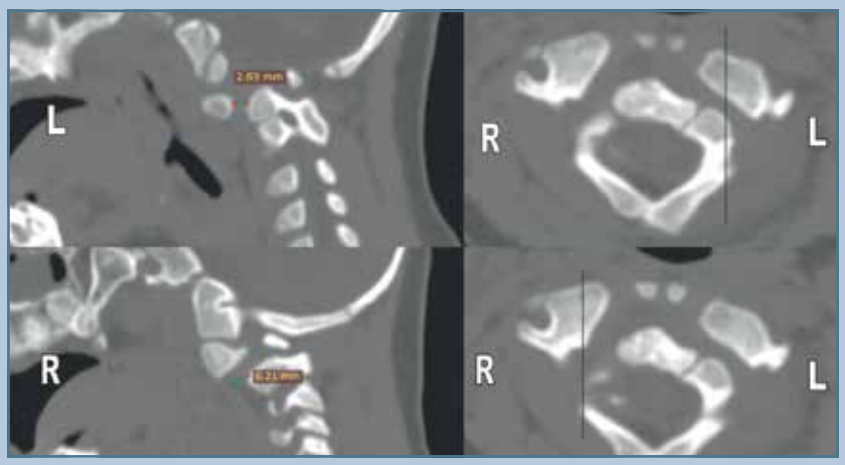

Fig. 1
Median sagittal sections through the right and left facets of C1 vertebra in a patient with triplanar dislocation: anterior displacement of the left facet by 2.9 mm, anterior displacement of the right facet by 6.2 mm; anterior Grade III displacement; Grade II displacement in the axial plane: the difference between displacement of the right and left facets is 3.3 mm

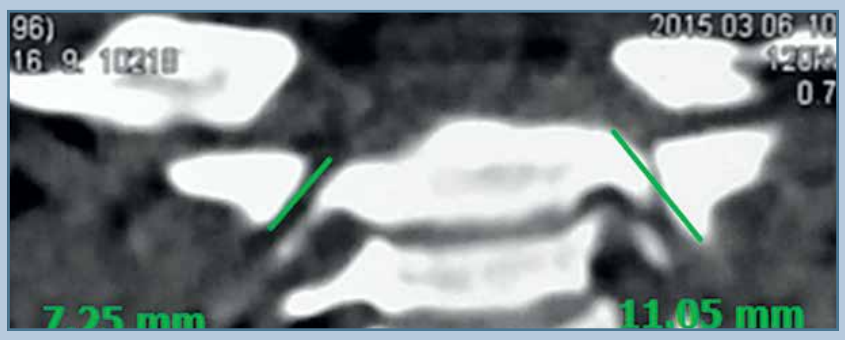

Fig. 2
Median frontal section through the right and left facets of C1 vertebra in a patient with undifferentiated bone dysplasia and odontoid hypoplasia: outward displacement of the right facet by 7.3 mm, outward displacement of the left facet by 11.1 mm, Grade III

accompanied by Binder phenotype (n = 3).

Nonsindromal AAD

Developmental anomalies of the odontoid process and *spina bifida* posterior C1 are characteristic of patients with non-syndromic AAD (Table 1). It should be noted that all patients with os odontoideum (OsO) from the non-syndromic AAD group had a dystopic OsO (Fig. 3a) with significant dislocation (3 to 10 mm in different planes).

In five patients, there was a chronic slowly progressing dislocation without triggering factors; in one patient with os odontoideum, manifestation was subacute after minimal trauma (torticollis, restriction of neck movements, neck pain).

All patients with non-syndromic AAD had localized pain syndrome (VAS 4.20 ± 2.64 points) accompanied by torticollis and restricted head movements, three of them had myelopathy (14.50 ± 4.00 on JOA scale as modified by Benzel).

Analysis of the tilt angle of C1 facets in patients with non-syndromic AAD showed that tilt angle of the inferior facet of C1 in the sagittal plane was 122.00° ± 71.00° on the right and 123.20 ± 72.44° on the left (2 patients with posterior dislocation had an acute, or inverse, tilt angle on both sides), the tilt angle of the inferior facet of C1 in the frontal plane was 149.00° ± 17.94° on the right and 142.30 ± 8.95° on the left. Displacement of the inferior facets of C1 with respect to the facets of C2 was 2.50 ± 3.33 mm on the right and 4.20 ± 2.73 mm on the left in the sagittal plane, 4.20 ± 1.41 mm on the right and 3.30 ± 1.47 mm on the left in the frontal plane. In most cases, anterior and outward displacement was observed, but two patients with an inverted tilt angle of the inferior facets of C1 in the sagittal plane demonstrated an asymmetric posterior displacement of up to 9 mm.

