Review Article

Os odontoideum: A comprehensive review

ABSTRACT

Os odontoideum (OO) is a rare craniocervical 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

Sia Cho, Nathan A. Shlobin, Nader S. Dahdaleh
Department of Neurological Surgery, Northwestern University Feinberg School of Medicine, Chicago, IL, USA

Address for correspondence: Dr. Nader S. Dahdaleh, 676 North St. Clair Street, Suite 2210, Chicago, IL 60611, USA.
E-mail: nader.dahdaleh@northwestern.edu

Submitted: 18-May-22 **Accepted:** 31-Jul-22
Published: 14-Sep-22

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.jcvjs_71_22	

How to cite this article: Cho S, Shlobin NA, Dahdaleh NS. Os odontoideum: A comprehensive review. J Craniovert Jun Spine 2022;13:256-64.

Table 1: Presentation and diagnostic imaging of os odontoideum

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

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

Table 2: Surgical management options for os odontoideum

Condition	Available surgical techniques
C1-C2 instability with normal vertebral anatomy	C1-C2 fusion Polyaxial screw-rod fixation Transarticular C1-C2 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 vertebral anatomy	C1-C2 fusion Polyaxial screw-rod fixation C2 translaminar screw fixation if aberrant vertebral artery OCF if dystopic OO with craniocervical instability + Transoral decompression if irreducible + C1 laminectomy if cord compression
Stable asymptomatic OO with compression	C1-C2 fusion Polyaxial screw-rod fixation + Transoral decompression if not reduced

OO - Os odontoideum, OCF - Occipitocervical fusion

management thus depend on the severity and type of clinical presentation in OO patients.[1,9] 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.

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.[18] 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%).[19,20] Although OO is most commonly found in the pediatric population, the disease can present clinically in adult patients with myelopathic or neurological symptoms.[2,21,22] 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.[23-26] 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.[24,27] Among upper cervical spine injuries, the estimated frequency of posttraumatic OO combined with AAD is <2%.[28]

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.[6,10,29,30] Increased frequency of OO has also been reported in patients with rheumatoid arthritis and bipartite atlas.[12,31,32] 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.[27] 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).[33] The same study found that 93% of patients had neck pain and headache, with all patients experiencing weakness, sensory disturbances, and myelopathy.[32] The exact proportion of OO patients with each of these symptoms is unclear, and many asymptomatic patients show no indication of pathological progression.[7,17] 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.[13,34,35]

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%.[36] 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.[37] In contrast, a Turkish study reported OO was present in 18 out of 16, 122 patients aged 20-70 years old (0.11%).[31] Although few studies have reported higher incidences of OO in males compared to females, the relationship between gender and OO frequency remains

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.[6,15] Early in development, the fourth occipital sclerotome, known as the proatlas, forms the apex of the dens and the apical ligament[6,10,38] The first and second sclerotomes, which are separated by an epiphyseal growth plate known as the neurocentral synchondrosis, form the odontoid and axis bodies.[2,38,39] 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.[1,6,10,38] The blood supply of the odontoid process significantly differs from the rest of the spine due to its dependence on the terminal apical arcade.[14,40] This arcade caudally anastomoses with the deep penetrating branches arising from the posterior ascending arteries originating in the vertebral artery.[2,41] 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.[2,40]

PATHOPHYSIOLOGY

Congenital theory

There are two main theories of the origin of OO: congenital and traumatic.[6] 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.[42,43] Other proposed mechanisms for this theory include incomplete caudal migration of the axis centrum, failure of segmentation, or nontraumatic osteonecrosis.[44] 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.[29,45] Patients with OO can lack aberrations of the BMP4, BMP2, and PTX1 genes.[46] Familial cases of OO with an autosomal dominant pattern of inheritance and case reports of OO in identical twins provide further evidence.[47-51] 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.[6,12]

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.[52-54] 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.[6,55] 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.[14,40] Numerous case reports of posttraumatic OO patients with previously documented normal odontoid processes further contradict the congenital theory and support an acquired etiology.[56-62]

CLINICAL PRESENTATION

OO has a wide range of clinical manifestations, with many cases presenting as incidental findings in asymptomatic patients [Table 1].[3,4,43] 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.[11,62] 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.[2,63] Repeated minor trauma to the spinal cord can also lead to lower limb weakness and gait impairment.[51] However, the association between musculoskeletal symptoms and OO may be difficult to establish due to their high prevalence in the general population.[40,64]

