

Os Odontoideum

A patient guide

In brief: Os odontoideum is a developmental or acquired anomaly of the upper neck in which a small, separate, well-corticated bone sits in place of the upper part of the odontoid peg (the dens) of the second cervical vertebra, with no bony continuity to the body below. Because the normal bony peg that the transverse ligament restrains is effectively replaced by a mobile ossicle, the top two neck bones (C1 and C2) can move more than they should, which is why the condition is chiefly important to a spinal surgeon as a cause of atlantoaxial instability. It is frequently discovered by chance, yet its potential to cause pressure on the spinal cord sets it apart from many other incidental imaging findings.

What os odontoideum is

Os odontoideum is best understood by first picturing the normal anatomy. The second cervical vertebra, the axis (C2), carries an upward bony projection called the odontoid process, or dens. This peg sits behind the front arch of the first cervical vertebra, the atlas (C1), and is held snugly against it by a strong band called the transverse ligament. The arrangement allows the head to rotate freely while keeping the atlas and axis securely aligned, protecting the spinal cord that runs immediately behind.

In os odontoideum this neat arrangement is disrupted. Instead of a continuous peg rising from the body of C2, there is a separate, rounded or oval ossicle that occupies the position of the upper dens, divided from a variably shortened (hypoplastic) dens base by a clear gap. The distinguishing feature is that the ossicle has a smooth, sclerotic cortical rim running the whole way around it, and the residual stump below is also corticated. In other words, both pieces of bone have a finished, rounded edge rather than the raw, jagged surface of a fresh break.

The practical consequence of this anatomy is mechanical. The transverse ligament is designed to restrain a solid peg. When that peg is replaced by a loose ossicle, the ligament has nothing competent to hold, and the atlas, together with the ossicle, may shift forwards, backwards or in several directions on the axis below. Movement of this kind narrows the space available for the spinal cord, and it is this potential for instability and cord compromise, rather than the mere presence of an extra ossicle, that gives os odontoideum its clinical significance.

Orthotopic and dystopic types

The classification still most widely used was set out by Fielding, Hensinger and Hawkins in 1980, and it divides os odontoideum into two anatomical types according to where the ossicle sits.

In the **orthotopic** type, the ossicle lies in roughly the anatomical position the dens tip would normally occupy. It tends to move in concert with the front arch of the atlas, so the two travel together as a unit. This shared movement is one reason the simple measurement of the gap between the dens and the atlas can be misleading in os odontoideum, a point returned to under assessment below.

In the **dystopic** type, the ossicle is displaced from the normal position, frequently sitting higher up near the base of the skull (the basion or clivus) and sometimes appearing functionally fused to the clivus. The dystopic pattern is the more commonly reported of the two and is generally regarded in the literature as carrying a greater tendency to instability.

A separate entity that is sometimes confused with os odontoideum is the **persistent ossiculum terminale** (os terminale persistens), in which the small secondary ossification centre at the very tip of the dens fails to fuse. This is usually a benign finding and should not be equated with os odontoideum, even though rare cases combining features of both with instability have been described.

Why it happens (congenital versus acquired/post-traumatic debate)

The origin of os odontoideum has been debated for decades and remains genuinely unsettled. Two broad explanations have been advanced, and the prevailing view is that both contribute.

The **congenital theory** holds that, in at least some cases, the odontoid fails to form or segment normally during development. This view is supported by the condition's well-recognised association with a range of inherited syndromes and skeletal dysplasias, including Down syndrome, Klippel–Feil anomaly, Morquio syndrome and other mucopolysaccharidoses, spondyloepiphyseal and multiple epiphyseal dysplasia, achondroplasia and pseudoachondroplasia, Larsen syndrome and Chiari I malformation. The clustering of os odontoideum with these conditions, particularly in younger patients, points to a developmental basis in a proportion of cases.

The **acquired or post-traumatic theory** was put forward by Fielding and colleagues, who proposed that an injury to the odontoid in early childhood can disrupt its blood supply. The segment above the level supplied by the body of C2 may then undergo avascular necrosis, gradually resorbing and remodelling into a separate, corticated ossicle. This mechanism is supported by reports of children with a documented normal odontoid who later developed an os after early trauma, including cases in which ligamentous injury was captured on MRI before the ossicle appeared on sequential imaging.

