A Review of the Diagnosis and Treatment of Atlantoaxial Dislocations

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Abstract

Study Design   Literature review.

Objective   Atlantoaxial dislocation (AAD) is a rare and potentially fatal disturbance to the normal occipital-cervical anatomy that affects some populations disproportionately, which may cause permanent neurologic deficits or sagittal deformity if not treated in a timely and appropriate manner. Currently, there is a lack of consensus among surgeons on the best approach to diagnose, characterize, and treat this condition. The objective of this review is to provide a comprehensive review of the literature to identify timely and effective diagnostic techniques and treatment modalities of AAD.

Methods   This review examined all articles published concerning "atlantoaxial dislocation" or "atlantoaxial subluxation" on the PubMed database. We included 112 articles published between 1966 and 2014.

Results   Results of these studies are summarized primarily as defining AAD, the normal anatomy, etiology of dislocation, clinical presentation, diagnostic techniques, classification, and recommendations for timely treatment modalities.

Conclusions   The Wang Classification System provides a practical means to diagnose and treat AAD. However, future research is required to identify the most salient intervention component or combination of components that lead to the best outcomes.

Keywords
► atlantoaxial dislocation
► atlantoaxial subluxation
► comprehensive review
► classification
► diagnosis
► treatment

Introduction

Atlantoaxial dislocation refers to a loss of stability between the atlas and axis (C1–C2), resulting in loss of normal articulation (►**Fig. 1**). The atlantoaxial joints can lose stable articulation from traumatic, inflammatory, idiopathic, or congenital abnormalities.[1] The mechanism of injury typically remains unidentified with several theories proposed in the literature.[2] Although it occurs in all age groups, atlantoaxial dislocation is most often seen in adolescents. Atlantoaxial dislocations have been previously studied extensively and reported in the literature with subsequent treatment recommendations published using older, less relevant classification systems. However, given recent advances in diagnostics and surgical techniques, this systematic review provides a concise summary of our current knowledge of atlantoaxial dislocation and offers treatment guidelines based on the most recent clinical evidence.

Anatomy of the Atlantoaxial Region

The atlantoaxial joint allows complex movements of the cervical spine while providing sufficient mechanical strength to stabilize the head. About 50% of total cervical spine flexion and extension
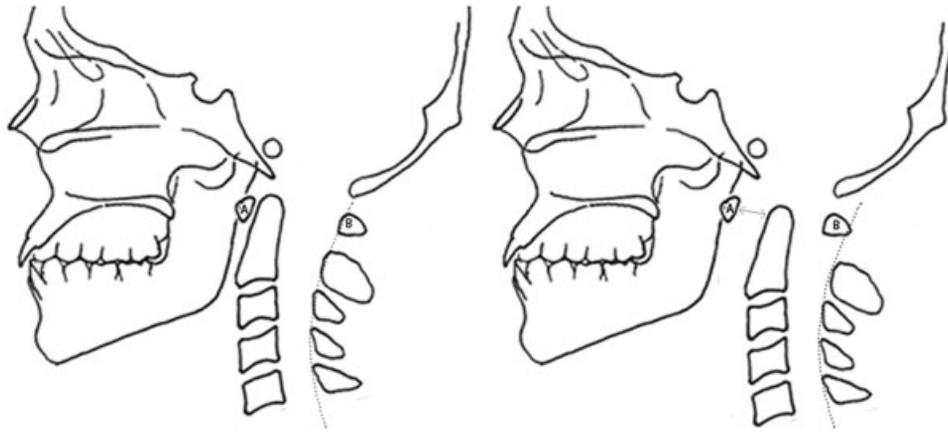
Fig. 1 Schematic representation of atlantoaxial dislocation. Lateral view of the normal cervical spine in relation to the occiput (left) compared with an abnormal relationship between the cervical spine and occiput representing an atlantoaxial dislocation (right). An increase in the distance between the anterior surface of the dens and the posterior surface of the C1 tubercle (A) as measured by the gray arrowed line is shown. The dotted line represents an imaginary line connecting the spinolaminar white lines (the junction between the lamina and the spinous process) and shows that the body of C1 (B) is displaced anteriorly relative to the cervical spine. The atlantodental interval (ADI) is measured between the posterior aspect of the anterior atlas ring and the anterior aspect of the odontoid process. The ADI is often constant in distance during movement of the head and generally does not exceed 3 mm for adults and 5 mm for children. Atlantoaxial dislocation is defined as ADI greater than 3 mm in adults older than 18 years of age and greater than 5 mm in children.

occurs at the occiput–C1 articulation.[3,4] Flexibility is provided by the dens (odontoid process) of C2 (the axis), which articulates with the C1 (atlas) and transverse ligaments, accounting for over 50% of all cervical spine rotation.[3,4] To provide this flexibility, the synovial joints at these segments do not have the same osseous and intervertebral disk-related stability mechanisms as the rest of the spinal column, relying more on ligamentous stabilizers.[5] The region is unique in that the occiput–C1 articulation and the C1–C2 articulation are the only vertebral segments without intervertebral disks, and the vertebral bodies do not directly bear the load distribution from the occiput. The occipital condyles transfer the load at the articulation with the C1 lateral masses, which transfers the load onto the C2 lateral masses.[4]

Stability against anterior translation is provided by the transverse ligament, which runs across the posterior dens, attaching on either side at the lateral masses of C1 and holding the dens in position. A small fascicular vertical portion of the transverse ligament articulates with the occiput superiorly and body of the axis inferiorly.[6] Additional stabilization of the dens comes from the alar ligaments, which extend off the odontoid in a lateral and cephalad direction to the basilar portion of the occiput. The transverse ligament is larger and stronger than the alar ligaments, thus providing a greater portion of the stability, with the alar ligaments providing secondary support.[6,7] Unlike in the lower cervical spine, the C1–C2 facets are oriented in the axial plane and have no bony structure between them preventing dislocation. This joint thus relies solely on the integrity of the transverse ligament to prevent anterior dislocation of C1 and the odontoid process abutting the anterior arch of the atlas to prevent posterior dislocation.

Etiology of Atlantoaxial Dislocation

Atlantoaxial dislocation can be broadly categorized into separate traumatic, congenital, or inflammatory etiologies, although the cause is commonly multifactorial.

