A novel technique to treat Grisel's syndrome: results of a simplified, therapeutical algorithm

Hakan Pilge · Boris Michael Holzapfel · Renee Lampe ·
Stefanie Pilge · Peter Michael Prodinger

Received: 7 January 2013 / Accepted: 3 April 2013 / Published online: 9 May 2013
© Springer-Verlag Berlin Heidelberg 2013

Abstract

Purpose Grisel's syndrome is a rare entity and usually develops in paediatric patients after otolaryngologic surgery or infection. It is defined as a fixed torticollis combined with a rotatory atlanto-axial subluxation. The success rate of physiotherapy is low. Conventional therapy concepts imply stage-related recommendations based on the Fielding classification (type I–IV). This classification was introduced in 1977 to assess the degree of subluxation between atlas (C1) and axis (C2). Thus, instability increases from type I to IV. Higher stages may require surgical intervention. The purpose of this study was to evaluate the value of an alternative, less invasive treatment protocol in Grisel's syndrome.

Methods Irrespective of the underlying Fielding type we treated five children (Fielding type I–III) by manual repositioning under general anaesthesia. Consecutively, the cervical spine was immobilized with a Minerva cast for four to eight weeks. Additional surgical treatment or immobilization in a Halo-Fixateur was not necessary.

Results Overall period of treatment was reduced, even in patients with delayed diagnosis. No case of recurrence was observed within a follow-up of six months.

Conclusions Current conventional recommendations suggest invasive treatment with Halo-Fixateur in patients with higher degrees of subluxation (e.g., Fielding type III) or after delayed diagnosis. Even in those patients, this novel therapy concept enables us to achieve excellent clinical results without surgical intervention.

Introduction

Grisel's syndrome is a very rare disease predominantly found in paediatric patients. It is characterized by a non-traumatic subluxation between atlas (C1) and axis (C2) leading to a painful torticollis. Onset of symptoms occurs usually in chronologically direct connection with a precedent otolaryngological intervention or infection after a symptom-free interval. Patients typically present with the following clinical signs: torticollis with the head in an oblique position, elevated ipsilateral muscular tension and stiffness of the sternocleidomastoid muscle.

Usually children under the age of 12 are affected, without any gender-specific predisposition [1, 2]. Manifestation in adults is exceptionally rare [3].

Pathogenesis of Grisel's syndrome remains unclear but there is virtually unanimous agreement that it is related to a previous otorhinolaryngologic intervention or infection. In an extensive literature review it was found that nearly every case reported was associated with either an infection of the ear, nose or throat, or a surgical intervention [4–6]. In 48 % of these cases a local infection was reported, whereas in 31 % the symptoms occurred after adenotonsillectomy. Less commonly, a history of local surgical interventions such as pharyngoplasty and ear operations was reported [5, 7].

H. Pilge · R. Lampe · P. M. Prodinger
Department of Orthopedics and Sports Medicine,
Technische Universität München, Munich, Germany

B. M. Holzapfel
Institute of Health and Biomedical Innovation,
Queensland University of Technology,
Brisbane, Australia

S. Pilge
Department of Anaesthesiology, Helios Clinic Wuppertal, Witten/
Herdecke University, Wuppertal, Germany

H. Pilge (✉)
Orthopedic Department, University Hospital Düsseldorf,
Düsseldorf, Germany
e-mail: Hakan.Pilge@med.uni-duesseldorf.de

In 1930, Grisel was the first to describe this entity and he assumed that a local infection would lead to a transmitted, infection-related reaction affecting the cervico-spinal ligaments [8]. Thus, the torticollis may be seen as an attempt of physical compensation to relieve the symptoms caused by the inflamed tissue. Others presume that local, infection-related hyperaemia and subsequent partial decalcification of the first and second cervical vertebra in combination with loosening of the anterior transverse ligament and the atlanto-axial ligaments to be the main reason for the atlanto-axial subluxation [9, 10]. In this context, Parke et al. described a previously unknown plexus of lympho-venous anastomoses through the pharyngobasilar fascia and the atlanto-occipital membrane that facilitates hematogenous conduction of local infections of the ear or the pharynx [11]. Constitutional laxity of atlanto-axial ligaments was also proposed to be crucial in the pathogenesis of Grisel's syndrome [3, 12, 13]. Why this pathology is almost solely confined to the paediatric population remains unclear. A hypermobility of C1 on C2 with an increased dens-atlas interval of up to 4.5 mm has been observed in children (adults range from 2.5 to 3 mm) [14]. Thus, Battiatà postulated a combination of ligamental laxity and an accompanying precipitator (e.g., infectious process, surgery) as a “two-hit” hypothesis [9].

