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Pathogenesis, dynamics, and management of os odontoideum

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Os odontoideum is an uncommon craniovertebral junction (CVJ) abnormality that exists as a separate ossicle apart from a hypoplastic dens. It usually moves with the clivus or the anterior arch of C-1 (dystopic) or rarely with the dens (orthotopic). Its genesis and natural history have been debated, and its proper treatment remains uncertain.

Two hundred and sixty patients, with symptomatic os odontoideum, were evaluated by the author over a 20-year period; the author performed surgery in 134 of these patients. In a prospective study the author evaluated the early childhood history of trauma, the dynamic studies of motion, and the effects of traction by using pleuridirectional tomography, computerized tomography (CT), CT myelography and magnetic resonance (MR) imaging. Operative findings were documented.

Early childhood trauma to the CVJ was recorded in 74 patients, in 30 of whom normal odontoid processes were documented at initial examination prior to the patient reaching age 4 years.

Acute neurological deterioration following trauma occurred in 63 of 134 patients. Symptoms were insidious in 71 of 134 patients. In six patients, who presented with acute neurological deterioration after trauma and in whom an abnormal spinal cord signal in the cervicomedullary junction was demonstrated on MR imaging, normal CVJ motion dynamics were shown. Motion dynamics varied and were unique to each patient. Irreducible ventral CVJ disease causing cervicomedullary compromise occurred in 28 patients in whom a transpharyngeal ventral decompressive procedure was necessitated. During the transoral operation, the transverse portion of the cruciate ligament was found to be located anterior to the axis body. All patients required dorsal CVJ arthrodesis, which, in 46, was limited to the C1-2 segment. Instability at the C1-2 joints was always multidirectional, as demonstrated on preoperative neuroimaging studies as well as at operation.

Sixteen patients presented after completed primary C1-2 dorsal fusion and with worsening deficits. They improved when the range of the fusion was extended to the occiput or if the ventrally located lesion was excised.

Os odontoideum is associated with early childhood trauma and is an acquired phenomenon. The presence of abnormal motion dynamics necessitates surgical intervention as do associated neurological deficits. Asymptomatic patients in whom os odontoideum is incidentally discovered and in whom no abnormal

motion dynamics are demonstrated should be followed closely.

Key Words * craniovertebral junction * os odontoideum * spinal cord compression * spinal fusion

A wide spectrum of congenital, developmental, and acquired abnormalities arise at the craniocervical junction because of the complex developmental anatomy, the mobile junction of the upper cervical spine and skull, and the complex transition between the spinal cord and the brainstem. These bony abnormalities involve not only osseous structures but the encompassed nervous system. The resulting compression and distortion of neural structures, as well as the vertebrobasilar vascular tree, lead to a constellation of symptoms and signs that often complicates the diagnosis of these disorders.[12,17,21,24,34,47] The term "os odontoideum" was coined by Giacomini in 1886,[19] and it refers to an independent bone observed cranial to the axis, in the place of the dens. Not an isolated dens, it exists apart from a hypoplastic dens. The os odontoideum, separated by a variable gap from a small odontoid process, is usually located in the position of the odontoid tip or near the basiocciput in the area of the foramen magnum where it may fuse with the clivus.[15,21,64] Os odontoideum is an infrequently occurring odontoid abnormality, and its pathogenesis has been variously explained on an embryological, traumatic, or vascular basis. In the past surgical intervention involved performing dorsal atlantoaxial arthrodesis in patients with neurological deficit, whereas those patients who were supposedly asymptomatic were treated expectantly because we lacked information on the natural history of this phenomenon.[36] At its worst, os odontoideum has serious implications regarding compression of the cervicomedullary junction (CMJ). Patients in the intermediate stages of spinal compression present with a variety of neurological syndromes referable to the cervicomedullary region. The questions that have been posed pertain to the origin of the disease, its occurrence in the general population, and whether all cases require surgical intervention.[3,8,9,11,14,26,28,30,39,41,48,57,63]

A prospective investigation of the presentation, radiological characterization, the biomechanics, and the treatment regimens of os odontoideum was undertaken in an effort to answer these questions.

