

OS ODONTOIDEUM

RECOMMENDATIONS

Diagnosis:

Standards: There is insufficient evidence to support diagnostic standards.

Guidelines: There is insufficient evidence to support diagnostic guidelines.

Options: Plain radiographs of the cervical spine (A-P, open mouth odontoid, and lateral) and plain dynamic lateral radiographs performed in flexion and extension are recommended. Tomography (computerized or plain) and/or MR of the craniocervical junction may be considered.

Management:

Standards: There is insufficient evidence to support treatment standards.

Guidelines: There is insufficient evidence to support treatment guidelines.

Options:

- Patients with os odontoideum without symptoms or neurological signs may be managed with clinical and radiographic surveillance.
- Patients with os odontoideum, particularly with neurological symptoms and/or signs, and C1-2 instability may be managed with posterior C1-2 internal fixation and fusion.
- Postoperative halo immobilization as an adjunct to posterior internal fixation and fusion is recommended unless successful C1-C2 transarticular screw fixation and fusion can be accomplished.
- Occipital-cervical fusion with or without C1 laminectomy may be considered in patients with os odontoideum who have irreducible cervicomedullary compression and/or evidence of associated occipital-atlantal instability.

- Transoral decompression may be considered in patients with os odontoideum who have irreducible ventral cervicomedullary compression.

RATIONALE

The definition of an os odontoideum (os) 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 (16,22). The etiology of os odontoideum remains debated in the literature with evidence for both acquired and congenital causes (18,23,25). The etiology of os, however, does not play an important role in its diagnosis or subsequent management.

Diagnosis

Os odontoideum can present with a wide range of clinical symptoms and signs, as well as be an incidental finding on imaging. The literature has focused on three groups of patients with os odontoideum, 1) those with occipital-cervical pain alone, 2) those with myelopathy, and 3) those with intracranial symptoms or signs from vertebrobasilar ischemia (4). Patients with os odontoideum *and* myelopathy have been subcategorized further into those with: 1) transient myelopathy (commonly following trauma), 2) static myelopathy, and 3) progressive myelopathy. (10) Because patients with occipital-cervical pain, myelopathy, or vertebrobasilar ischemia likely will have etiologies other than os, the diagnosis of os odontoideum is not usually considered until imaging is obtained. The presence of an os is usually first suggested after obtaining plain cervical spine radiographs. Most often plain cervical spine radiographs are sufficient to obtain a diagnosis.(15)

Os odontoideum has been classified into two anatomic types, orthotopic and dystopic. Orthotopic defines an ossicle that moves with the anterior arch of C1, while dystopic defines an ossicle that is functionally fused to the basion. The dystopic os may sublux anterior to the arch of C1.(10) Tomograms and computerized tomography have been used to better define the bony anatomy of the os 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 subluxing forward in relation to the body of C2. However, at times one will see either no discernible instability, or “posterior instability” with the os moving posteriorly into the spinal canal during neck extension.(10,20)

With respect to diagnosis, the issues regarding the imaging of os odontoideum are two: First, while plain radiographs are often diagnostic for os, the sensitivity and specificity of plain cervical radiographs for os odontoideum have not been reported. The utility of confirmatory studies such as computerized and plain tomography and MR has not been well defined. Second, following the diagnosis of os odontoideum on plain cervical x-rays, instability and osseous anomalies associated with os can influence clinical management. The best methods of further evaluating or excluding these complicating factors deserve definition.

Management

The natural history of untreated os 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. (22) Conversely, examples of sudden spinal cord injury in association with os following minor trauma have also been reported.(17) 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, os in association with occipital cervical pain alone and/or os in association with neurological deficit. (10,22) Other factors that may assist in determining the need for stabilization and/or decompression include C1-2 instability, associated deformities, and spinal cord compression. A variety of techniques have been used to stabilize C1 and C2 in patients with os odontoideum. (2,3,5,6,10,20,22,26,27) Fusion success rates and complication rates for these various procedures may provide evidence as to whether a preferred method of C1-2 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.(6,24) The literature will be examined in light of the risks and benefits these techniques may provide to patients with os odontoideum.

SEARCH CRITERIA

A National Library of Medicine computerized literature search from 1966 to 2001 was performed through MEDLINE using the key phrase “os odontoideum”. The search identified 121 articles. Articles written in English were reviewed. Twenty-seven articles that described the clinical aspects and management of patients with os odontoideum were identified and used to generate these guidelines. Not one of the articles meeting selection criteria represented Class I or

Class II studies. All 27 provided Class III evidence regarding the diagnosis and/or management of os odontoideum. These 27 articles represent the basis for this review and are summarized in Evidentiary Table format.