Traumatic Causes

A purely traumatic atlantoaxial dislocation in the absence of another predisposing risk factor is extremely rare. A literature review by Venkatesan et al in 2012 found only 12 adult case reports.[8] Traumatic atlantoaxial dislocation is due to forced displacement of the neck resulting in disruption of the transverse ligament. Rarely, injury of the transverse ligament can also involve simultaneous disruption of the alar and apical ligaments. Such injuries may be seen in head injuries, which may occur during tackling in football or rugby. In these ligamentous dislocations, the atlas will lose articulation with the dens, and the anterior atlantal arch may translate completely superiorly and posteriorly with significant damage to the ligaments.[9]

Traumatic osseous injuries may also result in atlantoaxial instability. Type II odontoid fractures occur at the base between the transverse ligament and body of C2 body.[10–12] Type II fractures are the most common odontoid fracture and the only type normally associated with atlantoaxial dislocation.[10] Traditionally believed to be a congenital anomaly, os odontoideum may in fact be caused by an early traumatic injury in which the odontoid is separated completely from the axis and then heals to resemble a separate ossicle. The resulting condition predisposes patients to dislocation.[13,14]

Congenital Causes

Certain congenital conditions are associated with craniocervical region abnormalities that predispose these populations to developing atlantoaxial dislocation.[15] One particularly well-studied condition is Down syndrome. Down syndrome (trisomy 21) is the most common inherited chromosomal disorder.[16] Predisposing sequela include hypermobility and instability caused by ligamentous laxity and osseous abnormalities,[17,18] resulting in an increased incidence of atlantoaxial dislocation (15 to 20%).[19] Ligamentous laxity may be due to an inflammatory process or to an intrinsic defect in collagen

fibers that form ligaments.[20] Alternatively, chronic environmental trauma seen in these population superimposed on congenitally weakened ligaments can lead to spondylitis with subsequent atlantoaxial dislocation.[18]

Skeletal dysplasias are a heterogeneous group of disorders with resulting abnormal cartilage and bone formation, growth, and remodeling. Craniocervical junction abnormalities, atlantoaxial dislocation, and kyphoscoliotic deformities are among the common spinal problems that are found in certain skeletal dysplasias.[16] Examples of skeletal dysplasias that have been linked with atlantoaxial dislocation include Goldenhar syndrome, spondyloepiphyseal dysplasia, and Morquio syndrome.[16]

Goldenhar syndrome is a clinically heterogeneous disorder characterized by spinal defects, hemifacial microsomia, and epibulbar dermoid appendages. An increased frequency of hypoplasia of the dens with atlantoaxial instability has been reported in children with Goldenhar syndrome.[16] In one series of eight children with Goldenhar syndrome, three had atlantoaxial instability greater than 5 mm with upward migration of the odontoid process. Two of the patients had atlantoaxial instability greater than 7 mm and required surgical treatment.[21]

Spondyloepiphyseal dysplasia encompasses several disorders characterized by abnormal growth of the spinal vertebrae and epiphysis. Typically, individuals with spondyloepiphyseal dysplasia have short-trunk dwarfism, with short proximal and middle limbs but normal-sized hands and feet.[16] Atlantoaxial instability associated with hypoplasia of the dens or ligamentous laxity is the most common spinal manifestation of congenital spondyloepiphyseal dysplasia in children. An increased incidence, as high as 35% of cervical myelopathy in children with congenital spondyloepiphyseal dysplasia, may be attributable to atlantoaxial dislocation.[22]

Mucopolysaccharidosis type IV (Morquio syndrome) is an autosomal recessive lysosomal storage disease characterized by an inability to metabolize keratan sulfate. These patients often have normal growth and development during the first 2 years of life with abnormalities progressing rapidly after 2 years of life.[16] Atlantoaxial dislocation has been identified in up to 42 to 90% of cases of Morquio syndrome,[23,24] likely due to an increased incidence of odontoid dysplasia (hypoplasia, aplasia, or os odontoideum) and increased ligamental laxity.[23] Other syndromes that have been associated with the development of atlantoaxial dislocation are listed in ►Table 1.

Congenital osseous abnormalities, in the absence of an underlying genetic abnormality, are also frequently associated with atlantoaxial dislocation. Wang et al documented a rate of 18% among 904 patients in the largest published series to date.[25] Failures in segmentation, such as occipitalized atlas, C2–C3 fusion, and asymmetrical occiput–C3 facet joints, can predispose to dislocation.[14,26] Occipitalization of the atlas occurs when the inferior neural arch and superior spinal sclerotome form improperly. Patients with occipitalization of the atlas may also have asymmetrical occipitoatlantal facet joints because the same sclerotomal segments contribute to development of the facets.

Inflammatory Causes

Another notable population that is disproportionately affected by atlantoaxial dislocation is chronic rheumatoid arthritis patients, particularly adults. The atlantoaxial joint is frequently affected in rheumatoid arthritis with studies showing incidence rates ranging from 23 to 86% of patients with rheumatoid arthritis.[27–29] The rates we present here are based on traditional rates. Although it is not well documented, it is likely the rate has declined in conjunction with advancements in medical treatment of rheumatoid arthritis (i.e., biologics and immunoregulatory medications). The cervical spine often becomes involved early in the course of rheumatoid arthritis, leading to three different patterns of instability: atlantoaxial dislocation, atlantoaxial impaction, and subaxial subluxation.[30] Chronic systemic inflammation in these patients leads to chronic synovitis resulting in bony erosion and ligamentous laxity that may result in instability

Table 1 Congenital conditions associated with atlantoaxial dislocation

Congenital condition	Class	Etiology	Incidence
Down syndrome[16,17]	Chromosomal disorder	Inflammatory process or an intrinsic defect in collagen fibers that form ligaments[20]	15–20%[19]
Goldenhar syndrome	Skeletal dysplasia	Hypoplasia of the dens with atlantoaxial instability[16]	3 of 8 in one case series[21]
Spondyloepiphyseal dysplasia	Skeletal dysplasia	Abnormal growth of the spinal vertebrae and epiphysis,[16] hypoplasia of the dens or ligamentous laxity[22]	35%[22]
Morquio syndrome (mucopolysaccharidosis type IV)	Skeletal dysplasia	Autosomal recessive lysosomal storage disease, resulting in odontoid dysplasia	42–90%[23,24]
Occipitalization of the atlas	Congenital osseous abnormalities	Abnormal motility in the joint region[26]	–

and atlantoaxial dislocation. In patients with rheumatoid arthritis, anterior atlantoaxial dislocation is the most frequently occurring deformity due to laxity of the ligamentous restraints.[3,31,32]