The sagittal diameter of the foramen magnum is 35.90 ± 8.49 mm, the sagittal diameter of C1 is 24.8 ± 2.3 mm. The sagittal diameter of the spinal canal at the level of C1 is 14.70 ± 4.50 mm, at the level of C2 — 14.70 ± 2.40 mm.

ADI and VAAI values could be reliably measured only in three patients without developmental abnormalities of the odontoid process: ADI was 1.70 ± 0.58 mm and VAAI was 0.74 ± 0.21.

Atlantooccipital dissociation (BAI and BDI of more than 12.0 mm) was observed in one patient.

There were developmental anomalies of the cardiovascular, respiratory, and genitourinary systems.

AAD with underlying Klippel – Feil syndrome

AAD with underlying Klippel – Feil syndrome often occurs in the case of C2—C3 block and C1 occipitalization, *spina bifida* posterior C1 occurs in almost half the cases (Table 2).

In patients with this type of AAD, there was chronic, slowly progressing dislocation without triggering factors dominated by local symptoms (Table 3): restriction of neck movements, torticollis, neck pain (VAS 2.40 ± 2.01 points). Three patients had myelopathy of 19.90 ± 2.00 points according to JOA as modified by Benzel.

The tilt angle of the lower facet of C1 in the sagittal plane averaged 132.50° ±

Table 1
The most common malformations of the craniovertebral junction associated with non-syndromic atlantoaxial dislocations

Anomaly	Patients, n (%)
Spina bifida C1	3 (50.0); 2 post, 1 ant
Os odontoideum	3 (50.0); dystopic
Occipitalization of C1	2 (33.3)

53.06° on the right and 139.30° ± 39.82° on the left, the tilt angle of the lower facet of C1 in the frontal plane was 148.60° ± 16.57° on the right and 149.30° ± 17.31° on the left. The displacement of the inferior facets of C1 was 2.60 ± 1.35 mm on the right and 4.90 ± 3.88 mm on the left in the sagittal plane, 2.50 ± 2.21 mm on the right and 2.80 ± 3.15 mm on the left in the frontal plane. The facets were most often displaced in anterior and outward direction. The sagittal diameter of the foramen magnum was 33.80 ± 5.04 mm, C1 — 29.70 ± 4.58 mm. The sagittal diameter of the spinal canal at the level of C1 was 17.60 ± 6.58 mm, at the level of C2 — 16.10 ± 4.12 mm. ADI and VAAI values were reliably measured in 8 patients without development anomalies of the odontoid process and amounted to 4.10 ± 1.92 mm and 0.76 ± 0.15, respectively. Atlantooccipital dissociation (BAI and BDI of more than 12.0 mm) was observed in 5 (45.5 %) patients.