Instability of the CVJ in OO can induce abnormal atlantoaxial motion in both anterior and posterior directions.[65] Patients with AAI may additionally experience numbness or stenosis in the occipitocervical region due to compression of the spinal cord or vertebrobasilar arteries.[10,11] Entrapment of the vertebral artery and subsequent vascular compromise can contribute to cervical myelopathy, along with tension on the spinal cord or bony compression.[63,66] Myelopathic deficits can range from mild paresis or transient myelopathy to progressive tetraplegia, bulbar signs, and even death.[6,67,68] 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.[6,7] 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

arrest.[14,69,70-72] 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, hypoanesthesia, allodynia, and hyperalgesia.[17,73]

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.[16,74,75] Vertebrobasilar ischemia can also occur from kinking or thrombosis of the vertebral arteries due to cervical instability in OO patients.[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.[21,76-81]

DIFFERENTIAL DIAGNOSIS

Although the clinical presentation of OO can resemble a multitude of other conditions, accurate differentiation is critical for weighing treatment options.[40] One alternate diagnosis to consider is an acute fracture of the odontoid process, which involves multiple adjacent fractures and soft tissue injury.[4,8] While OO is located above the atlantoaxial joint, odontoid fracture results in fracture ends located at the atlantoaxial joint level.[6,8] Another diagnosis that may be confused with OO is a persistent ossiculum terminale, which is caused by the nonunion of the apex at the secondary ossification center.[8,40] In addition to having a smaller ossicle than OO, ossiculum terminale is rarely associated with C1-C2 instability and does not require surgical correction.[6,8,40] 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.[2,40]

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.[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.[40,82] Varying radiographic definitions of AAI exist, with the consensus being an atlantodental interval (ADI) >3–5 mm on flexion-extension films.[3,40]

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.[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.[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.[2,6,14,40] However, MRI has superior soft-tissue resolution and can provide more preoperative information regarding spinal cord compression and pathology.[8,10,40] MRI should be performed in patients presenting with neurologic signs or painful OO indicative of synovial inflammation[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.[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.[6,86]

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.[1,45,80] Nonsurgical treatment modalities for incidentally diagnosed patients without evidence of AAI include clinical observation with serial imaging and longitudinal radiographic follow-up.[4,8,35,87] Immobilization consisting of collar fixation or cervical traction can also be utilized.[6,88] Numerous reports have documented successful cases of long-term conservative management for patients with stable OO.[1,7,89,90] Conversely, cases of neurological deterioration and sudden death have also been documented.[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.[92] Patient education regarding potential risks and avoidance of contact sports is also recommended.[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.[6,40] A sagittal spinal canal diameter <13 mm is strongly associated with myelopathy, with studies reporting

a 10% chance of its development.[1,7] 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%.[77] Dystopic configurations of OO and round types of morphology are also risk factors for myelopathy and instability.[33,40,93] 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.[4,16,89] 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 [Table 2].[6,94,95] 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.[25,26] All surgical approaches can be accompanied by transoral decompression in cases of irreducible AAS or ventral compression.[2,95]

Posterior screw fixation and fusion

The most common surgical approach is posterior C1-C2 fixation and fusion using screw instrumentation.[16,89,96] Transarticular C1-C2 screw fixation (TASF) is one such technique used to achieve biomechanical stability with high fusion rates and low-profile instrumentation.[10,80,97] However, TASF is limited to patients with reduced atlantoaxial joints and places the vertebral artery at risk due to its technical difficulty.[6,94,98] 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.[17,99] 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.[11,100] While this technique can be combined with TASF, it has superior fusion rates and versatility due to the possibility of joint manipulation.[6,11,99] 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.[2,26] Both TASF and the polyaxial screw-rod fixation techniques obviate the need for rigid external postoperative immobilization unless additional stability is desired.[6,99,100] Figure 2 depicts a case of symptomatic OO that was associated with C1-C2 instability and hence was treated with posterior C1-C3 fusion.

