

Eponym

Grisel syndrome

Graciela Ortega-Evangelio · Jose Juan Alcon ·
Julio Alvarez-Pitti · Vicente Sebastia · Maria Juncos ·
Empar Lurbe

Received: 8 April 2011 / Accepted: 4 May 2011 / Published online: 24 May 2011
© Springer-Verlag 2011

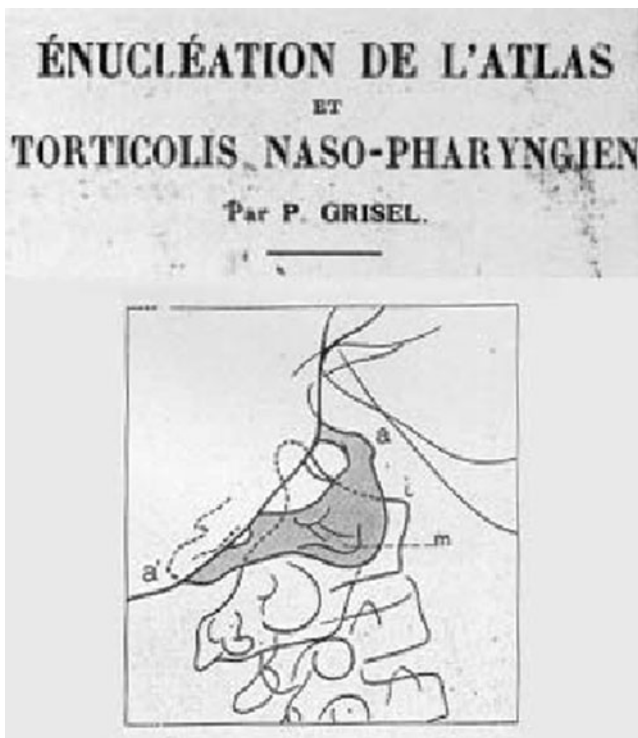


Fig. 1 Title of the paper published by Grisel in 1930, and the scheme that he drew to represent the pathology; *i* right articular surface angle, *a* and *a'* atlas anterior and posterior apophysis, *m* axis lateral body pushed

Abstract Grisel syndrome or atlantoaxial non-traumatic subluxation is a rare entity, poorly described in pediatric literature, although it is a pathology that usually appears in young children. The typical presentation is a torticollis with a previous surgical antecedent (mainly a surgery of the ear, nose, and throat area like adenoidectomy) or after an upper tract respiratory infection. A prompt diagnosis is essential for a successful evolution, and the treatment in most cases is conservative. We report a case of an 8-year-old girl with a typical evolution of this unusual complication. In our case, the most important element for diagnosis was the 3D CT scanner, and the treatment was conservative with a successful outcome. Pediatricians should be aware of this rare but potentially serious entity to establish the correct treatment and avoid complications.

Keywords Grisel syndrome · Torticollis · Atlantoaxial subluxation

Introduction and historical overview of the disorder

Grisel syndrome or atlantoaxial non-traumatic subluxation is a rare entity, poorly described in pediatric literature, although it is a pathology that usually appears in young children. As Patterson and Little described in 1943 [15], the spasmodic torticollis is an involuntary hyperkinesia which manifests as mobile, tonic, or clonic spasm of the neck musculature, and produces a more or less involuntary turn of the head into an abnormal position, the chin is rotated to one side or the head bends directly forward (antecollis) or backward (retrocollis). This is a frequent pathology in pediatric emergency and is usually associated with an abnormal position during sleep or mild traumas. As well as this typical cause, there are more than 80 different causes of torticollis including some serious and life-threatening forms. This is the reason that in a persistent torticollis, a complete study is necessary to discover the cause and reach a correct diagnosis. Grisel syndrome is the spontaneous and non-

G. Ortega-Evangelio (✉) · J. J. Alcon · J. Alvarez-Pitti ·
M. Juncos · E. Lurbe
Department of Pediatrics,
Consortio Hospital General Universitario,
Valencia, Spain
e-mail: graciela_ortega@hotmail.com