Resulting Pathology

When the upper cervical spine is destabilized and sagittal balance is compromised, the lower cervical spine compensates, which may lead to subaxial pathology and deformities.[1,33–35] When atlantoaxial dislocation causes diminished lordosis at the C0–C2 segment, the subaxial cervical region compensates with increased lordosis to maintain balance.[33,35] Some patients with end-stage changes can develop kyphosis at the occipitoaxial segment together with extreme hyperlordosis subaxially, resulting in swan neck deformity.[33]

Clinical Presentation

The presentation of atlantoaxial dislocation may range from minor axial neck pain to death. Approximately 50% of patients present with neck pain and/or neck movement restriction, 70% with weakness and/or numbness, and 90% with pyramidal signs.[33,36,37] Other preoperative clinical presentations include sphincter disturbances, lower cranial nerve dysfunction, and respiratory distress. Other serious sequelae include myelopathy, respiratory failure, vertebral artery dissection, neurologic compromise, and rarely quadriplegia or death if left untreated.[38–40] The differential diagnosis of atlantoaxial dislocation includes torticollis, atlantoaxial rotary fixation, and odontoid fractures without atlantoaxial dislocation.

Most cases of atlantoaxial dislocation appear in the adolescent population and should be considered in a child with inability or unwillingness to turn their head when history and physical examination are inconsistent with torticollis.[41] Typically, congenital atlantoaxial dislocation presents in childhood as chronically progressive spinal canal compression and associated with neurologic and respiratory symptoms.[42] Less commonly, a patient with a congenital yet asymptomatic unstable atlantoaxial joint may have an acute presentation after minor acute trauma. In one study, 9 of 15 patients who presented after acute trauma with congenital atlantoaxial dislocation presented with quadriplegia.[41] The symptoms may also present as relapsing and remitting episodes due to the motion of day-to-day life.[26,38,42]

Atlantoaxial dislocation presenting in adulthood has historically been the result of rheumatoid arthritis, with some patients experiencing few symptoms and others experiencing gross instability and neurologic compromise. Clinical findings can be confounded by the severity of systemic involvement. Radiographic changes are common, and the prevalence of neurologic injury is relatively low.[30] More recent series have found a decreased prevalence of atlantoaxial dislocation among patients with rheumatoid arthritis, especially with various immunoregulatory medications showing promising outcomes preventing cervical spine lesions in patients with rheumatoid arthritis.[43,44] One study that investigated a total of 904 patients with a diagnosis of atlantoaxial dislocation over a period of 12 years showed that only 36 patients (4%) had rheumatoid arthritis.[25] Regardless of the etiology, the clinical consequences of atlantoaxial dislocation have clear potential for neurologic compromise. Therefore, a careful history is critical to identify symptoms of cervical disease, and physical and neurologic examinations should be performed in all at-risk patients.[3]

Diagnosis

There are several different approaches to diagnosing atlantoaxial dislocation; however, no consensus exists. Atlantoaxial dislocation can be defined with radiographic measurements of atlantoaxial joint articulation using the atlantodental interval (ADI). The ADI is a small slitlike space between the posterior aspect of the anterior atlas ring and the anterior aspect of the odontoid process. Flexion and extension radiographs of the neck allow for the measurement of the ADI and to determine whether the atlantoaxial joint reduces itself in these positions.[38]

The ADI is measured from a line projected superiorly along the anterior border to the axis body to the anterior arch of the atlas (►Figs. 1 and 2).[45] The ADI is normally constant in
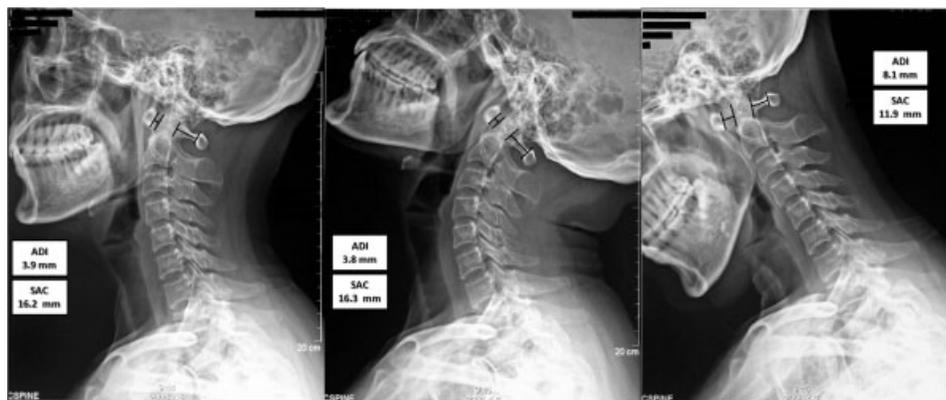

Fig. 2 Neutral (left), extension (center), and flexion (right) lateral X-rays showing the atlantodental interval (ADI) anterior to the odontoid process and the space available for spinal cord posteriorly. The ADI is above the average for adults of 3 mm and is slightly reduced in extension, but severely increased in flexion. This patient's space available for spinal cord (SAC) reducing to below 14 mm indicates risk of paralysis.

Table 2 Wang classification system of atlantoaxial dislocations

Type	Description	Diagnosis	Incidence (%)	Treatment
I	Instability	Reducible in dynamic X-rays	52.2	Posterior fusion procedure
II	Reducible	Reducible with skeletal traction under general anesthesia	17.7	Posterior fusion procedure
III	Irreducible	Irreducible with skeletal traction under general anesthesia	29.6	Transorally released anteriorly before posterior fusion
IV	Bony dislocations	Dislocations with bony anomalies that are visualized by reconstructive computed tomography scan	0.4	Transoral odontoidectomy

distance during movement of the head and generally does not exceed 3 mm for adults and 5 mm for children.[33,45,46] In this article, we define atlantoaxial dislocation as ADI greater than 3 mm in adults older than 18 years of age and greater than 5 mm in children. The majority (70%) of clinical atlantoaxial dislocation presentations are due to anterior dislocations.[41] Anterior dislocation increases the ADI, decreasing the space available for the spinal cord, which is measured from the posterior aspect of the dens to the anterior aspect of the posterior atlantal ring.[41] A decrease in the space available for the spinal cord increases the risk of spinal cord compression as well as neurologic sequelae. Of note, the space available for the spinal cord of less than 14 mm predicts the development of paralysis[47,48] and has been shown to correlate with severity of paralysis.[48]

