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# Non-traumatic Atlanto-axial Rotatory Subluxation- Grisel's Syndrome Case Report and Literature Review

Alqroom Y Rami<sup>1\*</sup>, Jeszenszky DJ<sup>2</sup>, Michalitsis G Sotirios<sup>3</sup> and Franke Jörg<sup>3</sup>

<sup>1</sup>Department of Neurosurgery, Royal Medical Services, King Hussein Medical Centre, Amman, Jordan

<sup>2</sup>Schulthess Clinic Spine Center, Zürich, Switzerland

<sup>3</sup>Spine Center, Hospital Klinikum Dortmund, Dortmund, Germany

# Abstract

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**Case Report** 

Study design: Case report of conservatively treated non-traumatic Atlanto-axial rotatory sublaxation.

**Objective**: To report our case with this rare condition (non-traumatic Atlanto-axial rotatory subluxation-Grisel Syndrome) in a 9-year old girl presenting with torticollis and neck pain and literature review.

Summary of background data: Atlanto-axial rotatory fixation or non-traumatic atlanto-axial subluxation which is usually secondary to an infection or an inflammation at the head and neck region is a rare subtype of subluxation in which a pathologic fixation of the atlas on the axis is present. It is most common in paediatric patients and it can be managed conservatively in the acute stage. In contrast, cases of chronic non-traumatic atlanto-axial rotatory subluxation are usually treated with operative reduction, according to literature data. Although high success rates have been achieved with operative reduction in chronic cases, a significant reduction in neck motion occurs due to the atlanto-axial fusion.

**Methods**: A 9-year-old girl developed torticollis established longer than 10 weeks as a result of an upper respiratory tract infection. Computed tomography showed a type 1 rotatory subluxation of the Atlanto-axial joint according to the classification of Fielding and Hawkins. The patient was treated by manual reduction according to Jeszenszky (Jeszenszky transoral Citation Nr 22) under general anaesthesia and fluoroscopy control, and then a halo vest had been applied for 6 weeks.

**Results**: We successfully treated this patient with chronic non-traumatic Atlanto-axial rotatory subluxation with manual reduction and immobilisation by applying a halo-body-jacket. She showed full recovery of neck motion and normal Atlanto-axial angle on clinical and radiological follow-up after 1 month and 3 months respectively.

**Conclusion**: Chronic non-traumatic Atlanto-axial rotatory subluxation is a rare condition which can easily escape the attention of physicians especially in children. Conservative treatment with complete reduction under anaesthesia and subsequent immobilisation via halo-body-jacket for at least 6 weeks is a viable option as presented in this case report. Surgical treatment should be reserved only for cases of failure of conservative management (recurrence or irreducible subluxation).

**Keywords:** Grisel Syndrome; Non-traumatic Atlanto-axial rotatory subluxation; Atlanto-axial rotatory fixation; Conservative management

## Introduction

Grisel's syndrome which is also known as the Non-traumatic Atlanto-axial Rotatory Subluxation (NAAS) or Atlanto-axial Rotatory Subluxation (AARS) without trauma or concomitant bone pathology, was first described by Sir Charles Bell in 1830 in a patient with syphilis and pharyngitis, who developed lethal outcome due to spinal compression [1,2]. The syndrome was named after Grisel who described two cases of pharyngitis and atlanto-axial subluxation in 1951 [1,2]. However, Grisel's syndrome is an uncommon condition of uncertain aetiology characterized by a non-traumatic Atlanto-axial rotatory subluxation, usually seen in children secondary to an infection and inflammation in the head and neck region or otolaryngeal procedures [1,3]. Patients generally complain about neck stiffness and pain, and sometimes dysphagia may occur. Diagnosis is established based on the clinical and radiological findings [1,4].