SCIENTIFIC FOUNDATION

Diagnostic Evaluation

There is no literature that describes the sensitivity and specificity of imaging studies for os odontoideum. Dai et al, in their review of 44 patients with os used tomography, CT, and MR, in addition to “routine” plain cervical radiographs (AP, lateral, open-mouth, flexion and extension x-rays) in 39, 27, and 22 patients, respectively.(6) Matsui et al, described only the plain radiographs of 12 patients with os.(16) They excluded patients with Down’s 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, (27) and Spierings and Braakman (22) described the plain radiographs of 34, and 37 patients respectively with os, without reference to other imaging studies. Fielding et al, described 35 patients with os odontoideum in which “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.(10)

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

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 reported clinical series.

Instability of C1-2 in association with os odontoideum has been investigated with multiple imaging modalities. Employing flexion and extension lateral cervical spine x-ray studies in 33 patients, Fielding et al, reported 22 patients (67%) with anterior instability who had a mean atlanto-dens interval (ADI) of 10.3 mm, five patients (15%) with posterior instability (mean posterior translation of the os during extension of 8.4 mm), three patients (9%) had less than three mm of C1-2 motion, and three patients (9%) with no detectable C1-C2 motion.(10) Eight patients (23%) had both anterior and posterior instability. The authors noted that cineradiography was helpful in examining range of motion at C1-C2 in these patients, but it was not of benefit in the measurement of the degree instability. Of note is that almost one fifth of the patients in their series manifested no radiographic evidence of C1-2 instability.

Spierings and Braakman 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 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 two groups, those with and without myelopathy.(22) The degree of C1-C2 instability did not correlate with neurological status, but the measured minimal spinal canal diameter was significantly smaller ($P < 0.05$) in the group with myelopathy. They identified 13 millimeters as the critical anterior-posterior (AP) spinal

diameter. Watanabe et al, made similar measurements in 34 patients using plain lateral cervical radiographs in flexion and extension.(27) Like Spierings and Braakman, the degree of instability in their patients did not correlate with the presence of myelopathy. Shirasaki et al, described radiographic findings on lateral flexion and extension radiographs in nine patients with os odontoideum.(20) They reported that a distance of 13 millimeters or less between the os and the dorsal arch of C1 “specifically defined severe cervical myelopathy” in their patients. They too, found that the degree of C1-C2 instability did not correlate with the presence of myelopathy. Yamashita et al, studied atlantoaxial subluxation with plain radiography and MR, and correlated the imaging studies with the degree of myelopathy in 29 patients (four 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.(28) The degree of cord compression on MR did correlate well with the degree of myelopathy measured clinically. Matsui et al, classified os odontoideum into three types according to the shape of the os on plain radiographs.(16) Three types were described: round, cone, and blunt-tooth. They compared these three os types to the degree of clinical myelopathy and found the degree of myelopathy correlated most closely with the “round” os type. Kuhns et al, described the MR appearance of os odontoideum in four children and identified signal changes within the posterior ligaments consistent with trauma.(14) They could not discern whether these changes represented a primary or secondary phenomenon with respect to atlanto-axial instability.

These studies provide two consistent conclusions: 1) the degree of C1-C2 instability does not appear to correlate with neurological status in patients with os odontoideum; and 2) sagittal spinal canal diameter on plain radiographs of 13 millimeters or less is strongly associated with 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.(11,17,24,26) 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 (e.g. 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. MR is the best modality for viewing cord compression even after apparent C1-C2 realignment.(28) 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.(19) Hosono et al, made interesting observations on the different motions of the posterior arch of C1 in relation to C2 in patients with os odontoideum. They observed two patterns of motion, linear and sigmoid. They felt that in those patients with a sigmoid shaped motion pattern posterior wiring techniques may not provide adequate stability.(13) The selection of and necessity for additional imaging studies in the evaluation of os odontoideum appears to be made on a patient-by-patient basis. The literature provides no convincing evidence as to which patients should undergo supplemental imaging (tomography or MR) 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 C1-2 levels. The earliest reports of os odontoideum describe small pediatric case series treated surgically. In 1978, Griswold et al, described four children with os odontoideum that underwent posterior C1-2 wiring and autologous iliac fusion (12). Three children had successful arthrodesis. The fourth child did not achieve fusion/stability despite three attempts. In the same year, Brooks and Jenkins described their technique of C1-2 wiring and fusion and reported three children with os who were immobilized postoperatively in Minerva jackets.(3) All three patients achieved successful fusion. In summary, six of the seven children with os odontoideum described in these two reports were successfully treated.