Conventional therapy concepts imply stage-related recommendations based on the Fielding classification (type I–IV). This classification was introduced in 1977 to assess the degree of subluxation between C1 and C2. Thus, instability increases from type I to IV and higher stages may require surgical interventions (Tables 1 and 2) [9, 12, 15, 16]. Adjuvant antibiotic treatment is considered to be beneficial [4, 5, 10].

The purpose of this study was to evaluate a simplified, less invasive therapeutical algorithm.

Patients

All parents or legal guardians gave informed written consent to the protocol that was approved by the ethics committee of the Technical University Munich. Alternative treatment methods were taken into consideration. The best applicable approach was chosen.

From January 2004 to December 2010 we identified five pediatric patients, aged eight to 13 years, with Grisel's syndrome (Table 3; Figs. 1, 2, and 3). In all cases, our department was asked for expert counselling because of a painful torticollis resistant to physiotherapy.

All patients showed at least five symptom-free days after surgery. Therefore, a mechanical cause for the subluxation of C1/C2—an impairment of the vertebral column while positioning of the patient peri-operatively—could be excluded.

Diagnosis was based on clinical examination supplemented with radiographs and high resolution computed tomography (three-dimensional reconstruction). All cases were assigned to Fielding types I to III (Tables 1 and 3; Figs. 2, 3, 4, and 5). A blood sample was taken, the white blood count and CRP values were determined.

All patients were treated with manual repositioning under general anaesthesia. For this purpose, the patients were intubated and received neuromuscular blocking agents (mivacurium 0.2 mg kg⁻¹). The repositioning technique used is based on well-established manual-therapeutic measures [18, 19] and implies a three-step approach. The first mobilization is a latero-lateral movement of the cervical spine in the coronal plane; the second step a lateral flexion to each side. As a third step we performed a rotation in the longitudinal axis (Fig. 4). During this procedure we noticed an audible repositioning of the subluxated vertebral joints in four cases. A mild manual distraction of the cervical spine was maintained during the whole procedure to prevent damage of osseous structures or the spinal cord. Interestingly, none of our patients showed spontaneous repositioning after induction of general anesthesia under muscle relaxation. The reposition manoeuvre was done under fluoroscopy control. Afterwards, but still under maintenance of general anaesthesia, a Minerva cast was adjusted (Fig. 4d). The Minerva cast was replaced by a Minerva orthosis after one week (Fig. 5) and physiotherapy was initiated. We administered analgetic medication (NSAID, ibuprofen 600 mg per day) and antibiotics (clindamycin 20 mg kg⁻¹ per day) and muscle relaxants (tetraepam 25 mg per day) to all patients. Four weeks after manual repositioning, free movement was allowed during the daytime and the Minerva orthosis was prescribed only at night for two more weeks. After this period, the orthosis was abandoned.

Results

Fielding classification

We identified types I to III according to Fielding (Table 3). In case number 5 we found a bilateral rotatory luxation with displacement of the vertebral body. So far this type has not been described; therefore, we considered it as a “modified type III” according to Fielding, because of the ventral dislocation of C1 of more than 5 mm.

