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Atlantoaxial rotatory dislocation: Surgical treatment in a pediatric patient cohort

Jan Štulík, Michaela Rybárová *, Michal Barna, Zdeněk Klézl

Center for Spinal Surgery, First Faculty of Medicine, Charles University and University Hospital Motol, Prague, Czech Republic

1. Introduction

Rotatory dislocation of the atlantoaxial complex was for the first time described by Bell (1830) in 1830 and subsequently by Corner (1907) in 1907. In 1968, Wortzman and Dewar (1968) introduced the term "atlantoaxial rotatory fixation". The most frequent synonyms used for rotatory pathology of the atlantoaxial complex of various etiologies include atlantoaxial rotatory dislocation (AARD), atlantoaxial rotatory fixation (AARF) or atlantoaxial rotatory subluxation (AARS). AARD most often occurs in childhood and in majority of cases it is successfully treated non-operatively in the primary care setting, especially during the first three weeks after the onset of the disease (Hill et al., 2021; Kinon et al., 2016; Pang, 2010; Pang and Li, 2004, 2005a, 2005b; Powell et al., 2017; Sae-Huang et al., 2020; Wang et al., 2016). The increased incidence of this pathology in children may be explained by specific anatomical features of an immature spine (Goel, 2021; Hill et al., 2021; Sae-Huang et al., 2020; Štulík, 2012). AARD is divided into acute and chronic lasting for more than 3 months (Ishii et al., 2011; Pang, 2010; Pang and Li, 2004, 2005a, 2005b). The clinical features include torticollis, neck pain and stiffness. The most obvious manifestation is the so-called cock-robin position of the head, with the chin tilted to one side and the neck flexed laterally to the other (Pang, 2010; Pang and Li, 2004, 2005a, 2005b) (Fig. 1). Torticollis in association with AARD is often linked to spreading of the laryngeal inflammation through the lymphatic and venous system and post-inflammatory ligament laxity and muscle spasm (Pang, 2010; Pang and Li, 2004, 2005a, 2005b). Other etiologic factors may be injuries in the region of the head and neck, the shoulder girdle, the chest or surgical procedures in this location (Powell et al., 2017). However, etiology remains unclear in up to a half of the patients (Beier et al., 2012; Powell et al., 2017). Non-traumatic atlantoaxial rotatory subluxation (AARS), often associated with infection in the region of the neck or head, is called the Grisel's syndrome (Barcelos et al., 2014). In 1977, Fielding and Hawkins (1977) published a generally accepted classification dividing AARD into 4 types. Wang et al. (2016) call the first two types "atlantoaxial rotatory fixation" (AARF) and the other two types "atlantoaxial rotatory fixed dislocation" (AARFD). Pang and Li (Pang, 2010; Pang and Li, 2004, 2005a, 2005b) developed their classification on the basis of a 3-position CT examination, i.e., a scan in displacement position and stress scans in zero position and in rotation of the head toward the other side as tolerated by the patient. Ishii et al. (2012) proposed a classification according to the degree of facet deformity and inclination of the atlas. Operative treatment is required of the patients after a failed non-operative therapy, repeated dislocations, and in the presence of anatomical defects of the atlantoaxial complex (Goel and Laheri, 1994; Goel and Shah, 2011; Goel, 2019, 2021; Hill et al., 2021; Kinon et al., 2016).

The aim of the present study was to evaluate radiological and clinical results of patients with AARD treated surgically at our center.

2. Methods

2.1. Patients

Between 2001 and 2021 we treated operatively 12 patients with AARD younger than 16 years at the Center for Spinal Surgery, 1st Faculty of Medicine, Charles University and University Hospital Motol, Prague. The group comprised 5 boys and 7 girls, with the mean age of 8.5 years (range, 5–16). All patients included in the study group had an intact odontoid peg, others, with odontoid peg defects or Klippel-Feil syndrome were excluded.

The study protocol was approved by the ethical committee of our hospital. All patients provided written consent (by parents) to be enrolled in the study and allowed publication of photographic documentation. Detailed patient characteristics and their corresponding atlantoaxial morphology are shown in Table 1 + 2. All patients underwent preoperative radiographic examination consisting of cervical spine radiographs in anteroposterior and lateral projection including the head, as well as MRI and CT scans of the craniocervical junction involving 3D reconstructions. Results of these examinations and patients' clinical status served as the basis for surgical planning.

