Diagnosis of Atlantoaxial Instability Requires Clinical Suspicion to Drive the Radiological Investigation

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Introduction

Atlantoaxial instability (AAI) occurs as a result of trauma, congenital conditions such as os odontoideum, neoplasm, infection and degenerative connective tissue disorders such as rheumatoid arthritis, genetic conditions such as HOX-D3 and Down syndrome, and heritable connective tissue disorders, emblematic of which are the Ehlers Danlos syndromes (EDS). Prototypical of disorders in which AAI is diagnosed, is rheumatoid arthritis (RA). Prior to the development of effective disease-modifying pharmacotherapies, 88% of RA patients exhibited radiographic evidence of C1-C2 involvement, in whom 49% were symptomatic and 20% myelopathic; ultimately, 10% may have suffered atlantoaxial dislocation and death [1-3].

In their report on pediatric patients undergoing C1-C2 transarticular screw fixation Gluf and Brockmeyer noted approximately one third of the cases of AAI resulted from trauma, one third from os odontoideum, and one third from congenital conditions such as Down syndrome, Stihl disease, dwarfism, Morquio syndrome, Klippel-Feil and others. Three patients were thought to have chronic instability, most likely to have resulted from ligamentous laxity, possibly EDS [4,5].

The diagnosis of AAI is not difficult in the presence of an abnormal ADI (Figure 1), but may be elusive in those cases where the transverse ligament is intact, and where there is incompetence of one or both the alar ligaments. In these cases, the diagnosis may require dynamic imaging and concordant clinical findings.

Etiology of Atlantoaxial Instability

Traumatic flexion of the neck may result in injury to the transverse odontoid ligaments and alar ligaments. An atlanto-dental interval (ADI) over 3 mm suggests possible instability in adults; an ADI exceeding 5 mm suggests instability in children. An ADI of 7 mm suggests rupture or incompetence of the transverse ligament, and/or of the cruciate ligament, and an ADI of 10 mm suggests loss or incompetence of the alar ligaments as well [6]. However, if the transverse ligament is intact, the ADI is normal despite the presence of alar ligament incompetence.

A proclivity to ligamentous incompetence renders the atlantoaxial joint at higher risk for instability. The atlantoaxial junction (AAJ) is the most mobile joint of the body. Held together by ligaments that allow a great degree of freedom of rotation, the AAJ is responsible for 50% of all neck rotation, 5° of lateral tilt, and 10° to 20° of flexion/extension [7]. It is not surprising therefore, that connective tissue disorders, such as Down syndrome and EDS, are more frequently visited by AAI. Motor delay [8,9], headache associated with "connective tissue pathological relaxation" and quadriparesis are attributed to ligamentous laxity and instability at the atlanto-occipital and atlantoaxial joints [10,11]. While the epidemiology of AAI in EDS-hypermobile type is unknown, AAI was seen in two thirds of patients with EDS-Vascular type [11]. A high risk of AAI is apparent in other connective tissue disorders, including 11% of Down syndrome patients [12].

The AAJ mechanical properties are determined by ligamentous structures [13,14], most prominent of which are the transverse and alar ligaments [15]. The alar ligaments limit axial rotation and lateral bending to the contralateral side, are often injured in motor vehicle collisions, and could be implicated in whiplash-associated disorders [15]. Failure of the alar ligament allows a 30% increased rotation to the opposite side [16]. The atlantoaxial joint is ill-equipped to handle the required multi-axial movements in the presence of ligamentous laxity or disruption [17]. Weakness of the muscles and ligaments, hormonal changes, infection, immunological problems, and congenital dysmorphism may contribute to the overall mechanical dysfunction at the C1-C2 motion segment.

Hypermobility of the AAJ is common in children, and up to 45° of rotation may be observed in each direction. However, in the adult there is substantially less than 40° of rotation [17-20]: at 35° of rotation of C1 upon C2 there is stretching and kinking of the contralateral vertebral artery [20]; at 45°, both vertebral arteries become occluded [21].

Diagnostic Findings

Diagnosis of AAI is based upon careful history, a detailed neurological exam and imaging of the upper cervical spine. The most common clinical features are neck pain and suboccipital headache, with the caveats that headache is present in 50% of patients with EDS [22], and that moderate pain is a common occurrence for most EDS patients. There may be symptoms referable to the vertebral artery blood flow, including visual changes, as well as headache associated with the vertebral artery itself. Syncopal and presyncopal events are frequent. Other symptoms include dizziness, nausea, sometimes facial pain, dysphagia, choking, and respiratory issues. There is usually improvement with a neck brace. Examination often demonstrates tenderness over C1-C2, altered mechanics of neck rotation, hyperreflexia, dysdiadochokinesia, hypoesthesia to pinprick. Weakness is not a constant feature of AAI [4,23-27].

Radiological features may show atlanto-dental interval (ADI) >2.5 mm in an adult, or 5 mm in a child, on lateral flexion x-rays (Figure 1) or rotation of C1 upon C2 >41° [4,28] (Figure 2). Excessive rotation occurs opposite to the side of an incompetent alar ligament. Alar ligament incompetence is the most frequent cause of AAI in the population of patients with hypermobility connective tissue disorders [4]. Thus diagnosis of AAI in EDS, or related connective tissue disorders, is best made with supine computerized tomography (CT), from occiput to C2, with full neck rotation (90° if possible) to left and to right. (Figure 2) The difficulty of recognizing rotary instability on standard x-ray, CT and MRI images has resulted in failure to diagnose [29].