Infection parameters

Patients 2 and 3 showed elevated infection parameters prior to the initial surgical intervention (C-reactive protein and

Table 1 Fielding classification of Grisel's syndrome

Fielding-Classification of Grisel's syndrome		
Type I	No subluxation. Fixed rotation of Atlas and Axis. Dislocation less than 3mm anteriorly	Type I
Type II	Unilateral, atlanto-axial subluxation of one atlanto-axial joint. The contralateral joint is pivotal. Ventral dislocation between 3 and 5 mm	Type II
Type III	Ventral subluxation of C1 in both joints. Ventral dislocation more than 5 mm	Type III
Type IV	Dorsal subluxation of C1. Combination with fractured Dens axis or congenital Dens-aplasia is possible.	Type IV

Table 2 Stage-related therapy of Grisel's syndrome according to the recommendations of [6, 12, 16, 17]

Stage-related therapy of Grisel's syndrome	
Type I	Soft neck brace
Type II	Conservative treatment, Philadelphia brace
Type III	Closed reposition, halo extension
Type IV	Operative treatment, fusion of C1-C2

leucocytes) due to a purulent mastoiditis and tonsillitis. After surgery both parameters decreased to normal range within a few days.

In patient number 4 the postoperative laboratory test revealed a prolonged declining CRP with normal parameters after the 13th postoperative day. Patients 1 and 5 did not show elevated parameters at all. At the time of the first expert counselling (Table 3) in our department, elevated infection parameters could not be verified in any of our patients.

Table 3 Patient's demographics and diagnostic details

Patient number	Age at first presentation (years)	Gender	Diagnosis	Time after surgery until first presentation in our clinic (days)	Classification (Fielding)	Treatment (duration) in weeks
1	13	Male	Cholesteatoma (resection)	10	III	7
2	12	Female	Mastoiditis (drainage)	8	I	4
3	8	Male	Tonsillitis (tonsillectomy)	14	II	6
4	11	Female	Labyrinthine hearing loss (cochlea-implant)	58	II	8
5	9	Female	Protruding ears (otoplasty)	7	III	6

Therapy

In our first patient we applied a soft-collar cast after repositioning. One week after manual repositioning, the patient presented a recurrence of the torticollis without subluxation of C1/C2 [4]. Though we decided not to reposition the spine again, restitution could be achieved in this case with regular physiotherapy. In the following cases we applied a more stable Minerva cast after repositioning in order to avoid recurrence. With this minor modification we were able to ensure good clinical results in all other patients. No case of recurrence was observed within a follow-up of six months.

Discussion

The aetiology of Grisel's syndrome is still unidentified. In the literature, the association to previous otorhinolaryngologic (surgical) interventions or infections of the ear, nose or throat has been discussed. Aetiologically there seems to be a conduction of local infection through the pharyngobasilar fascia and the atlanto-occipital membrane towards the cervical spine.

Pat No.5
9 y/o girl after otoplast (protruding ears). Three days after surgery she complains about a wryneck with the chin tilted to the left and her head flexed to the right

Fig. 1 Patient number 5, a nine-year-old girl after otoplasty (protruding ears). Three days after surgery she complained of a wryneck with the chin tilted to the left and her head flexed to the right

The syndrome can be caused even by subclinical, occult pharyngeal infections (visible only by MRT scan) [10, 12]. Therefore most authors recommend additional antibiotics. In our group all patients had otorhinolaryngeal surgery, prior to torticollis.

Treatment guidelines are still lacking. Recommendations are based on less than 80 scientific articles, which were published since 1950. A review of Karkos et al. [5] emphasizes that up-to-date recommendations for interventions—which are based on the underlying Fielding type—rely on case reports or small clinical studies. In general it is assumed that the degree of subluxation and instability increases with the Fielding type, and that the need for a surgical intervention is more likely with Fielding types III and IV. So far the following therapeutic approach is most commonly used: Fielding types I and II are conventionally treated via oral administration of antibiotics, muscle relaxants and anti-inflammatory medication in combination with immobilization of the cervical spine. In type III closed reposition and immobilization in a halo brace for up to 12 weeks is necessary, type IV is primarily treated surgically with fusion of C1

Radiograph taken at first presentation in our clinic two months after cochlea implantation (Pat No 4).

The chin is tilted to the right and the head shows a left-flexed position. The atlas (C1) shows a subluxation in relation to the odontoid process (#).