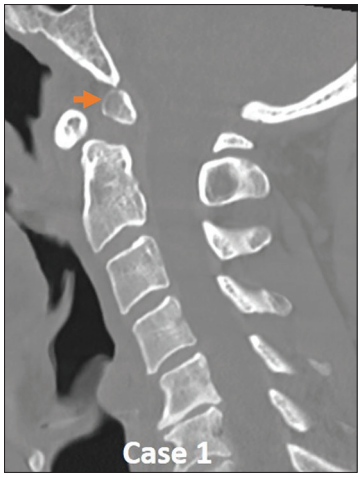

Figure 1: Sagittal CT showing an OO (red arrow) incidentally found in a patient 65-year-old man after a fall. The patient did not have any symptoms. This was managed with observation and clinic follow up. CT - Computed tomography, OO - Os odontoideum

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.[1,94,101] Prolonged immobilization consisting of postoperative halo traction or Minerva orthosis is recommended for 3 months following the procedure.[6,101] 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.[6,10,102]

Occipitocervical fusion

Indications for occipitocervical fusion (OCF) include cases involving unstable dystopic OO or congenital, inflammatory, and degenerative abnormalities affecting the CVJ.[8,103,104] 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.[6] OCF induces increased limitation of neck mobility by an additional 15°–20°.[6,103] 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.[2,105]

Surgical complications in os odontoideum patients

Reports of complication rates and operative morbidity for OO patients vary considerably.[4,98,106] Postoperative complications may include wound infection, cerebrospinal fluid leakage, ongoing muscular neck pain, neurological injury, anesthesia complications, vascular injury, pseudarthrosis, and hardware loosening.[2,89,97,107] In screw-based techniques, misplaced screws may induce nerve root or spinal cord injuries, while complication rates are even higher for OCF and sublaminar

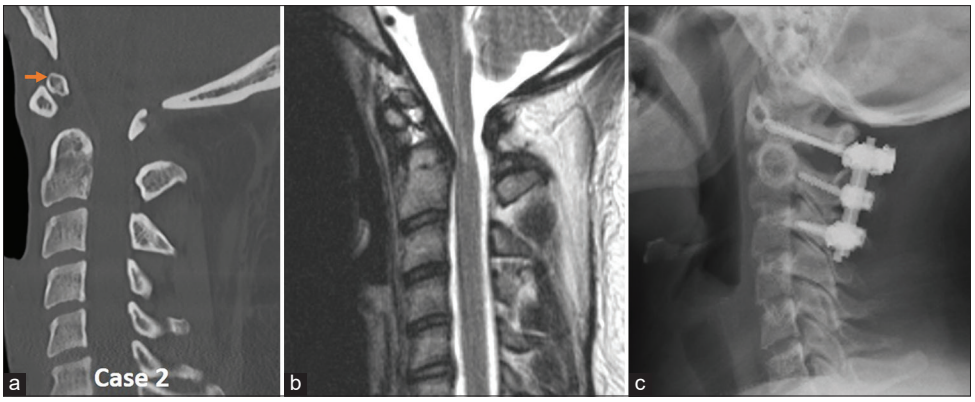

Figure 2: Sagittal CT (a) depicting an OO (arrow) in a 20-year-old female who presented with upper neck pain as well as upper and lower extremity weakness. She had brisk upper and lower extremity reflexes on exam. Sagittal T2 MRI (b) showed increased signal intensity at the level of C1-C2. X-rays (not shown) demonstrated C1-C2 instability. The patient was treated with Posterior C1-C3 fusion with resolution of her preoperative symptoms. Lateral cervical spine X-ray demonstrates the fusion (c). CT - Computed tomography, OO - Os odontoideum, MRI - Magnetic resonance imaging

wiring procedures.[6,94,98] 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.[4,108] Given osseous and vertebral artery anomalies are present in approximately one-fifth of patients with CVJ abnormalities, both anterior and posterior approaches pose significant risks of vertebral artery injury, with a reported incidence as high as 8.2% in TASF procedures alone.[106-108] 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.[109] Higher risk for perioperative complications is reported in patients with unstable OO involving cord compression or congenital ligamentous laxity.[16]