Two larger series, reported in the early 1980's, included adults and children with os odontoideum, and described both operative and nonoperative management strategies for these patients. Fielding et al, described 35 patients with os odontoideum, of which 27 had radiographic evidence of instability.(10) Twenty-six of these 27 patients underwent successful posterior C1-C2 fusion (Gallie type). Fusions were noted to be "solid" after two months of immobilization in children and three months in adults. One patient with instability refused surgery and remained well at two years follow-up. The eight remaining patients with no evidence of C1-C2 instability managed non-operatively remained well at last follow up of one to three years. Spierings and Braakman described 37 patients they managed with os. Seventeen were treated surgically.(22) 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. Nine of these 16 patients had flexion and extension radiographs. Of these nine patients, seven had abnormal motion of eight millimeters or greater. With a median follow-up of seven years none of these 16 patients developed a neurological deficit. Four

additional patients who presented with myelopathy were treated non-operatively with follow-up from six months to 14 years. Three of these four patients presented with transient myelopathy and had no recurrence at last follow-up, despite abnormal motion of C1 on C2 of eight mm to 16 mm. The fourth patient had a stable monoparesis at last follow-up. Of the 17 patients who underwent surgery, one patient had neurological worsening and two died. Eight of these 17 patients treated surgically had a posterior C1-2 fusion. Nine patients underwent occipital-cervical fusion with C1 laminectomy. The authors did not report a single failed fusion. They had a combined surgical morbidity and mortality of 18% (three of 17 patients). The authors conclude that patients with os odontoideum without C1-C2 instability can be managed without surgical stabilization and fusion with good result. While they did not provide operative treatment to every os patient with C1-C2 instability, those with myelopathy and greater amounts of instability were more likely to be operated upon. If these two 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 non-operatively with little or no morbidity over time. While patients with os odontoideum and myelopathy or C1-C2 instability have been managed conservatively, most patients with myelopathy or instability are treated surgically.

Clements et al, in 1995 reported a patient who had a documented os without instability who at five years follow-up developed symptomatic frank C1-2 instability which required surgical stabilization and fusion.(4) It appears that a lack of C1-C2 instability at initial diagnosis does not guarantee that instability will not develop in these patients. It is recommended,

therefore, that clinical and radiographic follow-up be provided to patients with os odontoideum who are found to have radiographic C1-C2 stability on initial assessment.

More recent series reported in the literature provide better descriptions of the operative procedures and postoperative immobilization techniques employed for patients with os odontoideum.(2,5-9,15,17,21-26) Smith et al, described 11 children with os who underwent posterior wiring and attempted fusion.(21) Autologous bone graft and halo immobilization were used in all children. Two children had fusion failure with non-union. One child incurred an intraoperative cord injury secondary to sublaminar wire passage. Lowry et al, also described eleven children with os odontoideum that were treated with C1-2 fusion and posterior wiring.(15) One child treated with a Gallie-type procedure had continued instability and fusion failure. The C1-C2 construct was revised successfully with a Brooks-type fusion procedure. The remaining ten children were successfully treated with Brooks C1-C2 wiring and fusion procedures. Coyne et al, in a review of posterior C1-2 fixation techniques described five patients with os odontoideum.(5) Three of these five had unsuccessful attempted posterior fusions despite halo immobilization. Two developed new neurological deficits after surgery. Dai et al, described 44 patients with os odontoideum with a mean follow-up of 6.5 years.(6) Seven patients were asymptomatic at presentation. Five of these seven refused surgery and were treated with a cervical collar only, and remained stable at last follow-up. The remaining 39 patients underwent successful fusion procedures following skeletal traction. The authors reported that nine patients underwent atlantoaxial fusion and 33 required occipitocervical fusion (42 operations in 39 patients). Symptoms and signs disappeared in 26 of their operative patients and improved in the remaining 13 at last follow-up. They employed occipital-cervical constructs

with fusion with or without C1 laminectomy in those patients with irreducible deformities because of the concern that sublaminar passage of wires or cables might result in neurological morbidity.

