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The Grisel Syndrome: early interdisciplinary treatment needed to prevent severe upper cervical deformity - Experience from a high-volume spine center and systematic review of existing literature

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ABSTRACT

Purpose: Grisel Syndrome (GS) describes a rare, non-traumatic atlantoaxial rotatory subluxation in children typically associated with upper respiratory tract infection, or otorhinolaryngological surgery. The aim of this study was to systematically review the literature regarding interdisciplinary treatment approaches, consequences of delayed or misdiagnosed GS, and upper cervical deformities resulting from misdiagnosed GS. Additionally, we share our institutional experience with this pathology.

Methods: We queried our institutional database for patients suffering from GS which were treated in our department between 2001 and 2024. A systematic literature search was conducted using Medline and the Cochrane Library, following PRISMA guidelines.

Results: Our database revealed three patients with GS. One patient with misdiagnosed of GS presented with rigid occipital rotation, ultimately progressing to occipito-atlantoaxial rotatory dislocation (OAARD). Two patients were early diagnosed and successfully treated with halo vest immobilization. The systematic literature review identified 13 cases of OAARD across 12 reports. Etiologies included upper respiratory infections, otorhinolaryngological surgery, juvenile idiopathic arthritis, and minor trauma. The average time to diagnosis of OAARD was 5.9 months (0.5–18 months). The treatment involved occipito-atlantal fusion (43 %), atlantoaxial fusion with halo vest (29 %), or traction followed by halo vest (21 %).

Conclusion: GS and OAARD are underrecognized and often diagnosed late, causing irreversible deformity and permanent loss of upper cervical spine mobility. Painful torticollis following upper respiratory tract infection, otorhinolaryngological surgery, or minor trauma should be treated as GS until proven otherwise. Early recognition and interdisciplinary treatment are crucial to prevent progression and avoid invasive surgical interventions.

1. Introduction

Grisel syndrome (GS) – also known as atlantoaxial subluxation (AARS) - describes a rare condition with fixed forward subluxation of the atlas on the axis [1]. This syndrome is typically seen in otherwise healthy children with a history of an inflammatory process, such as an upper respiratory tract infection, or otorhinolaryngological surgery [2–5]. Further, associations with trivial or severe trauma, autoimmune diseases, congenital deformations, and chromosomal aberrations have been described [4–7]. In clinical examination, the typical finding is a head held in “cock-robin” position: following the direction of the forward subluxated atlas, the head rotates to one side and side-bends to the opposite side with a slight flection, reminding of a robin turning his head to listen for a worm (Fig. 1) [8]. Diagnosis is confirmed by computed tomography (CT) scan – due to its ability to visualize bony alignment and rotational or tilting deformities - and classified by Fielding and Hawkins classification assessing the extent of atlas rotation and the atlanto-dental dissociation (Table 1) [8]. Magnetic resonance imaging (MRI) can be performed to exclude abscess formation, soft-tissue involvement, or basilar invagination if clinically suspected.

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Named after a French otorhinolaryngologist, Pierre Grisel (1869–1959), who described AARS in patients with pharyngitis in 1930, Aetius of Amida (502–576) was the first to associate a vertebral luxation and vertebral inflammation in a patient with retropharyngeal abscess [1,9]. However, the underlying pathophysiological causes have not been fully elucidated yet. Most of the patients suffering from GS are in a pediatric patient population, thus, Battiata and Pazos introduced a "two-hit hypothesis" to explain the obvious age dependency [3,10]. The "first hit" includes a predisposition for AARS in children due to biomechanical reasons, including an atlantoaxial hypermobility with increased atlas-dens interval, immature bone age and delayed facet-joint formation, ligamentous laxity, large synovial folds and weak cervical muscles [10–13]. The "second hit" includes an increased laxity of the transverse and the alar ligaments as well as muscle spasms resulting from a spread of inflammatory mediators carried by nasopharyngeal and pharyngovertebral veins to the periodontoid vascular plexus [10,14].

Current literature emphasizes the need to restore the atlantoaxial alignment and along with that the physiological joint function, with a focus of interest on conservative and surgical treatment options. [6,15]. However, we recognized a lack of evidence according to the natural course of non-treated GS [16] In 1959, Washington described a missed diagnosis of GS in a nine-year-old girl leading to counter backward rotation of the skull resulting in an irreversible AARS with additional atlanto-occipital dislocation (occipito-atlantoaxial dislocation, OAARD) [17,18]. Despite a reduction of head movement in rotation and extension, her head was in normal position with absent pain or neurological deficits after several years. Ever since, OAARD received little attention in published literature and is probably underdiagnosed. We believe that early diagnosis and structured interdisciplinary treatment are the key factors in case of GS, to restore normal function and to prevent irreversible spinal deformation.

Consequently, the aim of this study was to work up the evidence on OAARD resulting from GS. Therefore, we systematically reviewed the literature as well as our institutional database concerning three main questions.

