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Severe posttraumatic craniocervical instability in the very young

Report of three cases

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Children younger than 3 years of age represent a distinct subpopulation of patients at particular risk for high cervical and craniovertebral injuries. There are few descriptions of survivors of severe craniocervical trauma among the very young, and scarce data exist regarding management after initial emergency stabilization.

The authors describe three children, age 1 to 32 months, who presented with craniocervical junction injuries. Variable neurological findings were observed at presentation (cranial nerve deficits, obtundation, and moderate-to-severe quadriparesis). All three were treated with prolonged immobilization and have recovered with minimal to no neurological deficit.

KEY WORDS • occiput • atlas • axis • craniocervical instability • pediatric neurosurgery

SKELETALLY immature children are susceptible to cervical spine injury, complete neurological deficits, and a concomitant rise in mortality rates after traumatic spinal cord injury.[6,8,19,25,39] Atlantooccipital dislocation and AAI secondary to transverse atlantal ligament rupture or dens fracture represent two severe forms of SCI that are commonly fatal or result in severe SCI.[1,9,12,25,41,44] In several case reports, however, authors have described survivors of AOD and AAI with minimal or absent neurological deficit.[17,20,28,30,36,37,44] With improved patient retrieval and emergency management strategies, more survivors can be expected.[18,30]

Children younger than 3 years of age represent a distinct subpopulation of pediatric patients at particular risk for high cervical and craniovertebral junction injury.[22,39,42] Despite this, descriptions of AOD and AAI survivors among the very young are limited.[20,40,42,44] Similarly there is a paucity of data regarding how best to manage these unique patients after initial emergency stabilization. Although there is agreement that rigid internal fixation followed by immobilization therapy should be performed in adult and older-age pediatric survivors of AOD and AAI,[13,21,28] this approach is problematic in children younger than 3 years of age, whose spines are largely cartilaginous.

We describe three children, age 1 to 32 months, who sustained severe craniocervical injuries. The clinical findings, radiographic features, and factors contributing to delay or missed diagnosis are reviewed. The clinical outcomes after halo immobilization are also presented.

Case Reports

Case 1

Presentation. This 18 month-old girl was struck by a car traveling 30 mph, thrown 20 feet, and sustainied a brief loss of consciousness. On admission to the emergency room, she was awake and crying. She was able weakly to flex the right and extend the left upper extremities, while moving the lower extremities spontaneously. After a 48-hour delay in diagnosis, lateral radiography of the neck revealed evidence consistent with AOD and AAI, and spine-related precautions and a neck collar were ordered (Fig. 1 *left*). A head CT scan revealed intraventricular and subarachnoid hemorrhage in the posterior fossa and a depressed occipital skull fracture. An MR imaging study

Abbreviations used in this paper: AAI = atlantoaxial instability; ADI = atlas–dens interval; AOD = atlantooccipital dislocation; BDI = basion–dens interval; CCJ = craniocervical junction; CT = computerized tomography; MR = magnetic resonance; SCI = spinal cord injury.

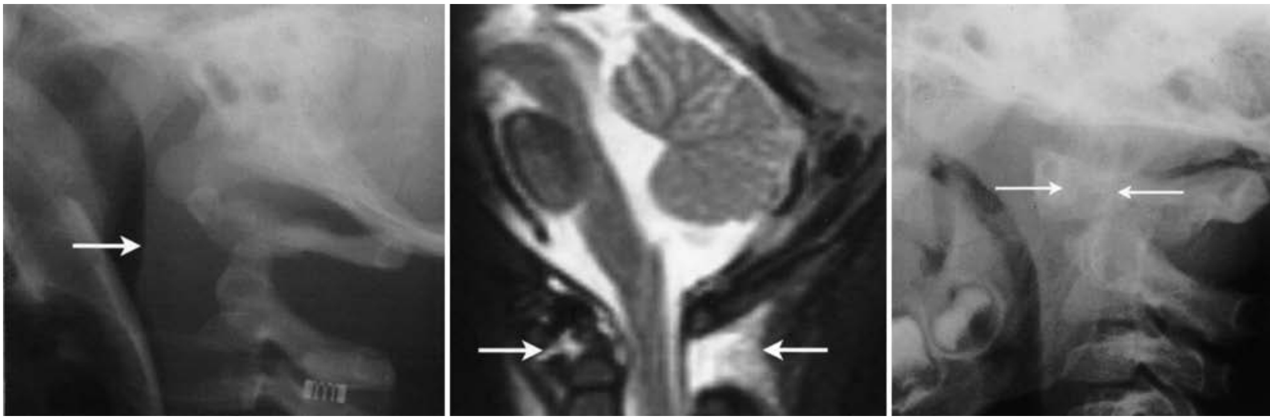

FIG. 1. Case 1. *Left:* Lateral cervical radiograph obtained 48 hours after injury demonstrating an abnormal BDI, marked soft-tissue swelling *(arrow)*, and widening of the ADI. *Center:* Sagittal T_2-weighted MR image of the CCJ, revealing ligamentous and soft-tissue injury *(arrows)*. *Right:* Lateral cervical radiograph obtained 72 months posttreatment demonstrating a normal BDI but continued widening of the ADI *(arrows)*, which remains asymptomatic.