CLINICAL MATERIALS AND METHODS

Two hundred and sixty patients, in whom symptoms of os odontoideum were demonstrated, were evaluated by the author at the University of Iowa Hospitals and Clinics between 1978 and 1998. Surgery was performed in 134 patients at the author's institution, and formed the basis of this prospective study. Of the 134 patients who ranged in age from 4 to 58 years, 102 were less than age of 16 years. There were 74 male and 60 female patients. A careful investigation of early odontoid or cervical trauma was undertaken, and the records and diagnostic studies obtained at the initial early evaluations were reviewed.[36] All patients underwent plain cervical radiography in the flexed, extended and lateral bending positions. This was supplemented with pleuridirectional dynamic tomography, computerized tomography (CT), CT myelography, vertebral angiography, and documentation of the effects of cervical traction.[38,45,62] Since 1984, myelography and pleuridirectional tomography have been replaced by CT scanning and magnetic resonance (MR) imaging.[56] Dynamic studies, including MR imaging, were always performed with the patient in the flexed and extended positions. Three-dimensional CT scanning of the craniocervical region has been performed in all patients since 1990. All patients were symptomatic at the time of examination and underwent surgical therapy according to the "reducibility" and the direction of encroachment previously described.[37,38]

The records of patient presentation, neurological status, neurodiagnostic investigations, and findings at

operation were stored in a database.

RESULTS

Radiological Findings

The os odontoideum was demonstrated to have a rounded, smooth cortical border that was separated by a variable gap from a small odontoid process, with a definite neurocentral synchondrosis always present in children. It was usually situated in the position of the normal odontoid tip at the base of the occiput in the region of the foramen magnum. The anterior arch of the atlas was usually hypertrophied. The ossicle was present in an "orthotopic position" in the place of the normal dens, and it moved with the axis and the atlas in six of the 134 patients. The ossicle was considered to be "dystopic" when it was attached to the clivus or moved in conjunction with the clivus and the anterior arch of C-1.[15] In 22 patients, it was partially fused to the clivus, and in the remaining 106 patients it moved in conjunction with the clivus and the anterior arch of the atlas. In 10 patients the os odontoideum was within the foramen magnum, behind the clivus, and the superior aspect of the axis body was above the plane of the foramen magnum (Fig. 1). In all patients, a small odontoid process was present separate from the ossicle. The gap between the os odontoideum and the remainder of the dens or axis body was always observed to be above the level of the superior facets of the axis vertebrae.

Fig. 1. Composite of lateral midline pleuridirectional tomography scans of the craniocervical junction (left) and frontal tomography scan obtained through the plane of the os odontoideum (right). This 18-year-old patient presented with quadriplegia, partial vocal cord paralysis, and difficulty with tongue motion. The anterior atlantal arch is hypertrophied. There is basilar invagination by the os odontoideum as well as the axis body.

Biomechanical Study

A lesion was considered to be "reducible" when traction or change in head position allowed for realignment so as to relieve compression of the CMJ.[38] This was possible in 106 of the 134 patients. An "irreducible" lesion was present both dorsally and ventrally in 28 patients.

In the neutral position, the os odontoideum compressed the ventral CMJ in 16 patients, and the axis body compressed the CMJ in 38 patients. Dystrophic spinal canal compromise occurred in 86 patients in the flexed-neck position, when the ossicle was displaced into the ventral aspect of the spinal canal and there the posterior atlantal arch caused posterior compression. While in the extended-neck position in this group of 86 patients, the posterior atlantal arch moved into a more caudal and ventral location while as the ossicle ventral compression was reduced. In these 86 patients, measurement of the motion of the atlas anterior arch (C-1) was more than 9 mm in the sagittal plane between flexion and extension positions. In

24 patients, spinal canal compromise worsened with the neck in the extended position; with the skull base and the dystopic os moving into the ventral spinal canal in the extended position. Thus, the motion dynamics varied with flexion and extension in each patient. Of 10 patients with extreme basilar invagination and with the axis body already within the foramen magnum, the application of skeletal traction only partially reduced the cranial settling in three patients.[36]

Coronal plane, lateral displacement was demonstrated with stress views on plain cervical radiographs as well as on CT scans in 26 individuals. This displacement was observed primarily at the atlantoaxial articulation. Of the 106 patients in whom lesions were considered to be reducible, there were 16 in whom worsening was demonstrated in extension due to a bifid anterior and posterior arch of C-1. Ten of these patients suffered from Down's syndrome, and in two Morquio's syndrome was present. We investigated retrospectively the following aspects: the minimum distance between the posterior border of the C-2 body and the posterior arch of the atlas (minimum sagittal diameter);[38,57] the difference between the atlantoaxial angle in flexion and extension (sagittal plane rotation angle); and the change of space for the spinal cord between the flexion and extension position (instability index).[1,65] All were found to be variable and noncontributory. Diagnostic parameters have previously been described.[37,56]

