ISSN: 2832-2584

Case Report

Journal of Clinical Pediatrics and Child Care Research

Decreased Arm Movement in Non-Verbal Male

Michael Arenson, William B Prince*

Pediatric and Emergency Medicine Departments of Seattle Children's Hospital, Seattle, Washington; and University of Washington, Seattle, Washington

***Corresponding author**
William B Prince, Seattle Children's Hospital, M/S: MB.7.520, ATTN: William Prince, PO Box 5371, Seattle, WA, USA

Submitted: 06 Jul 2022; **Accepted:** 11 Jul 2022; **Published:** 21 Jul 2022

Citation: *Michael Arenson, William B Prince. (2022). Decreased Arm Movement in Non-Verbal Male. J Cli Ped Chi Res, 3(1):88-90.*

Abstract

A 13-year-old non-verbal Trisomy 21 male presents to the emergency department with decreased arm movement four days after witnessed ground level fall. Patient intermittently using arm normally, ambulating, and neurologically at baseline. Arm imaging negative for fracture or dislocation. Review of past medical history reveals prior use of cervical collar for resolved subluxation. Cervical spine imaging reveals complete anterior dislocation of C1 on C2.

Introduction

Individuals with Down Syndrome (DS) are at increased risk of atlantoaxial instability (AAI), defined as excessive mobility of the articulation of the atlas (C1) and the axis (C2). AAI can be symptomatic or asymptomatic. Spinal cord compression due to subluxation or frank dislocation of the cervical spine is uncommon but can lead to acute spinal cord injury. A child with DS presenting with a focal neurologic deficit needs a cervical collar, an upright neutral lateral cervical spine radiograph, and an immediate referral to an orthopedic or neurosurgeon. In 2011, the American Association of Pediatrics (AAP) revised the recommendations regarding health surveillance of patients with DS [1]. The AAP guidelines state that routine radiographs are not indicated in the asymptomatic child at any age. Instead, the AAP recommends pediatricians conduct an annual clinical follow-up, medical history, and physical examination that focus on red flag neurologic symptoms and signs.

Case Report

A 13-year-old Caucasian non-verbal male with Trisomy 21 was brought to our emergency room with his father due to complaint of decreased left arm movement over last four days after witnessed ground level fall. Due to known gait instability his parents generally hold his arm when walking longer distances and report pulling patient's left arm up at time of fall to prevent patient from hitting the ground. Parents deny patient hitting head, loss of consciousness, or obvious seizure activity. Since that time the patient has been occasionally leaning to his left side and using his left arm less. He had two additional falls since then although his father this is not uncommon. Father denies other past medical history currently.

On exam patient was walking around exam room with a shuffling gait which is normal for him. Patient approached me smiling and lifting both arms up to give me a hug. The patient did not appear to be in any distress and his exam was unremarkable. Patient is non-verbal with significant developmental delay making a neurologic exam difficult, however cranial nerves were grossly intact. Dad suspected a left arm injury from initial fall although patient is not reliable in reporting pain. Patient had full passive and active range of motion of the left arm with no reproducible pain or obvious deformity. Tone in all extremities was grossly normal with seemingly symmetric strength. Sensation in extremities was difficult to elicit.

Left upper extremity imaging negative for fracture or dislocation. Upon further review of medical history, the patient's father stated that his son did have to wear a neck collar for a few weeks but was then told it wasn't necessary anymore. Dad was unsure as the reason. On further examination of the medical chart there were several notes from orthopedics and cervical spine x-rays noting resolved atlantoaxial subluxation.

Given their previous involvement with this patient, orthopedics was consulted regarding their preference for imaging in the setting of possible atlantoaxial instability. Cervical 2 view spine films were recommended, and radiology called to confirmed complete anterior dislocation of C1 on C2, with obliteration of the central canal space (Figure 1 & Figure 2). The patient was immediately placed supine in a cervical collar. Child life at bedside to assist father in keeping patient supine and calm.

