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Atlantoaxial Dislocation Related to Instilling Eyedrops in a Patient With Down's Syndrome

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PURPOSE: To alert ophthalmologists of the risk of hyperextending the neck of patients with Down's syndrome while holding them to instill eyedrops.

METHOD: We describe a case of atlantoaxial dislocation related to instilling eyedrops in a 6-year-old girl with Down's syndrome.

RESULT: Early recognition of the problem allowed prompt treatment and partial recovery of the patient.

CONCLUSION: Examination and clinical maneuvers in a child with Down's syndrome require gentleness and avoidance of hyperextension of the neck to prevent spinal cord injury.

ALTHOUGH ASYMPTOMATIC ATLANTOAXIAL INSTABILITY is a relatively frequent finding in individuals with Down's syndrome (10% to 20%),¹ symptomatic dislocation associated with medical maneuvers has been reported infrequently.² Atlantoaxial instability may be a manifestation of hypotonia and joint hyperextensibility caused by laxity of the transverse ligaments that hold the odontoid process of the axis close to the anterior arch of the atlas, allowing increased mobility between the atlas (first cervical vertebra) and the axis (second cervical vertebra) during flexion and extension of the neck. In ophthalmologic settings, individuals with Down's syndrome undergo a variety of procedures involving potentially stressful head and neck movements, including head rotation in the doll's head maneuver and neck extension for nasolacrimal duct probing. The following case report documents a dramatic occurrence of atlantoaxial dislocation caused by neck hyperextension.

A 6-year-old girl was diagnosed at birth as having Down's syndrome. Her developmental milestones were all delayed. At the time of our examination, the patient walked into our office alone with excellent stability, showing a normal sitting balance. After visual acuity testing and slit-lamp examination, we instilled two drops of cyclopentolate 1 minute apart in each eye. The second instillation required immobilization of the girl, during which the nurse hyperextended the neck of the patient. A few minutes later, the mother informed us of strange and incoordinate movements of the child's legs and noted that the child was "wet" despite good bladder and bowel control since age 3. The neurologic examination revealed upper motor neuron signs in all four extremities, such as tremor, hyperreflexia, and bilateral extensor plantar responses. A lateral radiograph of the cervical spine in neutral position showed an increased interval between the atlas and the odontoid process of the axis (Figure). The patient was immediately placed in halo traction and later had surgery to achieve bony

Figure. Lateral radiograph showing the odontoid process of the axis (arrow) displaced posteriorly, producing a significant decrease in dimension of the spinal canal. In the patient, the interval between the articular surfaces of the odontoid process (o) and of the anterior arch of the atlas (a) is markedly increased. In normal subjects, the areas identified by the asterisks overlap.

fusion. She is now hyperreflexic with very mild spasticity.

Several protocols have been proposed to detect atlantoaxial instability in patients with Down's syndrome. Some authors advocate cervical spine radiographic screening beginning at age 3 years,³ whereas others claim that physical examination with careful attention to neurologic signs is more predictive of impending dislocation.⁴ We suggest the following guidelines for avoiding atlantoaxial dislocation in Down's syndrome patients during ophthalmologic examination: be gentle and unhurried when dealing with these patients; administer topical ophthalmic medications by using a perfume sprayer as close (2 to 4 cm) to the superior eyelashes as possible, with the patient in an upright position (reference 5 and H. Freedman, MD, and R. Blocker, MD, written personal communication, March 1996); sedate uncooperative children rather than restraining them; and suspect cervical myelopathy, secondary to iatrogenic dislocation, when an otherwise normal Down's syn-

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drome child shows neurologic signs after head or neck manipulation. Early diagnosis and prompt treatment of atlantoaxial dislocation may prevent or minimize severe complications.

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