In this prospective study in which the history of cervical trauma in early childhood was examined, 38 patients were found. Radiographs of the neck demonstrated normal findings in 30 of the 38 children. Traumatic injury in childhood consisting of falls and motor vehicle accidents, as well as neck pain, was recorded in all. In addition, cervical traction for the treatment of neck injury or torticollis was noted to have been applied before the age of 4 years in 15 of these cases. In the children who experienced episodic torticollis, repeated nasopharyngeal infections were documented. In four of these children spondyloepiphyseal dysplasia was documented. A previous history of "neck problems in childhood" was documented in 36 other patients. In the children in whom initial cervical spine radiographs demonstrated a normal-appearing odontoid process, an os odontoideum was subsequently detected.

PRESENTING CHARACTERISTICS

Acute trauma precipitated neurological deterioration in 63 cases. The range of injuries included those sustained from football, gymnastics, removal of dental orthodontic appliances, and motor vehicle accidents. Wrestling injuries were particularly common in young children. Of 71 patients who presented with an insidious onset of symptoms, 14 were recognized on plain cervical spine radiographs at the time of minor injury. Six ventilator-dependent patients presented after undergoing a posterior fossa decompressive procedure and upper cervical spine laminectomy. Sixteen other patients had undergone previous failed atlantoaxial arthrodesis in which wire and bone construct was placed, and two patients experienced failure of transarticular screw fixation, which caused neurological worsening. In 12 of these patients with dystopic os odontoideum who had undergone a previous completed C1-2 dorsal arthrodesis, worsening of neurological symptoms occurred once the cervical immobilization was discontinued after a completed osseous fusion at the atlantoaxial joint. These patients recovered when the fusion site was extended to the occiput. In six other patients who had previously undergone a successful C1-2 dorsal arthrodesis, worsening occurred several years later after they sustained minor injuries; neuroimaging detected a ventral mass located behind the anterior arch of C-1. These patients recovered after undergoing a ventral decompression of the CMJ.

In six patients who experienced acute neurological deterioration following a motor vehicle accident (three patients) or wrestling activity (three patients) an abnormal signal was demonstrated on MR

imaging of within the spinal cord. In these six patients little or no motion of the CMJ was demonstrated on flexion or extension radiographs, or on MR images, in either immediate or delayed fashion.

The neurological symptoms consisted of neck pain, as well as headache in 86%. Limb weakness, ataxia, swallowing difficulties, and scoliosis were the major presenting symptoms. A significant number of patients presented with multiple symptoms.[34] There was evidence of limited neck motion in most patients who were experiencing neurological deficits. High cervical myelopathy was detected in 68 cases. Coarse vertical nystagmus as well as downbeat nystagmus was observed in six cases. In all 10 patients in whom extreme basilar invagination was present, vagal as well as hypoglossal nerve palsies occurred.

Operative Findings

In 28 patients in whom a transoral decompressive surgery was performed at the ventral CMJ removal of the os odontoideum as well as the superior aspect of the axis body were achieved. The apical as well as alar ligaments were still attached to the os odontoideum. The cruciate ligament was invariably in front of the anterior aspect of the axis body, and thus, the lesion was thought to be irreducible. In 10 cases the os odontoideum had migrated significantly upward into the foramen magnum, necessitating clival removal. Intense granulation tissue behind the ossicle was present in all 28 patients in whom an anterior decompressive procedure was required.

Of the 106 patients who underwent primary dorsal craniocervical arthrodesis, in 60 patients an occipitocervical fusion was required because of instability at the occipitoatlantal and atlantoaxial articulations. The instability occurred mainly at the atlantoaxial joint in 46 other cases. All patients were tested for instability in the vertical, sagittal, and lateral planes, despite the fact that they were in dynamic cervical traction and no paralyzing agent was being used for the anesthetic.

