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Original Article

Retrospective analysis of atlantoaxial rotatory fixation describing age distribution and gender ratio in children and adolescents: A preliminary report

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ABSTRACT

Introduction: Atlantoaxial rotatory fixation (AARF) in children presents with an acute onset of neck pain. Almost all cases heal within a few days of onset and are treated conservatively. Because few cases of AARF have been reported, the age distribution or gender ratio of AARF in the child population have not been described enough. In Japan, the social insurance system covers all citizens. Thus, we used insurance claims data to investigate the features of AARF. The aim of this study is to examine the age distribution, compare gender ratio and determine the recurrence proportion of AARF.

Methods: We used the JMDC database to search for claims data submitted between January 2005 and June 2017 for cases of AARF in patients aged <20 years.

Results: We identified 1949 patients with AARF, of which 1102 (56.5%) were male. The mean age was 98.3 ± 42.2 months and 91.6 ± 38.4 months in males and females, respectively, and males with AARF were significantly older at onset than females with AARF ($p < 0.001$). In both sexes, the highest frequency of AARF occurred when the patient was 6 years old. There were 121 (6.2%) cases of recurrent AARF (male: 61, 5.5%; female: 60, 7.1%), but the age differences between the sexes in these cases were not statistically significant.

Conclusions: This is the first report to describe the characteristics of the study population of AARF. Males were more likely to suffer from AARF than females. Furthermore, age (in months) at AARF onset was significantly higher in males than in females. Recurrence rate was not significant in both sexes.

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1. Introduction

Torticollis is a rare condition where the neck muscles contract resulting in the head to tilt to one side¹. As it affects very few children and no multicenter or nationwide studies are available, reports on its etiology are scarce. We used health care insurance claims data to perform a nationwide study on the features of atlantoaxial rotatory fixation (AARF) in children. According to Neal and Mohamed¹, because of the rarity of acute torticollis, case reports and case series have formed the basis for the nomenclature,

an etiology, natural history, and preferred treatments. Various names have been applied to the condition, including acute acquired torticollis², atlantoaxial rotatory subluxation³, atlantoaxial rotatory fixation⁴, atlantoaxial rotatory dislocation⁵, and similar variations. AARF caused by inflammation or surgery is known as Grisel syndrome, and a pediatric study revealed that the average age of onset was 8.5 years old⁶. Additionally, AARF is relatively rare¹ and the most common condition associated with acquired painful torticollis⁷, although the exact incidence rate remains unknown. So, therefore, to understand basic information such as the distribution or gender differences is important issue as per of epidemiologically.

This study aimed to retrospectively explore the age distribution and gender ratio of AARF in child and adolescent males and females aged <20 years by utilizing an insurance claims database.

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2. Materials and methods

2.1. Data source and study design

This retrospective study used the database of JMDC Inc. (Tokyo, Japan) to collect claims data (sex, age in months, and recurrence). The cumulative dataset contains approximately 13 million subjects (inpatients, outpatients, and pharmacy claims) aged ≤ 74 years from approximately 90% of all medical facilities in Japan⁸. All patients in the JMDC database have “social insurance” that covers the working member and their family. As of September 2021, the database represented approximately 10.0% of the Japanese population.

2.2. Patients' identification flow and definition

We obtained approximately 3 million general population between January 2005 and June 2017 from JMDC. We included cases of AARF (ICD10 code for M4361) that occurred in patients aged < 20 years from the dataset. No previous report discusses about clinical criteria of recurrent AARF, thus we defined recurrent AARF as cases reported for the same patient more than three month after the last claim in this study.

2.3. Statistical analysis

Data are expressed as mean \pm standard difference (SD). All statistical analyses were performed using JMP Pro v16.0 (SAS Institute, Cary, NC, USA). The Wilcoxon signed-rank test was used to determine differences in age at onset between the sexes, and the chi-square test was used to compare between with recurrence and without recurrence. Statistical significance was set at $p < 0.05$.

3. Results

Of the 1949 patients with AARF analyzed in our study, 1102 (56.5%) were males and 847 (43.5%) were females. Patient age and sex are shown in Table 1. In both sexes, the highest frequency of AARF occurred when the patient was 6 years old. The average age of onset was 98.3 ± 42.2 months in males and 91.6 ± 38.4 months in females ($p < 0.001$).

There were 121 cases (6.2%) of recurrent AARF (males: 61, 5.5% and females: 60, 7.1%; Table 2), but no significant difference was

Table 1
Age and sex of the patients with atlantoaxial rotatory fixation.

