Os odontoideum: A comprehensive review

ABSTRACT

Os odontoideum (OO) is a rare cranio cervical anomaly that is characterized by a round ossicle separated from the C2 vertebral body. With a controversial etiology and unknown prevalence in the population, OO may be asymptomatic or present in patients with myelopathic and neurological symptoms. In this literature review, we aimed to investigate epidemiology, embryology, pathophysiology, clinical presentation, and the role of diagnostic radiography in OO. By providing an overview of different management strategies, surgical complications, and postoperative considerations for OO, our findings may guide neurosurgeons in providing proper diagnosis and treatment for OO patients. A literature review was conducted using the PubMed, EMBASE, and Scopus databases. A search using the query “os odontoideum” yielded 4319 results, of which 112 articles were chosen and analyzed for insights on factors such as etiology, clinical presentation, and management of OO. The presentation of OO varies widely from asymptomatic cases to severe neurological deficits. Asymptomatic patients can be managed either conservatively or surgically, while symptomatic patients should undergo operative stabilization. Although multiple studies show different techniques for surgical management involving posterior fusion, the diversity of such cases illustrates how treatment must be tailored to the individual patient to prevent complications. Conflicting studies and the paucity of current literature on OO highlight poor comprehension of the condition. Further understanding of the natural history of OO is critical to form evidence-based guidelines for the management of OO patients. More large-center studies are thus needed to promote accurate management of OO patients with optimal outcomes.

Keywords: Atlantoaxial instability, atlantoaxial subluxation, craniocervical junction, os odontoideum, screw fixation

INTRODUCTION

Os odontoideum (OO) is a rare anomaly of the craniocervical junction (CVJ), in which a round ossicle is detached from a hypoplastic odontoid process at the body of the second cervical vertebrae (C2).[1,2] OO has a controversial etiology, natural history, and management, compounded by a paucity of literature.[3,4] The condition was first described by Giacomini in 1886 and is classified as either orthotopic or dystopic.[5,6] Orthotopic refers to an ossicle that moves in conjunction with the anterior arch of C1, while dystopic denotes an ossicle that is functionally fused to the basion.[3,7] Both types present variably, ranging from asymptomatic incidental findings to neurological dysfunction.[8,9] Such symptoms arise from the instability of the joint between the C1 and C2 vertebrae, which can further lead to atlantoaxial dislocation (AAD), occipital-cervical pain, or vertebrobasilar ischemia.[10-12] At the onset of the disease, compromised joint stability can be accompanied by neural and vascular compression, predisposing patients to sensory disorders, cervical myelopathy, quadriplegia, and other motor disturbances in addition to intracranial symptoms such as cerebellar infarction or brainstem damage.[6,13-15] In severe cases, the condition can lead to sudden death resulting from paralysis of the respiratory center.[16] Although neurological deterioration can occur in OO patients when cervical spine instability is left untreated, predictive factors for disease progression are unclear, especially in asymptomatic patients.[1,4,17] Treatment recommendations between conservative or operative
The exact proportion of OO patients with clinical symptoms is unknown. Although few studies have reported a high prevalence of OO, a comprehensive review of available surgical techniques is necessary to provide an overview of different management strategies. Surgical complications and postoperative considerations for OO patients are crucial.

### EPIDEMIOLOGY

The exact prevalence of OO is unknown due to the absence of large-scale epidemiological studies and the frequency of asymptomatic progression in patients. However, a magnetic resonance imaging (MRI) study of odontoid morphology in 133 patients aged between 19 and 81 years old found a single case (0.7%) of OO, while a retrospective study of abnormal cervical spine radiographs in 519 children detected OO in 16 patients (3.1%). Although OO is most commonly found in the pediatric population, the disease can present clinically in adult patients with myelopathic or neurological symptoms. OO can be associated with atlantoaxial joint abnormalities, inducing hypermobility, hence predisposing the cervical spine to atlantoaxial instability (AAI) or atlantoaxial subluxation (AAS), anterior subluxation (AS), or posterior subluxation (PS), and AAD. A study of 24 OO cases using MRI and high-definition computerized myelography determined that all patients had AAS, 12 had AS only (50%), and 12 had AS and PS (50%). Another study of 95 patients with Down syndrome and odontoid ossicles found that 77 (81%) patients had AAI. Among upper cervical spine injuries, the estimated frequency of posttraumatic OO combined with AAD is <2%.

