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Grisel's Syndrome: A Masquerader of Torticollis with a Hidden Atlanto-Axial Dislocation Agenda

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Abstract

Torticollis is a rather common presentation in pediatrics, with common etiologies. This is a rare presentation of a spinal pathology secondary to an underlying infection of the head and neck region causing anomalous head position masquerading as a torticollis. Painful torticollis, or abnormal head position described as "cock-robin head" position in children, draws attention. Grisel's syndrome (GS) is an inflammatory condition of a nontraumatic atlantoaxial rotatory subluxation or fixation (AARS/F). Diagnosis is derived clinically with radiological confirmation. The aim of this report is to raise attention of positional masquerade during intervals of radiographs, leading to the delay in diagnosis and heighten awareness of this syndrome that is needed to derive the diagnosis. In this case, the patient initially presented to the institution with a radiograph that had shown no overt abnormality and was requested for further detailed imaging, but the child's parents had refused. Upon a revisit to the clinic due to worsening of symptoms, a repeated radiograph was taken, a clear AARS was detected, and proceeded with a computed tomography (CT) cervical imaging to confirm the subluxation. The CT scan showed an overt subluxation with significant narrowing of the cord spaces with a retropharyngeal abscess. Otolaryngology and Spine services were consulted, and a surgical option was offered. However, the parents refused surgery and proceeded conservatively with antibiotics and a cervical halter. Early diagnosis and early treatment is fundamental to a better prognosis. Presence of GS is associated with late recovery in comparison to non-Grisel's causes of AARS/F. A treating surgeon should be aware of this condition and recognize it based on the "cock-robin" head position in absence of a triggering trauma with an underlying inflammatory pathology of the head and neck region. Presence of concomitant infection with AARS/F and Fielding and Hawkins classification types are prognostic factors for late recovery.

Keywords

- Grisel's syndrome
- atlantoaxial rotatory subluxation
- atlantoaxial rotatory fixation
- atlantoaxial instability
- retropharyngeal abscess
- cock robin
- torticollis

Introduction

Painful torticollis or abnormal head position in children draws specific attention. Several etiologies of torticollis occur, ranging from common causes like trauma, to more rare causes such as ocular causes and neoplasm or local infection, for example head and neck region infection like Grisel's syndrome (GS). GS is an inflammatory and nontraumatic atlantoaxial rotatory subluxation or fixation (AARS/F).[1] The anomalous head fixation, also known as "cock-robin head position," does not necessarily subluxate as in does in the present case, but frequently with

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only present with fixation which are within the normal range of movements.[1]

The pathogenesis of rotatory subluxation is the increased elasticity and destruction to the atlantoaxial structures due to the ongoing infection.[1] Diagnosis is derived clinically and confirmed radiologically. The aim of this report is to raise the awareness of positional masquerade during intervals of radiographs, leading to the delay or missed diagnosis. There should be awareness of this condition and recognizes it in patients with a "cock-robin" head position with absence of a triggering trauma with an underlying inflammatory pathology of the head and neck region.

Case

A 3-year-old boy presented with gradual worsening of neck pain for a month without history of overt infection: no fever, no recent fall or trauma, and no constitutional symptoms or tuberculous contact. He had multiple clinic visits for the similar complaint prior and was started on two courses of oral antibiotics with a provisional diagnosis of cervical lymphadenitis. The child was fretful with the neck in slight flexion, head tilted over to one side, and rotated to the opposite side suggesting a compensatory attempt to reduce the deformity and relieve infected tissues, resembling a "cock-robin" position.[2] His past medical and family history was unremarkable.

The cervical range of motion (neck rotation and lateral flexion) was deficit with bilateral sternocleidomastoid muscles that were still soft and not tender. There were multiple cervical, axillary, and inguinal lymph nodes palpable bilaterally. Neurovascular examination was unremarkable. Systemically, cardiorespiratory and abdominal examinations were normal. Initial laboratory results demonstrated elevated serum total white cell count of 16.10^9/L with inflammatory markers (erythrocyte sedimentation rate and C-reactive protein) of 40 mm/h and 2.2 mg/L, respectively (►**Fig. 1**).