* Corresponding author.
E-mail address: michaela.rybar@gmail.com (M. Rybárová).

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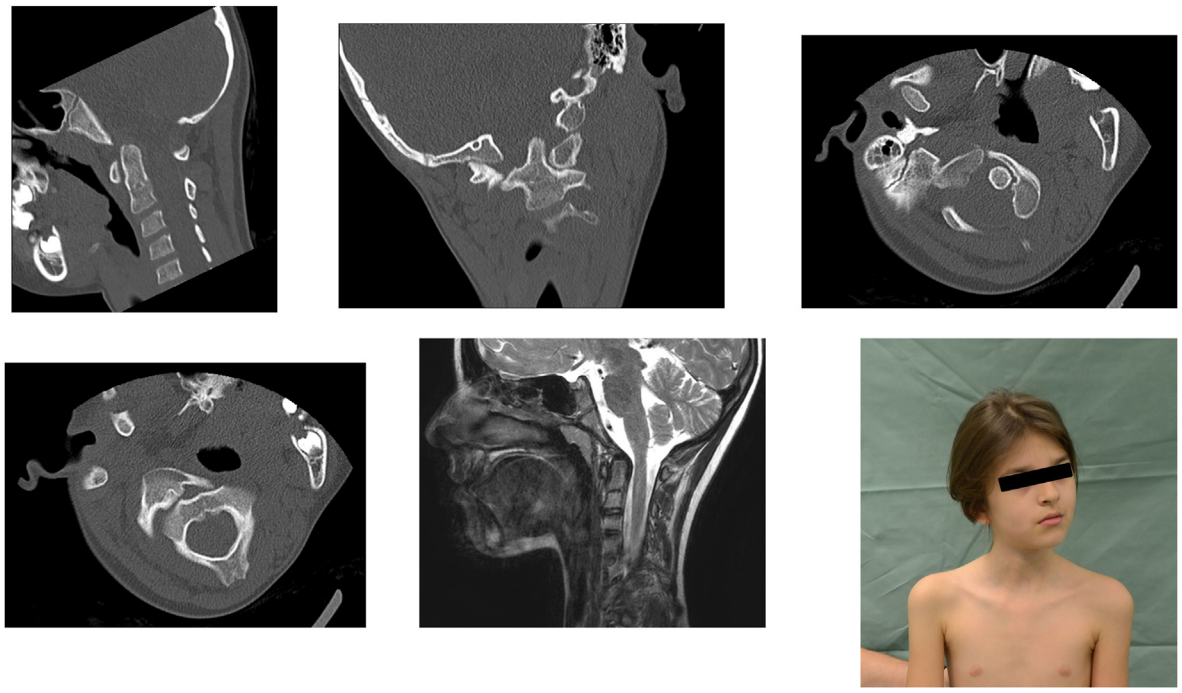

Fig. 1. Preoperative imaging studies and a clinical photograph of an 8 + 1 female patient with a history of a month long AARD type III according to Ishii et al. classification, previously treated non-operatively. CT reconstructions in a sagittal plane (a), coronal plane (b), axial cut at the level of C1 (c) and C2 (d) vertebra. Preoperative MRI T2-weighed sagittal image (e). The cock-robin head position (f).

Table 1
Surgical data.

Pt	Sex	Age	Type of surgery	Surgical time (min)	Blood loss (ml)	X-ray time (s)	FUP (m)	Etiology	Traction	Bone graft	Fusion
1	M	7	reduction + fixation	80	400	90	183	trauma	YES	YES	YES
2	F	7	reduction + fixation	120	500	13	168	respiratory infection	YES	YES	YES
3	F	16	reduction + fixation	80	500	36	161	trauma	YES	YES	YES
4	F	8	reduction + fixation	80	100	128	130	Down sy.	YES	YES	YES
5	F	5	reduction + fixation	52	100	42	130	trauma	YES	NO	NO
6	F	8	reduction + fixation	100	300	60	101	unknown	YES	YES	YES
7	M	14	reduction + fixation	40	200	52	101	unknown	YES	YES	YES
8	M	8	reduction + fixation	90	400	52	51	respiratory infection	YES	YES	YES
9	F	7	reduction + fixation	140	100	15	33	unknown	YES	YES	YES
10	F	8	reduction + fixation	100	50	20	34	trauma	YES	YES	YES
11	M	7	reduction + fixation	90	200	18	21	respiratory infection	YES	YES	YES
12	M	7	reduction + fixation	90	300	15	6	general anesthesia	YES	YES	YES
Average:		**8,5**		**88,5**	**262,5**	**47,1**	**93,5**				