Fig. 2 Radiograph taken at first presentation in our clinic two months after cochlea implantation (patient number 4). The chin is tilted to the right and the head shows a left-flexed position. The atlas (C1) shows a subluxation in relation to the odontoid process (#)

a: CT-Scan of the cervical spine in the frontal plane showing a rotatory luxation of C1/C2 (Patient No.5, Fielding Type III)
b: 3D-reconstruction of the same patient (Patient No.5)

+ Occiput
 # C1
 * Odontoid process of C2
 ★ C2

Fig. 3 **a** CT-scan of the cervical spine in the frontal plane showing a rotatory luxation of C1/C2 (patient number 5, Fielding type III). **b** 3D reconstruction of the same patient (patient number 5). + Occiput, # C1, * Odontoid process of C2, ★ C2

and C2 [9, 12, 15–17, 20]. After surgical stabilization, persistent neurological complications occur in up to 15 % of all patients [21]. Even fatal outcomes with tetraplegia have been described [3] in the literature.

Therefore, our aim was to develop a new therapeutic concept—irrespective of the underlying Fielding classification—in order to improve outcome with less invasive

After one week the Minerva cast is replaced by a Minerva-orthosis (Patient No. 5). The wound (after otoplasty) is still covered with a wound dressing

Fig. 5 After one week the Minerva cast is replaced by a Minerva orthosis (patient number 5). The wound (after otoplasty) is still covered with a wound dressing

techniques. Thereby the main focus relied on manual repositioning in a three-step approach under general anaesthesia followed by immobilization of the cervical spine. Tetracepam and NSAID were administered. In contrast to conventional recommendations, with this novel approach

Fig. 4 Repositioning under general anaesthesia. **a** First step: distraction, latero-lateral mobilization. **b** Second step: lateral flexion to both sides. **c** Third step: rotation to both sides

Repositioning under general anaesthesia

a First step: Distraction, latero-lateral mobilization

b Second step: Lateral flexion to both sides

c Third step: Rotation to both sides

d Fourth step: After repositioning a minerva-cast is applied

we were able to avoid cases of patients with Fielding type III, or halo extension after delayed diagnosis, or an even more severe surgical intervention such as internal fixation or fusion of C1 and C2. Furthermore, a more rapid recovery was possible. We achieved full restitution within four to eight weeks in all five patients.

It has to be mentioned that our study group did not include the very severe types of Grisel's syndrome: in almost seven years, we did not observe any patients with Fielding type IV. The combination of (sub-)luxation with a fractured dens axis or a congenital dens-aplasia implicates a major reduction of stability and contraindicates conservative treatment. Therefore, we recommend to primarily discuss surgical interventions in these cases [6, 12, 22].

Phillips et al. stated a close relation of rehabilitation time to the duration time of the untreated torticollis. Delayed treatment resulted in arthrodesis (C1–C2) in 60 % of the respective children [6]. Fielding et al. also recommended surgical fusion of C1 and C2 in cases with delayed diagnosis (of more than three months) to prevent recurrent subluxation [12]. The syndrome has been described to occur even months after local infections [10, 13]. If diagnosis is established within four weeks, manual reposition seems to be adequate [4, 6]. In the case of delayed diagnosis, many authors recommend immobilization with halo fixation for 12 weeks [6, 12, 16, 17].

In our study, one patient was diagnosed with a delay of eight weeks after the first symptoms had occurred. In this single case we achieved full recovery within the following eight weeks and surgical treatment could be avoided.

Conclusions

Grisel's syndrome should be considered in children who present themselves with an acquired torticollis, especially if there is a history of otorhinolaryngological infection or surgical intervention. Early diagnosis and therapy is essential to avoid neurological complications [3]. Blood samples can show elevated infectious parameters, but not always [10, 13, 23]. To confirm diagnosis, a CT scan of the cervical spine should be performed. Our new therapy concept enabled us to achieve excellent results in this rare disease. In a case with Fielding type III a surgical intervention could be avoided. Overall treatment times were reduced, even in patients with delayed diagnosis. No case of recurrence was observed in a follow-up of six months. However, this therapeutical approach with manual repositioning and the application of a Minerva cast under general anaesthesia should only be performed by an experienced specialist in a specialized centre.