(1) What are our experiences with GS and OAARD and are they in consistence with the published literature?
(2) Can delayed diagnosis of GS be considered as risk factor for OAARD?
(3) Are patients with OAARD at risk for increased severe events or worse outcomes, and what clinical consequences arise from this?

Table 1
Fielding and Hawkins classification. Classification of fixed rotatory subluxation of the atlantoaxial joint, also known as atlantoaxial rotatory subluxation (AARS), according to Fielding and Hawkins [8]. Type I-III typically occur in GS and are classified based on the amount of atlanto-dental dissociation, type IV with posterior displacement of the atlas can result from odontoid fracture.

Fielding and Hawkins classification of "fixed rotatory subluxation of the atlantoaxial joint"	
Type I	Rotatory subluxation of the atlas on the axis with no anterior displacement, the odontoid acting as the pivot
Type II	Rotatory subluxation with anterior displacement of 3–5 mm, one lateral articular process acting as the pivot
Type III	Rotatory subluxation with anterior displacement more than 5 mm, one lateral articular process acting as the pivot
Type IV	Rotatory subluxation with posterior displacement

2. Methods

This study was conducted at a DWG-certified Level 1 Spine Center at a tertiary university hospital, representing a high-volume spine center. We performed a comprehensive review of our institutional database to identity patients that were treated due to GS between 2001 and 2024. Cases of patients operatively treated due to the diagnosis of "Grisel syndrome" or "AARS" were included, as well as patients with the diagnosis of "OAARD" regardless of their surgical history. Patients who were treated for acute torticollis without radiological evidence of Grisel's syndrome or without requiring surgical treatment were not included. This study was performed in accordance with the institutional ethics committee that does not require approval for retrospective studies.

In addition, a systematic literature review was carried out following PRISMA guideline [19]. Accordingly, the Cochrane library and Medline electronic database were searched in October 2023 by using the following search strategy.

(1) Cochrane library [All text]: "Grisel OR AARS OR OAARD".
(2) Medline [All text]: "Pubmed [All text]: "((atlanto-occipital) OR (atlanto occipital) OR (occipital-atlantoaxial) OR occipitoatlantal OR (occipito atlantal)) AND rotatory AND (subluxation OR fixation OR dislocation) OR OAARD OR (natural course AND (Grisel OR AARS OR AARF))".

Inclusion criteria comprised systematic reviews, meta-analyses,

Fig. 1. The "cock-robin" position. Following the direction of the forward subluxated atlas, the head rotates to one side and side-bends to the opposite side with a slight flection. This reminds of a robin turning his head to listen for a worm (left picture: five-year-old boy with his head in "cock-robin" position due to GS; right picture, schematic illustration of a robin that listens to a worm).

prospective and retrospective studies, as well as case series and case reports that address occipital-atlantoaxial pathologies (OAARD) resulting from GS without restriction in language or publication date. The diagnosis had to be made by (1) a clinical presentation consistent with GS and (2) radiological confirmation. Case reports and studies describing isolated GS without radiologically confirmed OAARD were not included. Patients suffering from severe traumatic diseases, iatrogenic conditions, or syndromes, as well as patients with congenital deformities were excluded. The first and the third author reviewed titles and abstracts in October 2023 independently. After full texts were checked for suitability, bibliographies of included manuscripts were additionally searched for possible studies that meet our inclusion criteria. We planned to grade the level of evidence by Oxford Centre of Evidence-Based Medicine (OCEBM, 2011) and to assess the risk of bias by Cochrane Collaboration risk-of-bias tool, with retrospective studies - including case-series and case reports - graded as "high-risk" [20,21].

The data were collected using Microsoft Excel (Microsoft® Excel, version 16.78, © 2023 Microsoft, Redmond, WA). Statistical analysis was performed using „BiAS for Windows" (©Epsilon-Verlag GbR Hochheim, version 11.09, Darmstadt, Germany). Numerical data are presented as mean values and standard deviations with corresponding minimum and maximum values. Categorical data are presented as absolute and relative values.

3. Results

Institutional Database. There were three patients with GS identified in our database, of whom two patients suffered from GS and one patient from OAARD.

