Cervical Spine Considerations When Anesthetizing Patients with Down Syndrome

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DOWN syndrome (DS) is the most common chromosomal disorder, occurring in 1 of every 600–800 births. It is characterized by mental retardation, as well as craniofacial, upper airway, cardiovascular, and gastrointestinal anomalies. One manifestation of DS relevant to anesthesiologists is upper cervical spine instability produced by ligamentous laxity, skeletal anomalies, or both. This instability can result in neurologic impairment, including quadriplegia. However, there are no evidence-based practical guidelines to aid anesthesiologists in caring for these patients. The risk of spinal cord injury during anesthesia is unknown, as are the preoperative factors that might aid in accurately defining the risk in specific patients.

Review of Normal Upper Cervical Spine Anatomy

The structures relevant to this discussion include the base of the skull, the atlas (C1), and the axis (C2). The articulations between these structures are complex but are designed to facilitate maximal movement without compressing the spinal cord. The motions are flexion extension and rotation; little lateral bending occurs in this area.

The first cervical vertebrae is a ring, with articular surfaces on both the top and bottom aspects of its lateral masses. The occipital condyles rest on the superior surfaces. The inferior surfaces of C1 rest directly on C2, with no intervertebral disc. C1 is held to the base of the skull by capsular ligaments around the atlantooccipital joints, as well as the anterior and posterior atlantooccipital membranes. The primary motion between the occiput and C1 is extension (approximately 15°–20° relative to neutral). Most of the extension that occurs during direct laryngoscopy occurs at this interspace; there is almost no rotation at this level.

The second cervical vertebra is characterized by the odontoid process (or dens), a thumb-like upward protrusion from the body of the vertebra. The dens passes cephalad immediately behind the anterior arch C1 and is held in place by three ligaments (fig. 1). The transverse ligament (or the transverse component of the cruciate ligament) passes from side to side behind the odontoid and is anchored to the inside of the C1 ring. The atlantodental ligament bridges the small space between the anterior aspect of the odontoid and the posterior aspect of the anterior arch of C1 (this space is referred to as the anterior atlantodental interval [AADI]). The alar ligaments extend from the dens upward and laterally to the occipital condyles, and the apical ligament extends from the tip of dens upward to the anterior margin of the foramen magnum. The odontoid process is the primary axis for head rotation, and most motion occurs between C1 and C2. Under normal circumstances, there is almost no anterior–posterior subluxation between C1 and C2; the transverse ligament is primarily responsible for preventing such motion. There is roughly 20° of combined flexion/extension at C1–C2 (equally distributed between the two directions). The relatively large and funnel-shaped spinal canal from the foramen magnum to C2 minimizes the risk of cord compression during the extreme and complex motions of the upper cervical spine. In normal individuals, roughly one third of the space inside the C1 ring is occupied by the odontoid process, one third is occupied by the spinal cord, and the remaining one third is occupied by cerebral spinal fluid.

Causes of Craniocervical Instability in Down Syndrome

Craniocervical instability in DS involves the occiput–C1 or, more commonly, the C1–C2 level. Instability in these areas is usually termed atlantooccipital instability and atlantoaxial instability (AAI). This instability may be the consequence of a generalized ligamentous laxity. As noted above, the transverse ligament holds the odontoid process of C2 against the posterior aspect of the anterior arch of C1. If this ligament becomes lax, C1 (carrying the occiput) can sublux anteriorly on C2, compressing the spinal cord (fig. 2). If the odontoid and C1 are intact, posterior subluxation is prevented by impingement of the anterior arch of C1 on the dens. Laxity may also affect any of the atlantooccipital ligaments, resulting in other types of instability, including rotational. It is hypothesized that laxity of the cervical liga-
ments can be acquired or worsened by inflammation that is precipitated by upper respiratory infections, whereby inflammatory mediators travel retrograde via the common lymphatics.2

Bony anomalies can also contribute to instability at the atlantooccipital or atlantoaxial joints. Flattening or hypoplasia of the occipital condyles can cause a “rocker bottom” and lead to atlantooccipital instability. Anomalies of the atlas include hypoplasia of the posterior arch of C1, which can reduce the size of the cervical canal. Anomalies of C2 include an odontoid process that is absent, hypoplastic, or incompletely fused to the body of C2 (os odontoideum), making the transverse ligament ineffective, and resulting in instability.