Neutral and dynamic cervical radiographs are often used to diagnose atlantoaxial instability and dislocation, but the diagnostic sensitivity is low. Some studies report decreased false-negative diagnostic rate when using dynamic radiography, with some studies also reporting the necessity for magnetic resonance imaging (MRI) to obtain information on soft tissues, joints, and the spinal cord with high sensitivity and specificity and to possibly provide early warnings signs of instability.[41] When evaluating trauma patients with concerns about cervical spine involvement, it is important to most efficiently and effectively evaluate the cervical spine to avoid missing the injury. The NEXUS study group reported reliability of a standard three-view imaging of the cervical spine that included an open-mouth odontoid view, an anterior-posterior view, and a cross-table lateral view.[49] Another study supports the use of cervical computed tomography (CT) for diagnosis given the higher specificity.[50]

Classification of Atlantoaxial Dislocations

Atlantoaxial dislocation was initially classified by Greenberg into two subcategories—reducible and irreducible.[46] Greenberg further devised a treatment strategy based on this system. For irreducible atlantoaxial dislocation, Greenberg specifically stated that the treatment must be aimed at immediate decompression and achieving stabilization.[46] Greenberg's work has been considered a landmark publication and is considered by many to be the gold standard for atlantoaxial dislocation treatment in the subsequent litera-

ture.[51] Fielding and Hawkins subsequently developed a new classification system according to the direction of dislocation —anterior, posterior, lateral, and rotational.[52] This classification, known as the Fielding classification system, has been widely accepted for clinical application. Unfortunately, it was not found to have clinical significance in treatment or grading severity of injuries as the majority of clinical dislocations encountered were anterior.[45] Wang has recently proposed a novel classification system that aims to standardize atlantoaxial dislocation classification and treatment strategy.[51] Referred to as the "Wang classification system" in this article, it draws from Greenberg's system and is primarily based on classifying dislocations as reducible or irreducible atlantoaxial dislocation. According to this treatment algorithm, which includes preoperative evaluation using dynamic radiographs, reconstructive CT, and skeletal traction test, the Wang classification (►Table 2) categorizes atlantoaxial dislocation into four types: instability (type I), reducible dislocation (type II), irreducible dislocation (type III), and bony dislocations (type IV).[51] This novel classification system offers a unique diagnosis and treatment protocol for clinicians suspecting atlantoaxial dislocation in a patient.

Treatments

Treatment of atlantoaxial dislocation is aimed at the correction of the sagittal alignment of the upper cervical spine and the stabilization in near anatomical alignment.[45] There is no uniformly accepted method for determining treatment strategies, with extremely varying opinion on indications of nonoperative versus operative treatment modalities and on which operative techniques are more appropriate.[19,26,53–56] Some resources state that surgical treatment is recommended even in asymptomatic atlantoaxial dislocation to avoid development of myelopathy due to concerns about increased morbidity and mortality associated with atlantoaxial dislocation in patients presenting with myelopathy.[38–40,57] Recently, Wang et al have provided treatment recommendations based on the Wang classification system previously described.[25] Although not uniformly adopted, the classification system is unique in that it offers clear clinical and radiographic parameters for the clinician to reference.[51]

The main procedures reported in the literature are posterior occipitocervical/C1–C2 fusion after transoral odontoidectomy

or periodontoid tissue release. Fusion of the atlantoaxial joint greatly reduces the range of motion by preventing rotation of the occipitocervical region.[41] These procedures have many disadvantages, such as increasing hospitalization costs and length of stay, which may discount their effectiveness.[36] Surgical reduction and fixation of atlantoaxial dislocation is not without risk. One of the most serious complications is vertebral artery injury as it can have serious sequelae with poor prognosis such as exsanguination or catastrophic injury if not treated rapidly.[58] Both anterior and posterior approaches can pose a risk for vertebral artery injury, reported incidences ranging from 0 to 8.2% for posterior atlantoaxial transarticular screw fixation.[44] Dickman et al reported an overall complication rate of 9.4% during transoral surgery, even in experienced hands including cerebrospinal fluid leakage, wound dehiscence, wound infection, pneumonia, and death.[108]

Obtaining physiologic alignment is of particular importance. Normal upper cervical alignment differs between children, male adults, and female adults. Mean sagittal spinal angles according to group are provided in ►Table 3. Fusion of the atlantoaxial joint or occipitoaxial segment in hyperlordosis can place patients at high risk for postoperative iatrogenic subaxial loss of lordosis, or even kyphosis. According to a study by Matsunaga et al, a small number of patients (13%) with atlantoaxial dislocation developed postoperative kyphosis and swan neck deformity and more patients (32%) experienced subaxial subluxation after surgery.[34] High postoperative incidence of degenerative disk disease after fixation for atlantoaxial dislocation has also been reported in the setting of fusion in nonphysiologic lordosis.[35] Several authors have reported on the development of subaxial subluxation following occipitocervical fusion.[34,35] In such cases, degenerative disk disease was found near the apex of the subaxial sagittal curvature (C6–7) rather than at levels adjacent to the fusion.[33]

Nonoperative Treatment

Nonoperative treatment for atlantoaxial dislocation consists of cervical halter traction in the supine position and active range-of-motion exercises for 24 to 48 hours first, followed by ambulatory orthotic immobilization with active range-of-motion exercises until free motion returns.[59] Particular conditions and patient populations are recommended for nonoperative treatment. For example, children presenting acutely with evidence of transverse ligament disruption, diagnosed within 3 weeks, can be treated nonoperatively in the absence of neurologic injury.[60] The vast majority of patients with Grisel syndrome will improve spontaneously, and thus may be treated with halter traction if the atlantoaxial dislocation does not improve spontaneously. However, with persistent instability, stabilization with posterior arthrodesis is required.[21] Nonoperative treatment in symptomatic adults is generally not advised in the absence of surgical contraindications.

