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Case report

# "Orthotopic" ossiculum terminale persistens and atlantoaxial instability in a child less than 12 years of age: a case report and review of the literature

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## Abstract

We report ossiculum terminale persistens associated with atlantoaxial instability in a child less than 12 years of age. Static and dynamic X-rays, thin-cut computed tomography with sagittal and coronal reconstructions, and magnetic resonance imaging of the cervical spine showed atlantoaxial instability and an "orthotopic" ossiculum terminale persistens. This pathologic state was differentiated from the primary ossification center at the tip of the odontoid, which normally is not expected to fuse with the body of the odontoid until the age of 12 years. The patient was taken to the operating room for a posterior instrumented fusion of C1 and C2. The patient has done well in short- and long-term follow-up.

There have been only a few case reports and small case series regarding atlantoaxial instability, requiring surgical intervention, from ossiculum terminale persistens. Most have presented later in life or in association with Down syndrome. Furthermore, most cases have been of the "dystopic" variant. The terms - "orthotopic" and "dystopic" anatomic variants - have usually been reserved to describe os odontoideum. However, we introduce these terms in describing ossiculum terminale persistens and show a rare case of "orthotopic" ossiculum terminale persistens associated with atlantoaxial instability in a pediatric patient less than the age of 12.

## Case presentation

### *History and presentation*

This 10-year-old Caucasian boy from the USA with no history of Down's syndrome and no past medical history presented to our institution after a fall while skateboarding on a ramp. On presentation he reported mild headache, but no neck pain, back pain, weakness, or paresthesias. The patient reported no history of prior trauma, but he did

recall a tumbling episode while on the "moonwalk" at age 6 at which time he felt a "snapping" in his neck. No neurological deficits were elicitable on examination; he had full strength of his bilateral upper and lower extremities, reflexes were 2+ throughout, no clonus, no Hoffman's sign, and there were no deficiencies in the sensory exam including joint position sense. He did have tenderness to palpation along the midline cervical spine.

### *Diagnostic evaluation*

A CT scan of the head was performed after an episode of emesis. There were no intracranial abnormalities; however the scout film revealed an increased atlantodental interval. This prompted flexion and extension lateral X-rays of the cervical spine which demonstrated an 8 mm atlantodental interval in flexion, and a 3.5 mm atlantodental interval in extension (Figure 1). An unfused apical ossicle was also discernable on the plain films. Further investigation with thin cut CT of the cervical spine, defined the apical ossicle to have well defined and mildly sclerotic margins, consistent with an ossiculum terminale persistens (Figure 2). As the apical ossicle remained in anatomic position during dynamic X-rays, the abnormality was consistent with an orthotopic ossiculum terminale persistens. Subsequent MRI of the cervical spine delineated the remnant synchondrosis between the apical ossicle and superior odontoid process, and there was no evidence of ligamentous disruption (Figure 3). However, fluid was present within the atlantodental space.

**Operation** Given atlantodental instability in the presence of an orthotopic ossiculum terminale persistens, surgical intervention was recommended to the family. Our patient was taken to the operating room and underwent an awake fiberoptic intubation. Intraoperative neurophysiological monitoring was performed with somatosensory evoked potentials, electromyography, and transcranial motor evoked potentials. The patient was then placed in three point rigid skull fixation and positioned prone on the operating table. The exposure was continued until adequate lateral exposure of C1 and C2 was achieved. The C2 nerve roots bilaterally showed only minimal laxity and were consequently divided along with the surrounding venous plexus to allow accurate screw placement. The medial borders of the C1 lateral masses were identified and C1 lateral mass screws were placed bilaterally under

