Childs Nerv Syst (2003) 19:342–347
DOI 10.1007/s00381-003-0749-6

ORIGINAL PAPER

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Inflammatory atlanto-axial subluxation (Grisel's syndrome) in children: clinical diagnosis and management

Received: 14 August 2002
Revised: 25 February 2003
Published online: 3 June 2003

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Abstract *Background:* Grisel's syndrome is a uni- or bilateral subluxation of the atlas on the axis associated with an inflammatory condition of the head or neck. This uncommon entity usually affects children. The condition seems to be secondary to hyperemia and laxity of the atlanto-axial joints. *Patients and methods:* We report clinical and neuroimaging findings, treatment modalities, and outcomes in 4 children with atlanto-axial rotatory subluxation (AARS) associated with a previous upper respiratory tract infection treated at our Unit during the last 7 years. *Results:* All children presented with neck pain, head tilt, and reduced neck motion. In 3 patients, the condition was secondary to otitis media and in the remaining patient to viral pharyngitis. The diagnosis was established by clinical signs and symptoms and confirmed by neuroimaging studies, especially by computerized tomography and 3-D reconstructions. Early detection and treatment resulted in a good outcome in all patients. None of the patients required surgical treatment. *Conclusions:* Grisel's syndrome must be suspected in children with painful torticollis associated with an upper tract respiratory infection. The best results are obtained with early conservative treatment, which prevents chronic changes leading to persistent neck pain and deformity, namely atlanto-axial rotatory fixation (AARF). We emphasize the fact that the diagnosis of Grisel's syndrome is mainly clinical, while the diagnostic confirmation of AARF can only be attained by special neuroimaging techniques.

Keywords Atlanto-axial subluxation · Cervical spine pathology · Childhood spinal diseases · Inflammatory spinal diseases · Grisel's syndrome · Non-traumatic cervical subluxation

Introduction

Grisel's syndrome designates atlanto-axial rotatory subluxation (AARS) unrelated to trauma or bone disease [3, 5, 7, 15]. The condition is clearly predominant in children and characteristically follows an upper respiratory tract infection or surgery to the head or neck [3, 5, 7, 15]. In short, Grisel's syndrome is apt to occur in any condition that results in hyperemia and pathologic relaxation of the ligaments of the atlanto-axial joint [9, 15]. Several common entities, such as pharyngitis, adenoiditis, tonsillitis, tonsillar abscess, parotitis, cervical abscess, and otitis media constitute common causes of Grisel's syndrome. In addition, the syndrome has also been documented following diverse surgical ENT procedures like tonsillectomy, adenoidectomy, mastoidectomy, choanal atresia repair, and excision of neck tumors [1, 6, 7, 8, 13, 14, 15].

The current literature emphasizes that the diagnosis and treatment of Grisel's syndrome is frequently delayed [6]. In most cases, the atlanto-axial subluxation is easily reduced either spontaneously or by conservative means.

However, Fielding and Hawkins [2] reported a series of patients in whom the subluxation became irreducible, which they attributed to delayed diagnosis. They termed this condition atlanto-axial rotatory fixation (AARF) and recommended posterior arthrodesis for the relief of symptoms [2]. Apparently, this severe form of AARS appears when the initial transient changes in the atlanto-axial joints become permanent due to the production of irreversible structural changes.

We undertook a retrospective review of cases of non-traumatic atlanto-axial subluxation seen at our Unit during the last 7 years. Our aim was twofold: firstly, to draw attention to this incompletely understood entity, and secondly, to emphasize the importance of early recognition and treatment of the entity to patients' outcome. We suggest that the diagnosis of Grisel's syndrome is made primarily on clinical signs and symptoms and supported by radiographic findings. Left untreated, the syndrome may evolve and become the permanent variety of the deformity, namely AARF. The diagnosis of AARF can only be established by the special neuroimaging techniques already reported [11].

Patients and methods

The medical records of children diagnosed with cervical spine lesions seen at our Unit during the last 7 years (1995–2001) were searched for those corresponding to the diagnosis of AARS. Patients with previous cervical spine trauma (n=5) or with congenital conditions such as Down's syndrome, Morquio and Marfan diseases, or juvenile rheumatoid arthritis were excluded from the survey (n=2). Four patients fulfilled the diagnostic criteria for Grisel's syndrome [3, 5, 7]. We analyzed patients' age and sex, associated pathological conditions, duration of the symptoms before diagnosis, and clinical presentation. All the children were submitted to a thorough neurological examination to rule out other conditions apt to evolve with torticollis. Routine blood analysis, plain radiographs, and computerized tomography (CT) scans of the cervical spine were performed in all the children. Three-dimensional CT (3D-CT) scans were also available for all the patients and magnetic resonance imaging (MRI) for one. The diverse treatment modalities were also investigated. Follow-up time averaged 28 months (range 12 to 54 months). The review included clinical evaluation and repeat radiographic studies. The children's outcomes were determined from the patients' files of the Outpatient Clinic.