Surgical Procedure

All patients undergoing surgical treatment were placed in halo-vest cervical traction. Irreducible ventral disease was revealed in 28 patients who underwent both an anterior transoral decompressive as well as a dorsal occipitocervical fixation procedure. Of the remaining 106 patients, 40 underwent a C1-2 fusion procedure with bilateral interlaminar fixation in which titanium cable and rib graft were used. In eight patients a transarticular screw fixation construct was placed between C-2 and C-1. Dorsal occipitocervical fixation was the primary procedure in 60 individuals. Postoperative halo-vest immobilization was maintained for 3 months in patients in whom atlantoaxial arthrodesis had been performed and, on an average, 5 months in patients who underwent dorsal occipitocervical fixation. In 36 patients whom dorsal occipitocervical titanium loop instrumentation with rib graft was used, halo-vest immobilization was maintained for 4 months only. Patients who had undergone a transarticular screw fixation as well as the dorsal arthrodesis, were placed in an extended Philadelphia collar so that the occiput could be incorporated and used for immobilization. Plain cervical radiographs were obtained at 6-week intervals until fusion was complete. To obtain further documentation, CT scanning was used to identify the osseous integration. Dynamic flexion-extension radiographs were also obtained once fusion was believed to be complete. Postoperative MR imaging was performed in 16 patients in whom complex ventral compression was demonstrated and in whom a transoral procedure was performed. Intraoperative spinal cord monitoring was not reliable.[2,34]

ILLUSTRATIVE CASES

Case 1

This 16-year-old girl presented to an emergency facility at age 4 years following a motor vehicle accident. She complained of neck pain and limb paresthesias, and she experienced generalized hyperreflexia. Cervical spine radiographs demonstrated a dystopic os odontoideum with abnormal craniocervical motion (Fig. 2 left). She underwent an attempt at dorsal atlantoaxial arthrodesis in which a modified Gallie technique was utilized. She was subsequently referred to our facility for persistent C1-2 motion and persistent neurological symptoms. In investigating a possible source of early childhood trauma, we found an emergency-facility evaluation, performed after a motor vehicle accident at age 4, which cited the patient's severe neck pain with a diagnosis of C1-2 ligamentous injury. Figure 2 right demonstrates the presence of the intact odontoid. She underwent successful dorsal atlantoaxial fusion.

Fig. 2. Case 1. Left: Lateral cervical radiograph obtained at age 14 years revealing a dystopic os odontoideum with a narrow posterior atlantal arch and an increased predental space. Note the loss of cervical lordosis. Right: Lateral cervical radiographs obtained at age 4 years following motor vehicle accident. There is a widening of the interspinous distance between the atlas and the axis vertebrae. There is a partial exposure of the occipital condyles present. The odontoid process appears to be intact.

Case 2

This 15-year-old boy became acutely quadriplegic after sustaining an injury while wrestling. Cervical spine radiographs demonstrated a dystopic os odontoideum, with no motion detected on flexion and extension views. He recovered after 1 week of cervical collar therapy and was referred to our facility. At age 13 years, two years prior to the wrestling injury, he had experienced repeated episodes of neck pain, a stinging sensation between his shoulder blades, and episodic attacks of weakness in the upper and lower extremities that lasted a few minutes. Magnetic resonance imaging performed with the patient in the extended- and the flexed-neck position (Fig. 3 left and center) demonstrated no motion at the craniocervical junction. However, a high signal intensity was present on the T₂-weighted MR images obtained in the spinal cord and corresponding to the superior aspect of the axis body. He underwent

crown halo-vest cervical traction and a dorsal occipitocervical fusion. At surgery gross instability was demonstrated between the occiput and the atlas as well as between the atlas and the axis vertebrae. A threaded Steinmann pin was fashioned into an occipitocervical loop and anchored to the occiput as well as C-1 and C-2 laminae with stainless steel cables. The fusion construct was supplemented with autologous rib graft; at 4 months postoperatively, the construct was shown to have incorporated (Fig. 3 right).

Fig. 3. Case 2. Left and Center: Midsagittal T₂-weighted MR images obtained through the craniocervical junction in the flexed- (left) and extended-neck (center) positions. There is an os odontoideum present with patency of the subarachnoid space around the CMJ. High signal intensity within the spinal cord corresponds to the superior aspect of the axis body. There is little motion of the craniocervical junction. Right: Lateral cervical spine radiograph obtained 6 months postoperatively showing a dorsal occipitocervical fixation construct in which contoured stainless steel threaded Steinmann pin and autologous rib graft were used.