The first patient was a five-year-old boy with a history of sinus surgery and tonsillectomy due to chronic rhinosinusitis and recurrent upper respiratory tract infection. Two days after surgery, he complained of spontaneous torticollis and neck pain (Fig. 1A). The attending otorhinolaryngologist recommended an osteopathic treatment. The osteopath recommended to consult an orthopedic doctor. Due to history of otorhinolaryngological surgery and torticollis possible GS was suspected by the orthopedic doctor 4 days after onset of symptoms. However, radiological imaging was not performed at that time. Instead, the orthopedic doctor recommended physiotherapy, and, after another otorhinolaryngological consultation, pain medication and antibiotics were given in suspicion of a delayed wound healing. There were no signs of a systemic infection such as fever or blood test abnormalities. As the torticollis did not improve, 17 days after the onset of symptoms, MRI was performed for further evaluation. Here, a vertebral subluxation of the upper spine was suspected, however, in absence of spinal cord compression or pharyngeal abscess the *watch and wait* approach was continued. Finally, after the torticollis did not resolve after five weeks of outpatient treatment, the patient was admitted to an otorhinolaryngologist working at a university hospital. At that time, the patient had his head held in cock-robin position, there were still no signs of infection, and a CT confirmed the diagnosis of GS (Type Fielding II). Finally, the boy was admitted to our Department of Orthopedics and Trauma Surgery. As we did not manage to receive a normal function of the atlantoaxial joint, despite additional neck immobilization, high-dosed pain medication and additional administration of muscle relaxants, we recommended a closed reduction under general anesthesia followed by halo vest immobilization for 6 weeks. Under general anesthesia, and application of slight traction, extension, and rotation the cervical spine regained free range of motion. Transoral closed reduction was not needed. The cervical spine was stabilized with a hard neck collar and the reduction was confirmed with a CT scan. Still in general anesthesia, we adjusted the halo vest (Fig. 3A and B). This was performed 6 weeks after the onset of symptoms. The patient did well in the clinical examinations and 6 weeks postoperatively the halo vest was changed to a hard collar immobilization for another 6 weeks (Fig. 3C). At clinical examination half a year later, the boy held his head in a neutral position (Fig. 3D). Cervical range of motion was without restriction, and he had no complaints.

The second patient was an eight-year-old boy who suffered from an upper respiratory tract infection. One week after the onset of the symptoms he developed a torticollis with the head in a cock-robin position. Similar to the course of the first patient - after unsuccessful conservative outpatient treatment - the boy was transferred to our department approximately eight weeks after symptom onset. A CT scan confirmed the diagnosis of GS (Type Fielding I, Fig. 4). As immobilization, pain medication, and muscle relaxants did not reduce the AARS, closed reduction with halo vest immobilization for three months was performed. Several days after removing the halo vest, the boy moved his cervical spine without any restrictions, and further follow-ups were uneventful.

The third patient was a 14-year-old boy who presented due to a reduced capability of head movement. Examination revealed a slight facial scoliosis with his head held in a mild cock-robin position (Fig. 5). Rotation was possible up to 55° to the left side, 35° to the right side, extension up to 30°, and flexion up to 50°. The patient reported that he experienced the torticollis in connection with an uncomplicated pharyngitis approximately 18 months ago. In outpatient treatment he was given pain medication and antibiotics, but a relief was not achieved. After several weeks of persisting torticollis, chiropractic manipulation was performed. As this was an unpleasant memory, no further doctor was consulted. We assumed a chronic GS and initiated a CT scan, which showed a fixed forward subluxation of the atlas on the axis, with deformation and pseudarthrosis of the atlantoaxial joint (Fig. 5B). Interestingly, we also recognized the head in a nearly neutral position despite a 45-degree rotated atlas – due to a counter backward rotation of the skull. This finally led to the diagnosis of OAARD. There were no signs of infection of fracture. However, due to structural changes and resulting atlantoaxial pseudarthrosis, a closed reposition with halo vest was no promising option. We neither found clinical nor radiological signs of high-grade instability, as well as in the absence of neck pain, spinal stenosis, or basilar invagination, the patient in accordance with his family refused an open reduction with atlantoaxial (or occipito-atlantoaxial) fusion. However, in the short-term follow-up he had no complaint despite the known restricted cervical range of motion, the further development is uncertain.

Systematic review. Our search strategy identified 51 records. After reviewing titles and abstracts, 13 reports remained for full-text assessment. Following our inclusion and exclusion criteria, 9 reports were included in our analysis. Additionally, we reviewed five full texts resulting from citation searching, and 3 cases were added after searching our institutional database. Finally, 13 reports remained to be analyzed consisting of 12 case reports and one case series (Fig. 2) [17,18,22–31]. There are neither systematic reviews, nor meta-analyses, prospective or retrospective studies that have been published with respect to OAARD, thus, level of evidence was graded 4 with high-risk of bias.

In total, the 14 reported cases of OAARD consist of 9 female and 5 male patients with an average age of 12 years (range 8–17 years). All patients had GS prior to OAARD. In 10 patients (71 %) the diagnosis of GS was initially missed, in two patients GS was diagnosed after two months (14 %), and in another case the treatment of GS was unsuccessful. The assumed causes for GS included upper respiratory tract infection, surgery, juvenile idiopathic arthritis, trivial traumata and idiopathic genesis. Average time to diagnosis of OAARD was 5.9 months (range 0,5–18 months). At that time, all 14 patients presented with neck pain (100 %), eight patients showed mild CR position (57 %) and six patients (43 %) had a neutral head position. Neurologic symptoms were absent in 13 patients (93 %) with one patient suffering from idiopathic left C5-neuropathy. The treatment consisted of occipitoaxial (C0-C2) fusion in six cases (43 %), atlantoaxial fusion (C1-2) with halo vest in four cases (29 %), traction and halo vest in three cases (21 %) and one patient refused treatment (Table 2).