In conclusion, with our novel therapy regimen, in contrast to conventional recommendations, good clinical results

can be obtained without surgical intervention in Fielding types I–III or after delayed diagnosis. More patients need to be investigated to confirm our results and establish this novel therapeutic regimen.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Wilson BC, Jarvis BL, Haydon RC 3rd (1987) Nontraumatic subluxation of the atlantoaxial joint: Grisel's syndrome. *Ann Otol Rhinol Laryngol* 96(6):705–708
2. Grobman LR, Stricker S (1990) Grisel's syndrome. *Ear Nose Throat J* 69:799–801
3. Wilson MJ, Michele AA, Jacobson EW (1940) Spontaneous dislocation of the atlanto-axial articulation, including a report of a case with quadriplegia. *J Bone Joint Surg Am* 22:698–707
4. Pilge H, Prodinger P, Burklein D, Holzapfel BM, Lauen J (2010) Non-traumatic subluxation of the atlanto-axial joint as rare form of acquired torticollis: Diagnosis and clinical features of the Grisel's syndrome. *Spine (Phila Pa 1976)* 36:E747–E751. doi:10.1097/BRS.0b013e3181fab0e4
5. Karkos PD, Benton J, Leong SC, Mushi E, Sivaji N, Assimakopoulos DA (2007) Grisel's syndrome in otolaryngology: a systematic review. *Int J Ped Otorhinolaryngo* 71:1823–1827
6. Phillips WA, Hensinger RN (1989) The management of rotatory atlanto-axial subluxation in children. *J Bone Joint Surg Am* 71:664–668
7. Durst F, Staudenmaier R, Pilge H, Lauen J, Prodinger PM, Holzapfel K, Pickhard A (2011) Grisel's syndrome after otoplasty. *HNO* 60:135–140
8. Grisel P (1930) Énucléation de l'atlas et torticollis nasopharyngien. *La Presse Med* 38:50–53
9. Battista AP, Pazos G (2004) Grisel's syndrome: the two-hit hypothesis—a case report and literature review. *Ear Nose Throat J* 83(8):553–555
10. Harth M, Mayer M, Marzi I, Vogl TJ (2004) Lateral torticollis on plain radiographs and MRI: Grisel syndrome. *Eur Radiol* 14:1713–1715
11. Parke WW, Rothman RH, Brown MD (1984) The pharyngovertebral veins: an anatomical rationale for Grisel's syndrome. *J Bone Joint Surg Am* 66:568–574
12. Fielding JW, Hawkins RJ (1977) Atlanto-axial rotatory fixation. *J Bone Joint Surg Am* 59:37–44
13. Doshi J, Anari S, Zammit-Maempel I, Paleri V (2007) Grisel syndrome: a delayed presentation in an asymptomatic patient. *J Laryngol Otol* 121:800–802
14. Jackson H (1950) The diagnosis of minimal atlanto-axial subluxation. *Br J Radiol* 23:672–674
15. Hirth K, Welkoborsky HJ (2003) Grisel's syndrome following ENT-surgery: report of two cases. *Laryngorhinootologie* 82(11):794–798. doi:10.1055/s-2003-44547
16. Kasten P, Zeichen J, Gosling T, Krettek C (2002) Grisel syndrome—a trauma surgery rarity. *Unfallchirurg* 105(6):565–568
17. Gourin CG, Kaper B, Abdu WA, Donegan JO (2002) Nontraumatic atlanto-axial subluxation after retropharyngeal cellulitis: Grisel's syndrome. *Am J Otolaryngol* 23:60–65
18. Bischoff HP, Moll H (2007) Short textbook of manual medicine. Spitta, Balingen

19. Greenman PE (2003) Principles of manual medicine. Lippincott Williams & Wilkins, Philadelphia
20. Wetzel FT, LaRocca H (1989) Grisel's syndrome. Clin Orthop 240:141–152
21. Rinaldo A, Mondin V, Suarez C, Genden EM, Ferlito A (2005) Grisel's syndrome in head and neck practice. Oral Oncol 41 (10):966–970
22. Yu KK, White DR, Weissler MC, Pillsbury HC (2003) Nontraumatic atlantoaxial subluxation (Grisel syndrome): a rare complication of otolaryngological procedures. Laryngoscope 113:1047–1049
23. Derkay CS, Kenna MA, Pang D (1987) Refractory torticollis: an uncommon complication of adenotonsillectomy. Int J Ped Otorhinolaryngol 14:87–93