

## Grisel's syndrome—a rare complication following “small” operations and infections in the ENT region

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**Abstract** The atlanto-axial subluxation (Grisel's syndrome) is a rare complication following operative procedures and/or infections in the upper aerodigestive tract. Pathogenetically the higher flexibility of the ligaments during the inflammation causes a subluxation between axis and atlas. When the inflammation heals, this can probably result in a fixation in the rotated position. The purpose of the present study was to describe the clinical and radiological characteristics of this rare disease in children. The clinical files of 12 patients with Grisel's syndrome were examined retrospectively. The clinical files of these patients were reviewed and analyzed along with the results of radiographic and laboratory examinations. The clinical parameters were compared with the patient's therapy and outcome. Of 12 children (6 males, 6 females, average age of 7.1 years), Grisel's syndrome developed following surgery in 8 patients, and in another 4 patients following a severe infection of the upper aerodigestive tract. The duration of complaints differed from 2 days to 6 months. All patients had a torticollis as the first symptom of atlanto-axial dislocation and three children had accompanying cervical lymphadenopathy. All patients were given antiphlogistic therapy either with diclofenac or ibuprofen. Additionally, 11 patients were treated with intravenous antibiotics (amoxicillin, ampicillin, clindamycin or cephalosporins)

and 1 child with oral antibiotics. In eight patients a remission during antibiotic therapy occurred. In four cases (2 with Grisel's syndrome following surgery, 2 following infection), however, the torticollis persisted despite adequate conservative treatment and required reposition of the atlanto-axial joint along with external fixation. In all of them, starting of therapy was delayed. An early diagnosis of Grisel's syndrome and immediate therapy is most important. Grisel's syndrome must be taken under consideration in children with acute torticollis following either an infection or operative procedure in the upper aerodigestive tract. Early adequate antibiotic and antiphlogistic therapy is mandatory and leads to a high remission rate.

**Keywords** Grisel's syndrome · Atlanto-axial subluxation · Torticollis · Complications · Adenotomy · Tonsillectomy

### Introduction

Grisel's syndrome is a non-traumatic subluxation of the atlanto-axial joint usually caused by pharyngitis, which is accompanied by an edematous hyperemia of the longitudinal anterior ligament and the alaria ligament. Pathogenetically the higher flexibility of the ligaments during the inflammation causes a subluxation between axis and atlas. When the inflammation heals, this can probably result in a fixation in the rotated position [1, 2]. The Grisel's syndrome underlying pharyngitis can be on the one hand caused by an infection in the upper aerodigestive tract (i.e. tonsillitis, adenoiditis) and on the other can occur following operative procedures in this region with consecutive reactive pharyngitis [3].

The atlanto-axial dislocation has been first described by Sir Charles Bell in 1830 in a patient with syphilitic ulceration

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of the pharynx [4]. Pierre Grisel published two cases of atlanto-axial subluxation in children in 1930 [5]. Basically according to Fielding and Hawkings [2] Grisel's syndrome is classified into four types (cp. Table 1), with type I meaning a rotary fixation with an anterior displacement of atlas of <3 mm, and type IV a most severe rotary fixation with a posterior displacement of atlas.

Although described some 70 years ago, currently there are only few studies published on Grisel's syndrome in children. In the present study the clinical and radiological characteristics of 12 children with Grisel's syndrome are described along with therapeutic management of this rare complication.

## Patients and methods

From 2002 through 2009, 12 patients with Grisel's syndrome were admitted in the Department of Otorhinolaryngology. Clinical files of these patients were reviewed retrospectively and analyzed along with the results of radiographic and laboratory examinations. The clinical parameters were compared with the patient's therapy and outcome. The clinical parameters of all patients are detailed in Table 2.