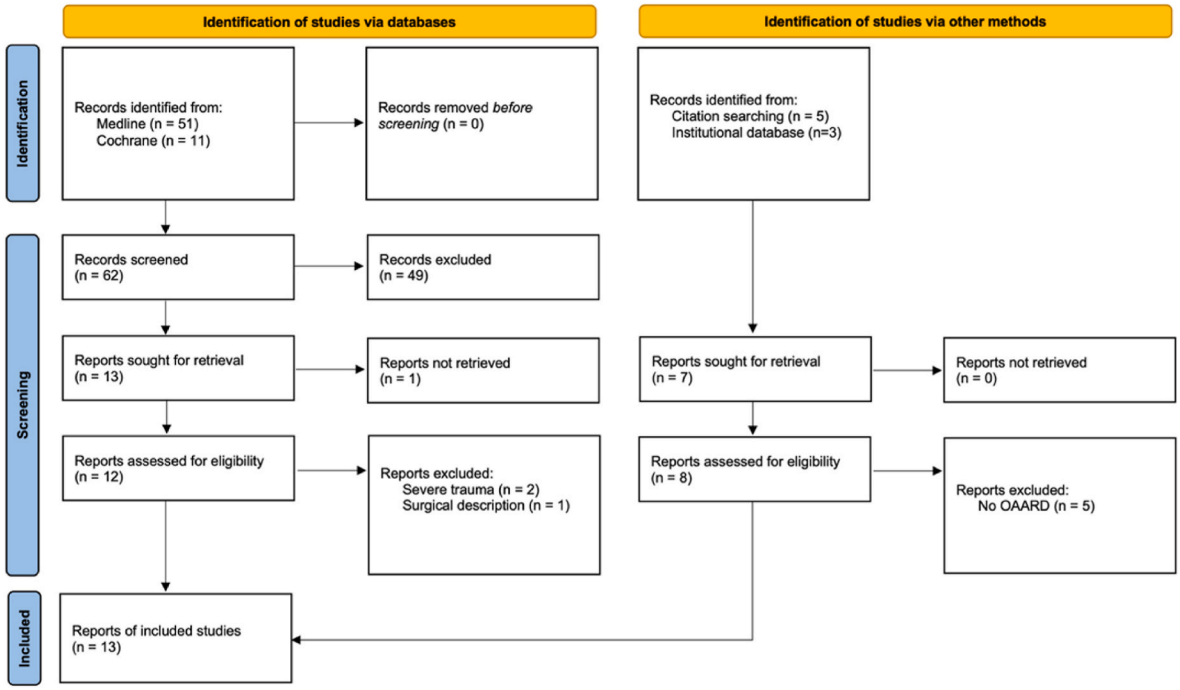

Fig. 2. PRISMA 2020 flow diagram. PRISMA flow diagram for new systematic reviews which included searches of databases, and other sources (from:http://www.prisma-statement.org).