Case 3

This 45-year-old woman sustained a severe neck injury in a motor vehicle accident when she was 3 years of age; quadriparetic, she was hospitalized for 4 months and wore a plaster cast. She had been diagnosed as having a "neck fracture." She recovered full function. At age 16 years she underwent a C1-2 fusion procedure in which wiring and iliac crest bone graft were used to treat severe neck pain. She presented at age 45 years with right arm weakness, persistent occipital-frontal headaches, and urinary incontinence. On examination moderate cervical myelopathy was demonstrated. A plain lateral cervical radiograph demonstrated the dorsal atlantoaxial bone arthrodesis with suboptimum ventral reduction (Fig. 4 upper left). Three-dimensional CT with sagittal reconstruction and axial CT scanning through the superior aspect of the axis body revealed an hourglass constriction of the spinal canal between the anterior aspect of the posterior atlantal arch and the superior posterior aspect of the axis. A dystopic os odontoideum was also observed. Other findings included gross fixed atlantoaxial dislocation and a bony protuberance pointing dorsally from the superior aspect of the axis body (Fig. 4 upper right and center). Magnetic resonance imaging confirmed the cervicomedullary dorsal and ventral compression (Fig. 4 lower left). The patient underwent transpharyngeal ventral decompression of the CMJ, with removal of the os

odontoideum and the offending superior aspect of the axis. Postoperative T₂-weighted sagittal MR imaging confirmed that decompression had been achieved. The patient experienced marked improvement of neurological symptoms and resolution of the limb weakness (Fig. 4 lower right).

Fig. 4. Case 3. Imaging studies. Upper Left: Lateral cervical radiograph revealing a completed dorsal atlantoaxial arthrodesis in which bone and wire were used. The posterior atlantal arch appears to be displaced ventrally. The anterior arch of the atlas and the odontoid process is poorly visualized. Upper Center: Three-dimensional CT scan, with midsagittal reconstruction, revealing the presence of dystopic os odontoideum. The anterior atlantal arch is displaced forward and downward. There is fixed atlantoaxial dislocation. The posterior atlantal arch, and fusion mass around it, projects into the spinal canal just below foramen magnum. Upper Right: Axial CT scan obtained through the plane of the lateral atlantal masses. The dystopic os odontoideum can be seen attached to the anterior atlantal arch. The axis body has a bony protuberance that projects dorsally into the space available for the spinal canal near the midline. Lower Left: Midsagittal T₂-weighted MR image showing the craniocervical junction and upper cervical spine in which an hourglass constriction of the CMJ has developed with a ventral tethering and compression caused by the superior aspect of the axis body. Lower Right: Postoperative midsagittal T₂-weighted MR image obtained 1 week after ventral transoral decompressive surgery of the craniocervical junction that included the superior aspect of the axis body. Note the improved caliber of the subarachnoid sac.

Case 4

This 27-year-old man had fallen from a 30-foot-high barn at 4 years of age suffering severe neck pain that required immobilization in a cervical collar for 3 months. At age 17 he underwent a C1-2 decompressive procedure to treat neck pain as well as weakness in his upper extremities and difficulty with his gait. Postoperatively he became acutely quadriplegic. Over the next 2 years he was gradually able to walk; however it was with significant difficulty. At age 26 years he underwent a dorsal occipitocervical fusion procedure in which iliac crest bone graft was used. This procedure failed. Computerized tomography myelography with Iohexol demonstrated the posterior decompression at C-1 (Fig. 5 upper) with the os odontoideum displacing the cervical spinal cord dorsally and indenting it from a ventral aspect. During a ventral transoral operation (Fig. 5 lower) the os odontoideum was observed to be situated above the cruciate ligament. The transverse portion of the cruciate ligament was anterior to the axis body. The patient subsequently underwent a dorsal occipitocervical fusion procedure. Postoperatively he was able to walk. However, he made only moderate recovery from his spastic quadriparesis.

Fig. 5. Case 4. Upper: Axial CT scan obtained through the plane of the anterior arch of C-1. The subarachnoid space is outlined by Iohexol. The superior aspect of the axis body grossly indents into the ventral CMJ. A C-1 laminectomy had been previously performed. The dystopic os is fused to the anterior atlas arch. Lower: Intraoperative photograph obtained during the ventral transoral-transpharyngeal decompression of the craniocervical junction. The os odontoideum (Os) can be seen with the cruciate ligament (Cr) below it and in front of the axis body.

