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Atlanto-Axial Rotatory Fixation in Children：Comparison of Clinical Findings and Outcomes by Etiology

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Abstract：**Background**: Atlantoaxial rotatory fixation（AARF）is an uncommon cause of childhood torticollis and is generally associated with trauma or inflammation of the upper respiratory tract and neck. We aimed to clarify the clinical findings and outcomes of AARF by etiology to provide pediatricians with valuable for effective treatment.

Methods: Sixteen patients, admitted to our hospital from 2005 to 2013 with AARF, were assigned to 2 groups according to etiology; traumatic（n = 10, age 6.8 ± 0.6 years）or atraumatic（n = 6, age 6.3 ± 0.4 years）. Patient backgrounds, treatment, and prognosis were evaluated retrospectively and compared between the two groups.

Results: The median length of persistent torticollis and time from the initiation of treatment to remission were longer（but not significant）in the atraumatic group than in the traumatic group（16 days vs. 10.5 days; $p = 0.07$, 7.5 days vs. 5 days; $p = 0.09$）. There was a significant correlation between the time to initiate Glisson traction and length of Glisson traction in only those who underwent Glisson traction（$r^2 = 0.45$, $p = 0.0001$）.

Conclusions: Pediatricians should immediately refer AARF patients to surgeons who specialize in atlantoaxial junction surgery.

Introduction

Atlantoaxial rotatory fixation（AARF）is an uncommon cause of childhood torticollis and is generally associated with trauma or inflammation of the upper respiratory tract and neck. Children with torticollis present with asymmetrical（“cock-robin”）head position and restricted movement.

Since Bell（1830）reported the first case of rotation deformity of the atlantoaxial joint, produced after pharyngeal ulceration in a patient with torticollis, many cases have been reported[1)]. The term *rotatory fixation* was used for the first time by Wortzman et al.（1968）as *rotatory fixation of the atlantoaxial joint*[10)]. In addition, this clinical condition has been described using multiple terms such as rotatory subluxation, ro-

Key words：atlantoaxial rotatory fixation, childhood torticollis, Fielding classification, Glisson traction
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tatory dislocation, spontaneous hyperemic dislocation, inflammatory torticollis (named after its etiology), and Griselís syndrome.[3)6)] The term AARF, coined by Fielding and Hawkins in 1977, is preferred because fixation of the axis (C2) may not occur with subluxation or dislocation[3)]. The etiology may be trauma; however, upper respiratory tract infections and cervical lymphadenitis as well as inflammatory diseases of the neck, including rheumatoid arthritis and cervical lymphadenopathy due to Kawasaki disease, may trigger AARF[8)]. Many studies have addressed AARF in the field of orthopedics. However, in pediatrics, only few authors have reported about AARF, and none have compared cases with different etiologies.

The diagnosis of AARF is generally based on cervical CT, particularly 3-dimensional (3D)-CT, whereas MRI can identify ligamentous damage[8)]. Repositioning is possible using an elastic cervical orthosis and Glisson traction. However, in recurrent cases or those in which repositioning may be impossible, operative treatment may be required[2)]. Unfortunately, delayed treatment may lead to neurological complications, requiring elaborate operative intervention[7)]. Subach et al. (1998) reported a relationship between the initiation of primary care and treatment success, recurrence, complications, and prognosis. Therefore, early diagnosis and treatment of AARF are essential[9)].

While AARF is a childhood disease, pediatricians do not have the necessary information and educational tools to diagnose the disease. This ability is generally restricted to orthopedic specialists. Therefore, the present study aimed to investigate and compare clinical findings and outcomes of AARF patients by etiology to provide pediatricians with valuable for effective treatment.

METHODS

Cases of AARF

From January 2005 to December 2013, 16 cases of AARF were diagnosed at Sagamihara Kyodo Hospital, Kanagawa, Japan. Patients were retrospectively divided into traumatic and atraumatic groups according to etiology and clinical findings of AARF in the medical records. AARF was diagnosed on the basis of painful torticollis and CT or 3D-CT scans through C1-C2 in patients with clinically diagnosed AARF who demonstrated rotatory subluxation of C1 on C2. Outpatients and patients with a bone fracture or congenital deformity were excluded. Traumatic factors were defined as an external injury, for example, sudden rotation or fall. Atraumatic factors were defined as medical conditions or diseases such as inflammation or cervical lymphadenopathy.