Certain patient populations predisposed to atlantoaxial dislocation also have recommendations for preventative treatment and screening as well. For example, it is recommended that patients with Down syndrome obtain cervical radiographs between 3 and 5 years of age and have their cervical canal width measured.[61] Controversy exists regarding optimal management in the subset of patients with Down syndrome with asymptomatic atlantoaxial dislocation. The Special Olympics mandated screening all potential participants with Down syndrome in 1983 for atlantoaxial dislocation and limiting the participation of those patients found to have radiographic evidence of instability.[16] Although supported by the Committee on Sports Medicine of the American Academy of Pediatrics, controversy about appropriate interventions in these asymptomatic patients still remains.[62] In fact, it is unclear if asymptomatic patients with Down syndrome with an ADI greater than 4.5 mm are at higher risk for neurologic sequelae.[61]

Management of patients with rheumatoid arthritis is aimed at preventing permanent neurologic injury while avoiding potentially dangerous and unnecessary surgery. Strategies include patient education, lifestyle modification, regular radiographic follow-up, and early surgical intervention when indicated. MRI is indicated when myelopathic symptoms are present or when plain radiographs show atlantoaxial dislocation with the space available for the spinal cord less than or equal to 14 mm, any degree of atlantoaxial impaction, or subaxial stenosis with a canal diameter less than or equal to 14 mm.[30] There is evidence that the incidence of upper cervical abnormalities is lessened by early, aggressive drug treatment, specifically administration of disease-modifying antirheumatic drugs in combination with newer immunologic mediators such as tumor necrosis factor-α blockers.[3,32] When administered before the onset of cartilage destruction, these drugs may actually prevent or significantly mitigate the development of changes in the cervical spine.[3] These patients generally have better outcomes when treated before the onset of myelopathy.[3,32] Not as well established, however, is whether immunoregulatory drugs have the potential to interrupt the onset of

Table 3 Mean sagittal spinal angles according to age group

	Oc–C2 (°)	C1–C2 (°)	C2–C7 (°)	C1–C7 (°)	C7 slope
Pediatric (average 8.8 y)[106]	−15.2 (±6.7)	−26.0 (±6.2)	−6.5 (±11.7)	−32.7 (±11.3)	21.3 (±6.9)
Pediatric (average 14.2 y)[106]	−18.3 (±6.1)	−30.3 (±6.0)	−0.7 (±11)	−30.5 (±10.1)	17.4 (±6.6)
Male adults[107]	14.5 (±8)	26.5 (±7)	16.2 (±12.9)	n/a	n/a
Female adults[107]	16 (±8.5)	28.9 (±6.7)	10.5 (±10.3)	n/a	n/a

ongoing degenerative changes. Radiographs of the cervical spine with lateral flexion-extension dynamic views should be obtained periodically and used to evaluate the cervical spine before elective surgery requiring general anesthesia. Advanced imaging, such as MRI, myelography, and CT, may be necessary to evaluate neuraxis.[3]

Patients with Goldenhar syndrome with an ADI less than 6 mm should get cervical flexion-extension films every 6 months and be advised against contact sports. Surgical treatment is recommended in any child with instability greater than 6 mm to reduce the possibility of catastrophic spinal cord impingement. Moreover, the relatively high frequency of cervical malformations in patients with Goldenhar syndrome may warrant extensive radiographic investigation and planning before surgery.[21] Likewise, not all patients with atlantoaxial dislocation secondary to Morquio syndrome have spinal cord compression or require surgery. Asymptomatic patients or patients with a less than 50% reduction in spinal cord diameter may not require surgical treatment. However, the preferred time for elective surgery is between 3 and 8 years of age when skeletal anomalies are well developed.[24]

Indications for Surgical Treatment

Surgical treatment for patients with symptomatic atlantoaxial dislocation is widely accepted to protect against potential respiratory failure, progressive neurologic symptoms, and death.[63] However, there is a lack of consensus on surgical indications for asymptomatic atlantoaxial dislocation as these patients are at risk for new-onset severe neurologic and respiratory compromise from a minor acute trauma.[42,64] Different suggestions exist in the literature for these asymptomatic patients depending on patient age and underlying diagnosis. For adults, surgical treatment can be considered if ADI is greater than 5 mm. Surgical fusion is indicated in children when one or more of the following is present: neurologic involvement, persistent anterior displacement with ADI greater than 4 mm,[65] deformity present for more than 3 months, or recurrence of deformity following 6 weeks of immobilization. In young adults, fusion is recommended when moderate displacement is seen in flexion and extension cervical radiographs or instability with or without pain is present.[51,60,66,67]

In patients with rheumatoid arthritis, surgery should be considered promptly in an asymptomatic patient with atlantoaxial dislocation for any of the following: chronic neck pain in the setting of radiographic instability that does not respond to nonnarcotic pain medication, any degree of atlantoaxial impaction or cord stenosis seen radiographically, the space available for the spinal cord less than or equal to 14 mm, atlantoaxial impaction represented by odontoid migration less than or equal to 5 mm rostral to McGregor line, sagittal canal diameter < 14 mm, or cervicomedullary angle < 135 degrees.[30] For patients with Down syndrome or os odontoideum, there is no clear surgical indication in the literature, but it is recommended to monitor with annual lateral and flexion/extension cervical radiograph due to the possibility of sudden change from spinal cord compression in this patient population.[68,69] Due to the paucity of universally accepted guidelines in the literature, more research on the relative risks of surgery versus neurologic deterioration due to an asymptomatic atlantoaxial dislocation is needed given the lack of patient-derived outcome data.[42,64]

Methods for Surgical Treatment

Attempt at Conversion of Irreducible to Reducible Atlantoaxial Dislocation via Traction

The first step in correcting atlantoaxial dislocation is attempting reduction of the dislocation. When the dislocation is not easily reduced by neck flexion or extension, different intraoperative or preoperative approaches can be utilized to attempt reduction via traction (►**Fig. 3**). Traction lengthens and relaxes muscles around the dislocation, thereby enabling settling into normal anatomical position. Some surgeons choose to curarize back and neck muscles to alleviate tension
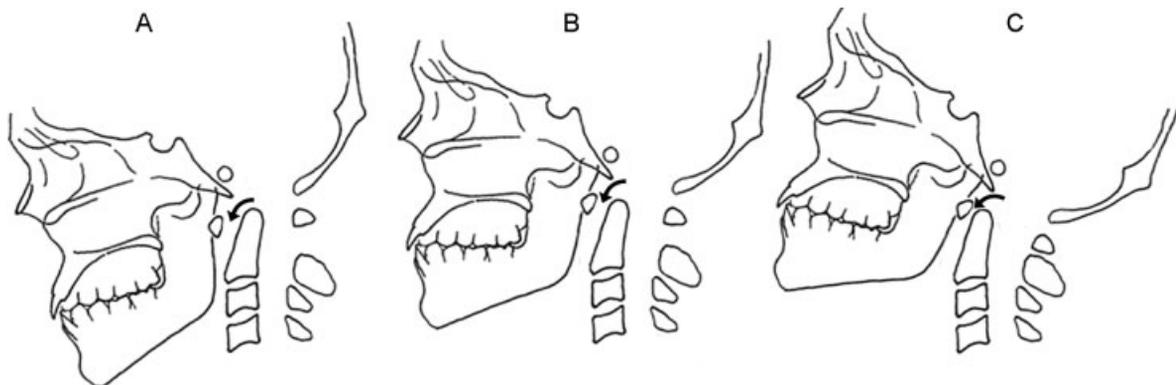

