Occipitocervical fusion of traumatic atlanto-occipital dissociation in a patient with autofused cervical facet joints: illustrative case

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BACKGROUND Patients who survive traumatic atlanto-occipital dissociation (AOD) may present with normal neurological examinations and near-normal-appearing diagnostic images, such as cervical radiographs and computed tomography (CT) scans.

OBSERVATIONS The authors described a neurologically intact 64-year-old female patient with a degenerative autofusion of her right C4–5 facet joints who presented to their center after a motor vehicle collision. Prevertebral soft tissue swelling and craniocervical subarachnoid hemorrhage prompted awareness and consideration for traumatic AOD. An abnormal occipital condyle–C1 interval (4.67 mm) on CT and craniocervical junction ligamentous injury on magnetic resonance imaging (MRI) confirmed the diagnosis of AOD. Her autofused right C4–5 facet joints were incorporated into the occipitocervical fusion construct.

LESSONS Traumatic AOD can be easily overlooked in patients with a normal neurological examination and no associated upper cervical spine fractures. A high index of suspicion is needed when evaluating CT scans because normal values for craniocervical parameters are significantly different from the accepted ranges of normal on radiographs in the adult population. MRI of the cervical spine is helpful to evaluate for atlanto-occipital ligamentous injury and confirm the diagnosis. Occipitocervical fusion construct may need to be extended to incorporate spinal levels with degenerative autofusion to prevent adjacent level degeneration.

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KEYWORDS atlanto-occipital dissociation; atlanto-occipital dislocation; degenerative facet fusion; facet joint autofusion; occipitocervical fusion

Atlanto-occipital dissociation (AOD) is an uncommon injury, with an incidence of 0.2% to 0.6%, whereby the skull is forcibly separated from the upper cervical spine due to a tremendous amount of traumatic blunt force.[1,2] AOD is historically considered a fatal diagnosis, but the advent of improved prehospital systems and shorter transportation times have increased the number of survivors of this devastating injury.[3] Early treatment with occipitocervical fixation or halo vest immobilization is paramount because of the severe spinal instability that is associated with AOD. Nearly all patients who are not treated develop some form of neurological decline, and many never fully recover neurological function.[4]

Proper initial diagnosis of AOD can be challenging for several reasons. First, AOD is foremost a ligamentous injury that is not always associated with coexisting upper cervical spine fractures. Second, cervical radiographs may be inadequate or difficult to interpret, and even established radiographic indices on computed tomography (CT) may be normal depending on the severity and type of AOD. Third, patients with AOD may be neurologically intact, which further decreases the suspicion of upper cervical spine injuries. Finally, the upper cervical spine has a complex anatomical configuration; therefore, the radiographic criteria for instability in this region of the spine in patients with a normal-appearing radiograph or CT scan remain controversial.[5] The following illustrative case highlights a neurologically intact patient who presented with AOD after a motor vehicle collision and was found to have autofused facet joints on CT. The rationale for diagnostic work-up and the proposed surgical plan in light of this patient's cervical facet autofusion are discussed.

Illustrative Case

The patient was a 64-year-old woman without any significant medical history who presented to the emergency department after a motor vehicle collision. She was a restrained driver in a vehicle that was struck on the driver's side by a truck traveling approximately 40 to 45 miles per hour at an intersection. Her airbags were deployed, and she had to be extricated from the vehicle. On evaluation she was awake, alert, and following commands. She endorsed intractable nausea and sharp, diffuse neck pain. She did not exhibit any cranial nerve palsies. On strength examination, she exhibited pain-limited 4+ movements in her bilateral upper extremities and full strength in her bilateral lower extremities. Sensation was intact to light touch in all four extremities. Other than 3+ reflexes in her patellar reflexes, she did not have other signs of hyperreflexia on the rest of her deep tendon reflexes. Hoffman's sign, Babinski's reflex, and clonus were not present.