Clinical Assessment and Treatment Parameters

Patient backgrounds and clinical symptoms (age, gender, history, height and weight on admission, torticollis direction, and rotation direction), cervical CT findings [atlantodental interval (ADI) and rotation angle], Fielding classification, duration of treatment (time to initiate treatment, persistence of torticollis, and time from the initiation of treatment to the remission of torticollis), therapeutic modality (compliance with and duration of Glisson traction, initiation of use of an elastic harness, and operative treatment), and prognosis (recurrence and complications) were examined. The outcomes for each parameter were compared between the groups. Moreover, the patients were evaluated for disease severity using Fielding classification. The rotation angle was determined to be right or left

from the neutral position of the C1–C2 joint on cervical CT.

The time to initiate treatment was defined as the time from the onset of torticollis to the application of an elastic harness or the initiation of Glisson traction. The persistence of torticollis was defined as the time from the onset to the remission of torticollis. The time from the initiation of treatment to the remission of torticollis was defined as the time from the application of an elastic harness or the initiation of Glisson traction to the remission of torticollis.

We examined the correlation between the time to initiate treatment and persistence of torticollis in all the patients and examined the correlation between the time to initiate Glisson traction and duration of Glisson traction in only those who underwent Glisson traction. We defined remission as the time when AARF resolved and persistence when there was no remission.

Fielding Classification

AARF is classified into 4 types: (i) type I, rotatory fixation without anterior displacement of the atlas; (ii) type II, rotatory fixation with anterior displacement of 3–5 mm; (iii) type III, rotatory fixation with anterior displacement of more than 5 mm; and (iv) type IV, rotatory fixation with posterior displacement[3].

Statistical Analysis

Data were analyzed using graphpad prism 5 for windows (GraphPad Software, La Jolla, CA, USA). Data are presented as the mean ± standard error of mean (SEM) or as the median and range. Parameters between the groups were compared using Fisherís exact test and Mann–Whitney U-test. Non-continuous variables were compared using Spearman rank correlation coefficients. A p value less than 0.05 indicated statistical significance.

Results

Sixteen patients were assigned to a traumatic (n = 10, age 6.8 ± 0.6 years) or an atraumatic (n = 6, age 6.3 ± 0.4 years) group. In the traumatic group, AARF was caused by sudden rotation (n = 5), neck sprain (n = 4), or fall (n = 1). In the atraumatic group, torticollis was caused by acute upper respiratory infection (n = 2), parapharyngeal abscess (n = 1), cervical lymphadenitis (n = 1), or Kawasaki disease (n = 2) and AARF occurred secondarily (Table 1). In the traumatic and atraumatic groups, 4 (40%) and 4 (67%) patients were males, respectively. All the patients had no neurological complication during treatment. The direction of torticollis was toward the right in 8 (80%) and 4 (67%) patients in the traumatic and atraumatic groups, respectively (Table 2). There were no significant differences in other aspects of patient profiles or clinical findings between the groups. Because 1 patient had a medical history of attention deficit/hyperactivity disorder, this patient could not undergo Grison traction.

There were no significant differences in ADI (traumatic group, 3.5 ± 0.3 mm; atraumatic group, 2.7 ± 0.2 mm) or the angle of rotation on cervical CT (traumatic group, 21.5 ± 3.4°; atraumatic group, 17.7 ± 43.8°; Table 2) between the groups. Moreover, there were no significant differences in Fielding classification (type I, II, III: 2, 7, 1 patients in the traumatic group and 3, 2, 0 patients in the atraumatic group). There were no patients with Fielding type IV, and none required operative treatment. The use of 3D-CT imaging for AARF diagnosis has increased in the last few years (Table 1), and for example we present 3D-CT findings for cases 13 (Figure.

