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CERVICAL SPINE

Atlantoaxial Rotatory Fixed Dislocation

Report on a Series of 32 Pediatric Cases

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Study Design. Retrospective case series of atlantoaxial rotatory fixed dislocation (AARFD).

Objective. To describe clinical features and the surgical treatment of AARFD.

Summary of Background Data. The classification and treatment strategy for atlantoaxial rotatory fixation (AARF) were previously described and remained controversial. AARF concomitant with atlantoaxial dislocation has different clinical features and treatment strategy with the most AARF. Due to deficiency of the transverse ligament or odontoid, the atlantoaxial remains unstable even after the torticollis relieved or cured. Because of the rarity, treatment strategy for this special condition has not been specialized and fully explored in the literatures.

Methods. Thirty-two children with AARFD (sustained torticollis more than 6 weeks and atlanto-dental internal more than 5 mm) were retrospectively reviewed. Treatment methodology, pearls, and pitfalls of the treatment were discussed.

Results. Thirty-two cases had sustained torticollis for an average of 5.7 months. ADI of them ranged from 8 to 22 mm, with a mean of 11.3 mm. Eight cases presented with signs and symptoms of spinal cord dysfunction. All 32 cases underwent surgery and had no spinal cord or vertebral artery injury. The surgery included posterior reduction and fusion (reducible dislocation and torticollis, 16 cases), and transoral release followed by posterior reduction and fusion (irreducible dislocation and torticollis, 16 cases). The average follow-up time was 42 months. Solid fusion and torticollis healing were achieved in 31 patients (96.9%) as detected radiologically. Two cases (6.3%, 2/32) suffered complications (cerebrospinal fluid leakage and recurred torticollis followed by revision).

Conclusion. AARFD had distinct clinical features relative to common presentations of AARF. Because of deficiency of the transverse ligament or odontoid and subsequent atlantoaxial dislocation, surgical treatments are applied for this condition, including transoral release and posterior C1–2 reduction and fusion. AARFD cases were successfully managed surgically without preoperative traction, with few complications seen.

Key words: atlantoaxial dislocation, atlantoaxial fusion, atlantoaxial rotatory fixation, atlantoaxial rotatory fixed dislocation, posterior reduction, surgery, torticollis, transoral release, transverse ligament deficiency.

Level of Evidence: 4

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Atlantoaxial rotatory fixation (AARF) occurs predominantly in children. Patients typically present with torticollis, neck pain, and stiffness. Clinically, children presenting with AARF most commonly exhibit the "Cock-robin" position,[1] in which the chin is turned toward one side and the neck is laterally flexed in the opposite direction. It has been postulated that torticollis causing AARF is due to the spread of inflammation from anastomotic lymphatic and venous channels of the pharynx, together with postinflammatory ligament loosening, and paravertebral muscle spasms.[2,3]

Fielding and Hawkins[4] first described AARF as a fixed clinical torticollis that developed as a result of atlantoaxial dysfunction. They further separated AARF into four different types based on the axial images of the C1 and C2 vertebrae.[4] In this widely accepted classification, Types I and II have a normal atlantoaxial interval (less than 5 mm). However, Type III displays bilateral anterior facet displacement with an atlantoaxial interval greater than 5 mm due to a deficiency of the transverse ligament. Type IV is an unusual type, with a deficient odontoid and posterior dislocation of the atlas. Unlike Types I and II which are rotatory displacements around an intact odontoid articulation, Types III and IV have planar instability which allows translation between the atlas and the axis particularly in the sagittal plane and can result in narrowing of the space available for the cord at the atlantoaxial level. In Type I

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or II AARF, the atlantoaxial stabilizing structures, particularly the transverse ligament or the odontoid, remain intact. In Type III, the intrinsic atlantoaxial structures (including the transverse ligament and alar ligaments) have been affected by the pathological process and atlantoaxial stability is thus compromised. Type IV cases with a deficient odontoid also result in atlantoaxial instability. These two latter conditions present as torticollis with an unstable atlantoaxial joint in the sagittal plane and have very different biomechanical features compared to Types I and II. Collectively, we refer to clinical torticollis secondary to atlantoaxial rotation, together with an unstable atlantoaxial joint as determined by the atlantodental interval, as atlantoaxial rotatory fixed dislocations (AARFD) in this series. In the following study, AARFD is defined as a clinical torticollis secondary to atlantoaxial rotation with an unstable atlantoaxial joint as determined by the widened atlantodental interval.