## Results

Among the 12 children, 6 patients each were male and female, respectively with an average age of 7.1 years (range 4–11 years). In eight patients a Grisel's syndrome developed following surgery, in the other four following severe infection of the upper aerodigestive tract. Among the group of Grisel's syndrome following surgery, most frequent operation was adenotomy/paracentesis (5 patients). Among the group of post-infection Grisel's syndrome, tonsillitis was the most frequent cause (3 patients). In one patient each, the atlanto-axial dislocation followed tonsillectomy, functional neck dissection due to unclear lymphadenopathy, tympanoplasty and gastrointestinal infection.

**Table 1** Classification of Grisel's syndrome according to Fielding and Hawkings [2]

Type I	Rotary fixation without anterior displacement of atlas ( $\leq 3$ mm)
Type II	Rotary fixation with anterior displacement of atlas of 3–5 mm
Type III	Rotary fixation with anterior displacement of atlas $>5$ mm
Type IV	Rotary fixation with posterior displacement of atlas

Grisel's syndrome was estimated when (cp. Table 3) (1) the torticollis occurred some days after a typical surgical procedure (i.e., adenotomy, tonsillectomy) or an infection in the upper aerodigestive tract (i.e., tonsillitis), (2) a rotation and slight flexion of the head to the contralateral side was obvious, (3) initial laboratory examinations showed elevated values for CRP and leucocytes with a normalization within few days, (4) X-ray of the cervical spine showed the atlanto-axial subluxation with a space between atlas and dens axis of  $>5$  mm, (5) a CT-scan (when performed) displayed the atlanto-axial subluxation and rotation, and (6) a mastoiditis, meningitis, mechanical subluxation during surgery, or immobility resulting from painful lymphadenopathy could be excluded.

The duration of complaints differed from 2 days to 6 months. All patients had a torticollis as the first symptom of atlanto-axial dislocation and three children had accompanied moderate and not painful cervical lymphadenopathy. The severity of pharyngitis and the elevation of C-reactive protein (CRP) values in peripheral blood differed depending on the duration of the history. The earlier the patients were admitted after the first onset of symptoms the higher was their CRP in peripheral blood. This corresponded with the activity of their infection.

All patients were given antiphlogistic therapy either with diclofenac or ibuprofen. Additionally, 11 patients were treated with intravenous antibiotics (amoxicillin, ampicillin, clindamycin or cephalosporins) and only 1 child with oral antibiotics. In eight patients a remission during antibiotic therapy occurred. In four cases (2 with Grisel's syndrome following surgery, 2 following infection), however, the torticollis persisted despite adequate conservative treatment. Three of these children were admitted several weeks or even months after onset of first symptoms of atlanto-axial dislocation. In these patients a reposition of the atlanto-axial joint in general anesthesia and an external fixation by halo-fixation for 6 weeks each was necessary. After this procedure, all of these patients had a remission.

Prior to any therapeutic management, all patients underwent clinical examination. Because of the radiation exposure a computed tomography (CT) scan was not performed in all cases. The indication for a CT-scan was recommended in all patients with a persisting torticollis after i.v.-antibiotic therapy. This was done in five cases. Four of them underwent external reposition and fixation by a halo frame. In these cases, CT-scans prior to the procedure are mandatory. The fifth case had a remission during the planning of surgical reposition. In the CT-scan the atlanto-axial subluxation can be seen in the two-dimensional slices (Fig. 1). The 3D reconstruction shows the severity of the luxation in the three-dimensional view (Fig. 2).