Diagnosis of Craniocervical Instability

The definitive diagnosis of cervical spine instability can be difficult. The most common screening test is intended to detect AAI, the most common cervical spine disorder in DS, and involves lateral flexion and extension films and odontoid views. AAI is defined by a wider-than-normal AADI, which is evidence that C2 is not firmly affixed to C1, thus allowing C1 and the skull base to dislocate anteriorly on C2. Although there is some debate regarding the upper limit for a normal AADI, work based on x-ray images in normal children3 suggests that an AADI greater than 4–5 mm in any lateral view is abnormal. The argument against plain films as a screening tool is their poor reliability in predicting cervical cord compression symptoms, as well as their poor reproducibility.4 White et al.5 compared plain films to magnetic resonance imaging and found that the neural canal width on plain films is a better predictor of potential cord compression. They recommend the routine measurement of neural canal width as a more appropriate screening tool when interpreting c-spine films of patients with DS.

Most studies of AAI report greater subluxation when the neck is flexed, with normalization during extension. However, extension, particularly when associated with a concomitant “lifting” of the skull and C1 as occurs during laryngoscopy, may result in movement particularly in patients with atlantooccipital instability.6 Rotation of the head may also result in C1–C2 subluxation, sometimes locking the neck in a rotated position. Clinically, rotary subluxation is often associated with significant pain and immobility of the neck. It may cause kinking of the ipsilateral vertebral artery and stretching of the contralateral vessels, producing symptoms of dizziness. It may also worsen any canal compromise and place increased stress on ligaments.7

There has been considerable debate among the medical community and interest groups pertaining to whether cervical radiography should be used as a screening test of cervical instability in children with DS. The Special Olympics convened a panel of health professionals to review the subject and issued a position statement in 1983 requiring all DS participants to be screened with lateral cervical radiography before participation. The American Academy of Pediatrics (Elk Grove Village, Illinois) supported this in 1984 but later retracted their support in 1995.8 The American Academy of Pediatrics Committee on Sports Medicine concluded that there was no published evidence that screening radiographs prevented the extremely rare catastrophic c-spine injuries during sports. The Special Olympics Medical Committee, an international group of healthcare professionals, continues to require radiographs and argues that more cases of cervical spine instability would be detected if the medical community were better informed of the issue. Of interest is that the American Academy of Pediatrics

Fig. 1. The anatomical relations and major ligamentous structures of the first cervical vertebra (C1) and the second cervical vertebra (C2) area.

Fig. 2. The top two images are lateral x-ray images taken in extension (left) and flexion of a patient with marked atlantoaxial instability. The bottom two drawings are intended to clarify the changes occurring in the x-ray images. Note the increased anterior atlantoental interval (AADI) with flexion and the corresponding decrease in the space between the posterior aspect of odontoid and the posterior arch of C1 (through which the spinal cord passes).
continues to recommend radiologic screening with lateral x-rays for all patients with DS once between the ages of 3 and 5 yr, especially if they are involved in contact sports. However, there are no published policies regarding the cervical spine evaluation before a general anesthetic. Furthermore, there may be a significant lack of awareness of potential c-spine issues in patients with DS, judging from a 1995 survey of pediatric anesthesiologists by Litman et al. Only half of the survey respondents would maintain the patient’s head and neck in a neutral position during general anesthesia if a patient presented with symptoms of AAI.