demonstrated ligamentous damage at the CCJ (Fig. 1 *center*). The patient was transferred to our institution for further management.

Examiation and Treatment. Atlantooccipital dislocation was diagnosed in association with moderate quadriparesis. After induction of general anesthesia, a halo vest was placed; eight evenly spaced pins, with 2 in/lbs of torque per pin, were inserted. Bed rest was ordered while the patient was in halo fixation for 2 months; supine lateral cervical radiographs were frequently obtained to ensure normal CCJ alignment. After 1 month, the patient was allowed to sit up in bed, and supine and upright lateral cervical x-ray films continued to demonstrate no displacement. Activity was gradually increased during the 2nd month, and after 3 months of halo fixation, dynamic studies, including lateral cervical fluoroscopic images (flexion, extension, and manual distraction [~15 lbs]) were obtained. Thereafter, the child remained in a Minerva brace for 3 months; when imaging studies demonstrated stability and symptoms were absent, brace therapy was discontinued.

Posttreatment Course. The patient's most recent imaging studies, obtained 72 months after injury, demonstrated widening of the ADI (Fig. 1 *right*), which has remained asymptomatic and unchanged on serial radiographs (Table 2). With no apparent neck deformity or impairment of range of motion, operative intervention has been deferred. Findings on neurological examination were normal at most recent follow-up evaluation.

Case 2

Presentation. This 32-month-old girl was struck by a car traveling 35 mph and was found unconscious and unresponsive at the scene. She was intubated and chemical paralysis was induced in the field.

Examination. Her pupils were small and nonreactive on admission to the emergency room. A lateral cervical radiograph demonstrated AOD (Fig. 2 *left*), which was confirmed by emergency MR and CT imaging. Axial CT scans also revealed a Type I occipital condyle fracture.[2] Her neurological status was notable for 10th and 12th cranial nerve palsies and severe quadriparesis (Grade 1–2/5 strength in all extremities).

Treatment. The patient was treated with halo immobilization and remained at rest in bed; frequent screening lateral cervical radiographs were obtained. After approximately 1 week, malalignment of the craniocervical region was noted, and the patient was returned to the operating room where the halo device was adjusted using fluoroscopic guidance. With an adequate reduction restored, she remained at bed rest for 1 month with her spine in stable alignment. Thereafter, upright lateral cervical spine radiographs documented continued stability; she remained hospitalized and underwent screening radiography frequently for another 2 months. She was discharged from the hospital after dynamic flexion, extension, and distraction x-ray films documented healing and CCJ stability, with a full range of motion (Fig. 2 *center* and *right*).

TABLE 1
*Summary of patient demographic and clinical data obtained in three children with SCI**

Case No.	Mechanism of Injury	Initial Finding	Time to Diagnosis	Follow-Up Finding	Follow-Up Period (mos)
1	MVA vs ped	quadriparesis	4 days	normal	72
2	MVA vs ped	quadriparesis, 10 & 12 CN palsies	1 hr	slight lt 12 CN palsy	72
3	NAT	obtunded, flaccid, quadriparesis	2 mos	developmental delay, Grade 4/5 LUE, paresis	32

* CN = cranial nerve; LUE = left upper extremity; MVA =motor vehicle accident; NAT = nonaccidental trauma; ped = pedestrian.

TABLE 2
*Summary of radiological findings on lateral cervical radiographs**

	Radiological Measurement (mm)					
	Case 1		Case 2		Case 3	
	Presentation	FU	Presentation	FU	Presentation	FU
BDI	14	8	14	8	11	10
ADI	5	1–8	3	2	8	6
SAC	12	16	19	19	6	10
STS	8	2	12	2	10	2
Wackenheim line	+4		−4		+1	
Power ratio	0.4		0.8		0.667	

* FU = follow up; SAC = space available for the cord; STS = soft-tissue swelling.