Table 1. Patient background

Patient No.	Age (years)	Gender	Etiology	3D-CT	Fielding classification	ADI (mm)	Rotation angle(°)	Time to initiation of treatment (days)	Glisson traction
1	5.4	Male	Kawasaki disease	No	−	−	−	19	No
2	5.7	Female	Sudden rotation	No	II	3.7	5.4	2	Yes
3	5.2	Male	Fall	No	II	3.2	−	6	Yes
4	4.2	Male	Neck sprain	No	II	3.9	−	9	Yes
5	5	Female	Fall	No	I	2.3	17.7	1	Yes
6	9.8	Male	Neck sprain	No	II	4.5	11.1	3	Yes
7	5	Female	Sudden rotation	No	I	2.5	21.1	2	Yes
8	6.5	Male	URI	Yes	II	3.6	25.4	10	Yes
9	7.6	Female	URI	No	II	2.8	23	5	Yes
10	6	Male	Paraphary ngeal abscess	No	I	2.2	6.64	3	No
11	8.8	Female	Neck sprain	Yes	II	3.5	32	4	Yes
12	7.2	Female	Sudden rotation	Yes	I	2.4	24.5	9	Yes
13	6	Female	Kawasaki disease	Yes	I	2.7	28.3	11	Yes
14	8.4	Female	Sudden rotation	Yes	I	2.3	30.8	12	Yes
15	3.6	Male	Cervical lymphadenitis	No	I	2.3	12.4	3	No
16	6.5	Male	Sudden rotation	Yes	III	5.5	29.5	5	Yes

Traumatic group(n＝10), atraumatic group(n＝6)
ADI, atlantodental interval；URI, Upper respiratory tract infection

1a)and 16(Figure. 1b).

The median time to initiate treatment was greater in the atraumatic group(7.5 days, range 3-19 days)than in the traumatic group(4.5 days, range 1-12 days; p = 0.09; Figure. 2a). The median length of persistence of torticollis was longer in the atraumatic group(16 days, range 7-47 days)than in the traumatic group(10.5 days, range 5-30 days; p = 0.07; Figure. 2b). The median time from the initiation of treatment to the remission of torticollis in the atraumatic group was 7.5 days(range 4-28 days)and was longer than that in the traumatic group(5 days, range 3-51 days; p = 0.09; Figure. 2c). However, each interval was no significant differences between the groups(Figure. 2). Furthermore, no significant differences were observed for these intervals between the groups when compared by Fielding classification. In case of 14 patients in whom Fielding classification could be determined, the median time to initiate treatment was 3 days in type I(range 1-11 days)and 5 days in type II(range 2-12 days). The persistence of torticollis was 13 days in both type I

Table 2. Comparison of backgrounds, imaging, Fielding classification, and therapeutic modality between traumatic and atraumatic groups.

Variable	Traumatic group (n＝10)	Atraumatic group (n＝6)	*p*
Patient background/clinical symptoms			
Age(years)	6.6±0.6	5.9±0.5	ns
Gender: Male; n(%)	4(40)	4(67)	ns*
History; n(%)	1(10)	0(0)	−
Height(m)	1.2±0.04	1.1±0.03	ns
Body weight(kg)	19.8±2.1	20.0±1.1	ns
Torticollis direction(right); n(%)	6(60)	3(50)	ns*
Persistence of torticollis(days)	13±2.8	21.2±5.9	0.09
Imaging			
ADI(mm)	3.5±0.3	2.7±0.2	ns
rotation angle(°)	21.5±3.4	17.7±3.8	ns
Fielding classification			
type I; n(%)	2(20)	3(50)	ns
type II; n(%)	7(70)	2(33)	ns
type III; n(%)	1(10)	0(0)	−
Therapeutic method			
Glisson traction; n(%)	10(100)	3(50)	0.06*
Duration of Glisson traction(days)	7.4±1.4	6.7±0.8	ns
Elastic harness; n(%)	10(100)	5(83)	ns*
Surgery; n(%)	0(0)	0(0)	−
Complications; n(%)	0(0)	0(0)	−

Traumatic group(n＝10), atraumatic group(n＝6)
Mean±standard error of the mean(SEM)
Mann-Whitney U-test；*Fisher's exact test; ns, not significant
ADI, atlantodental interval

(range 7-28 days)and type II(range 5-30 days). The median time from the initiation of treatment to the remission of torticollis was 6 days in type I(range 4-25 days)and 4 days in type II (range 3-24 days).

