Case Report

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Occipitocervical Dissociation in Three Siblings: A Pediatric Case Report and Review of the Literature

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Abstract

This report includes three cases of occipitocervical (OC) dissociation in three siblings involved in a single, head-on, motor vehicle accident. The oldest sibling, a 9-year-old girl, required surgical fixation. The second sibling, a 6-year-old boy, was treated nonsurgically with a neck brace. The youngest sibling, a 5-year-old boy, sustained a fatal OC dissociation. CT and MRI findings are used to assess the reliability of the currently recommended diagnostic modalities for OC dissociation. The literature review focuses on the diagnosis of OC dissociation via CT measurements and proposed treatment recommendations using CT and MRI classification systems. In the 9-year-old girl, the mean atlanto-occipital joint width, the interspinous ratio, and space available for cord measurements indicated OC dissociation. However, on MRI evaluation, OC dissociation was observed in both living siblings and severe ligamentous injury was noted in the girl. The proposed CT measuring techniques for the diagnosis of OC dissociation were inconsistent in our cases. In the setting of clinically suspected OC dissociation with normal CT measurements, we recommend obtaining a cervical spine MRI to properly assess the extent of ligamentous injury.

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JAAOS Glob Res Rev 2018;2:e067

DOI: 10.5435/JAAOSGlobal-D-17-00067

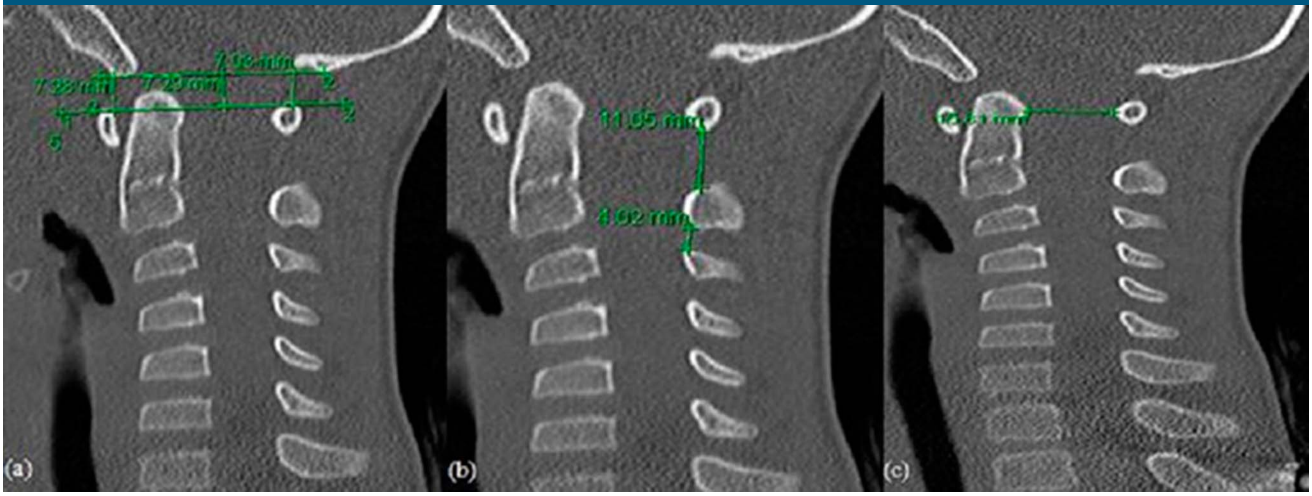
Occipitocervical (OC) dissociation, first described by Blackwood,[1] was generally considered fatal and rare throughout the last century. However, recent innovations in resuscitation, spinal immobilization, emergency transport services, and imaging modalities have resulted in more children presenting with OC dissociation.[2,3] The improved outcomes of OC dissociation have increased awareness of this injury, yet it is still linked to substantial rates of morbidity and mortality.[4]

Case 1

A 9-year-old girl (patient 1) was a restrained backseat passenger in a motor vehicle accident (MVA). Emergency personnel found the patient responsive and stable at the scene. Upon arrival at the emergency department (ED), her initial assessment revealed a Glasgow Coma Scale score of 15.

Cervical CT examination revealed the following abnormal values: an atlanto-occipital junction width of

Figure 1



Cervical CT scans of patient 1 showing a mean width of the AO joint of 7.2 mm (normal < 5 mm) (**A**), an interspinous ratio of 2.75 mm (occipitocervical dislocation > 2.5 mm) (**B**), and a space available for the cord of 15.81 mm (normal > 19 mm) (**C**).

Figure 2

Sagittal magnetic resonance image of the cervical spine of patient 1 showing evolving anterior upper cervical epidural and retroclival hematoma (long arrow) and tectorial membrane and apical ligament disruption (short arrow).

Figure 3

Lateral radiograph of the cervical spine of patient 1 after C2 posterior spinal fusion with instrumentation harvest of right iliac crest bone graft.

7.2 mm (Figure 1, A), an interspinous ratio of 2.75 mm (Figure 1, B), and space available for the cord (SAC) of 15.81 mm (Figure 1, C). Normal measurements included a basion-dens interval (BDI) of 6.27 mm, the Wackenheim line, a Powers ratio of 0.79, a condyle-C1 interval (CCI) of 3.17 mm, and an atlanto-dens interval (ADI) of 2.94 mm.