Age	Male	Female	Total
0	0	0	0
1	6	3	9
2	34	23	57
3	58	58	116
4	112	95	207
5	124	118	242
6	146	130	276
7	105	83	188
8	102	79	181
9	93	69	162
10	75	49	124
11	72	47	119
12	49	30	79
13	48	24	72
14	34	20	54
15	15	9	24
16	14	2	16
17	9	4	13
18	5	3	8
19	1	1	2
Total	1102	847	1949

Table 2
Age at first onset in patients with recurrent atlantoaxial rotatory fixation.

Age	Male	Female	Total
0	0	0	0
1	0	0	0
2	0	0	0
3	2	4	6 (5.2%)
4	7	8	15 (7.2%)
5	6	7	13 (5.4%)
6	6	9	15 (5.4%)
7	10	9	19 (10.1%)
8	9	5	14 (7.7%)
9	4	6	10 (6.2%)
10	5	3	8 (6.5%)
11	3	1	4 (3.4%)
12	2	2	4 (5.1%)
13	2	1	3 (4.2%)
14	1	4	5 (9.3%)
15	3	1	4 (16.7%)
16	1	0	1 (6.3%)
17	0	0	0
18	0	0	0
19	0	0	0
Total	61 (5.5%)	60 (7.1%)	121 (6.2%)

noted in AARF recurrence between the two sexes (N.S.). The age at onset in patients with recurrent AARF was 98.3 ± 42.2 in males and 91.6 ± 38.4 months in females (N.S.). The median duration between the initial onset and the first recurrence was 11 (interquartile range: 24–6) months.

Six cases (0.31%) claimed invasive treatment: two cases (4 years 11 months girl and 12 years 6 months boy) needed spinal surgery, one case (9 years 6 months girl) treated manipulation and Halo vest fixation, one case (15 years 5 month boy) treated Halo vest fixation, one case (18 years 1 month boy) treated manipulation one time and one boy treated manipulation twice at the age of 6 years 4 months and 13 years 5 months.

4. Discussion

During the COVID-19 pandemic, Kawasaki disease and other common podiatric infectious diseases decreased⁹, and physical activity during these periods also decreased due to lockdown, a state of emergency period, school closure period, social distancing period and/or other conditions. Thus, we assume the frequency of AARF might be decreased due to decreases in inflammation and trauma triggers. Hence, further study of etiology was required.

Congenital torticollis is the third most common musculoskeletal abnormality in infants with hip dysplasia and clubfoot¹⁰. According to Cheng, the incidence of congenital torticollis is 0.3%–1.9%, and although the etiology remains uncertain, the influence of birth trauma has been described and was found to occur in almost all patients younger than 12 months at onset¹¹. The incidence of congenital torticollis was reported to be higher in boys than in girls, but this was not statistically significant¹². Thus, we believe that congenital torticollis and AARF are distinct diseases. In our study of AARF, the youngest patient was 14 months old, and the gender ratio was higher in males (56.5% vs. 43.5%). As already proposed by Tang et al., we agree that the reason for the occurrence of torticollis in children or teenagers rather than adults could be the larger synovial folds, which may become entrapped, leading to oedema and subsequent subluxation of the atlantoaxial joints¹³.

Sex differences have not been described in previous studies and reviews of AARF. We found that the ratio of AARF was higher in male patients than in females. Furthermore, the average age of males at AARF onset was 6 months older than that of females. Even after adjusting for sex in Japan, where there are 5% more males than

females, the ratio of AARF in males remained relatively higher than in females.

Fielding classified AARF into four types: from type 1, the most common, to type 4, the rarest⁴). Various treatment methods have been reported¹. In our study, symptoms onset and type were not considered; thus, we do not discuss it; likewise, we were unable to consider treatment method by the type and chronic or acute. In chronic cases, some patients choose elective surgery¹, whereas others are managed using remodeling therapy¹⁴. However, in our study only 6 cases (0.3%) claimed invasive treatment and others (1943 cases occupied 99.7%) did not claimed treatment fee. Thus, we assume 1943 cases (99.7%) treated conservatively. So, this study might be first report to treatment ratio invasive and conservative.

Reports of recurrent AARFs are rare. In our study, the recurrence was observed in 6.2%, and the average duration between initial onset and first recurrence was 17.8 months. However, the distinguishing features of recurrent AARFs remain unclear and require further investigation.

Our study had several limitations. First, patients who did not visit a doctor, were misdiagnosed or did not claim health insurance were excluded from our study population. Second, the data collected in our study were limited, and we did not record data pertaining to the AARF type, rotation direction (right or left), diagnostic method, symptom duration, treatment method, trigger event that caused torticollis, duration of symptoms before visiting a doctor or other insurance case information. Thus, we were unable to discuss treatment options and duration or other factors related to these. Furthermore, to understand the etiology of AARF, population-based cohort studies are needed.

