Os Odontoideum

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RECOMMENDATIONS

Diagnosis:

Level III:

Plain radiographs of the cervical spine (anterior-posterior, open mouth-odontoid, and lateral) and plain dynamic lateral radiographs performed in flexion and extension are recommended to diagnose and evaluate os odontoideum, with or without tomography (computerized or plain) and/or magnetic resonance imaging of the craniocervical junction.

Management:

Level III:

- Clinical and radiographic surveillance or posterior C1-C2 internal fixation and fusion are recommended for patients with os odontoideum without symptoms or neurological signs.
- Posterior C1-C2 internal fixation and fusion are recommended for patients with os odontoideum with neurological symptoms, signs, or C1-C2 instability.
- Postoperative halo immobilization is recommended as an adjunct to posterior internal fixation and fusion unless rigid C1-C2 internal fixation is accomplished.
- Occipital-cervical internal fixation and fusion with or without C1 laminectomy is recommended in patients with os odontoideum who have irreducible dorsal cervicomedullary compression and/or evidence of associated occipital-atlantal instability.
- Ventral decompression should be considered in patients with os odontoideum who have irreducible ventral cervicomedullary compression.

RATIONALE

The definition of an os odontoideum is uniform throughout the literature: an ossicle with smooth circumferential cortical margins representing the odontoid process that has no osseous continuity with the body of C2.[1] The origin of os odontoideum remains debated in the literature with evidence for both acquired and congenital causes.[2-4] The etiology of os odontoideum, however, is not relevant to its diagnosis or subsequent management.

Diagnosis

Os odontoideum can present with a wide range of clinical symptoms and signs or as an incidental finding on imaging. The literature has focused on 3 groups of patients with os odontoideum: those with occipital-cervical pain alone, those with myelopathy, and those with intracranial symptoms or signs from vertebrobasilar ischemia.[5] Patients with os odontoideum and myelopathy have been subcategorized further into those with transient myelopathy (commonly after trauma), those with static myelopathy, and those with progressive myelopathy.[6] Because patients with occipital-cervical pain, myelopathy, or vertebrobasilar ischemia likely will have etiologies other than os odontoideum, the diagnosis of os odontoideum is not usually considered until imaging is obtained. The presence of an os odontoideum is usually first suggested after plain cervical spine radiographs are obtained. Most often, plain cervical spine radiographs are sufficient to obtain a diagnosis.[7]

Os odontoideum has been classified into 2 anatomic types: orthotopic and dystopic. Orthotopic defines an ossicle that moves with the anterior arch of C1, and dystopic defines an ossicle that is functionally fused to the basion. The dystopic os odontoideum may sublux anterior to the arch of C1.[6] Tomograms and computerized tomography (CT) have been used to better define the bony anatomy of the os odontoideum and the odontoid process. Plain dynamic radiographs in flexion and extension have been used to depict the degree of abnormal motion between C1 and C2. Most often, there is anterior instability, with the os odontoideum translating forward in relation to the body of C2. However, at times, one will see either no discernible instability or "posterior instability" with the os odontoideum moving posteriorly into the spinal canal during neck extension.[6,8]

With respect to diagnosis, there are 2 issues regarding the imaging of os odontoideum. First, although plain radiographs are often diagnostic for os odontoideum, the sensitivity and specificity of plain cervical radiographs for os odontoideum remain unreported. The utility of confirmatory studies such as CT and magnetic resonance imaging (MRI) has not been well defined. Second, after the diagnosis of os odontoideum on plain cervical x-rays, instability and osseous anomalies associated with os odontoideum can influence clinical management. The best methods of further evaluating or excluding these complicating factors deserve definition.

Management

The natural history of untreated os odontoideum covers a wide spectrum. The literature provides many examples of both asymptomatic and symptomatic patients with known os odontoideum who have never been treated and who have had no reported new problems in follow-up over many years.[1] Conversely, examples of sudden spinal cord injury in association with os odontoideum after minor trauma have also been reported.[9,10] The natural history of os odontoideum is variable, and predictive factors for deterioration, particularly in the asymptomatic patient, have not been identified. Indications for surgical stabilization include simply the existence of an os odontoideum, os odontoideum in association with occipital cervical pain alone, and/or os odontoideum in association with neurological symptoms and signs.[1,6,10-12] Other factors that may assist in determining the need for stabilization and/or decompression include C1-C2 instability, associated deformities, and spinal cord compression. A variety of techniques have been used to stabilize C1 and C2 in patients with os odontoideum.[1,6,8,13-26] Fusion success rates and complication rates for these various procedures may provide evidence as to whether a preferred method of C1-C2 arthrodesis is supported by the literature.

Finally, neural compression is an important consideration in patients with os odontoideum. Neural compression may be anterior from a combination of bone and soft tissue or posterior from the dorsal arch of C1. Surgical techniques to stabilize and fuse across the craniocervical junction with or without C1 laminectomy and techniques that provide ventral decompression have been reported in the treatment of os odontoideum with irreducible neural compression.[18,21,27]

The guidelines author group of the Joint Section on Disorders of the Spine and Peripheral Nerves of the American Association of Neurological Surgeons and the Congress of Neurological Surgeons previously produced a medical evidence-based guideline on this topic.[28] The present review is undertaken to update the medical evidence on the diagnosis and management of patients with os odontoideum since that 2002 publication.