2.2. Preoperative protocol

Prior to surgery, radiographs were performed in two views, as well as CT scans including the sagittal and coronal reconstructions, 3D reconstructions and MRI in 3 planes. In all patients, traction with Glisson loop was applied for 3–5 days, initially with a load corresponding to 5% of the patient's body weight, gradually increasing over 24 h to 10% of the body weight as tolerated; patients with persisting rotation were indicated for operative treatment. All patients were operated on by a single surgeon from the posterior approach (Fig. 2).

2.3. Surgical technique

The patient under general endotracheal anesthesia and after application of muscle relaxants was placed on a standard operating table in the prone position and fixed with adhesive tapes in the maximum possible correction. A midline incision was made to expose the region of the first and second cervical vertebrae, the segment between the superior edge of the posterior arch of the atlas and the C2–C3 intervertebral joint was exposed and orientation of vertebrae assessed. Spontaneous reduction did not occur in any of the patients. The release of the both atlantoaxial joint capsules was necessary in 6 patients. Subsequently, partially threaded 3.5 mm or 4.0 mm shaft screws were inserted into C1 according to Goel (Goel and Laheri, 1994; Štulík, 2012) and fully threaded screws into C2 according to Harms (Harms and Melcher, 2001; Štulík, 2012). C1–C2 intervertebral joints were exposed and the fixator was assembled (6x S4Cervical, Aesculap, Germany, 1x Moss Miami, Johnson and Johnson, USA, 5x Virage, Zimmer, USA), with the following correction of the deformity and final tightening of the fixator. Screws served as joy-sticks to facilitate the correction of the deformity. Appropriate direction of screw insertion is crucial for final correction. This surgical technique was unified at our department; we used neither neuromonitoring nor 3D intraoperative guidance imaging.

Correct position of the atlantoaxial complex was verified by the relation between the posterior tubercle of atlas and the C2 spinous process. In absence of anatomical reference points, the position was assessed based on anatomical symmetry. In two cases it was necessary to insert an auxiliary temporary wire loop below the C1 arch and correct the deformity simultaneously by both the fixator and the wire loop that was removed after reduction and tightening of the assembly. In a majority of