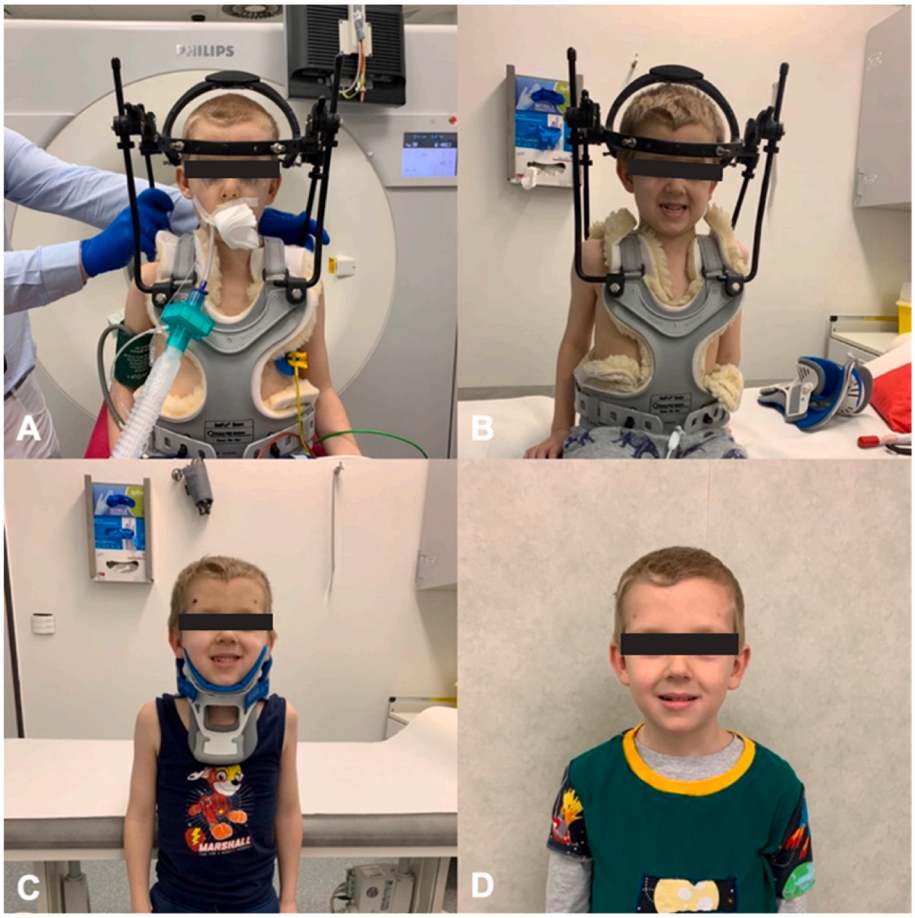

Fig. 3. Treatment of GS (same patient as in Fig. 1A**). A** After closed reduction, a halo vest was placed in general anesthesia. **B** Patient at clinical examination after 6 weeks. **C** Patient at follow-up wearing a hard collar. **D** Patient after 12 weeks.

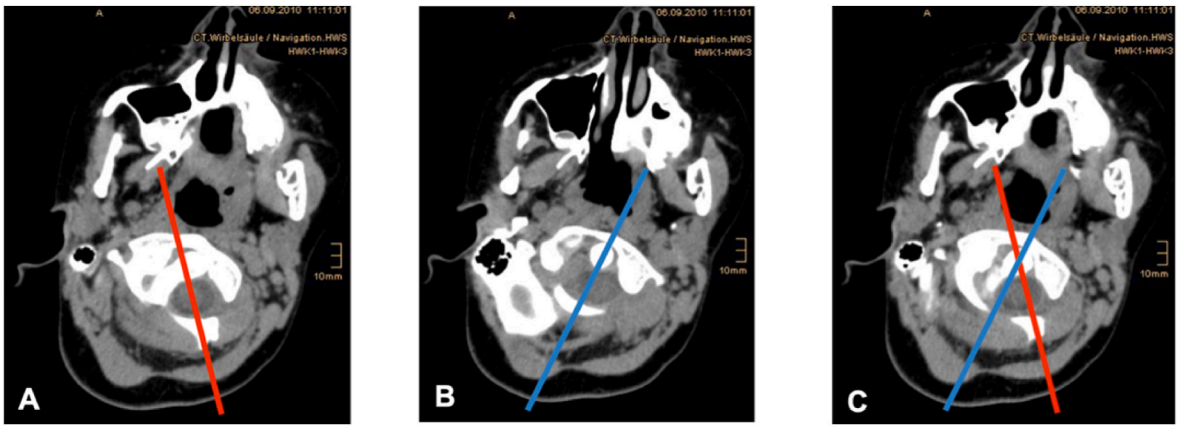

Fig. 4. CT scan of an 8-year-old boy with GS. A Cut at level of C2 (axis). The red line marks the sagittal plane at the middle of the axis. **B** C1 level (atlas). The blue line marks the sagittal plane of the middle of the atlas. **C** (Middle picture) Cut at level of atlantoaxial (C1/2) joint. The atlas is fixed in right rotation and left side-bending with contact of the odontoid to the ventral arch of the atlas (classified as Fielding and Hawkins Type I). The side of the rotation can be seen by the direction of the nasal septum. As the nasal septum is not in line with the direction of the atlas (blue line and axis of the nasal septum), and as there is maximum of 3° rotational movement in a physiological occipito-atlantal joint, an occipito-atlantal hypermobility, or slight occipito-atlantal counter rotation with beginning occipital condyle deformation (finally resulting in OAARD), can be expected. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