The etiopathogenesis and the underlying pathomechanics have not been clearly explained. A haematogenous spread of infection from the posterior pharynx to the cervical spine, according to the recent literature, with hyperaemia and abnormal relaxation of the Atlanto-axial ligaments is a widely accepted theory [1,5]. The vascular plexus providing the drainage of poster superior pharyngeal area is responsible. The periodontoid plexus is connected with posterior nasopharyngeal veins via the pharyngovertebral vein. Any infective embolism may spread from superior pharyngeal area to upper cervical joints due to this plexus which has not any lymph node thus providing an anatomical explanation for the Atlanto-axial hyperaemia reported in Grisel's syndrome.

## **Case Report**

A 9-year-old female patient was referred to our out-patient clinic, having complained of neck pain and stiffness for the last 10 weeks. No history of trauma was mentioned. The parents mentioned an upper respiratory tract infection 3-4 weeks previously to the development of this new condition. The general neurological examination did not reveal any focal deficits and the otorhinolaringeal examination was at that time normal. The range of motion in her neck was markedly limited, and the girl suffered from severe neck pain and stiffness.

The clinical and laboratory examination revealed: There was no fever, the erythrocyte sedimentation rate (ESR) was 69 mm/h, the

\*Corresponding author: Rami Alqroom Y, M.D., Spine surgeon, Department of Neurosurgery, Royal Medical Services, King Hussein Medical Center, Amman, Jordan, Tel: 00962797017870; Fax: 004923195321020; E-mail: dr.alqroomrami@gmail.com

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C-reactive protein (CRP) was 24.98 mg/dl, and the white blood cell count (WBC) was 7.100 U/ml. All other laboratory tests were normal. Repeated oropharyngeal cultures showed normal flora.

The radiological examination showed: Anterior view of the cervical radiography revealed tilted position of the head over neck (Figure 1).

Lateral view showed thickening of the Para pharyngeal soft tissue (Figure 2). On the computed tomography (CT), head in neutral position, the distance between the axis and dens was within normal values for age (ADI= 2-4 mm) (Figure 3).

The CT scan with 3D reconstruction view showed 30 degree of rotation (Figure 4). This is confirming our diagnosis of type 1 Atlantoaxial subluxation according to Fielding & Hawkins classification. We have managed our patient using the Jeszenszky transoral closed reduction technique (after his personal consultation) which consists of locking the spinous process of the axis with the left hand (between thumb and index finger) while the index finger of the other hand presses in counter rotation to the dislocation on the lateral mass of the atlas through the posterior wall of the pharynx (Figure 5).

After confirmation of the reduction using the CT scan, while the patient was still under anaesthesia (Figure 6) (a- coronal section, b-axial section) which showed the full reduction just after the manoeuvre, the halo ring and vest was placed and the reduced position in neutral position maintained. She has been followed-up clinically at 0, 1 week, 4weeks, and then at 6 weeks intervals. At the 6 weeks clinical follow-up we performed a cervical radiograph (Figure 7) (a- anterior-posterior









Figure 4: CT scan, 3D reconstruction view, showing more than 30 degree of atlanto-axial rotational subluxation.



**Figure 5:** Sketch of transoral closed according to Jeszenszky. The image illustrates that the spinous process of the axis is locked by one hand, while the index finger of the other hand presses in counter rotation to the dislocation on the lateral mass of the atlas through the posterior wall of the pharynx.

view, b- lateral view) and due to the excellent position we decided to remove the halo and put the patient on a soft collar for another 4 weeks.

The last radiographic follow-up so far, 12 weeks after the removal of the halo, showed a sustained reduction as demonstrated with (Figure 8) (cervical radiograph: a- anterior-posterior view, b- lateral view, c-flexion , d- extension). The girl was fully pain free and had a normal range of motion of her cervical spine.