Wang et al, reported 16 children with C1-2 instability of which four had os odontoideum.(26) These four children were treated with C1-2 transarticular screw fixation with posterior C1-C2 wiring and fusion. The youngest child was four years old. All achieved stable fusion arthrodesis without complications. Halo immobilization devices were not used. Brockmeyer et al, as well, reported 31 children they treated with C1-2 transarticular screw fixation and fusion.(2) Twelve of these children had os odontoideum. Bilateral screws were placed successfully without complication in all children with os odontoideum. They did not comment on the type of postoperative immobilization devices they employed. In 1991 Dickman and colleagues reviewed their experience with fusion plus twelve weeks of halo immobilization in the treatment of C1-C2 instability.(7) They described 36 patients with C1-2 instability, four of whom had os odontoideum. Three of four os patients they treated in this way developed osseous union. One had a stable fibrous union at last follow-up. In a subsequent report in 1998, Dickman et al, compared their series of patients undergoing C1-2 transarticular screw fixation with posterior wiring and fusion to those patients who were treated with posterior wiring and fusion alone. The fusion rates in the two groups were 98% and 86%, respectively.(8) No patient with os treated with C1-C2 transarticular fusion techniques failed to fuse. Only one of eight patients with os odontoideum in the posterior wiring and fusion group developed a nonunion (previously described). In contrast to the posterior wiring and fusion only patients, no patient treated with transarticular screw fixation required postoperative halo immobilization. Menezes and Ryken described four children with os odontoideum and Down's syndrome that they

successfully treated with posterior wiring and fusion, utilizing full thickness autograft rib, and at least three months of postoperative halo immobilization.(17) Dyck reported eight children with os odontoideum, six of whom were treated with posterior C1-3 wiring and fusion techniques.(9) All were externally immobilized in a four-poster brace “usually” for three to four months postoperatively. Two of six children required reoperation for nonunions.

Apfelbaum et al, described their experience in treating recent and remote (≥ 18 months after injury) odontoid injuries with anterior screw fixation.(1) 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 expected to be poor. Anterior C1-C2 trans-facet fixation techniques may have merit in the surgical treatment of os odontoideum, but there are no descriptions of its application for os odontoideum in the literature.

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 employed, 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. While the numbers are small, transarticular C1-2 screw fixation and fusion has been associated with higher rates of fusion compared to posterior wiring and fusion techniques alone. Of note is that patients treated with transarticular screw fixation have been managed in hard collars postoperatively obviating the need for halo immobilization devices. If transarticular screw fixation is not utilized 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.(24) Reports of the management of ventral compression and os odontoideum are scant. In a review of 36 patients with Down's syndrome and craniovertebral junction abnormalities, twelve patients were described with os odontoideum.(24) Eleven patients of the 36 reported were noted to have basilar invagination. Five of these eleven 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 five patients; however, the total number of patients who had basilar invagination due to os odontoideum was not described. The report implies however, that selected patients with atlantoaxial instability and irreducible symptomatic ventral cervicomedullary compression may benefit from ventral decompression. On the other hand, Dai and colleagues reported the successful use of occipital cervical fusion with or without C1 laminectomy in cases of 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.(6) While it may seem intuitive to remove ventral neural compression in association with os odontoideum, the literature suggests that 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 C1-2 instability. Tomography (computerized or plain)

may be helpful to define the osseous relationships at the skull base, C1 and C2 in patients where 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 C1-2 level of less than 13 millimeters does correlate with myelopathy detected on clinical examination. MR can depict spinal cord compression and signal changes within the cord that correlates 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 have no instability at C1-2 on flexion and extension studies can be managed without operative intervention. Even patients with documented C1-C2 instability and neurological deficit have been managed non-operatively without clinical consequence during finite follow-up periods. Most investigators of 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. While not supported by Class I or Class II evidence from the literature, multiple case series (Class III evidence) suggest that stabilization and fusion of C1-C2 is meritorious in this circumstance.(6,15,24,26) Because a patient with an initially stable os odontoideum has been reported to develop delayed C1-C2 instability, and because there are rare examples of patients with stable os odontoideum who have developed neurological deficits following minor trauma, longitudinal clinical and radiographic surveillance of patients with os odontoideum without instability is recommended. (4,10)

Posterior C1-2 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.(3,5,6,22,26) Atlantoaxial transarticular screw fixation and fusion appears to have merit in the treatment of C1-2 instability in association with os odontoideum, and appears to obviate the need for postoperative halo immobilization. Neural compression in association with os odontoideum has been treated with reduction of deformity, dorsal decompression of irreducible deformity, and ventral decompression of irreducible deformity, each in conjunction with C1-C2 or occipital cervical fusion and 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 C1-2 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 C1-2 transarticular screw fixation for patients with os odontoideum and C1-2 instability would help to definitely identify the risks and merits of each of the two procedures in this patient population.

