

ATLANTOAXIAL INSTABILITY IN CHILDREN WITH DOWN SYNDROME

INSTABILIDADE ATLANTOAXIAL EM CRIANÇAS COM SÍNDROME DE DOWN

INESTABILIDAD ATLANTOAXOIDEA EN NIÑOS CON SÍNDROME DE DOWN

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ABSTRACT

Atlantoaxial instability (AAI) occurs in 10 to 30% of Down Syndrome (DS) patients, with symptomatic disease ranging from 1 to 2%. Based on a rare clinical case, this article aims to highlight the main aspects of AAI screening and treatment. The authors report the case of a 4-year-old patient with DS who presented with quadriplegia after minor cervical trauma. Imaging studies revealed atlantoaxial dislocation associated with *os-odontoideum* (OsO). The patient was submitted to posterior C1-C2 arthrodesis (Harms technique) with an iliac bone graft. At three months post-surgery, the patient exhibited complete neurological recovery and remained neurologically intact and asymptomatic over the actual 10 years of follow-up. This case report highlights the importance of screening DS patients for myelopathic signs and symptoms. Cervical imaging should be reserved for symptomatic patients and surgery for patients with neurological compromise or an atlanto-dens interval superior to 10mm. When warranted, stable fixation with posterior screws appears to be the surgical technique of choice. **Level of Evidence V; Case Report.**

Keywords: Atlanto-Axial Joint; Down Syndrome; Diagnosis; Therapeutics.

RESUMO

A instabilidade atlantoaxial (IAA) ocorre em 10 a 30% dos doentes com Síndrome de Down (SD), sendo que 1 a 2% dos casos são sintomáticos. A propósito de um caso clínico, este artigo pretende destacar os principais aspetos do rastreio e tratamento da IAA. Os autores reportam o caso de uma criança de 4 anos com SD internada com um quadro de tetraparésia na sequência de trauma cervical menor. Os exames de imagem revelaram uma luxação atlantoaxial, associada à presença de *os-odontoideum* (OsO). A doente foi submetida a artrodese posterior C1-C2 (técnica de Harms), suplementada com enxerto de osso ilíaco. Três meses após a cirurgia, a doente apresentava recuperação neurológica completa, mantendo-se sem compromisso neurológico e assintomática durante os 10 anos de seguimento. Este caso clínico realça a importância de vigiar sinais e sintomas de mielopatia em doentes com SD. O estudo imagiológico encontra-se indicado em doentes sintomáticos e a cirurgia em casos de compromisso neurológico ou distância atlanto-odontoideia (DAO) superior a 10 mm. Quando indicada, a fixação posterior rígida com parafusos deverá ser a técnica de eleição. **Nível de Evidência V; Relato de Caso.**

Descritores: Articulação Atlantoaxial; Síndrome de Down; Diagnóstico; Terapêutica.

RESUMEN

La inestabilidad atlantoaxoidea (IAA) ocurre en el 10-30% de los pacientes con Síndrome de Down (SD), de los cuales el 1-2% presentan síntomas. A propósito de un caso clínico, este artículo busca destacar los principales aspectos del cribado y tratamiento de la IAA. Los autores reportan el caso de una niña de 4 años con SD ingresado con un cuadro de tetraparesia tras un trauma cervical leve. Los exámenes de imagen revelaron una luxación atlantoaxoidea asociada a la presencia de *os-odontoideum* (OsO). La paciente fue sometida a artrodese posterior C1-C2 (técnica de Harms), suplementada con injerto de hueso ilíaco. Tres meses después de la cirugía, la paciente presentaba una recuperación neurológica completa, permaneciendo sin compromiso neurológico y asintomática durante los 10 años de seguimiento. Este caso clínico resalta la importancia de vigilar los signos y síntomas de mielopatia en pacientes con DS. El estudio por imágenes está indicado en pacientes sintomáticos y la cirugía en casos de compromiso neurológico o distancia atlanto-odontoidea (DAO) superior a 10 mm. Cuando esté indicada, la fijación posterior rígida con tornillos debe ser la técnica de elección. **Nivel de Evidencia V; Informe de Caso.**

Descriptor: Articulación Atlantoaxoidea; Síndrome de Down; Diagnóstico; Terapêutica.