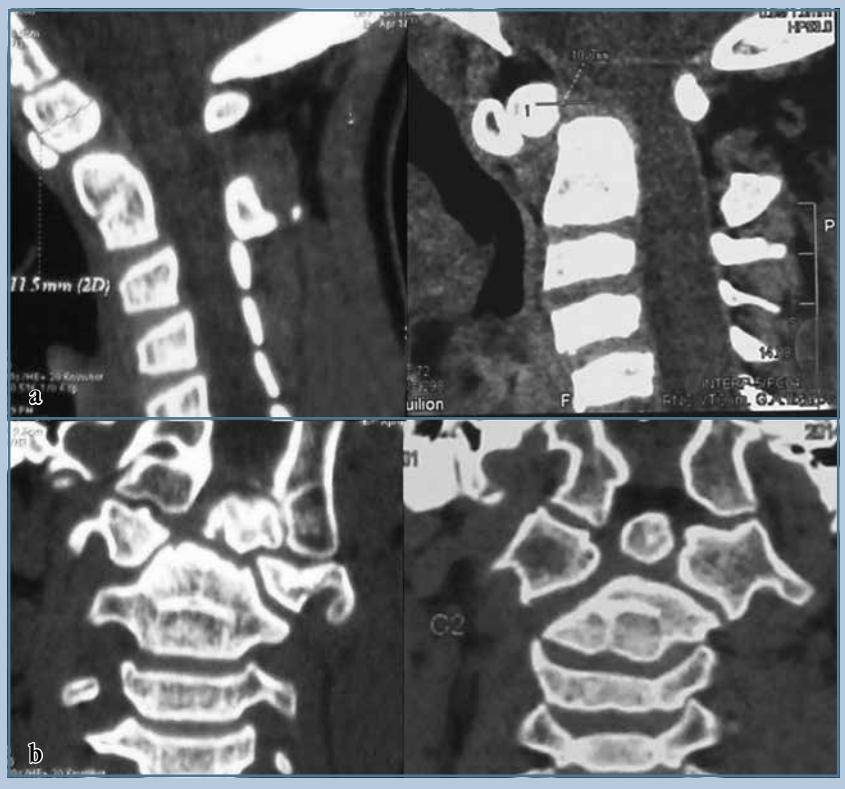

Fig. 3
Median frontal and sagittal CT sections of a patient with dystopic os odontoideum from the group of non-syndromic atlantoaxial dislocations: anterior and left displacement of the os odontoideum **(a)**; median frontal and sagittal CT sections of the patient with orthotopic os odontoideum from the group of syndromic atlantoaxial dislocations: anterior and left displacement of the os odontoideum **(b)**

Seven patients had hemifacial microsomia (Goldenhar phenotype), three patients with myelopathy had combined progressive scoliosis. Concomitant heart abnormalities were detected in 3 patients and one patient with each of the respiratory system anomaly, mental retardation, grade 2 obesity, celiac disease, and chronic tonsillitis.

Syndromal AAD

Os odontoideum was observed in 7 patients in the group of syndromic AAD, it was orthotopic in 6 patients (Fig. 3b); displacement of the odontoid process from 3 to 12 mm was observed in 4 patients (Table 4).

There was chronic slowly progressing dislocation in 6 patients with syndromic AAD without triggering factors; manifestation was subacute or acute after minimal trauma (tetraparesis) in 3 patients with developmental anomalies of the odontoid process.

Symptoms of myelopathy (8.30 ± 5.32 on JOA as modified by Benzel) and local pain syndrome (VAS 2.30 ± 1.94 points, Table 5) dominated in patients with syndromic AAD.

In patients with syndromic congenital AAD, tilt angle of the inferior facet of C1 was 149.10 ± 14.63° on the right and 145.60° ± 21.80° on the left in the sagittal plane, 145.80° ± 12.86° on the right and 141.20° ± 16.22° on the left in the frontal plane. The displacement of C1 facet was 5.30 ± 4.00 mm on the right and 4.90 ± 3.82 mm on the left in the sagittal plane, 5.20 ± 2.11 mm on the right and 5.30 ± 2.40 mm on the left in the frontal plane; displacement most often occurred in anterior and outward direction. The sagittal diameter of the foramen magnum was 32.00 ± 6.04 mm, the sagittal diameter of C1 was 27.00 ± 7.11 mm. The sagittal diameter of the spinal canal at the level of C1 was 7.80 ± 2.35 mm, at the level of C2 — 11.10 ± 3.13

Table 2
The most common malformations of the craniovertebral junction associated with atlantoaxial dislocations with underlying Klippel — Feil syndrome

Anomaly	Patients, n (%)
C2—C3 block	9 (81.8)
Spina bifida C1	5 (45.5); 2 post, 3 ant + post
Occipitalization of C1	5 (45.5)
Odontoid hypoplasia	2 (18.2)
Spina bifida C2	2 (18.2)
Os odontoideum	1 (9.1); дистопическая
Chiari malformation	1 (9.1)
Syringomyelia	1 (9.1)

Table 3
Symptoms in the group of patients with atlantoaxial dislocations associated with Klippel — Fail syndrome

Symptoms	Patients, n (%)
Restricted neck movement	11 (100.0)
Torticollis	10 (90.1)
Neck pain	8 (72.7)
Hemifacial microsomia	7 (63.6)
Myelopathy	3 (27.3)

Table 4
The most common malformations of the craniovertebral junction associated with syndromic atlantoaxial dislocations

Anomaly	Patients, n (%)
Os odontoideum	7 (77.8); 6 orthotopic, 1 dystopic
Spina bifida C1	4 (44.4); 4 post, 3 ant
Odontoid hypoplasia	2 (22.2)
Spina bifida C2	1 (11.1)

mm. Reliable measurement of ADI and VAAI was impossible, since all patients from this group had developmental anomaly of the odontoid process of C2. Atlantooccipital dissociation (BAI and BDI of more than 12.0 mm) was detected in 6 patients.

