

## Grisel's Syndrome: A Rare Cause of Atlantoaxial Subluxation

<sup>id</sup> Zehra Serpil USTALAR ÖZGEN<sup>a</sup>, <sup>id</sup> Murat BAŞARIR<sup>b</sup>, <sup>id</sup> Seher İrem KIRAN<sup>a</sup>,  
<sup>id</sup> Selin ÜNSALER<sup>c</sup>, <sup>id</sup> Hakan AĞIR<sup>d</sup>

<sup>a</sup>Acıbadem Mehmet Ali Aydınlar University Faculty of Medicine, Department of Anesthesiology and Reanimation, İstanbul, TURKEY

<sup>b</sup>Acıbadem Altunizade Hospital, Department of Pediatric Neurosurgery, İstanbul, TURKEY

<sup>c</sup>Acıbadem Mehmet Ali Aydınlar University Faculty of Medicine, Department of Ear, Nose Throat Surgery, İstanbul, TURKEY

<sup>d</sup>Acıbadem Mehmet Ali Aydınlar University Faculty of Medicine, Department of Plastic and Reconstructive Surgery, İstanbul, TURKEY

**ABSTRACT** Grisel's syndrome is nontraumatic atlantoaxial subluxation secondary to inflammatory conditions or surgery in the head and neck region due to anomalous head posture. In this case report, a 6-year-old female patient, diagnosed as Grisel's syndrome following the surgery for the repair of cleft palate with velopharyngeal insufficiency is presented. This is to note the importance of perioperative positioning and postoperative stabilization. 6 years old female child, with a weight of 23 kg admitted to the hospital for speech disorders and velopharyngeal insufficiency associated with cleft palate. She underwent surgery for the repair of cleft palate and three weeks after surgery, she was readmitted to the hospital with abnormal head posture, pain and limitation in neck movements. After cervical CT scanning, atlantoaxial subluxation was confirmed. She was diagnosed as Grisel's syndrome and treated with conservative therapy by stabilization of the neck with a hard cervical collar and medical treatment. Extreme head positions are sometimes necessary for cleft lip and palate surgeries. Early diagnosis of Grisel's syndrome is critical for a successful treatment.

**Keywords:** Grisel's syndrome; atlantoaxial subluxation; torticollis; cleft palate repair

Grisel's syndrome is an atlantoaxial subluxation due to nontraumatic inflammatory conditions, surgery in the head or neck region due to anomalous head posture. After inflammatory conditions or surgeries in which extreme positions are necessary, the spasm of the neck muscles occurring in reaction to these conditions causes atlantoaxial subluxation at the end. This syndrome had been described in 1830 by Sir Charles Bell in a patient who had pharyngeal ulceration due to syphilis and had undiagnosed atlantoaxial subluxation which caused spinal cord compression and eventually death of the patient.<sup>1,2</sup> In 1930, a French physician Grisel had reported two cases of atlantoaxial subluxation due to nasopharyngeal inflammation.<sup>2,3</sup> He had attributed the atlantoaxial subluxation to the increased flexibility of the atlantoaxial joint ligaments which could easily be affected by extreme positions and/or inflammatory processes. The

pediatric age group, surgical trauma, genetic disorders, inflammatory processes like pharyngitis, adenotonsillitis, any upper respiratory tract infection in patient history, recent abscess of peritonsillar or deep neck or cervical regions, otitis media, and head and neck surgery which might be at extreme head positions or might cause inflammation are described as the risk factors so far. Surgical procedures like tonsillectomy, uvulectomy, adenoidectomy, repair of cleft lip and palate requires positions with extreme head extension and the extreme positions and inflammation may cause postoperative morbidities ranging from head and neck pain to injury of the great vessels located at the base of the skull, i.e. posterior vertebral artery. Grisel's syndrome which presents with torticollis, neck stiffness and pain in neck movements is also a rare postoperative complication seen after these surgical procedures. Complete remission is possible with early diagnosis,

**Correspondence:** Zehra Serpil USTALAR ÖZGEN

Acıbadem Mehmet Ali Aydınlar University Faculty of Medicine, Department of Anesthesiology and Reanimation, İstanbul, TURKEY

**E-mail:** serpozgen@gmail.com



Peer review under responsibility of Türkiye Klinikleri Journal of Case Reports.