Spinal stability remained at her most recent follow-up examination 72 months after treatment (Table 2). Neurological examination demonstrated a persistent left 12th cranial nerve palsy and a positive right Babinski sign. Otherwise, cranial nerve, sensation, gait, and balance functions were normal.

Case 3

Presentation. This 24-day-old girl was the victim of violent shaking. Sixteen hours after the incident, the mother noticed that the baby was cyanotic and unresponsive and took the child to a local hospital where the infant was intubated and transferred to a trauma center.

Examination. Several injuries including multiple rib fractures, a liver laceration, and a hip dislocation were noted. On neurological examination, the infant had minimal lower-extremity tone, minimal upper-extremity spontaneous movement, and would only open eyes to voice. Findings on initial cervical spine radiographs were interpreted as normal, but head CT scanning revealed subdural and intraventricular hemorrhage.

The patient remained ventilator dependent and hospitalized in an intensive care unit, suffering from prolonged sepsis. An MR image revealed ligamentous injury at the occipitocervical junction, with atlantoaxial subluxation resulting in marked narrowing of the space available for the spinal cord (Fig. 3 *upper*). Imaging findings were consistent with disruption of the osseocartilaginous ring of C-1 and sagittal atlantoaxial subluxation with rupture of the C-1 transverse ligament of the atlas (Fig. 3 *lower*).

Treatment. The patient was kept in a body cast and underwent head immobilization for 2 months, at which time dynamic radiographic studies revealed widening of the atlantodental interval unchanged in flexion and extension. On dynamic studies, which included gentle manual distraction, the BDI remained unchanged (Table 2).

Posttreatment Course. Follow-up studies, 2 years after removal from the body cast, demonstrated persistent fixed widening of her ADI, and she underwent posterior fusion at another institution. Her neurological condition had stabilized and she was asymptomatic at the time of fusion. Although the patient has significant developmental delay and Grade 4/5 strength in the right upper extremity, she was ambulating without assistance at latest follow-up examination (32 months) (Table 1).

Discussion

Craniocervical stability is provided by a complex arrangement of ligaments. Traumatic disruption of these vital soft tissues, as seen in high-energy blunt trauma, can have fatal consequences. In the past, chances for survival of these injuries were deemed to be scant, at best.[1,9] This, perhaps, has led to a decreased emphasis on these injuries. As is the case with any predominantly ligamentous injury, radiographs obtained in survivors after craniocervical disruption may reveal only subtle abnormalities that may be easily missed.[30,31]

It has been postulated that pediatric patients are more susceptible to traumatic craniocervical disruption,[8,22,25] yet reports on infant survivors of these injuries have been infrequent. In this limited case series we have provided detailed descriptions of traumatic occipitocervical instability in the very young, a distinct subpopulation of children with respect to SCI.[39] The recognition and treatment of craniocervical instability in these patients presents several diagnostic and treatment challenges not commonly encountered in older children and adults.

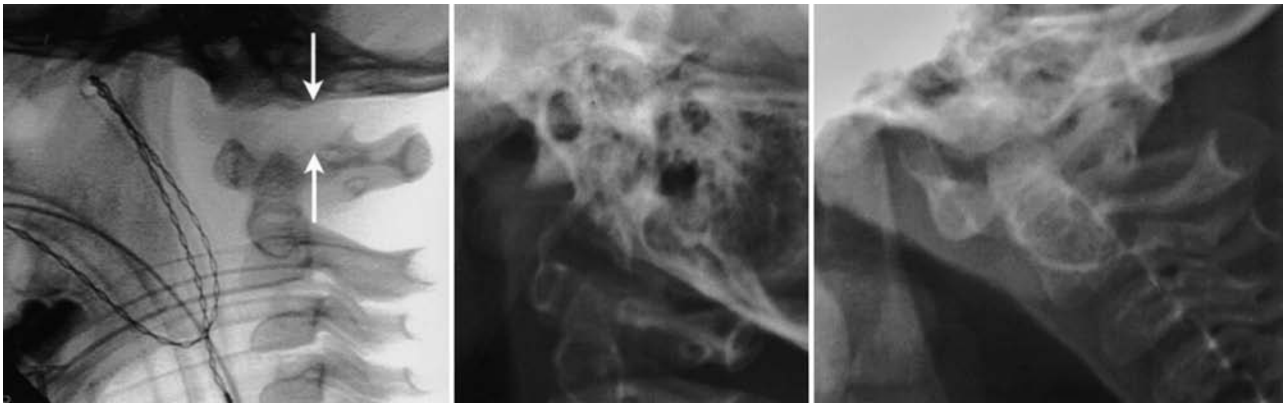

FIG. 2. Case 2. *Left:* Lateral cervical radiograph obtained 1 hour after injury, demonstrating an abnormal BDI, soft-tissue swelling, and a widened atlantooccipital interval *(arrows)*. *Center* and *Right:* Lateral cervical radiographs obtained 72 months posttreatment in extension *(center)* and flexion *(right)*. Normal BDI and ADI are maintained.