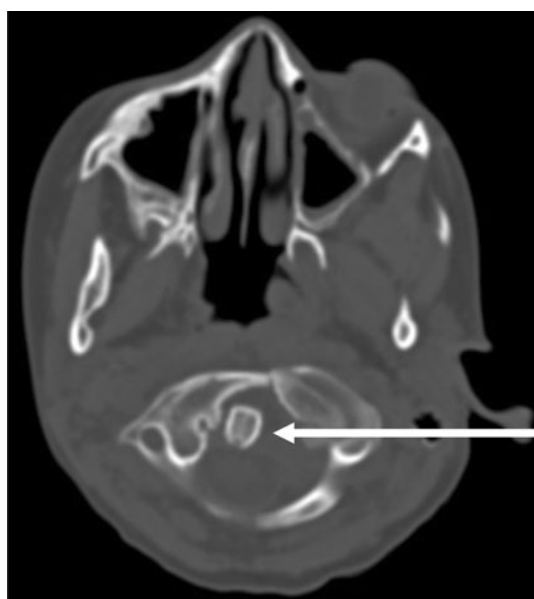
**Table 2** Clinical parameters of patients with Grisel’s syndrome

Age at diagnosis	Gender	Medical history and probable cause for Grisel’s syndrome	Duration of symptoms	Diagnostics	Laboratory findings	Therapy
4 7/12	♂	AT/PC	2d	History, clinical examination, X-ray cervical spine	CRP 23.5 ng/dl; WBC 13.7	i.v.-antibiotics Amoxicillin + Ibuprofen
4 1/12	♂	AT/PC	2d	History, clinical examination	CRP 0.92 ng/dl; WBC 14.4	i.v.-antibiotics Cefotaxime + Diclofenac
4 1/12	♀	AT/PC	2d	History, clinical examination	CRP 2.41 ng/dl; WBC 7.9	i.v.-antibiotics Clindamycin + Ibuprofen
9 1/12	♂	AT/PC	8d	History, clinical examination	CRP 0.75 ng/dl; WBC 11.6	i.v.-antibiotics Clindamycin + Diclofenac
4 1/12	♀	AT/PC	2d	History, clinical examination		Oral antibiotics Amoxicillin + Ibuprofen + Paracetamol, oral antibiotics sine effectu
7 6/12	♂	tymanoplasty type IIIa left; inspection of the nasal pharynx + microscopy of the right ear	6d/14d	History, clinical examination, CT	CRP 0.23 ng/dl; WBC 7.6	Oral antibiotics with Co-trimoxazole sine effectu, i.v.-antibiotics Cefotaxime + Ibuprofen,
11 3/12	♀	tonsillectomy	~2 weeks	History, clinical examination, MRI; CT	WBC 12.0; BSR 52/73	i.v.-antibiotics ==> halo-fixation
7 9/12	♀	neck dissection because of unclear lymphadenopathy	~6 months	History, clinical examination, ultrasonography, CT		Halo-fixation
4 4/12	♀	tonsillitis	~2-3 h	History, clinical examination, ultrasonography	CRP 5.87 ng/dl; WBC 24.3	i.v.-antibiotics Cefuroxim + Ibuprofen
10 5/12	♂	tonsillitis	5d	History, clinical examination, ultrasonography	CRP 8.77 ng/dl; WBC 17.0	i.v.-antibiotics Ampicillin + Ibuprofen
8 0/12	♂	tonsillitis	5 months	History, clinical examination, X-ray cervical spine, CT	CRP 0.05 ng/dl; WBC 7.8	i.v.-antibiotics Ampicillin + Ibuprofen + Paracetamol ==> halo-fixation
10 4/12	♀	gastrointestinal infection	~1 ¼ months	History, clinical examination, X-ray cervical spine, CT	CRP 6.66 ng/dl	Halo-fixation

Age years and months, AT/PC adenoidectomy and paracentesis, BSR blood sedimentation rate, CRP C-reactive protein in peripheral blood, CT computed tomography, Halo-fixation external fixation with halo frame, i.v. antibiotics intravenous application of antibiotics, MRI magnetic resonance imaging, WBC white blood cell count (leucocytes in peripheral blood)