Initial cervical radiograph showed no overt abnormalities. Subsequently, an ultrasound was ordered which reported bilateral cervical lymphadenopathy with no other mass noted. The parents were counseled for a contrast-enhanced computed tomography (CECT) of the neck, but they refused and requested for "at-own-risk" discharge to seek traditional treatment. The child was brought back to the hospital 4 days later due to worsening pain but without neurological deficit. A repeated cervical X-ray showed C1/C2 subluxation (►**Fig. 2**).

CECT neck was ordered which revealed $1.5 \times 4.5 \times 2.8$ cm low density fluid collection at the peri-vertebral region extending superiorly to the base of clivus and encasing the anterior arch of C1 and posteriorly to C4 vertebra level with bony erosion of C1 and C2 causing the atlantoaxial rotatory subluxation (AARS; ►**Figs. 3–5**).

The patient was diagnosed with GS, an AARS secondary to a retro-pharyngeal abscess. The otolaryngology division offered surgical drainage for the retropharyngeal abscess transorally with an occipito-spinal laminectomy and fixation under the orthopedics spine division. However, the patient's parent refused any surgical intervention and was discharged home with a sternal-occipital-mandibular immobilization (SOMI) brace and started on antitubercular treatment with a monthly follow-up till removal of the brace. A repeated CECT scan was arranged in 2 months, which noted resolving retropharyngeal abscess with bony sclerosis C1 and C2 and with no further displacement of atlantodental (►**Figs. 6** and **7**).

The orthotics was kept for a total duration of 18 weeks. Prior to removal of the orthotics a final dynamic radiograph in flexion and extension was taken, showing callus formation and stability. Postremoval, the patient is active, painless with resolution of the "cock-robin" head positioning (►**Fig. 8**).

Discussion

GS was first described by Charles Bell in 1830 and refers to "AARS untriggered by trauma, affecting patients with infections of the head and neck region or occurring following otorhinolaryngology procedures."[3] AARS is characterized by the loss of stability between the atlas and axis articulation secondary to

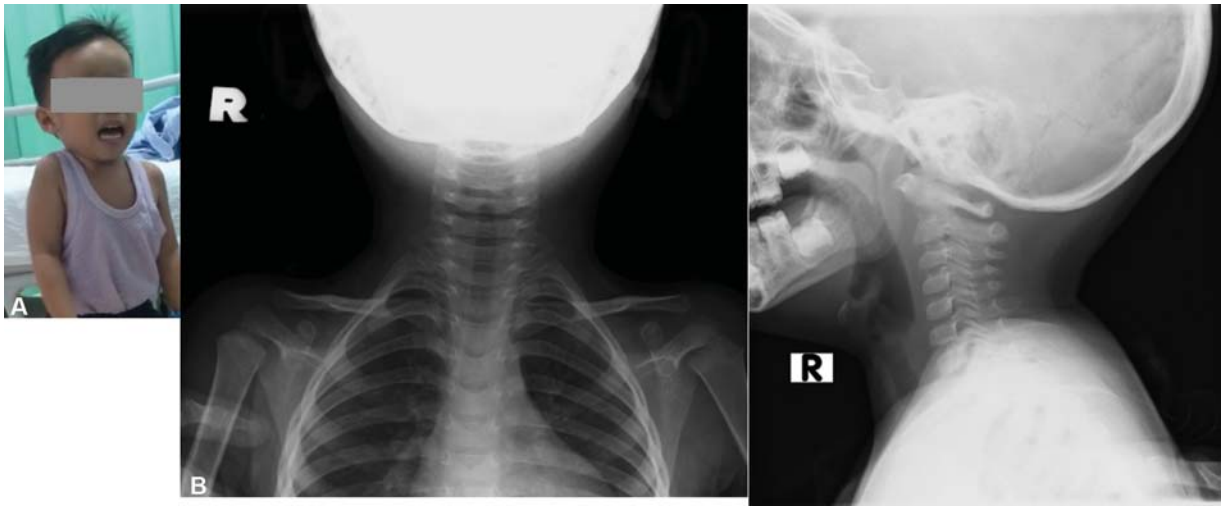

Fig. 1 (**A**) Clinical image showing the patient with an anomalous head position known as cock-robin head position. (**B**) Initial anteroposterior and lateral radiograph of the neck revealed no overt abnormalities.