V. Sebastia
Department of Physical Medicine and Rehabilitation,
Hospital Universitario La Fe,
Valencia, Spain

traumatic subluxation of the atlantoaxial joint following periharyngeal inflammation. It was first described by Sir Charles Bell [2] in 1830. Bell reported a case of death after an atlantoaxial subluxation which resulted in spinal cord compression in a patient with a syphilitic ulceration of the pharynx. The autopsy showed erosion of the transverse ligament of the axis. The eponymous name has its origin in the French physician otolaryngologist P. Grisel who described two cases in 1930, one of them after a nasopharyngitis [9]. A picture from an original article is attached, which is drawn by himself, in which (Fig. 1) he described the anatomic changes associated with the syndrome. Nowadays, this rare syndrome has been described in ENT, neurosurgery, or orthopedic literature, but it is rare in pediatrics, even though it mainly affects the infant population (68% are patients younger than 12 years old, and 90% are younger than 21 years old) [19]. In most cases, there is a surgical (for example mastoidectomy, adenoidectomy, or tonsillectomy) or infectious history (usually pharyngitis, otitis, or mastoiditis). According to Deichmueller and Welkoborsky [5], the most frequent cause of Grisel syndrome is surgery (in his experience 67% was after surgery). On the other hand, Karkos et al. [11, 5], in a review article which analyzed all the Grisel syndromes that were published from 1950 to 2005 (with a total of 103 cases), found that 48% was caused by infections, and surgery was the second cause with 40% of the cases. It has also been described after impact with a hard object, and the subsequent complication of a retropharyngeal abscess. The subluxation usually appears after a previous period of torticollis and fever. This period, with rotational deformity but without subluxation, is of great importance [14] because it is during this initial period of time when conservative therapy can be undergone (analgesia, immobilization, and rest). If the muscle spasm persists over 24 h, diazepam can be added to the treatment until improvement. This early management could avoid bad progression. If it is not treated promptly, subluxation may develop. The pathogenesis of the process is controversial. There are two undeniable facts: first, there is a higher frequency of presentation in pediatric population (children present higher ligament laxity between C1–C2 and especially the crossed ligament) and second, usually, presentation is after important infectious or inflammatory processes. Both facts cause ligament hypermobility with distension and abnormal laxity of ligaments surrounding the articulation, resulting from the direct spread of inflammation from ENT area, which joined to the muscle spasm, may cause the subluxation [1, 3, 6, 17]. The laxity of ligaments can explain the association in reported cases with Down syndrome and Marfan syndrome [10, 12, 16]. Other high-risk groups include Klippel–Feil syndrome, osteogenesis imperfecta, neurofibromatosis, and any syndromes associated with spinal instability [11]. Other pathologies with similar symptoms must always be ruled out, like cervical bone abnormalities, posterior fossa tumors, or cervical trauma.

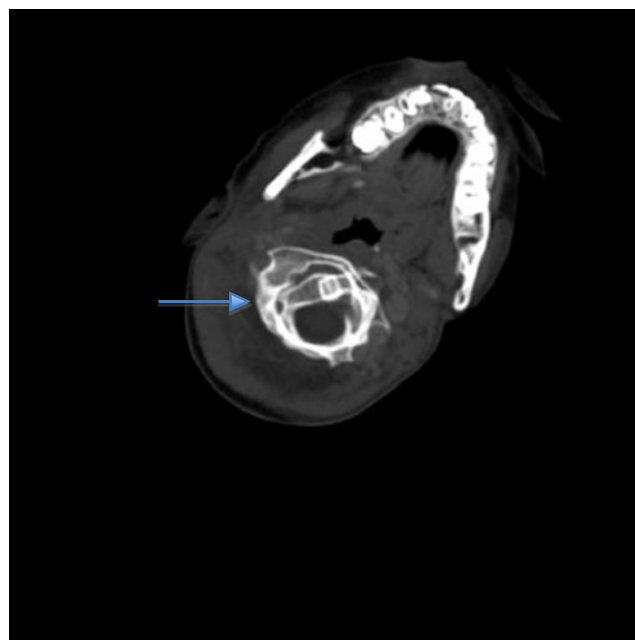


Fig. 2 CT image, rotatory atlantoaxial subluxation, with a twisting C1 over C2, making an angle of 38°. There was no anterior displacement, and the atlantoaxial space was less than 3 mm

Clinical presentation: case report

An 8-year-old girl was treated at the pediatric emergency area for a torticollis with an 8-day-history of painful cervical deviation without improvement with usual analgesic treatment. She had suffered an acute pharyngitis the week before, which was initially treated with azithromycin. The antibiotic was changed to amoxicillin–clavulanic due to the persistence of fever after 3 days of treatment. No history of trauma was