Fig. 3 A schematic representation of the traction technique involving three stages to reposition the joint. In the initial distraction phase (A), the patient is placed in slight flexion to keep the ring of C1 opposed to the posterior odontoid and to avoid hitting the spinal cord as traction weight is gradually added. After full distraction, the realignment phase (B) will occur when C1 slips back over the odontoid. Excessive flexion during distraction could cause the ring to slip too far forward. The release phase (C) consists of switching to an extension posture and slowly releasing traction over several hours with gradual weight reduction. Note the gradual realignment of the occiput to the cervical spine, as denoted by the decreasing atlantodental interval (black arrows).

and to briefly apply skeletal traction with the patient under general anesthesia for a rapid reduction before fixation in the same surgical session.[19,55] The entire reduction procedure with curarization takes ~10 minutes. Traction weight should start at 7 to 8% of body weight and gradually increase to a maximum of ~7 kg, and reduction should be monitored by sequential lateral radiographs.[26,64]

It is also possible to apply traction over a longer period preoperatively without general anesthesia and muscle curarization to slowly elongate muscles and reduce the joint.[19,53] Reductions without such relaxation techniques are less likely to be successful and have been shown to take longer traction durations even when successful.[9] Three stages of reduction have been described in complete C1–C2 dislocations. In the initial distraction phase, the patient is placed in slight flexion to keep the ring of C1 opposed to the posterior odontoid and to avoid hitting the spinal cord as traction weight is gradually added. After full distraction, the realignment phase will occur when C1 slips back over the odontoid. Excessive flexion during distraction could cause the ring to slip too far forward, so care must be taken through these stages to properly position C1 and C2. Finally, the release phase consists of switching to an extension posture and slowly releasing traction over several hours with gradual weight reduction.[9]

Traction may induce an increase in canal diameter due to the distraction of the odontoid from the foramen magnum.[70,71] Several studies report successful reduction of irreducible atlantoaxial dislocation up to 80% with traction,[71] with others reporting equally high failure rates.[15] If the reduction procedure is successful, defined by achieving an ADI measuring less than 3 mm in adults and 5 mm in children, the operation may proceed with a posterior fusion to stabilize the reduced reducible atlantoaxial dislocation. Reducible atlantoaxial dislocation requires posterior fusion alone while an irreducible atlantoaxial dislocation requires an anterior transoral decompression plus posterior fusion.[14,51,72] Posterior decompression and fusion alone are not sufficient in irreducible atlantoaxial dislocation as they may only achieve partial decompression while stabilizing the upper cervical spine in a suboptimal position. Assessment of the reducibility of the atlantoaxial dislocation is critical prior to proceeding with posterior fusion alone to optimize adequate decompression and restoration of physiologic alignment.[73]

Anterior Methods

An irreducible dislocation with persistent compression causing profound neurologic deficit after traction may necessitate an anterior approach.[51] Although it is possible that neurologic status can improve simply with a successful fusion, it is generally accepted that irreducible atlantoaxial dislocation must be either released or decompressed prior to fixation. Currently, the most accepted treatment for irreducible atlantoaxial dislocation is transoral odontoidectomy.[74] Most techniques that are approached anteriorly, such as transoral anterior release and transoral atlantoaxial reduction plate (TARP), have shown positive results (►Table 4).

In the transoral odontoidectomy, the apical and alar ligaments are detached with a curette before removing the

dens from top down.[74] More recent advances have made it possible to do a less invasive odontoidectomy endoscopically via a transnasal, transoral, or retropharyngeal approach. The endoscopic transoral odontoidectomy procedure obtains access through an incision in the midline pharyngeal wall, avoiding any palatal splitting or use of self-retaining retractors. The transoral endoscopic approach has yielded promising results with improvement in neurologic status after surgery.[75] However, cerebrospinal fluid leakage, incomplete decompression, infection, abscess formation, vertebral artery injury, and spinal cord injury have been reported during transoral decompression, and other more serious complications such as death from vertebral artery injury and respiratory distress should be considered and discussed with the patient.[1,55,74,76] Moreover, excessive bone removal of the odontoid and parts of the C2 body is a possible adverse effect as it compromises the C2 pedicle, which can disrupt posterior fixation.[55] Less invasive endoscopic approaches may become more popular as the techniques advance. An endoscopic expanded endonasal approach has been explored previously in a cadaveric study.[77,78] Similarly, an endoscopically assisted retropharyngeal approach with a small incision in the angle of the mandible has been performed with positive results.[79] All 21 patients undergoing the retropharyngeal approach in one prospective study had an uneventful recovery with significant improvement in neurologic function and radiographic parameters.[79]

The TARP technique eliminates the need for a two-step operation. This technique uses a reduction plate capable of anterior decompression, reduction, and fusion for anterior irreducible atlantoaxial dislocation in one stage.[80–82] In this procedure, incision of the posterior pharynx and exposure of C1 and C2 are completed similar to the other transoral approaches. The TARP is placed on the anterior aspect of C1, and the reduction screw is then inserted into the C2 body. One notch of the TARP system engages the TARP at the midline and the other engages the protruding reduction screw, causing distraction forces between C1 and C2, pushing C1 superiorly with the reduction screw serving as a base. The joint is then fused with bone graft.[80,81] This procedure is advantageous because it is able to reduce and provide adequate internal fixation and stabilization, making a posterior procedure unnecessary and reducing the duration of hospitalization and surgery as well total cost. In a 2- to 4-year follow-up study, adequate anatomical reduction was seen in 100% of patients with 73.3% having improved spinal cord function.[81]

Anterior transarticular screw fixation is yet another option to reduce fixed atlantoaxial dislocation. It provides a safer and more appropriate environment with high fusion rates and only minor complications for challenging cases such as patients with osteolysis in infectious or tumorous conditions, aberrant vertebral artery, and/or narrow pars interarticularis.[83] However, there are concerns about safe screw dimension, insertion, and trajectory. One study analyzed the feasibility of anterior transarticular screw fixation in 100 patients and recommended reliable fluoroscopic landmarks with minor complications and threshold lengths of C1 purchase screws during this procedure.[83]