One patient with undifferentiated bone dysplasia had cardiac malformation, 2 patients had deformities of the upper respiratory tract of varying severity (constriction and deformation of the trachea and bronchi), 3 patients had underdevelopment of the midface (Binder phenotype), 6 patients with underlying myelodysplasia and bone dysplasia developed combined progressive scoliosis, 3 patients had talipes valgus with articular contractures, 3 patients had mental retardation.

VAA1 in patients with developmental abnormalities of the odontoid process

VAA1 was 0.73 ± 0.53 in 4 patients with odontoid hypoplasia, 0.59 ± 0.16 in 11 patients with OsO (the apex of the odontoid process was considered as the apex of the os odontoideum). Normal VAA1 values (0.71 to 0.80) were observed in four patients out of 26; 0.81 to 1.00 in 5 patients, while outward displacement of both facets of C1 with respect to C2, i.e. axial dislocation, was observed in 4 patients.

Displacement of the os odontoideum

In the case of non-syndromic AAD with underlying dystopic OsO (3 patients), displacement in different planes up to 3 mm was observed. In the case of AAD with underlying Klippel – Feil syndrome, dystopic OsO was detected in only one patient in the form of anterior and left dislocation by 8 mm. In the case of syndromic AAD with OsO (7 patients), orthotopic OsO was observed in 6 patients, and dystopic OsO was observed in 1 patient; 3 patients with orthotopic OsO had no bone dislocation. Os odontoideum was displaced in one of three planes (by 3 to 9 mm) in 3 patients, simultaneously in two planes (by 3 to 12 mm) — in 5 patients.

Discussion

Dislocation direction. We used several classifications which turned out to be quite non-uniform when analyzing the literature.

Anterior and posterior AAD. Anterior dislocations can be associated with transverse ligament laxity, where C2 tilts in posterior direction during flexion and straightened during extension. Its anterior motion is restricted by C1 arch, so that only anterior displacement of C1 is observed in this case [1–3, 31]. Dislocation in two planes is possible in the case of the os odontoideum or odontoid aplasia, when C2 body movements are not restricted by the transverse ligament and the anterior arch of C1, so that C2 body can be displaced in two directions, anterior and posterior [1–3, 31].

The proportion of anterior dislocations in our patient setting was much higher compared to the proportion of posterior ones (22 and 2 patients, respectively), which is most likely associated with the anatomical features of the craniovertebral junction, the physiological downward slanting of the plane of C1 facet in the posterior-

Table 5
Symptoms in patients with syndromic atlantoaxial dislocations

Symptoms	Patients, n (%)
Scoliosis	8 (88.9)
Tetraparesis	8 (88.9)
Neck pain	7 (77.8)
Pelvic dysfunction	3 (33.3)
Midface underdevelopment	3 (33.3)
Equinovarus	2 (22.2)

anterior direction: reverse orientation of the facet plane (downward slanting in the anterior-posterior direction) was observed in two patients with a posterior dislocation.

Rotational AAD. As a rule, this type of dislocation occurs in one plane (axial) due to pterygoid ligament insufficiency in children, as well as congenital malformations of the cervical spine [4].

There are several types of rotational AAD [13, 21], and Samartzis et al. [33] extended this classification for the use in patients with congenital pathology of the cervical spine. This classification evaluates C1 dislocation in both sagittal and axial planes simultaneously, which is its advantage: type I implies a pure rotational dislocation, types II and III — rotational and anterior.

Normal ADI in children is 4 mm or less, which necessitates its modification for pediatric patients and it is a drawback of this classification. Furthermore, despite the fact that it implies gradation of rotational AADs, it does not directly evaluate the degree of C1 rotation, but rather the degree of the anterior dislocation of C1.

Central (vertical) AAD. As noted earlier, the adjacent C1—C2 facets are normally horizontal and parallel; in the case of their slant orientation in the frontal plane, C2 body tends to slide upward with respect to the head (due to the weight of the patient's head during flexion). Therefore, high tilt angle of these joints in the frontal plane leads to telescoping of the C2 body into C1 ring, central dislocation, and posterior displacement of C2. In the case of true basilar invagination (Goel type A), basal skull structures forming the foramen magnum are raised or invaginated into the posterior cranial fossa. Invagination of the foramen magnum automatically leads to superior displacement of C1 and C2; nevertheless, C1 and C2 have normal structure and relationships in the case of type A basilar invagination. When the relationships between C1 and C2 are abnormal, the correct diagnosis is either "central AAD" (in the case of mild dislocation), or "axial invagination" (in extreme forms), or "type B basilar invagination".