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fielding JW, Hensinger RN, Hawkins RJ. Os Odontoideum. J Bone Joint Surg Am 1980;62:376-83.
2. Pereira Duarte M, Das JM, Camino Willhuber GO. Os Odontoideum. In: StatPearls. StatPearls Publishing Copyright © 2020.Treasure Island (FL): StatPearls Publishing LLC; 2020.
3. 3 Zhao D, Wang S, Passias PG, Wang C. Craniocervical instability in the setting of os odontoideum: Assessment of cause, presentation, and surgical outcomes in a series of 279 cases. Neurosurgery 2015;76:514-21.
4. 4 Rozzelle CJ, Aarabi B, Dhall SS, Gelb DE, Hurlbert RJ, Ryken TC, *et al.* Os odontoideum. Neurosurgery 2013;72 Suppl 2:159-69.
5. Perrini P, Montemurro N, Iannelli A. The contribution of Carlo Giacomini (1840-1898): The limbus Giacomini and beyond. Neurosurgery 2013;72:475-81.
6. Arvin B, Fournier-Gosselin MP, Fehlings MG. Os odontoideum: Etiology and surgical management. Neurosurgery 2010;66:22-31.
7. Spierings EL, Braakman R. The management of os odontoideum. Analysis of 37 cases. J Bone Joint Surg Br 1982;64:422-8.
8. Wang Q, Dong S, Wang F. Os odontoideum: Diagnosis and role of imaging. Surg Radiol Anat 2020;42:155-60.
9. Helenius IJ, Bauer JM, Verhofste B, Sponseller PD, Krengel WF, Hedequist D, *et al.* Os odontoideum in children: Treatment outcomes and neurological risk factors. J Bone Joint Surg Am 2019;101:1750-60.
10. Hedequist DJ, Mo AZ. Os odontoideum in children. J Am Acad Orthop Surg 2020;28:e100-7.
11. Goel A, Patil A, Shah A, Dandpat S, Rai S, Ranjan S. Os Odontoideum: Analysis of 190 surgically treated cases. World Neurosurg 2020;134:e512-23.
12. Altun I, Yuksel KZ. An uncommon case mimicking cervical trauma: Os odontoideum. Turk J Emerg Med 2017;17:22-4.
13. Paeng SH. What causes severe myelopathy resulting in acute quadriplegia