CT of the brain demonstrated a scant amount of subarachnoid hemorrhage in the left posterior Sylvian fissure as well as subarachnoid blood anterior to the medulla at the level of the foramen magnum (Fig. 1). CT of the cervical spine demonstrated splaying of the right occiput to the C1 condylar space and diastasis of the right C1–2 facet as well as significant prevertebral edema along the entire cervical spine (Fig. 2). Of note, the patient had a degenerative autofusion of the right C4–5 facet joints. The basion-dens interval (BDI) was 10.1 mm, basion-posterior axial line (PAL) distance was 11.9 mm, Powers ratio was 1.13, and right condyle–C1 interval (CCI) was 4.67 mm (Fig. 3). CT angiography of the neck was negative for internal carotid artery or vertebral artery dissection. Magnetic resonance imaging (MRI) of the cervical spine demonstrated T2-weighted imaging and short tau inversion recovery (STIR) signal hyperintensity at the craniocervical junction, suggesting ligamentous injury. Specifically, injury to the atlanto-occipital ligament, apical ligament, tectorial membrane, transverse ligament, and posterior ligamentous complex was observed. There was also significant prevertebral edema spanning from the basion to C5 as well as evidence of hemorrhage in the cervicomedullary junction (Fig. 4). Together, these imaging findings were concerning for a Type 2 AOD.

Surgery was recommended for stabilization of the occipitocervical junction, to which the patient and her spouse consented. The plan was made for occiput to C5 posterior instrumentation and arthrodesis with tricortical ileum structural allograft. The patient was
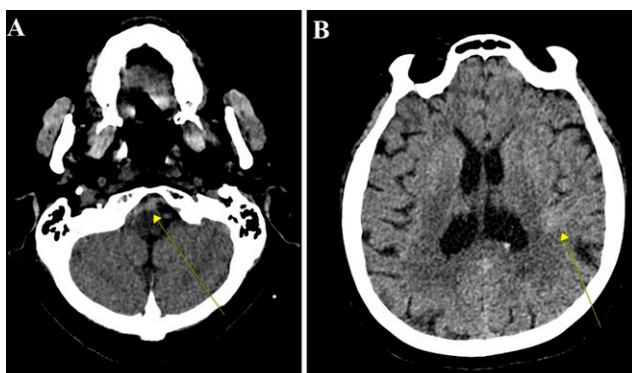
FIG. 1. Initial CT of the head demonstrating subarachnoid hemorrhage ventral to the medulla (*arrow*; **A**) and within the left posterior Sylvian fissure (*arrow*; **B**).
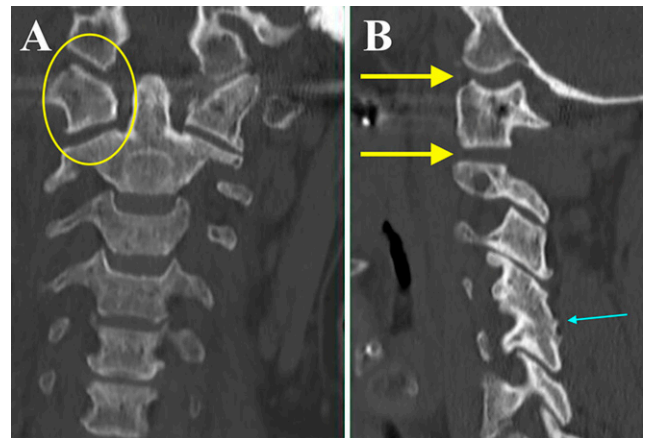
FIG. 2. A: Coronal CT of the cervical spine demonstrating splaying of right occiput–C1 and C1–2 space relative to left side (*circle*). **B:** Sagittal CT demonstrating splayed segments on right side with some air in occiput–C1 space (*yellow arrows*) as well as fused right C4–5 facet (*blue arrow*).

secured in Mayfield pins and carefully placed prone, with confirmation of neutral head positioning with relation to her shoulders and the rest of her body through both visual inspection and occipitocervical alignment on fluoroscopy. Motor evoked potentials and somatosensory evoked potentials were monitored and remained stable throughout the procedure. A midline incision and exposure were performed from occiput to C5, with the posterior arch of C1 exposed. A reference frame was sterilely attached to the Mayfield pins and an intraoperative O-ARM stereotactic navigation (Medtronic) spin was obtained, followed by registration of instruments and verification of navigational accuracy. Pilot holes were first made for bilateral C2 pedicle screws and bilateral lateral mass screws at C3, C4, and C5. Two 20-mm pedicle screws were placed in C2 while 14-mm lateral mass screws were placed in C3, C4, and C5. To achieve a smoother and less acute rod angle, no instrumentation was placed at C1. A small occipital plate was fixed to the occipital bone using three 12-mm screws along the midline keel and two 6-mm screws laterally. The occipital bone, posterior arch of C1 and lateral masses, and facet joints of C2 to C5 were decorticated. A tricortical ilium graft was wedged from the occipital bone over the C1 bone to the top of the decorticated C2 and fashioned with troughs bilaterally in preparation for prebent rods. Postoperative radiographs showed good alignment of the occiput to the cervical spine (Fig. 5). The patient recovered well from the surgery, her nausea resolved after the procedure, and she was discharged to a rehabilitation center after an uneventful postoperative course and stable neurological examination.