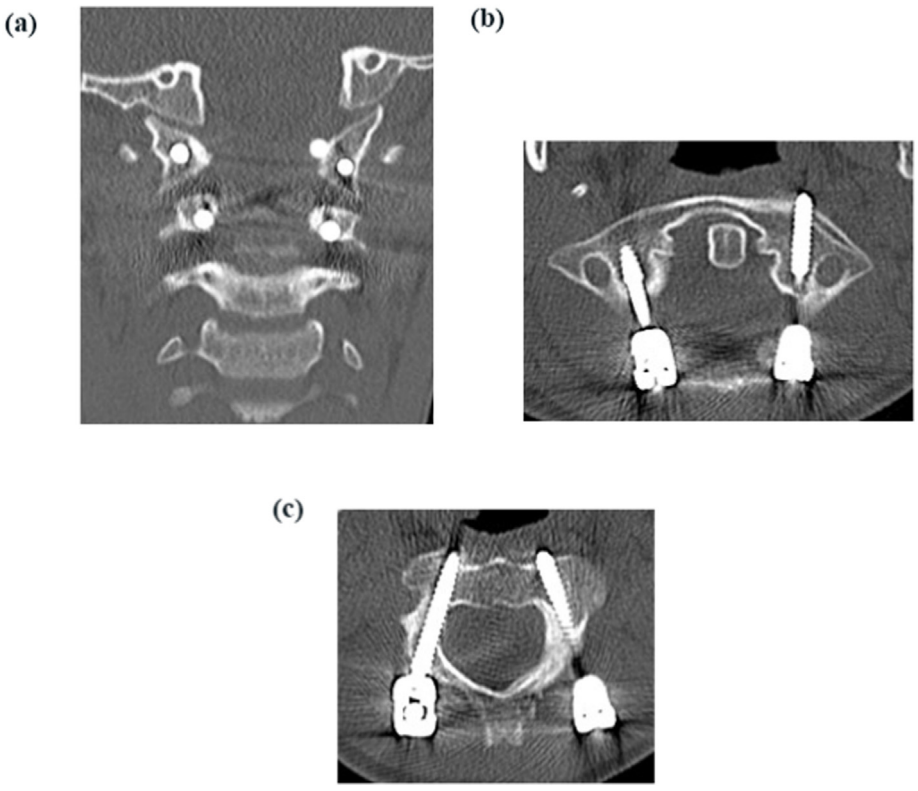

Fig. 2. Imaging studies of the same patient after instrumented C1–C2 reduction and fusion (Virage, Zimmer, USA). CT reconstructions in coronal and axial planes (a–c) show correct placement of the screws.

cases cancellous bone grafts harvested from the iliac wing were applied between the posterior arch of the atlas and the C2 lamina. In one case a temporary fixation without bone fusion was used as an alternative, which was indicated due to less rigid rotation with a clear trauma etiology, minimal bone changes and subjectively low intraoperative reduction force. In both cases the operation was completed by insertion of a suction drain and suture of the surgical wound in layers**.**

2.4. Postoperative protocol

Patients wore Philadelphia or a custom-made rigid collar for 3 months. After removal of the drain, the patients underwent radiographic and CT examination and were discharged from the hospital. Standard follow-up, including radiographic examination, was scheduled at 6 and 12 weeks, 6 and 12 months and then annually, and CT and MRI follow-up at 4–6 months postoperatively (Fig. 3). In one patient with temporary fixation, implants were removed 6 months postoperatively.

2.5. Clinical and radiographic measurements

In addition to the standard follow-up schedule, all patients were examined also at the end of the study, i.e., at the mean interval of 93.5 months (range, 6–183) postoperatively. Our study focused on evaluation of anatomy of the atlantoaxial complex and the craniocervical junction, the course of the operative procedure, correction of the deformity, bone fusion, the patient's clinical condition and complications.

Clinical evaluation was based on the *Visual Analogue Scale* (VAS) for neck pain. Because NDI has not been adapted for pediatric population, we

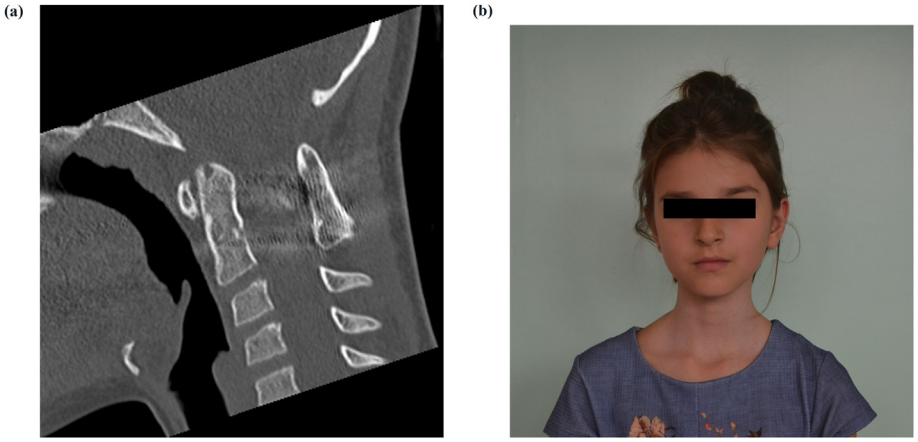

Fig. 3. Imaging studies and a clinical photograph of the same patient 4 months after surgery. CT reconstructions in sagittal plane showing successful bone fusion (a) and final head position (b).

have evaluated functional results of 5 main activities of daily living, which were derived from the NDI. This included evaluation of 5 main points, which relate to the ablity of the child to participate in normal activities of daily living like: 1. ability to dress and take care of personal hygiene, 2. read, 3. play with toys or participate in sport activities, 4. attend school on regular basis and 5. quality of sleep. Each of the 5 parameters were assigned 0 to 5 points (maximum 5 points in 5 categories – 25 points, which is equal to 100%, 1 point is equal to 4%, 0 points means no problems).

This evaluation was done with the patients and their parent(s). As with other treatment of spinal pathologies, parents play an indispensable role in communication with the children patients. Informing the parents about the evaluation of the procedure outcome is part of the consent form, when both parents consent to close cooperation with us.