5. Conclusion

Our study revealed that the gender ratio of AARF was higher in males than in females. Furthermore, the age at onset (months) was higher in males than in females. The highest frequency was observed in patients aged 6 years. Recurrent AARF was present in 6.2% of the cases, and the median duration between the initial onset and the first recurrence was 11.0 months. Further etiological investigations are required to better understand AARF.

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None.

Author contributions

Y.O., K.T., and R.S. designed the study; Y.O., K.T., R.S., T.U., and K.M. performed the analyzed the data; T.U. supervised the literature search; Y.O., T.U., and K.M. wrote the manuscript.

Ethical approval

The commercial database of JMDC. Inc. used in this study is anonymized processed information based on Japan's Personal Information Protection Law, and individual informed consent is not required for its provision and use.

In addition, according to the ethical guidelines for clinical research in Japan, research using anonymized processed information should not be reviewed by an ethical review committee.

Informed consent

N/A.

Declaration of competing interest

Y.O. received lecture fees from Daiichi Sankyo Co. Ltd and Asahi Kasei Pharma. K.M. and JMDC Inc. collaborate other project according to the collaborative research agreement. JMDC Inc. did not intervene in data implementation according to the analyzed results of this study. K.M. received honorarium fees for presentations from JMDC Inc. Nippon-Kayaku and Abbvie Inc. K.T. received a research grant from Bayer Yakuhin, Ltd. Department of Hospital Pharmaceutics received budget from Ono with a contract research project according to the collaborative research agreement.

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References

- [1] Neal KM, Mohamed AS. Atlantoaxial rotatory subluxation in children. *J Am Acad Orthop Surg* 2015 Jun;23(6):382–92.
- [2] Hicazi A, Acaroglu E, Alanay A, Yazici M, Surat A. Atlantoaxial rotatory fixation-subluxation revisited: a computed tomographic analysis of acute torticollis in pediatric patients. *Spine (Phila Pa 1976)* 2002 Dec 15;27(24):2771–5.
- [3] Phillips WA, Hensinger RN. The management of rotatory atlanto-axial subluxation in children. *J Bone Joint Surg Am* 1989 Jun;71(5):664–8.
- [4] Fielding JW, Hawkins RJ. Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). *J Bone Joint Surg Am* 1977 Jan;59(1):37–44.
- [5] Landi A, Pietrantonio A, Marotta N, Mancarella C, Delfini R. Atlantoaxial rotatory dislocation (AARD) in pediatric age: MRI study on conservative treatment with Philadelphia collar—experience of nine consecutive cases. *Eur Spine J* 2012 May;21(Suppl 1):S94–9.
- [6] Anania P, Pavone P, Pacetti M, Truffelli M, Pavanello M, Ravegnani M, et al. Grisel syndrome in pediatric age: a single-center Italian experience and review of the literature. *World Neurosurg* 2019 May;125:374–82.
- [7] Herring JA. Tachdjian's pediatric orthopaedics: from the Texas scottish rite hospital for children. In: E-book. fifth ed. Elsevier Health Sciences; 2013. p. 167–205 [Chapter 11], Disorders of the neck.
- [8] Nagai K, Tanaka T, Kodaira N, Kimura S, Takahashi Y, Nakayama T. Data resource profile: JMDC claims database sourced from health insurance societies. *J Gen Fam Med* 2021 Feb 14;22(3):118–27.
- [9] Ae R, Shibata Y, Kosami K, Nakamura Y, Hamada H. Kawasaki disease and pediatric infectious diseases during the coronavirus disease 2019 pandemic. *J Pediatr* 2021 Dec;239: 50-58.e2.
- [10] Kaur S. Congenital torticollis and its physiotherapy management. *Int J Health Sci Res* 2020 Feb 10;(2):94–101.
- [11] Cheng JC, Tang SP, Chen TM, Wong MW, Wong EM. The clinical presentation and outcome of treatment of congenital muscular torticollis in infants—a study of 1,086 cases. *J Pediatr Surg* 2000 Jul;35(7):1091–6.
- [12] Chen MM, Chang HC, Hsieh CF, Yen MF, Chen TH. Predictive model for congenital muscular torticollis: analysis of 1021 infants with sonography. *Arch Phys Med Rehabil* 2005 Nov;86(11):2199–203.
- [13] Tang XY, Liu LJ, Yang HJ, Peng MX, Liao SH. Anatomic study of the synovial folds of the occipito-atlanto-axial joints. *Clin Anat* 2007 May;20(4):376–81.
- [14] Ishii K, Matsumoto M, Momoshima S, Watanabe K, Tsuji T, Takaishi H, et al. Remodeling of C2 facet deformity prevents recurrent subluxation in patients with chronic atlantoaxial rotatory fixation: a novel strategy for treatment of chronic atlantoaxial rotatory fixation. *Spine (Phila Pa 1976)* 2011 Feb 15;36(4):E256–62.