Results

Clinical findings

There were 3 boys and 1 girl in the study group whose ages, at the time of consultation, ranged from 4 to 13 years and averaged 7.5 years. Symptoms had been present for a mean of 65 days (range 1–210 days). All the children presented with neck pain, head tilt, and diverse degrees of restricted cervical mobility. All children were neurologically intact and exhibited the typical "cock-robin" position (head tilted to one side, rotated to the opposite side, and in slight flexion (Fig. 1). Three patients had a close antecedent of otitis media and had been treated with antibiotics. One child had been diagnosed with viral pharyngitis and had not been given treatment. Hospital admission was not

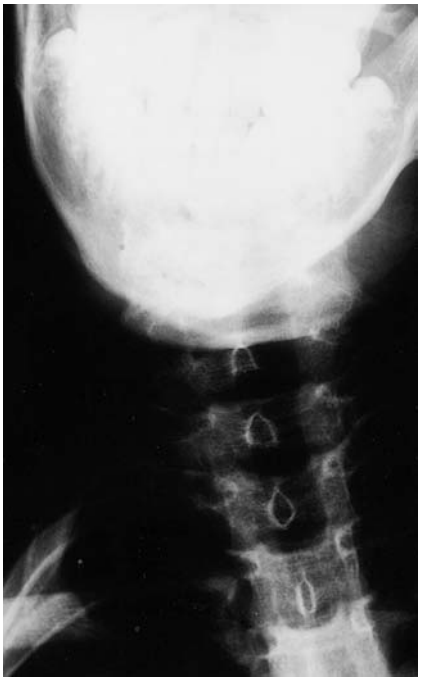

Fig. 1 Antero-posterior radiographs of the cervical spine showing the typical head tilt ("cock-robin" position) in Patient 3

Table 1 Clinical features of 4 patients with Grisel's syndrome (*NSAID* non-steroid anti-inflammatory drugs, *BZD* benzodiazepines)

Number	Age, sex	Association	Duration	Symptoms	Treatment	Outcome
1	5, boy	Otitis media	7 months	Pain, restricted neck mobility	Collar, SOMI brace	Mild restriction of neck mobility
2	8, girl	Otitis media	20 days	Pain, head tilt, limited neck mobility	Collar, SOMI brace, early physiotherapy	Asymptomatic
3	13, boy	Viral pharyngitis	1 month	Pain, head tilt, limited neck mobility	Collar, NSAID, BZD, early physiotherapy	Asymptomatic
4	4, boy	Otitis media	1 day	Pain, head tilt, limited neck mobility	Collar, NSAID, early physiotherapy	Asymptomatic

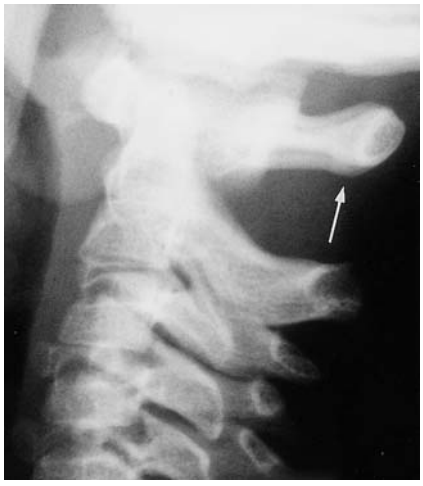

Fig. 2 Lateral cervical radiograph of Patient 1 depicting a widened inter-spinous distance C1–C2 (*arrow*)

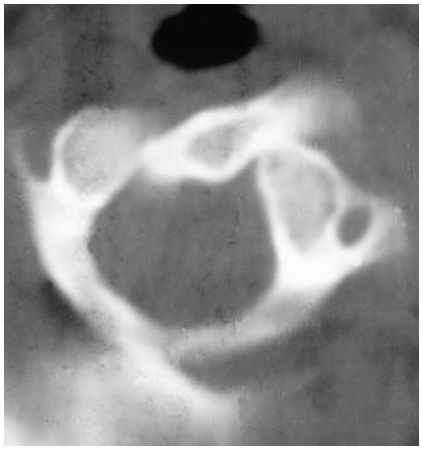

Fig. 4 Axial CT scan of Patient 2 illustrating the rotational component of the atlanto-axial subluxation