3. Results

3.1. Radiographic and surgical results

All 12 patients underwent failed non-operative treatment prior to operation. The mean interval between the onset of rotation and operation was 5.7 months (range, 4 days to 22 months). In 6 patients their head was rotated to the left, in 6 patients to the right. The mean anterior atlantodental interval (ADI) was 4.3 mm (range, 2–7). ADI was less than 5 mm in 8 cases and greater in 4 cases. According to the Fielding and Hawkins classification, 3 cases were type I, 6 cases type II and 3 cases type III. With the use of the classification by Ishii et al. we identified 4 cases of type I, 5 cases of type II and 3 cases of type III. Medical history of 9 patients revealed predisposing factors, in 3 patients the etiology was unclear (Table 1). Neurologic deficit was recorded in one case (mild quadriparesis, Frankel Grade D) which improved postoperatively to Frankel Grade E.

The mean operative time was 88.5 min (range, 40–140), the mean blood loss 262.5 ml (range, 50–500) and the mean radiation exposure 47.1 ss (range, 15–128); the mean preoperative mutual C1–C2 rotation was 30° (range, 18–51), the mean post-correction value was 3.3° (range, 2–7); the mean preoperative lateral inclination was 9.4° (range, 5.2–14.3), the mean post-correction value was 1.1° (range, 0.7–3.9); the mean postoperative ADI was 2 mm (range, 1–3). Bone fusion of C1–C2 was achieved in all the relevant cases (N = 11, 100%), temporary fixation was used in one patient. Injuries to the neural structures or the vertebral artery, or other complications were not identified (Table 1).

3.2. Clinical and radiograhic results

The mean VAS value for neck pain was 5.8 preoperatively, 0.4 at one year and 0.1 at the final follow-up. Comparison of VAS values for cervical neck pain and functional results values before surgery and at the end of the study (N = 12) showed major improvement (Table 3). VAS values improved at 6 months, 1 year and 2 years postoperatively, while further follow-ups did not reveal additional improvement. In functional results, considerable improvement was observed at 6 months postoperatively, and no major changes occurred at further follow-ups (Table 3). Analysis of the values of radiological indicators of the atlantoaxial anatomy (ADI, inclination angle, C1–C2 rotation) before surgery and after healing (N = 12) showed extensive improvement (Table 2).

4. Discussion

In 1977, Fielding and Hawkins (1977) published an AARD classification that has been generally accepted and widely used. It divides rotatory dislocation into 4 types. In Type I, the center of rotation is in the region of the dens, there is no atlantodental displacement and the transverse ligament of the atlas is intact. It is typical for transient dislocations that can be well managed by non-operative treatment. In Type II, the center of rotation is in the region of one atlantoaxial joint, the contralateral joint is displaced anteriorly, and the atlantodental interval is less than 5 mm. Type III is characterized by atlantodental interval of more than 5 mm and insufficiency of the transverse atlantal ligament and the alar ligaments, both articular surfaces of C1 are displaced anteriorly, one more than the other. Type IV is associated with asymmetric displacement of both articular surfaces posteriorly and deficiency of the dens (e.g., RA, os odontoideum, aplasia of the dens etc.). Levine and Edwards (1986) added to the above-mentioned classification the fifth, traumatic type of AARD and called it “frank rotatory atlantoaxial dislocation”. Wang et al. (2016) consider type IV of the Fielding classification with dens deficiency and posterior displacement of C1 as another nosological entity, particularly in relation to rheumatoid arthritis. In their cohort of 32 pediatric patients with AARFD, they found 4 cases with dens deficiency and always with anterior displacement of C1. We also take dens deficiency for another nosological entity and therefore we have excluded patients with this pathology from our study group. Pang and Li (Pang, 2010; Pang and Li, 2004, 2005a, 2005b) published a classification on the basis of a 3-position CT examination, i.e., a scan in displacement (with the head in the presenting position), and stress scans in zero position and in rotation of the head toward the other side as tolerated by the patient (scans with the nose pointing straight forward (i.e., neutral), and with the head turned as far to the opposite side as tolerated by the patient). CT scans in all positions are then used to measure rotation of C1 and C2 relative to the vertical, with rotation of C1 and the C1–C2 separation angle (C1 minus C2°) being considered as essential. The authors compare the normal and pathological motion curves. Type I, with the correction of the separation angle during reduction of less than 20%, is characterized by the highest rigidity or locking of the atlantoaxial

Table 2
Classifications, anatomical characteristics and radiological data.