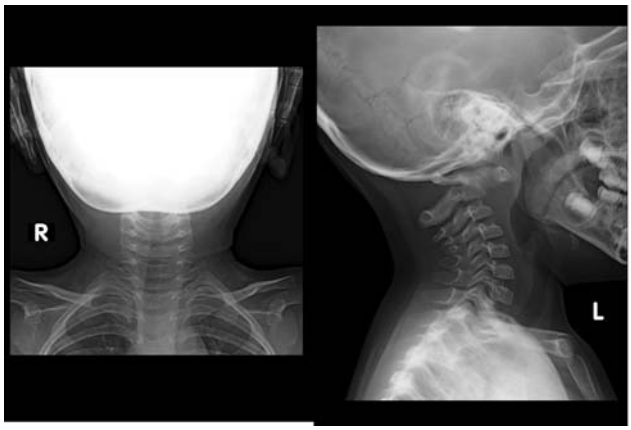

Fig. 2 Repeated anteroposterior and lateral cervical radiograph noted subluxation of C1/C2.

traumatic, inflammatory, congenital, or idiopathic causes.[4] Infection of the neck or otorhinolaryngology procedures may be a common cause for GS.[4] Local inflammation with ligamentous laxity and spontaneous muscle spasm occurs when inflammatory exudates are carried through pharyngovertebral veins to the cervical ligaments of the atlantoaxial joint. It connects the veins of the posterosuperior pharynx to the periodontal venous plexus, a lymphovenous anastomotic system.[4]

Few cases of GS have been described. GS presenting with a painful wryneck and swelling and cervical lymphadenitis.[5] Some patients complains of fever, sore throat, coryza, and tenderness at C2 spinous process. Usually, the neurological examination will be unremarkable. Clinically, the ipsilateral sternocleidomastoid muscle will have spasm, to which the neck will be in slight flexion with the head tilted over to one side and rotated toward the opposite side suggesting a compensatory attempt to reduce the deformity and relieve infected tissue, resembling a "cock-robin" position, which is similar to the way a Robin holds its head while listening for a worm.[2] In this case, absence of rotational deformity in the plain radiograph despite a cock-robin head positioning suggest a gross positional instability, which masked the deformity at the initial radiographs.

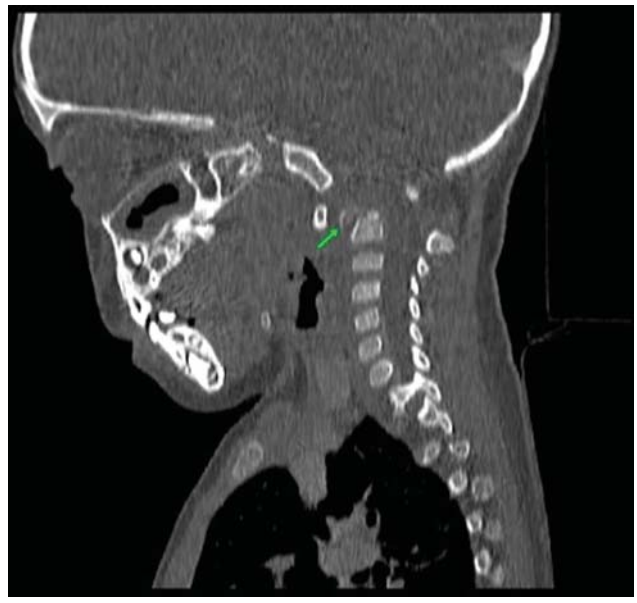

Fig. 4 Contrast-enhanced computed tomography neck sagittal view; arrow showed increased in atlanto-dental interval with irregular lytic erosion over odontoid process and anterior vertebra body C2.

