**Atlantoaxial Instability Screening**

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Atlantoaxial instability (AAI) is subluxation or slippage of the first neck vertebrae in relation to the second. It is more common in people with Down syndrome (DS) and there are unique screening considerations for people with DS compared to people without DS.

Here is what the American Academy of Pediatrics (AAP) writes about AAI or neck instability:

“Bones in the neck or spine can be unstable in some people with Down syndrome. There are almost always visible signs when there are problems. Daily physical activity is important to your child and should not be limited by unneeded worries. X-rays are not needed unless there is pain or changes in the use of hands, walking, or bowel or bladder function. If x-rays are done, and the results are abnormal, your child may be referred to a spine or neck specialist. Special neck positioning may be needed for some medical procedures. Let your child’s doctor know if your child is having: stiff or sore neck; change in stool or urination pattern; change in walking; change in use of arms or legs; numbness (loss of normal feeling) or tingling in arms or legs; or head tilt.”

At the Adult Down Syndrome Center (ADSC), we screen all our patients at their annual exam with questions about change in function or symptoms that could be related to AAI (similar to those described by the AAP). We also complete a neurological exam. In the past, we also screened all adults with DS on a regular, scheduled basis (usually every 10 years) with X-rays. However, after the AAP came out with their updated guidelines for children with DS in 2011, some healthcare providers at the ADSC stopped doing X-rays on a regular basis (see AAP quote above).

A common reason to do the X-rays is to complete a Special Olympics form. While the X-rays are still required in Illinois, nationally Special Olympics is not requiring the X-rays and they are phasing that into states over time. The Illinois Special Olympics staff report that, ultimately, they will not be requiring the X-rays.

We continue to do X-rays and often an MRI and/or CT scan of the cervical spine if a person with DS is having symptoms or signs of AAI. We have a strong index of suspicion for any symptoms that could be related to AAI and, therefore, we still do a fair number of X-rays. For example, we have found headaches to be a relatively uncommon complaint by our patients but, in those with the complaint, AAI was a fairly common finding. In addition, our overall experience with AAI is similar to that described in the literature that about 1% of our patients have required surgery to correct AAI.
In addition, there is the potential for cervical spine injury with extension of the neck during surgery and/or intubation in people with Down syndrome, even when they have a normal lateral cervical spine X-ray. When we evaluate patients for pre-operative clearance (for any surgery requiring anesthesia), caution is advised and fiberoptic visualization during intubation is recommended.

Efforts are underway to update the healthcare guidelines for adults with Down syndrome. AAI screening is one of the first 10 topics that are being addressed. Links to more information about these efforts are included in the References section below.

References

Pediatric Healthcare Guidelines for Families

Pediatric Healthcare Guidelines for Healthcare Providers
- [http://pediatrics.aappublications.org/content/128/2/393](http://pediatrics.aappublications.org/content/128/2/393)

Adult Healthcare Guidelines for Families

Updating the Adult Healthcare Guidelines