Therefore, type A basilar invagination is an abnormal relationship between the foramen magnum and the posterior cranial fossa, whereas AAD (type B basilar invagination) is the central dislocation of C2 or axial invagination. These states can occur concomitantly [11].

This concept of AAD is quite new, and therefore, central AAD is considered as a form of basilar invagination rather than individual pathology in most sources.

Various ways to determine the degree of basilar invagination based on radiological parameters have been proposed: Chamberlain line [5], McGregor line [22], McRae line [24]. Kulkarni and Goel [15] recommend using VAAI to determine the degree of vertical AAD. Despite the variety of indices and measurements, normal values for various age groups of children are not defined. Furthermore, our experience shows that the measured data are not reliable in the case of anomalies of the C2 odontoid process (hypoplasia, aplasia, OsO). The central AAD can be estimated based on the disposition of the lateral masses of C1 with respect to C2 in the frontal plane on the frontal section through the middle of the lateral masses of C1.

Hybrid AAD. In our group, 23 (88.5 %) patients had a hybrid AAD.

We found some patterns characteristic of different AAD groups:

1) patients with non-syndromic AAD had moderate to severe dislocation (Grade II—III) in three planes, 4 (66.6 %) cases;

2) most patients with AAD associated with Klippel—Feil syndrome had a biplanar dislocation: anterior Grade II—III (91.0 %) and rotary Grade I—II (63.6 %) dislocation;

3) most patients with syndromic AAD had Grade II—III dislocation in the sagittal and frontal planes (anterior and central) (88.9 %).

Unilateral and bilateral AAD. Lutsik et al. [1, 2] classify C1 dislocations into unilateral and bilateral. All our patients had bilateral dislocations.

AAD reducibility and stability of the craniovertebral junction.

AAD is reducible, when C1 and C2 can be reduced by flexion/extension of the neck or traction. Possible reasons of non-reducibility of AAD (fixed or superstable AAD) include bone or fibrous block of joints and ligaments, inadequate traction, pain and muscle spasm, which can hinder reduction even in the case of adequate traction, transligamentous dislocation of the odontoid process [12, 16, 31, 37, 40, 41].

Deepak et al. [7] found a number of patterns for reducible and non-reducible AADs: the incidence of segmentation disorders (Klippel – Feil syndrome) was significantly higher in the case of non-reducible AADs compared to reducible ones; at the same time, os odontoideum was much more common in the case of reducible AADs than with non-reducible ones. Further, non-reducible AADs are more often associated with developmental abnormalities of the vertebral artery.

Salunke et al. [31] and Behari et al. [3] found that patients with reducible AADs are characterized by slightly more acute sagittal angle of the lower facet of C1 compared to that in healthy individuals. In the case of non-reducible AADs with

underlying congenital abnormalities of the craniovertebral junction, facets of C1—C2 joints are more inclined in the sagittal and coronal planes [32]. The more acute is (craniocaudal) inferior sagittal facet angle of C1, the earlier the non-reducibility stage arises. Additionally, age-related changes make the lower cervical joints rigid. This gradual restriction of movement increased load on the ligaments and capsules of C1—C2 joints and enhances instability [32].

Several criteria have been proposed to determine atlantoaxial instability: ADI of more than 4 mm in children and more than 3 mm in adults [10] with a neutral head position, a difference between ADI in flexion and extension of 3.5 mm [18] or 3 mm (3—6 mm — grade I, more than 6 mm — grade II) [1, 11, 12], in the case of rotation, depending on the angle of the atlantoaxial rotation (40 to 60° — grade I, more than 60° — grade II) [1]. The individual morphology of the defect and the age of a child should be kept in mind, since ADI up to 4 mm is normal in children [18]; hypermobility in the atlantoaxial articulation is also normal in patients with Down syndrome [20, 23, 26, 30].

The information content of the functional radiographs is quite low compared to CT and MRI. It is recommended to carry out functional flexion-extension CT and MRI of the craniovertebral junction in all patients with os odontoideum, odontoid hypoplasia and aplasia, as well as genetic syndromes [6, 8, 19]. We did not carry out routine functional CT and MRI of the craniovertebral junction to determine stability and reducibility of dislocations and were guided by the presence of developmental abnormalities of the odontoid process, the degree of C1 dislocation in prone position, the presence of stenosis of the spinal canal, and signs of myelopathy.