INTRODUCTION

Etiology and Clinical presentation

Atlantoaxial instability (AAI) occurs in 10% to 30% of Down Syndrome (DS) patients, with symptomatic disease ranging from 1% to 2%.^{1,2} It is the most common radiographically observed abnormality of the craniovertebral junction in DS patients.¹

Previous studies have concluded that AAI in DS patients is mainly attributed to transverse ligament laxity secondary to collagen defects and bony anomalies, such as *os-odontoideum* (OsO). Other factors include low bone mineral density, low muscle tone, and weakening of the ligamentous structures by a chronic inflammatory state.³ OsO occurs in approximately 6% of children with DS.⁴ It consists of an ossicle of varying size and shape, separated from

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the hypoplastic odontoid process by a gap that extends above the superior articular facets of C2. Instability in DS patients with OsO is more likely to develop during childhood and adolescence compared to adulthood, when ligament laxity is less prominent.⁵

Clinical presentation of AAI can range from acute neurological compromise after minor trauma to progressive neurological deterioration.⁶⁻⁸ Signs and symptoms suggestive of AAI include neck pain, changes in gait, lack of coordination, changes in bowel and bladder function, hyperreflexia, extensor-plantar reflex, muscle weakness, quadriparesis/quadriplegia, and/or torticollis.^{9,10} The latter is associated with C1-C2 rotatory subluxation.

Screening guidelines

With the growing participation of DS individuals in sports events, in 1983, the Special Olympics mandated that athletes with DS undergo radiographic screening of the cervical spine before joining a Special Olympics event. This became the stepping stone towards later establishing a standard routine radiographic screening for all children with DS. In 2001, the American Academy of Pediatrics (AAP) recommended that routine lateral cervical radiographs be obtained during preschool years (between 3-5 years).¹¹ Normal cervical radiographs before this age would not rule out AAI due to insufficient bone formation. Lateral cervical radiographs were taken in the neutral, flexed, and extended position, the atlanto-dens interval (ADI) was measured, and AAI was considered when the ADI was superior to 4.5 mm.³ Since they were issued, these recommendations have been criticized over the years, and some authors have attempted to suggest alternative screening guidelines. Performing routine screening radiographs for potential atlantoaxial instability in asymptomatic children is no longer recommended.¹²

Treatment guidelines and surgical approaches

To our knowledge, there are no current guidelines for treating AAI. Indications for surgery remain controversial and appear to be based on individual clinical experience. Early fusion is recommended in patients with neurological symptoms.^{13,14} Various techniques have been described in the surgical management of AAI. Early methods involving posterior wiring yielded discouraging results, with a high incidence of non-union.^{15,16} Meanwhile, posterior screw fixation has become the mainstay in surgical treatment of symptomatic AAI in DS patients.¹⁷⁻¹⁹ In 2002, Harms and Melcher presented a technique for posterior fusion using polyaxial screws placed in the lateral masses of C1 and pedicles of C2.²⁰ This technique overcame some anatomical difficulties related to previous posterior fixation techniques and proved a safe and efficient approach for C1-C2 fusion.

This article reviews the evolution of screening guidelines and presents the main aspects of AAI treatment based on a rare clinical case.

Case description

A 4-year-old female patient with Down Syndrome presented to the local hospital with a two-week course of gait instability and lower limb weakness, following a sudden neck movement while having her hair brushed by her mother. The child fell over from the lavatory where she was seated, and the mother unintentionally pulled her hair to prevent her from falling on the floor.

Her clinically relevant personal medical history included congenital cardiopathy with ventricular septal defect, atrial septal defect, and a patent ductus arteriosus, which was surgically corrected two months after birth. She had no history of previous radiographic screening for AAI.

After initial clinical evaluation at the local hospital, a cervical CT scan and MRI were performed, demonstrating anterior C1 dislocation with a rotatory component, associated with the presence of OsO (Figure 1), and spinal cord injury (Figure 2). She was transferred to our hospital on a rigid spine board. Upon arrival, the patient was hemodynamically stable and eupneic. She presented with torticollis associated with a neck “cock robin” posture and quadriparesis (grade 2/5 on her upper extremities and grade 3/5 on her lower extremities).