OO is frequently associated with congenital syndromes such as Morquio’s disease, multiple epiphyseal dysplasia, Down syndrome, achondroplasia, Klippel–Feil syndrome, Larson syndrome, pseudoachondroplasia, Wolcott–Rallison syndrome, and chondrodystrophic calcification. Increased frequency of OO has also been reported in patients with rheumatoid arthritis and bipartite atlas. Pediatric patients with connective tissue disorders are more susceptible to OO than adults, given ligament relaxation is reduced in adults and cervical spine instability progresses more slowly. A study comparing pediatric and adult cohorts of OO patients found that Down syndrome was more common in younger individuals (6 out of 16 patients) than in adults (none out of 25 patients). The same study found that 93% of patients had neck pain and headache, with all patients experiencing weakness, sensory disturbances, and myelopathy. The exact proportion of OO patients with each of these symptoms is unclear, and many asymptomatic patients show no indication of pathological progression. However, symptomatic patients with cervical myelopathy or other neurological deficits may experience morbidity and mortality, with rare cases of acute progression into quadriplegia or sudden death.

While no large-scale screening studies have been performed, variable incidences of OO by study population and ethnic origin have been noted. A study of 15 patients of Korean ethnicity with a mean age of 23.7 years and OO-associated pseudoachondroplasia reported an OO incidence of 60%. Another study of 62 patients of Indian ethnicity with variable symptoms ranging from neck pain to limb weakness detected OO in 14.51% of the patients. In contrast, a Turkish study reported OO was present in 18 out of 16, 122 patients aged 20–70 years old (0.11%). Although few studies have reported higher incidences of OO in males compared to females, the relationship between gender and OO frequency remains unclear.

### Table 1: Presentation and diagnostic imaging of os odontoideum

| OO Type          | Presentation                        | Imaging Modality                               |
|------------------|------------------------------------|-----------------------------------------------|
| Asymptomatic     | Absence of compression/instability  | Plain radiographs + CT or MRI for confirmation |
|                  | Compression or radiographic         | or prophylactic surgery                        |
|                  | evidence of AAI                     |                                               |
| Symptomatic      | Local symptoms                      | Plain radiographs followed by CT and MRI       |
|                  | Occipitocervical pain               |                                                |
|                  | Restricted neck movement            | MRI + multiplanar reconstruction and angiography for |
|                  | Numbness/weakness/gait impairment   |                                                |
| Myelopathy       | Paresis/tetraplegia                 | Surgical planning + kinetic MRI for spinal cord |
|                  | Central cord syndrome               |                                                |
|                  | Bulbar signs                        |                                                |
|                  | Intracranial symptoms               |                                                |
|                  | Brainstem damage/cerebellar         |                                                |
|                  | infarction                          |                                                |
|                  | Cervical vertigo/seizures           |                                                |
|                  | Vertebrobasilar ischemia            |                                                |

**Table 2: Surgical management options for os odontoideum**

| Condition                        | Available surgical techniques                                      |
|----------------------------------|-------------------------------------------------------------------|
| C1-C2 instability with normal    | C1-C2 fusion, Polyaxial screw-rod fixation, Transarticular C1-C2   |
| vertebral anatomy                | screw fixation if reducible, Sublaminar wiring if vertebral artery |
|                                  | anomalies, or pediatric patient + Transoral decompression if      |
|                                  | irreducible + C1 laminectomy if cord compression                  |
| C1-C2 instability with abnormal  | C1-C2 fusion, Polyaxial screw-rod fixation, C2 transaminar screw  |
| vertebral anatomy                | fixation if aberrant vertebral artery, OCF if dystopic OO with    |
|                                  | craniovertebral instability + Transoral decompression if          |
|                                  | irreducible + C1 laminectomy if cord compression                  |
| Stable asymptomatic OO with      | C1-C2 fusion, Polyaxial screw-rod fixation + Transoral decompression |
| compression                       | or if not reduced                                                 |