Table 4 Select clinical series

Authors	Study design	Demographics	Etiology	Surgery	Outcomes and complications
Wang et al (2007)[54]	Retrospective clinical and radiologic case series	$n = 57$; 35 M, 22 F; mean age: 35 y (range, 13–66 mo); average follow-up 47 mo (range, 24–76)	Reducible atlantoaxial dislocation (AAD); os odontoideum ($n = 26$)	Transarticular screw fixation with morselized autograft ($n = 57$)	2 screw malplacements: 1 penetrated to occipitoatlantal joint, 1 slightly breached vertebral artery groove, no clinical sequelae
			Reducible AAD: transverse ligament disruption/relaxation ($n = 20$)		
			Reducible AAD: odontoid fracture nonunion ($n = 6$)		
			Irreducible AAD: os odontoideum ($n = 1$)	Transoral anterior AA joint release and reduction for irreducible AAD ($n = 5$)	No screw fracture, loosening, or backing out
			Irreducible AAD: transverse ligament relaxation ($n = 1$)		No pseudarthrosis
			Irreducible AAD: malunion of odontoid fracture ($n = 3$)		2 cases of postoperative iatrogenic C2–C3 instability
Harms and Melcher (2001)[88]	Retrospective clinical and radiologic case series	$n = 37$; 19 M, 18 F; mean age: 49 y (range, 2–90)	Fractures ($n = 20$)	Polyaxial C1L-C2PSF ($n = 37$)	1 deep wound infection
			Symptomatic os odontoideum ($n = 6$)		No cases of implant failure
			Rheumatoid arthritis ($n = 4$)		Solid fusion in all patients
			Rotary subluxation ($n = 3$)	Transoral odontoidectomy ($n = 2$)	
			Osteoarthritis C1–C2 ($n = 2$)		Satisfactory screw placement in all patients
			Congenital malformation ($n = 2$)		No dural laceration or vertebral artery injury

Abbreviations: AA, atlantoaxial; AAD, atlantoaxial dislocation; C1L-C2PSF, C1 lateral mass screw and C2 pedicle screw fixation.

Table 5 Summary of posterior surgical techniques

Method	Stability	Complications	Surgical procedure
Transarticular screw fixation with sublaminar wiring	Very high fusion rates, up to 100%, due to direct fusion and stability	Neurologic damage can occur due to wire loosening, which has been reported in up to 50% of patients (Matsumoto et al 2005[95])	Transarticular screw fixation placement risks damage to vertebral artery, hypoglossal nerve; most difficult posterior procedure
	One of the gold standard treatments	Procedure risks damage to vertebral artery	Articular screw placement requires steep screw angle and joint exposure that can cause C2–C3 instability
Transarticular screw fixation with C1 hook	Added stability compared with transarticular screw fixation alone, three-point fixation	Minimizes risk of neurologic damage (no wire) and subaxial instability (from dissection for wire insertion)	Fluoroscopic imaging required for proper placement of screws to avoid complications
Transarticular screw fixation with morselized autograft	Less stable than other transarticular screw fixation constructs	Minimizes risk of neurologic damage (no wire)	Wiring risks neurologic damage, and tissue dissection for wiring can cause weakness or longer healing process
	Sufficient biomechanical stability	Instrumentation breakage has been reported, maybe due to screw	C1 hook or autograft obviates risk of tissue dissection
C1L-C2PSF	Best overall stability; more stable than intact spine on axial rotation, lateral bending, and AP translation; less stable on flexion/extension	No complications reported in main clinical series of this procedure	Extension to occiput or subaxially is simple if necessary
			C1L is compatible with most anatomical variations, imaging unnecessary
			C2P pedicle variations make placement challenging; ~20% of patients have insufficient pedicles for placement
			Intraoperative reduction is possible
C1L-C2LSF	Not as stiff as transarticular screw fixation and C1L-C2PSF, especially on lateral bending	Early hardware failure	Weaker in extension to occiput than C1L-C2PSF construct
	Less stable than C1L-C2PSF on axial rotation as well	Relatively safe regarding neurovascular injury, probably safest of the techniques	C1L is compatible with most anatomical variations; intraoperative imaging unnecessary
	Should be reserved for when anatomy restricts to this construct (Finn et al 2008[63])		C2L key structures can be visualized during procedure, less challenging
			Technically less complicated procedure, but less stability

Abbreviations: AP, anteroposterior; C1L, C1 lateral mass screw; C1L-C2PSF, C1 lateral mass screw and C2 pedicle screw fixation; C2L, C2 lateral mass screw; C1L-C2LSF, C1 lateral mass screw and C2 laminar screw fixation; C2P, C2 pedicle screw.

Posterior Methods

The most common posterior surgical methods include C1–C2 transarticular screw fixation, C1 lateral mass screw–to–C2 pedicle screw fixation, and C1 lateral mass screw–to–C2 laminar screw fixation. Posterior techniques can be used alone for reducible atlantoaxial dislocation treatment or in conjunction with anterior transoral decompression to treat certain types of irreducible atlantoaxial dislocation. In many cases, the posterior instrumentation extends to the occiput or fuses the occiput to the axis, especially if there is a congenital anomaly causing instability that extends beyond the atlantoaxial joint or if anatomical anomalies preclude screw placement into the atlas (►**Table 5**).[33] For cases in which placement of a C1 lateral mass screw is difficult, recently described procedures such as open, posterior-only, and instrumented reduction of a fixed C1–2 subluxation using occipital and C2/C3 fixation as reported by Meals et al provide surgeons more options.[84]

Historical Methods

Some surgical procedures that have been used in the past are highlighted here as historical notes and in comparison with contemporary procedures. The earliest surgical treatments for atlantoaxial dislocation used the Gallie or Brooks methods of wiring and bone grafting to fuse and stabilize the atlantoaxial joint.[85] This practice has fallen out of favor because posterior wiring requires external immobilization (bracing) of the head and neck postoperatively and longer hospitalizations and has a nonunion rate of up to 30%.[12,86–88] Sublaminar wiring has also been implicated as a cause of neural injury and disruption of ligamentous tissue that supports the spinal column.[13] Posterior wiring may still be used for secondary fixation with transarticular screws in certain salvage procedures,[89] or in cases with highly abnormal anatomy.

