Atlantoaxial Instability: Why the Sudden Concern?

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The conditions of atlantoaxial instability and subsequent dislocation have raised much concern in recent years. Lack of knowledge in the medical and educational professions hampered efforts by Special Olympics, Inc. in 1983 to encourage screening of all Down syndrome participants prior to athletic competition. As a result, numerous articles on these conditions have been published in the last few years attempting to promote awareness. This paper looks at atlantoaxial instability and dislocation in depth, with the hope of providing knowledge to those who do not understand the concern surrounding these conditions.

Prior to 1983 few persons had heard of atlantoaxial instability. Early in that year, Special Olympics, Inc., issued a bulletin, effective immediately, restricting the participation of all athletes with Down syndrome until they had been examined for atlantoaxial instability (Committee on Sports Medicine, 1984; Shriver, 1983). Special Olympics directors were given 6 weeks to initiate compliance with the policy if they wished to remain an accredited member of Special Olympics.

A panel of medical personnel, lawyers, and sports medicine experts issued this policy with guidelines for coaches, directors, and physicians. Coaches were instructed to restrict participation of Down syndrome athletes in activities which, by their nature, could cause undue stress on the upper cervical spine region. Activities considered to be potentially dangerous were heading in soccer, high jumping, diving, gymnastics, the butterfly stroke in swimming, pentathlon, tumbling, and warm-up exercises which apply pressure to the head and neck area (Shriver, 1983). In addition, contact sports, trampoline activities, and somersaults were to be avoided.

Illustrative of articles printed in several journals to acquaint professionals with atlantoaxial instability was one prepared by Cooke (1984). More information is needed on atlantoaxial instability to motivate school personnel, as well as Special Olympics coaches, to individualize assessment and programming for students who may have this condition. Educators need also to understand the difference between atlantoaxial instability and dislocation. Atlantoaxial instability is a condition of the first and second cervical vertebrae in which there is abnormal-
ly increased mobility within the joint. A dislocation involves an actual displacement of the bone from its normal position in the joint (Thomas, 1985) and is a potentially life-threatening condition (Howard, 1985; Hreidarsson, Magram, & Singer, 1982). Awareness of the significance of atlantoaxial instability can aid in the prevention of critical injuries at the upper cervical spine level.

Atlantoaxial instability is not exclusively associated with Down syndrome. In able-bodied persons, atlantoaxial dislocation has been reported as a result of trauma or cervical-area infections (El-Khoury, Clark, & Gravett, 1984; Fielding, Fietti, & Mardam-Bey, 1978; Jacobson & Adler, 1956; Sullivan, 1949). Detailed incidence figures for able-bodied individuals are not available. Atlantoaxial instability has also been reported in persons with Morquio’s syndrome, Scott’s syndrome, pseudoachondroplastic dysplasia congenita, cartilage-hair hypoplasia (Finerman, Sakai, & Weingarten, 1976; Pueschel, Scola, Perry, & Pezzullo, 1981), rheumatoid arthritis, ankylosing spondylitis (Giblin & Micheli, 1979; Wortzman & Dewar, 1968), dwarfism, and Klippel-Feil syndrome (Roach, Duncan, Wenger, Maravilla, & Maravilla, 1984).

The purpose of this paper is to present facts relative to atlantoaxial instability and dislocation: their prevalence, causes, symptoms, diagnostic procedures, treatment, and implications for adapted physical education personnel. The earliest reference to atlantoaxial instability in medical journals appeared in 1949 (Sullivan). This paper reviews the literature published since that date.

Prevalence

There is wide variance in figures used to report prevalence of atlantoaxial instability among the Down syndrome population. The earliest study of incidence was reported in 1966 (Martel & Tishler). Fourteen (20%) out of a sample of 70 Down syndrome persons had C1-C2 joint instability. Semine, Ertel, Goldberg, and Bull (1978) examined 85 children with Down syndrome for atlantoaxial instability. In their sample, 10 patients (12%) exhibited abnormal joint intervals. An additional 5 patients (6%) had abnormal odontoid processes. This resulted in 18% of the total number exhibiting joint abnormalities. Pueschel et al. (1981) detected atlantoaxial instability in 15 (9.5%) out of 158 patients with Down syndrome. In 1983, Pueschel examined 236 Down syndrome individuals for evidence of atlantoaxial instability. In this selected sample, 40 (17%) had a gap of 5 mm or more between the joint surfaces of the atlas and the odontoid process. The accepted normal joint interval is less than 4 mm (Roach et al., 1984; Semine et al., 1978).