**Table 3** Diagnostic algorithm for Grisel's syndrome

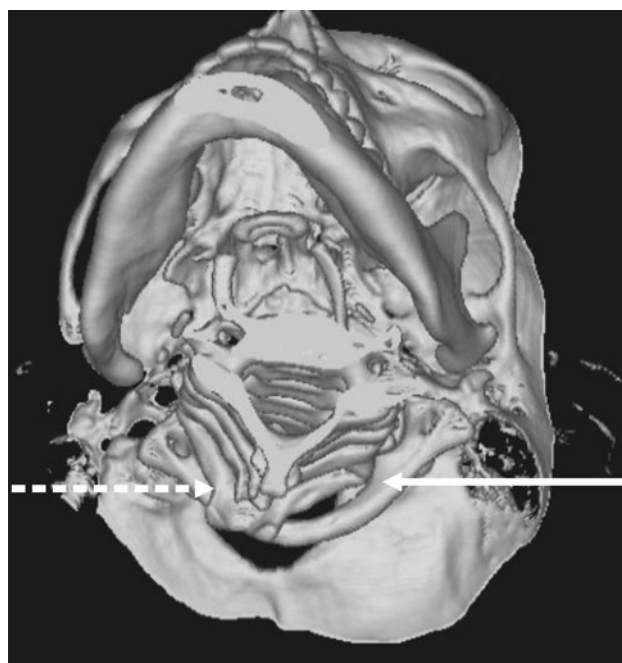
History of ENT surgery or ENT infection
Torticollis some days after onset of infection/operation
Rotation and slight flexion of the head, chin shows to the contralateral side
Painful active and passive rotation of the head
In the first days of torticollis elevated CRP and leucocytes, later on normalization of these parameters and usually no fever
X-ray of the cervical spine: space between atlas and dens axis >5 mm
CT-scan: atlanto-axial subluxation and rotation

**Fig. 1** CT-scan in axial planes shows atlanto-axial dislocation (arrow)

## Discussion

Grisel's syndrome is a rare complication after surgical procedures in the ENT region or following infections in the upper aerodigestive tract (i.e., tonsillitis, pharyngitis, adenitis). Karkos et al. [6] reported 96 cases with non-traumatic atlanto-axial rotary subluxation. Out of these, Grisel's syndrome occurred following infections in 48% and after surgery (of these 78% after adenotonsillectomy) in 40%. The syndrome most frequently occurs in children up to 12 years of age, which might be due to more frequent infections in the upper aerodigestive tract and probably a minor tissue density in children. There are only rare cases described in adults [7, 8].

In the present study, atlanto-axial subluxation occurred in 67% of our patients after surgery (5 out of 12 following adenotomy) and 33% after an ENT infection. The torticollis

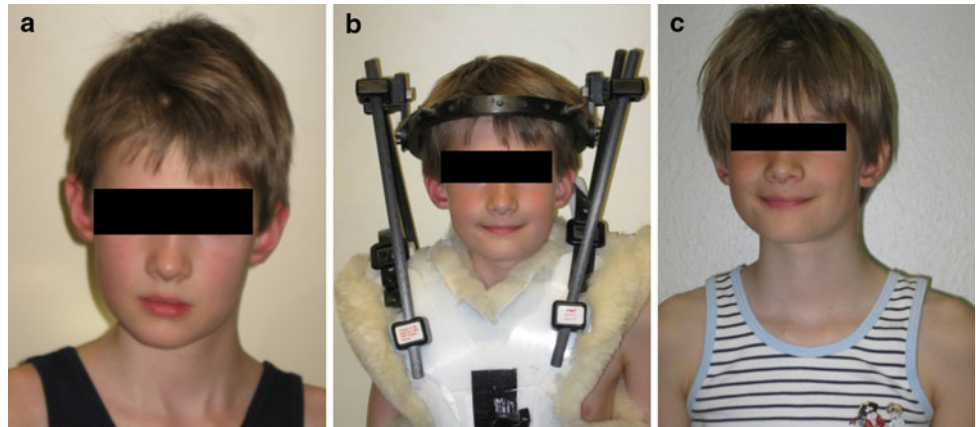
**Fig. 2** 3D-reconstruction of the CT-scan gives a three dimensional impression of the severity of the dislocation. The atlas (solid arrow) is in normal position to the foramen and the head. The axis (dashed arrow) is rotated to the right side. The following vertebral segments of the cervical spine make a slight rotation back to the midline

