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# Atlanto-axial Subluxation and Trisomy-21: Another Perioperative Complication

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Atlanto-axial instability or subluxation in children is a rare disorder, usually occurring secondary to infections, collagen vascular disorders, or steroid therapy.[1] The incidence of atlanto-axial instability in children with trisomy-21, however, is 14–22%.[2–6] Although only a small percentage of these children with atlanto-axial instability will become symptomatic (15% of the original 22%),[3,4] many organizations require all trisomy-21 children be screened for altanto-axial instability prior to participation in vigorous physical activities.[7,8]

A significant proportion of children with trisomy-21 require surgical intervention for correction of developmental abnormalities of the eye, ear, nose, throat, and heart. The experience which we now report suggests that all trisomy-21 children over 4 yr of age who present for surgery should also be screened for this atlanto-axial instability. This will hopefully avoid the appearance of atlanto-axial subluxation during the perioperative period.

## CASE REPORT

A 6-yr-old boy had a symptomatic atrial septal defect (ASD). The past medical history included trisomy-21 with attendant dysmorphic features, juvenile rheumatoid arthritis (currently inactive), and progressive exercise intolerance for the past 4 yr. He had a history of two previous general endotracheal anesthetics for ophthalmic procedures without complications.

On physical examination, the patient was a well-nourished, well-developed boy with the typical features of trisomy-21. Physical examination was significant for nonobstructive macroglossia, normal dentition, and adequate neck mobility in all planes. The lungs were clear to auscultation and percussion. Cardiac examination revealed a grade II/VI murmur, but was otherwise unremarkable. Examination of his joints demonstrated no effusions or stiffness.

In the operating room, a peripheral iv catheter, arterial catheter, endotracheal tube, and right internal jugular catheter were inserted without incident. Following successful repair of his ASD, the child was taken to the pediatric intensive care unit, where the trachea was extubated the following morning without complication.

The patient did well until the fifth postoperative day, when the child was noted to be holding his head to the right. This was treated with analgesics, physical therapy, and local heat applications, and the patient was discharged the following day.

The child returned approximately 1 month later, complaining of severe right-sided neck pain. On neurologic examination, the patient had extreme pain on any attempted motion of the neck, and maintained his neck slightly flexed and turned to the right. Sensory and motor examinations were unremarkable.

Rotatory subluxation of the atlanto-axial joint was demonstrated on both computerized tomographic scan and antero-posterior neck roentgenograms without evidence of fractures or abnormal articulations. Somatosensory evoked potentials were normal bilaterally.

The child was placed in a hard cervical collar and scheduled for C1-C2 fusion and placement of a halo brace. The anesthetic was induced without incident, and the remainder of the operation was uneventful. The child was placed in the halo brace and the trachea extubated after establishment of spontaneous ventilation. The patient had a stable postoperative course, and was discharged on the seventh postoperative day. Follow-up at 1 and 3 months demonstrated no motor or sensory abnormalities, and the child has had no further problems.

## DISCUSSION

An increased incidence of atlanto-axial dislocation in children with trisomy-21 was first noted by Tishler and Martel in 1965. This child's presumed atlanto-axial instability was asymptomatic until the fifth postoperative day, and was manifested by deviation of the head to the right. It is impossible at present to determine exactly when or why this child's presumed instability progressed to become symptomatic subluxation. There are no studies which detail the natural history of asymptomatic altanto-axial instability. Furthermore, no preoperative roentgenographic evaluation was done, and, thus, no special perioperative precautions were taken.

Even though clinically silent atlanto-axial instability is the rule in these children, it should be a major concern to those medical professionals involved in their care. For instance, the Committee on Sports Medicine endorsed routine screening of all children with trisomy-21 for atlanto-axial instability at 5–6 yr of age, and recommended that all of these children should have screening

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roentgenograms of the neck prior to participating in high risk sports.[7]

Screening for atlanto-axial instability should include lateral roentgenograms of the neck in flexed, extended, and the neutral positions. If the distance between the anterior arch of the atlas and the adjacent odontoid process exceeds 5.0 mm, the diagnosis of atlanto-axial instability can be made with relative certainty.

In asymptomatic patients, the diagnosis of atlanto-axial instability requires certain precautions. Not only should their participation in contact sports be eliminated, but they should also be evaluated for surgical fusion, and, where appropriate, the use of cervical bracing. Not all children will require surgical procedures, but it is necessary to thoroughly evaluate their treatment needs.

There are only two other case reports that describe atlanto-axial subluxation associated with trisomy-21 occurring during the perioperative period.[13,14] Kobel *et al.*[13] recommend that "the anaesthetists should be particularly careful to avoid forceful flexion of the neck."

We feel that this is too lenient, in veiw of the potential for disaster should subluxation occur. In a prospective study of 236 trisomy-21 children, 35 patients (15%) underwent surgery for atlanto-axial subluxation.[3] Only eight patients (23%) made a complete recovery. The remainder made either no symptomatic improvement or died (31%), or demonstrated only mild improvement (40%). The primary factors which determined outcome were severity of injury and time from symptom onset to surgery.

We hope to accomplish several goals with our recommendation that all children over the age of 4 yr who present for surgery be screened for atlanto-axial instability. First, we aid parents by defining their childrens' risk for atlanto-axial instability. Second, we perform the service which will be required before the child is allowed to enter into any form of vigorous physical activity.[8] Last, we aid anesthesiologists by alerting them to the possibility of atlanto-axial instability and subsequent subluxation. Posterior cervical spine fusion is required prior to any other surgical intervention in those children who are symptomatic from their subluxation.[3] Recommendations for asymptomatic patients are not so easily defined, although prudence dictates neurosurgical consultation regarding the degree of cervical stabilization required prior to and following anesthesia.

In summary, atlanto-axial instability in trisomy-21 is an important but little appreciated disorder. Detailed but appropriate preoperative evaluations should include lateral roentgenograms of the neck in the flexed, neutral, and extended positions. Should any defect be found, appropriate neurosurgical consultation will aid in determining the need for further therapy.

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