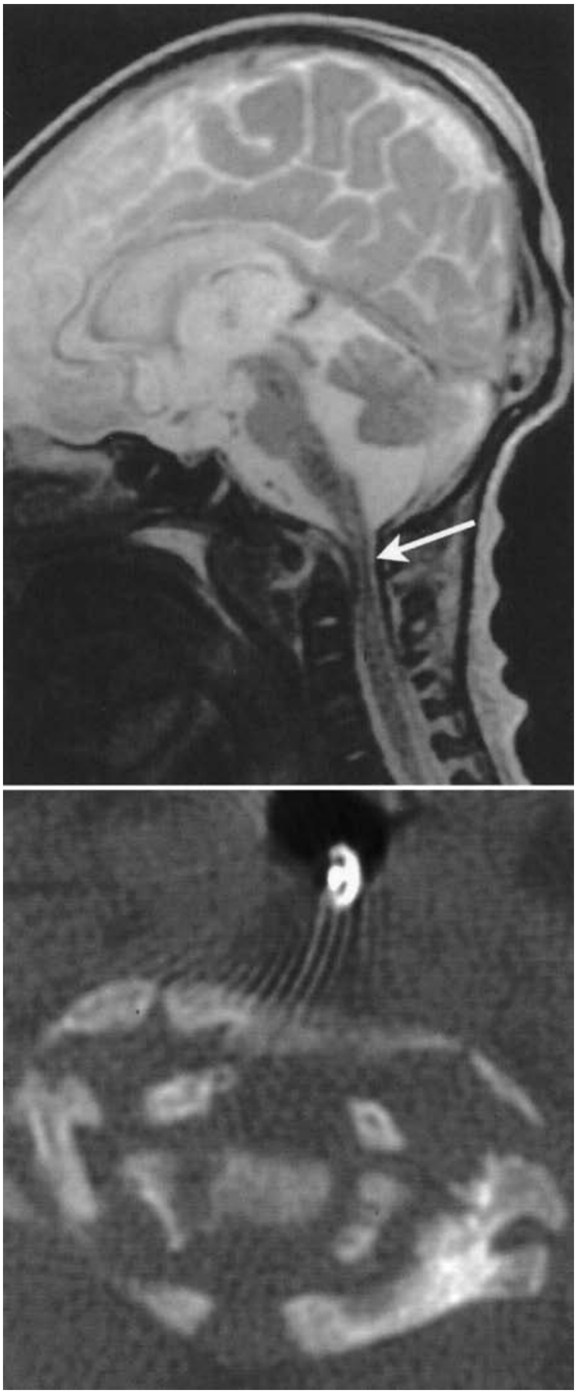

FIG. 3. Case 3. *Upper:* Sagittal T_2-weighted MR image of the CCJ, revealing marked narrowing *(arrow)* of the space available for the cord and AAI. *Lower:* Axial CT scan obtained through C-1 8 weeks after injury, demonstrating disruption of the osteocartilaginous ring of C-1, as well as atlantoaxial subluxation.

Developmental Anatomy and Radiology of the Occipitocervical Junction

In two of the three patients diagnosis of craniocervical instability was delayed (Table 1). In the 18-month-old child (Case 1) the delay may have been influenced by a low index of suspicion because the patient was presumed to be neurologically intact at the time of initial evaluation. An uncertain interpretation of the neurological and radiographic findings, as well as other systemic injuries, led to the missed diagnosis in the 6-week-old child in Case 3. Conversely, clinical findings of brainstem signs and the initial radiographic findings in the 2 year-old child in Case 2 (Fig. 2 *left*) contributed to establishment of a timely diagnosis. Review of these three cases emphasizes that infants require special attention whe interpreting data derived from neurological/physical examination and diagnostic imaging studies, as well as a high index of suspicion based on mechanism of injury.[22,39]

Because of the unique anatomy, biomechanics, and ossification pattern of the occipitocervical segments, radiographic diagnosis in infants can be difficult. The pediatric spine in the very young is placed at increased risk of flexion, distraction, and rotational injuries because of its more horizontally oriented facet joints, incompletely formed uncovertebral joints, increased laxity of the surrounding ligaments, immature paraspinous musculature, and a higher head/body mass ratio with an upper cervical fulcrum for maximal flexion.[43] The rostral location of the cervical center of rotation in infancy renders victims of high-energy blunt trauma at increased risk for occipitocervical instability.[22,35,39]