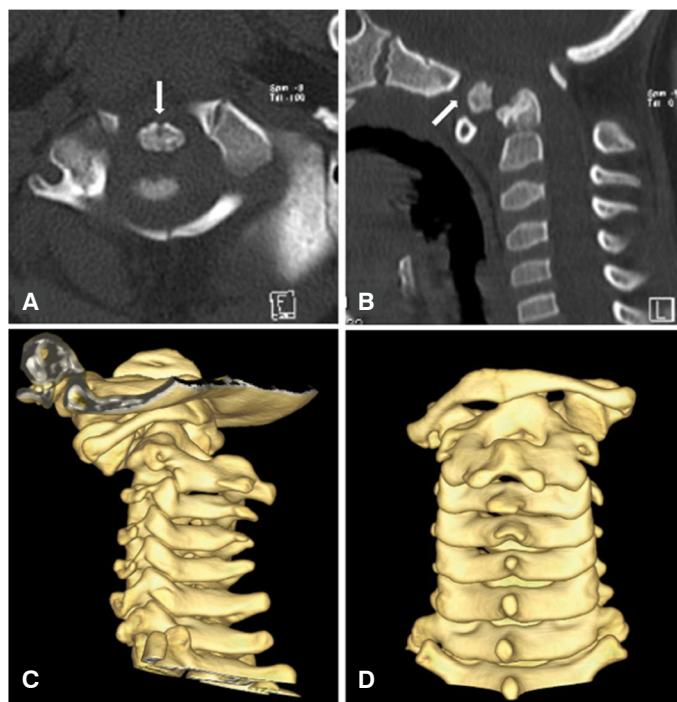


Figure 1. Preoperative cervical CT scan. (A) Axial and (B) sagittal images demonstrating C1 anterior dislocation associated with OsO (white arrows); the measured ADI was 6.1mm. Three-dimensional reconstruction with (C) lateral and (D) posterior cervical spine views highlights anterior C1 dislocation with a rotatory component.

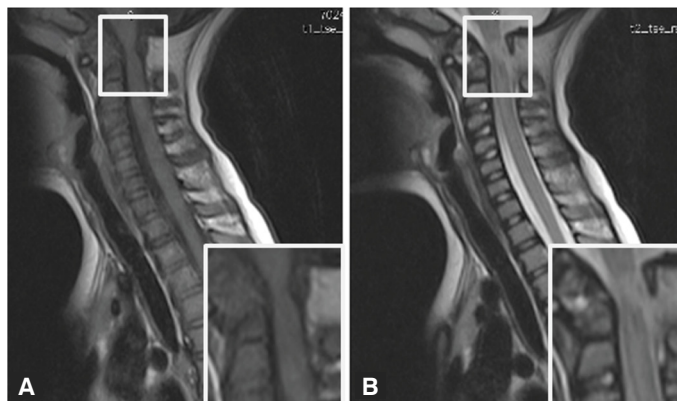


Figure 2. Preoperative MRI demonstrating spinal canal narrowing on a T1-weighted scan (A) and intramedullary hyperintensity at the level of the craniovertebral junction on a T2-weighted image (B), consistent with spinal cord injury.

The patient was admitted under the care of the Neurosurgery Department with a cervical collar, spinal injury precautions, and intravenous dexamethasone. At one week, she had regained full motor function in the lower extremities and left upper limb while maintaining motor deficits in her right upper limb (grade 3/5). Despite asymptomatic she kept the “cock robin” deformity. At two weeks, the case was discussed with our team at the Orthopedics Department, and the patient was scheduled for surgery. By then, the patient regained normal motor function in the right C5/C6 myotomes, although maintaining a muscle function grade 3/5. A posterior reduction followed by C1-C2 fixation (Harms technique) was performed, supplemented with autologous iliac bone graft (Figure 3). Surgery went uneventfully with complete restoration of the cervical alignment, and three months postoperatively, the patient displayed a normal neurological exam. Follow-up radiographs confirmed successful C1-C2 fusion with maintenance

of appropriate cervical alignment (Figure 4). The patient remained neurologically intact (ASIA grade E) and asymptomatic. At 10 years of follow-up, the patient remains asymptomatic, exhibiting a solid fusion and no junctional instabilities (Figure 5).

DISCUSSION

Evidence of pathological motion at the atlantoaxial segment in DS patients was first described in 1965 by Tischler and Martel,²¹ With a case of symptomatic atlantoaxial subluxation being reported for the first time the following year.²² Significant effort has been made to identify anatomical abnormalities present at the craniovertebral junction that predispose to subluxation and subsequent neurological compromise. In several studies conducted on AAI in DS,

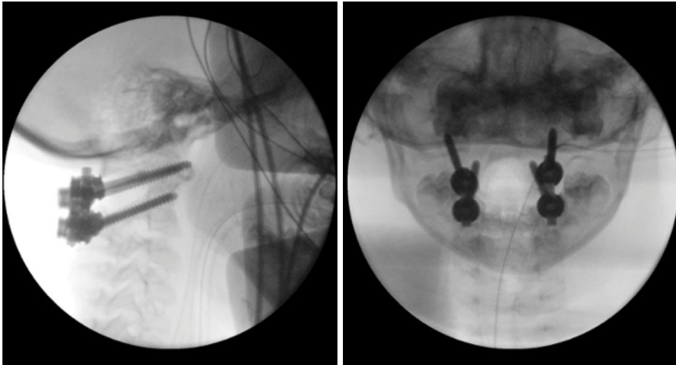


Figure 3. Intraoperative imaging documenting the placement of polyaxial screws in the lateral masses of C1 and pedicles of C2 (Harms technique).

