

Atlantoaxial Instability

A patient guide

In brief: Atlantoaxial instability describes excessive or abnormal movement between the first two bones of the neck, the atlas (C1) and the axis (C2). It is distinct from instability lower in the neck because this joint has no cushioning disc and relies almost entirely on ligaments and a small peg of bone, the dens, for its stability. The condition can arise from inherited differences, inflammatory disease, injury or developmental variations, and it is defined by particular measurements that describe both the movement itself and the room left for the spinal cord.

What atlantoaxial instability is

The atlantoaxial complex is the most mobile segment of the entire spine. Roughly half of all the rotation of the head occurs at this single level, where the atlas turns around the axis like a wheel around a pivot. Because so much of this movement is built into the joint by design, the boundary between normal mobility and abnormal instability is defined with care, and it rests on specific anatomical landmarks rather than on a general impression of looseness.

Atlantoaxial instability is the term used when the structures that restrain the atlas allow it to displace on the axis beyond the limits that are considered safe. The displacement is most often forwards, with the atlas sliding anteriorly relative to the axis, but it can also be rotatory, sideways, upwards (a vertical settling, where the upper neck telescopes downward) or, less commonly, backwards. What unites these patterns is that the lesion sits specifically at the C1–C2 articulation, and that the consequences are judged not simply by how much the joint moves but by how much space remains for the spinal cord as it passes through the ring of the atlas.

This focus on the cord is what gives the condition its clinical weight. The upper cervical canal is generously sized in health, which means a degree of instability can exist without any narrowing of the cord's working space. The same generous canal, however, can mask significant instability until the reserve is used up, at which point the margin for the cord may be lost comparatively suddenly.

The C1–C2 joint and what normally holds it stable (dens, transverse ligament)

The atlas, C1, is an unusual bone. It is a simple ring with no vertebral body of its own, designed to carry the skull and to swivel. The axis, C2, carries upward from its body a strong peg of bone called the odontoid process, or dens. The dens projects up into the front of the ring of the atlas and forms the pivot around which the head rotates. Unlike every typical level of the spine below it, the C1–C2 joint contains no intervertebral disc, so it draws none of its stability from the disc or from interlocking vertebral bodies.

Instead, stability here is overwhelmingly ligamentous, anchored to the dens. The single most important structure is the transverse ligament of the atlas, a thick band that arches across the back of the dens and holds it firmly against the anterior arch of C1. This ligament is the primary restraint against the atlas sliding forwards on the axis. As long as it is competent and the dens is intact, forward translation is tightly controlled.

Supporting this are the alar ligaments and the apical ligament, which run from the dens upward to the occipital condyles and the rim of the foramen magnum. These limit rotation and side-to-side and vertical movement, and they act as secondary restraints if the transverse ligament is compromised. The tectorial membrane, a broad band that is the upward continuation of the posterior longitudinal ligament, lies behind these structures and contributes further restraint. The practical point is that two elements carry most of the load: an intact dens to act as the pivot, and an intact transverse ligament to hold that pivot in place. Instability tends to follow when one or both of these fail.

What can make it unstable (congenital/syndromic incl. Down syndrome, rheumatoid, traumatic, os odontoideum)

The causes of atlantoaxial instability fall into recognisable groups, and the group matters because it shapes the mechanical nature of the problem.

****Congenital and syndromic causes.**** Some people are born with ligaments that are inherently more lax, or with a dens that has formed differently. Down syndrome (trisomy 21) is the best-known example: ligamentous laxity, sometimes combined with an underdeveloped or separated dens, produces measurable atlantoaxial instability in a notable minority of individuals, the large majority of whom have no symptoms. Several connective-tissue and skeletal conditions, including Morquio syndrome, spondyloepiphyseal dysplasia, Marfan syndrome, Larsen syndrome and neurofibromatosis, are also recognised associations.

****Os odontoideum.**** This is a particular developmental variation in which the upper part of the dens exists as a separate ossicle, with no bony continuity to the body of the axis. Because the dens is the pivot to which the transverse ligament anchors, a dens that is detached cannot do its job: the transverse ligament may itself be perfectly intact and yet be mechanically useless, because the bone it holds is no longer fixed to C2. The atlas, carrying the loose ossicle with it, can then translate on the axis.

****Inflammatory causes.**** Rheumatoid arthritis is the classic acquired cause. Long-standing inflammation produces synovial tissue, known as pannus, which erodes the transverse ligament, the dens and the small lateral mass joints. The result is first a forward atlantoaxial subluxation and, over time, a vertical settling in which the upper neck migrates downward and the dens rises toward the skull base. Pannus itself can also take up space around the cord.