**AAI - Atlantoaxial dislocation, CT - Computed tomography, MRI - Magnetic resonance imaging**

management thus depend on the severity and type of clinical presentation in OO patients. In this review, we discuss epidemiology, embryology, pathophysiology, clinical presentation, and the role of diagnostic radiography in OO. We also provide an overview of different management strategies, surgical complications, and postoperative considerations for OO patients.
inconclusive due to low sample sizes within and poor quality of limited existing literature.[31]

**EMBRYOLOGY**

The embryologic development of the atlas and axis derives from the sequential fusion of different sclerotomes at three primary ossification centers. Early in development, the fourth occipital sclerotome, known as the proatlas, forms the base of the dens and the apical ligament. [32] The first and second sclerotomes, which are separated by an epiphyseal growth plate known as the neuralcental synchondrosis, form the odontoid and axis bodies. While this vestigial disc postnatally separates the dens and the body, disappearing by 8 years of age, the body of the dens fuses with the proatlas to form the odontoid process during gestation. [33] The blood supply of the odontoid process significantly differs from the rest of the spine due to its dependence on the terminal apical arcade. [34] This arcade caudally anastomoses with the deep penetrating branches arising from the posterior ascending arteries originating in the vertebral artery. [35] The relative deficiency of the odontoid blood supply and its susceptibility to obstruction due to its complex vascular anatomy predisposes it to ischemia and necrosis during embryologic development or traumatic events. [36]

**PATHOPHYSIOLOGY**

**Congenital theory**

There are two main theories of the origin of OO: congenital and traumatic. [37] The congenital hypothesis attributes the lesion to segmental defects involving the incomplete fusion of the dens and the axis vertebral body during embryonic development due to developmental failure between ossification centers at the synchondrosis. [38] Other proposed mechanisms for this theory include incomplete caudal migration of the axis centrum, failure of segmentation, or nontraumatic osteonecrosis. [39] Findings in patients without a history of trauma and the higher frequency of OO in patients with congenital malformations such as neurofibromatosis or skeletal dysplasia support the congenital theory. [40] Patients with OO can lack aberrations of the BMP4, BMP2, and PTX1 genes. [41] Familial cases of OO with an autosomal dominant pattern of inheritance and case reports of OO in identical twins provide further evidence. [42] In addition, a radiological study suggested that a joint between the odontoid and the atlas anterior arch called the “jigsaw sign” is a reliable indicator of congenital etiology for OO. [43]

**Traumatic theory**

The posttraumatic theory supports an acquired pathology of OO in which an unrecognized odontoid fracture, followed by contraction of the alar ligament leads to avascular necrosis and osseous remodeling that contributes to the formation of the ossicle. [44] Deficiencies in arterial blood supply and trabecular bone in the base of the dens predispose the dens to stress fractures induced by repeated microtrauma, which can occur in prenatal or postnatal periods or in utero through fracture of the cartilage dens. [45] Given OO is most commonly found at the base of the dens and not at the synchondrosis, existing evidence favors a traumatic etiology over congenital etiology. [46] Numerous case reports of posttraumatic OO patients with previously documented normal odontoid processes further contradict the congenital theory and support an acquired etiology. [47]

**CLINICAL PRESENTATION**

OO has a wide range of clinical manifestations, with many cases presenting as incidental findings in asymptomatic patients [Table 1]. Common local symptoms may include neck pain and stiffness, torticollis, ataxia, shoulder pain, headache, restricted neck movement, fatigue, hoarseness of voice, respiratory dysfunction, and swallowing difficulties. [48] Isolated occipitocervical pain can result from static or dynamic compression of the occipital nerves, along with upper-extremity paresthesias involving intermittent tingling and numbness in the neck and upper limbs. [49] Repeated minor trauma to the spinal cord can also lead to lower limb weakness and gait impairment. [50] However, the association between musculoskeletal symptoms and OO may be difficult to establish due to their high prevalence in the general population. [51]