When measuring the tilt angles of the facets of C1 in the sagittal plane, the following patterns were found:

1) the tilt angle is more acute in the group of non-syndromic AADs and AADs with underlying Klippel—Feil syndrome;

2) the flattest position of the plane of the inferior facets of C1 and the smallest variation of the values was observed in the group of syndromic AAD, i.e., they were the most unstable and mobile, which confirms the presence of the development anomalies of the odontoid process in all patients in this group;

3) the average tilt angle of the facets is 136.40° ± 48.61° on the right and 136.90° ± 44.6° on the left in all groups, that is, less than 150°, which explains the higher incidence of C1 dislocation in the sagittal plane.

Measurement of the tilt angles of C1 facets in the frontal plane showed the following pattern: the tilt angle was more acute in the group of syndromic AAD, which may explain the higher incidence of Grade II and Grade III central dislocations in this group.

Syndromic and non-syndromic AADs

There are a number of differences between syndromic and non-syndromic AADs with underlying congenital pathology of the craniovertebral junction: for example, AADs with underlying genetic syndromes are often reducible; non-reducible AADs are more common in patients with the Klippel-Feil syndrome or with an isolated abnormality of the craniovertebral junction. In this case, myelopathy and stenosis of the spinal canal are more characteristic of syndromic AADs [23, 34]. All the aforementioned features were confirmed in our study.

Conclusion

It is important to differentiate between the syndromic and non-syndromic forms of AADs and AADs with underlying Klippel – Feil anomaly, since the prognosis in these patients is different. Myelopathy is more common in the case of syndromic AADs, these patients require early and aggressive surgical treatment. AADs with underlying Klippel – Feil syndrome and non-syndromic AADs often manifest in the form of local symptoms.

The existing classifications of AADs are not always suitable for patients with congenital AADs, since developmental abnormalities of the odontoid process are common in this group. We proposed a new classification based on the evaluation of the displacement of the inferior facets of C1 with respect to C2 in the sagittal and frontal planes as shown by CT.

The study was not sponsored. The authors declare no conflict of interest.