SEARCH CRITERIA

A National Library of Medicine (PubMed) computerized literature search from 1966 to 2011 was performed through MEDLINE using the key phrase "os odontoideum." The search identified 224 articles. Articles written in English were reviewed. Thirty-eight articles that described the clinical aspects and management of patients with os odontoideum were identified and used to generate these guidelines. None of the articles meeting selection criteria provided Class I or Class II medical evidence. All 38 citations offered Class III medical evidence on the diagnosis and/or management of os odontoideum. These 38 articles represent the basis for this review and are summarized in Evidentiary Table format (Table).

SCIENTIFIC FOUNDATION

Diagnostic Evaluation

There is no literature that describes the sensitivity and specificity of imaging studies for os odontoideum. Dai et al[18] in their review of 44 patients with os odontoideum used tomography, CT, and MRI, in addition to "routine" plain cervical radiographs (anterior-posterior, lateral, open mouth, flexion and extension x-rays), in 39, 27, and 22 patients, respectively. Matsui et al[29] described only the plain radiographs of 12 patients with os odontoideum. They excluded patients with Down syndrome and Klippel-Feil anomalies. The authors made no mention of any other studies to obtain or confirm the diagnosis in these 12 patients. Likewise, Watanabe et al[20] and Spierings and Braakman[1] described the plain radiographs of 34 and 37 patients, respectively, with os odontoideum, without reference to other imaging studies. Fielding et al[6] described 35 patients with os odontoideum: "Each patient had extensive roentgenographic investigation, including multiple roentgenograms of the cervical spine and *often* flexion-extension lateral roentgenograms and flexion-extension laminagrams." No mention was made as to whether additional studies beyond static plain c-spine x-rays were necessary to confirm the diagnosis of os odontoideum in their series of patients.

The literature supports the ability of plain cervical spine radiographs to establish the diagnosis of os odontoideum. There is no compelling medical evidence in the literature that supports the need for additional studies to confirm the diagnosis of os odontoideum.

Specific characteristics or associated abnormalities of os odontoideum, including C1-C2 instability, soft-tissue masses, spinal canal diameter, associated osseous anomalies, spinal cord appearance, and vertebral artery compromise, have been investigated with a variety of imaging studies. The imaging of abnormal motion and spinal cord compression in association with os odontoideum has received the most attention in the literature.

Instability of C1-C2 in association with os odontoideum has been investigated with multiple imaging modalities. Using flexion and extension lateral cervical spine x-ray studies in 33 patients, Fielding et al[6] reported 22 patients (67%) with anterior instability who had a mean atlanto-dens interval of 10.3 mm, 5 patients (15%) with posterior instability (mean posterior translation of the os odontoideum during extension of 8.4 mm), 3 patients (9%) with < 3.0 mm of C1-C2 motion, and 3 patients (9%) with no detectable C1-C2 motion. Eight patients (23%) had both anterior and posterior instability. The authors noted that cineradiography was helpful in examining the range of motion at C1-C2 in these patients, but it was not of benefit in the measurement of the degree of instability. Of note is that almost one fifth of the patients in their series manifested no radiographic evidence of C1-C2 instability.

Klimo et al[10] retrospectively reviewed flexion/extension lateral cervical spine radiographic findings in 60 patients with os odontoideum as part of their surgical case series. Defining C1-C2 instability as "a change in C-1 translation from the neutral position" either anteriorly or posteriorly, they found only 4 patients (7%) who demonstrated no radiographic evidence of instability. Anterior instability was observed in 42 patients (70%), ranging from 3 to17 mm (mean, 8.8 mm); posterior instability ranging from 4 to 13 mm (mean, 7.7 mm) was demonstrated in 6 patients (10%); and 8 patients (13%) had both anterior and posterior instability. Only 1 of their 14 "asymptomatic" patients was found to have no radiographic evidence of instability.

Spierings and Braakman[1] studied 21 of their 37 patients with os odontoideum with flexion and extension cervical spine radiographs or tomograms. They measured the maximal distance the os odontoideum moved in the sagittal plane, the inner diameter of the atlas, and the minimal spinal canal diameter (the distance between the posterior aspect of the C2 body and the dorsal arch of C1 during flexion). They compared these measurements in 2 groups: those with and those without myelopathy. The degree of C1-C2 instability did not correlate with neurological status, but the measured minimal spinal canal diameter was significantly smaller ($P < .05$) in the group with myelopathy. They identified 13 mm as the critical anterior-posterior spinal diameter. Watanabe et al[20] made similar measurements in 34 patients using plain lateral cervical radiographs in flexion and extension. Like Spierings and Braakman, the degree of instability in their patients did not correlate with the presence of myelopathy. Shirasaki et al[8] described radiographic findings on lateral flexion and extension radiographs in 9 patients with os odontoideum. They reported that a distance of ≤ 13 mm between the os odontoideum and the dorsal arch of C1 "specifically defined severe cervical myelopathy" in their patients. They also found that the degree of C1-C2 instability did not correlate with the presence of myelopathy. Yamashita et al[30] studied atlantoaxial subluxation with plain radiography and MRI and correlated the imaging studies with the degree of myelopathy in 29 patients (4 with os odontoideum). They found that the degree of myelopathy did not correlate with the distance of subluxation of C1 on C2 on plain radiographs. The degree of cord compression on MRI correlated well with the degree of myelopathy determined clinically. Matsui et al[29] classified os odontoideum into 3 types according to the morphology of the os odontoideum on plain radiographs: round, cone, and blunt tooth. They compared these 3 os odontoideum types to the degree of clinical myelopathy and found the degree of myelopathy correlated most closely with the "round" os odontoideum type. Kuhns et al[31] described the MRI appearance of os odontoideum in 4 children and identified signal changes within the posterior ligaments consistent with trauma. They could not discern whether these changes represented a primary or secondary phenomenon with respect to atlantoaxial instability.