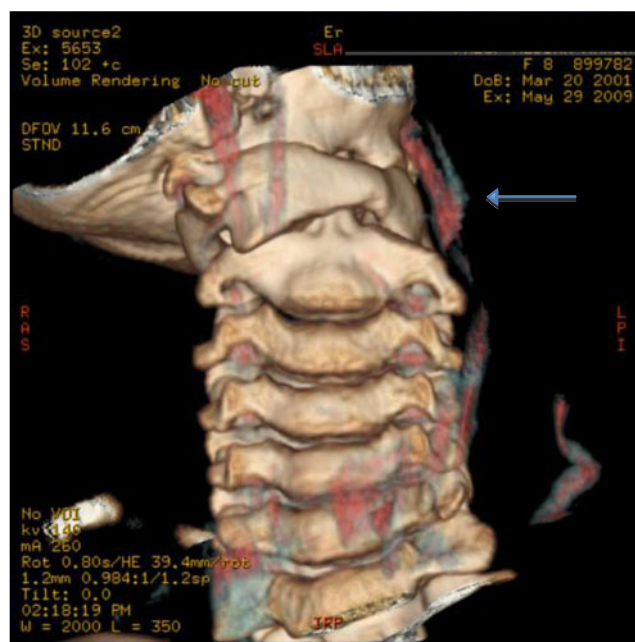


Fig. 3 Three-dimensional CT. Posterior view

Table 1 Diagnosis of Grisel syndrome

Diagnosis of Grisel syndrome
1. History of ENT surgery/infection
2. Torticollis after [1] with torpid evolution
3. Trans-oral or latero-cervical plain radiograph
4. CT scan with or without three-dimensional study

ENT eyes, nose, and throat

reported. In the physical examination, a right cervical twist was evident with an important muscle spasm, as well as bilateral and painful lymphadenopathies. No other alterations were found. The cervical ultrasound scan reported non-confluent inflammatory lymphadenopathies, all of them smaller than 1 cm. The cell blood count, C-reactive protein, and biochemical parameters were normal. The cervical CT scanner showed a rotatory atlantoaxial subluxation, with a twisting C1 over C2 (Figs. 2 and 3), making an angle of 38°. There was no anterior displacement, and the atlantoaxial space was less than 3 mm. Treatment was started with cervical immobilization, analgesia with ibuprofen and nightly diazepam during 5 days and rest. The outcome was successful with complete recovery after 2 weeks.

Diagnostic work-up

This syndrome may be found through clinical diagnosis; however, the confirmation is always radiological. The first

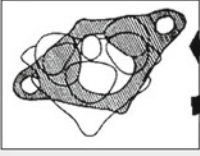
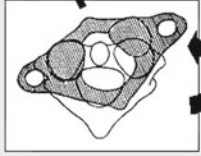
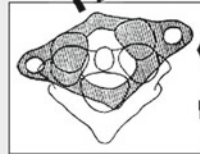
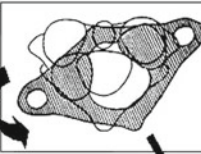
exploration may be a transoral or laterocervical plain radiograph (this can classify the stage according to rotational deformity) and later a cervical CT scanner with or without a three-dimensional study [4]. We performed a three-dimensional reconstruction CT, which gave us the diagnosis, classified the case, and helped to establish the therapeutic protocol (Table 1). The classification of Grisel syndrome according to Fielding [7,8], depends on the degree of the atlas above the axis displacement, and this can be discovered through radiological study (Table 2).

Treatment

The management of this condition is not universally accepted. Several authors (Battiata et al.) [1], consider a conservative treatment for type I (immobilization, antibiotics, rest, and analgesia), as well as for type II (including cervical traction or muscle relaxants) using a more invasive treatment in types III and IV (halo immobilization, arthrodesis, and C1–C2 cervical fusion). Wetzel and Larocca [18] propose another treatment algorithm: type I soft collar, type II rigid collar, type III close fixation with halo, and type IV open fixation with halo. Nevertheless, these algorithms may be adapted in each case, and patients should be controlled and managed individually.

The correct management of Grisel syndrome is based on three aspects: specific treatment of the infectious disease, correction of the bone deformity, and prevention of

Table 2 Fielding classification. Table reprinted with permission from [3]

		Type I	Simple rotatory displacement without anterior shift. The transverse ligament is intact and the dens acts as a pivot point.
		Type II	Rotatory and anterior displacement between 3-5 mm. The transverse ligament is injured and the opposite facet acts as the pivot point.
		Type III	Rotatory and anterior displacement greater than 5 mm with both lateral atlantoaxial joints anteriorly subluxed. The transverse and alar ligaments are injured.
		Type IV	subluxation of both lateral atlantoaxial joints.

neurological damage. Also, neurosurgical consultation is paramount in all cases, even in mild ones [11].

In our case, there was no anterior displacement, and the atlantoaxial space was less than 3 mm so it could be considered as a type I subluxation in Fielding classification (Fig. 3 and Table 2). Following the recommendations, we admitted the patient to the hospital for 5 days, resting. We applied a soft collar to cause cervical immobilization, ibuprofen as analgesic and anti-inflammatory, and we also added diazepam as muscle relaxant medication. As the girl had been treated with antibiotics over the previous 7 days, we decided not to continue them. The patient was followed-up in the outpatient clinic for 2 weeks until complete recovery.

Prognosis

When treatment is effective and correct, the prognosis is excellent. Early subluxation (Fielding types I and II) is not typically associated with neurologic impairment; Fielding type III and IV lesions have been reported to have a 15% incidence of neurologic impairment ranging from radiculopathy to paralysis and death [13]. Most of the cases reported in the current literature, have a successful outcome with conservative treatment, including our patient. Without treatment, the atlantoaxial subluxation becomes permanent, and will result in chronic torticollis, with the possibility of affection of facial flattening and neurological complications.