after a surgery for atlantoaxial subluxation due to Os Odontoideum? A case report. Korean J Neurotrauma 2019;15:50.
14. Raj A, Srivastava SK, Marathe N, Bhosale S, Purohit S. Dystopic Os odontoideum causing cervical myelopathy: A rare case report and review of literature. Asian J Neurosurg 2020;15:236-40.
15. Choy WJ, Shivapathasundram G, Cassar L, Mobbs RJ. Dynamic myelopathy as a result of C1 posterior arch aplasia and os odontoideum. A case report. J Surg Case Rep 2020;2020:rjaa019.
16. Kim IS, Hong JT, Jang WY, Yang SH, Sung JH, Son BC, *et al.* Surgical treatment of os odontoideum. J Clin Neurosci 2011;18:481-4.
17. Visocchi M, Di Rocco C. Os Odontoideum syndrome: Pathogenesis, clinical patterns and indication for surgical strategies in childhood. In: Di Rocco C, Akalan N, editors. Pediatric Craniovertebral Junction Diseases: Surgical Management of Craniovertebral Junction Diseases in Children. Cham: Springer International Publishing; 2014. p. 273-93.
18. Warner W. Pediatric cervical spine. In: Canale ST, Beaty JH, editors. Campbell's Operative Orthopaedics. 11th ed. Philadelphia, PA: Mosby; 2007. p. 1879-98.
19. Perdikakis E, Skoulikaris N. The odontoid process: Various configuration types in MR examinations. Eur Spine J 2014;23:1077-83.
20. Sankar WN, Wills BP, Dormans JP, Drummond DS. Os odontoideum revisited: The case for a multifactorial etiology. Spine (Phila Pa 1976) 2006;31:979-84.
21. Klassov Y, Benkovich V, Kramer MM. Post-traumatic os odontoideum-Case presentation and literature review. Trauma Case Rep 2018;18:46-51.
22. Robson K. Os odontoideum: Rare cervical lesion. West J Emerg Med 2011;12:520-2.
23. Dross P, Rizvi A. Traumatic transverse atlantal ligamentous injury with posterior atlantoaxial instability in os odontoideum: Magnetic resonance features. Emerg Radiol 1998;5:442-5.
24. Stevens JM, Chong WK, Barber C, Kendall BE, Crockard HA. A new appraisal of abnormalities of the odontoid process associated with atlanto-axial subluxation and neurological disability. Brain 1994;117 (Pt 1):133-48.
25. Pluemvitayaporn T, Kunakornsawat S, Piyaskulkaew C, Pruttikul P, Pongpinyopap W. Chronic posterior atlantoaxial subluxation associated with os odontoideum: A rare condition. A case report and literature review. Spinal Cord Ser Cases 2018;4:110.
26. Wu X, Wood KB, Gao Y, Li S, Wang J, Ge T, *et al.* Surgical strategies for the treatment of os odontoideum with atlantoaxial dislocation. J Neurosurg Spine 2018;28:131-9.
27. Sergeenko OM, Dyachkov KA, Ryabykh SO, Burtsev AV, Gubin AV. Atlantoaxial dislocation due to os odontoideum in patients with Down's syndrome: Literature review and case reports. Childs Nerv Syst 2020;36:19-26.
28. Moreau PE, Nguyen V, Atallah A, Kassab G, Thiong'o MW, Laporte C. Traumatic atlantoaxial dislocation with odontoid fracture: A case report. Orthop Traumatol Surg Res 2012;98:613-7.
29. Nader-Sepahi A, Casey AT, Hayward R, Crockard HA, Thompson D. Symptomatic atlantoaxial instability in Down syndrome. J Neurosurg 2005;103:231-7.
30. Stevens JM, Kendall BE, Crockard HA, Ransford A. The odontoid process in Morquio-Brailsford's disease. The effects of occipitocervical fusion. J Bone Joint Surg Br 1991;73:851-8.
31. Öğüt E, Şekerci R, Şen H, Çakın H, Gediz T, Keles-Celik N. Anatomo-radiological importance and the incidence of os odontoideum in Turkish subjects: A retrospective study. Surg Radiol Anat 2020;42:701-10.
32. Garg A, Gaikwad SB, Gupta V, Mishra NK, Kale SS, Singh J. Bipartite atlas with os odontoideum: Case report. Spine (Phila Pa 1976) 2004;29:E35-8.
33. Dlouhy BJ, Policeni BA, Menezes AH. Reduction of atlantoaxial dislocation prevented by pathological position of the transverse ligament in fixed, irreducible os odontoideum: Operative illustrations and radiographic correlates in 41 patients. J Neurosurg Spine 2017;27:20-8.
34. Minderhoud JM, Braakman R, Penning L. Os odontoideum, clinical, radiological and therapeutic aspects. J Neurol Sci 1969;8:521-44.
35. Klimo *P* Jr., Coon V, Brockmeyer D. Incidental os odontoideum: Current management strategies. Neurosurg Focus 2011;31:E10.
36. Shetty GM, Song HR, Unnikrishnan R, Suh SW, Lee SH, Hur CY. Upper cervical spine instability in pseudoachondroplasia. J Pediatr Orthop 2007;27:782-7.
37. Dhadve RU, Garge SS, Vyas PD, Thakker NR, Shah SH, Jaggi ST, *et al.* Multidetector computed tomography and magnetic resonance imaging evaluation of craniovertebral junction abnormalities. N Am J Med Sci 2015;7:362-7.
38. Menezes AH. Craniocervical developmental anatomy and its implications. Childs Nerv Syst 2008;24:1109-22.
39. Hita-Contreras F, Roda O, Martínez-Amat A, Cruz-Díaz D, Mérida-Velasco JA, Sánchez-Montesinos I. Embryonic and early fetal period development and morphogenesis of human craniovertebral junction. Clin Anat 2014;27:337-45.
40. Jumah F, Alkhdour S, Mansour S, He P, Hroub A, Adeeb N, *et al.* Os Odontoideum: A comprehensive clinical and surgical review. Cureus 2017;9:e1551.
41. Akobo S, Rizk E, Loukas M, Chapman JR, Oskouian RJ, Tubbs RS. The odontoid process: A comprehensive review of its anatomy, embryology, and variations. Childs Nerv Syst 2015;31:2025-34.
42. Currarino G. Segmentation defect in the midodontoid process and its possible relationship to the congenital type of os odontoideum. Pediatr Radiol 2002;32:34-40.
43. Choit RL, Jamieson DH, Reilly CW. Os odontoideum: A significant radiographic finding. Pediatr Radiol 2005;35:803-7.
44. McHugh BJ, Grant RA, Zupon AB, DiLuna ML. Congenital os odontoideum arising from the secondary ossification center without prior fracture. J Neurosurg Spine 2012;17:594-7.
45. Nguyen JC, Pollock AN. Os odontoideum. Pediatr Emerg Care 2015;31:225-7.
46. Greenberg AD. Atlanto-axial dislocations. Brain 1968;91:655-84.
47. Sato H, Shimokawa N, Matsumoto H, Takami T. Familial os odontoideum: Proatlas segmentation abnormality. World Neurosurg 2019;130:146-9.
48. Morgan MK, Onofrio BM, Bender CE. Familial os odontoideum. Case report. J Neurosurg 1989;70:636-9.
49. Prasad A, Shah A, Sasane S, Goel A. Familial os odontoideum. World Neurosurg 2020;141:215-8.
50. Straus D, Xu S, Traynelis VC. Os odontoideum in identical twins: Comparative gene expression analysis. Surg Neurol Int 2014;5:37.
51. Tang X, Tan M, Yi P, Yang F, Hao Q. Atlantoaxial dislocation and os odontoideum in two identical twins: Perspectives on etiology. Eur Spine J 2018;27:259-63.
52. Fielding JW, Griffin PP. Os odontoideum: An acquired lesion. J Bone Joint Surg Am 1974;56:187-90.
53. Dias RP, Buchanan CR, Thomas N, Lim S, Solanki G, Connor SE, *et al.* Os odontoideum in wolcott-rallison syndrome: A case series of 4 patients. Orphanet J Rare Dis 2016;11:14.
54. Schuler TC, Kurz L, Thompson DE, Zemenick G, Hensinger RN, Herkowitz HN. Natural history of os odontoideum. J Pediatr Orthop 1991;11:222-5.
55. Hawkins RJ, Fielding JW, Thompson WJ. Os odontoideum: Congenital or acquired. A case report. J Bone Joint Surg Am 1976;58:413-4.
56. Ricciardi JE, Kaufer H, Louis DS. Acquired os odontoideum following acute ligament injury. Report of a case. J Bone Joint Surg Am 1976;58:410-2.
57. Wang S, Wang C. Acquired os odontoideum: A case report and literature review. Childs Nerv Syst 2012;28:315-9.
58. Sakaida H, Waga S, Kojima T, Kubo Y, Niwa S, Matsubara T. Os