Pt	Sex	Age	Before surgery			After surgery			Classification			rotation direction	duration of rotation (m)
			ADI (mm)	LI (°)	Rotation (°)	ADI (mm)	LI (°)	Rotation (°)	FD	Ishii	FH		
1	M	7	5	8,7	45	1	1,1	2	NO	I	I	RIGHT	22
2	F	7	2	9,1	24	1	0,7	2	NO	I	I	LEFT	6
3	F	16	2	7,1	37	2	3,9	5	NO	I	I	LEFT	1
4	F	8	4	7,6	18	1	1,1	7	YES	II	II	LEFT	12
5	F	5	4	5,2	26	3	2,1	4	NO	I	II	LEFT	0,1
6	F	8	7	9	42	3	1,3	4	YES	II	III	RIGHT	3
7	M	14	4	12,4	35	3	3,3	2	YES	II	II	RIGHT	1
8	M	8	6	14,3	23	2	1,5	2	YES	II	III	RIGHT	8
9	F	7	3	11,4	51	1	1,7	2	YES	III	II	RIGHT	2
10	F	8	4	10,1	38	3	2,4	4	YES	III	II	LEFT	3
11	M	7	4	10,7	22	2	1,2	3	YES	II	III	LEFT	6
12	M	7	6	6,7	18	2	0,7	3	YES	III	II	RIGHT	4
Average:		**8,5**	**4,3**	**9,4**	**31,6**	**2,0**	**1,8**	**3,3**					5,7

Abbreviations: ADI *= atlantodental interval,* ***LI*** *= lateral inclination angle of the C1–C2 lateral mass,* ***FD*** *= facet deformity,* ***Ishii*** *=* (Ishii et al., 2011)*,* ***FH*** *=* (Fielding and Hawkins, 1977).

Table 3
Clinical outcomes.

Pt	VAS					functional results questionnaire				
	pre-op	6M	12M	24M	Out	pre-op	6M	12M	24M	Out
1	5	2	1	0	0	44%	20%	4%	8%	8%
2	6	0	0	0	0	16%	0%	0%	0%	0%
3	9	1	1	1	1	84%	12%	12%	12%	12%
4	0	0	0	0	0	0%	0%	0%	0%	0%
5	8	0	0	0	0	72%	0%	0%	0%	0%
6	4	0	0	0	0	12%	0%	0%	0%	0%
7	7	6	3	0	0	60%	36%	4%	0%	0%
8	10	0	0	0	0	84%	0%	0%	0%	0%
9	6	0	0	0	0	64%	0%	0%	0%	0%
10	8	0	0	0	0	80%	0%	0%	0%	0%
11	2	0	0	0	0	48%	0%	0%	0%	0%
12	5	0	x	x	0	64%	0%	x	x	0%
Average: 5,8		**0,8**	**0,5**	**0,1**	**0,1**	**52,3%**	**6,2%**	**1,8%**	**1,8%**	**1,8%**

Abbreviations: VAS *- Visual Analogue Scale,* ***pre-op*** *- preoperative values,* ***6M*** - 6-month *follow-up values,* ***12M*** - 12-months *follow-up values,* ***24M*** **-** 24-months *follow-up values,* ***Out*** *- values at the time of the end of the study.*

complex. Type II, with the separation angle of more than 20%, shows "pathologic stickiness" without crossover of C1 on C2. Type III is least severe, with a typical crossover of C1 on C2 during rotation of the head to the other side. The classification of 2005 is too complicated in terms of its practical use and we do not use it at our center. Ishii et al. (2011) proposed a classification according to the degree of facet deformity and inclination of the atlas. Type I is characterized by intact facets, zero inclination, and by the presence of rotation only. Type II is associated with facet deformity and inclination of less than 20°, and Type III also with facet deformity, but with inclination of more than 20°. Goel and Shah (2011) divide facetal locking into two types: rotatory and translatory. Rotatory locking has been further subdivided into partial, when the facet of atlas of only one side is dislocated anteriorly, and complete, when the facets of atlas on both sides are dislocated, one anteriorly and the other posteriorly. Translatory dislocation has been described as a condition when facets of atlas on both sides are dislocated anteriorly, which results in a fixed flexion neck deformity.