A routine laboratory test will show an elevation of leucocyte, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP).[5,6] Raffaele Falsaperla et al (2018) suggested to include mycoplasma pneumonia antibodies as GS could be one of the extrapulmonary complications.[6] Plain cervical radiograph may show an increase atlanto-odontoid distance of more than 5 mm. A cervical computed tomography (CT) scan is much more sensitive in detecting early subluxation and rotation degrees of the C1 and C2 vertebrae.[3] Magnetic resonance imaging (MRI) is useful to detect any inflammation, collection, or edema at the retropharyngeal region (►**Table 1**).[3]

Recent studies by Aladag et al (2017) demonstrated successful conservative treatment for GS patients, which were diagnosed early within a month of having synptoms.[5] Surgical treatment should be the main mode of treatment in delayed cases depending on the radiological findings, which is indicated for those with an atlanto-dens interval of more

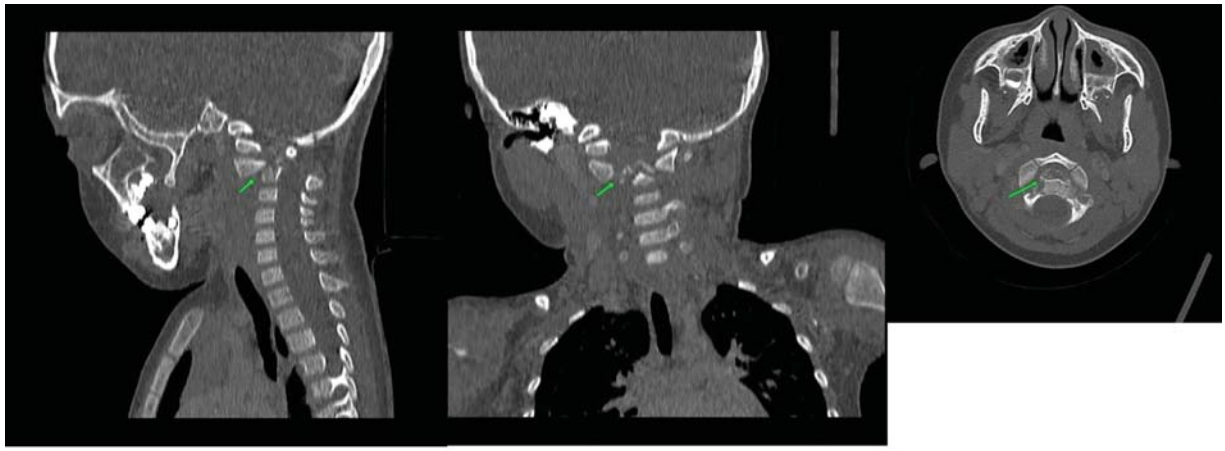

Fig. 3 Contrast-enhanced computed tomography neck; arrow showed subluxation of C1/C2 (sagittal, coronal, and axial view).

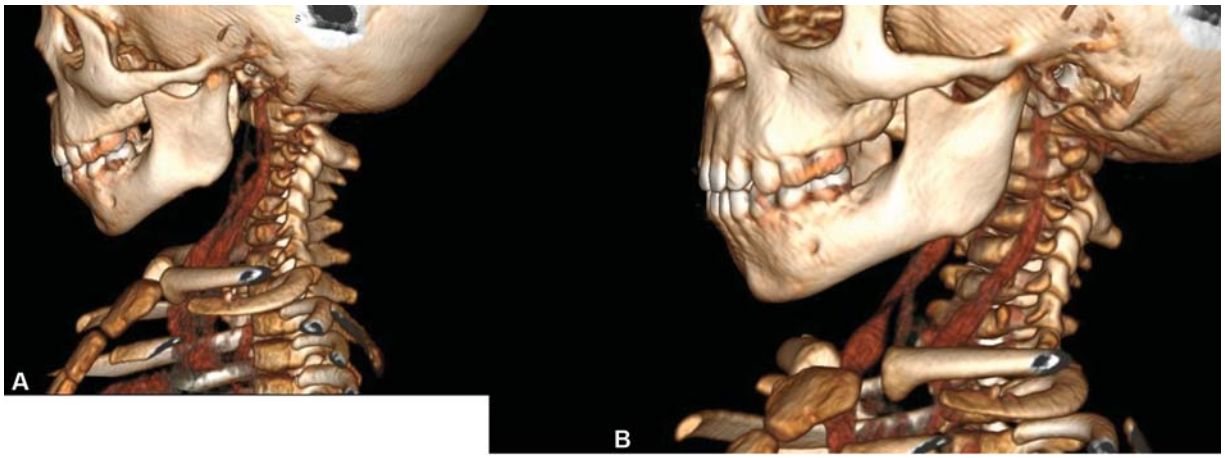

Fig. 5 Contrast-enhanced computed tomography neck (three-dimensional reconstruction) showing subluxation of C1/C2 (**A**) lateral view and (**B**) oblique lateral view.