odontoideum associated with hypertrophic ossiculum terminale. Case report. J Neurosurg 2001;94 Suppl 1:140-4.
59. Verska JM, Anderson PA. Os odontoideum. A case report of one identical twin. Spine (Phila Pa 1976) 1997;22:706-9.
60. Hammerstein J, Russo S, Easton K. Atlantoaxial dislocation in a child secondary to a displaced chondrum terminale. A case report. J Bone Joint Surg Am 2007;89:413-7.
61. Zygourakis CC, Cahill KS, Proctor MR. Delayed development of os odontoideum after traumatic cervical injury: Support for a vascular etiology. J Neurosurg Pediatr 2011;7:201-4.
62. Kim TY, Ratnayake K. Os odontoideum discovered after minor cervical trauma. Pediatr Emerg Care 2017;33:104-6.
63. Shirasaki N, Okada K, Oka S, Hosono N, Yonenobu K, Ono K. Os odontoideum with posterior atlantoaxial instability. Spine (Phila Pa 1976) 1991;16:706-15.
64. Mintken PE, Metrick L, Flynn TW. Upper cervical ligament testing in a patient with os odontoideum presenting with headaches. J Orthop Sports Phys Ther 2008;38:465-75.
65. Chrobak K, Larson R, Stern PJ. Varied clinical presentation of os odontoideum: A case report. J Can Chiropr Assoc 2014;58:268-72.
66. Zhang Z, Zhou Y, Wang J, Chu T, Li C, Ren X, *et al.* Acute traumatic cervical cord injury in patients with os odontoideum. J Clin Neurosci 2010;17:1289-93.
67. McGoldrick JM, Marx JA. Traumatic central cord syndrome in a patient with Os odontoideum. Ann Emerg Med 1989;18:1358-61.
68. Kirlew KA, Hathout GM, Reiter SD, Gold RH. Os odontoideum in identical twins: Perspectives on etiology. Skeletal Radiol 1993;22:525-7.
69. Campbell E, Brown J. Case report of os odontoideum causing Ondine's curse. Br J Neurosurg 2013;27:836-7.
70. Zhang Z, Wang H, Liu C. Acute traumatic cervical cord injury in pediatric patients with os odontoideum: A series of 6 patients. World Neurosurg 2015;83:1180.e1-6.
71. Lee DY, Jeong ST, Lee TH, Kim DH. Brown-Sequard syndrome caused by hyperextension in a patient with atlantoaxial subluxation due to an os odontoideum. Acta Orthop Traumatol Turc 2018;52:240-3.
72. Brecknell JE, Malham GM. Os odontoideum: Report of three cases. J Clin Neurosci 2008;15:295-301.
73. Rahimizadeh A, Malekmohammadi Z, Karimi M, Rahimizadeh A, Asgari N. Unstable os odontoideum contributing to cervical myelopathy and obstructive sleep apnea. Surg Neurol Int 2019;10:125.
74. Sasaki H, Itoh T, Takei H, Hayashi M. Os odontoideum with cerebellar infarction: A case report. Spine (Phila Pa 1976) 2000;25:1178-81.
75. Fukuda M, Aiba T, Akiyama K, Nishiyama K, Ozawa T. Cerebellar infarction secondary to os odontoideum. J Clin Neurosci 2003;10:625-6.
76. Takakuwa T, Hiroi S, Hasegawa H, Hurukawa K, Endo S, Shimamura T. Os odontoideum with vertebral artery occlusion. Spine (Phila Pa 1976) 1994;19:460-2.
77. Watanabe M, Toyama Y, Fujimura Y. Atlantoaxial instability in os odontoideum with myelopathy. Spine (Phila Pa 1976) 1996;21:1435-9.
78. Chang H, Park JB, Kim KW, Choi WS. Retro-dental reactive lesions related to development of myelopathy in patients with atlantoaxial instability secondary to Os odontoideum. Spine (Phila Pa 1976) 2000;25:2777-83.
79. Galli J, Tartaglione T, Calo L, Ottaviani F. Os odontoideum in a patient with cervical vertigo: A case report. Am J Otolaryngol 2001;22:371-3.
80. Klimo P Jr., Kan P, Rao G, Apfelbaum R, Brockmeyer D. Os odontoideum: Presentation, diagnosis, and treatment in a series of 78 patients. J Neurosurg Spine 2008;9:332-42.
81. Buntting CS, Dower A, Seghol H, Kohan S. Os odontoideum: A rare cause of syncope. BMJ Case Rep 2019;12:e230945.
82. Abe H, Tsuru M, Mitsumori K. Atlanto-axial dislocation-Instability index and indications for surgery (author's transl). No Shinkei Geka 1976;4:57-72.