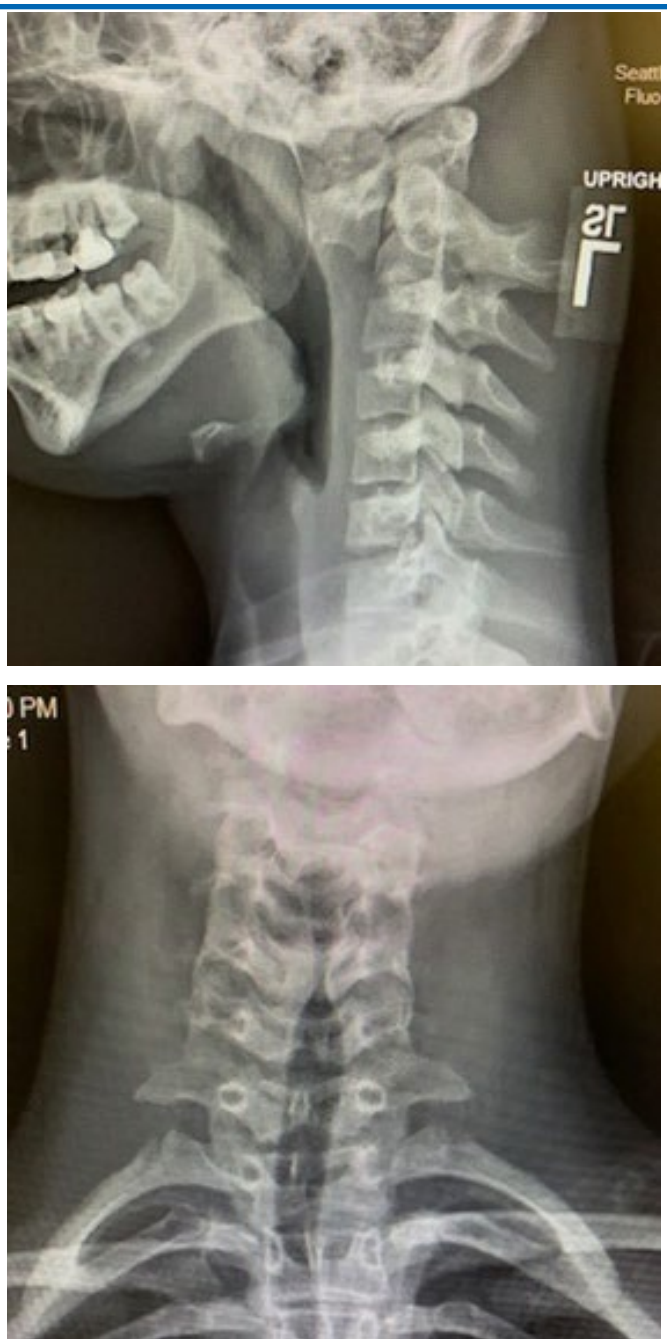

Figure 1&2: Complete anterior dislocation of C1 on C2

Neurology consulted who observed clonus in left foot. Orthopedics made aware of imaging results. Neurosurgery admitted the patient. Patient received CT cervical spine angiogram and MRI spine confirming diagnosis of C1 anterior and rotary subluxation on C2, spinal canal decreased to 5mm, with cord edema at that level. Patient was placed in cervical halter traction which resulted in gradual decrease in left foot clonus and improved frequency of left arm movement. He underwent 0-C3 fusion of posterior cervical spine, C1 posterior decompression, and halo placement/removal (Figure 3). POD #6 he was discharge home with clinical resolution of previous neurologic deficits.

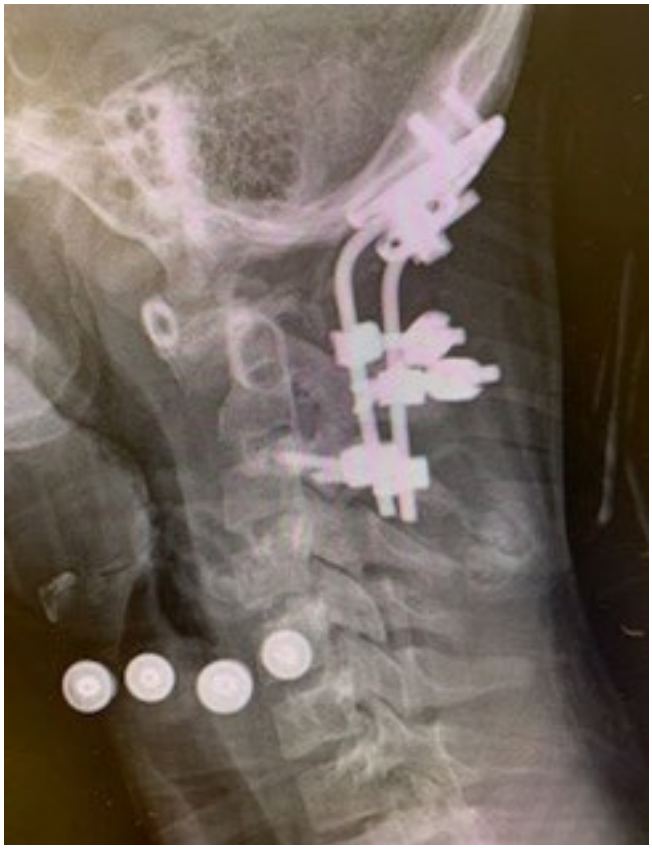

Figure 3: C1 laminectomy and posterior spinal fusion from the occiput through C3.

Discussion

Down syndrome is the most common genomic disorder of intellectual disability and is caused by trisomy of chromosome 21. The eponym of the syndrome is from Down, who described the clinical aspects of the syndrome in 1866 [2]. This case is an important reminder that children with DS have ligamentous laxity and osseus abnormalities which can occur in the upper cervical spine. This can lead to several complications; including atlantoaxial instability (AAI), which occurs in 10% to 30% of patients with DS, and atlanto-occipital instability (AOI) which has been reported in 8% to 63% of patients with DS. The majority of these cases are asymptomatic, with estimates of symptomatic disease ranging from 1% to 2% [3,4].