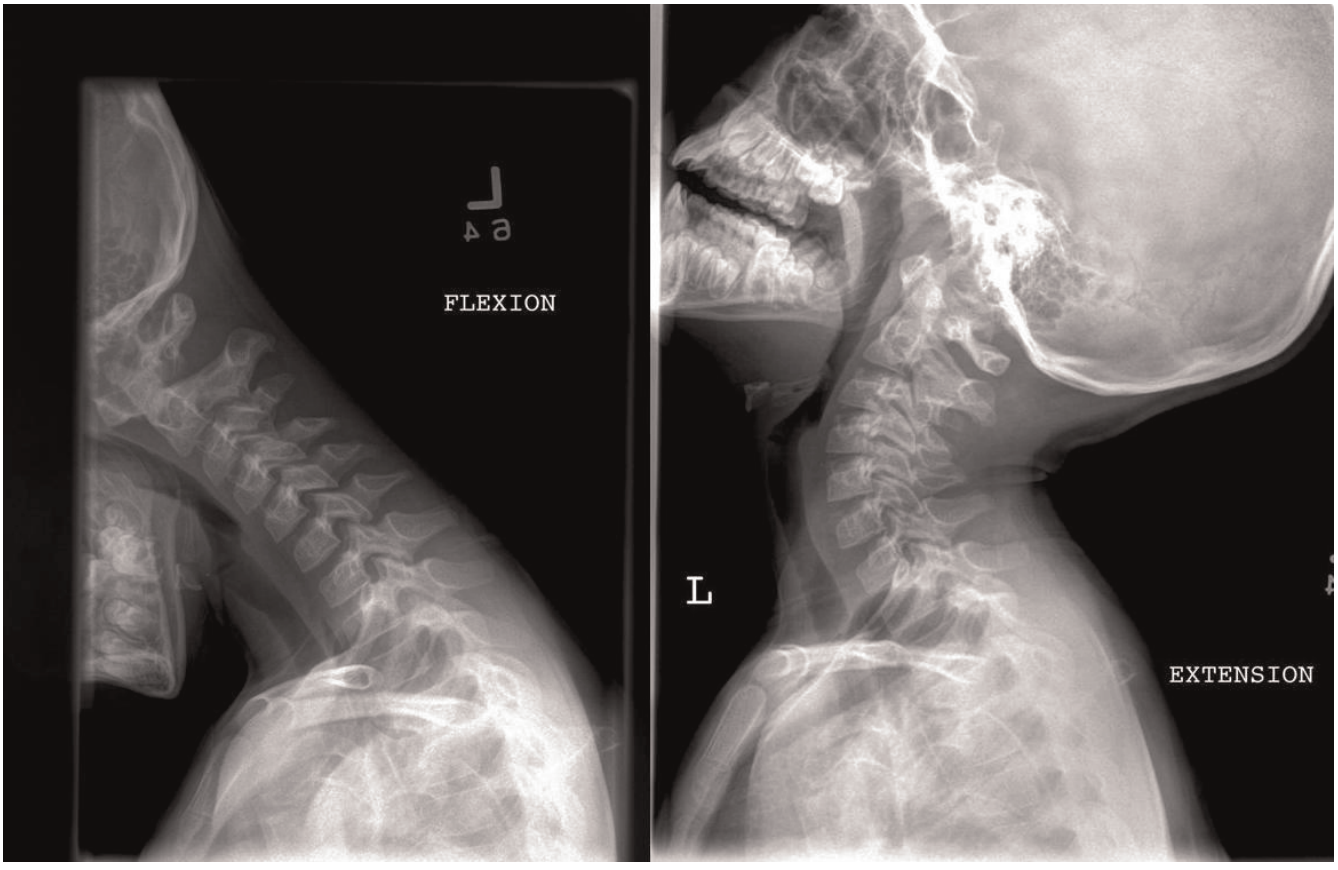


**Figure 1. (A)** Flexion and **(B)** extension cervical spine X-rays show an increased atlantodental interval of up to 8 mm.

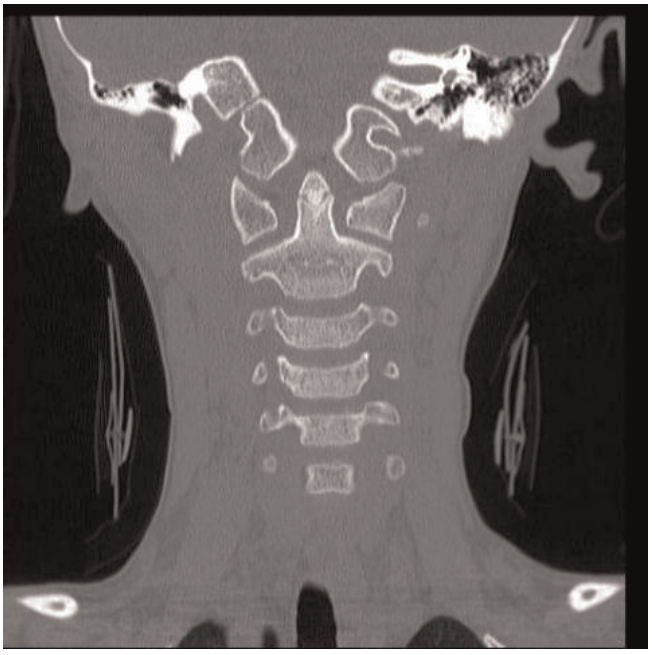

**Figure 2.** Coronal reconstructed CT of the odontoid complex demonstrates clear and intact cortical margins of the apical ossicle separate from the odontoid proper. There is no radiographic evidence of fusion.