**Received:** 28 Dec 2019

**Received in revised form:** 26 Apr 2020

**Accepted:** 29 Apr 2020

**Available online:** 05 May 2020

2147-9291 / Copyright © 2020 by Türkiye Klinikleri. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

antibiotics, anti-inflammatory therapy, stabilization and muscle relaxants. If it is not diagnosed in the early period, surgery may be necessary.

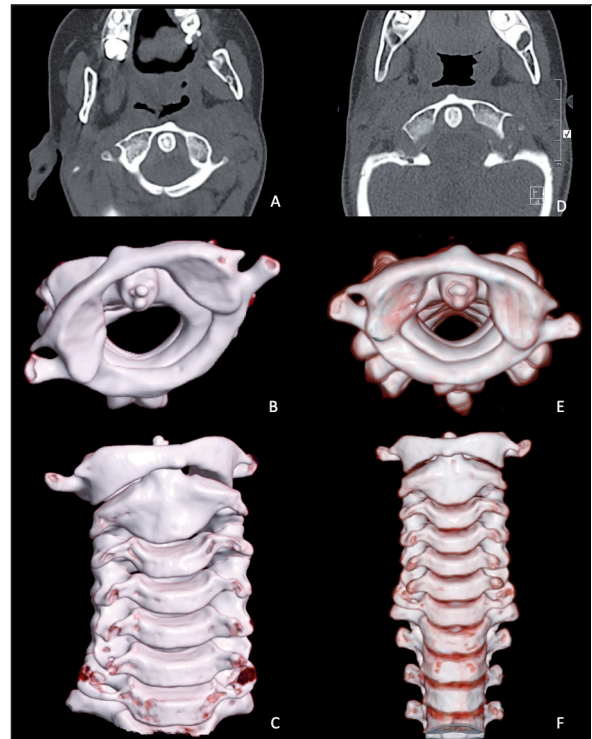
This case report is to present a child who had been operated for her velopharyngeal insufficiency due to her cleft palate. She had been diagnosed as Grisel's syndrome which could have been mistreated if remained undiagnosed. This is to note the importance of perioperative positioning and postoperative stabilization.

## CASE REPORT

Six years old female patient, with a weight of 23 kg was admitted to the hospital for speech disorder and velopharyngeal insufficiency associated with cleft palate. Surgery was planned as double opposing z-plasty and pharyngeal flap and informed consent of her parents had been taken. The operation which had lasted two and a half hours was uneventful, either for surgery and anaesthesia. Three weeks after surgery, abnormal head posture, pain and limitation of neck movements occurred. In cervical CT scanning, atlantoaxial subluxation was suspected and the patient was consulted to pediatric neurosurgery department. The patient's radiological findings and neurological status were evaluated. C1 was rotated 10 degrees left compared with the occiput. There was a 20 degrees rotation between C1 - C2. In the lower cervical vertebrae (C6 - C7), this rotational degree was decreasing to 11 (Figure 1). Because she had no neuromotor deficit, a cervical collar (Philadelphia) stabilization treatment was started. Clindamycin therapy was mandatory for the retropharyngeal inflammation and had been administered for 2 weeks. The patient was examined every one month with cervical 3D CT scanning. After 2 months, atlantoaxial subluxation was totally regressed and the alignment of the vertebrae were normal. Cervical MRI was done 3 months after starting the treatment. The collar was removed after seeing the cervical ligaments intact. She was followed-up for one year with neurological examinations. There was no neck pain or deformity at that time.