During the last half century, normal values of the CCJ in children have been documented using plain radiography, CT scanning, and MR imaging,[4,10,11,23,25–27,43] yet errors in evaluation are not uncommon. In a limited study conducted at our institution's regional pediatric trauma unit, 37 children with cervical spine injuries were evaluated and a surprisingly high diagnostic error rate of 19% was found.[3] Most were false-negative errors (that is, fractures misread as normal) occurring in young children at the craniovertebral junction. More recently at our institution, routine spiral screening head and cervical spine CT scanning has been performed in cases of severe craniovertebral trauma.[7] Because half the children with SCI will also have an associated head injury, this examination is a cost-effective and expeditious means of evaluating both head injury and avoiding the inherent difficulty of interpreting findings derived from plain radiographs in young children. The use of MR imaging is also recommended when neurological and radiological findings do not correlate.[3,23]

Treatment Options and Pitfalls

In older children[28,40] and adults,[13] urgent occipitocervical fusion with rigid internal fixation is recommended after AOD or AAI. In children who have sustained cervical spine injury, instrumentation-augmented arthrodesis has become increasingly common.[16] In the very young, however, this strategy carries significant perioperative and long-term risks.

In infants and very young children, the upper cervical spine is mainly cartilaginous and unable to support rigid internal fixation devices to any significant degree. Onlay bone graft techniques have been proposed as a viable alternative in infants, but postoperative and long-term morbidity rates can be significant. Acquired subaxial instability[24] and degenerative changes,[32] abnormal curvature of the spine or the so-called crankshaft phenomenon,[15,38] growth disturbance,[34] and impairment of mobility are well-recognized phenomena that may occur after occipitocervical fusion in children. We suspect the risk of these complications is probably higher in infants and the very young in whom the skeleton is even more immature.

Based on our three cases, nonoperative management is a reasonable alternative to surgical intervention in children younger than 3 years of age after traumatic cervical injury. Each case was managed with cast or halo immobilization, and there was no associated morbidity nor a need for subsequent fusion in two of the three cases by the 72-month follow-up examination. Neurological status significantly improved compared with posttraumatic baseline level in all cases. All three patients also regained satisfactory atlantooccipital stability (Table 2) as demonstrated by dynamic radiographic studies and physician-supervised traction tests. This may imply that healing of the CCJ occurred with fibrous or osseous ankylosis. In contrast, the transverse atlantal ligament healed insufficiently in two of three patients. The patient in Case 3 underwent fusion 2 years postinjury when her neurological status had stabilized and her spine had matured from infancy. With persistent widening of the ADI in the patient in Case 1, fusion may become necessary. Nevertheless, the time interval between injury and fusion will have allowed further development of her immature spine.

Despite the increasing trend toward undertaking fusion in children after trauma, in a recent series of 102 cases involving cervical spine injuries in children, Eleraky, et al.,[16] described 72 patients who were successfully managed by immobilization therapy alone. Their results supported the opinion held by Osenbach and Menezes[35] who advocated halo immobilization to treat upper cervical spine injuries in children. Although halo brace therapy is deemed a safe option in children as young as 12 months of age,[33] complications, including pin site–related infection, loosening, and vest excoriation, are not uncommon, warranting close monitoring and care of the pin site.[5,14] We further emphasize the need for vigilance in the examination of serial cervical radiographs to ensure that anatomical reduction has been maintained during the healing process, because movement is possible after initial halo placement. In the infant in Case 3, we stabilized the spine by using a tailored body cast rather than a halo vest, which was impractical in a child this young.

Conclusions

We have reported on three children younger than 3 years of age who presented with severe traumatic occipitocervical instability; this subpopulation of pediatric patients represents diagnostic and treatment challenges not encountered in older children and adults with similar injury. The unique radiographic and biomechanical features of the CCJ in the very young warrant special consideration when these patients present with head or neck injuries. The clinician must maintain a high index of suspicion for occipitoatlantoaxial injury, and we recommend CT and MR imaging to rule out osseous, ligamentous, or neural damage if AOD or AAI is suspected based on the history or mechanism of injury. Once the diagnosis of occipitocervical instability is established, prompt immobilization in a properly fitting halo vest or body cast is indicated; the patient should undergo surveillance cervical radiography to ensure stable alignment. Immobilization may obviate the need for urgent operative intervention and its attendant complications in this unique age group.

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