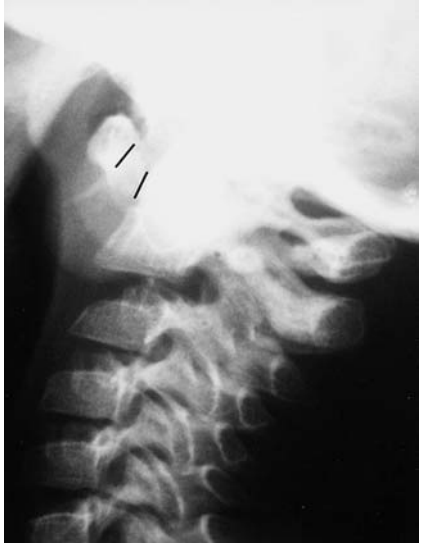

Fig. 3 Lateral radiograph of the cervical spine in Patient 4 depicting a widened atlas-odontoid distance (*lines*)

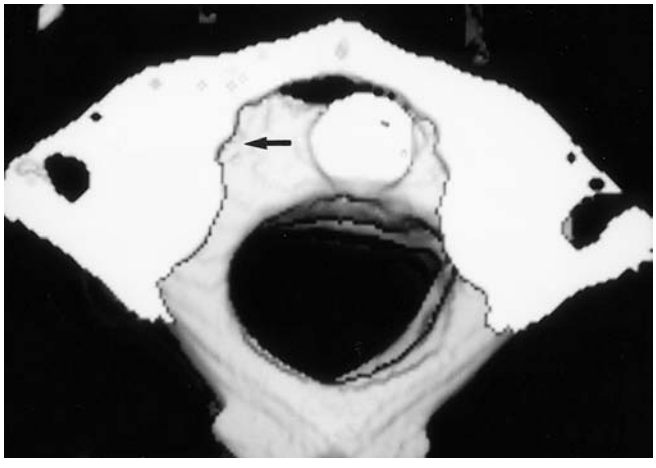

Fig. 5 Axial CT reformatted view of Patient 1 showing lateral separation and rotation of C1 on C2 (*arrow*)

deemed necessary for any of the patients. Clinical findings are summarized in Table 1.

Diagnostic procedures

The results of hematological and biochemical tests were within the normal range in all instances. Rheumatoid antibodies performed in Patient 1 were negative. Cervical spine plain radiographs were normal in 2 patients. The inter-spinous distance was widened in 2 patients (Fig. 2). The atlas-dens distance was less than 3.5 mm in 3 and 5 mm in the 4th child (Fig. 3). Antero-posterior views always showed the patients' head tilt (Fig. 1). Open-mouth projections, made in two instances, were worthless due to the odd head position that the children held to avoid pain. Flexion-extension radiographs were also obtained but gave no additional information to that given by the standard cervical films. CT scans of the cervical spine showed the abnormal position of C1 on C2 and especially the rotational component of the subluxation (Fig. 4). CT scans, with axial, coronal and 3-D reconstructions, were the most reliable technique for establishing a diagnosis of AARS (Figs. 5, 6). According to Fielding and Hawkins' classification of AARS, 3 patients had a type 1 and another had a type 2 subluxation [2].

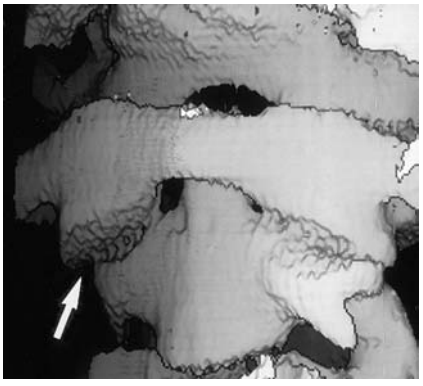

Fig. 6 Three-dimensional CT reconstruction of Patient 1 showing the forward and downward sliding of the articular process of C1 on the body of C2 (*arrow*)

Management

Patients 2, 3 and 4 were treated with the use of cervical immobilization, anti-inflammatory agents, and physiotherapy, including gentle head traction. Patient 3 was also given benzodiazepines at the onset. A SOMI brace was used in 2 patients. Patient 1 was initially managed with a hard collar for 2 weeks, and then with a SOMI device to achieve further immobilization. No physiotherapy, head traction or anti-inflammatory agents were used in this child. Given the good evolution of the children, surgery for atlanto-axial fusion was deemed unnecessary.