MRI of the cervical spine revealed an evolving retroclival and anterior upper cervical epidural hematoma, as well as tectorial membrane and apical ligament disruption (Figure 2). Four days after the accident, the patient underwent occiput-to-C2 posterior spinal fusion with instrumentation harvest of right iliac crest bone graft (Figure 3). At the 2-year follow-up, she denied any pain or tenderness in the paravertebral areas. Her range of motion was limited because of the fusion; however, she maintained 50% of her rotation and lateral bending. Cervical flexion and extension radiographs obtained at the 1- and 2-year follow-up visits revealed a good fusion mass at the occiput to C2 with intact instrumentation and no evidence of OC instability.

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Case 2

The 6-year-old brother (patient 2) of the 9-year-old female patient was also a restrained backseat passenger in the same vehicle involved in the MVA. The patient was placed in a cervical immobilization device and transported to the ED. His initial assessment revealed a Glasgow Coma Scale score of 15.

Cervical CT examination revealed the following normal values: an atlanto-occipital junction width of 3.83 mm, an interspinous ratio of 2.14, SAC of 20.05 mm, a BDI of 8.76 mm, the normal Wackenheim line, a Powers ratio of 0.79, a CCI of 3.21 mm, and an atlanto-dens interval (ADI) of 2.47 mm (Table 1).

MRI of the cervical spine revealed the tectorial membrane as intact, although elevated by retroclival hematoma (Figure 4, A). There was no evidence of severe posterior ligamentous injury. The injury was thought to be treatable with a neck brace.

MRI of the brain 5 days after the MVA revealed that the hematoma in the retroclival region was improved, with reduction in the elevation of the tectorial membrane compared with the previous study (Figure 4, B). The

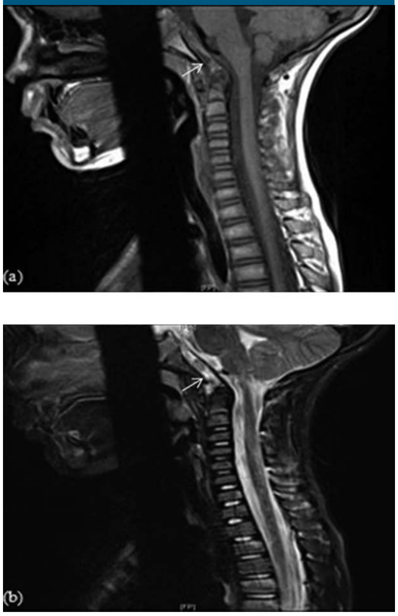
Table 1

Results of Cervical CT Examinations

Atlanto-Occipital Junction Radiographic Measurements	Radiographic Measurement Technique	Measurement Indicating OC Dissociation	Patient 1 Measurements	Patient 2 Measurements
Atlanto-occipital junction	Measurement of the atlanto-occipital joint width	Width >5 mm indicates OC dissociation[5]	7.2 mm	3.83 mm
Interspinous ratio	Ratio of the interspinous distance between C1-C2 divided by C2-C3	Ratio >2.5 indicates OC dissociation[6]	2.75	2.14
SAC	The distance between the posterior surface of dens to the anterior surface of the posterior arch of the atlas	SAC >19 mm is normal, and <14 mm indicates anterior subluxation of the atlas and probable spinal cord injury[7,8]	15.81 mm	20.05 mm
BDI	Measure from the inferior aspect of the basion to the superior aspect of the dens	Interval ≥12 mm indicates OC dissociation[7,9]	6.27 mm	8.76 mm
Wackenheim line	A line that runs along the back of the clivus and should intersect with the posterior half to third of the odontoid process	Wackenhein line out of place	Normal Wackenheim line	Normal Wackenheim line
Powers ratio	Distance between the basion and midpoint of the posterior arch of the atlas divided by the distance between the opisthion and midpoint of the anterior arch of the atlas	Ratio >1 indicates OC dissociation[9]	0.79	0.79
CCI	On either sagittal or coronal CT, four evenly placed marks are made on the articulating surface on the atlanto-occipital joint, with their mean distance equaling the CCI	CCI ≥4 mm indicates OC dissociation[10]	3.17 mm	3.21 mm
ADI	The distance between the anterior arch of the atlas and the dens of the axis	ADI >5 mm on lateral radiographs indicates instability[7]	2.94 mm	2.47 mm

ADI = atlanto-dens interval, BDI = basion-dens interval, CCI = condyle-C1 interval, OC = occipitocervical, SAC = space available for the cord

Figure 4



A, Sagittal cervical spine magnetic resonance image of patient 2 showing the intact tectorial membrane, although elevated by retroclival hematoma (arrow). **B**, Sagittal cervical spine MRI of patient 2 showing reduction in the elevation of the tectorial membrane (arrow) 5 days after the initial cervical spine MRI.

patient was discharged with a neck brace in stable condition. At the 2-week follow-up, the patient had developed a right, lateral gaze palsy, which resolved in 4 months. At the 3-month follow-up, he lacked 20% of lateral rotation to the left. At the 5-month follow-up, physical examination revealed normal rotation and flexion of the neck. He was instructed to remove his cervical collar and given permission to resume physical activities. Cervical flexion and extension radiographs at the 1- and 2-year follow-up periods revealed no OC instability and normal alignment.