Discussion

Observations

Summary of Case

This illustrative case focuses on a 64-year-old woman who had a traumatic Type 2 distraction-type AOD after a motor vehicle collision. She presented with a near normal neurological examination and autofused facet joints. AOD can be frequently missed even by experienced clinicians because it is primarily an injury to the ligaments between the occiput and upper cervical spine. Furthermore, there is not always an associated upper cervical fracture that draws
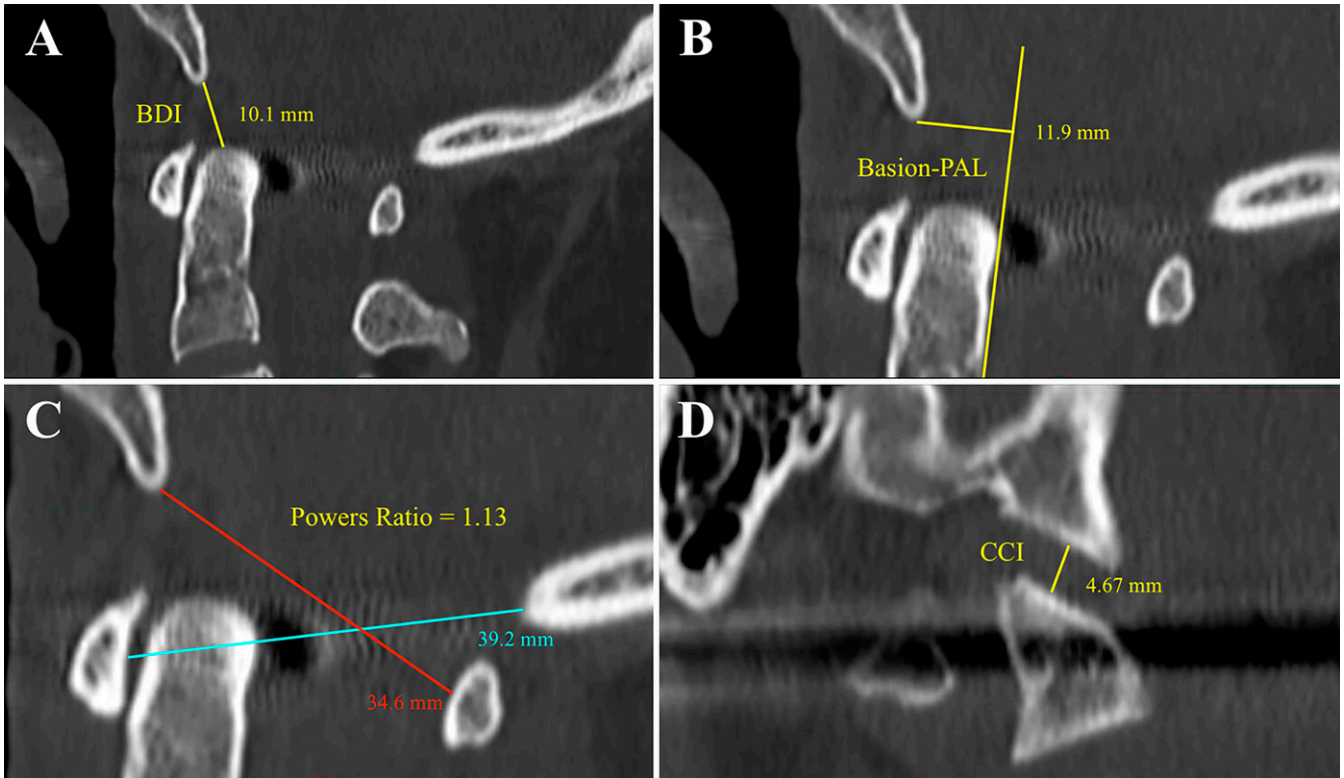
FIG. 3. A: Calculation of BDI is 10.1 mm. **B:** Calculation of basion-PAL is 11.9 mm. **C:** Powers ratio, which equals the ratio between the distance of the basion to the posterior arch of C1 (*red line*) over that of the opisthion to the anterior arch of C1 (*blue line*), is 1.13. **D:** Calculation of the occipital CCI is 4.67 mm.