****Traumatic causes.**** A sufficient injury can rupture the transverse ligament directly, fracture the dens, or fracture the ring of the atlas (a Jefferson fracture) in a way that pulls the ligament's bony attachments apart. Traumatic rotatory subluxation, in which the joint becomes locked in a rotated position, is a further pattern.

****Other causes.**** A retro-odontoid pseudotumour (a mass of tissue behind the dens), infection, and tumour can all undermine the joint. Grisel syndrome is a specific inflammatory rotatory subluxation that can follow a head or neck infection or surgery in that region, particularly in children.

How it tends to present

The presentation of atlantoaxial instability is varied, and a striking feature is that a substantial degree of instability can be present with very little to show for it. In some people the finding is identified incidentally on imaging performed for another reason.

When symptoms do occur, neck pain and stiffness, often felt at the back of the head and upper neck, are common. In rotatory fixation the head may be held in a characteristically tilted and turned posture, sometimes described as a cock-robin position. Pain in the back of the head over the territory of the C2 nerve, a form of occipital neuralgia, is also recognised.

The more significant group of features relates to the spinal cord. When the cord's working space is encroached upon, signs of myelopathy may appear: disturbance of gait and balance, clumsiness of the hands, weakness, brisk reflexes, changes in sensation, and sometimes alterations in bladder or bowel control. A brief electric-shock sensation running down the spine on bending the neck, known as Lhermitte's phenomenon, can occur. With high or vertical disease affecting the region of the brainstem and the vertebral arteries, symptoms such as dizziness, drop attacks, problems with the lower cranial nerves or, in severe cases, effects on breathing are described in the literature.

A theme that runs through the published accounts is that neurological change can be either gradual or comparatively abrupt. The generous reserve of space in the upper cervical canal can conceal severe instability until a minor injury removes the remaining margin, a pattern that is particularly emphasised in os odontoideum and in

Down syndrome.

How it is assessed and measured (ADI, PADI/space available for the cord, dynamic CT/MRI)

Atlantoaxial instability is one of the conditions in which precise measurement genuinely defines the diagnosis, and several numbers recur in the literature.

Plain radiographs, including lateral views taken in flexion and extension and an open-mouth view of the dens, are the traditional first-line images. They allow the joint to be examined while it moves.

The atlanto-dental interval (ADI) is the gap between the back of the anterior arch of the atlas and the front of the dens. In adults this is normally no more than about 3 mm, and a little more is accepted in children. Widening beyond these limits indicates laxity of the transverse ligament; greater widening implies that the ligament has failed, and very large intervals imply that the secondary restraints, including the alar ligaments, have failed as well. A particular caveat is that the ADI becomes unreliable in chronic erosive disease such as advanced rheumatoid arthritis, where the dens itself is being destroyed and the interval can paradoxically narrow even as instability worsens.

The posterior atlanto-dental interval (PADI), also called the space available for the cord (SAC), measures from the back of the dens to the front of the posterior ring of the atlas. This is widely regarded as the more clinically informative figure, because it describes the room actually left for the spinal cord rather than the looseness at the front of the joint. The literature links smaller values to a greater likelihood of neurological involvement and poorer recovery.

Steel's rule of thirds is a helpful way of understanding why this is so: at the level of the dens, the ring of the atlas is occupied roughly one-third by the dens, one-third by the cord and one-third by free space. That free third is the buffer that allows modest displacement to be tolerated, and its consumption is what explains why decompensation can be sudden.

The rule of Spence applies to Jefferson fractures: a combined sideways overhang of the lateral masses of the atlas on the axis beyond about 6.9 mm on the open-mouth view implies that the transverse ligament has ruptured.

CT, particularly dynamic CT taken in flexion and extension or with the head rotated, gives detailed pictures of the bone and characterises rotatory fixation; it is central to judging whether a displacement can be reduced.

MRI shows the spinal cord itself, any pannus, and the true space available for the cord, and it can reveal signal change within the cord that carries prognostic weight. **CT or MR angiography** is used to map the course of the vertebral arteries, which can run in unusual positions, when surgery is being considered.

How it is distinguished from related conditions

Three neighbouring conditions are worth setting apart, because the distinctions are precise and they change how the problem is understood.

Versus subaxial cervical instability (C3–C7). Instability lower in the neck involves discs, facet joints and uncovertebral joints, and it is judged by the forward translation and the angulation between vertebral bodies, using the long-established White and Panjabi thresholds of more than 3.5 mm of translation or more than 11 degrees of angulation. Atlantoaxial instability has none of this machinery: there is no disc, the stability is purely ligamentous and dens-dependent, and the relevant measurements are the ADI and the space available for the cord rather than body translation and angulation. The pivot mechanics of rotation around the dens are unique to C1–C2 and have no equivalent in the lower neck.