The Magerl technique, or C1–C2 transarticular screw fixation with posterior wiring, was first described in 1986 and became popular for cervical fixations including treatment of atlantoaxial dislocation because of its superior biomechanical strength and high fusion rates compared with posterior wiring,[12] with fusion rates as high as 100% reported.[90–92] This technique is successful because it provides immediate and direct fixation at the atlantoaxial joint. A midline incision is made to expose the posterior arch of C1 and lamina of C2.[90,91] The screw is positioned anteriorly through the C2 pars, across the C1–C2 joint, and into the anterior aspect of the lateral mass of the atlas. In 1995, McGuire and Harkey introduced a modification of this technique to minimize tissue dissection in which a smaller midline incision is supplemented with two bilateral 1-cm caudal percutaneous incisions ~2 cm lateral to the T1 spinous process.[90,93,94] Once the C1–C2 joint is positioned and adequately reduced and the screws are placed, a bone graft is placed between C1 and C2 and wiring goes around the posterior arches to resist torsional and flexion forces.[91] However, one study found that up to 50% of the wires were loosened after using the Magerl technique with a 95% fusion rate after a mean follow-up of 45 months,[95] suggesting that posterior wiring is not required for successful fusion. Loosening of posterior wiring can violate the spinal cord and cause neurologic deficits.[95]

Recently, a modified Magerl technique that uses transarticular screw fixation with morselized autograft without posterior wiring has been described.[54] Although this technique has less biomechanical strength than the Magerl technique, the authors reported that patients maintained a high fusion rate and avoided the risk of neurologic complications and loosening of cables associated with posterior wiring.[54] Similarly, the Magerl technique has recently been used in combination with a transarticular screw fixation with C1 hook fixation for reducible atlantoaxial dislocation.[13,96,97] With this method, transarticular screws are placed through the C2 pars and connected ipsilaterally with a rod to a hook grasping the posterior arch of C1. Bone graft is placed in between the C1 posterior arch and C2 lamina. This modified technique is easier and less invasive than sublaminar wiring because the posterior arch of C1 does not need to be exposed.[13] Additionally, this three-point fixation provides high biomechanical stability while also preserving the ligamentous tissue between C2 and C3. However, ~20% of patients requiring atlantoaxial fusion have anatomical variations that would interfere with safe placement of transarticular screws and must undergo a different fusion technique.[88]

The C1 lateral mass screw and C2 pedicle screw fixation (C1L-C2PSF) utilizes a C1 lateral mass screw technique in combination with a C2 pedicle screw fixation connected by rods to stabilize the atlantoaxial joint, although certain centers prefer plates connecting the fixation points.[88,98,99] The technique enables extension to the occiput or subaxially if needed, and screw placement is more precise.[89,100] Additionally, it allows for intraoperative reduction after screw fixation.[88] C1 lateral mass screw placement has an advantage over the Magerl technique because it is compatible with most anatomical variations and does not require posterior wiring.[89,101] The trajectory and placement of the C1 lateral mass screws is guided mostly by anatomical landmarks and does not require fluoroscopic imaging as does the Magerl technique.[88] Additionally, C1L-C2PSF permits individual placement of screws in C1 and C2 to allow direct intraoperative manipulation for corrective alignment of the head and reduction of the dislocation.[88,89] Neural complications associated with posterior wiring of the Gallie and Magerl techniques are mostly eliminated because sublaminar wiring is not required for C1L-C2PSF.[88] Although longer fusion is required due to the involvement of levels adjacent to the atlantoaxial dislocation, this technique can also be applied for patients with an underlying rheumatic or ligamental cause for their atlantoaxial dislocation.[88] In up to 20% of patients, the pedicles of C2 are too narrow for pedicle screws to be placed, and overall great care must be taken in pedicle screw placement. Additionally, the close proximity of the vertebral artery laterally, the spinal cord medially, and nerve roots vertically put patients at high risk for neurovascular complications.[102] Yeom et al compared the risk of vertebral artery injury by C1–C2 transarticular screw versus C2 pedicle screw and found that the two techniques did not differ in risk of vertebral artery injury but recommended the use of pedicle screw over transarticular screw in cases of high-riding vertebral artery.[41]

Alternatively, the C1 lateral mass screw and C2 laminar screw fixation (C1L-C2LSF) technique was described in 2004 by using a crossed screw approach through the C2 lamina. This technique minimizes the risk of vertebral artery injury compared with transarticular screw fixation and C1L-C2PSF and allows direct visualization of the lamina to guide screw placement intraoperatively.[8,20,37] Because there is no need for intraoperative fluoroscopy, the C1L-C2LSF procedure minimizes radiation exposure and reduces intraoperative time.[103] It appears to be the safest of all available constructs with regards to vertebral artery injury. However, the biomechanical strength is similar to that of the transarticular screw fixation and C1L-C2PSF construct, but it cannot resist lateral bending as well.[63,103,104] Additionally, there is a high incidence of early hardware failure with this technique.[41]

A 2008 report compared the three most commonly used constructs for atlantoaxial dislocation fixation (transarticular screw fixation, C1L-C2PSF, C1L-C2LSF), determining that both transarticular screw fixation and C1L-C2PSF showed greater stability, relative to C1L-C2LSF, with no significant biomechanical differences between these two constructs.[63] Thus, the two are interchangeable depending on the opinion of the surgeon and the anatomy of the patient. A detailed biomechanical analysis of the C1L-C2PSF versus C1L-C2LSF techniques across four domains of motion (anteroposterior translation, lateral bending, axial rotation, and flexion/extension) revealed that the C1L-C2PSF is a structurally superior construct because it is stiffer in lateral bending and axial rotation when compared with C1L-C2LSF.[105] Both of these constructs were found to be stronger than an intact spine. However, the C1L-C2LSF was only stronger in anteroposterior translation and lateral bending, whereas the C1L-C2PSF construct also resisted axial rotation better.[105] C2 laminar screws have also been demonstrated to be weaker in occipitocervical construct extensions.

Conclusion

Atlantoaxial dislocation is a rare but dangerous condition that affects particular patient populations disproportionately. A variety of etiologies contributes to atlantoaxial dislocation, and the underlying causes or predisposing factors must be taken into consideration to maximize timely and effective treatment. The recent Wang classification system offers a practical and clinically proven means to effectively diagnose and treat atlantoaxial dislocation. Several anterior and posterior surgical techniques are suitable for atlantoaxial fusion. Each technique has its own indications, contraindications, risks, and technical difficulty. Future prospective clinical trials are necessary to more adequately determine which surgical technique is optimal based for respective clinical presentations.

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