Other diagnostic methods such as computerized tomography (CT) or magnetic resonance imaging have better sensitivity but are not practical for preoperative screening because of the expense, scheduling issues, and potential need for general anesthesia to accomplish the studies. These more detailed diagnostic studies are usually reserved either for symptomatic patients or for follow-up of patients with abnormal plain films.

Incidence

Since the first report describing atlantoaxial subluxation was published in the early 1960s, a number of studies have looked for radiographic evidence of AAI in patients with DS and found an incidence approximating 15%. The three referenced studies used lateral films of the cervical spine in neutral, flexion, and extension. A wider range of 7–36% incidence can be found across the literature, probably because of differences in the definition of AAI, the age group studied, the filming technique, or the patient positioning. The incidence of atlantoaxial instability in DS also varies widely in the literature; however, Parfenchuck et al. found an incidence of 8.5% by calculating the Powers ratio from plain films (ratio between the measured distance from the basion to the anterior surface of the posterior arch of C1 and the measured distance from the opisthion to the posterior surface of the anterior arch of C1). The incidence of cervical skeletal anomalies approximates 50% of patients with DS in studies that have used both plain films and CT images; there is a significantly higher incidence (79%) in those with an abnormal AADI. Importantly, 26% of toddlers with DS were found to have a hypoplastic posterior arch of C1.

The anatomical abnormalities described above are sometimes associated with neurologic disorders. Deterioration in function most often occurs gradually but may occur suddenly. There are two reports in the literature of children with DS whose symptoms were preceded by upper respiratory infections. Another report describes a 4-yr-old child with DS who was found dyspneic and, after arrival to the hospital, became apneic and quadriplegic, without any known precipitating event. However, the patient was known to have abnormal cervical spine films 1 yr previously. In 1998, Peuschel stated that he had found 126 cases in the medical literature of symptomatic AAI in patients with DS.

Natural History

Several authors have tried to determine the natural history of AAI in persons with DS. The duration of follow-up in four separate studies varied from 5 to 13 yr. The percentage of patients with DS whose lateral cervical films differed from their initial evaluation (enough to change the assessment from stable to unstable or unstable to stable) ranged from 3 to 24%. In a study by Burke et al., most of the changes were from a normal AADI to an abnormal AADI, whereas the other studies found the opposite trend—a tendency for the AADI to normalize. However, Morton et al. reported one patient with a normal AADI of 3 mm during the first screen who subsequently underwent otolaryngologic surgery 2 yr later and developed acute torticollis with a 7-mm rotary dislocation, necessitating surgical posterior fusion. Larger longitudinal studies are clearly needed.

Cervical Spine Injuries Associated with Surgery and Anesthesia

Although there are no published data on the fraction of patients with DS who require anesthesia and surgery, clinical experience suggests that the value is high. The University of Iowa Hospitals (Iowa City, Iowa), which is a major referral center, has performed approximately 170 surgical procedures in patients with DS during the past 3.5 yr, making up approximately 0.25% of the total surgical procedures. Unfortunately, there has been no reported large series of anesthetics performed in patients with DS, and hence the incidence of perioperative neurologic deterioration is unknown. Therefore, information can only be gained indirectly. We first searched the American Society of Anesthesiologists Closed Claims Database, both for all cases suggestive of spinal cord and/or other neurologic injury and for all cases involving patients with DS or trisomy 21. Of the 6,449 claims in the database, only one contained any suggestion of an injury related to the cervical spine in a patient with DS. This was a teenage girl who underwent a right tympanoplasty during general anesthesia. After being discharged 24 h later, she reported neck and arm pain. A suit was filed for a “herniated cervical disk.” The anesthesiologist was dropped from the suit, but without more details and radiologic data, it is impossible to draw any conclusions.