mostly occurred only a few days after surgery or infection. As much as 50% of the patients were seen in our clinic within 5 days after the first symptoms, 17% had a history up to 10 days. The remaining 4 patients (33%), however, had the first contact in our clinic after 2 weeks, 1¼ months, 5 months, and 6 months, respectively.

In this study one patient had already started an antibiotic therapy prior to admittance. Detailed patient history revealed that the torticollis had already decreased. With additional antiphlogistic therapy a complete remission was seen. The other seven patients who had a remission after i.v.-antibiotic therapy and oral antiphlogistic therapy had a history of torticollis from few hours up to 8 days. The four patients with complaints  $\geq 2$  weeks underwent a long medical treatment by pediatricians, orthopedic specialists, physiotherapist and osteopaths without any recovery. At the first contact they had a very distinctive torticollis (Fig. 3a) and up to this point no sufficient therapy.

In these four patients no remission after intravenous antibiotic therapy and oral antiphlogistic therapy occurred. These patients required reposition with general anaesthesia followed by a halo-fixation (Fig. 3b), which led to remission in all cases (Fig. 3c). The other eight patients had a remission after antibiotic therapy. Only one case in which the oral antibiotic therapy was started 6 days after the first symptoms had a prolonged course. Because of non-complaints of the patient, the i.v.-antibiotic therapy started

**Fig. 3** A 9-year-old boy with Grisel's syndrome. Duration of torticollis: 4 months. No remission with conservative treatment and so reposition and external fixation was required. **a** Patient before therapy, **b** with halo-fixation, **c** after therapy



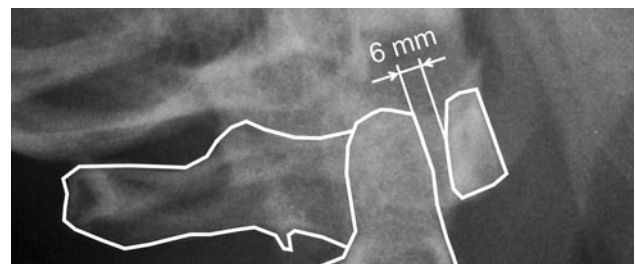
just after 14 days. During conservative treatment and planning of halo-fixation the patient had a remission. Thus it is important to note that in this series, conservative therapy with intravenous antibiotics plus antiphlogistics, which began immediately after the first signs of atlanto-axial subluxation, helped reversing the problem in certain number of patients.

This is in accordance with the literature. The few studies on Grisel's syndrome in children also revealed a good prognosis following early therapy. Nevertheless, some authors discussed whether an atlanto-axial subluxation and fixation after infection frequently requires a reposition and halo-fixation. However, a surgical fusion of the atlanto-axial joint is described only in very few cases, when the joint stays unstable [9]. Mathern, Fielding and Gourin described neurological symptoms caused by a compression of the cervical spine [2, 10, 11]. The authors stated that in these cases a fusion of C1 + C2 might be adequate. Park et al. described a case of a 9-year-old girl who was only treated with cervical traction, first with halter traction of 5-lb weight for 6 weeks and then with a brace for 4 months and collar for 2 more months. After this therapy, the patient had a full remission [12]. However, it seems reasonable to note that this very invasive therapy is recommended only in very few cases when all other, especially conservative and minimal invasive, procedures do not lead to a remission. In our study such more invasive treatment modalities were not necessary.