These studies provide 2 consistent conclusions: The degree of C1-C2 instability does not appear to correlate with neurological status in patients with os odontoideum, and sagittal spinal canal diameter on plain radiographs of ≤ 13 mm is strongly associated with clinical symptoms and signs of myelopathy.

Beyond plain spine radiographs and flexion-extension x-rays, imaging to assist with operative planning of unstable os odontoideum receives brief mention in several reports.[9,14,19,27,32] Important factors to consider before proceeding with surgical intervention for this disorder are the ability to reduce C1-C2, spinal cord compression, an assimilated atlas, an incomplete C1 ring, the course of the vertebral arteries at C1 and C2, and the presence of an associated congenital fusion of the cervical spine (eg, Klippel-Feil). Plain radiographs, tomography, and CT scans provide information regarding the ability to achieve anatomic alignment of C1 on C2 and the presence or absence of a congenital fusion. CT can provide important information about the bony anatomy at the craniocervical junction, including the completeness of the atlas ring and the position of the transverse foramina at C1 and C2.[33] Hosono et al[34] made interesting observations on the different motions of the posterior arch of C1 in relation to C2 in patients with os odontoideum. They observed 2 patterns of motion: linear and sigmoid. They felt that in those patients with a sigmoid-shaped motion pattern, posterior wiring and fusion techniques may not provide adequate stability. MRI is the best modality for viewing cord compression even after apparent C1-C2 realignment.[30] The selection of and necessity for additional imaging in the evaluation of patients with os odontoideum appear to be made on a patient-by-patient basis. The literature provides no convincing evidence as to which patients should undergo supplemental imaging (CT or MRI) after the diagnosis of os odontoideum has been made.

Management

The universal theme of the various management strategies offered in the treatment of patients with os odontoideum has been either confirming or securing cervical spinal stability at the C1-C2 levels. The earliest reports of os odontoideum describe small pediatric case series treated surgically. In 1978, Griswold et al[35] described 4 children with os odontoideum who underwent posterior C1-C2 wiring and autologous iliac fusion. Three children had successful arthrodesis. The fourth child did not achieve fusion/stability despite 3 attempts. In the same year, Brooks and Jenkins[16] described their technique of C1-C2 wiring and fusion and reported 3 children with os odontoideum who were immobilized postoperatively in Minerva jackets. All 3 patients achieved successful fusion. In summary, 6 of the 7 children with os odontoideum described in these 2 early reports were successfully treated.

Two larger series, reported in the early 1980s, included adults and children with os odontoideum and described both operative and nonoperative management strategies for these patients. Fielding et al[6] described 35 patients with os odontoideum, of whom 27 had radiographic evidence of instability. Twenty-six of these 27 patients underwent successful posterior C1-C2 internal fixation and fusion (Gallie type). Fusions were noted to be "solid" after 2 months of immobilization in children and after 3 months in adults. One patient with instability refused surgery and remained well at the 2-year follow-up. The 8 remaining patients with no evidence of C1-C2 instability managed nonoperatively remained well at last follow-up of 1 to 3 years. Spierings and Braakman[1] described 37 patients with os odontoideum whom they managed. Seventeen were treated surgically. They provide 20 patients for analysis of the natural history of os odontoideum. Information about radiographic stability was provided for only 21 of the 37 patients they reported. Sixteen patients in their series presented with neck pain only or had an incidentally discovered os odontoideum. Nine of these 16 patients had flexion and extension radiographs. Of these 9 patients, 7 had abnormal motion of ≥ 8.0 mm. With a median follow-up of 7 years, none of these 16 patients developed a neurological deficit. Four additional patients who presented with myelopathy were treated nonoperatively with follow-up from 6 months to 14 years. Three of these 4 patients presented with transient myelopathy and had no recurrence at last follow-up despite abnormal motion of C1 on C2 of 8 to 16 mm. The fourth patient had a stable monoparesis at last follow-up. Of the 17 patients who underwent surgery, 1 patient had neurological worsening and 2 died. Eight of the 17 patients treated surgically underwent posterior C1-C2 internal fixation and fusion. Nine patients underwent occipital-cervical internal fixation and fusion with C1 laminectomy. The authors did not report a single failed fusion. They had a combined surgical morbidity and mortality of 18% (3 of 17 patients). The authors concluded that patients with os odontoideum without C1-C2 instability can be managed without surgical stabilization and fusion with good result. Although they did not provide operative treatment to every os odontoideum patient with C1-C2 instability, those with myelopathy and greater amounts of instability were more likely to be operated on.