All the patients had complaints regarding wearing an elastic harness or regarding Glisson traction. There were no significant differences between the groups when compared for therapeutic modality(Table 2).

In all the patients, there was a significant correlation between the time to initiate treatment and persistence of torticollis(r^2 = 0.60, p = 0.0005). In addition, there was a significant correlation between the time to initiate Glisson traction and duration of Glisson traction in only those who underwent Glisson traction(r^2 = 0.45, p = 0.0001; Figure. 3).

Discussion

We retrospectively performed a medical record-based study to compare clinical findings and outcomes of 16 AARF cases by etiology. To the best of our knowledge, this is the first comparative study on pediatric AARF cases in Japan. We demonstrate that it is important for pe-

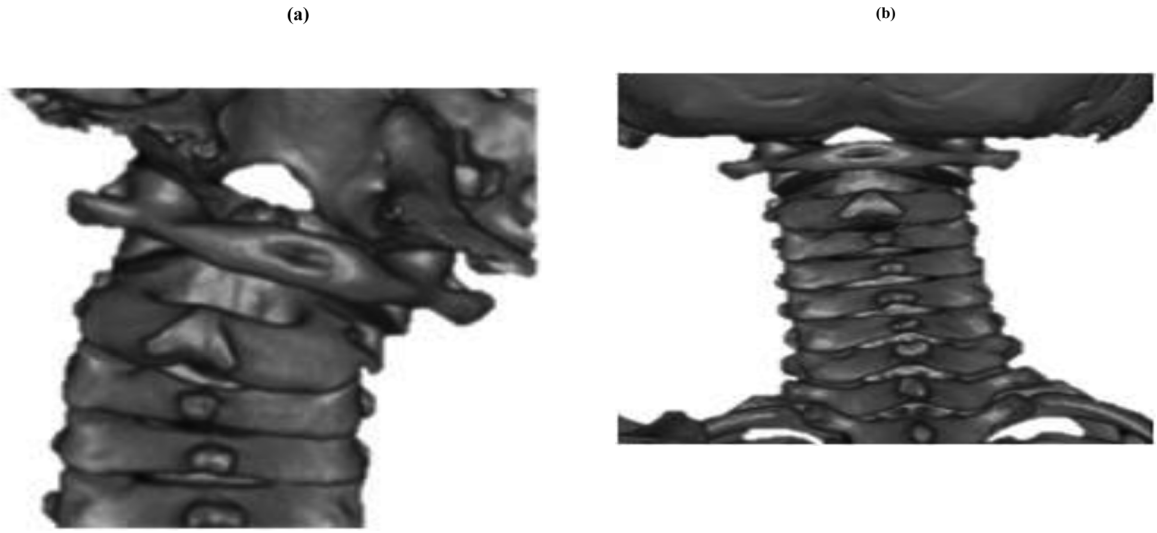

Fig. 1. 3D-CT images of cases 13 and 16.
Torticollis direction was toward the right on admission in case 13 and can also be observed from the front(a). In case 16, torticollis direction was toward the right on admission ; however, the neck was in the median position during remission(b ; back view).

diatricians to suspect AARF when a patient presents with painful torticollis so that early diagnosis can be made and treatment can be initiated as soon as possible.

Various conditions may lead to AARF development. In the present study, 6 of 16 cases developed AARF caused by cervical inflammatory diseases, such as inflammation of the upper respiratory tract or neck and Kawasaki disease. Hicazi et al.(2002)reported that 6 of 33 AARF cases were caused by upper respiratory tract and tonsil inflammation and that 18 cases developed from unknown causes and did not present with lymph node swelling[4]. Thus, these independent studies have consistently suggested that AARF is caused by atraumatic conditions.

While various conditions may lead to AARF, the mechanisms underlying AARF have not been clearly identified till date. Fielding and Hawkins(1977)proposed possible clues to solve this problem[3]. They hypothesized that early articular capsule injury, sustained muscle spasm, and synovial swelling could cause fixation of the joint in a rotated orientation. Some pediatric diseases, particularly, those causing swelling of the lymph nodes, present with inflammation, articular capsule development, synovial inflammation, and swelling were developed AARF. It is assumed that atraumatic AARF is caused by such inflammatory conditions in conjunction with sustained movement restriction caused by cervical pain. However, some cases assessed in the present study did not present with lymph node swelling, suggesting that AARF was caused by another type of inflammation.