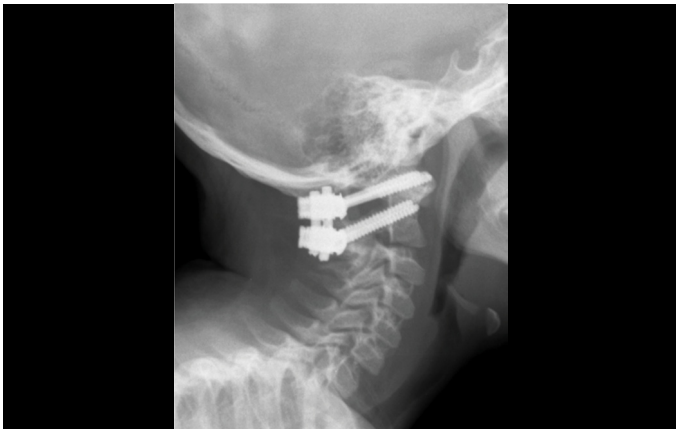


Figure 4. Follow-up radiography demonstrating adequate cervical alignment and successful C1-C2 fusion.

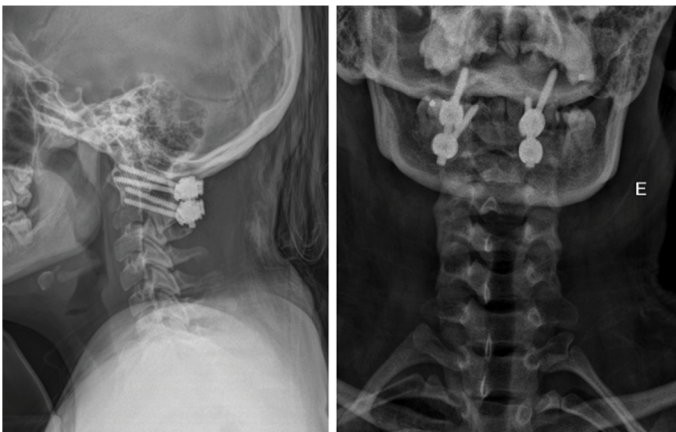


Figure 5. Cervical radiographs at the 10-year follow-up appointment.

OsO has been identified in a significant percentage of patients.^{2,23} With growing evidence that the incidence of OsO is higher in patients with conditions characterized by instability at the craniovertebral junction, discussion on whether this is the cause or effect arose. Since the traumatic etiology for OsO is gaining general acceptance, the association can be explained by the frequent hypermobility at the atlantoaxial complex in DS patients, causing repeated minor trauma and fracturing the odontoid process.^{2,24} Because the OsO is not attached to the odontoid, which produces a gap that extends above the superior articular facets of C2, the transverse ligament becomes incompetent.²

Due to the potentially catastrophic consequences of AAI, significant concerns were raised regarding the participation of DS patients in sports events. Consequently, in 1983, the Special Olympics required lateral cervical radiographs to be performed before DS patients participated in athletic competitions. Those with radiographic evidence of instability were banned from activities associated with increased risk of atlantoaxial dislocation.²⁵

In 2001, the AAP published recommendations that suggested screening for AAI in *all* children with DS between 3 and 5 years with lateral cervical radiographs in the neutral, flexed, and extended positions.¹¹ Since then, controversy has developed around the value of lateral cervical radiographs in detecting patients at risk for developing myelopathy. In fact, research conducted over the following years provided evidence that argued against it. Arguments included the rarity of neurological compromise in patients with documented AAI, the possibility that a patient's radiologic status can change over repeated screening, and the absence of evidence that radiographic screening is effective in preventing the development of symptomatic AAI.²⁶ Finally, the inherent technical difficulties associated with the lack of cooperation from young patients also posed a limitation.

It was only in 2011 that the AAP published new guidelines for health supervision in children with DS, promoting radiographic evaluation only in patients with clinical evidence of AAI.²⁷ Finally, in 2015, the Special Olympics replaced radiographic screening with a focused neurological evaluation.²⁸

A recent study has provided evidence supporting the de-escalation of screening recommendations.²⁹ Of 1566 DS patients included in the study, the authors identified only three children (0.19%) who might have benefited from radiographic screening before developing neurological symptoms. This study also reported on a patient who underwent radiographic screening with normal cervical radiographs and later developed AAI, requiring surgical intervention.