References

1. **Vetrile ST, Kolesov SV.** Craniovertebral Pathology. Moscow, 2007. In Russian.
2. **Lutsik AA, Ratkin IK, Nikitin MN.** Craniovertebral Junction Lesions and Diseases. Novosibirsk, 1998. In Russian.
3. **Behari S, Bhargava V, Nayak S, Kiran Kumar MV, Banerji D, Chhabra DK, Jain VK.** Congenital reducible atlantoaxial dislocation: classification and surgical considerations. Acta Neurochir (Wien). 2002;144:1165–1177. DOI: 10.1007/s00701-002-1009-3.
4. **Behari S, Jain VK, Phadke RV, Banerji D, Kathuria M, Chhabra DK.** C1–C2 rotary subluxation following posterior stabilization for congenital atlantoaxial dislocation. Neurol India. 2000;48:164–169.
5. **Chamberlain WE.** Basilar impression (platybasia): a bizarre developmental anomaly of the occipital bone and upper cervical spine with striking and misleading neurologic manifestations. Yale J Biol Med. 1939;11:487–496.
6. **Da Silva OT, Ghizoni E, Tedeschi H, Joaquim AF.** Role of dynamic computed tomography scans in patients with congenital craniovertebral junction malformations. World J Orthop. 2017;8:271–277. DOI: 10.5312/wjo.v8.i3.271.
7. **Deepak AN, Salunke P, Sahoo SK, Prasad PK, Khandelwal NK.** Revisiting the differences between irreducible and reducible atlantoaxial dislocation in the era of direct posterior approach and C1–C2 joint manipulation. J Neurosurg Spine. 2017;26:331–340. DOI: 10.3171/2016.8.SPINE16408.
8. **El Asri AC, Akhaddar A, Gazzaz M, Okacha N, Boulhroud O, Baallal H, Belfquih H, Belhachmi A, Mandour C, El Mostarchid B, Boucetta M.** Dynamic CT scan of the craniovertebral junction: a role in the management of os odontoideum. Neurol Neurochir Pol. 2010;44:603–608. DOI: 10.1016/S0028-3843(14)60159-6.
9. **Fielding JW, Hawkins RJ.** Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg Am. 1977;59:37–44.
10. **Fielding JW, Cochran GV, Lawsing JF 3rd, Hohl M.** Tears of the transverse ligament of the atlas. A clinical and biomechanical study. J Bone Joint Surg Am. 1974;56:1683–1691.
11. **Goel A, Bhatjiwale M, Desai K.** Basilar invagination: a study based on 190 surgically treated patients. J Neurosurg. 1998;88:962–968. DOI: 10.3171/jns.1998.88.6.0962.
12. **Goel A, Shah A, Rajan S.** Vertical mobile and reducible atlantoaxial dislocation. Clinical article. J Neurosurg Spine. 2009;11:9–14. DOI: 10.3171/2009.3.SPINE08927.
13. **Ishii K, Toyama Y, Nakamura M, Chiba K, Matsumoto M.** Management of chronic atlantoaxial rotatory fixation. Spine. 2012;37:E278–E285. DOI: 10.1097/BRS.0b013e31823cc2ea.
14. **Jain VK.** Atlantoaxial dislocation. Neurol India. 2012;60:9–17. DOI: 10.4103/0028-3886.93582
15. **Kulkarni AG, Goel AH.** Vertical atlantoaxial index: a new craniovertebral radiographic index. J Spinal Disord Tech. 2008;21:4–10. DOI: 10.1097/BSD.0b013e31804856d7.
16. **Laheri V, Chaudhary K, Rathod A, Bapat M.** Anterior transoral atlantoaxial release and posterior instrumented fusion for irreducible congenital basilar invagination. Eur Spine J. 2015;24:2977–2985. DOI: 10.1007/s00586-015-3836-y.
17. **Li L, Yu X, Wang P, Chen L.** Analysis of the treatment of 576 patients with congenital craniovertebral junction malformations. J Clin Neurosci. 2012;19:49–56. DOI: 10.1016/j.jocn.2011.03.036.
18. **Locke G, Gardner J, Van Epps E.** Atlas-dens interval (ADI) in children: a survey based on 200 normal cervical spines. Am J Roentgenol Radium Ther Nucl Med. 1966;97:135–140.
19. **Mackenzie WG, Dhawale AA, Demczko MM, Ditro C, Rogers KJ, Bober MB, Campbell JW, Grissom LE.** Flexion-extension cervical spine MRI in children with skeletal dysplasia: is it safe and effective? J Pediatr Orthop. 2013;33:91–98. DOI: 10.1097/BPO.0b013e318279c51f.
20. **Martich V, Ben-Ami T, Yousefzadeh D, Roizen N.** Hypoplastic posterior arch of C-1 in children with Down syndrome: a double jeopardy. Radiology. 1992;183:125–128. DOI: 10.1148/radiology.183.1.1532260.
21. Mazzara JT, Fielding JW. Effect of C1–C2 rotation on canal size. Clin Orthop Relat Res. 1988;(237):115–119.
22. **McGregor M.** The significance of certain measurements of the skull in the diagnosis of basilar impression. Br J Radiol. 1948;21:171–181. DOI: 10.1259/0007-1285-21-244-171.
23. **McKay SD, Al-Omari A, Tomlinson LA, Dormans JP.** Review of cervical spine anomalies in genetic syndromes. Spine. 2012;37:E269–E277. DOI: 10.1097/BRS.0b013e31823b3ded.
24. **McRae DL, Barnum AS.** Occipitalization of the atlas. Am J Roentgenol Radium Ther Nucl Med. 1953;70:23–46.
25. **Mehrotra A, Nair AP, Das K, Chunnilal JS, Srivastava AK, Sahu R, Kumar R.** Congenital paediatric atlantoaxial dislocation: clinico-radiological profile and surgical outcome. Childs Nerv Syst. 2012;28:1943–1950. DOI: 10.1007/s00381-012-1801-1.
26. **Menezes AH, VanGilder JC, Graf CJ, McDonnell DE.** Craniocervical abnormalities. A comprehensive surgical approach. J Neurosurg. 1980;53:444–455. DOI: 10.3171/jns.1980.53.4.0444.
27. **Parikh RN, Muranjan M, Karande S, Sankhe S.** Atlas shrugged: cervical myelopathy caused by congenital atlantoaxial dislocation aggravated by child labor. Pediatr Neurol. 2014;50:380–383. DOI: 10.1016/j.pediatrneurol.2013.11.014.
28. **Passias PG, Wang S, Kozanek M, Wang S, Wang C.** Relationship between the alignment of the occipitoaxial and subaxial cervical spine in patients with congenital atlantoaxial dislocations. J Spinal Disord Tech. 2013;26:15–21. DOI: 10.1097/BSD.0b013e31823097f9.
29. **Pedachenko EG, Slynko EI, Malysheva TA, Robak OP, Chernenko OG.** Structural features and diagnostics of axis maldevelopments, a differentiated choice of surgical tactics. Ukrainian Neurosurgical Journal. 2015;2:25–31. DOI: 10.25305/unj.45292.
30. **Pizzutillo PD, Herman MJ.** Cervical spine issues in Down syndrome. J Pediatr Orthop. 2005;25:253–259.
31. **Salunke P, Behari S, Kirankumar MV, Sharma MS, Jaiswal AK, Jain VK.** Pediatric congenital atlantoaxial dislocation: differences between the irreducible and reducible varieties. J Neurosurg. 2006;104(2 Suppl):115–122. DOI: 10.3171/ped.2006.104.2.115.
32. **Salunke P, Sharma M, Sodhi HB, Mukherjee KK, Khandelwal NK.** Congenital atlantoaxial dislocation: a dynamic process and role of facets in irreducibility. J Neurosurg Spine. 2011;15:678–685. DOI: 10.3171/2011.7.SPINE1152.
33. **Samartzis D, Shen FH, Herman J, Mardjetko SM.** Atlantoaxial rotatory fixation in the setting of associated congenital malformations: a modified classification system. Spine. 2010;35:E119–E127. DOI: 10.1097/BRS.0b013e3181c9f957.
34. **Sardhara J, Behari S, Jaiswal AK, Srivastava A, Sahu RN, Mehrotra A, Phadke S, Singh U.** Syndromic versus nonsyndromic atlantoaxial dislocation: do clinico-radiological differences have a bearing on management? Acta Neurochir (Wien). 2013;155:1157–1167. DOI: 10.1007/s00701-013-1717-x.
35. **Syre P, Petrov D, Malhotra N.** Management of upper cervical spine injuries: a review. J Neurosurg Sci. 2013;57:219–240.
36. **Wang S, Wang C, Leng H, Zhao W, Yan M, Zhou H.** Cable-strengthened C2 pedicle screw fixation in the treatment of congenital C2–C3 fusion, atlas occipitalization, and atlantoaxial dislocation. Neurosurgery. 2012;71:976–984. DOI: 10.1227/NEU.0b013e31826cdd3b.
37. **Wang S, Wang C, Yan M, Zhou H, Jiang L.** Syringomyelia with irreducible atlantoaxial dislocation, basilar invagination and Chiari I malformation. Eur Spine J. 2010;19:361–366. DOI: 10.1007/s00586-009-1208-1.