The majority of cases of AARF can be treated successfully by nonoperative modalities.[1,5–7] Only recurrent cases require surgery.[1,8–10] As a result of the deficiency of the transverse ligament or odontoid, Types III and IV dislocations (AARFD) remain unstable even after the clinical torticollis has resolved. To date, this condition had not been fully investigated in the literature, and thus the treatment strategy remains unclear. In this report, AARFD were specifically chosen in order to clearly evaluate the clinical presentation, treatment algorithm, and surgical outcomes. We reviewed a consecutive series of pediatric AARFD cases, which, to our knowledge, represents the largest case series to date for this condition.

MATERIAL AND METHODS

Clinical Materials

This investigation was a retrospective analysis of a prospectively collected database. All cases were performed at a single institution. Institutional board approval was obtained prior to initiating this study. Radiographic findings and clinical notes were reviewed from 2001 until March 2012.

AARF was clinically diagnosed by the presence of neck pain, stiffness, and sustained torticollis exhibiting the "Cock-robin" position.[1,4] Following the clinical algorithm of atlantoaxial rotatory deformity (Figure 1), atlantoaxial dislocation (AAD) was then further assessed according to the lateral radiograph and with three-plane computed tomography (CT) as seen in Figure 2 (C–E). Dynamic radiographs (flexion and extension) were utilized to identify evidence of atlantoaxial instability, defined as an atlantodental distance greater than 5 mm in children (<18 years). For patients with an incomplete odontoid process, the same criteria were adopted by measuring the distance between the inferior rim of the C1 anterior arch and the remaining attached part of the odontoid or C2 anterior-superior edge.[11] Atlantodental rotation was evaluated on axial CT because measurements on the anteroposterior plain radiograph were deemed unreliable for the torticollis cases.[12]

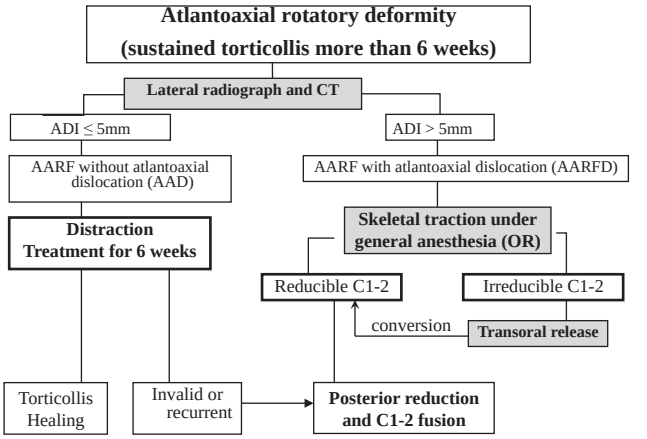

Figure 1. Algorithm for atlantoaxial rotatory deformity. AAD indicates atlantoaxial dislocation; AARF, atlantoaxial rotatory fixation; AARFD, atlantoaxial rotatory fixed dislocation; ADI, atlantodental internal; CT, computed tomography; OR, operating room.

Patients with acute or subacute AARF (torticollis less than 6 weeks) were excluded. AARF cases without AAD were excluded from this study (Figure 1, left branch).

Surgical Treatment

All surgical procedures were performed by the same surgeon. Prior to each procedure, the patient was laid in the supine position. Skull traction (one-sixth of the body weight) was applied after the patient was placed under general anesthesia and administered muscle relaxants.[13,14] Reducible deformity was diagnosed intraoperatively by (a) a fully corrected torticollis and (b) normal ADI and satisfactory atlantoaxial alignment as determined on fluoroscopy. Otherwise, irreducible deformity was determined. Sixteen cases were deemed to have an irreducible AARFD and underwent transoral atlantoaxial release first in supine position, using the technique previously described.[13] The bilateral longus colli and longus capitis muscles, together with the anterior longitudinal ligament, were dissected first. Then, the capsule of lateral mass joints was dissected to expose the joints. We released the articular adhesions with a curette to unlock and reduce rotatory deformity (Figure 3). Following this, the posterior approach with an atlantoaxial fusion was performed immediately.