## DISCUSSION

In this case report, a 6 years old female child who had been diagnosed as Grisel's syndrome 3 weeks after



**FIGURE 1:** Cervical MRI of the patient. Axial **A**) and 3D remodeling of the cervical spine **B,C**) showing 20 degrees of C1 – C2 rotation before applying the cervical collar (Philadelphia). After the stabilization of the cervical vertebrae for 2 months, a new CT was done. Axial **D**) and 3D remodeling shows that the atlantoaxial subluxation is totally regressed and the alignment of the cervical vertebrae were normal

her cleft palate repair for velopharyngeal insufficiency has been presented. Grisel's syndrome, usually reported between ages 5 and 12 years, can also be seen in adult patients with equal incidence between males and females.<sup>2</sup> The patient presented in this report is also 6 years old child with velopharyngeal insufficiency.

Grisel's syndrome may have both inflammatory and nontraumatic causes. During cleft palate repair, nasopharyngeal veins may be affected. This plexus has not any lymph node, and any infective embolism may spread from superior pharyngeal area to the cervical joints via this plexus.<sup>4,5</sup> Vascular plexus of posterosuperior pharyngeal area draining this region hold responsible. Pharyngovertebral vein connects periodontoid plexus with posterior nasopharyngeal veins. Inflammatory mediators arising in relation to surgery or trauma may cause synovial and vascular congestion, inflammation around the transverse and alar lig-

aments which are the primary and secondary stabilizers of the atlantoaxial joint, and oedema of this region, resulting in weakening of the articular capsules, and eventually subluxation. Finally, it may cause imbalance of cervical spine which may be complicated by adverse neurological outcomes.<sup>4,6</sup>

Patients generally suffer from neck stiffness and painful neck movements, eventually the patient is seen at torticollis posture. This syndrome can also be seen after rheumatic conditions, rhinopharyngitis, cervical osteomyelitis, and surgical procedures such as adenoidectomy, tonsillectomy, repair of choanal atresia and mastoidectomy. Grisel's syndrome has also been reported after shock, mumps, or retropharyngeal abscess.<sup>2,3,6</sup> Down and Marfan-like syndromes which are associated with increased ligament laxity may increase the predisposition to Grisel's syndrome. Since children with Klippel-Feil, osteogenesis imperfecta, Morquio's syndrome, and neurofibromatosis have spinal instability, they have also higher risk for atlantoaxial subluxation. Majority of the reported cases were secondary to surgery of adenotonsillectomy, whereas our patient had cleft palate revision with double opposing Z-plasty and pharyngeal flap.

Torticollis, pain in neck motion, neck pain, cockrobin position, can be the early signs of Grisel's syndrome. Our patient has presented with torticollis, and painful neck motion after three weeks from her surgery. Having no consensus criterion for the diagnosis and treatment, early diagnosis is essential for good prognosis. Late diagnosis and inadequate or late treatment may cause neurological sequelae and/or painful and lasting deformity of the neck or surgical treatment.<sup>3,7,9</sup> Sensitive palpable C2 spinous processes (Sudeck's sign), tingling and neck pain at the upper and lower extremities in the neck flexion (Lhermitte's sign) are indicative of atlantoaxial subluxation.<sup>3,7,8</sup> Quadriplegia and sudden death had also been reported, though these are extreme consequences.<sup>6</sup> In radiological evaluation, which is critical for early diagnosis, increased atlantodens distance ( $> 3$  mm in adults and  $>5$  mm in children) could be detected in the sagittal sections.<sup>9</sup> CT and magnetic resonance imaging (MRI) are gold standard of diagnostic tools since the deep neck infection and re-

lationship between ligamentous and bone structure of the spine can be shown. The 3D (dimensional) reconstruction of the CT images is also useful. Dynamic CT can be demonstrative but it may cause neurological complications. There are no inflammatory indicators which are specific for diagnosis of this syndrome.<sup>3</sup>

Immediate treatment should be started in accordance with consultations with relevant departments in order to avoid any neurological complications which necessitate surgical interventions. Our patient had been consulted to pediatric neurosurgery and otorhinolaryngology and the treatment had begun as soon as the diagnosis had been verified with radiological findings. Treatment should be quickly initiated to avoid any neurological complications. The treatment of the acute syndrome is conservative; bed rest, immobilization, anti-inflammatory agents, muscle relaxants, antimicrobial therapy with antibiotics, external fixation, cervical traction, and/or simple traction with hard collar is mandatory at this period. If spasm of the cervical muscles persists longer than 24 hours, diazepam can be administered.