EVIDENTIARY TABLES

First Author Reference	Description of Study	Data Class	Conclusions
Apfelbaum RI, et al, 2000 <i>J Neurosurg</i>	18 patients with odontoid fractures incurred \geq 18 months prior to treatment, who were treated with anterior odontoid screw fixation.	III	16 patients with follow-up. 25% fusion rate. Three with screw fracture and one with screw pull-out.
Brockmeyer DL et al, 2000 <i>J Neurosurg</i>	Review of transarticular screw placement in 31 children. 12 children with os odontoideum (ages four to 16 years).	III	55 of 62 possible sites deemed suitable for transarticular screws. All children with os odontoideum were able to have two screws placed.
Dai L et al, 2000 <i>Surg Neurol</i>	A review of 44 patients ages seven to 56 years with os odontoideum. Mean follow-up of 6.5 years.	III	7 patients were asymptomatic. five of these seven 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 DA et al, 2000 <i>J Neurosurg</i>	A review of craniovertebral junction abnormalities in 36 Down's Syndrome patients. Os odontoideum present in 12.	III	Twenty-seven underwent surgical procedures. Of 11 with basilar invagination, it was irreducible in five and transoral decompression was performed.
Dickman CA et al, 1998 <i>Neurosurgery</i>	Review of 121 patients treated with posterior C1-2 transarticular screws and wired posterior bone struts. Os odontoideum was present in 9. This group was compared to 74 patients treated with posterior wiring techniques alone.	III	2 failures in the transarticular group. The etiology of the C1-2 instability was not stated for these two failures. One of eight patients with os odontoideum in the posterior wiring group had a nonunion. Overall fusion rate for transarticular was 98% versus 86% for posterior wiring techniques.
Wang J, et al, 1999 <i>Pediatr Neurosurg</i>	16 children treated for atlanto-axial instability. Four of which had os odontoideum who were treated with C1-2 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-years
Kuhns LR, et al, 1998 <i>J Pediatr Ortho</i>	4 children with os odontoideum underwent MR examinations.	III	All four children had changes in the nuchal cord consistent with injury.
Lowry DW et al, 1997 <i>J Neurosurg</i>	A review of 25 children requiring upper cervical fusions. 11 children had os odontoideum.	III	10 underwent a Brooks type C1-2 fusion. two of these children did not fuse. one underwent a Gallie type fusion. This child remained unstable and was revised to a Brooks type fusion which was successful.
Matsui H, et al, 1997 <i>Spine</i>	Review of the plain radiographic morphology of C2 was evaluated in 12 patients (15 to 71 years-old) with os odontoideum unrelated to any syndrome.	III	Three configurations described from an anteroposterior view: Round, cone, and blunt-tooth. Myelopathy was more severe in the group with a round configuration.
Verska JM 1997 <i>Spine</i>	Report of a pair of identical twins, one with os odontoideum, and one without an os odontoideum	III	History of trauma in the twin with an os odontoideum. Fell at age three years, had torticollis and neck pain for several months.

First Author Reference	Description of Study	Data Class	Conclusions
Watanabe M et al, 1996 <i>Spine</i>	Review of 34 patients with os odontoideum (5 to 76 years-old). Divided into groups by Rowland Classification (1=local symptoms, two =post-traumatic transient myelopathy, 3,4=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 degrees or instability index of > 40% correlates with myelopathy.
Clements WD et al, 1995 <i>Injury</i>	Report of nonoperative treatment of an incidentally discovered os odontoideum without C1-2 instability at diagnosis	III	After five years profound C1-2 instability and symptoms had developed necessitating posterior instrumentation and fusion.
Coyne TJ et al, 1995 <i>Neurosurg</i>	Review of posterior C1-2 fusion and instrumentation techniques. Five of 32 patients had os odontoideum.	III	3 of five with os odontoideum failed with posterior wiring techniques. All were immobilized in halos. two of five developed new neurological deficits as operative complications.
Stevens JM et al, 1994 <i>Brain</i>	Review of abnormal odontoids and C1-2 instability. 24 of 62 patients with os odontoideum. nine children and 15 adults.	III	Periodontoid soft tissue thickening was only present in those with Morquio's disease. Following fusion the odontoid was noted to partially or completely regenerate in cases of Morquio's disease.
Menezes AH et al, 1992 <i>Neurosurg</i>	Review of 18 Down's syndrome patients with symptomatic cervicomedullary compromise. Four had os odontoideum.	III	All four had gross instability on dynamic radiographs. Successful fusion with posterior wiring techniques and full thickness rib grafts. Immobilized for a "minimum of three months".
Dickman CA et al, 1991 <i>J Neurosurg</i>	Review of 36 patients treated with C1-2 posterior wiring and fusion for various reasons. Four patients had os odontoideum (ages 16,25,38,43). All patients were maintained in a halo for 12 weeks after surgery.	III	Of the four with os odontoideum, three developed osseous unions and one had a stable fibrous union (follow-up of 15 to 44 months). No complications for these four patients.
Hosono N et al, 1991 <i>Spine</i>	Cineradiographic evaluation of six 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 MD et al, 1991 <i>Spine</i>	Review of 17 children operated on for C1-2 instability. 11 had os odontoideum. Posterior wiring techniques, autologous bone, and halo used in all.	III	2 of the 11 with os odontoideum had non-unions. One cord injury thought secondary to sublaminar wire passage.