Fig. 6 Patient on sternal-occipital-mandibular immobilization brace: anterior and side view.

than 4 mm and those with neurological involvement.[4] Another study by Pasquale Anania et al (2019) recommends Halo-vest with C1 to C2 arthrodesis or occipito-cervical arthrodesis for patients with late diagnosis depending on the degree of subluxation.[7] In our case, the patient had a good outcome from the SOMI brace alone with no further displacement.

A multidisciplinary approach in its treatment modality is suggested (►**Table 2**).

The characteristics of GS is nearly exclusive in the pediatric age group.[8] Hence, an early diagnosis is vital to ensue early commencement of treatment. The patient's history and clinical presentations are important cues to deriving the diagnosis, with further difficulty in history taking and examination of a child. First, the atlantoaxial rotatory subluxation/fixation (-AARS/F) should be immediately identified. Three clinical signs defining an AARS/F are (1) a palpable deviation at the spinous process, (2) spasm of the ipsilateral sternocleidomastoid muscle as a positional compensation secondary to pain, and (3) the inability of head to turn beyond the midline in the opposite direction to the injury.[8]

Not all entities might present simultaneously, and a higher suspicion is crucial to point toward the diagnosis, which can easily be missed as common causes of torticollis are still more common. Most of these patients had been reported to have normal neurology with normal biochemistry work-ups during presentation.[8] Biochemical findings are not a useful diagnostic

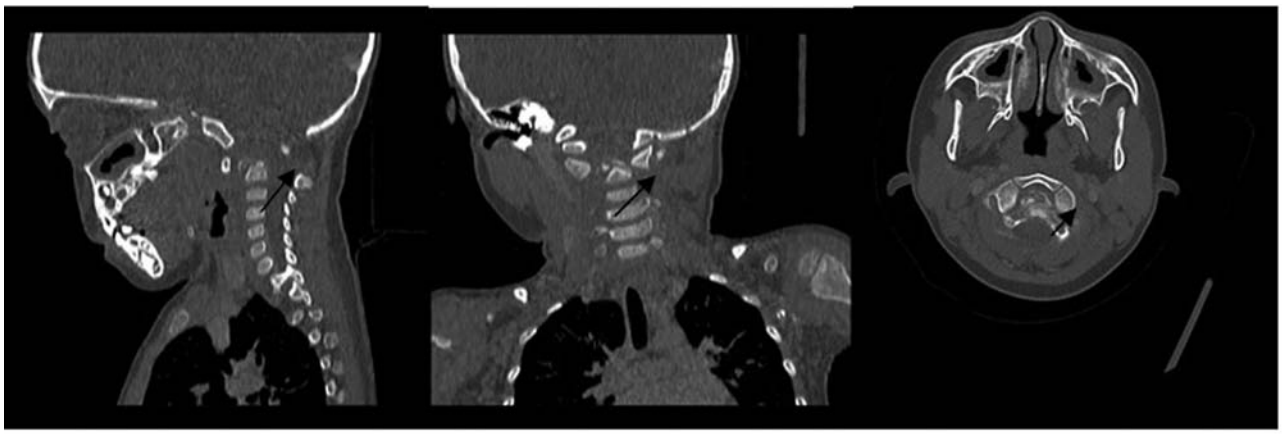

Fig. 7 Repeated contrast-enhanced computed tomography neck; arrow showed sclerosis of odontoid process with no further displacement of atlantodental interval (sagittal, coronal, and axial view).