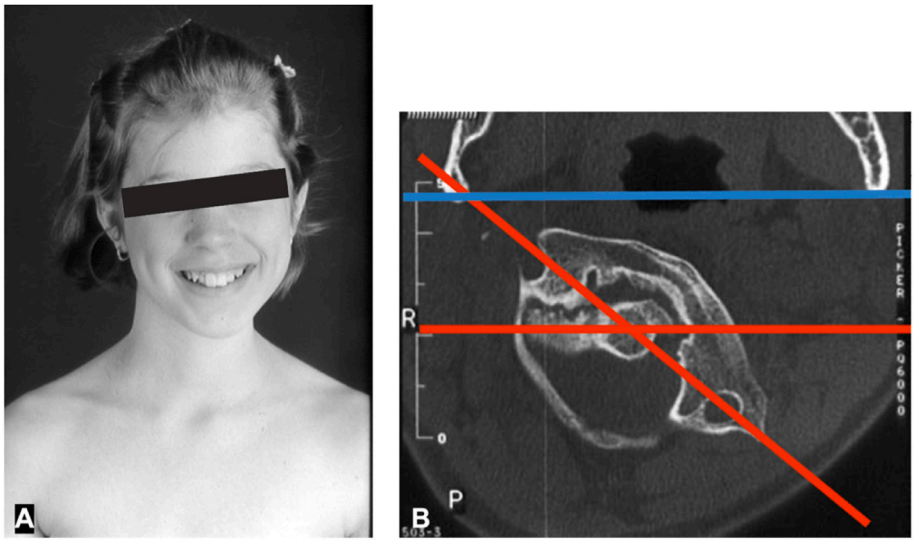

Fig. 5. Chronic GS resulting in OAARD. 14 years old boy with head in a mild "cock-robin" position. **A** Front view: a slight facial scoliosis can be observed when focusing on the axial deviation between the nose and the front teeth. This is a result from occipital counter rotation secondary to GS. **B** CT scan with at the C1/2 level (atlantoaxial joint). Fixed forward subluxation of the atlas on the axis (45°), with deformation and pseudarthrosis of the right atlantoaxial joint resulting from chronic GS (straight and oblique red line). The atlas line (oblique red line) is approximately 45° to the mandibular line (blue line). This is pathognomic for occipito-atlantal dislocation, leading to the diagnosis fixed OAARD with a head in a nearly neutral position - despite a subluxated atlas in fixed 45° rotation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

4. Discussion

GS is a rare disease in children. Throughout the years, since Bell, Grisel, and Watson-Jones sensitized for non-traumatic AARS, much has been written in terms of pathophysiology, conservative, and surgical treatment options [1,4,32,33]. On the other side, little is known in terms of OAARD [18]. This is the first study to systematically analyze the existing literature considering OAARD, to try to find a link between GS and occipital counter rotation resulting from GS.

Our main findings from our institutional cohort revealed that two children who underwent halo vest treatment after failed outpatient therapy for 6 and 8 weeks respectively, achieved favorable outcomes with restored head mobility. This highlights the efficacy of closed reduction in general anesthesia with halo vest immobilization in cases where conservative management is ineffective. The optimal surgical treatment strategy, after failure of conservative treatment of GS which should include antibiotics, pain medication, and a soft neck immobilization, is an often intensely debated question in clinical departments [34,35]. Our experience shows, that often-recommended halter traction for several weeks prior to closed reduction might not be necessary in most cases of GS. Rather, we believe that, after the failure of conservative therapy, prompt closed reduction in general anesthesia with halo vest immobilization should be considered the gold standard in the therapy of primary GS. This eliminates the need for immobilization for children for an unnecessary long period of time, and further promotes the remodeling of incipient atlantoaxial or occipitoatlantal joint changes.

In addition, within our institutional cohort we also encountered a case of a 14-year-old boy with OAARD after delayed diagnosis and insufficient treatment. This case underscores the potential long-term

Table 2
Published cases of patients treated with OAARD found by systematic literature analyisis. AARS atlantoaxial rotational subluxtion; C0 occiput, C1 atlas, C2 axis, CR cock-robin position; ENT ears, nose, and throat; f female; JIA juvenile idiopathic arthritis; m male; OAARD ocipito-atlantoaxial dislocation; post-op postoperatively; cRoM cervical range of motion.