Versus basilar invagination. Basilar invagination is upward migration of the dens into the foramen magnum, a problem of the vertical dimension, assessed by reference lines such as Chamberlain's, McRae's and McGregor's lines. Atlantoaxial instability is predominantly a horizontal and rotatory problem at the C1–C2 joint itself. The two

can coexist, especially in rheumatoid disease and in some congenital syndromes, and an influential body of work has argued that many cases of basilar invagination are in fact driven by underlying atlantoaxial instability and can be reduced by distracting and fixing the joint. Separating fixed bony invagination from reducible, instability-driven invagination is precisely what dynamic imaging is intended to do.

****Versus os odontoideum as an isolated finding.**** Os odontoideum is a description of the bone, a dens that exists as a separate ossicle. It is a cause of atlantoaxial instability rather than a synonym for it. The clinical problem it produces is secondary instability at C1–C2 and a recognised risk of delayed, sometimes serious, neurological injury after minor trauma. The defining feature is that the instability sits at the joint, but the failed element is the dens itself, so that the transverse ligament can be intact yet ineffective.

The summary point is that in atlantoaxial instability the lesion is specifically at the C1–C2 joint, frequently involving the dens or the transverse ligament or both, and the load-bearing measurements are the ADI and the space available for the cord, not the lines and translations used for the lower neck or for vertical settling.

What the evidence shows about management (immobilisation vs C1–C2 fusion) — impersonal

The published literature describes a spectrum of approaches, and the recurring theme is that the mechanical nature of the lesion, specifically whether it is ligament or bone, and whether it is reducible or fixed, shapes what the studies report.

****External immobilisation.**** Where a joint is stable, where the restraining ligament is competent, or where a bony injury is expected to consolidate, immobilisation in a rigid collar or a halo is described as a means of allowing healing. A frequently cited distinction is between the two types of transverse ligament injury: bony avulsion injuries, in which the ligament pulls a fragment of bone away, are reported to heal without surgery in a substantial majority of cases, whereas intrasubstance tears within the ligament itself do not heal in a collar and require fixation. In Down syndrome, asymptomatic radiographic instability is generally described as being observed rather than operated upon.

****Surgical stabilisation by C1–C2 fusion.**** For instability that is irreducible or progressive, for myelopathy, or for unstable ligamentous or dens lesions, the literature describes fusing C1 to C2 so that the unstable joint is held securely. The techniques have evolved over several decades. The earliest methods used posterior wires and bone graft, the Gallie midline technique and the Brooks–Jenkins wedge-compression technique among them; these are effective but achieve lower fusion rates and poorer control of rotation than modern constructs and depend heavily on external support afterwards. The introduction of Magerl transarticular screws was a substantial advance in rigidity and fusion rate, though it carries a risk to the vertebral artery in patients whose artery runs in an anomalous position. The Goel–Harms construct, using screws in the lateral mass of C1 and in the pedicle of C2 joined by rods, is now widely regarded as the workhorse technique: it achieves high fusion rates, is biomechanically strong, can be used when vascular anatomy precludes transarticular screws, and allows the joint to be reduced during surgery.

Across the comparative evidence, several themes are consistent. Screw-and-rod constructs outperform wiring for fusion and rotational control. The anatomy of the vertebral artery is the principal technical hazard and is therefore imaged before surgery; systematic reviews place the risk of vertebral artery injury in posterior C1–C2 fusion at roughly two per cent per patient. In myelopathy, the space available for the cord and the presence of signal change within the cord carry the strongest prognostic weight, and intervention before irreversible cord change is reported to yield better outcomes. Finally, in basilar invagination accompanied by instability, the approach of distracting and fixing the joint has reframed many cases once considered irreducible as reducible.

Outlook

The outlook in atlantoaxial instability depends heavily on the cause, on whether the spinal cord has been affected and on whether the joint can be realigned. Many people with radiographic instability, particularly those identified incidentally and without symptoms, remain stable over long periods, and observation is the course described for asymptomatic findings in conditions such as Down syndrome. Where the cord has not been compromised, the published outcomes after stabilisation are generally favourable, with high rates of bony fusion using modern screw-and-rod techniques and reliable control of the unstable segment.

Where myelopathy is already established, the literature consistently indicates that the degree of recovery relates to how much cord damage has occurred before the joint is stabilised, which is why the space available for the cord and the presence of signal change within it are emphasised as prognostic markers. The trade-off inherent in fusing C1 to C2 is a loss of roughly half of the neck's rotation, although this is often tolerated better than might be expected because the lower cervical segments compensate to a degree. As with much of spinal surgery, the balance of benefit and risk is individual, and the outlook is best understood in the context of the specific cause and the specific imaging in each case.