The remaining 16 reducible cases underwent a single posterior surgery, as the AARFD was deemed reducible. The patient was put into the prone position, maintaining cranial traction throughout the procedure. A posterior midline incision was made to expose the C1 posterior arch and the C2 lamina. The vessels and C2 nerve roots were retracted to expose the entry points of the C1 lateral mass screws (LMS) and C2 pedicle screws. The entry points were created with a burr. C1 lateral mass and C2 pedicle were prepared with a 2.5-mm hand drill before screw placement.[15] The C1 posterior tubercle and C2 spinous process were used as landmarks for reduction (Figure 4A). Cranial traction and transoral release (if necessary) was able to partially reduce the rotatory deformity. Two reconstruction

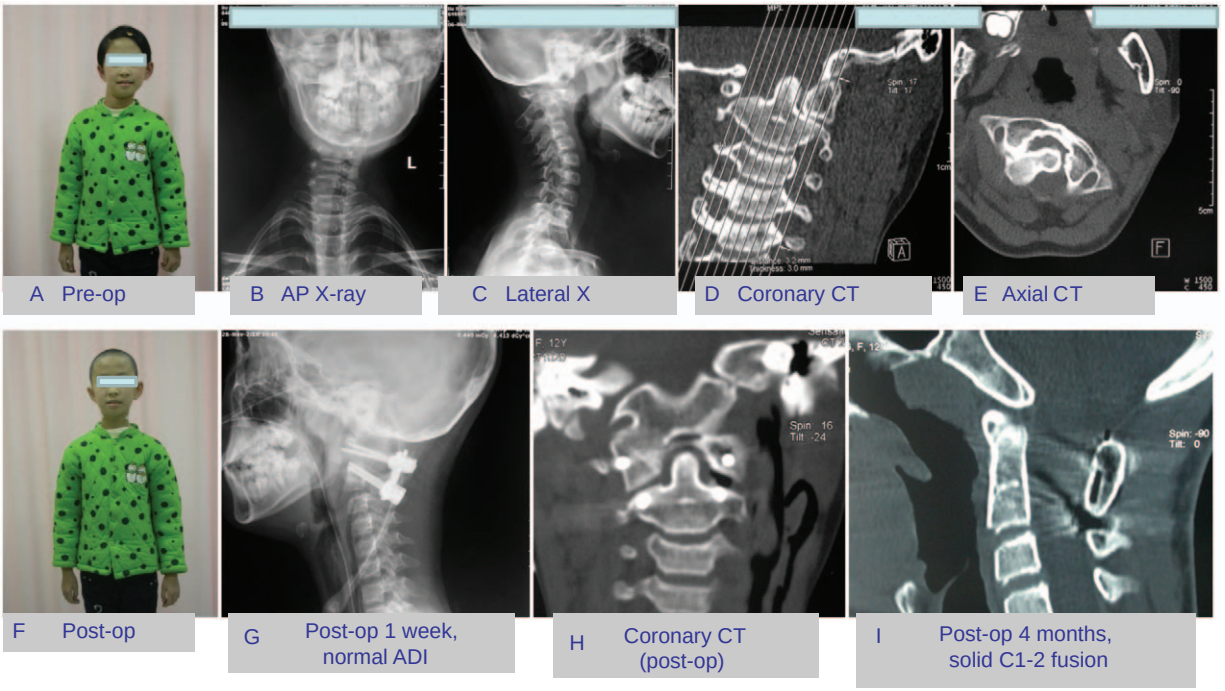

Figure 2. A, A 12-year-old female sustained torticollis (after a thyroid adenoma excision); **B**, the preoperative radiography (AP); **C,** the preoperative radiography (lateral) showed abnormal ADI was 12 mm; **D,** the coronary CT showed severe torticollis; **E,** the axial CT showed rotatory fixed C1; **F,** after the transoral C1–2 release and posterior C1–2 reduction, the torticollis was corrected; **G,** the postoperative radiography (lateral) showed the ADI was 1 mm; **H,** the coronary CT showed torticollis was completely corrected and the screws were placed accurately; **I**, after 4 months, solid fusion was achieved between C1 and C2. ADI indicates atlanto-dental internal; CT, computed tomography.