The diagnosis of a Grisel's syndrome can most frequently be based on clinical examinations alone. Often, a subluxation can be seen in a conventional X-ray of the cervical spine. The distance between dens axis and the back-side of the anterior arch of the atlas is normally 2–3 mm; in children it can physiologically be up to a maximum of 5 mm [13]. If this space exceeds 5 mm, an atlanto-axial subluxation is very probable (Figs. 4, 5). The subluxation, however, can finally be verified only in a CT-scan. The advantage of CT-scans is that bony structures are visible



**Fig. 4** Conventional X-ray in a child with Grisel's syndrome



**Fig. 5** Schematic view of the atlanto-axial joint of the same child

very clearly and so the relation between C1 and C2 can be estimated. An algorithm of diagnostic parameters is listed in Table 3. On the first view, the magnetic resonance imaging (MRI) seems to be the preferred diagnostic procedure especially in children, because of lack of radiation exposure. However, in MRI scans the soft tissue structures



are better visible than in CT-scans. This can, on the other hand, lead to a wrong diagnosis, because the bony structures of the vertebral spine are not properly shown [3, 14, 15]. Due to this difficulty, hyperemia of the longitudinal or alaria ligaments, which is characteristic for Grisel's syndrome might mimic an abscess in the retropharyngeal space and therefore might lead to an unnecessary operation [16].

The correct diagnosis of Grisel's syndrome is challenging as important differential diagnosis which could also cause a torticollis must be taken into consideration (i.e., meningitis, mastoiditis, painful lymphadenopathy, or mechanical subluxation during surgery).

In this study, the diagnosis was mostly done on the basis of the history, the clinical examination and clinical signs. In addition in some cases X-ray of the cervical spine was performed. A CT-scan was indicated in children, when an i.v.-antibiotic and antiphlogistic therapy showed no effects in the first few days.

The differential diagnosis of meningitis can be done based on clinical signs. In meningitis, a rotation of the head is possible whereas the flexion is limited and painful. Typical for Grisel's syndrome is the fixed rotation to one side with only a slight flexion. The chin shows to the contralateral side. Especially the rotation is very painful. When clinical examination and patient's history remain unclear, a liquor puncture should be necessary, which reveals typical values i.e. leucocytes in meningitis and normal values in Grisel's syndrome. In this series, however, liquor puncture was not necessary in any case to exclude meningitis. Finally X-ray or CT-scan shows no atlanto-axial subluxation in meningitis.

If the atlanto-axial subluxation occurs following surgical procedures, a malpractice might be discussed, i.e. hyperflexion of the cervical spine or incorrect placement of the patient's head during operation. Especially, the adenotomy is a very short operation without hyperflexion of the head. In addition, the torticollis in Grisel's syndrome occurs usually 1 day up to several days after operation, whereas it should be obvious immediately after surgery when caused by mechanical distraction. Therefore, an intraoperative malpractice is not likely [17].

Furthermore, an immobility resulting from big and painful lymphadenopathy can be misdiagnosed as Grisel's syndrome. In lymphadenopathy, the active rotation of the head can be painful, but there is no fixation of the cervical spine, so that a passive rotation (if so under pain) is possible. Also the palpation of lymph nodes in a lymphadenitis is often painful. In Grisel's syndrome the palpation of nodes is only slightly painful.

Finally, Bezold's mastoiditis leads to a torticollis. However, Bezold's mastoiditis is attended by the typical signs of an acute mastoiditis (i.e. distant from the auricle, doughy

swelling of the mastoid in most cases). In addition there is a swelling at the beginning of the sternocleidomastoideus muscles, whereas in Grisel's syndrome the muscle is slim and X-ray shows no atlanto-axial dislocation.

## Conclusion

An early diagnosis of a Grisel's syndrome and immediate onset of therapy is most important. Although very rare, Grisel's syndrome must be taken into consideration in children with acute torticollis following either an infection or operative procedure in the upper aerodigestive tract. On the basis of this series, early adequate antibiotic plus antiphlogistic therapy seems to be mandatory. With this, Grisel's syndrome has a good prognosis with a high remission rate.

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