Outcome

After intervals of 1–4 weeks of conservative management with physiotherapy, anti-inflammatory therapy, and halter traction, patients 2, 3, and 4 were asymptomatic and had attained full-range cervical mobility. Plain radiographs and CT scans showed a full recovery of the position of atlas on the axis. After a follow-up of 4.5 years, Patient 1 was also asymptomatic but showed a moderate restriction for rotation movements of the neck. He had been treated by prolonged neck immobilization using a SOMI collar, but had not been given physiotherapy or head traction. This patient's plain radiographs and CT scan showed a reduction of the atlanto-axial subluxation. There were no cases of recurrence or of fixed deformity among our patients.

Discussion

Etiopathogenesis

There are some morphological and functional peculiarities in children that render them more susceptible to traumatic lesions of the cervical spine:

1. Larger head size in relation to the trunk
2. Cervical muscles are weaker
3. Ligaments and joints are looser
4. Facet joints, especially those of C1 and C2, are shallower and more horizontally placed
5. The cartilages of these joints have a convex surface that allows a wider range of motion
6. The uncinate processes in children are still scantily developed

All these features displace the fulcrum of head mobility to the upper segment of the cervical spine, give the spine a larger elasticity, but at the same time permit excessive displacements, and may ultimately lead to atlanto-axial subluxation [5, 15].

AARS is an uncommon acquired entity that produces displacement and limited motion of C1 on C2 [5, 15]. One lateral joint of the atlas slips on the body of the axis, usually forward and downward (Fig. 6) [6, 15]. Initially the dislocation can easily be reduced, but with time, the lesion may become fixed in the abnormal position, a condition usually recognized as AARF. For practical purposes, we have classified AARS into four groups according to its cause:

1. Trauma of variable intensity. In these instances, traumatic forces injure ligaments and joints initially producing subluxation and later abnormal fixation of the vertebrae.
2. Atlanto-axial dislocation associated with diseases of bone and connective tissues, such as Morquio disease, Down's and Marfan's syndromes, and juvenile rheumatoid arthritis. These conditions evolve with an increased laxity of the ligaments and joints that enables dislocation after trivial injuries or forceful cervical movements.
3. Grisel's syndrome is an inflammatory, non-traumatic subluxation of the atlanto-axial joint that follows a peripharyngeal infection. It has been attributed to laxity of the transverse and alar ligaments caused by hyperemia [9, 13]. A venous drainage system, from the posterosuperior pharyngeal region to the cervical epidural venous sinuses, has been documented [9, 13]. Because of the direct connection with the periodontoid venous plexus, the pharyngovertebral veins may provide a hematogenous route for the transport of peripharyngeal septic exudates to the upper cervical spinal structures thus providing an anatomical explanation for the atlanto-axial hyperemia reported in Grisel's syndrome [9, 13]. Pharyngeal inflammation may weaken the transverse ligaments and the joint capsules resulting in atlanto-axial instability and dislocation. Grisel's syndrome has been documented in otitis media, pharyngitis, retropharyngeal abscess, and viral infections of the upper respiratory tract. Despite these investigations, there is still some controversy about the pathogenesis of the syndrome [14]. Grisel's syndrome

occurred in approximately one-third of the patients diagnosed with AARS in several series [2, 10, 12].

4. Some patients develop AARS following surgical procedures on the head and neck [15]. These cases are most probably iatrogenic in nature and may have resulted from a forced position of the head under the effects of muscle relaxants used during anesthesia [6]. There is a report of AARS occurring after the insertion of a central-venous catheter [1].

Diagnosis

The usual presentation of Grisel's syndrome is with cervical pain, head tilt, and restricted and painful neck movements [5, 6, 7, 15]. Typically, there is an antecedent of previous upper respiratory tract infection or of a neck surgical procedure [5, 6, 7, 15]. The infection may be so unimportant, or remote, as to go unnoticed by the parents. The antecedent of a head or neck infection constitutes a crucial clue for the diagnosis of Grisel's syndrome and must be deliberately sought. The characteristic head position is 20° of tilt to one side, 20° of rotation to the opposite side, and slight flexion. This posture has been compared to that of a robin listening for a worm and has been named the "cock-robin" position [6, 15]. A thorough neurological examination is mandatory to rule out other causes of torticollis, especially posterior fossa and spinal cord tumors.