plates were precontoured and placed between the C1 LMS and C2 PS (Figure 4B). Locking caps were installed on the screws and used to tighten the plates, which further reduced the rotatory deformity and AAD (Figure 4C). The plates were subsequently locked to retain reduction. At the end of the procedure, the C1 arch, C2 lamina, and spinous process were decorticated with a high-speed burr. Morselized cancellous grafts (15 grams) harvested from the posterior iliac crest were bridged between the C1 arch and C2 lamina. The wound was closed following placement of a deep drain.

In three patients (case 16, 18, and 31), occiput-C2 fixation was performed due to failure of the C1 LMS placement. However, in these cases the morselized cancellous grafts were placed between C1 and C2. After 4 months, solid fusion was confirmed by reconstructive CT. The hardware was then removed in order to release the occipital-atlanto joint.

Radiographic Evaluation

All patients had postoperative x-rays and reconstructive CT scans of the cervical spine obtained at 3 to 5 days and 4 months after the surgery (Figure 2G–I). X-ray examination was repeated at 12 months and annually thereafter. Screw position and atlantoaxial alignment were analyzed using the postoperative reconstructive CT (Figure 2H). Fusion was confirmed by postoperative reconstructive CT when there was definite osseous union between the C1 and C2 lamina (Figure 2I).

RESULTS

From 2001 to March 2012, 32 pediatric patients (under 18 years) were diagnosed with AARFD at our institution. The series included 11 males and 21 females, and the mean age was 11.3 years (range: 6–17).

Clinical Features

All 32 cases had sustained torticollis for an average of 5.7 months (1.5–12 months) prior to treatment. Nineteen of the torticollis cases displayed right chin direction, while 13 cases

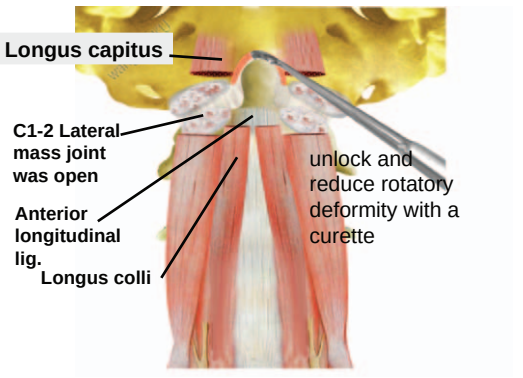

Figure 3. A sketch map of the transoral atlantoaxial release: the bilateral longus colli, longus capitis, and anterior longitudinal ligament were dissected first. Then, the capsule of lateral mass joints was dissected to expose the joints. The articular adhesions were released with a curette to unlock and reduce rotatory deformity.

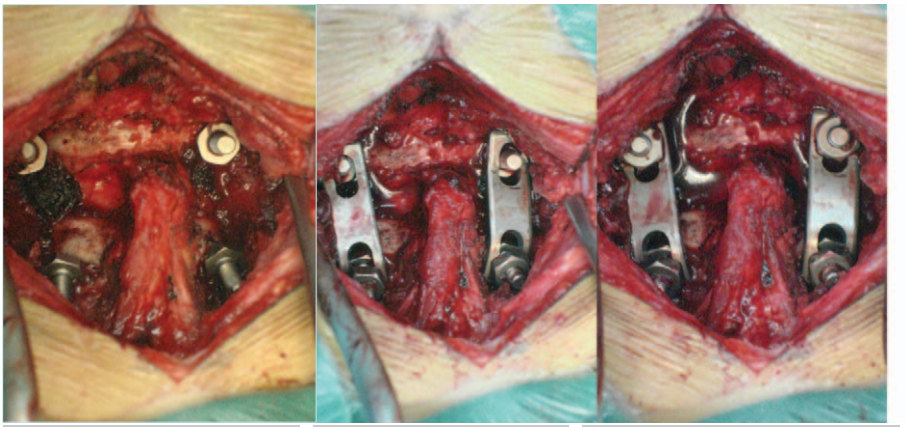

Figure 4. The reduction steps during the posterior procedure (the atlas had left rotation). **A,** After screw placement; **B,** Plates were placed, partly reduced; **C,** Locking the screws with plates completely corrected the rotation and dislocation between C1 and C2.