The prevalence of actual C1-C2 dislocation is much less than that of instability. There have been 17 documented cases of atlantoaxial dislocation in Down syndrome individuals reported in the literature (Aung, 1973; Curtis, Blanks, & Fisher, 1968; Dawson & Smith, 1979; Dzenitis, 1966; Finerman et al., 1976; Giblin & Micheli, 1979; Groborscheck & Strohecker, 1985; Hreidarsson et al., 1982; Hungerford, Akkaraju, Rawe, & Young, 1981; Martel & Tishler, 1966; Semine et al., 1978; Sherk & Nicholson, 1969; Sherk, Pasquariello, & Watters, 1982; Shield, Dickens, & Jensen, 1981; Whaley & Gray, 1980). However, these cases represent only a small number of the actual dislocations that have occurred.
Antony

Anatomy of the Atlantoaxial Joint

Knowledge of the anatomy of the atlantoaxial joint is requisite to an understanding of its anomalies. The atlantoaxial joint has four synovial spaces: the two between the atlantal and axial articular masses, the bursa between the odontoid and the transverse ligament, and the bursa between the odontoid and the anterior arch of the atlas. Movements at the atlantoaxial joint include rotation, flexion, and extension (Wortzman & Dewar, 1968). Flexion and extension occur in a hinge-like manner. Lateral movement is prevented by circumferential fibers of the transverse ligament. The transverse ligament forms a strong band passing behind and along the sides of the odontoid process and attaches to tubercles in the midmedial aspect of each atlantal lateral mass, thus preventing forward subluxation during head flexion.

The alar ligaments attach to the posterolateral portion of the head of the odontoid and run up laterally and superiorly to the medial aspect of the occipital condyles. They are lax with the head in neutral position and tighten with rotation, preventing excess rotation at the atlantoaxial joint (Wortzman & Dewar, 1968). Thus, the alar ligaments are crucial in preventing rotational dislocation. Anterior-posterior x-rays, in addition to lateral views, are helpful in detecting a rotational dislocation. Figures 1 and 2 display key parts of the atlantoaxial joint.

Etiology

Although the etiology of atlantoaxial instability is not well understood, several causes have been hypothesized. Sullivan, Bruwer, and Harris (1958) suggested that upper respiratory tract infections induced hyperemic decalcification of the

Figure 1 — Anatomical drawing of the atlantoaxial joint in neutral position (anteroposterior view) (adapted from Jacobson & Adler, 1956).
upper cervical vertebrae, allowing loosening of the ligaments supporting the atlantoaxial joint. In 1965, Tishler and Martel postulated that generalized ligament laxity found in Down syndrome individuals was the cause of subsequent instability leading to dislocation.

Today most researchers agree on four probable etiologies: (a) ligament laxity (Giblin & Micheli, 1979; Howard, 1985; Hreidarsson et al., 1982; Pueschel et al., 1981; Semine et al., 1978), (b) pharyngeal infection/inflammation (Howard, 1985; Hreidarsson et al., 1982; Semine et al., 1978), (c) abnormalities of the odontoid process (Dawson & Smith, 1979; Hungerford et al., 1981; Roach et al., 1984), and (d) trauma to the head and neck region.

The stability of the atlantoaxial joint anteriorly depends mainly on the ligaments and an eccentrically placed odontoid process (Fielding, Hawkins, & Ratzan, 1976). Pharyngeal inflammations tend to weaken ligaments and, in turn, increase their laxity. Ligament laxity allows forward subluxation of the atlas on the axis. Abnormalities of the odontoid process are responsible for a loss of stability in the joint itself. Hypoplasia of the odontoid is a relatively common defect in individuals with Down syndrome.

**Symptoms**

Many symptoms of atlantoaxial instability have been listed in the literature. Parents, teachers, and other professionals need to be aware of the following symptoms: increased clumsiness, sudden preference for sitting, changes in gait pattern, change in neck posturing, neck pain, limited neck movement, weakness in ex-

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Figure 2 — Anatomical drawing of atlantoaxial joint showing the transverse ligaments.
tremities, hyperactive reflexes, spasticity, or changes in bowel or bladder function (Cooke, 1984; Howard, 1985). Recognition of symptoms can lead to early detection of this condition and, in turn, increase the chance of remediation.

Screening/Diagnosis

Screening for atlantoaxial instability requires the use of radiographic technology. Lateral roentgenograms (x-rays) taken of the cervical spine in positions of flexion, extension, and neutral are examined, and the distance between the posterior rim of the anterior arch of the atlas and the anterior surface of the odontoid process is measured in millimeters. Measurements exceeding 4.5 mm are considered abnormal (Roach et al., 1984; Sernine et al., 1978). See Figure 3 in this regard.

Computerized tomography (CT) scans are used when the distorted anatomy of certain anomalies of the cervical spine make it difficult to interpret plain radiographs (Roach et al., 1984). The scan is also helpful in detecting abnormal motion not occurring between the body of the axis and the posterior ring of the atlas.

Figure 3 — With an intact odontoid process, measurements are taken from the anterior arch of the atlas and the odontoid process (adapted from Fielding et al., 1976, p. 402).
In a CT scan, the patient is examined while supine, with cervical spine in flexion. Transverse cuts of the odontoid process and the anterior ring of the atlas are obtained. The procedure is then repeated with the cervical spine in extension. The transverse cuts allow comparison of the subarachnoid space of the spinal cord in flexion and extension (Roach et al., 1984). Cord compression can be detected using the CT scan. Giblin and Micheli (1979) noted that preoperative contrast studies could be completed to determine the site and extent of cord compression. This procedure is invasive, however, and subjects the patient to an increased risk. The presence or absence of neurological disturbances may be a more reliable and safe indicator of cord compression.