We next searched MEDLINE in an attempt to find all published cases of possible perioperative changes. By searching the English-language reports from 1966 until November 2003 pertaining to DS and including the Medical Subject Headings terms atlantoaxial instability or atlantoaxial instability or craniocervical instability or intra/postoperative complications or general anesthesia, eight cases were found. Only two describe events in which there is a possible relation to laryngoscopy and
intubation. The first was a 3-yr-old child with DS who underwent general anesthesia for strabismus surgery, but the operation was canceled when apneoa and severe cardiopulmonary instability developed. There is no other information about the anesthetic. After emergence, the patient refused to walk, and right hemiparesis followed 1 month later. No diagnostic studies were performed for 3 more years, at which time the AADI was 7 mm. The second case was an 18-yr-old patient with DS who underwent intubation for postoperative pneumonia more than a day after an Achilles tendon release. After the pneumonia cleared, she could not be weaned from the ventilator. A subsequent film of her cervical spine revealed craniovertebral settling with atlantoaxial subluxation and a spinal canal diameter of only 2.5 mm.

There have been other cases where cord compression may have occurred intraoperatively. A 4-yr-old child with DS who underwent an unremarkable ventricular septal defect closure was noted postoperatively to have diffuse hypotonia of his lower extremities and was unable to walk. A CT image, which was obtained after the patient returned for a follow-up visit, revealed posterior subluxation of C2 with flattening of the spinal cord. From the report, it is impossible to know at what point the injury occurred. In another case, a 3-yr-old child with DS became severely quadriparetic with pharyngeal dysfunction after myringotomy tube placement during general anesthesia. Information on airway management is not available, but lateral rotation of the head and neck likely occurred during the myringotomy. Interestingly, a lateral cervical spine radiograph obtained 1 yr before surgery revealed a 10-mm dislocation of the atlantoaxial joint. A postoperative CT scan identified a dystopic os odontoideum with occipitocervical instability as well as atlantoaxial subluxation. Both of these latter patients underwent boney fusion with partial or complete recovery.

There are three case reports of rotary subluxation occurring in the perioperative period in patients with DS. Two of the cases occurred in conjunction with otolaryngologic surgery (tympanomastoidectomy and an unspecified surgery). The second patient had neck films 2 yr previously, which showed an AADI of only 3 mm (normal); after surgery, the AADI measured 7 mm. The third case occurred in relation to surgical repair of an atrial septal defect. Although it is likely that the first two cases were related to surgical positioning, it is unclear when the injury occurred with the atrial septal defect repair. Head tilt was noted on postoperative day 5 and was still present 1 month later, when the diagnosis was made. All three patients, who ranged in age from 6 to 14 yr, underwent upper cervical fusion. Although rotary subluxation is unlikely to be attributed to laryngoscopy and intubation, it may be due to positioning of the patient during general anesthesia or during postoperative sedation in the intensive care unit.

The final case represents the first case report of a neonate with DS to experience atlantooccipital subluxation, but the timing of the injury is unclear. The infant had undergone ligation of a large patent ductus arteriosus on the first day of life. Postoperatively, she was kept sedated and paralyzed. When the medications were stopped, she was found to have neither spontaneous movements in her extremities nor respiratory effort. A CT image on day 16 revealed atlantooccipital subluxation. The patient underwent occiput-C2 fusion with gradual and complete resolution of strength and sensation by the third postoperative week.

**Comment**

The preceding material describes the anatomical changes that can be present in patients with DS and demonstrates that neurologic complications can occur during or after surgery. There is little doubt that general anesthesia has the potential for allowing increased joint mobility and obliterating any protective reflexes compared with the awake state. However, surgery and anesthesia are common, and complications seem to be extremely rare. Because of such rarity, it is impossible to develop a standard of care for preoperative assessment and intraoperative care that can be followed for each patient with DS. The approach described below is based on our best judgment. It is admittedly conservative, but without more evidence, it seems prudent to err on the conservative side. However, we will try to point out those areas of greatest uncertainty.