Case 3

The youngest sibling (patient 3), a 5-year-old brother of the aforementioned children, was a restrained backseat passenger in the same vehicle involved in the MVA. This child was killed during the accident. The autopsy report obtained from the trauma service revealed that he died with OC dissociation.

Discussion

A delay in the diagnosis and stabilization of OC dissociation can result in secondary neurologic injury, as well as increased morbidity and mortality.[11,12] With advances in imaging modalities, physicians now use several CT measurement techniques of the cervical spine, each with its own drawbacks, to identify OC dissociation. Such methods include the BDI, atlanto-occipital junction width, Wackenheim line, Powers ratio, interspinous ratio, ADI, and SAC. A more recent method, the CCI, was described by Pang et al[10] as having 100% sensitivity and specificity for OC dissociation.

Several classification systems have been proposed to help physicians decide between nonsurgical management and surgical fixation. In 2003, Steinmetz et al[13] described a system using MRI to grade OC dissociation. The injury is deemed either grade I (incomplete) or grade II (complete) depending on the integrity of the tectorial membrane. Steinmetz grade I injuries can be managed nonsurgically, whereas Steinmetz grade II injuries require surgical fixation. Astur et al[14] proposed using surgeon-supervised flexion/extension/distraction imaging with live lateral fluoroscopy when physicians are unable to assess the extent of ligamentous injury via both CT and MRI evaluation.

In 2007, Horn et al[15] proposed a grading system using both CT and MRI. Grade I injuries have normal CT measurements, such as the Powers ratio and other previously mentioned methods, but they do have moderately abnormal findings on MRI (high posterior ligaments or edema in OC joints). Grade II injuries have at least one abnormal finding on the established CT diagnostic criteria or exhibit grossly abnormal MRI findings in the atlanto-occipital joints, tectorial membrane, alar or cruciate ligaments. Grade I injuries qualify for nonsurgical management, whereas grade II injuries warrant surgical fixation.

In our study, patient 1 sustained a Steinmetz grade II injury; MRI showed significant tectorial membrane disruption. Furthermore, according to the Horn classification system, patient 1 sustained a grade II injury (abnormal CT measurements of a 7.2-mm mean atlanto-occipital joint width, an interspinous ratio of 2.75 mm, and SAC of 15.81 mm, as well as significant MRI findings of an evolving retroclival and anterior upper cervical epidural hematoma, with a torn tectorial membrane and apical ligament). In light of these radiographic findings, patient 1 underwent occiput-to-C2 posterior spinal fusion with instrumentation and harvest of right iliac crest bone graft. At the 2-year follow-up, she showed no evidence of OC instability, maintained 50% of her neck rotation and lateral bending, and had an entirely normal neurologic examination.

Patient 2 sustained a Steinmetz grade I injury; MRI revealed a mildly elevated but intact tectorial membrane. His injury was deemed a Horn grade I injury (no abnormal CT measurements and moderate MRI abnormalities revealing a retroclival hematoma without additional evidence of ligamentous injury). The literature reports only a limited number of patients with OC dissociation were successfully treated nonsurgically.[16] Patient 2 was successfully treated with a neck brace for 5 months, resuming normal activities thereafter. At the 2-year follow-up, he showed no evidence of OC instability, had no deficits in his cervical spine range of motion, and had an entirely normal neurologic examination.

Posterior OC fusion is usually performed in patients with OC instability.[14,17,18] Bone graft, usually harvested from the iliac crest, as was performed on patient 1, is commonly used. Because children are more commonly presenting with OC dissociation in the inpatient setting, physicians must be prepared to successfully manage this previously unsalvageable injury.

Conclusion

The measurements collected from our two patients, the mean AO joint width, the interspinous ratio, and SAC measurement indicated OC dissociation in the older sister. The Powers ratio, BDI, and Wackenheim line were all within normal limits. Of note, the CCI was also within normal limits for both of our patients. However, on MRI evaluation, OC dissociation was observed in both living siblings, and severe ligamentous injury was noted in the older sister. In comparison, despite an elevated tectorial membrane, the younger brother's MRI did not reveal the torn ligaments seen in his sister's case. This occurrence demonstrates the inaccuracies of the various CT measuring techniques used for the diagnosis of OC dissociation. The CT measurement methods may prove useful when determining the injury grade based on the classifications proposed by Steinmetz et al[13] and Horn et al.[15] However, in the setting of clinically suspected OC dissociation with normal CT measurements, we recommend obtaining a cervical spine MRI to properly assess the extent of ligamentous injury. The successful treatment of our two patients supports the treatment recommendations proposed by these grading systems. The injuries sustained by these three siblings during the same MVA—ranging from moderate, requiring conservative management; severe, requiring surgical intervention, and fatal—further highlight the spectrum of disease observed in OC dissociation.

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