## CONCLUSION

Extreme head positions are sometimes necessary for cleft lip and palate surgeries and inflammatory conditions as well as protective head positions may accompany afterwards. Early diagnosis of Grisel's syndrome is very important for a successful treatment. This process requires multidisciplinary teamwork, meticulous clinical evaluation and confirmation by radiographic imaging.

### Source of Finance

*During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.*

### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

**Authorship Contributions**

**Idea/Concept:** Zehra Serpil Ustalar Özgen; **Design:** Zehra Serpil Ustalar Özgen, Seher İrem Kıran; **Control/Supervision:** Hakan Ağır; **Data Collection and/or Processing:** Seher İrem Kıran;

**Analysis and/or Interpretation:** Murat Başarır, Selin Unsaler; **Literature Review:** Seher İrem Kıran; **Writing the Article:** Zehra Serpil Ustalar Özgen, Murat Başarır; **Critical Review:** Hakan Ağır, Murat Başarır.

## REFERENCES

1. Bucak A, Ulu S, Aycicek A, Kacar E, Miman MC. Grisel's syndrome: A rare complication following adenotonsillectomy. *Case Reports in Otolaryngology*. 2014;1-4. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]
2. Serdaroğlu Beyazal M, Demirok D, Çapkin E, Usul H, Tosun M, Sarı A. Grisel's syndrome: a case report. *Turk J Rheumatol*. 2011;26(3):243-7. [[Crossref](#)]
3. Karkos PD, Benton J, Leong SC, Mushi E, Sivaji N, Assimakopoulos DA. Grisel's syndrome in otolaryngology: a systematic review. *Int J Pediatr Otorhinolaryngol*. 2007;71(12):1823-7. [[Crossref](#)] [[PubMed](#)]
4. Harma A, Firat Y. Grisel syndrome: nontraumatic atlantoaxial rotatory subluxation. *J Craniofac Surg*. 2008;19(4):1119-21. [[Crossref](#)] [[PubMed](#)]
5. Martínez-Lage JF, Morales T, Cornejo VF. Inflammatory C2-3 subluxation: a Grisel's syndrome variant. *Arch Dis Child*. 2003;88(7):628-9. [[Crossref](#)] [[PubMed](#)] [[PMC](#)]
6. Bociolini C, Dall'Olio D, Cunsolo E, Cavazuti PP, Laudadio P. Grisel's syndrome: a rare complication following adenoidectomy. *Acta Otorhinolaryngol Ital*. 2005;25(4):245-9. [[PubMed](#)]
7. Park SW, Cho KH, Shin YS, Kim SH, Ahn YH, Cho KG, et al. Successful reduction for a pediatric chronic atlantoaxial rotatory fixation. (Grisel syndrome) with long-term halter traction: case report. *Spine (Phila Pa 1976)*. 2005;30(15):E444-9. [[Crossref](#)] [[PubMed](#)]
8. Youssef K, Daniel S. Grisel syndrome in adult patients. Report of two cases and review of the literature. *Can J Neurol Sci*. 2009;36(1):109-13. [[Crossref](#)] [[PubMed](#)]
9. Dagtekin A, Kara E, Vayisoglu Y, Koseoglu A, Avci E, Talas D, et al. The importance of early diagnosis and appropriate treatment in Grisel's syndrome: report of two cases. *Turk Neurosurg*. 2011;21(4):680-4. [[PubMed](#)]