attention to the craniocervical junction. Thus, correctly diagnosing this uncommon injury requires a high index of suspicion and a thorough investigation of the craniocervical junction. At first glance, the upper cervical spine may sometimes appear normal on CT in some patients. The craniocervical parameters in our patient were abnormal based on CT criteria of the BDI, Powers ratio, and CCI. Her prevertebral soft tissue swelling and craniocervical subarachnoid hemorrhage also prompted awareness and consideration for AOD. The abnormal craniocervical junction ligamentous injury on MRI

further confirmed the diagnosis of AOD. She was kept immobilized in a hard cervical collar with cervical c-spine precautions until surgery for internal fixation and fusion.

Rationale for Surgical Plan

The patient's existing right congenital C4–5 facet fusion had to be taken into consideration when planning our surgical approach. We believed it was important to include the C4 and C5 levels in the posterior fusion construct to reduce the risk of accelerated
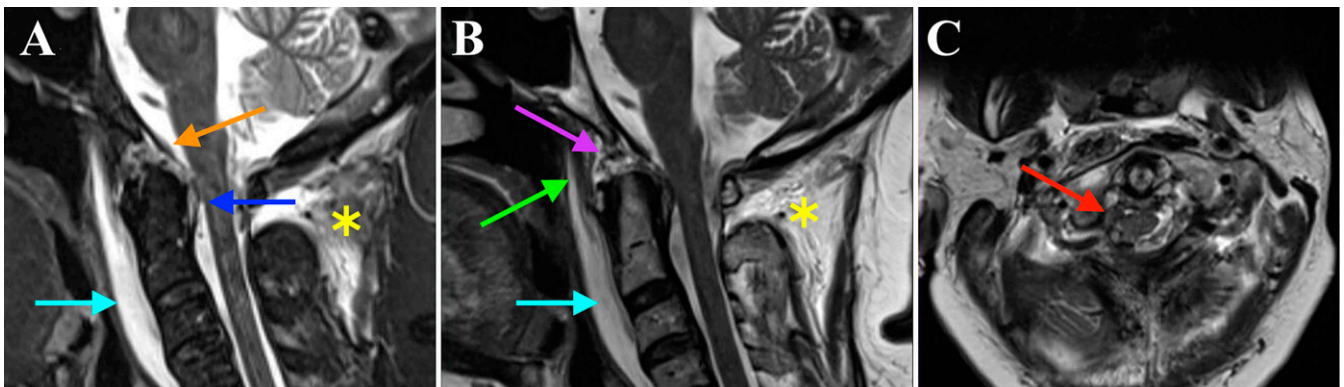

FIG. 4. Sagittal MRI of the cervical spine without gadolinium showing STIR (**A**) and T2-weighted imaging (**B**) signal hyperintensity at the craniocervical junction, suggesting ligamentous injury. Specifically, injury to the atlanto-occipital ligament (**B,** *green arrow*), apical ligament (**B,** *purple arrow*), tectorial membrane (**A,** *orange arrow*), transverse ligament (**A,** *dark blue arrow*), and posterior ligamentous complex (**A and B,** *yellow asterisks*) was observed. Prevertebral edema from basion to C5 is also apparent (**A and B,** *light blue arrows*). Axial MRI (**C**) of the cervical spine without gadolinium demonstrating epidural blood (*red arrow*) seen in the cervicomedullary junction toward the right side.
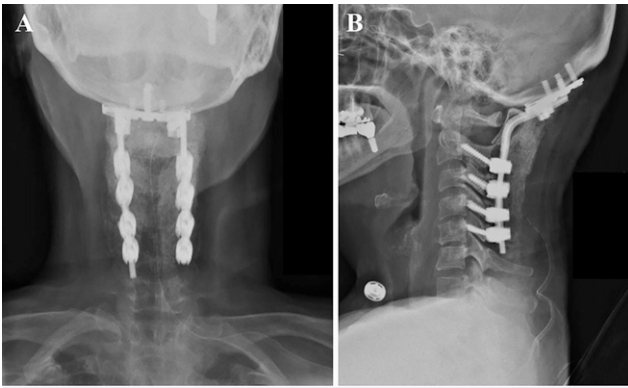
FIG. 5. Postoperative upright radiographs showing good alignment of occiput to cervical spine in the anteroposterior (**A**) and lateral (**B**) views.