Although the clinical case we present dates to early 2012, shortly after the new screening guidelines were implemented, our patient had no history of previous radiographic screening for AAI. Therefore, it is impossible to make any tentative conclusions on whether radiographic screening could have led to an earlier diagnosis or prevented the development of neurological symptoms in this case.

According to the latest AAP guidelines issued in 2022, the current norm remains careful screening for signs and symptoms of myelopathy over standard radiographic screening. Of note, patients who are symptomatic should first undergo cervical spine radiography in the neutral position; flexion and extension radiographs are warranted if no significant radiographic abnormalities are present in the neutral position.¹² Correspondingly, in a retrospective study reviewing the efficiency of neutral position lateral cervical radiographs compared to flexion/extension radiographs, only one of 88 patients presented with abnormal findings on flexion/extension that were not detected on neutral position cervical radiographic imaging.³⁰

While screening guidelines have been the point of debate regarding AAI in DS patients, literature concerning treatment guidelines is contrastingly sparse. Although many authors have suggested protocols based on their individual clinical experience, to our knowledge, there are no current guidelines for treatment. The main indication for surgical treatment of AAI is the presence of associated neurological symptoms.^{2,13,14,31} According to some authors, an ADI greater than 10mm, despite the presence of symptoms, also constitutes an indication for surgery.³¹ While there is no established

timing for surgery, early fixation is recommended.^{13,32} Nevertheless, placing the patient under traction preoperatively has been proposed to aid in reduction, particularly in patients with OsO.^{13,32} In fact, the presence of OsO in association with C1-C2 subluxation may pose a significant challenge to reduction.²

Case series describing the outcomes of traditional implants, including cables and wires, demonstrated high postoperative complications, such as nonunion and high reoperation rates.¹⁵ Neurological compromise was often reported, particularly in sublaminar wiring.^{33,34} Although postoperative halo-vest immobilization could augment the fusion rates of traditional implants, this has been associated with significant comorbidities.¹⁷

Since the development of recent implants, such as screws and screw-rod constructs, studies describing the outcomes of these techniques have been limited to small sample sizes. Nevertheless, clinical outcomes appear to be promising. In a case series of twelve patients with DS, B.W. Yang et al. reported a complication rate of 41.7% (nonunion, wound breakdown, and vertebral artery injury), with a revision rate of 33.3%. At the final follow-up, 100% of patients obtained radiographic union.³¹ Li et al. reported on five DS patients with the diagnosis of OsO and spinal cord injury secondary to AAI treated with posterior screw fixation. At follow-up, all patients presented with evidence of fusion and an ASIA grade E.³⁵ Similarly, our case illustrates a successful example of neurological recovery and stable fixation after posterior screw instrumentation.

In most cases of AAI requiring surgical treatment, C1-C2 posterior screw fixation is currently recommended.^{2,17,20,35} The first posterior screw fixation techniques were developed using transarticular screws. The use of this technique presented a challenge in cases of anatomic variants of the vertebral artery, in which bilateral placement of these screws might be precluded.^{2,17,20} The first screw-rod system

developed by Harms and Melcher, using polyaxial screws for C1 lateral mass and C2 pedicle fixation, presented as a safe alternative with high fusion rates, overcoming the anatomical limitations associated with transarticular screw placement.^{17,20} This consists of adapting the biomechanical principles behind Goel's screw-plate system into a screw-rod construct.³⁶ even though several modifications to the original screw-rod technique have been reported, the technique developed by Harms and Melcher remains the most popular for posterior atlantoaxial fixation.³⁷ Therefore, the surgical technique and clinical outcomes described in our illustrative case align with current tendencies in the management of AAI.

The clinical case we present constitutes a paradigmatic example of AAI in DS. Despite the scarcity of cases of atlantoaxial dislocation in DS patients described in literature and treated in our hospital, the potential severity of this injury renders this topic highly relevant. Considering the current understanding that radiographic screening for AAI is not a strong predictor of the risk of atlantoaxial dislocation and neurological injury, our general recommendation is that cervical spine radiographs should be performed only in cases of neurological symptoms. Patients and their families must be alerted to potential signs of AAI, and cervical spine precautions in daily activities must be discussed.

In our experience, the prognosis after surgical treatment is favorable even in cases of atlantoaxial instability with associated myelopathy. Finally, we strongly believe that this knowledge is of the utmost relevance to any physician caring for DS patients.

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