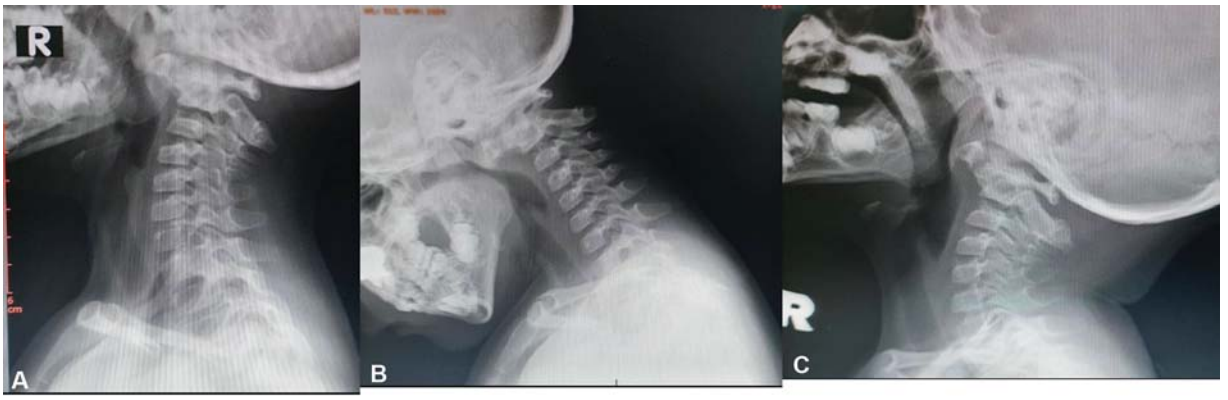

Fig. 8 Dynamic radiograph: lateral view in (**A**) neutral (**B**) flexion (**C**) extension at 18 weeks postorthotics application.

Table 1 Fielding and Hawkins classification of Grisel's syndrome

Type I	No subluxation Fixed rotation of atlas and axis Dislocation less than 3 mm anteriorly
Type II	Unilateral, atlanto-axial subluxation of one atlanto-axial joint Contralateral joint is pivotal Ventral dislocation between 3 and 5 mm
Type III	Ventral subluxation of C1 in both joints Ventral dislocation more than 5 mm
Type IV	Dorsal subluxation of C1 Combination with fractured dens axis or congenital dens-aplasia is possible

Table 2 A suggested treatment protocol according to the Fielding and Hawkins recommendations

Type I	Conservative treatment Antibiotics, muscle relaxant Immobilization with a soft collar
Type II	Conservative treatment Neck brace/Philadelphia-brace
Type III	Close reposition Halo-vest extension
Type IV	Operative treatment (decompression and arthrodesis of C1–C2)

tool in deriving the diagnosis; blood cultures are mostly negative and ESR are always elevated.[8] Radiographic findings play an important role in early detection of AARS/F: look for facet joints and available spaces asymmetry. The present case showed that this asymmetry could be altered by position; hence, a dynamic study might have been useful. Both CT or MRI are used diagnostically here, as a retropharyngeal soft tissue swelling was demonstrated.

Other differential diagnosis, for example, sternocleidomastoid tumors, muscular torticollis, vertebral neoplasm, or axial cervical fractures should be considered, as common causes are still more common. The rotatory subluxation is then graded with Fielding-Hawkins classification, which helps in deciding the management protocol.[9]

AARS/F with presence of concomitant infection or inflammation in the head and neck region has been shown to be an important prognostic factor associated with late recovery.[4] A study by Spinnato et al (May 2020) classifies recovery of patients into two groups: early recovery (i.e., healing at 3 months follow-up) and late recovery (i.e., persistence or relapse at 3 months follow-up and beyond).[4] The study series suggested the presence of GS is associated with late recovery versus noninfective causes of AARS/F. The Fielding-Hawkins classification significantly shows late recovery in higher Fielding-Hawkins grading.[4] C1–C2 rotational degrees show significant associations with patient recovery time ($p = 0.027$). Hence, presence of concomitant infection with AARS/F, Fielding-Hawkins classification, and C1–C2 rotational degrees are prognostic factors for late recovery.

Conclusion

GS is a rare cause for AARS/F leading to abnormal head posture (cock-robin head position) masked as torticollis. Early diagnosis and early treatment is fundamental to a better prognosis. The treating surgeon should be aware of this condition and recognize it in patients with a "cock-robin" head position with absence of a triggering trauma, but associated with an underlying infection of the head and neck region.

GS should still be in consideration in patients presenting with acute torticollis with or without history of prior infection or post aerodigestive tract surgery.

Note

The usage of the patient's clinical images is consented by the parents for publications and academic purposes. A written consent was granted and documented in the case notes.

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None.

Conflict of Interest

None declared.

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