degeneration of the C3–4 level. It is apparent that hypermobility at the adjacent levels of a fusion construct may generate degenerative changes in nonfused segments.[6] Therefore, performing an occiput–C2 or occiput–C3 fusion in this patient would expose the C3–4 level to further accelerated changes and hypermobility due to the autofusion of the caudal C4–5 level. Biomechanical studies have shown that intradiscal pressure at intervertebral levels adjacent to cervical fusions increases by approximately 50% in the proximal adjacent level and 125% in the distal adjacent level.[7] Furthermore, the additional segmental fixation at C4 and C5 acts to decrease the lever arm of the bone–screw cantilever model. This ultimately reduces hardware failure because the screw–bone interface experiences a moment force proportional to the length of the lever arm, as determined by the length of rod between fixation points.[8] Adding intervening lateral mass screws at the C3, C4, and C5 levels serves to decrease the length of this lever arm and reduce of magnitude of the applied moment force.

Diagnosis of AOD

An accurate and timely diagnosis as well as proper treatment of AOD are of utmost importance. Historically, AOD was classified into three types based on the direction of dislocation as described by Traynelis et al.: anterior (Type I), longitudinal (Type II), and posterior (Type III) dislocation of the occiput from the cervical spine.[9] Despite the classifications, there is a lack of consensus regarding the best measurement technique in the diagnosis of AOD. Original descriptions of the diagnosis of AOD were based on radiographs and include BDI >10 mm by Wholey et al.[10] and the Powers ratio measuring the basion to posterior atlas distance divided by the distance between the opisthion and anterior atlas (Powers ratio >1).[11] These two methods are the most commonly encountered; however, alternative measurements describe distances between the mandible and the atlas or axis, as described by Dublin et al.[12] The sensitivity of the various radiographic diagnostic techniques have been considered a point of debate. A study by Lee et al. determined that the sensitivity of the Wholey method, Powers ratio, and Dublin's method was 50%, 33%, and 25%, respectively,[13] whereas Harris et al. determined that the basion-axial interval (BAI)-BDI method was 100% sensitive on lateral cervical spine radiographs.[14] Specifically, Harris et al. described that improved detection of AOD is achieved by measuring both BAI and BDI, where BAI is the distance from the basion to the C2 posterior line and BDI is the distance from the basion to the tip of the dens (abnormal BAI and BDI >12 mm).[14,15] The improved sensitivity likely stems from the combination of these two measurements, in which BDI more consistently captures Type II dislocations and BAI identifies Types I and III dislocations. Trauma guidelines by Theodore et al. describe level III evidence that lateral cervical radiographs are the preferred imaging modality to obtain BAI-BDI, and when taken together, the Harris method is the most reliable radiographic analysis in the diagnosis of AOD.[4] If radiographs are inadequate, then additional imaging with CT and MRI is needed to confirm the diagnosis of AOD. It is important to keep in mind that normal values for craniocervical parameters are significantly different from the accepted ranges of normal on radiographs in the adult population. For instance, the normal values of BDI and Powers ratio on CT scan are <8.5 and 0.9, respectively.[16] The BAI is highly variable in adults and is associated with a high standard error, making it difficult to ascribe a normal value based on CT. A CT-based occipital CCI of 1.5 mm (condylar sum of 3.0 mm) cutoff value had the highest sensitivity and specificity for the diagnosis of AOD in the adult population, according to Martinez-Del-Campo et al.[17]