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In the hypermobility disorders, there may be abnormal facet overlap on full neck rotation<20% [30,31] (Figure 3); lateral translation of the facet joints: translation in aggregate >7 mm on coronal imaging as seen on open mouth odontoid views. Open mouth odontoid views are very effective in identifying AAI [32] (Figure 4).

Dvorak showed in cadavers that the mean axial rotation between the axis and the second cervical vertebra was 31.1°, increasing to 35° after contralateral rupture of the alar ligament; CT imaging thus demonstrated increased angular rotation of 4° to the side opposite the alar ligament injury [16].

The diagnosis of AAI can sometimes be seen on three dimensional CT, where there may be a clear demonstration of subluxation [31]. Increased ligament signal intensity on high-resolution proton density-weighted MRI may be seen, with the caveats that alar ligaments of asymptomatic patients may show high signal intensity, and that there is variable inter-examiner reliability of MRI evaluation [33]. Other radiological indiations of AAI include compromise of the vertebral arteries based upon abnormal mechanics at the C1-C2 junction; anomalous joints; retro-odontoid pannus [23,27,34] (Table 1).

Criteria for Surgery

The decision to proceed to surgery rests upon the presence of severe neck or suboccipital pain, the presence of cervical medullary syndrome or syncopal (or pre-syncopal) episodes, demonstrable neurological findings and radiological evidence of instability or compression of the neuraxis.

Figure 3: Axial CT scan through C1-C2 showing loss of facet overlap on full neck rotation, consistent with AAI.
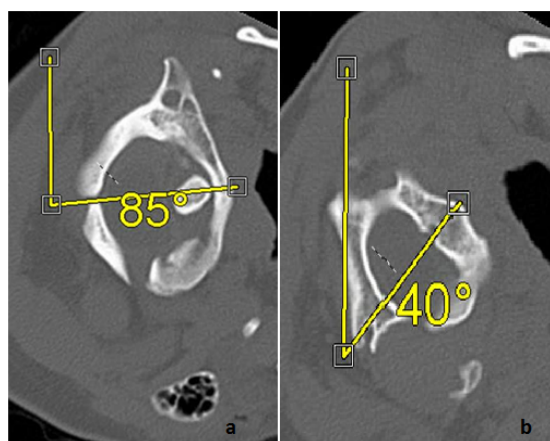

Figure 1: ADI >3 mm in an adult.

Figure 2: Excessive (>41°) rotation between C1 and C2. Axial CT views through (a) C1 and (b) C2 on full neck rotation demonstrate that the angle between C1 and C2 is (85°-40°) = 45°, and therefore unstable.

Figure 4: AP open-mouth radiograph of the C1-C2 levels showing pathological loss of overlap.

Treatment

For milder form of instability, the patient should be considered for treatment with neck brace, physical therapy and avoidance of activities that provoke exacerbation of the AAI symptoms. If the non-operative treatment fails, fusion-stabilization at C1-C2 is required. Failure of any of the components of the atlantoaxial ligament complex requires dorsal surgical fusion [35]. This is most often accomplished with posterior screw constructs, transarticular screw fixation [24], or C1-C2 lateral mass/pedicle screws and interposed graft [4,25,26] (Figure 5). Aberrant vertebral artery anatomy may preclude the desired screw placement in 18% to 23% of patients [36,37], and the surgery may be complicated in EDS by small bone architecture.

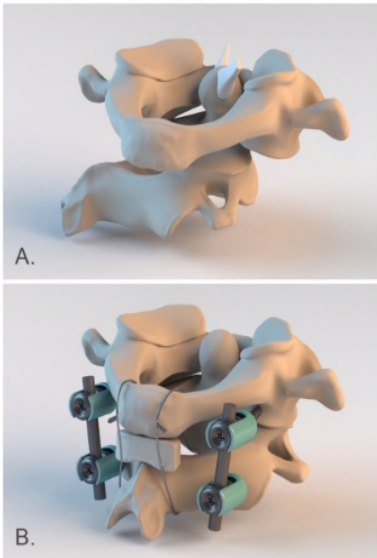| ADI (Atlanto-dental interval) >3 mm in an adult, or 5 mm in a child, on lateral flexion x-rays. |
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| Lateral translation of the facet joints: translation in aggregate >7 mm on coronal imaging as seen on open mouth odontoid views. Open mouth odontoid views are very effective in identifying AAI [32]. |
| Rotation of C1 upon C2, >41° [4,28] (Figure 1). |
| Facet overlap on full neck rotation <20% [30,31] (Figure 2). |
| Greater than 8° difference between the left and right axial rotation, on axial CT with full neck rotation [37]. |
| Three-dimensional CT demonstrating subluxation (Figure 3). |
| Increased ligament signal intensity on high-resolution proton density-weighted MRI, with the caveats that alar ligaments of asymptomatic patients may show high signal intensity and that there is variable inter-examiner reliability of MRI evaluation [33]. |
| Compromise of the vertebral arteries based upon abnormal mechanics at the C1-C2 junction. |
| Anomalous joints. |
| Retro-odontoid pannus [23,27,34]. |

Table 1: Radiological findings of AAI.

Figure 5: (a) Sagittal model of atlanto-axial rotary sub-luxation; (b) Treatment of AAI.

Occiput to C1/C2 fusion should be considered in the presence of craniocervical instability, basilar invagination or complex Chiari malformation.

Conclusion

AAI results from trauma, congenital conditions, neoplasm, infection, degenerative connective tissue disorders, genetic conditions such as the HOX-D3 or Down syndrome, and heritable connective tissue disorders, emblematic of which are the Ehlers Danlos syndromes. AAI in the hypermobility disorders usually requires dynamic imaging to demonstrate ligamentous incompetence. Radiological findings which are concordant with clinical findings should prompt consideration of surgery.

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