**Preoperative Assessment**

In several of the reported cases, anesthesia and surgery were associated with worsening of previously unrecognized abnormalities, abnormalities that probably reflected some degree of preexisting cord compression. Therefore, a thorough history and physical examination should be conducted to look for signs or symptoms suggestive of cord compression.

Specific questions to be addressed include the following:

1. Has the patient’s behavior changed (refusal to participate in usual activities)?
2. Has his or her ability to ambulate worsened?
3. Has fine motor function decreased?
4. Has there been any change in bladder or bowel function?
5. Does the patient report any pain in the head or neck area?
6. Does the patient refuse or seem unable to turn his or her head?
7. Have there been any episodes of dizziness or syncope?

During the physical examination, these specifics should be sought:
1. Abnormal range of motion of the head and neck, and examine for neck tenderness
2. Abnormal gait
3. Weakness, spasticity, increased deep tendon reflexes, a positive Babinski reflex, or clonus of the lower extremities
4. If the planned surgical procedure requires the head and neck to be in any position other than neutral, one may want to repeat the radiography. This is most commonly an issue during otolaryngologic procedures such as tonsillectomies, laryngoscopy/bronchoscopy, and mastoidectomies.

Positioning for otolaryngologic procedures such as tonsillectomies, direct laryngoscopies, or ear procedures deserves special care. For myringotomies, our procedure is to securely strap the patient to the operating room table, place supports alongside the head, and roll the table side to side, rather than turning the head. For airway procedures that usually involve head extension, it is important to communicate with the surgeon preoperatively about minimizing head and neck movement. It may be possible for the surgeon to perform the laryngoscopy or tonsillectomy without using suspension.

It is obvious from some of the case reports that we have done a poor job of recognizing and diagnosing cervical cord compression postoperatively. At least five of the eight cases associated with surgery had a significant delay before diagnostic studies were considered. Anesthesiologists and surgeons should look for neurologic changes in patients with DS postoperatively and keep AAI and atlantooccipital instability in their differential diagnosis. When patients require continued ventilation after surgery, consideration should be given to allow them to awaken enough after the procedure, if possible, to test for signs of myelopathy before sedation is continued. If cord compression is suspected, treatment is more likely to be successful if it is detected and treated early.

Conclusion

Given the relatively high incidence of cervical spine abnormalities, the number of complications in the literature is surprisingly low. Perhaps we are inappropriately creating concern about an event (spinal cord injury during laryngoscopy) that is exceedingly rare. Recommendations regarding the use of caution during airway management and surgical positioning may be vague and serve only to create anxiety. Conversely, it is possible that anesthetic and surgical complications are not being recognized, or they are not being reported because of medicolegal concerns.

There is a similar lack of information regarding the value of screening radiographs for asymptomatic patients. Many groups and individuals have made recommendations regarding the care of these patients. However, no general agreement exists. Although the Special Olympics requires cervical spine radiographs before allowing children to participate, the American Academy of Pediatrics does not support this recommendation but still recommends routine radiography between the ages...
of 3 and 5 yr. Some clinicians insist that every patient with DS needs radiography before any anesthetic, others believe that radiography is needed only before certain procedures, and still others take no position. No broad consensus exists, and clinicians should probably view any rigid recommendations with some skepticism. The only issue about which we feel strongly is the need for careful preoperative evaluation to identify symptomatic patients. If there is any suspicion of signs or symptoms consistent with cervical cord compression (see directed questions and physical examination findings under Preoperative Evaluation), radiologic evaluation and surgical consultation should be considered before proceeding with elective surgery.

For asymptomatic patients, we must find a cost-effective screening tool to better determine their risk of injury. Some of these issues could be answered with a large prospective multicenter epidemiology study. We would gain an appreciation for the number of patients with DS who undergo surgery, we could correlate preoperative history and physical findings with radiologic screening, and we could determine the incidence of postoperative neurologic changes due to cervical cord compression. With such information, evidence-based guidelines might be possible.

References