## Discussion

Grisel syndrome refers to concurrence of non-traumatic Atlantoaxial joint rotatory subluxation with infectious, inflammatory processes such as Familial Mediterranean Fever or after an otolaryngological procedure, including tonsillectomy, adenoidectomy, mastoidectomy, choanal atresia repair, and cleft palate repair of the head and neck region [5]. The etiopathogenesis of this clinical entity is not clear yet [6,7]. Citation: Algroom RY, Jeszenszky DJ, Michalitsis GS, Franke J (2016) Non-traumatic Atlanto-axial Rotatory Subluxation- Grisel's Syndrome Case Report and Literature Review. J Spine 5: 312. doi:10.4172/2165-7939.1000312

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Figure 6: CT scan, (a) coronal section, (b) axial section showing post manipulation reduction.



Figure 7: cervical radiograph, (a) anterior-posterior view, (b) lateral view, with halo vest, 6 weeks later.



Figure 8: cervical radiograph, (a) anterior-posterior view, (b) lateral view, (C) flexion, (d) extension, and 3 months later after Halo removal and 5 months after the initial reduction

Systemic review conducted by Karkos et al, identified 103 patients with Grisel syndrome published in seventy-one papers between 1950 and 2006. They concluded that the main causes of Grisel's syndrome were infection (48%) and post-adenotonsillectomy (31%). Less common causes included other postoperative cases such as pharyngoplasty and ear operations [8,9].

The fundamental primary stabilizer of the Atlanto-axial joint is the transverse ligament and as secondary stabilizers are the alar ligaments in addition to joint capsule [6,8]. High-risk group of patients for developing Atlanto-axial subluxation are children with greater laxity of Atlanto-axial ligaments and increased Atlanto-axial space such as children with Down's syndrome, Klippel-Feil syndrome, osteogenesis imperfecta, neurofibromatosis and any syndromes associated with spinal instability. Although, Atlanto-axial subluxation may also occur in non-syndromic cases with no underlying spinal laxity and early diagnosis is required to prevent significant morbidity and/or mortality [10-12]. However, Martínez-Lage et al. [13] reported a child who presented with clinical manifestations of Grisel's syndrome but had a C2-C3 subluxation.

The underlying mechanism has not been clearly explicated yet, but the most valid theory is the haematogenous spread of infection from the posterior pharynx to the cervical spine. According to the recent literature, hyperaemia and abnormal relaxation of the Atlanto-axial ligaments, is a widely accepted theory [1,6]. Any infective embolism may spread from superior pharyngeal area to upper cervical joints due to this plexus which lacks lymph nodes thus providing an anatomical explanation for the Atlanto-axial hyperaemia reported in Grisel's syndrome [14].

One other theory proposed by Tedesco et al. suggests that a cervical lymphadenitis caused by a nasopharyngeal infection may lead to irritative spastic contraction of sub-occipital and paravertebral muscles causing torticollis [10,15].

History and clinical signs are fundamental hints for the diagnosis of Grisel's syndrome. For patients with painful head tilt and restricted neck movements in addition to a previous history of an upper respiratory tract infection, the suspicion for the diagnosis should be raised. Patients generally complain about progressive throat and neck pain at attempted motion. The nuchal pain radiates to the head or ears, and the head is held immobile in a pathological position (known as the "cock-robin" position) in which the head is tilted and the chin is rotated away from the affected side (Figure 1).

Radiological control with simple X-ray may indicate the condition and computed tomography scans of the cervical spine will establish the diagnosis. Nevertheless, the gold standard exam for the diagnosis of Atlanto-axial rotatory subluxation is the CT scan with 3D reconstruction. The 3D- CT scan permits a clear visualization of C1–C2 rotation (Figure 4), including the loss of congruence of the articular facets. The sagittal/axial images give the ability to measure the ADI, also to evaluate the location of the dens in relation to the atlas, thus to give us better hints regarding the grade of subluxation according to Fielding and Hawkins (Figure 3). Early management-consisting of cervical immobilisation, medical treatment, and physiotherapy- was considered satisfactory till recently [16]. Inappropriately handled cases may result in a fixed and painful neck deformity that may even require surgical fusion [17].