MRI for AOD

The principal structures that confer stability to the atlanto-occipital joint are the alar ligaments, tectorial membrane, and atlanto-occipital joint capsules.[18] Deciding on stability of the upper cervical spine remains a challenge. There are several different classification systems, yet none of them consider combination injuries of the upper cervical spine.[5] The Harborview group proposed a new classification for AOD involving the combination of static and dynamic imaging plus MRI.[19] However, many clinicians may be deterred from this classification given the need to perform dynamic testing to detect instability of an injured upper cervical spine. Horn et al. proposed a simple grading system for atlanto-occipital injuries to determine stability of the upper cervical spine that incorporated abnormal findings on CT and ligamentous injury on MRI.[20] Based on this classification system, Grade I injuries are designated by normal CT findings in relation to established methods of diagnosis (the Powers ratio, BDI, BAI-BDI, and X-line) but have moderately abnormal MRI findings (high posterior ligaments or atlanto-occipital signal). The authors support nonoperative treatment in patients with Grade I injuries. Grade II injuries include a minimum of one abnormal finding on CT based on established diagnostic criteria or grossly abnormal MRI findings in the atlanto-occipital joints, tectorial membrane, alar ligaments, or cruciate ligaments. In these patients, surgical fixation is recommended. Using the Horn et al. classification, our patient was deemed to have a Grade II injury and therefore considered to have upper cervical spine instability based on CT and MRI criteria.

Treatment of AOD

Among the available treatment modalities, external immobilization using a halo orthosis and craniocervical fusion are the mainstays of treatment. Traction is not a recommended treatment option for patients with AOD because there is a high risk (10%) of neurological deterioration.[4] Based on the 2013 Congress of Neurological Surgeons guidelines and systematic review by Theodore et al., treatment with internal fixation and fusion remains a level 3 evidence-based recommendation.[4] Importantly, of the 12 patients who were treated with external orthosis, 4 (33%) worsened transiently and required subsequent craniocervical fusion. Of the remaining 8 patients managed

with external immobilization alone, 3 (37.5%) remained unstable after 6 to 22 weeks of immobilization. Comparatively, only 1 of 29 patients (3.4%) with planned early craniocervical fusion worsened neurologically after surgery. In this group, no patients experienced late instability that required another operation. Failure to treat AOD resulted in neurological worsening in 7 of 13 patients (54%). Therefore, early diagnosis and treatment with craniocervical fusion are recommended in patients with AOD. Treatment with external immobilization alone is associated with unacceptably high rates of neurological deterioration and nonunion.

Prognosis of Patients with AOD

Autopsy studies report an AOD rate of incidence up to 8% among patients who die on scene after a high-energy mechanism of injury.[21] Because the annual incidence of patients with AOD who present to the emergency department ranges only from 0.2% to 0.6%, it is readily apparent that AOD is associated with a high mortality rate.[1,2] AOD is often commonly associated with traumatic brain injury. Patients who survive AOD may present with neurological deficits such as lower cranial nerve palsies, unilateral or bilateral weakness, or quadriplegia. However, nearly 20% of patients with traumatic AOD have a normal neurological examination, which may delay or cause clinicians to miss the diagnosis, particularly in patients who also have normal-appearing initial cervical radiographs.[4] To emphasize the difficulty of making the diagnosis, consider a report by Souslian et al. that described a 37-year-old polytrauma patient who had traumatic AOD after a high-speed motor vehicle accident.[22] The patient had normal occiput–C1 craniometric parameters and unexplained perimesencephalic subarachnoid hemorrhage. Cervical MRI showed evidence of disruption of more than two atlanto-occipital ligaments, which led to the diagnosis and subsequent halo immobilization followed by occipitocervical fusion.[22] Schellenberg et al. queried the National Trauma Data Bank in 2018 and showed that traumatic AOD is not as devastating as previously considered because 78% of patients who arrived alive to the hospital survived to discharge.[23] In that database study, 1,489 patients were diagnosed with AOD almost exclusively after blunt traumatic mechanisms (97%), particularly after motor vehicle collisions (66% of cases). Lower age, lower Injury Severity Score, and higher Glasgow Coma Scale score on admission independently predicted survival, but time to neurosurgical intervention did not. A 15-year retrospective study by Mendenhall et al. at Vanderbilt University reported 22 patients with early diagnosis of AOD followed by surgery with occipitocervical fusion and 9 patients with a failed diagnosis of AOD.[2] Missed AOD was the strongest predictor of mortality, and patients with better American Spinal Cord Injury Association scores were associated with missed AOD cases.

Lessons

Traumatic AOD can be easily overlooked in patients with a normal neurological examination and no associated upper cervical spine fractures. A high index of suspicion is needed when evaluating CT scans because normal values for craniocervical parameters are significantly different from the accepted ranges of normal on radiographs in the adult population. MRI of the cervical spine is helpful in evaluating for atlanto-occipital ligamentous injury and confirming the diagnosis. Occipitocervical fusion may need to be extended to incorporate spinal levels with degenerative autofusion to prevent adjacent level degeneration.

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