Case 5

This 27-year-old man became quadriparetic at age 14 years following a motor vehicle accident. An os odontoideum was detected, and the patient refused surgical treatment. He experienced significant upper- and lower-extremity weakness but was able to ambulate with a walker. Lateral cervical radiographs observed with the neck in the flexed and extended positions revealed an unstable dystopic os odontoideum (Fig. 6 upper). Flexion-extension T₂-weighted MR imaging revealed that the os odontoideum was in a posterior location, and images obtained with the neck in the flexed position showed it to be in the anterior arch of the atlas. In the extended position, the ventral extradural abnormality corrected itself; however, the posterior arch of the atlas indented into the spinal canal (Fig. 6 center). Three-dimensional CT, with sagittal and coronal reconstructions, demonstrated instability in the sagittal as well as the coronal plane that signified multidirectional instability (Fig. 6 lower left). He underwent cervical traction as well as a procedure in which dorsal transarticular C2-1 screws were placed into the lateral mass of C-1 bilaterally. There was gross atlantoaxial instability in all planes. The atlantooccipital articulation appeared to be firm. The construct was supplemented with bilateral interlaminar rib grafts. Postoperatively significant improvement in neurological status was shown (Fig. 6 lower right).

Fig. 6. Case 5. Imaging studies. Upper: Lateral cervical radiographs obtained in the extended- (left) and flexed-neck (right) positions. Note the dystopic os odontoideum with gross excursions in the sagittal plane of the atlas vertebrae. Center: Midsagittal T₂-weighted MR image of the craniovertebral junction the flexed- (left) and extended-neck (right) position. In the flexed position (left) the os odontoideum together with the anterior atlantal arch moves posteriorly into the ventral spinal canal, indenting the CMJ. This is corrected when the neck is in the extended position. However, in the extended position (at right) the posterior atlantal arch encroaches into the spinal canal. Lower Left: Three-dimensional CT scan showing the craniocervical junction and upper cervical spine in the neutral-neck position. There is a stub to the odontoid process present located above the superior articular facets of the axis vertebrae. There is gross atlantoaxial dislocation in the sagittal as well as in the lateral planes. Lower Right: Postoperative lateral radiograph of the craniocervical junction obtained 4 weeks after undergoing a transarticular C1-2 screw fixation procedure in which bilateral interlaminar rib graft fusion was performed. The craniocervical alignment is satisfactory.

DISCUSSION

The pathogenesis of os odontoideum and its treatment have elicited considerable discussion. Numerous classifications of os odontoideum have been proposed[9,21,26,61] as well as radiographic parameters for prognostication of neurological deficit and cervical myelopathy,[1,29,54,57,64] which have been based on small groups of patients and have not stood the test of time. The present prospective study includes 134 patients in whom the author performed surgery from an overall group of 260 patients who were evaluated for the disease. The results of this analysis appear to be provocative.

Origins of Os Odontoideum

In a series reported by McRae[33] there were 17 patients with os odontoideum, including two children. Together with several other authors, McRae believed that os odontoideum had embryological origin, but required the ongoing stresses of life to trigger instability and produce symptoms after childhood.[8,10,13,17,18,20,27,41,44,50,55,66] It is important to review embryological development and the blood supply to the axis vertebra.[15,37,49] The caudal-most occipital sclerotome or the proatlas divides into a ventral rostral segment and a corresponding dorsal caudal portion. The ventral component of the proatlas forms the anterior "U"-shaped margin of the foramen magnum as well as the occipital condyles. A portion of this ventral component of the proatlas detaches to incorporate with the pleuricentrum of the atlas vertebra that develops as a dens. The core of the proatlas centrum transforms into the apical ligament of the dens. The atlas and the check ligaments as well as the transverse ligament of the atlas derived from the proatlas as unossified tissue. The dens component of the odontoid process represents the atlas centrum.[8,21] At birth the odontoid process is separated from the body of the axis by

a cartilaginous band that represents a vestigial disc referred to as a neurocentral synchondrosis. This lies below the level of the superior articular facets of the axis and does not represent the anatomical base of the dens. The neurocentral synchondrosis is present in nearly all children by age 3 years and is absent in most by the age of 6 to 8 years.[15] At birth there should be a recognizable odontoid process that has not yet fused to the base of the axis. The tip of the odontoid process is not ossified and is represented by a separate ossification center usually observed in children by age 3 years and fuses with the body of the dens by age 12 years. When this may occasionally fail to fuse with the odontoid process, it is termed an *ossiculum terminale persistens* and has little clinical significance.