fluoroscopic guidance. Following this, translaminar screws were placed at C2. The instrumentation and arthrodesis were completed with no intraoperative complications.

#### *Follow-up*

Postoperatively, the patient awoke with no neurological deficits. He was maintained in an Aspen collar, and postoperative lateral X-rays and thin cut CT scan showed the instrumentation to be in solid position with good alignment of the cervical spine (Figure 4). He was discharged home on postoperative day 3, and at greater than 1 year follow-up he has continued to do well.

## Discussion

#### *Embryology*

The axis ossifies from five primary ossification centers - two forming the body of the odontoid process, one for the body of the vertebra, and one each for each side of the posterior neural arch - and one secondary ossification center, giving rise to the tip of the odontoid. The odontoid process begins to ossify between the first and fifth months of gestation [1]. The apex of the odontoid process develops from a separate ossification center called the ossiculum terminale or apical odontoid epiphysis, derived from the most caudal occipital sclerotome. It usually appears by 3 years of age, continuously enlarging and fusing with the body of the odontoid process by the age of 12. Nonfusion

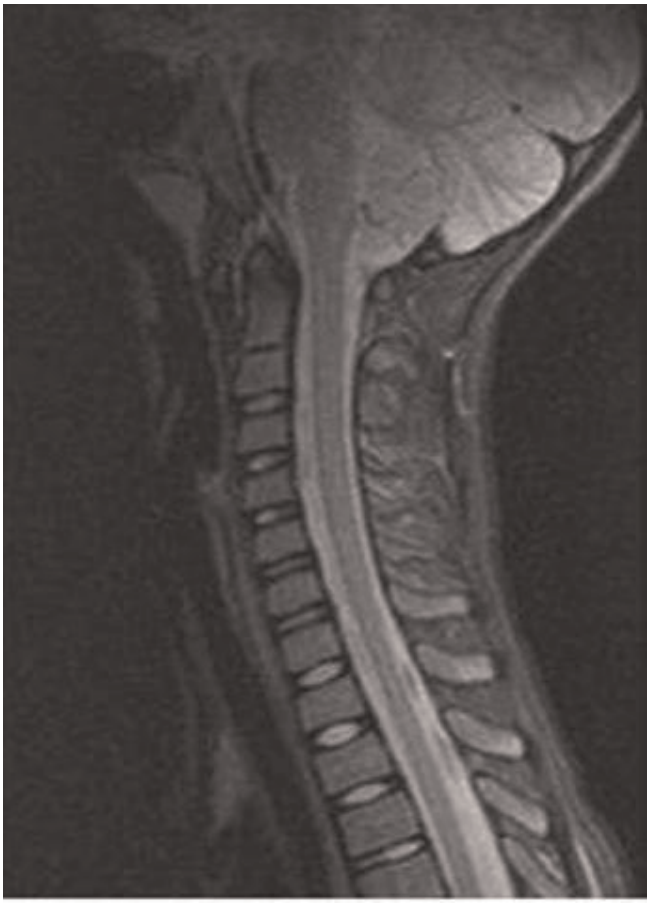

**Figure 3.** STIR sagittal MRI of the cervical spine shows the remnant synchondrosis or fibrous plate between the apical ossicle and remainder of the odontoid process. There is also a minimal amount of fluid within the atlantodental interval.

of the terminal ossicle beyond the age of 12 is termed ossiculum terminale persistens. To the best of our knowledge, prior to this report, no cases of ossiculum terminale persistens in non-Down syndrome patients less than 12 years of age has been described in the literature.

#### *Clinical presentation*

Signs and symptoms associated with atlantoaxial instability from ossiculum terminale peristens are similar to instability from any other cause - traumatic, neoplastic, degenerative, or inflammatory. These include recurrent torticollis, Lhermitte's sign, and neck pain with limited head movements. Half of all cases present with signs of lower brainstem or upper cervical cord compression [2,3]. Vertebral insufficiency may be encountered in a few cases [4]. There is often a history of significant remote or recent trauma and this often makes it difficult to distinguish between an odontoid fracture and a congenitally separate odontoid [5].