Schools, recreation programs, and camps may wish to require screening of Down syndrome individuals prior to participation in vigorous physical activities (Committee on Sports Medicine, 1984). Down syndrome individuals should be examined for evidence of atlantoaxial instability at age 2 years and older. The Committee on Sports Medicine (1984) suggested that children be screened by age 5 or 6 years, before beginning school. The need for repeat roentgenograms is questionable at the present time for those with no evidence of instability.

**Treatment**

Individuals with atlantoaxial joint intervals indicating instability have several treatment options. Persons with asymptomatic instability need only be restricted from activities that could result in hyperflexion or hyperextension of the cervical spine and monitored very closely for symptoms of neurologic disturbance. At this time, it is usually not recommended that surgical stabilization be carried out when no neurologic symptoms are present.

Persons with symptomatic atlantoaxial dislocation, in most cases, undergo spinal fusion to stabilize the joint. Reduction of a dislocation is usually attempted prior to surgery using skeletal traction followed by placement of a neck brace (i.e., Halo cast, Minerva jacket). Recent developments indicate that attempts to reduce a chronic dislocation at the time of surgery rather than prior to surgery may result in a poor outcome. In some instances, skeletal traction followed by a neck brace is the only treatment needed.

Extreme mobility of the joint resulting from damaged ligaments or a deficient odontoid process requires spinal fusion to stabilize the joint. Spinal fusion of the upper cervical spine involves grafting a bone segment to the first and second vertebrae. The procedure (Gallie fusion) involves passing a wire loop from below up under the arch either directly or with the aid of a silk suture or an aneurysm needle. A cortical graft taken from the iliac crest is notched to conform to the spine of the second cervical vertebra. . . . A loop of wire is passed over the graft and around or through the spinous process of the second cervical segment. Grafts of the cancellous strips are then added. (Fielding et al., 1976. p. 402)

A Minerva jacket is then applied until fusion is complete. The use of wire in the stabilization procedure has recently presented some concern.
Rotation of the atlantoaxial joint following fusion diminishes an average of 13% in patients under the age of 20 years. In those 20 to 40 years old, the loss is approximately 25%, and in persons over 40 years of age a 29% decrease in rotation is expected (Fielding et al., 1976).

The following is a list of indications for spinal fusion: (a) evidence of central nervous system involvement, (b) persistent cervical symptoms, and (c) a measured joint interval greater than 5 mm (Fielding et al., 1976). The third indication is debatable when the instability is asymptomatic.

**Implications for Adapted Physical Education Personnel**

Within the realm of legal liability, professionals need to be conscious of potential injury-inducing activities and/or situations. Awareness of conditions reported in the literature can decrease the possibility of serious injury by helping persons to know when to restrict an activity for the safety of the participant. Therefore, atlantoaxial instability is an issue that needs to be brought to the attention of schools, parents, and sport and recreation personnel who are involved with persons with Down syndrome, dwarfism, rheumatoid arthritis, and other syndromes in which atlantoaxial instability occurs.

Instructors should be aware of the student’s medical status including the condition of the atlantoaxial joint. Results of examination should be kept in the student’s file. When evidence of instability exists, appropriate restrictions should be written into the individualized educational plan (IEP) and strictly adhered to in classes, at recess, and in athletic competition.

Failure to consult the student’s medical records can lead to serious problems for both the child and the teacher. Allowing students to participate in tumbling and other contraindicated activities when they have a diagnosis of atlantoaxial instability can lead to devastating results. Some injuries are unavoidable, but injury resulting from failure to consult medical files is due to negligence.

From the literature reviewed, the following guidelines from Special Olympics, Inc., the Committee on Sports Medicine, and the National Down Syndrome Congress appear warranted.

1. Determine which individuals have conditions in which atlantoaxial instability is known to occur.
2. Check medical files for a record of atlantoaxial joint screening.
3. If record is present and positive—
   a. restrict from activities that may cause undue stress on the head and neck area;
   b. have the parents sign a consent form allowing the child to participate with the restrictions;
   c. watch for development of symptoms indicating a possible dislocation;
   d. adhere to physician’s recommendations.
4. If record is present and negative, allow participation in all activities with appropriate supervision.
5. If record of screening is not present—
a. contact parents and explain importance of the screening;
b. restrict the child from all contraindicated activities until a record of screening has been received and placed in the file.

If instability has been demonstrated by x-ray, frequent mini-neurological examinations should be carried out to determine the development of the earliest possible signs of cord compression.

**Summary**

The literature reviewed in this paper, as well as the resultant guidelines, supports the APAQ viewpoint by Iyriboz (1985), who addressed the adverse effects of nonindividualized physical activity. Medical screening should be an important part of the IEP process. Only when information on atlantoaxial instability is available can school personnel individualize physical education instruction so as to provide optimal motor, fitness, and leisure experiences for all handicapped children and youth.

**References**