More recently, Zhang et al[12] reviewed 10 cases of os odontoideum diagnosed following acute spinal cord injury. All were treated with posterior internal fixation and fusion (8 C1-C2, 2 O-C2). No complications were reported, and all patients' American Spinal Injury Association scores improved. If these 3 series are considered representative of patients with os odontoideum, the implication is that minimally symptomatic or asymptomatic patients with os odontoideum without C1-C2 instability can be managed nonoperatively with little or no morbidity over time. Although patients with os odontoideum and myelopathy or C1-C2 instability have been managed conservatively, most patients with myelopathy and/or instability are treated surgically.

Clements et al[5] in 1995 reported a patient who had a documented os odontoideum without instability who at 5 years of follow-up developed symptomatic frank C1-C2 instability that required surgical stabilization and fusion. It appears that a lack of C1-C2 instability at initial diagnosis does not guarantee that instability will not develop in these patients. In the largest published os odontoideum case series to date, Klimo et al[10] described 3 individuals who sustained neurological deterioration after a known os odontoideum was left untreated. Therefore, it is recommended that patients diagnosed with os odontoideum be counseled regarding the risk of delayed instability/late neurological deterioration and that clinical and radiographic follow-up be provided to patients with untreated os odontoideum.

The literature reviewed on the surgical treatment of os odontoideum for the original "Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injury" described posterior C1-C2 fusion procedures using sublaminar cables with or without transarticular screw fixation.[28] All reports provided Class III medical evidence. The conclusions drawn were that posterior C1-C2 fusion is a treatment option for os odontoideum and that postoperative halo immobilization should be used unless successful transarticular screw fixation was accomplished. Since the publication of the original guidelines, the reported transarticular screw fixation experience has grown, and several new techniques for atlantoaxial internal fixation have been described for patients with os odontoideum. Gluf et al[13] reported 191 C1-C2 transarticular screw fixation and fusion procedures in patients > 16 years of age. Although only 4 patients had os odontoideum as the cause of their instability, the overall radiographic fusion rate was 98%. Five vertebral artery injuries occurred in their series, including bilateral injuries in 1 patient resulting in death. Simultaneously, the same authors reported a series of C1-C2 transarticular screw fixation and fusion procedures in patients < 16 years of age. Os odontoideum was the surgical indication in 22 of 67 procedures they accomplished. Radiographic fusion was documented in all cases, and only 2 vertebral artery injuries were observed, both asymptomatic. The overall complication rate was 10.4%, predominantly wound infections.

More recently, Reilly and Choit[11] described their experience with transarticular screw fixation/posterior C1-C2 fusion for pediatric C1-C2 instability. Nine of the 12 cases reported were performed for os odontoideum-associated instability. All achieved radiographic fusion. There were no vertebral artery injuries or other major complications.

Klimo et al[10] reviewed 78 patients with os odontoideum who were 1.5 to 73 years of age (mean, 20.5 years), all treated surgically. Posterior fusion with transarticular screw fixation was performed in 76 patients (C1-C2 in 74 and O-C2 in 2). One patient underwent odontoid screw fixation. One posterior C1-C2 fusion was supplemented with a C1 lateral mass/C2 pedicle screw construct.

All patients achieved radiographic fusion in 2 to 17 months (median, 4.8 months). Approximately 90% of patients had improvement or resolution of presenting symptoms. The only reported complication was a superficial wound infection that was treated successfully with antibiotics. The authors' high success rate and negligible complication rate and the 3 patients who experienced neurological deterioration following nonsurgical management led them to recommend consideration of C1-C2 posterior fusion with internal screw fixation for all patients diagnosed with os odontoideum.

Review of the recent literature identified 2 series of polyaxial screw/rod fixation and fusion of C1-C2, first described by Harms et al[41] for instability caused by os odontoideum. Brecknell and Malham[25] reported successful C1-C2 internal fixation and fusion for 3 adult patients with os odontoideum using polyaxial C1 lateral mass and C2 pedicle screws placed with image guidance. No complications were reported. Haque et al[22] subsequently reviewed 17 cases of screw fixation of the upper cervical spine in pediatric patients (3-17 years of age) without intraoperative image guidance. Five C1-C2 posterior fusions were performed in patients with os odontoideum, all with bilateral C1 lateral mass and C2 pars screws. Intentional bilateral C2 nerve root sacrifice was performed in 2 of the 5 cases to improve visualization of the C1 lateral mass. There were no unintended neurovascular injuries, wound infections, or hardware revisions, and all patients achieved radiographic stability. C2 nerve root sacrifice reportedly did not cause postoperative pain. Polyaxial screw/rod constructs appear to be comparable to transarticular screw fixation for the treatment of atlantoaxial instability.

Other alternative posterior fusion techniques have been reported recently. Visocchi et al[23] reported 7 children with unstable os odontoideum associated with Down syndrome (n = 6) or Morquio syndrome (n = 1). All underwent posterior C1-C2 or O-C2 fusions with rod and sublaminar wire internal fixation constructs. All patients were immobilized postoperatively with halo or sterno-occipital mandibular immobilizer devices for a minimum of 4 months. Six patients had no complications and documented radiographic fusion after 6 months. The seventh patient required a revision fusion procedure following treatment of a wound infection and cerebrospinal fluid leak. Chamoun et al[24] described their experience using translaminar screw fixation and fusion followed by nonhalo immobilization for pediatric upper cervical spine instability. "Os odontoideum/terminale" was present in 2 of the 7 cases reviewed. All 7 accomplished subsequent fusion documented radiographically. The only reported complication was dysphagia resulting from a malpositioned C1 lateral mass screw. Ni et al[14] used C1 laminar hooks and C2 pedicle screws for internal fixation in a series of 13 C1-C2 posterior internal fixation and fusion procedures. Os odontoideum was the cause of instability in 4 patients and chronic odontoid fracture was the pathology in another 4 patients. After 3 months in a Philadelphia collar, radiographic fusion was achieved in all 13 cases with no report of vertebral artery injury or new neurological deficits. These reports provide additional Class III medical evidence that posterior fusion procedures are an effective treatment for C1-C2 instability, regardless of the internal fixation construct applied.