Instability of the CVJ in OO can induce abnormal atlantoaxial motion in both anterior and posterior directions. [52] Patients with AAI may additionally experience numbness or stenosis in the occipitocervical region due to compression of the spinal cord or vertebrobasilar arteries. [53] Entrapment of the vertebral artery and subsequent vascular compromise can contribute to cervical myelopathy, along with tension on the spinal cord or bony compression. [54] Myelopathic deficits can range from mild paresis to transient myelopathy to progressive tetraplegia, bulbar signs, and even death. [55] Such symptoms depend on the degree of compression and morphology at the OO site, varying between acute cord compression, chronic static compression, repetitive microtrauma, and chronic progressive damage. [56] Increased motion at the C1 to C2 level combined with chronic mechanical stress on the craniocervical ligament can further induce symptoms of central cord syndrome, hypoventilation syndrome (Ondine’s curse), Brown-Sequard syndrome, Lhermitte’s phenomenon, and cardiorespiratory
In addition to the myelopathic sensory motor and cardiorespiratory disturbances resulting from impingement of the bulbospinal junction, other symptoms of OO associated with CVJ instability can include sleep apnea, lower cranial nerve dysfunctions, hyperesthesia, bowel and bladder dysfunction, hypoesthesia, alodinia, and hyperalgesia.\textsuperscript{[14,69,70-72]} Although late neurological deterioration occurs in only 4\% of patients, OO may present with extreme intracranial symptoms such as brainstem damage or cerebellar infraction, leading to embolization.\textsuperscript{[16,74,75]} Vertebrobasilar ischemia can also occur from kinking or thrombosis of the vertebral arteries due to cervical instability in OO patients.\textsuperscript{[10]} In severe cases, vertebral artery occlusion and ischemia of the brainstem and posterior fossa structures can lead to seizures, cervical vertigo, syncope, visual disturbances, or sudden death.\textsuperscript{[21,76-81]}

\textbf{DIFFERENTIAL DIAGNOSIS}

Although the clinical presentation of OO can resemble a multitude of other conditions, accurate differentiation is critical for weighing treatment options.\textsuperscript{[8,40]} One alternate diagnosis to consider is an acute fracture of the odontoid process, which involves multiple adjacent fractures and soft tissue injury.\textsuperscript{[4,8]} While OO is located above the atlantoaxial joint, odontoid fracture results in fracture ends located at the atlantoaxial joint level.\textsuperscript{[6,8]} Another diagnosis that may be confused with OO is a persistent ossicum terminale, which is caused by the nonunion of the apex at the secondary ossification center.\textsuperscript{[8,40]} In addition to having a smaller ossicle than OO, ossicum terminale is rarely associated with C1-C2 instability and does not require surgical correction.\textsuperscript{[8,40]} Other conditions with similar clinical manifestations include degenerative disc disease of the cervical spine, dens morphologic abnormalities, cervical spondylosis, neurofibromatosis, rheumatoid arthritis, transverse ligament injury, and Grade II mechanical neck pain.\textsuperscript{[6,40]}

\textbf{RADIOGRAPHY}

Diagnosis of OO can be made using plain radiographs with open mouth, anteroposterior, lateral, and dynamic flexion-extension views for the assessment of atlantoaxial stability.\textsuperscript{[6,14,77]} Although several indicators for evaluating instability have been suggested, including the direction of AAI (anterior, posterior, or multidirectional) and the instability index for measuring the range of space available for the spinal cord, such parameters are often poor indicators of the true degree of instability in symptomatic patients.\textsuperscript{[40,82]} Varying radiographic definitions of AAI exist, with the consensus being an atlantoaxial interval (ADI) >3–5 mm on flexion-extension films.\textsuperscript{[5,40]}