Although it is far less common than instability, atlantoaxial dislocation (as seen in our patient) is life-threatening and presents with signs and symptoms of cord compression. This can include abnormal gait, neck pain, limited neck mobility, head tilt, incoordination, clumsiness, and changes in bowel and bladder control. A neurologic examination may reveal sensory deficits, spasticity, hyperreflexia, clonus, and the presence of a Babinski sign [5]. Nearly all people with AAI who have suffered a catastrophic injury to the spinal cord have had preceding neurologic symptoms [6].

A child with DS presenting with a focal neurologic deficit needs at minimum an upright neutral lateral cervical spine radiograph [1].

The patient should be placed in a collar and referred immediately to an orthopedic or neurosurgeon. Flexion and extension radiographs may be performed prior to referral if no significant radiographic abnormalities are present. The AAP Committee on Sports Medicine and Fitness recommends that symptomatic children have magnetic resonance imaging (MRI) to clarify the extent of spinal cord compression and that appropriate surgical consultation be obtained to evaluate the need for definitive treatment [6]. The goal of surgery is to prevent further injury to the spinal cord by stabilizing the upper segment of the cervical spine.

As the biomechanics of the pediatric cranio-cervical junction are incompletely understood, the natural history of an individual's condition is difficult to predict. Evidence indicates that asymptomatic ligamentous instability in the absence of osseous abnormalities rarely progresses to clinical relevance [7]. Symptomatic instability is rare among patients who have DS. It is not even clear that radiographically demonstrated asymptomatic instability is a significant risk factor for either symptomatic instability or atlantoaxial dislocation [5].

No evidence has established that screening with lateral cervical radiographs is effective in preventing symptomatic atlantoaxial instability [1]. Given this lack of clarity, the American Academy of Pediatrics Committee on Genetics and the AAP Committee on Sports Medicine and Fitness recommend careful neurologic evaluation for signs and symptoms consistent with spinal cord injury as the most important clinical predictor of symptomatic atlantoaxial instability and dislocation [1,6]. The evaluating clinician should take a careful history and perform a thorough physical examination, looking for evidence of neurologic involvement. This clinical screening process should be done at least annually. Caution regarding contact sports and trampoline use should be discussed with families as they increase the risk of spinal cord injury in the child.

The AAP guidelines stated that routine radiographs are not indicated in the asymptomatic child at any age. Cervical radiographs are inaccurate before 3 years of age due to the lack of adequate vertebral mineralization, and they have low predictive value regarding the risk of developing AAI [1]. However, children who are found to have AAI on radiographs but lack neurologic symptoms should be followed closely with repeat neurologic examinations (at least annually) [6]. Radiologic screening should be performed in children with DS prior to procedures that require extremes of head position during induction of anesthesia or surgery [3]. Despite the AAP guidelines, the Special Olympics continues to mandate, in addition to a physical examination, preparticipation radiographic screening of all children who have Down syndrome.

Abbreviations

DS: Down Syndrome
AAI: Atlantoaxial Instability
AAP: American Academy of Pediatrics
POD: Post-operative day
CT: Computed tomography
AOI: Atlanto - occipital instabilit
MRI: Magnetic resonance imaging

Table of Contents Summary

Case report of adolescent male with Down Syndrome who presents with decreased arm movement after fall with complete anterior dislocation of C1 on C2 vertebrae.

Contributors' Statement

Dr. William Prince and Dr. Michael Arenson were both medical providers who treated the patient in this case report. They jointly discussed the case, wrote and revised the manuscript. All authors approve of the final manuscript as submitted.

Conclusion

This case is an important reminder that children with DS have ligamentous laxity and osseus abnormalities which can occur in the upper cervical spine. This can lead to AAI, AOI, dislocation and ultimately catastrophic injury to the spinal cord. A child with DS presenting with a focal neurologic deficit needs an upright neutral lateral cervical spine radiograph. The patient should be placed in a collar and referred immediately to an orthopedic or neurosurgeon. Per AAP guidelines, the routine use of cervical X-ray should be discouraged, especially in children less than 3 years of age, due to its low reliability. Parents should be educated about concerning focal neurologic signs and symptoms.

References

1. Bull, M. J., & Committee on Genetics. (2011). Health supervision for children with Down syndrome. Pediatrics, 128(2), 393-406.
2. Down, J. L. H. (1866). Observations on an ethnic classification of idiots. London hospital reports, 3(1866), 259-262.
3. Hankinson, T. C., & Anderson, R. C. (2010). Craniovertebral junction abnormalities in Down syndrome. Neurosurgery, 66(suppl_3), A32-A38.
4. Pueschel, S. M., & Scola, F. H. (1987). Atlantoaxial instability in individuals with Down syndrome: epidemiologic, radiographic, and clinical studies. Pediatrics, 80(4), 555-560.
5. Braganza, S. F. (2003). Atlantoaxial dislocation. Pediatrics in Review, 24(3), 106-107.
6. Committee on Sports Medicine and Fitness. (1995). Atlantoaxial instability in Down syndrome: subject review. Pediatrics, 96(1), 151-154.
7. Brockmeyer, D. (1999). Down syndrome and craniovertebral instability. Pediatric neurosurgery, 31(2), 71-77.