Wang et al. (2016) claim that AARD is rare in adults and markedly dominates in pediatric patients, which may reflect the effect of immaturity of the pediatric skeleton, namely the shallower and more horizontally oriented joint surface, the relative elasticity of the ligaments, the not yet fully developed neck muscles, and a relatively large head in children (Goel and Laheri, 1994). The above-mentioned characteristics support the concept of a more frequent development of atlantoaxial instability rather than fixed torticollis; nevertheless, due to their active growth, the bone and soft tissue structures of the pediatric spine show tendency to remodeling. The same remodeling processes may result also from infection in the region of the neck, or minor trauma or surgical procedure in this location. Atlantoaxial joints are fixed by rapid remodeling in the pathological position, persisting also after subsidence of the initial deficiency. In adult patients, the remodeling process is much longer and the initial deficiency as a rule subsides faster. In rare cases, often associated with a high-energy trauma, healing may be delayed and result in development of AARF or AARFD. Wang et al. (2016) present yet another theory of onset of pediatric AARF, i.e., that fast healing and fibrosis of joint capsules and ligaments cause contracture of joint capsules in children, particularly when the facets are in an extreme, displaced or locked position. Štulík et al. (2019) reported surprisingly high number of adult patients in their previous study a (46.7%), which they explain by a higher concentration of adult patients with a craniocervical junction disorder at their department.

The repeatedly described predisposing factors for onset of AARF include nasopharyngeal infection, upper respiratory tract infection, trauma, head and neck surgery (Hill et al., 2021; Kinon et al., 2016; Powell et al., 2017; Sae-Huang et al., 2020). Wang et al. (2016) report 50% of cases with a known etiology, with the highest share (25%) of moderately severe injuries. Beier et al. (2012) described association with trauma in 50% of their cases. Akbay et al. (2014) identified initial trauma in up to 92% of cases. Ishii et al. (2012) report upper respiratory tract infection as the most frequent etiology, in 71% of the patients. In our study group, we found initial injury in 33,3% and evident infection in 25% of cases; in the remaining cases etiology was unclear.

Fielding and Hawkins (1977), as well as Pang (Pang, 2010; Pang and Li, 2004, 2005a, 2005b) reported higher incidence rate of the disorder in men; Beier et al. (2012) identified in a large cohort the male:female ratio of 3:2. By contrast, according to Wang (Wang et al., 2016) the incidence rate was twice higher in women than in men, Akbay et al. (2014) reported 3 women per 1 man and Ishii et al. (2011) even 100% of females. Our study group comprised 58% of females.

The common AARD types I and II are rarely associated with neurologic deficit, including myelopathy. According to Wang et al. (2016), the onset of myelopathy is associated especially with AARFD and AAD (25% cases in their study). We recorded neurologic deficit in 8.3% of our study group.

Non-operative therapy by manipulation, external fixation or traction is mostly successful in functional and less severe types of the disorder (Akbay et al., 2014; Barcelos et al., 2014; Beier et al., 2012; Goel, 2019; Hill et al., 2021; Ishii et al., 2012; Kinon et al., 2016; Pang, 2010; Pang and Li, 2004, 2005a, 2005b; Sae-Huang et al., 2020). About 35% of authors use cervical collar or brace in combination with painkillers and myorelaxants; 30% of authors favor halter traction (Sae-Huang et al., 2020). Beier et al. (2012) evaluated 40 children with AARS, treated initially non-operatively, of these only one patient required operative treatment due to a severe anatomical stenosis of the spinal canal and another two patients underwent surgery after a failed conservative therapy, including halo fixation. Ishii et al. (2012) described therapeutic algorithm for a chronic AARF and recommended, in mobile segments of the spine, closed manipulation followed by halo fixation for 2–4 months, the so-called remodeling therapy. Complete remodeling was observed in 10 of 12 patients, partial remodeling in the remaining 2 cases. Operative treatment was indicated only after failure of non-operative treatment, and as a primary procedure it was recommended only in patients with C1–C2 bone fusion, dens deficiency or congenital anomalies. Akbay et al. (2014) prefer manual reduction under general anesthesia and application of brace for 4 weeks; only 1 of 12 pediatric patients required subsequent atlantoaxial fixation and fusion after repeated failures of non-operative treatment. In another study, Ishii et al. (2011) reported in 71% of patients a normal cervical range of motion 2 weeks after removal of the halo vest and no recurrence at the final follow-up.

