

Images in Spine Surgery: Atlantoaxial Instability in Down syndrome

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A 6 year old boy with Down syndrome, presented with a 6 month history of neck pain. The pain was located in the sub-occipital region with referral to the posterior part of the occiput, rostrally. His family had noticed that he had difficulty in walking and had unsteadiness of gait for a few months, which had led to a few falls. This was progressively getting worse. He did not report any loss of motor strength, sensory disturbance or bladder/ bowel incontinence. Examination showed that his neck pain was accentuated with flexion and extension. He had mildly increased tone in all extremities. His motor strength and sensory examination was within normal limits. He demonstrated hyper-reflexia in all four extremities and had a positive Babinski sign bilaterally. Sustained left sided clonus was also noted. On the flexion X-rays of his cervical spine an increased ADI (Atlanto-Dens interval) of 11 mm was noted. His CT scan confirmed this finding and also demonstrated an associated Dystropic Os Odontoideum. MRI examination showed an extensive area of high signal intensity at the cervico-medullary junction. The patient had application of a halo ring under fluoroscopic guidance and an awake intubation. He was placed in a halo vest for positioning prone. The prone positioning was performed under neuromonitoring with SSEP (Somatosensory evoked potentials) and MEP (Motor evoked potentials). This patient underwent a posterior occipitocervical instrumentation and fusion in the reduced position. He was instrumented using sublaminar wires as the anatomy of the Axis was deemed inappropriate for screw fixation. His Postoperative C-spine x-rays showed a reduced ADI and on flexion/extension views there was no sign of Atlanto-Axial instability. Post operatively he remained in a halo vest for 3 months. Resolution of patient's symptoms was noted on periodic follow-up exams with complete restoration to a stable gait.

Commentary

"Let me win, but if I cannot win, let me be brave in the attempt" - Special Olympics Oath.

Though first reported by Spitzer, the association of cervical spine instability with Down syndrome was highlighted by the Special Olympics. The three main types of cervical spine pathologies described in Down syndrome are Occipito-Atlantal and Atlanto-axial instability and disc or facet degeneration / instability at subaxial level. The causes of instability are due to bony anomalies and ligamentous laxity. Commonly associated bony cervical spine anomalies in Down syndrome are Os Odontoideum and hypoplasia of

various craniovertebral elements. Atlanto-axial instability usually co-exists with occipitocervical instability.¹ Neutral, flexion and extension cervical spine X-rays should be obtained and critically evaluated for Occipito-atlantal junctional integrity, Atlanto-Dens Interval (ADI) and at each cervical level for signs of disc or facet degeneration and instability.

About 10 to 30% of patients with Down syndrome will demonstrate radiographically confirmed evidence of atlantoaxial instability (increased ADI) on dynamic X-rays, although only about 1-2 % of Down syndrome patients may be symptomatic. Symptomatic patients often present between ages 5 and 15 years and present with neck pain, sensory deficits, or difficulty with bladder control. Progressively decreasing level of activities, ambulation and changes in gait can also be some of the subtle presenting symptoms. Physical examination may demonstrate a variety of upper motor neuron signs such as a wide-based gait, abnormal neck posturing, decreased neck range of motion, hyperreflexia, an abnormal Babinski reflex, ankle clonus, or weakness.²

The radiographic criteria of instability for other diagnoses cannot be applied to Down syndrome patients. Despite decades of work, the radiographic interpretation and determination of instability in Down syndrome has remained controversial. Some authors describe this phenomenon as hypermobility as compared to instability since most of the radiographic abnormalities are asymptomatic. The Atlanto-Dens Interval provides indirect information about the Space available for Spinal cord. Radiographic guidelines^{3,4} for treatment of an increased Atlanto-Dens Interval (ADI) due to Atlanto-Axial instability are as follows: with an ADI less than 4.5 mm no activity restriction is needed; with an ADI > 4.5 mm but less than 10 mm without neurological symptoms, it is recommended that patients avoid contact sports and high risk activities. In patients with an ADI of >4.5 mm and neurological symptoms, activity limitation is advised and an MRI examination should be performed. If the MRI shows any signs of spinal cord compression or signal change, surgical treatment is advised. In the case of a normal MRI with an ADI of >4.5 mm but <9.9 mm, observation is advised. If the ADI is found to be > 9.9 mm, surgical treatment is advised regardless of the presence or absence of neurological symptoms. In such cases the space available for spinal cord is just marginal and a sudden flexion of the neck due to any cause can lead to spinal cord injury.



Figure 1. (A) X-ray cervical Spine lateral view in Neutral position does not show any signs of instability. Normal ADI (Atlanto-Dens Interval can be noted) (B) Extension X-rays also do not show any instability. (C) Flexion X-rays show an exaggerated ADI and decreased space available for spinal cord posteriorly.

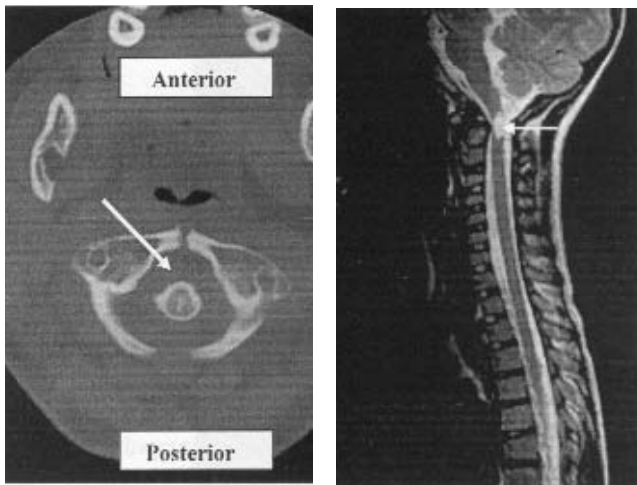


Figure 2. (A) Axial views CT scan cervical spine at C1-2 level show the increased ADI (arrow). (B) Sagittal section, T2 weighted MRI scan, shows high signal intensity (arrow) at the site of Atlanto-axial instability.

References

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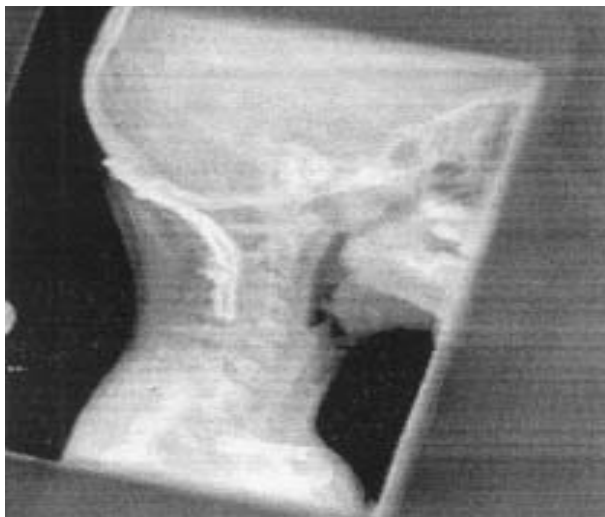


Figure 3. Postoperative films show Occipito-cervical instrumentation and fusion and a reduced Atlanto-Axial interval.