displayed a left direction. ADI ranged from 8 to 16 mm, with a mean of 10.3 mm. Twenty-eight cases had an intact odontoid with more than 5-mm anterior displacement of the atlas (matching Fielding's Type III), while four had evidence of a deficient odontoid (os odontoideum) and anterior displacement of the atlas (Figure 5). All four cases that presented with anterior displacement were unable to be classified according to Fielding's system, as Type IV has rotatory fixation with posterior displacement of the atlas. Sixteen cases had valid predisposing factors: mild cervical trauma (8 cases), cervical surgery (4), and cervical or nasopharyngeal infection (4). All cases underwent unsuccessful conservative treatments prior to surgery: collar application (12 cases), cervical belt traction (9), skeletal traction (8), and bracing (2). Eight cases presented with signs and symptoms of spinal cord dysfunction (Japanese Orthopaedic Association [JOA] score from 7 to 15, averaged 11.9). Among the eight cases, two had preoperative dyspnea (case 17 and 31). One case had a previously failed atlantoaxial fusion (case 8). Clinical features are summarized in Table 1.

Surgical Results

All 32 cases were treated surgically. No spinal cord or vertebral artery injuries were recognized. The surgery included posterior reduction and fusion (reducible dislocations with torticollis, 16 cases), and transoral release followed by posterior reduction and fusion (irreducible dislocations with torticollis, 16 cases). Atlantoaxial fixation was performed in 29 cases during the posterior procedure. In three cases (case 16, 18, and 31), "salvage" occiput-C2 fixation was performed because the C1 LMS placement was unsuccessful. The mean operation time was 135 minutes (from 90 to 200 minutes). The estimated blood loss ranged from 50 to 300 ml with a mean of 137 ml.

Follow-up Results

The mean postoperative follow-up interval was 42 months (25–120 months). Complete atlantoaxial reduction, satisfactory torticollis correction (Figures 2A, B, F and Fig. 6), and solid atlantoaxial fusion were achieved in 31 patients (96.9%). For the eight cases with preoperative myelopathy, the mean JOA score improved from 11.9 to 16.0.

Two of the 32 cases (6.3%) suffered complications. Case 9 had immediate postoperative resolution of torticollis; however, the torticollis recurred 1 month after the surgery. This patient underwent successful revision (transoral release combined with posterior C1–2 reduction and fusion). Case 16 had CSF leakage during the transoral release and was treated successfully by lumbar CSF drainage.

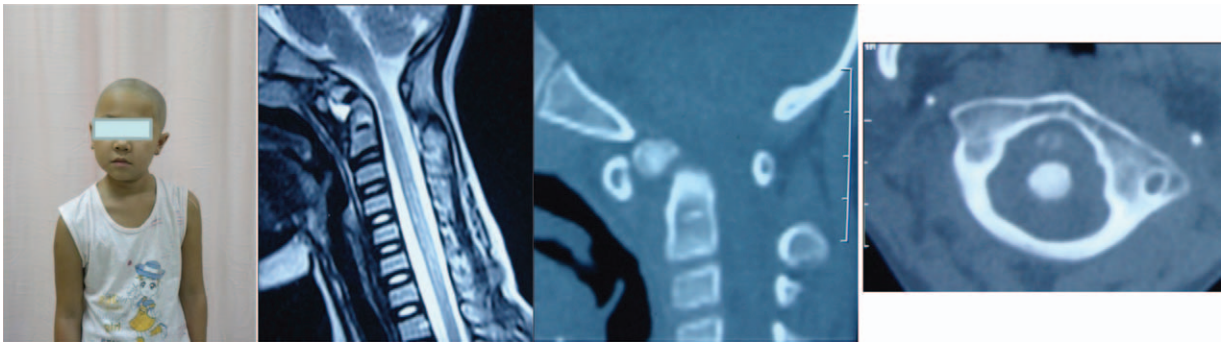

Figure 5. A 9-year-old girl sustained torticollis and was diagnosed with AARFD. She had deficient odontoid and anterior displacement (modified ADI = 14 mm). AARFD indicates atlantoaxial rotatory fixed dislocation; ADI, atlanto-dental internal.