Complex imaging modalities such as computed tomography (CT) and MRI can confirm the diagnosis of OO and provide more detailed anatomical illustrations of the CVJ.\textsuperscript{[10,40,43,83]} CT scans with multiplanar reconstruction can be used for surgical planning through delineation of the osseous anatomy and allows for detection of CVJ malformations.\textsuperscript{[2,8,10,70]} CT angiography can be utilized to identify anomalies commonly found in syndromic patients and visualize the arrangement of the vertebral arteries in relation to adjacent structures of the cervical spine.\textsuperscript{[2,6,14,40]} However, MRI has superior soft-tissue resolution and can provide more preoperative information regarding spinal cord compression and pathology.\textsuperscript{[8,10,40]} MRI should be performed in patients presenting with neurologic signs or painful OO indicative of synovial inflammation.\textsuperscript{[10,84]} Kinetic MRI can also directly visualize the motion of joint components and surrounding soft tissues to demonstrate AAD and signal changes of the spinal cord.\textsuperscript{[85]} However, the high sensitivity of MRI may lead to the added diagnosis of common pathologies such as disc prolapses, cervical spondylotic myelopathy, spinal cord tumors, and synovial cysts.\textsuperscript{[6,86]}

\textbf{CONSERVATIVE MANAGEMENT}

Management of asymptomatic OO is controversial due to limitations in the understanding of its natural history and may involve conservative measures or prophylactic surgical treatment.\textsuperscript{[1,45,83]} Nonsurgical treatment modalities for incidentally diagnosed patients without evidence of AAI include clinical observation with serial imaging and longitudinal radiographic follow-up.\textsuperscript{[4,8,35,87]} Immobilization consisting of collar fixation or cervical traction can also be utilized.\textsuperscript{[6,88]} Numerous reports have documented successful cases of long-term conservative management for patients with stable OO.\textsuperscript{[1,7,89,90]} Conversely, cases of neurological deterioration and sudden death have also been documented.\textsuperscript{[24,80,89,91]} Given that initially stable OO may progress to develop AAI and associated symptoms, serial observation and aggressive medical management is required to detect notable signs of cervical involvement or neurological deficit.\textsuperscript{[92]} Patient education regarding potential risks and avoidance of contact sports is also recommended.\textsuperscript{[26,80]}

Although conservative management should only be used in cases of stable OO without myelopathy, predictive risk factors of neurological involvement in asymptomatic patients remain unclear.\textsuperscript{[6,40]} A sagittal spinal canal diameter <13 mm is strongly associated with myelopathy, with studies reporting
Survival complications in os odontoideum patients

Reports of complication rates and operative morbidity for OO patients vary considerably. Postoperative complications may include wound infection, cerebrospinal fluid leakage, ongoing muscular neck pain, neurological injury, anesthesia complications, vascular injury, pseudarthrosis, and hardware loosening. In screw-based techniques, misplaced screws may induce nerve root or spinal cord injuries, while complication rates are even higher for OCF and sublaminar wiring.

Figure 1: Sagittal CT showing an OO (red arrow) incidentally found in a patient 65-year-old man after a fall. The patient did not have any symptoms. This was managed with observation and clinic follow up. CT - Computed tomography, OO - Os odontoideum

a 10% chance of its development. Compared to patients without neurological symptoms, patients with transient or progressive myelopathy had a sagittal plane rotation that was 20° larger and an instability index of more than 40%. Dystopic configurations of OO and round types of morphology are also risk factors for myelopathy and instability. Figure 1 represents a case of OO found incidentally in an adult patient who was asymptomatic and hence was treated conservatively with close follow-up.

SURGICAL MANAGEMENT

Surgical decompression and stabilization are necessary for patients with radiological indications of AAI, dynamic myelopathy, or neurological dysfunction. There are a multitude of operative techniques that can be tailored to each individual case, including posterior and anterior approaches or a combination of both. The decision paradigm depends on the anatomic variability of OO patients, including factors such as the area for arthrodesis, bone quality, and the location of spinal cord compression along with other morphological features of the CVJ. All surgical approaches can be accompanied by transoral decompression in cases of irreducible AAS or ventral compression.