The **current synthesis**, articulated by Sankar and colleagues in 2006 and echoed by later database analyses, is that a **multifactorial aetiology** fits the evidence best. Some cases appear congenital, some acquired, with the balance shifting according to the population studied: younger, syndromic patients at one end and older, post-traumatic patients at the other.

How it relates to atlantoaxial instability

Atlantoaxial instability simply means excessive or abnormal movement between the atlas and the axis. It has many causes, including incompetence of the transverse ligament, rheumatoid pannus eroding the joint, trauma, and the general ligamentous laxity seen in Down syndrome. Os odontoideum is a distinctive cause because the instability arises from a **structurally deficient dens** rather than from a ligament problem in isolation.

The logic is mechanical. The transverse ligament may be entirely intact, but it has no solid peg to restrain. With the dens replaced by a mobile ossicle, the C1 and ossicle complex can translate on C2, most often forwards, but sometimes backwards or in more than one direction. Because the ossicle commonly moves with the front arch of the atlas, the conventional anterior atlantodental interval, the gap usually measured between dens and atlas, can stay deceptively normal even when there is significant movement. For this reason, attention shifts to how much room remains for the spinal cord and to imaging taken in different neck positions, rather than to a single static measurement.

This is the heart of why os odontoideum is treated as more than a curiosity. The combination of a deficient peg and a competent-but-unanchored ligament creates a setting in which the cord can be compromised, particularly during movement, and occasionally after relatively minor trauma.

How it tends to present (often incidental; instability; myelopathy)

Os odontoideum presents along a wide spectrum. A great many cases are **incidental and asymptomatic**, picked up on imaging performed for an unrelated reason or after a minor neck injury. The ossicle may have been present and silent for years.

When the condition does produce symptoms, the commonest complaints are **neck pain and occipital headache**. Neurological features form a separate and more serious group. These range from transient episodes of weakness, numbness or clumsiness following minor trauma, through to progressive myelopathy, the medical term for spinal cord dysfunction, which may show itself as difficulty with fine hand movements, altered sensation,

problems with balance or gait, or changes in bladder function. Less often, dynamic compromise of the vertebral arteries produces vertebrobasilar symptoms such as vertigo or fainting.

In the classic Fielding series, roughly one-third of patients had neural signs or symptoms, most reported neck pain, and most showed radiographic instability. The feared event throughout the literature is **catastrophic spinal cord injury after relatively minor trauma**, and it is this possibility that underlies the precautionary, surveillance-minded attitude towards a known os odontoideum, and the recurring discussion in the literature of avoiding contact and collision sports.

How it is assessed (dynamic imaging, canal measurements, instability index)

Assessment combines static and dynamic imaging, each contributing a different piece of the picture.

Plain radiographs, including open-mouth (odontoid) and lateral views, often first reveal the hallmark appearance: a smooth-margined, corticated ossicle sitting above a hypoplastic dens stump.

Dynamic flexion–extension lateral radiographs, or dynamic CT, are central to the assessment because they demonstrate the abnormal translation that static films may miss. Importantly, they allow both anterior and posterior subluxation to be examined and, critically, they allow the **space available for the cord** to be quantified, rather than relying on the anterior atlantodental interval alone, which is confounded because the ossicle moves with the atlas.

CT provides the best bony detail of the ossicle, the residual dens and the surrounding architecture, and it defines the vertebral artery anatomy and the bony corridors of C1 and C2, information that is essential where stabilising surgery is being planned.

MRI is the best way to assess the spinal cord itself. It shows any compression, reveals signal change within the cord (myelomalacia) that indicates established injury, and demonstrates the state of the ligaments. It remains valuable even when the bones appear realigned.