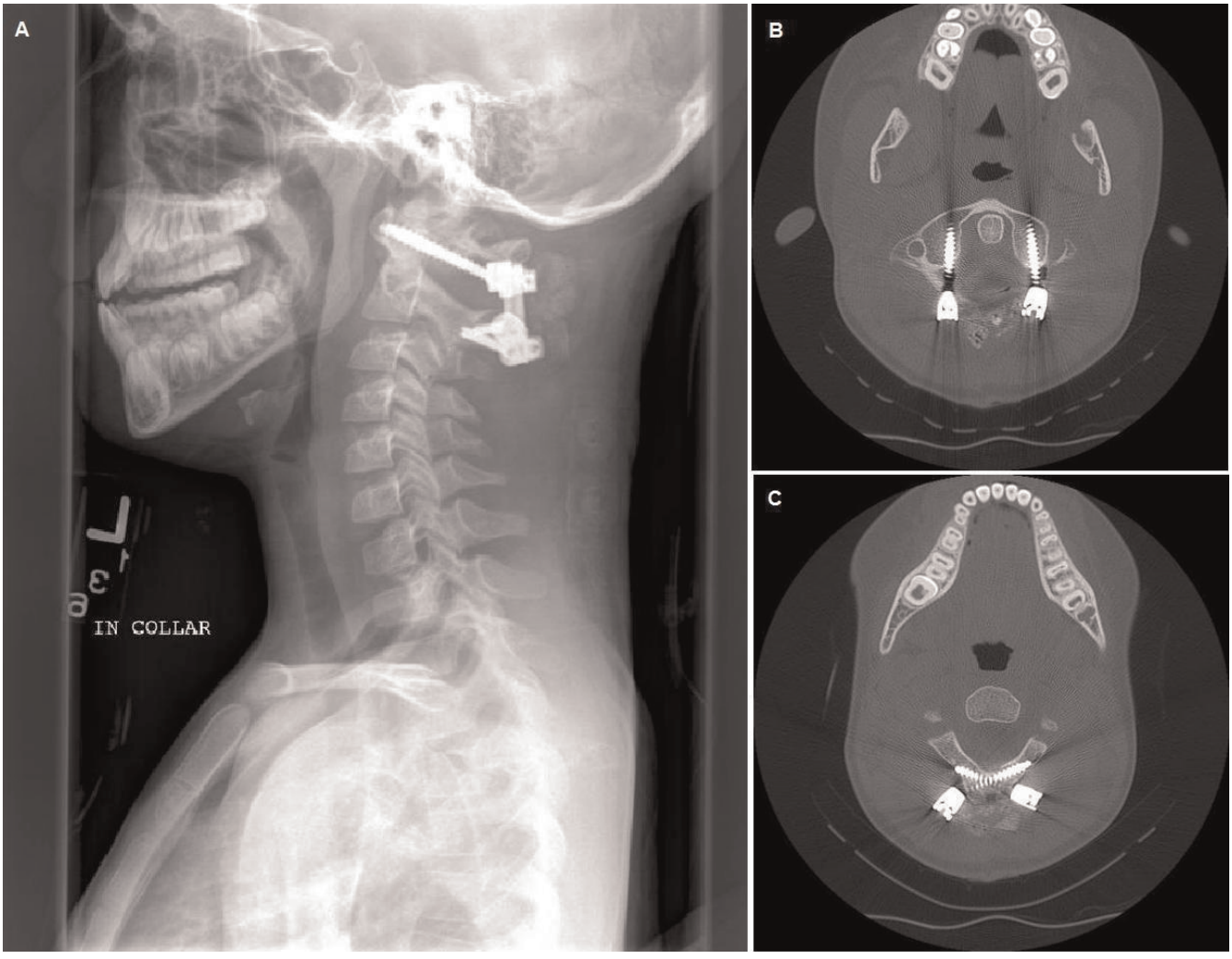


**Figure 4. (A)** Postop lateral cervical spine X-ray and axial CT of the cervical spine shows **(B)** C1 lateral mass screws and **(C)** bilateral crossing C2 translaminar screws.