Apfelbaum et al[36] described their experience in treating recent and remote ($\geq$ 18 months after injury) odontoid injuries with anterior screw fixation. They reported a fusion rate of 25% in 16 "remote" odontoid injuries. If an os odontoideum were considered anatomically similar to a remote odontoid fracture, then the rate of fusion for os odontoideum treated with an odontoid screw fixation would likewise be expected to be poor. Anterior C1-C2 transfacet fixation techniques may have merit in the surgical treatment of os odontoideum, but there are no descriptions in the literature of its application for os odontoideum.

The surgical treatment of patients with C1-C2 instability in association with os odontoideum has been demonstrated to be successful when combined fusion and internal fixation techniques are used, usually in conjunction with postoperative halo immobilization. Fusion success rates and reports of operative morbidity varied considerably among the clinical case series reported in the literature. Although the numbers are small, rigid C1-C2 internal fixation and fusion (whether transarticular screw, C1-C2 screw/rod, C1 hook/C2 screw, etc) have been associated with higher rates of fusion compared with posterior wiring and fusion techniques alone. Of note, patients treated with rigid C1-C2 screw fixation have been managed in hard collars postoperatively, obviating the need for halo immobilization devices. If a rigid internal fixation construct is not used in the treatment of unstable os odontoideum, postoperative halo immobilization as an adjunct to dorsal internal fixation and fusion is recommended.

Ventral or transoral decompression for irreducible ventral cervicomedullary compression in association with os odontoideum has been suggested.[27] Reports of the management of ventral compression and os odontoideum are scant. In a review of 36 patients with Down syndrome and craniovertebral junction abnormalities, 12 patients with os odontoideum were described. Eleven of the 36 patients were noted to have basilar invagination. Five of these 11 patients with basilar invagination had irreducible ventral spinal cord compression and were treated with transoral decompression. The authors reported stable to excellent outcomes without complications following transoral decompression in all 5 patients; however, the total number of patients who had basilar invagination resulting from os odontoideum was not given. The report implies, however, that selected patients with atlantoaxial instability and irreducible symptomatic ventral cervicomedullary compression may benefit from ventral decompression. Recently, Lü et al[21] described endoscopically assisted anterior release and reduction of fixed atlantoaxial dislocations in 21 consecutive patients. Seven of their patients had os odontoideum and another 8 had "late odontoid fractures." Anatomic reduction was attained in all patients followed by successful posterior internal fixation and fusion. They reported no complications. On the other hand, Dai and colleagues[18] reported the successful use of occipital cervical internal fixation and fusion, with or without C1 laminectomy, in cases of

TABLE. Evidentiary Table: Os Odontoideum

Reference	Description of Study	Evidence Class	Conclusions
Lü et al,[21] *Spine*, 2010	Retrospective series of 21 fixed atlantoaxial dislocation cases treated with endoscope-assisted anterior release and reduction followed by posterior fixation	III	Fixed atlantoaxial dislocation was due to os odontoideum in 7 cases, all of which achieved anatomic reduction with no complications reported.
Ni et al,[14] *European Spine Journal*, 2010	Retrospective case series of 13 posterior C1-C2 fusion procedures using bilateral C1 hook and C2 pedicle screw fixation	III	Os odontoideum was the indication in 4 cases and chronic odontoid fracture in another 4. Radiographic fusion was documented in all 13, and no new neurological deficits or vertebral artery injuries were observed.
Zhang et al,[12] *Journal of Clinical Neuroscience*, 2010	Review of 10 patients 22-52 y of age treated with posterior fusion for os odontoideum; mean follow-up of 20.7 mo	III	All 10 patients presented with acute spinal cord injury following minor trauma and underwent posterior C1-C2 (n = 8) or O-C2 (n = 2) fusion. No complications were reported, and all patients' American Spinal Injury Association scores improved.
Haque et al,[22] *Journal of Neurosurgery: Pediatrics*, 2009	Retrospective case series of 17 patients 3-17 y of age treated surgically with posterior C1-C2 or O-C2 fusion using screw fixation	III	Lateral mass screws were placed in C1, whereas C2 fixation was with pars or laminar screws. Os odontoideum was the indication in 5 cases, radiographic stability was achieved in 100%, and there were no vertebral artery injuries or major complications.
Visocchi et al,[23] *Acta Neurochirurgica*, 2009	Retrospective series of 7 children with os odontoideum (5 with Down syndrome, 1 with Morquio syndrome) treated with posterior sublaminar wiring/fusion	III	Six patients achieved radiographic fusion within 6 mo; the seventh required repeat bone grafting to achieve fusion (after treatment of cerebrospinal leak and wound infection). No other complications and no new neurological deficits were reported.
Chamoun et al,[24] *Neurosurgery*, 2008	Review of translaminar screw fixation for upper cervical spine instability in 7 pediatric (< 15 y of age) patients (follow-up, 4-21 mo)	III	Os odontoideum/terminale was the cause of instability in 2 patients; radiographic fusion was achieved in 100%; and 1 patient experienced prolonged dysphagia resulting from C1 lateral mass screw malposition.
Klimo et al,[10] *Journal of Neurosurgery: Spine*, 2008	Retrospective case series of 78 patients 1.5-73 y of age with os odontoideum; median follow-up of 14 mo	III	All were treated surgically (77 posterior rigid screw fixation/fusion and 1 odontoid screw; no mortality or major morbidity); 13 of 14 asymptomatic patients had C1-C2 instability on dynamic x-rays. Neurological deterioration following conservative management of os odontoideum was seen in 3 cases.
Brecknell and Malham,[25] *Journal of Clinical Neuroscience*, 2008	Report of 3 cases of os odontoideum treated surgically with posterior C1-C2 fusion and polyaxial screw/rod fixation	III	C1 lateral mass and C2 pedicle screw fixation was achieved using image guidance. All 3 patients achieved radiographic fusion without complications.
Reilly and Choit,[11] *Journal of Pediatric Orthopedics*, 2006	Retrospective case series of 12 patients 5-17 y of age treated surgically with posterior C1-C2 fusion and transarticular screw fixation; mean follow-up of 5.1 y	III	9 had os odontoideum as the cause of C1-C2 instability, radiographic fusion was achieved in 100%, and there were no vertebral artery injuries or other major complications.
Gluf and Brockmeyer,[13] *Journal of Neurosurgery: Spine*, 2005	Retrospective case series of 67 C1-C2 transarticular screw fixations in patients < 16 y of age	III	Os odontoideum was the indication in 22 cases; radiographic fusion was achieved in 100%; and 2 asymptomatic vertebral artery injuries were observed.