Several radiographic measures recur across published series as markers of risk. A **minimum sagittal canal diameter of less than 13 millimetres** is associated with a high risk of permanent cord damage. An **Instability Index greater than 40 per cent** and a sagittal-plane rotation around 20 degrees greater than in asymptomatic patients have been used to distinguish those with myelopathy, though the Instability Index assumes movement in the sagittal plane and under-captures multidirectional instability. Dystopic morphology and rounded ossicle types are reported to carry greater risk.

How it is distinguished from a dens fracture and related conditions

Three conditions are weighed quite differently by a spinal surgeon, and distinguishing them matters.

An acute or non-united odontoid (dens) fracture is the most important look-alike. A fresh fracture has sharp, non-corticated, irregular edges that match across the gap, as a broken biscuit does. Os odontoideum, by contrast, shows a smooth, corticated rim on both the ossicle and the residual stump, with a wider, often well-rounded gap between them. This cortical rind is the single most important discriminator. The aetiological debate, namely whether some examples of os odontoideum are in fact remodelled childhood fractures, does not change the radiographic position: the appearance of os odontoideum is settled and chronic, not acute.

Atlantoaxial instability in general describes excessive C1–C2 movement from any cause. Os odontoideum is one specific cause, defined by a structurally deficient dens rather than, for instance, an inflamed and eroded joint or an isolated ligament rupture. The mechanism, a mobile ossicle that the transverse ligament cannot anchor, is what marks it out from other forms of instability.

Basilar invagination is a separate craniovertebral problem in which the odontoid, or its remnant, migrates upwards through the foramen magnum and indents the brainstem; the difficulty is one of cranial settling. In os odontoideum the primary issue is a replaced or segmented dens predisposing to translational C1–C2 instability at

the level of the cord, not upward migration, although the two can coexist within the broader spectrum of craniovertebral junction anomalies.

What the evidence shows about management (observation versus fusion) — impersonal

The points below describe what the published literature reports, and are general information rather than guidance for any individual. There are no randomised trials in this field; the evidence consists of retrospective case series and reviews.

For **symptomatic or unstable disease**, there is little controversy in the literature. Patients with neurological signs, demonstrable cord compression, or clear atlantoaxial instability are reported to be treated with **posterior atlantoaxial (C1–C2) stabilisation and fusion**. Modern series favour rigid segmental screw fixation, such as the C1 lateral mass to C2 pedicle or pars construct widely known as the Goel–Harms technique, which permits realignment during surgery and is reported to achieve high fusion rates with low rates of non-union and acceptable vertebral-artery risk. Occipitocervical fusion is described where C1–C2 fixation is not feasible because of unfavourable anatomy or a dystopic, irreducible deformity.

The distinction between **reducible and irreducible** deformities recurs throughout. Reducible deformities are reported to be managed by posterior fusion alone, whereas irreducible cases may call for combined anterior and posterior approaches, transoral surgery, or staged reduction. Reported outcomes include significant reduction of the atlantodental interval and improvement in myelopathy scores after stabilisation, with solid fusion achieved in the great majority.

The genuinely **debated** group is incidental, asymptomatic, stable disease. Here the literature describes case-by-case decision-making. Some authors favour surveillance, with serial clinical and dynamic radiographic review, reserving surgery for emerging instability or symptoms. Others argue for earlier fixation in younger patients with radiographic instability and favourable screw anatomy. Counselling about the risk of catastrophic injury, and discussion of avoiding contact sport, recur across the published accounts.

Outlook

The outlook in os odontoideum depends heavily on which part of the spectrum a given case occupies. Many people with an incidental, stable os odontoideum remain well, and the ossicle may never cause trouble. Where instability or cord involvement is present, the published surgical literature is broadly encouraging: rigid posterior fixation is reported to produce solid fusion in the large majority, to reduce abnormal movement, and to improve myelopathy scores, generally with low rates of non-union and serious complication in experienced hands.

The central concern that shapes the cautious tone of the literature is the possibility of significant spinal cord injury after relatively minor trauma in an unstable neck. It is this, rather than the day-to-day symptoms, that explains the emphasis on accurate characterisation, dynamic imaging and ongoing review. As a general principle, the literature frames os odontoideum as a condition to be understood and monitored carefully, with the balance between watchful surveillance and stabilising surgery decided individually.