Patient backgrounds, clinical symptoms, and diagnostic imaging results were not significantly different between the traumatic and atraumatic groups. These findings indicated that the clinical state was not affected by etiology.

AARF diagnosis is necessary to evaluate rotatory subluxation of C1 on C2 by cervical CT, to assess the direction and range of rotation and migration of the atlantoaxial joint, as well as to diagnose severity on the basis of Fielding classification.

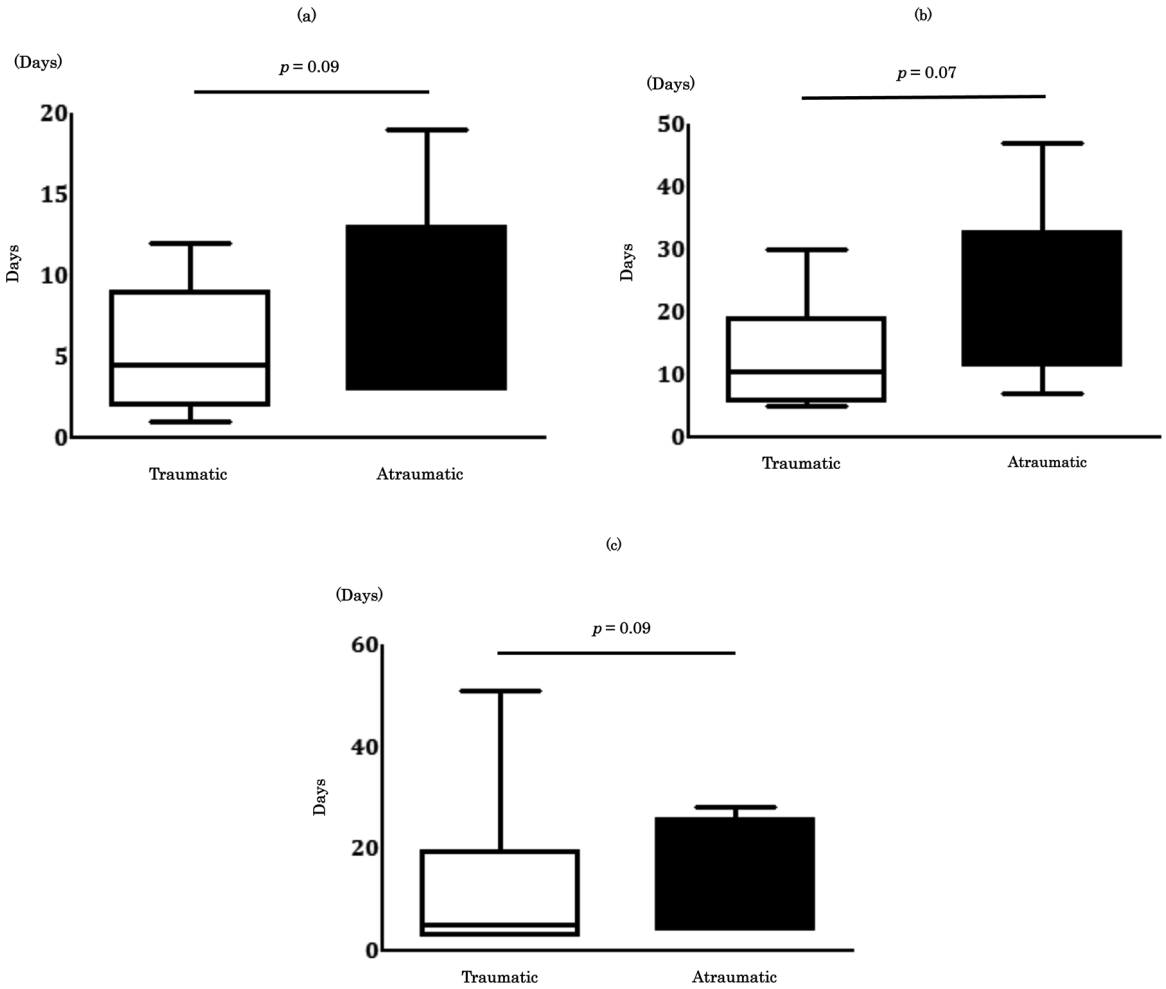

Fig. 2. Relationship of treatment and torticollis in traumatic (n = 10) and atraumatic (n = 6) groups. (a) Initiation of treatment: 3 days (5-11 days) vs. 10 days (3-19 days; $p = 0.09$)；(b) Persistence of torticollis: 7.5 days (5-30 days) vs. 18 days (13-47 days; $p = 0.07$)；(c) Time from the initiation of treatment to remission of torticollis: 4.5 days (3-24 days) vs. 8 days (4-28 days; $p = 0.09$). Data are presented as median (range). Mann-Whitney U-test, $p < 0.05$.

The mean age of the patients in the present study was 6.6 ± 0.6 years in the traumatic group and 5.9 ± 0.5 years in the atraumatic group. This is similar to the median age of 6.5 years of patients in the study conducted by Mihara et al. (2001)[5] AARF is indeed observed more frequently in infants and school-age children; some reasons for this may include features pertaining to physical development and anatomy in children, such as (i) insufficient bony structural support; (ii) loose joint capsule and large torsion angle as well as large proportion of soft tissue in the atlantoaxial joint and its susceptibility to inflammation; (iii) horizontal articular surfaces of the lower cervical spine; (iv) invaginated synovial walls with clear crescentic vasculature in the occiput, atlantoaxial joint, and outer atlantoaxial joint; and (v) direct connection of the pharyngeal lymph vessel to the venous plexus and the resultant susceptibility to inflammation.

Our results revealed that the duration of tor-

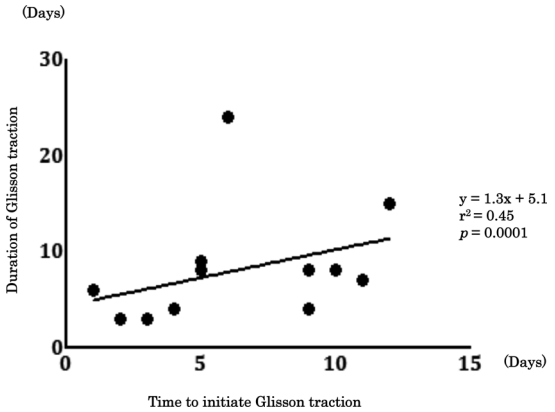