TABLE 1. Clinical Data of 32 Cases

No	Age	Sex	Torticollis Direction	Predisposing Factors	Duration (Months)	Myelopathy (JOA score)	Previous Treatments	Operations	Blood Loss	Time (minutes)
1	17	M	Left	Upper respiratory tract infection	2	N	Failed skull traction (4 kg for 5 days)	C1–2 transarticular fixation	200	100
2	14	F	Left	NA	7	+ (14)	Collar	TOR& C1–2 transarticular	200	200
3	6	F	Left	Fall to the ground	1.5	N	Collar	C1–2 pedicle fixation and fusion	50	115
4	11	F	Right	NA	5	N	Skull traction (3 weeks) and torticollis recurrent after 1 month	TOR& C1–2 pedicle fixation and fusion	300	140
5	12	F	Right	NA	7	+ (13)	Collar	TOR& C1–2 pedicle fixation and fusion	200	185
6	11	F	Right	Clavicle fracture surgery	6	N	Failed cervical belt traction for 3d	TOR& C1–2 pedicle fixation and fusion	200	170
7	10	F	Right	Cervical cyst resection	10	+ (14)	Collar	TOR& C1–2 pedicle fixation and fusion	200	180
8	7	F	Right	Upper respiratory tract infection	12	N	Failed skull traction (6 weeks) Failed C1–2 fusion	TOR& C1–2 pedicle fixation and fusion	200	200
9	9	M	Left	NA	8	N	Cervical belt traction for 50 days and torticollis recurrent after 4 weeks	C1–2 pedicle fixation and fusion	200	140
10	12	F	Right	Cervical sebaceous cyst resection	11	N	Cervical belt traction for 2 weeks	TOR& C1–2 pedicle fixation and fusion	200	160
11	11	M	Right	Fall to the ground	4	N	Collar	TOR& C1–2 pedicle fixation and fusion	200	120
12	15	M	Right	NA	4	N	Collar	C1–2 pedicle fixation and fusion	100	130
13	10	M	Right	Neck sprain by a boy	1.5	N	Collar	C1–2 pedicle fixation and fusion	100	110
14	17	M	Right	NA	6	N	Skull traction (2 weeks)	C1–2 pedicle fixation and fusion	100	165
15	12	F	Right	Thyroid cyst resection	8	N	Cervical belt traction for 2 weeks	TOR& C1–2 pedicle fixation and fusion	100	140
16	11	F	Left	with Down syndrome	8	+ (11)	Collar	TOR& occiput-C2 fixation	200	200
17	15	F	Right	Fall (3 meters)	1.5	+ (7) Dyspnea	Collar	C1–2 pedicle fixation	100	110
18	9	M	Left	NA	12	N	Collar	TOR& occiput-C2 fixation	150	160
19	7	F	Left	Fall from a small stool	1.5	N	Cervical brace using for 1 month	C1–2 pedicle fixation	80	100
20	9	F	Right	Cervical herpes zoster	8	N	Skull traction (4 kg, 2 months)	TOR& C1–2 pedicle fixation	150	166
21	12	M	Left	Neck injury by a boy's blow	1.5	N	Collar	C1–2 pedicle fixation	200	100
22	12	F	Left	NA	6	N	Cervical belt traction (4 kg, 3 weeks)	C1–2 pedicle fixation	50	90
23	15	F	Right	NA	3	+ (12)	Skull traction for 1 week	C1–2 pedicle fixation	50	110
24	14	M	Right	Fall to the ground	8	N	Collar	C1–2 pedicle fixation	100	90
25	10	M	Left	NA	7	N	Skull traction (3.5 kg, 1 week)	C1–2 pedicle fixation	50	90
26	8	F	Right	NA	6	N	Cervical belt traction (3 kg, 4 weeks)	C1–2 pedicle fixation	50	120
27	11	M	Left	NA	18	N	Cervical belt traction for 2 weeks	TOR& C1–2 pedicle fixation	100	200
28	15	F	Right	NA	5	N	Collar (2 months)	C1–2 pedicle fixation	150	90
29	9	F	Left	NA	9	N	Cervical belt traction for 2 weeks	C1–2 pedicle fixation	50	90
30	13	F	Right	Nasopharyngeal infection	6	N	Cervical belt traction for 3 weeks (failed)	C1–2 pedicle fixation	100	90
31	9	F	Left	Fall to the ground	1.5	+ (9) Dyspnea	Skull traction (1 kg, 2 weeks), Brace	Occiput-C2 fixation	100	120
32	8	F	Right	NA	3	+ (15)	Massage	TOR& C1–2 pedicle fixation	150	140