In patients with developed anatomical changes, non-operative treatment usually fails and surgical correction and stabilization should be considered (Goel, 2019; Ishii et al., 2012; Pang, 2010; Pang and Li, 2004, 2005a, 2005b; Štulík et al., 2019; Wang et al., 2016). Landi et al. (2012) in a group of 9 children with AARF treated non-operatively, found lesions

of alar ligaments, the transverse atlantal ligament and capsular ligaments on MRI. They subsequently recommended operative treatment. Goel and Shah (2011), in their study of 14 patients, also recommend operative treatment in patients with AARF who fail to respond to non-operative therapy for two months. Wang et al. (2016) published a therapeutic guideline for treatment of atlantoaxial rotatory deformity in children. In the presence of torticollis lasting for more than 6 weeks, they perform radiographic examination of the cervical spine in the lateral view. Patients with an atlantodental interval (ADI) of less than 5 mm are treated with traction for 6 weeks and in case of its failure, posterior fixation of C1–C2 is indicated. In patients with ADI of more than 5 mm they use skeletal traction under general anesthesia and subsequently, after successful reduction, again posterior fixation of C1–C2. In patients with irreducible dislocation, this procedure is preceded by transoral release of both atlantoaxial joints. Pang (Pang, 2010; Pang and Li, 2004, 2005a, 2005b) recommends treating irreducible dislocations with fixation and fusion in situ with the best possible correction. Wang et al. (2016) consider in situ fixation as esthetically unacceptable and point out a satisfactory correction in all their patients. Our center also prefers correction in all patients since in situ fixation, particularly in children, aggravates the gradual development of face asymmetry and deformity the cervical spine.

Venkatesan et al. (2012) described 2 adult patients with purely traumatic atlantoaxial rotatory subluxation (TAARS) successfully managed by halo traction for 4–5 days, followed by external fixation in a rigid collar applied for 8 weeks. By contrast, Guo et al. (2015) prefer to use in children with TAARS and a defect of the transverse atlantal ligament reduction and atlantoaxial fixation with fusion as the initial treatment.

In a majority of cases, Harms/Goel or Magerl technique, in young children tension band wiring techniques or a combination thereof, are used for atlantoaxial fixation and fusion [(Akbay et al., 2014; Guo et al., 2015) (Pang, 2010; Pang and Li, 2004, 2005a, 2005b), (Štulík et al., 2019; Wang et al., 2016)]. Wang et al. (2016) favor fixation of C1–C2 according to Harms/Goel and achieved successful fusion in 96.6% of cases. In 9.4% of their patients, fixation had to be extended to involve the occipital bone. Guo et al. (2015) published 5 pediatric patients with acute TAARS and a modified application of 3.5-mm screws into the lateral masses of C1 through the posterior arch with subsequent atlantoaxial fixation and fusion. They encountered no complications and C1–C2 bone fusion was achieved in all cases. Goel (2021) recommends, for the sake of a better orientation and assessment of the position of the atlantoaxial joint, resection of the C2 ganglion and filling of the joint space with bone grafts from the iliac crest. Under favorable anatomical conditions he prefers transarticular insertion of screws, the so-called double insurance technique. In our study group we have confirmed a high success rate of bone fusion (11/12, 11 = 100%) without a risk of AARD recurrence. In one patient with traumatic etiology, we successfully used a temporary atlantoaxial fixation.

5. Conclusion

After failure of non-operative therapy, surgical correction of AARD is an appropriate method ensuring a favorable position of the atlantoaxial complex, improving the patient's clinical condition and preventing development of asymmetric face in children. Full reduction and fixation of C1–C2 by Harms/Goel technique with bone fusion is for us the method of choice in children and adolescents with AARD. In patients with traumatic etiology of AARD and simple reduction we recommend to consider temporary fixation.

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Authors' contributions

Jan Štulík and Zdeněk Klézl contributed to the study conception and design. Michaela Rybárová and Michal Barna collected radiographic and clinical data. All authors contributed to the analysis and interpretation of acquired data. Jan Štulík obtained administrative and technical support necessary for successful completion of the project. Jan Štulík and Michaela Rybárová were responsible for drafting of the first version of the manuscript. Finally, all authors read, reviewed and approved the final version of the manuscript.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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