Fielding and Hawkins had described 4 types of rotatory Atlantoaxial subluxations. The first type is the rotation of atlas above axis without anterior displacement. The second type is the rotation of atlas above lateral reticular process with 3–5 mm anterior displacement. The third type is the rotation of atlas with anterior dislocation more than 5 mm and the fourth type is the rotation of atlas with posterior dislocation [18]. Types 1 and 2 of subluxation are most common and associated with no neurological symptoms. Our patient had type one Atlanto-axial subluxation.

In our case differential diagnosis had included tumours of the posterior fossa and spinal Cord, cervical spine trauma, ocular and vestibular disorders, and dystonic torticollis associated with drugs side effects which were all excluded. The management of this condition still poses a controversial issue, especially in chronic cases.

Principles of management include: (a) confirmation of primary pathology with bacteriological diagnosis and appropriate management (b) reduction of the osseous deformity, then appropriate assurance of reduction with collar or vest (c) prevention of neurological damage.

Initial treatment in acute cases in the agreement of many authors is usually conservative, including bed rest, a cervical collar, analgesics, anti-inflammatory medications, muscle relaxants and, antibiotic therapy, when appropriate, also abscess drainage [8,19]. Barcelos et al. has recommended the use of a soft cervical collar molded to the height of the neck at acute phase due to cervical posture which prevents the use of a rigid cervical collar. As greater relaxation of the cervical muscles occurs, the authors recommend increasing the height of the collar until reduction of the lesion occurs. Then, the soft collar is replaced with a rigid one. As there is no agreement regarding the waiting and follow-up time, they recommend a rigid collar for at least 2 weeks to treat pain and also to prevent early recurrence. A return to normal life activities and sports after 4 to 6weeks is suggested [8]. However, most cases usually resolve spontaneously up to 7 days after the beginning of treatment [20]. Once the luxation is reduced and the inflammatory process is completely solved, stability recovers.

What about the case where no spontaneous reduction of the lesion occurs, or in case of late diagnosis?! In fact, closed reduction under general anaesthesia is recommended, using Jeszenszky transoral reduction technique. Definitive treatment should be established after reduction [16,21,22]. Here we would like to emphasize the fact that the most important step is to apply rigid retention and immobilization such as a Halo vest [19].

One treatment protocol for non-traumatic Atlanto-axial rotatory subluxation (Grisel syndrome) was proposed by Wetzel and La Rocca which was based on the classification of Fielding and Hawkins. According to this protocol: Conservative treatment for type I is a soft collar, for type II a rigid collar, and a halo-vest for type III lesions. For type IV lesions, they recommend surgical treatment [23].

Park et al reported successful reduction for a paediatric chronic Atlanto-axial rotatory fixation (Grisel syndrome) with long-term immobilisation-traction [21,24]. Surgical treatment is reserved and indicated in cases of failure of conservative treatment, recurrence of subluxation and irreducible subluxations, although, according to Jeszenszky, rarely needed [19].

We have managed and reported a case of a symptomatic patient with a chronic Atlanto-axial subluxation in after an upper respiratory tract infection resulting in inflammation of the ligaments and joints of Atlanto-axial region. The patient was managed by manual closed reduction under general anaesthesia and dynamic CT control followed by Halo vest for 6 weeks (Figure 7).

#### Conclusion

Non-traumatic Atlanto-axial rotatory subluxation is a rare but major complication that can often go unnoticed in its early phase and which can be a major cause of morbidity following upper respiratory tract infections, head and neck procedures and/or any head and neck surgeries thus early recognition and diagnosis is mandatory especially in children. Conservative treatment is a viable option in children even in chronic cases. Surgical treatment should be reserved only for cases of failure of conservative management, in case of recurrences or irreducible subluxation.

#### **Declaration of Conflicting Interests**

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

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