JOA indicates Japanese Orthopaedic Association; F, female; M, male; N, negative; NA, not applicable; TOR, transoral release.

Figure 6. A 7-year-old girl sustained torticollis (4 days after upper respiratory infection) and was diagnosed with AARFD. She underwent transoral C1–2 release and posterior C1–2 reduction and fusion. AARFD indicates atlantoaxial rotatory fixed dislocation.

DISCUSSION

Despite being commonly used, there are several limitations with Fielding's classification for AARF. Firstly, Fielding's classification does not make a clear distinction between rotatory deformity and atlantoaxial instability, thereby assuming the two occurrences are equivocal. The common Type I and Type II AARF in pediatric patients is typically linked with simple torticollis from exogenous inflammation, cervical ligament or joint involvement, and muscle spasm.[1,3,16] The atlantoaxial joint remains stable with an unaffected spinal cord. In AARFD cases, however, the transverse ligament or odontoid is deficient, thereby causing atlantoaxial instability and often resulting in clinical myelopathy. Secondarily, in Fielding's initial report,[4] only one 68-year-old woman with rheumatoid arthritis was classified as Type IV. Notably, atlantoaxial displacement in rheumatoid arthritis is a distinctly different entity from AARF or AARFD. Thirdly, Fielding's Type IV classification does not consider anterior displacement, as most AARFD cases with deficient odontoid have anterior displacement (all four cases in this report). Rotatory deformity itself refers only to a symptom or a sign, but is in reality a pathological entity with multiple etiologies. Fielding's classification is a radiological classification encompassing different etiologies which may lead to diagnostic confusion. Therefore, the purpose of this series was to test a diagnosis and treatment algorithm of atlantoaxial instability with rotatory deformity, and refer to it as "AARFD," as indicated in this report.

AARF in adults is rare, and has been reported mostly in a pediatric population.[17] Similarly, in our study, AARFD was noted in only one adult patient, during the same consecutive time period, while 32 pediatric cases were observed. In the literature, this predominance in children has been attributed to the shallower and more horizontally oriented joint surface in the atlantoaxial joint, the relative elasticity of the ligaments, the incompletely developed neck muscles, and a relatively larger head.[8,18] From a theoretical perspective, however, these features more specifically predispose children to atlantoaxial instability rather than a fixed torticollis. The pediatric cervical spine and related muscles and ligaments are actively growing, with a rapid remodeling process occurring, and abundant adaptive capacity.[19] Inflammatory stimulation, due to infection or slight trauma or surgery, can also cause painful contractions of neck muscles that arise from the cervical vertebrae as well as the sternocleidomastoid and trapezius muscle.[16,20] In the setting of atlantoaxial inflammation, postoperative or post-trauma muscle spasms in the pediatric cervical spine can result in a self-protect posture (to relieve pain) and initiate the torticollis. The rapid remodeling process inherent to the pediatric cervical spine, related muscles and ligaments readily adapt to the rotatory deformity. The pediatric atlantoaxial joints fix to the maladaptive posture due to the rapid remodeling process, and cannot be corrected even after the inflammation resolves. In an adult, the remodeling process takes much longer than the process of inflammatory resolution and does not lead to a rotatory fixation. In very rare circumstances, the neck inflammation process is prolonged and results in adult AARF or AARFD. In the literature, adult onset AARF is most commonly caused by high-energy trauma.[17] Presumably, a high-energy trauma resulting in a longer reparative inflammatory process has the same effect as a shorter remodeling process in children. In addition, another explanation for children's rotatory fixation is that rapid healing and fibrosis of the capsule and ligaments in a child causing capsular contracture, especially if the facets are in an extreme, dislocated, or locked position. However, these theories represent only hypotheses without direct supported objective evidence.