First Author Reference	Description of Study	Data Class	Conclusions
Shirasaki N et al, 1991 <i>Spine</i>	9 patients with os odontoideum and posterior instability had three radiographic parameters measured. Distance between the os and C2 spinous process in extension (Dext), distance between the os and posterior C1 arch (Dat1), and “degree of instability” (Inst). These findings were compared to their neurological status.	III	Those without history or evidence of myelopathy had a Dext of > 16 mm. Dext was ≤ 16 mm in those with myelopathy. The presence or absence of myelopathy was not related to the Inst. In those with myelopathy and a Dat1 > 13 mm there was reversible cord compression in extension, in those with a Dat1 of ≤ 13 mm the cord remained compressed in flexion and extension.
Morgan MK et al, 1989 <i>J Neurosurg</i>	Report of three family members with C2-3 Klippel-Feil abnormalities and os odontoideum	III	Ages 16 (index case), 39 (father), and 64 (paternal grandmother). None with neurological signs or symptoms.
Yamashita Y et al, 1989 <i>Acta Radiologica</i>	Correlation of clinical status, MR, and radiographs in 29 patients with C1-2 instability. four had os odontoideum.	III	The atlanto-dens interval did not correlate with the degree of myelopathy but MR degree of cord compression did correlate with degree of myelopathy.
French HG, et al, 1987 <i>J Pediatr Ortho</i>	Review of dynamic cervical spine radiographs in 185 patients with Down’s Syndrome	III	Six had abnormal odontoids consistent with os odontoideum for an incidence of 3%. Three had prior radiographs showing no abnormality. One had an exaggerated ADI of six mm.
Spierings EL et al 1982 <i>J Bone Joint Surg (Br)</i>	37 patients with os odontoideum. 20 treated conservatively.	III	Of 20 managed conservatively, one was lost to follow-up. 15 had no myelopathy (median f/u of five years) and none developed myelopathy. Of four with myelopathy (f/u of 0.5, 1, 7, and 14 years) one is dead from cancer, one has neck pain, one has neck pain and paresthesias, and one has headaches.
Fielding HG et al, 1980 <i>J Bone Joint Surg (Am)</i>	35 patients (3 to 65 years-old) with os odontoideum. 25 patients were symptomatic.	III	22 patients had anterior instability with a mean ADI of 10.3 mm. five had posterior instability. three had no detectable motion. three had less than three mm of C1-2 motion. 26 underwent posterior fusion successfully. Five were not operated, three were asymptomatic with no instability. They remained well with no instability at 1,2, &3 years, respectively. One patient with instability refused surgery but was well at two years follow-up. One patient died of renal failure.
Brooks AL et al, 1978 <i>J Bone Joint Surg (Am)</i>	3 children (8,11,12 years-old) with os odontoideum treated with sublaminar C1-2 wires and autologous iliac crest graft. Minerva cast immobilization	III	All fused. Spontaneous extension of fusion to C3 in one child.

First Author Reference	Description of Study	Data Class	Conclusions
Dyck P, 1978 <i>Neurosurg</i>	Review of eight children (ages seven to 17 years) with os odontoideum. six were treated with posterior wiring and fusion of C1-3. External immobilization for “usually” three to four months.	III	6 children underwent posterior fusion by the author. Two required reoperation.
Griswold DM et al, 1978 <i>J Bone Joint Surg (Am)</i>	4 patients with os odontoideum treated with sublaminar C1-2 wires and autologous iliac crest.	III	3 fused. One did not fuse after three attempts.

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