(Continues)

TABLE. *Continued*

Reference	Description of Study	Evidence Class	Conclusions
Gluf et al,[13] *Journal of Neurosurgery: Spine*, 2005	Retrospective case series of 191 C1-C2 transarticular screw fixations in patients >16 y of age	III	Os odontoideum was the indication in 4 cases; radiographic fusion was achieved in 98%; and 5 vertebral artery injuries occurred—bilateral in 1 case resulting in patient death.
Apfelbaum et al,[36] *Journal of Neurosurgery*, 2000	18 patients with odontoid fractures incurred ≥ 18 mo before treatment who were treated with anterior odontoid screw fixation	III	16 patients with follow-up. 25% fusion rate. 3 with screw fracture and 2 with screw pullout.
Brockmeyer et al,[15] *Journal of Neurosurgery*, 2000	Review of transarticular screw placement in 31 children, 12 children with os odontoideum (age, 4-16 y)	III	55 of 62 possible sites deemed suitable for transarticular screws. All children with os odontoideum were able to have 2 screws placed.
Dai et al,[18] *Surgical Neurology*, 2000	A review of 44 patients 7-56 y of age with os odontoideum; mean follow-up, 6.5 y	III	7 patients were asymptomatic. 5 of these 7 were treated with a cervical collar only and have remained stable. 39 underwent fusion successfully (9 atlantoaxial and 33 occipitocervical). Symptoms and signs disappeared in 26 and improved in 13.
Taggard et al,[27] *Journal of Neurosurgery*, 2000	A review of craniovertebral junction abnormalities in 36 patients with Down syndrome; os odontoideum present in 12	III	Twenty-seven underwent surgical procedures. Of 11 with basilar invagination, it was irreducible in 5 and transoral decompression was performed.
Dickman and Sonntag,[16] *Neurosurgery*, 1998	Review of 121 patients treated with posterior C1-C2 transarticular screws and wired posterior bone struts; os odontoideum was present in 9; this group was compared with 74 patients treated with posterior wiring techniques alone	III	2 failures in the transarticular group. The cause of the C1-C2 instability was not stated for these 2 failures. 1 of 8 patients with os odontoideum in the posterior wiring group had a nonunion. Overall fusion rate for transarticular was 98% vs 86% for posterior wiring techniques.
Wang et al,[19] *Pediatric Neurosurgery*, 1999	16 children treated for atlantoaxial instability, 4 of whom had os odontoideum and were treated with C1-C2 transarticular screws and posterior wiring and fusion techniques	III	All fused. No halo immobilization. Transarticular screws were successfully used in children as young as 4 y of age.
Kuhns et al,[31] *Journal of Pediatric Orthopedics*, 1998	4 children with os odontoideum underwent magnetic resonance imaging examinations	III	All 4 children had changes in the nuchal cord consistent with injury.
Lowry et al,[7] *Journal of Neurosurgery*, 1997	A review of 25 children requiring upper cervical fusions; 11 children had os odontoideum	III	10 underwent a Brooks-type C1-C2 fusion. 2 of these children did not fuse. 1 underwent a Gallie-type fusion. This child remained unstable and was revised to a Brooks-type fusion. which was successful.
Matsui et al,[29] *Spine*, 1997	Review of the plain radiographic morphology of C2 in 12 patients (15-71 y of age) with os odontoideum unrelated to any syndrome	III	3 configurations described from an anteroposterior view: round, cone, and blunt tooth. Myelopathy was more severe in the group with a round configuration.
Verska and Anderson,[4] *Spine*, 1997	Report of a pair of identical twins, 1 with and 1 without os odontoideum	III	History of trauma in the twin with an os odontoideum. Fell at 3 y of age and had torticollis and neck pain for several months.