In Fielding and Hawkins's[4] and Pang's[1] AARF reports, there were more male patients than female. However, the current AARFD report displays a predominance of females (21 females to 11 males). With regard to clinical presentation, AARFD had a greater risk for myelopathy due to AAD. In this report, myelopathy was observed in 25% of patients. Several predisposing factors for AARF have been reported, including nasopharyngeal infection, upper respiratory tract infection, trauma, and surgery of head and neck.[1,4,21–23] Some AARF cases can occur, without an obvious predisposing cause.[23] In this report, 50% (16/32) of AARFD cases had valid predisposing factors including mild cervical trauma (8 cases), cervical surgery (4), and

cervical or nasopharyngeal infection (4). These predisposing factors are common to other published studies.

It is important to note that in AARFD there is both fixed rotation and instability between the two vertebrae. Skull traction trials or closed manipulation followed by use of a halo-vest may correct the torticollis. However, in our opinion, the atlantoaxial joint remains unstable even after the torticollis has been relieved or cured because of deficiency of the transverse ligament or odontoid. In an MRI study of AARF cases with atlantoaxial subluxation (correspond to AARFD in the current paper), Landi *et al*[6] found abnormal lesions affected the alar, capsular, and transverse ligaments. Thereafter, atlantoaxial reduction and fusion were recommended for AARFD, as described in the current report. This surgical indication has been supported by the literature. In a report of 14 cases by Goel and Shah,[8] the AARF was named "facet locking," and surgery was recommended for those who had failed 2 months of conservative treatment. The third case in Goel and Shah's[8] report can be diagnosed as AARFD by our criteria. The delay of surgical intervention may result in local osseous fusion.[24] In Ishii *et al*'s[25] report, six AARFD cases (chronic Fielding's Type III) underwent awakened skull traction, and all of them had failed reduction or sustained a recurrence. All six patients ultimately underwent posterior fusion. In Ishii *et al*'s[22] algorithm, surgery was indicated for chronic AARF, autofused but uncorrected, and cases with a deficient dens. Moreover, patients were subjected to a difficult course of prolonged skeletal traction while awake or a halo-vest period. In this report, we used heavy skull traction under general anesthesia to correct the torticollis and identify the reducibility. If fully corrected torticollis and satisfactory atlantoaxial alignment were acquired, the patients underwent posterior C1–2 reduction and fixation. If not, the transoral atlantoaxial release was performed in the supine position. The transoral release converted the irreducible deformity to reducible AARFD, followed by posterior reduction and fixation. The procedure in this report abandoned painful awakened skull traction or halo-vest usage, and achieved satisfactory results.

For the irreducible patients, Pang[1] recommended fusion *in situ* having obtaining the best achievable alignment. However, atlantoaxial fusions fixed *in situ* implies uncorrected torticollis. For young patients, the uncorrected torticollis was particularly unacceptable because of the appearance. In the current report, half the cases of AARFD were irreducible. Satisfactory reduction was achieved by performed a transoral atlantoaxial release. Notably, satisfactory reduction was achieved for all irreducible patients.

CONCLUSION

In the current case series, pediatric patients presenting with AARFD had distinct clinical features relative to common presentations of AARF. Notably, a higher prevalence of cervical myelopathy was observed. Because of deficiency of the transverse ligament or odontoid and subsequent AAD, surgical treatments were recommended for these cases. Surgery involved intraoperative traction-based assessment of the irreducibility, with subsequent transoral release (when indicated) and posterior C1–2 reduction and fusion. In this report, AARFD cases were successfully managed surgically without preoperative traction, with few complications seen.

➢ Key Points

- ❑ AARFD was used to address the pathological state of fixed torticollis concomitant with AAD.
- ❑ Thirty-two children with AARFD (sustained torticollis more than 6 weeks and ADI more than 5 mm) were retrospectively studied.
- ❑ AARFD had distinct clinical features relative to common presentations of AARF.
- ❑ Because of deficiency of the transverse ligament or odontoid and subsequent AAD, surgical treatments were applied for all 32 patients.
- ❑ Solid fusion and torticollis healing were achieved in 31 patients (96.9%). The complications included CSF leakage and torticollis recurrence.

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