Posterior screw fixation and fusion

The most common surgical approach is posterior C1-C2 fixation and fusion using screw instrumentation. Transarticular C1-C2 screw fixation (TASF) is one such technique used to achieve biomechanical stability with high fusion rates and low-profile instrumentation. However, TASF is limited to patients with reduced atlantoaxial joints and places the vertebral artery at risk due to its technical difficulty. Another technique that can be employed for atlantoaxial stabilization is C1-C2 fusion with polyaxial screw-rod fixation, also known as the Goel-Harms technique. Independent placement of the C1 and C2 lateral mass and pars allows for intraoperative reduction and decreases the risk of vertebral artery injury, rendering the procedure safer than TASF or sublaminar wiring. While this technique can be combined with TASF, it has superior fusion rates and versatility due to the possibility of joint manipulation. Disadvantages of C1-C2 fusion include the loss of normal neck rotation by up to 50% but are outweighed by the high proportion of OO patients for whom neurological compromise is prevented. Both TASF and the polyaxial screw-rod fixation techniques obviate the need for rigid external postoperative immobilization unless additional stability is desired.

Sublaminar wiring of C1-C2

Earlier techniques include posterior C1-C2 wiring combined with autologous iliac fusion, which requires an intact posterior arch of C1 and C2. Prolonged immobilization consisting of postoperative halo traction or Minerva orthosis is recommended for 3 months following the procedure. Although sublaminar wiring is recommended in cases of severe vertebral artery anomalies and pediatric patients, it is becoming less common due to its association with higher rates of pseudarthrosis and morbidity compared to newer screw-based techniques.

Occipitocervical fusion

Indications for occipitocervical fusion (OCF) include cases involving unstable dystopic OO or congenital, inflammatory, and degenerative abnormalities affecting the CVJ. These conditions may lead to posterior arching of the atlas and axis, destructive or absent occipital condyles, and cranial settling, especially in OO patients with Down syndrome. OCF induces increased limitation of neck mobility by an additional 15°-20°. Failed attempts of atlantoaxial fusion or cases of poor bone quality with increased risk of screw displacement also necessitate OCF with or without C1 laminectomy.

Surgical complications

Surgical complications of OCF include wound infection, cerebrospinal fluid leakage, ongoing neck pain, neurological injury, anesthesia complications, vascular injury, pseudarthrosis, and hardware loosening. In screw-based techniques, misplaced screws may induce nerve root or spinal cord injuries, while complication rates are even higher for OCF and sublaminar wiring.
Although postoperative neurological decline arising from direct neural tissue trauma or decreased blood pressure can lead to poor prognosis, vertebral artery injury is another severe complication in OO patients.\textsuperscript{[4,108]} Screening for additional perioperative complications including spinal cord injury, incision infection, cardiovascular stroke, pulmonary embolism, wound dehiscence, deep venous thrombosis, and donor site pain is paramount to decrease the risk of morbidity and mortality in OO patients.\textsuperscript{[109]} Higher risk for perioperative complications is reported in patients with unstable OO involving cord compression or congenital ligamentous laxity.\textsuperscript{[16]}

Although fusion rates are typically high with the advent of contemporary screw-based techniques, some OO patients undergo revision surgery.\textsuperscript{[10,26,96,110]} Anterior lesions such as synovial cysts may regress following successful posterior stabilization.\textsuperscript{[111,112]} Postoperative development of subaxial subluxation and degenerative disk diseases like kyphosis have also been reported after C1-C2 fixation and fusion.\textsuperscript{[107]}

CONCLUSIONS

The presentation of OO varies widely from asymptomatic cases to severe neurological deficits. Asymptomatic patients can be managed either conservatively or surgically, while symptomatic patients should undergo operative stabilization. Although there are numerous techniques for posterior fusion, segmental screw-rod fixation is generally the preferred option. However, treatment must be tailored to the individual patient to promote optimal outcomes and prevent complications. Further understanding of the natural history of OO may provide the foundation for evidence-based guidelines for the management of OO patients.

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Conflicts of interest
There are no conflicts of interest.

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