83. Sardi JP, Iwanaga J, Oskouian RJ, Tubbs RS. First gross anatomic findings of an os odontoideum. World Neurosurg 2017;101:813.e1-3.
84. Mahmoud B, Hammad W, Abdrabou A. Os odontoideum: Report of eight cases and review of the literature. Open J Modern Neurosurg 2020;10:464-74.
85. Hughes TB Jr., Richman JD, Rothfus WE. Diagnosis of Os odontoideum using kinematic magnetic resonance imaging. A case report. Spine (Phila Pa 1976) 1999;24:715-8.
86. Gigante PR, Feldstein NA, Anderson RC. C1-2 instability from os odontoideum mimicking intramedullary spinal cord tumor. J Neurosurg Pediatr 2011;8:363-6.
87. Hadley MN, Walters BC, Grabb PA, Oyesiku NM, Przybylski GJ, Resnick DK, *et al*. Os odontoideum. Neurosurgery 2002;50 Suppl 3:S148-55.
88. Verhofste BP, Glotzbecker MP, Birch CM, O'Neill NP, Hedequist DJ. Halo-gravity traction for the treatment of pediatric cervical spine disorders. J Neurosurg Pediatr 2019;25:1-10.
89. Dai L, Yuan W, Ni B, Jia L. Os odontoideum: Etiology, diagnosis, and management. Surg Neurol 2000;53:106-8.
90. Wilson JR, Dettori JR, Vanalstyne EM, Fehlings MG. Addressing the challenges and controversies of managing os odontoideum: Results of a systematic review. Evid Based Spine Care J 2010;1:67-74.
91. Michaels L, Prevost MJ, Crang DF. Pathological changes in a case of os odontoideum (separate odontoid process). J Bone Joint Surg Am 1969;51:965-72.
92. White D, Al-Mahfoudh R. The role of conservative management in incidental os odontoideum. World Neurosurg 2016;88:695.e15-7.
93. Shaffrey C, Chenelle A, Abel M, Menezes A, Wiggins G. Anatomy and physiology of congenital spinal lesions. In: Spine Surgery: Techniques, Complication Avoidance, and Management. London: Churchill Livingstone; 2005. p. 61-87.
94. Huang DG, Hao DJ, He BR, Wu QN, Liu TJ, Wang XD, *et al.* Posterior atlantoaxial fixation: A review of all techniques. Spine J 2015;15:2271-81.
95. Govindasamy R, Preethish-Kumar V, Gopal S, Rudrappa S. Is transoral surgery still a relevant procedure in atlantoaxial instability? Int J Spine Surg 2020;14:657-64.
96. Wu Z, Xu J, Wang Z, Xia H, Zhang Q, Ma X, *et al.* Transoral approach for revision surgery of os odontoideum with atlantoaxial dislocation. Orthopedics 2014;37:e851-5.
97. Ni B, Zhou F, Xie N, Guo X, Yang L, Guo Q, *et al.* Transarticular screw and C1 hook fixation for os odontoideum with atlantoaxial dislocation. World Neurosurg 2011;75:540-6.
98. Gluf WM, Schmidt MH, Apfelbaum RI. Atlantoaxial transarticular screw fixation: A review of surgical indications, fusion rate, complications, and lessons learned in 191 adult patients. J Neurosurg Spine 2005;2:155-63.
99. Du YQ, Li T, Ma C, Qiao GY, Yin YH, Yu XG. Biomechanical evaluation of two alternative techniques to the Goel-Harms technique for atlantoaxial fixation: C1 lateral mass-C2 bicortical translaminar screw fixation and C1 lateral mass-C2/3 transarticular screw fixation. J Neurosurg Spine 2020; 32 (5): p. 682-688.
100. Harms J, Melcher RP. Posterior C1-C2 fusion with polyaxial screw and rod fixation. Spine (Phila Pa 1976) 2001;26:2467-71.
101. Brooks AL, Jenkins EB. Atlanto-axial arthrodesis by the wedge compression method. J Bone Joint Surg Am 1978;60:279-84.
102. Bach CM, Arbab D, Thaler M. Treatment strategies for severe C1C2 luxation due to congenital os odontoideum causing tetraplegia. Eur Spine J 2013;22:29-35.
103. Kukreja S, Ambekar S, Sin AH, Nanda A. Occipitocervical fusion surgery: Review of operative techniques and results. J Neurol Surg Part B Skull Base 2015;76:331-9.
104. Wu A, Jin H, Dou H, Pan X, Sheng S, Huang C, *et al.* Anterior decompression through transoral axis slide and rotation osteotomy for salvage of failed posterior occipitocervical fusion: A novel technique note. Ann Transl Med 2020;8:129.
105. Abd-El-Barr MM, Snyder BD, Emans JB, Proctor MR, Hedequist D.