There are three clinical signs that may help to distinguish AARS from muscular torticollis of other etiologies. The first consists of the palpable deviation of the spinous process of the axis in the same direction of head rotation [12]. In normal head rotation, the process of C2 deviates to the opposite side. This is a very specific physical finding in AARS. The second is the spasm of the ipsilateral sternocleidomastoid muscle that the children exhibit to avoid pain [12]. The third sign is the inability to turn the head beyond the midline in the direction opposite to that of the injury [6, 12]. However, the diagnosis of Grisel's syndrome is made primarily on the basis of clinical signs and symptoms, namely the coexistence of AARS and an antecedent upper respiratory tract infection.

Differential diagnosis should be established against muscular torticollis, tumors of the posterior fossa and spinal cord, Chiari malformation, syringomyelia, trochlear nerve paralysis causing visual dysfunction, vertebral neoplasms, and fractures of the C1–C2 segment. In some cases, the deformity inflicts permanent changes and becomes fixed, at which point the condition is named AARF.

Neuroimaging

The radiographic diagnosis of AARS—and of Grisel's syndrome—is difficult, as standard radiographic projections alone do not confirm the diagnosis [4, 11]. The odd position of the patients' neck increases the difficulties involved in obtaining and interpreting the images. Static CT scans may achieve a greater resolution of the C1–C2 joint but they may also fail in disclosing the subluxation. Static CT scans and reconstructions aided in establishing the diagnosis in our 4 patients. However, even sagittal, coronal, and 3-D reconstructions may fail in differentiating normal rotation from AARS [4]. Because abnormal C1–C2 relationships can be observed in radiographs of children without evidence of AARS, neuroimaging studies cannot be the sole basis of the diagnosis. At present, MRI is being used to evidence lesions in transverse and alar ligaments and joint capsules. Rinaldi et al. performed dynamic CT studies in AARF and outlined the criteria for the diagnosis of the condition [11]. The technique involves three fine-cut axial CT scans of the upper cervical spine. The first scan is obtained with the head in neutral position and the next two scans are obtained with the patient's head rotated laterally in each direction until limited by discomfort [11]. The diagnosis of AARF is made by the presence of an absence of motion between the atlas and axis during rotation [11]. Rinaldi et al.'s technique for the diagnosis of AARS is a cumbersome procedure and is better reserved for those cases suspected of having fixation of the atlanto-axial joint, especially when surgical treatment is being considered.

Management

Once the diagnosis is established, treatment must be initiated soon and is directed toward reduction and stabilization of the joint [10, 12]. The length of time until reduction has been directly related to the failure of conservative treatment and to an increased risk of recurrence [10]. Cases diagnosed early can be managed with a 1–2-week trial of neck immobilization, anti-inflammatory therapy, and benzodiazepines [10, 12]. If there is no improvement in the patient's symptoms, gentle head traction, benzodiazepines, and physiotherapy should be employed [12]. After reduction, the children's neck is best immobilized with a collar for 6 weeks [12].

Patients with unreduced AARS for longer than 3 weeks are at higher risk of recurrence or permanent deformity, even after treatment, due to the development of chronic changes in the transverse and alar ligaments. These changes might explain the long-term sequelae of Patient 1 in our series. If a recurrence should occur, a second trial of closed reduction followed by prolonged neck immobilization (3 months) has been advocated [10, 12]. Patients with fixation or more than two episodes of recurrence might benefit from cervical fusion [10, 12].

Conclusions

Grisel's syndrome is an uncommon condition that we must bear in mind when neck pain, head tilt, and decreased neck mobility occur in a child with the antecedent of an upper respiratory tract infection or recent ENT surgery. A strong index of suspicion is required to diagnose this entity. Diagnosis is mainly based on clinical findings and is confirmed by a combination of neuroimaging studies (cervical plain radiographs, CT scans, and sagittal, coronal, and 3-D CT reconstructions). Once the diagnosis is established, treatment should be initiated soon, with intensive conservative therapy, including physiotherapy and head traction, and always in consultation with a pediatric neurosurgeon. Surgery is best reserved for cases of failed conservative management or recurrences with irreducible or painful neck deformity.

Acknowledgement The authors wish to thank Mr. Saturnino Espín, of the Hospital's Unit of Photography, for preparing the illustrations.

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