Author	Sex/Age (years)	Assumed cause	AARS diagnosis initially missed	Duration of symptoms to diagnosis AARS/ OAARD	Clinical presentation (time OAARD diagnosed)	Treatment	Outcome (last follow-up)
Washington [17] (1959)	f, 11	upper respiratory tract infection	yes	5 months	neck pain: yes head position: neutral neurologic deficit: no	calot jacket + cervical brace	2 years after treatment: neutral position, cROM rotation 40°-0–40°, flexion/ext 45°-0–35°
Clark et al. [22] (1986)	f, 16	trivial trauma	yes	8 months	neck pain: yes head position: CR neurologic deficit: no	C0-C2 fusion	6 months post-op: pain free, 50 % cervical rotation and flexion
Altongy et al. [23] (1990)	m, 9	trivial trauma	no (diagnosis after two months)	2 months	neck pain: yes head position: CR neurologic deficit: no	C1/2 fusion + halo vest	36 months post-op: neutral position, cRoM rotation 70°-0–70°
Cowan et al. [24] (1996)	f, 13	ENT surgery	yes	9 months	neck pain: yes head position: CR neurologic deficit: no	C1/2 fusion	6 months post-op: residual rotation of the head, good cRoM
Hettiaratchy et al. [25] (1998)	f, 13	upper respiratory tract infection	yes	4 months	neck pain: yes head position: neutral neurologic deficit: no	C1/2 fusion + halo vest	3 months post-op: good reduction, 65 % cervical flexion/ext., 55 % rotation
Bouillot et al. [26] (1999)	f, 17	spontaneous torticollis, cervical manipulation	yes	0,5 months	neck pain: yes head position: mild CR neurologic deficit: no	C0-C2 fusion	Post-op: satisfactory reduction
Fusco et al. [27] (2011)	f, 8	JIA with recurrent cervical lymphadenopathy	yes	9 months	neck pain: yes Head position: neutral neurologic deficit: no	C0-C2 fusion	3 months post-op: 50 % of normal cRoM, fusion C0-2, no basilar invagination
Oshima et al. [28] (2012)	f, 11	spontaneous torticollis	no (diagnosis after two months)	2 months	neck pain: yes head position: mild CR neurologic deficit: no	Glisson traction + halo-vest	5-years after treatment: pain free, CT-scan: OAARD remained unchanged
Krengel et al. [29] (2015)	m, 11	spontaneous torticollis, cervical manipulation	yes	9 months	neck pain: yes head position: CR neurologic deficit: yes	halo vest with spontaneous C0-C2 fusion	6 months after treatment: mild CR left, left deltoid weakness improved, no severe pain
Kashii et al. [30] (2017)	f, 12	spontaneous torticollis	no (insufficient treatment)	6 months	neck pain: yes head position: neutral neurologic deficit: no	C0-C2 fusion	30 months post-op: “only a small restriction of neck motion”
Kashii et al. [30] (2017)	m, 13	clavicle fracture	yes	1,7 months	neck pain: yes head position: CR neurologic deficit: no	C0-C2 fusion	3-years post-op: “only a small restriction of neck motion”
Shimizu et al. [31] (2019)	m, 9	trivial trauma	no (but missed C0-condyle fracture)	4 months	neck pain: yes head position: neutral neurologic deficit: no	C0-C2 fusion	1-year post-op: pain free, cRoM rotation r/l 40°-0°-50°, extension 50°
Koljonen et al. [18] (2021)	f, 11	spontaneous torticollis	yes	not described (several weeks to month)	neck pain: yes head position: neutral neurologic deficit: no	C1/2 fusion + halo vest	1,5-years post op: pain free, cRoM rotation 60°-0–60°, C0/C1 remodeling
This report	m, 14	upper respiratory tract infection, cervical manipulation	yes	18 months	neck pain: yes head position: mild CR neurologic deficit: no	no further treatment	2 years after diagnosis: pain free, cRoM slightly limited in all directions

consequences of untreated GS and is in line with the findings of our systematic literature analysis: the average time to diagnosis in the literature was approximately 6 months, with persisting neck pain being the most common presenting symptom (100 %). Mild cock-robin position was observed in eight patients (57 %), while six patients (43 %) had a neutral head position caused by occipital counter rotation. Neurologic symptoms were absent in the majority of cases, except for one patient who presented with idiopathic left C5 neuropathy [29]. However, this palsy can be contributed to an outpatient chiropractic manipulation, e.g. considered as iatrogenic complication due to improper management of GS. We recognized that one out of four children had early chiropractic manipulation of the cervical spine, resulting in a negative experience without resolving GS, but potentially rather contributing to the development of OAARD (see Table 2). According to our review, the management for OAARD included occipito-axial fusion (C0-C2), atlantoaxial fusion (C1-2) with halo vest, or cervical traction and halo vest. Independent of the treatment performed, patients typically suffered from a persisting reduction range of cervical motion in the last follow-up.

The findings of our institutional cases together with the systematic review emphasize several important points. Firstly, early recognition and diagnosis of GS are crucial to prevent adverse events and improve outcomes. Prompt initiation of appropriate treatment can lead to successful resolution of symptoms and restoration of head mobility and function. Thus, a prerequisite for this is the knowledge of the condition and increased awareness among healthcare professionals in different medical specialties. Secondly, our results suggest that younger children have the potential of joint remodeling especially for the occipital condyle. This phenomenon was also observed by Kolionen et al. [18]. Therefore, we cannot recommend occipito-axial fusion in a first step, and if a surgery is chosen, atlantoaxial fusion with halo-vest treatment should be the treatment of choice to preserve the C0/1 joint function. Nevertheless, independently of the treatment modality chosen, patients and parents must be informed about permanent limitation in head movement due to loss of occipito-atlantal joint function, leading to an increased load of the lower cervical spine in the long-term. Despite one patient that suffered from concomitant asymptomatic basilar invagination, no case reported in the literature observed a severe neurological adverse event, and long-term reduced quality of life due to OAARD was not observed. Furthermore, our systematic literature review identified various causes for OAARD. These causes included upper respiratory tract infections, otolaryngological surgeries, juvenile idiopathic arthritis, trivial traumas, and idiopathic origins. The diverse etiologies underline the fact that GS can arise from multiple underlying factors, necessitating individualized treatment approaches based on the specific etiology and time after diagnosing. In cases following otolaryngological surgery, the pathogenesis of GS can be related to perioperative malposition, including hyperextension or rotation. However, GS often occurs only several days after surgery. This, together with the observation that children are more prone to GS than adults and that patients with congenital hypermobility of the craniovertebral joints – such as in trisomy syndromes – are at higher risk, supports the "two-hit hypothesis" proposed by Battiata and Pazos. According to this concept, increased joint laxity of the craniovertebral junction combined with inflammatory changes of the upper cervical ligaments represents the main risk factor for subluxation, rather than the surgical indication itself. However, especially patients with AARS arising from minor trauma or early, unspecific outpatient chiropractic manipulation seem to be at risk for OAARD – due to late diagnosing, or inappropriate treatment.