The etiology for the structural anomaly of the odontoid in our patient is unclear. However, it may relate to his history of remote flexion trauma to the neck in a tumbling accident at the age of 6, resulting in traumatic disruption of the apical odontoid epiphysis [6] or ligamentous injury with loss of the tightening effect of the transverse ligament to the odontoid process [7].

### *Radiographic evaluation*

Plain X-rays and tomograms may reveal separate ossicles with intact cortical margin above the remainder of the odontoid process. Flexion-extension radiographs may demonstrate the ossicles move independently and relative to the remainder of the odontoid process; static or dynamic radiographs more consistently show an increased atlantodental interval [3,6]. The ossiculum usually rides with the anterior arch of C1, unless the transverse ligament is damaged by an associated injury [2]. A remnant of the synchondrosis can be seen between the apical ossicles and odontoid body [8].

Thin-cut CT with sagittal and coronal reconstructions is the test of choice for diagnosing ossiculum terminale persistens. Coronal reconstructed CT scan may reveal a separated odontoid process with either proper anatomic alignment or scoliotic axial malalignment of the odontoid fragments [8]. The ossicle in ossiculum terminale peristens has intact sclerotic cortical margins circumferentially [3,9] and varies in size from a few millimeters to 12 mm [2].

Magnetic resonance imaging may demonstrate ventral compression of the cervicomedullary junction caused by

the anomalous ossicula. A remnant of the synchondrosis may also be seen at the junction between the apical ossicula and odontoid proper [8]. The presence of abundant soft tissue between the ossiculum and the dens suggests a dislocated hypertrophied transverse ligament [2] or a fibrous nonunion.

In this report, we propose the presence of ossiculum terminale persistens in a 10-year-old child. Coronal CT in this patient clearly demonstrated a terminal ossicle with intact and sclerotic cortices and nonfusion of this epiphysis. This helped differentiate true pathologic ossiculum terminale persistens from a normal apical primary ossification center that has not yet fused to the odontoid peg in a child less than 12 years of age.

In some patients with ossiculum terminale persistens, good spinal stability is demonstrated because bone fusion has developed between the ossicle and odontoid process [9]. In cases of atlantoaxial instability, true separation between the ossicles and the odontoid process has been present and confirmed at autopsy examination [10].

#### *Ossiculum terminale persistens classification*

We extrapolated from the anatomic classification of os odontoideum to ossiculum terminale persistens. Most cases of atlantoaxial instability reported in the literature have been associated with dystopic ossiculum terminale late in life or associated with Down syndrome. Ligamentous laxity in combination with an anomaly of the odontoid process places the patient with Down syndrome at an even higher risk for dislocation and possible spinal cord compression [11]. Cases of ossiculum terminale persistens and atlantoaxial instability with good anatomic alignment of the odontoid tip and odontoid proper - orthotopic ossiculum terminale persistens - may be considered a subtype of this disease entity. In such cases, radiography may suggest typical anatomic alignment of the odontoid complex, but in fact the ossicle is unfused to the odontoid process. However, similar to cases of os odontoideum, our review of the literature found dystopic ossiculum terminale persistens more likely to be symptomatic than the orthotopic variant.

#### *Treatment*

The aims of management are to reduce the dislocation and prevent further recurrence of dislocation by fusion. To achieve these goals, several fusion procedures have been developed. The screw-plate fixation of C1 and C2 using unicortical C1 lateral mass screws and C2 pedicle screws was first described by Goel and Laheri [12] and then modified and popularized by Harms [13] to a screw-rod construct with bicortical C1 lateral mass screws and C2 pars screws. We felt the Harms technique was ideally suited for surgical treatment of this type of odontoid anomaly in our pediatric patient with anatomy less amenable to transarticular screw placement.

## Abbreviation

CT, computed tomography.

## Consent

Written informed consent was obtained from the patient's parent for publication of this case report and accompanying images. A copy of the written consent is available for review from the journal's Editor-in-Chief.

## Competing interests

The authors declare that they have no competing interests.

## Authors' contributions

AV was responsible for the concept and design of the manuscript and for writing and editing the manuscript. WEW aided in the editing of the manuscript. TGL aided in the editing of the manuscript. AI analyzed and interpreted the radiographic data related to the case. AJ was responsible for the concept and design of the manuscript and for writing and/or editing the manuscript. All authors read and approved the final manuscript.

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