The vertebral as well as the carotid arteries provide the blood supply to the odontoid process. The vertebral arteries provide anterior and posterior ascending vessels that pass ventral and dorsal to the body of the axis and the odontoid, and they anastomose in an apical arcade in the region of the alar ligament. These vessels supply the small perforating branches of the body of the axis and the odontoid process.[37,49] In addition the anterior ascending arteries in the apical arcade receive blood from the carotid vessels by way of the the skull base and the alar ligament via occipital branches. Thus, the arrangement of the blood supply has an embryological basis.

According to those who maintain that the formation of the *os odontoideum* is embryologically based, it occurs because the dens and the axis body fail to fuse together. If this were the case, the gap between the *os odontoideum* and the axis should be located below the level of the superior axis facet. However, this has never been demonstrated. In addition, in all patients in whom an *os odontoideum* is present, a neurocentral synchondrosis is nearly always visible. Thus, the *os odontoideum* cannot be explained on an embryologic basis.

Trauma alone cannot explain the formation of an *os odontoideum*. An unhealed fracture with a sequestrum of the odontoid does not explain the hypertrophy of the anterior arch of the atlas nor the hypoplastic dens. Conversely, our experiences as well as those of several other authors have shown that an *os odontoideum* can form after bony or ligamentous injury to the atlas and the axis in early childhood, at which time a complete odontoid process can be demonstrated.[22,23,26,44,46,51,59] Riccardi, et al.,[46] have reported that Tredwell and O'Brien documented 13 cases of avascular necrosis of the cephalad portion of the odontoid process following prolonged halo-vest pelvic traction, in conjunction with spine fusion. They suggested that this was caused by ligament disruption, which impaired the blood supply to the odontoid process, and warned against "excessive distraction." Riccardi, et al., reported that "overpull" was evident in several roentgenograms obtained while a 15-month-old infant was placed in traction, with traumatic atlantoaxial dislocation; the child subsequently developed an *os odontoideum*. Fielding, and colleagues[15] have argued that, as the most common cause of *os odontoideum*, the evidence favored an unrecognized fracture in the region of the base of the odontoid process. Following the fracture injury in early childhood or acute ligamentous injury, a slight separation of the fracture fragments may occur. With time and contracture of the alar ligaments, a distraction force pulls the odontoid fragment away from the axis centrum and closer to the occiput. The blood supply to the odontoid process is easily traumatized and may contribute to poor healing or callus formation that now would retard closure of the gap. The blood supply to the ossicle is then supplied via the proximal arterial arcade, and this could account for hypertrophy of the anterior atlantal arch, which shares the same blood supply.[25,64] There is now increasing evidence that *os odontoideum* is more frequently associated with trauma and upper respiratory infections and, hence, is also found in conditions such as the Down's syndrome, Morquio's syndrome, and spondyloepiphyseal dysplasia[4,5,16,23,31,50,52,53,58-60] because these children are more prone to trauma in this region.

Radiological Parameters

Careful review of the previous case reports on symptomatic ossiculum terminale indicates that those lesions were actually os odontoideum.[8,13,21,38] Several radiographic indices have been proposed. Spierings and Braakman[57] have analyzed the radiographic features of the upper cervical spine in 21 patients with os odontoideum. They found that for the subarachnoid space at C-1 a sagittal diameter of less than 13 mm was associated with a high risk of permanent spinal cord damage. They thought that there was no correlation with the degree of instability. Abe,et al.,[1] in 1976 devised the instability index as an indicator of atlantoaxial instability; this is the change of rate of space available for the spinal cord between flexion and extension. The instability index is calculated by subtracting the maximum distance for the spinal cord between the body of C-2 and the anterior portion of the posterior atlantal arch, as well as the minimum distance on flexion-extension radiographs. This sum is divided by the maximum distance and multiplied by 100, and the final value is an indicator of anteroposterior instability. However, the instability with os odontoideum is multidirectional, as indicated by Watanabe, et al.[65] These investigators devised a sagittal plane rotation angle by which the change of difference of the atlantoaxial angle between the flexed and extended position can be documented. In a recent study of 34 patients with os odontoideum, they found that myelopathy occurred in 90% of patients in whom more than 40% of the instability index was documented.[65] In patients in whom more than 20° of sagittal plane rotation was present, myelopathy was observed in 86% of the cases. In 1983 Kobayashi, et al.,[29] discussed similar values. However, our own experience has shown that the excursions between flexion and extension, as documented on plain radiographs as well as MR images, do not actually reflect the true degree of instability that one observes in the patient after induction of general anesthesia or in cases in which the clamping muscle action of the cervical muscular prevents abnormal spinal motion. Thus, the indices do not truly reflect the extent of instability.