Combined preoperative traction with instrumented posterior occipitocervical fusion for severe ventral brainstem compression secondary to displaced os odontoideum: Technical report of 2 cases. J Neurosurg Pediatr 2016;25:724-9.
106. Chang H, Park JB, Choi BW, Kang JW, Chun YS. Posterior sublaminar wiring and/or transarticular screw fixation for reducible atlantoaxial instability secondary to symptomatic os odontoideum: A neglected technique? Asian Spine J 2019;13:233-41.
107. Yang SY, Boniello AJ, Poorman CE, Chang AL, Wang S, Passias PG. A review of the diagnosis and treatment of atlantoaxial dislocations. Global Spine J 2014;4:197-210.
108. Chen Q, Brahimaj BC, Khanna R, Kerolus MG, Tan LA, David BT, *et al.* Posterior atlantoaxial fusion: A comprehensive review of surgical techniques and relevant vascular anomalies. J Spine Surg 2020;6:164-80.
109. Huang DG, Wang T, Hao DJ, He BR, Liu TJ, Ma XW, *et al.* Posterior C1-C2 screw-rod fixation and autograft fusion for the treatment of os odontoideum with C1-C2 instability. Clin Neurol Neurosurg 2017;163:71-5.
110. Qureshi MA, Afzal W, Malik AS, Ullah JS, Aebi M. Os-odontoideum leading to atlanto-axial instability-Report of surgery in four cases. J Pak Med Assoc 2008;58:640-2.
111. Aksoy FG, Gomori JM. Symptomatic cervical synovial cyst associated with an os odontoideum diagnosed by magnetic resonance imaging: Case report and review of the literature. Spine (Phila Pa 1976) 2000;25:1300-2.
112. Chang H, Park JB, Kim KW. Synovial cyst of the transverse ligament of the atlas in a patient with os odontoideum and atlantoaxial instability. Spine (Phila Pa 1976) 2000;25:741-4.