(Continues)

TABLE. *Continued*

Reference	Description of Study	Evidence Class	Conclusions
Watanabe et al,[20] *Spine*, 1996	Review of 34 patients with os odontoideum (5-76 y of age). Divided into groups by Rowland classification (1 = local symptoms, 2 = posttraumatic transient myelopathy, 3 and 4 = progressive myelopathy or intracranial symptoms); lateral neutral and dynamic radiographs obtained; sagittal-plane rotation angle, minimum distance, and instability index were measured	III	Low correlation between sagittal-plane rotation angle and instability index. Sagittal-plane rotation angle of $>$ 20° or instability index of $>$ 40% correlates with myelopathy.
Clements et al,[5] *Injury*, 1995	Report of nonoperative treatment of an incidentally discovered os odontoideum without C1-C2 instability at diagnosis	III	After 5 y, profound C1-C2 instability and symptoms had developed, necessitating posterior instrumentation and fusion.
Coyne et al,[17] *Neurosurgery*, 1995	Review of posterior C1-C2 fusion and instrumentation techniques; 5 of 32 patients had os odontoideum	III	3 of 5 with os odontoideum failed with posterior wiring techniques. All were immobilized in halos. 2 of 5 developed new neurological deficits as operative complications.
Stevens et al,[3] *Brain*, 1994	Review of abnormal odontoids and C1-C2 instability; 24 of 62 patients with os odontoideum: 9 children and 15 adults	III	Periodontoid soft-tissue thickening was present only in those with Morquio disease. Following fusion, the odontoid was noted to partially or completely regenerate in cases of Morquio disease.
Menezes and Ryken,[9] *Neurosurgery*, 1992	Review of 18 Down syndrome patients with symptomatic cervicomedullary compromise; 4 had os odontoideum	III	All 4 had gross instability on dynamic radiographs. Successful fusion with posterior wiring techniques and full-thickness rib grafts. Immobilized for a "minimum of 3 mo."
Dickman et al,[38] *Journal of Neurosurgery* 1991	Review of 36 patients treated with C1-C2 posterior wiring and fusion for various reasons; 4 patients had os odontoideum (16, 25, 38, 43 y of age); all patients were maintained in a halo for 12 wk after surgery	III	Of the 4 with os odontoideum, 3 developed osseous unions and 1 had a stable fibrous union (follow-up, 15-44 months). No complications for these 4 patients.
Hosono et al,[34] *Spine*, 1991	Cine-radiographic evaluation of 6 patients with os odontoideum	III	2 types of C1 posterior arch translation: straight (vertical; n = 4) and sigmoid (n = 2). Correlated abnormal motion with biomechanics of posterior wiring techniques.
Smith et al,[39] *Spine*, 1991	Review of 17 children operated on for C1-C2 instability; 11 had os odontoideum; posterior wiring techniques, autologous bone, and halo used in all	III	2 of the 11 with os odontoideum had nonunions. 1 cord injury thought secondary to sublaminar wire passage.
Shirasaki et al,[8] *Spine*, 1991	9 patients with os odontoideum and posterior instability had 3 radiographic parameters measured: distance between the os odontoideum and C2 spinous process in extension, distance between the os odontoideum and posterior C1 arch, and "degree of instability"; these findings were compared with their neurological status	III	Those without history or evidence of myelopathy had a distance between the os odontoideum and C2 spinous process in extension of $>$ 16 mm. It was $\leq$ 16 mm in those with myelopathy. The presence or absence of myelopathy was not related to the degree of instability. In those with myelopathy and a distance between the os odontoideum and posterior C1 arch $>$ 13 mm, there was reversible cord compression in extension. In those with a distance between the os odontoideum and posterior C1 arch of $\leq$ 13 mm, the cord remained compressed in flexion and extension.
Morgan et al,[2] *Journal of Neurosurgery*, 1989	Report of 3 family members with C2-3 Klippel-Feil abnormalities and os odontoideum	III	Ages: 16 y (index case), 39 y (father), and 64 y (paternal grandmother). None with neurological signs or symptoms.

(Continues)