BIOMECHANICAL STUDY

Atlantooccipital ligamentous laxity was recognized in a significant number of patients with symptomatic os odontoideum in whom atlantoaxial instability was evident. This finding is significant for surgical management.[15] Patients who deteriorate following successful atlantoaxial arthrodesis may harbor occult atlantooccipital instability or persistence of ventral compressive disease at the craniocervical border, as indicated in our series. Multidirectional laxity at the atlantoaxial articulation signifies an defective cruciate ligament as well as the capsular ligaments. It is for this reason that transarticular C1-2 screw fixation would be an ideal treatment.[11,34,42]

The biomechanical processes at the craniocervical junction in patients with os odontoideum have already been elucidated earlier under the heading of results. It is important to keep in mind that extension alone is not sufficient to try to correct the alignment. In short, treatment should be tailored to the unique case each patient presents.

Treatment Options

The patients in our series were all symptomatic and showed evidence of atlantoaxial or occipitoatlantoaxial instability or extreme basilar invagination as a result of the os odontoideum. Should the lesion show reduction in size either with the patient in the flexed- or extended-neck position or when placed in cervical traction, then the need for stabilization is of paramount importance.[38] Conversely, if the size of the lesion cannot be reduced with these methods, then a decompressive procedure and subsequent fixation are essential. The decompressive surgery may need to be performed dorsally or

ventrally. In our experience, a dorsal fixation achieved the best results. Of 134 surgical procedures, there were two cases in which the fusion failed. One was in a patient with Down's syndrome and another in a patient who had undergone transoral decompression and dorsal occipitocervical fixation in which titanium loop instrumentation was used. This latter patient sustained a postoperative wound infection that necessitated a revision. Thus, the incidence of fusion success is approximately 98.5%. However, there is a high incidence of fusion failure if instrumentation has not been placed. In a report on the Toronto experience by Coyne, et al.,[7] there was a 75% incidence of fusion failure when the C1-2 wiring technique was used. Lowry, et al.,[32] have reported that fusion failed in three (27%) of 11 patients with os odontoideum. In an in vitro study of posterior atlantoaxial arthrodesis Naderi, et al.,[42] pointed out that it was mechanically advantageous to include as many fixation points as possible when treating gross atlantoaxial instability. Transarticular screws prevented better lateral bending as well as axial rotation than the use of posterior cable and graft placement technique. Should the occiput need to be incorporated, an occipitocervical titanium threaded custom-made loop has been used in this series.

The presence of the transverse ligament in front of the axis body makes alignment difficult, and thus adventurous odontoid screw fixation will not be feasible. In those patients with os odontoideum in whom the size of an atlantoaxial dislocation can be reduced, transarticular C1-2 screw placement would not be indicated. Based on the poor operative results intervention obtained by several authors they themselves have indicated that the best course of treatment for patients, in whom neurological deficit is present but instability is not, is watchful expectancy.[40,57] We disagree with this opinion, as do several other authors, having gained experience in treating this region.[6,11,12,15,24,35,43,59]

The natural history of os odontoideum is one in which minor trauma is commonly associated with the onset of symptoms that may vary from transient paretic episodes to severe myelopathy. This has been well documented, and hence stabilization should be offered 1) to patients with neurological deficits whether or not one can demonstrate instability; 2) to those in whom instability and a minimum spinal canal diameter of 13 to 14 mm are present; and 3) to those in whom grossly abnormal motion is demonstrated. In patients in whom an incidental os odontoideum is found, close follow-up review is made imperative.

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