To summarize the answers to our initial questions, occipital counter rotation can be considered as natural course of untreated GS, ultimately to rebalance the eyes again. This leads to OAARD, with a naturally reduced cock-robin head position. Hereby, delayed diagnosis can be considered as the main risk factor for OAARD and a diminished clinical outcome. The most important complications include a permanent limitation of head movement and irreversible upper cervical joint deformation. Severe adverse events, such as neurologic deficits, or a reduced quality of life are extremely rare, even in non-treated patients. However, evidence for long-term complications following OAARD is missing, but premature degeneration of the cervical spine can be expected due to hypermobility of the adjacent joints. According to our experience, and supported by our literature analysis, the key factor in treating GS is an early interdisciplinary treatment with a prompt diagnose – to prevent OAARD and reduce the invasiveness of therapy. Further, in case of infection an early diagnosis enables the possibility of intravenous antibiotic treatment, which may reduce the failure of conservative attempts – if started within the first 2 week after onset of symptoms, as reported by Deichmüller and Welkoborsky [15]. Thus, the treatment decision should be made according to the causing pathology, with pain medication and soft collar immobilization as standard therapy accompanied by antibiotics whenever a bacterial genesis is possible. If GS is suspected, the diagnosis should be confirmed by CT scan, whereas MRI can provide additional information if abscess formation or basilar invagination is suspected [10]. We now can assume that patients with a delay of more than two month after onset of symptoms to diagnose and appropriate treatment can be considered to have an increased risk for OAARD.

It is important to acknowledge the limitations of our study. Due to the nature of rare pathologies, the relatively small number of cases limits the generalizability of our findings, as such we graded the level of evidence level 4. The retrospective nature of case reports according to OAARD, as well as the small number of retrospective studies concerning GS introduce confounding factors, implicating a high risk of bias. Additionally, long-term follow up studies of patients suffering from GS are rare, and in case of OAARD they are missing. It is possible that cases exist in which radiologically confirmed GS with AARS — i.e., not isolated torticollis due to muscular spasm — may spontaneously resolve without any form of treatment. The authors are not aware of such cases, and our study did not specifically search for them. Thus, future prospective studies with larger cohorts and long-time follow up are needed to validate our results and provide more robust evidence. Because of the rarity of GS and OAARD, this should be done across pediatricians, ENT, and spine centers.

5. Conclusion

Although GS is a rare disease and reliable data on incidence rates are lacking, misdiagnosis can lead to severe consequences. Based on a systematic literature review and our own experiences, this study emphasizes the importance of early diagnosis and appropriate treatment. Therefore, a painful torticollis following cervical inflammatory processes, otorhinolaryngological surgery, or minor trauma should be considered as cardinal sign of GS and treated as such until proven otherwise. This highlights the need to increase the awareness and sensitization especially among pediatric doctors and otorhinolaryngologists. Early interdisciplinary treatment is essential in order to restore normal head mobility and reduce the probability of irreversible upper cervical deformation such as atlantoaxial pseudarthrosis, spontaneous ankylosis, or OAARD - ultimately to prevent the need for atlantoaxial fusion or severe complications in these young patients.

CRediT authorship contribution statement

Vincent J. Heck: Writing – original draft, Methodology, Conceptualization. **Peer Eysel:** Writing – review & editing, Formal analysis, Conceptualization. **Tobias Prasse:** Writing – original draft, Methodology, Data curation. **Max J. Scheyerer:** Writing – review & editing, Visualization, Investigation, Conceptualization. **Katrin Eysel-Gosepath:** Writing – review & editing, Supervision, Methodology.

Data collection

All collected data are presented in the manuscript.

Declaration of generative AI in scientific writing

During the preparation of this work the authors used ChatGPT in order to improve the language by correction and identifying errors in spelling, punctuation, grammer, and phrasing. After using this tool the authors reviewed and edited the content as needed and take full responsibility for the content of the published article.

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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.

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