Outcome

It is important to highlight that this syndrome can develop a catastrophic progression, so an early diagnosis is essential to avoid the presentation of complications (neurologic and functional deficits, great aesthetic deformities, and paralysis secondary to medullar compression). Morbidity is significant in those cases where diagnosis is delayed, with the most devastating consequence being a permanent neurological deficit. However, if the disease is identified and promptly treated, the outcome in the majority of cases will be successful

Key points

1. The main causes of Grisel syndrome are infection (48%) and post-adenotonsillectomy (31%).
2. Less common causes include other postoperative cases such as pharyngoplasty and ear operations.
3. Neurosurgical consultation is paramount in all cases, even the mild ones.
4. In the majority of cases, conservative management in the form of bed rest, antibiotics, muscle relaxants, and collar with or without traction, is effective.

5. In a few cases only, surgery in the form of arthrodesis is deemed necessary.
6. Morbidity is significant in those cases where diagnosis is delayed, with the most devastating consequence, a permanent neurological deficit.

Acknowledgments The authors gratefully acknowledge the assistance of Rachael Dix in manuscript's edition.

References

1. Battiatà AP, Pazos G (2004) Grisel syndrome: the two-hit hypothesis—a case report and literature review. *Ear Nose Throat J* 83:553–555
2. Bell C (1830) *The nervous system of the human body: embracing papers delivered to the Royal Society on the subjects of nerves*. Longman, Rees, and Orme, London, p 403
3. Boccionlini C, Dall'olio D et al (2005) Grisel's syndrome: a rare complication following adenoidectomy. *Acta Otorhinolaryngol Ital* 25:245–249
4. Cowan TA, Inglis GS (1996) Atlanto-axial rotatory fixation: improved demonstration using spiral CT. *Australas Radiol* 40:119–124
5. Deichmueller CMC, Welkoborsky HJ (2010) Grisel's syndrome—a rare complication following “sml” operations and infections in the ENT region. *Eur Arch Otorhinolaryngol* 267:1467–1473
6. Feldmann H, Meister EF, Kuttner K (2003) From the experts office. Atlanto-axial subluxation with spastic tortico resp. tonsillectomy in rose position—malpractice surgeon or anesthesiologist? *Laryngorhinootologie* 82:790–804
7. Fielding JW, Hawkins RJ (1977) Atlanto-axial rotatory fixation (fixed rotatory subluxation of the atlanto-axial joint). *J Bone Joint Surg* 59A:37–44
8. Fielding JW, Hawkins RJ, Hensinger RN, Francis WR (1978) Atlanto-axial rotator deformities. *Orthop Clin North Am* 9:955–967
9. Grisel P (1930) Eucleation de l'atlas et torticollis nasopharyngen. *Presse Méd* 38:50
10. Herzka A, Sponseller PD, Pyeritz RE (2000) Atlanto-axial rotatory subluxation in patients with Marfan syndrome. A report of three cases. *Spine* 25(4):524–526
11. Karkos PD, Benton J, Leong SC, Mushi E, Sivaji N, Assimakopoulos DA (2007) Grisel's syndrome in otolaryngology: a systematic review. *Int J Pediatr Otorhinolaryngol* 71:1823–1827
12. Martínez L, Morales T, Comejo EF (2002) Inflammatory C2-C3 subluxation: a Grysel syndrome variant. *Laryngoscope* 112:1445–1449
13. Meek MF, Hermens RAEC, Robinson PH (2001) La maladie de Grisel: spontaneous atlantoaxial subluxation. *Cleft Palate-Craniofac J* 38(3):268–270
14. Mezue WC, Taha ZM, Bashir EM (2002) Fever and acquired torticollis in hospitalized children. *J Laryngol Otol* 116:280–284
15. Patterson R, Little S (1943) Spasmodic torticollis. *J Nerv Ment Dis* 98:571–599
16. Tseng SH, Cheng Y (1998) Occiput-cervical fusion for symptomatic atlantoaxial subluxation in a 32-month-child with Down syndrome: a case report. *Spinal Cord* 36(7):520–522
17. Welinder NR, Hoffmann P, Hakansson S (1997) Pathogenesis of non-traumatic atlanto-axial subluxation (Grisel's syndrome). *Eur Arch Otorhinolaryngol* 254:251–254
18. Wetzel FD, LaRocca H (1989) Grisel's syndrome: a review. *Clin Orthop Relat Res* 240:141–152
19. Wilson BC, Jarvis BL, Haydon RC III (1987) Nontraumatic subluxation of the atloaxial joint: Grisel's syndrome. *Ann Otol Rhinol Laryngol* 96:705–708