TABLE. *Continued*

Reference	Description of Study	Evidence Class	Conclusions
Yamashita et al,[30] *Acta Radiologica*. 1989	Correlation of clinical status, magnetic resonance imaging, and radiographs in 29 patients with C1-C2 instability; 4 had os odontoideum	III	The atlanto-dens interval did not correlate with the degree of myelopathy, but magnetic resonance imaging degree of cord compression correlated with degree of myelopathy.
French et al,[32] *Journal of Pediatric Orthopedics*, 1987	Review of dynamic cervical spine radiographs in 185 patients with Down syndrome	III	Six had abnormal odontoids consistent with os odontoideum for an incidence of 3%. 3 had prior radiographs showing no abnormality. 1 had an exaggerated atlanto-dens interval of 6 mm.
Spierings and Braakman,[1] *Journal of Bone and Joint Surgery: British Volume*, 1982	37 patients with os odontoideum; 20 treated conservatively	III	Of 20 managed conservatively, 1 was lost to follow-up. 15 had no myelopathy (median follow-up, 5 y), and none developed myelopathy. Of 4 with myelopathy (follow-up, 0.5, 1, 7, and 14 y), 1 died of cancer, 1 has neck pain, 1 has neck pain and paresthesias, and 1 has headaches.
Fielding et al,[6] *Journal of Bone and Joint Surgery: American Volume*, 1980	35 patients (3-65 y of age) with os odontoideum; 25 patients were symptomatic	III	22 patients had anterior instability with a mean atlanto-dens interval of 10.3 mm. 5 had posterior instability. 3 had no detectable motion. 3 had le$<$ 3 mm of C1-C2 motion. 26 underwent posterior fusion successfully. 5 were not operated, 3 were asymptomatic with no instability. They remained well with no instability at 1, 2, and 3 y, respectively. 1 patient with instability refused surgery but was well at 2 y of follow-up. 1 patient died of renal failure.
Brooks and Jenkins,[16] *Journal of Bone and Joint Surgery: American Volume*, 1978	3 children (8, 11, 12 y of age) with os odontoideum treated with sublaminar C1-C2 wires and autologous iliac crest graft; Minerva cast immobilization	III	All fused. Spontaneous extension of fusion to C3 in 1 child.
Dyck,[40] *Neurosurgery*, 1978	Review of 8 children (7-17 y of age) with os odontoideum; 6 were treated with posterior wiring and fusion of C1-3; external immobilization for "usually" 3 to 4 mo	III	6 children underwent posterior fusion by the author. Two required reoperation.
Griswold et al,[35] *Journal of Bone and Joint Surgery: American Volume*, 1978	4 patients with os odontoideum treated with sublaminar C1-C2 wires and autologous iliac crest	III	3 fused. 1 did not fuse after 3 attempts.

irreducible deformity with cervicomedullary neural compression in 33 patients with os odontoideum. They described improvement in all patients and no complications related to their dorsal only approach. Although it may seem intuitive to remove ventral neural compression in association with os odontoideum, the literature suggests that treatment with dorsal stabilization and fusion without ventral decompression is an effective management option.

SUMMARY

Plain cervical spine radiographs appear adequate to make a diagnosis of os odontoideum in the vast majority of patients with this disorder. Lateral flexion and extension radiographs can provide useful information regarding C1-C2 instability. Tomography (CT or plain) may be helpful to define the osseous relationships at the skull base, C1, and C2 in patients in whom the craniovertebral junction is not well visualized on plain radiographs. The degree of C1-C2 instability identified on cervical x-rays does not correlate with the presence of myelopathy. A sagittal diameter of the spinal canal at the C1-C2 level of $<$ 13 mm does correlate with myelopathy detected on clinical examination. MRI can depict spinal cord compression and signal changes within the cord that correlate with the presence of myelopathy.

Surgical treatment is not required for every patient in whom os odontoideum is identified. Patients who have no neurological deficit and no instability at C1-C2 on flexion and extension studies can be managed without operative intervention. Even patients with documented C1-C2 instability and neurological deficits have been managed nonoperatively without clinical consequence during finite follow-up periods. Most investigators who study and treat this disorder favor operative stabilization and fusion of C1-C2 instability associated with os odontoideum. The concern exists that patients with os odontoideum with C1-C2 instability have an increased likelihood of future spinal cord injury. Although not supported by Class I or Class II medical evidence from the literature, multiple case series (Class III medical evidence) suggest that stabilization and fusion of C1-C2 is meritorious in this circumstance. Because a patient with an initially stable os odontoideum has been reported to develop delayed C1-C2 instability and because there are examples of patients with untreated stable os odontoideum who have developed neurological deficits following minor trauma, surgical consideration and longitudinal clinical and radiographic surveillance of patients with os odontoideum without instability are recommended.

Posterior C1-C2 internal fixation with arthrodesis in the treatment of os odontoideum provides effective stabilization of the atlantoaxial joint in the majority of patients. Posterior wiring and fusion techniques supplemented with postoperative halo immobilization provided successful fusion in 40% to 100% of cases reported. Rigid internal screw fixation and fusion appear to have merit in the treatment of C1-C2 instability in association with os odontoideum and appear to obviate the need for postoperative halo immobilization. Neural compression in association with os odontoideum has been treated with a reduction of deformity, dorsal decompression of irreducible deformity, and ventral decompression of irreducible deformity, each in conjunction with C1-C2 or occipital cervical fusion with internal fixation. Each of these combined approaches has provided satisfactory results. Odontoid screw fixation has no role in the treatment of this disorder.

KEY ISSUES FOR FUTURE INVESTIGATION

A cooperative, multi-institutional, natural history study of patients with os odontoideum without C1-C2 instability would provide demographic and clinical information that may provide predictive factors for the development of subsequent instability. In a related study, the prevalence of os odontoideum as an incidental finding should be established.

The literature supports essentially no treatment for os odontoideum, even with C1-C2 subluxation. Whether activity restriction is called for in these patients would be helpful and should be studied.

A cooperative, multi-institutional, prospective, randomized trial of posterior wiring and fusion techniques with and without C1-C2 rigid internal screw fixation for patients with os odontoideum and C1-C2 instability would help to definitively identify the risks and merits of each of the 2 procedures in this patient population.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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