38. **Wang C, Yan M, Zhou HT, Wang SL, Dang GT.** Open reduction of irreducible atlantoaxial dislocation by transoral anterior atlantoaxial release and posterior internal fixation. Spine. 2006;31:E306–E313. DOI: 10.1097/01.brs.0000217686.80327.e4.
39. **Yang SY, Boniello AJ, Poorman CE, Chang AL, Wang S, Passias PG.** A review of the diagnosis and treatment of atlantoaxial dislocations. Global Spine J. 2014;4:197–210. DOI: 10.1055/s-0034-1376371.
40. **Zhao D, Wang S, Passias PG, Wang C.** Craniocervical instability in the setting of os odontoideum: assessment of cause, presentation, and surgical outcomes in a series of 279 cases. Neurosurgery. 2015;76:514–521. DOI: 10.1227/NEU.0000000000000668.
41. **Yin QS, Wang JH.** Current trends in management of atlantoaxial dislocation. Orthop Surg. 2015;7:189–199. DOI: 10.1111/os.12196.

Address correspondence to:
Pavlova Olga Mikhailovna
Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics,
M. Ulyanovoy str., 6, Kurgan, 640014, Russia,
pavlova.neuro@mail.ru

Received 28.08.2017
Review completed 26.09.2017
Passed for printing 05.10.2017

Olga Mikhailovna Pavlova, neurosurgeon, orthopedist-traumatologist, junior researcher in the Laboratory of Axial Skeletal Pathology and Neurosurgery, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, M. Ulyanovoy str., 6, 640014, Kurgan, Russia, pavlova.neuro@mail.ru;
Sergey Olegovich Ryabykh, DMSc, pediatric surgeon, orthopedist-traumatologist, vertebrologist, Head of the Clinic of Spine Pathology and Rare Diseases, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, M. Ulyanovoy str., 6, 640014, Kurgan, Russia, rso_@mail.ru;
Aleksandr Vladimirovich Burtsev, MD, PhD, orthopedist-vertebrologist, researcher, Laboratory of Axial Skeletal Pathology and Neurosurgery, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, M. Ulyanovoy str., 6, 640014, Kurgan, Russia, bav31rus@mail.ru;
Alexandr Vadimovich Gubin, DMSc, orthopedist-traumatologist, Director, Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, M. Ulyanovoy str., 6, 640014, Kurgan, Russia, shugu19@gubin.spb.ru.