Fig. 3. Correlation between the time to initiate Glisson traction and duration of Glisson traction in only those who underwent Glisson traction (n = 13, r^2 = 0.45, p = 0.0001). Spearman rank correlation coefficient, $p < 0.05$.

ticollis in the atraumatic group tended to be longer than that in the traumatic group. This could be caused by delayed treatment in the atraumatic cases, wherein the primary disease is preferentially treated before treating AARF. Roche et al.(2001) demonstrated that the ease of treatment of AARF depends on the elapsed time from onset to diagnosis[7]. When diagnosis is made within 1 week of disease onset, the case can be simply treated with an elastic cervical orthosis. When diagnosis takes over 1 week, traction is required, and when diagnosis takes over 3 months, surgical treatment is required. We therefore conclude that early diagnosis, including assessment for severity, is important.

Subach et al.(1998) reported that longer durations in initiating treatment resulted in lower rates of treatment success and higher rates of recurrence and complication (13.8 ± 12.1 days vs. 69.7 ± 52.4 days)[9]. In the present study, we also observed a significant correlation between the time to initiate Glisson traction and remission of torticollis. In addition, in only those who underwent Glisson traction, we observed a significant correlation between the time to initiate Glisson traction and duration of Glisson traction. Previous and current results consistently suggest the effectiveness of early treatment of AARF.

Conclusion

Pediatric AARF can be caused by traumatic as well as atraumatic events. We provide key diagnostic information to aid pediatricians in an early diagnosis of AARF in children presenting with painful torticollis so that treatment can be initiated as soon as possible. For effective treatment, we propose that pediatricians should immediately refer pediatric AARF patients to surgeons who specialize in atlantoaxial junction surgery.

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Conflict of Interest

The authors have no conflicts of interest.

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