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Diagnosis and treatment of craniocervical dislocation in a series of 17 consecutive survivors during an 8-year period

Carlo Bellabarba, M.D., Sohail K. Mirza, M.D., M.P.H., G. Alexander West, M.D., Ph.D., Frederick A. Mann, M.D., Andrew T. Dailey, M.D., David W. Newell, M.D., and Jens R. Chapman, M.D.

Departments of Orthopaedics and Sports Medicine, Neurological Surgery, and Radiology, Harborview Medical Center, University of Washington School of Medicine, Seattle, Washington; and Department of Neurological Surgery, Oregon Health and Science University, Portland, Oregon

Object. Craniocervical dissociation (CCD) is a highly unstable and usually fatal injury resulting from osseoligamentous disruption between the occiput and C-2. The purpose of this study was to elucidate systematic factors associated with delays in diagnosing and treating this life-threatening condition and to introduce an injury-severity classification with therapeutic implications.

Methods. In a retrospective evaluation of institutional databases, the authors reviewed medical records and original images obtained in 17 consecutive surviving patients with CCD treated between 1994 and 2002. Images and clinical results of treatment were evaluated, emphasizing the timing of diagnosis, clinical effect of delayed diagnosis, potential clinical or imaging warning signs, and response to treatment.

Craniocervical dissociation was identified or suspected on the initial lateral cervical spine radiograph acquired in two patients (12%) and was diagnosed based on screening computerized tomography findings in two additional patients (12%). A retrospective review of initial lateral x-ray films showed an abnormal dens–basion interval in 16 patients (94%). The 2-day average delay in diagnosis was associated with profound neurological deterioration in five patients (29%). Neurological status declined in one patient after a fixation procedure was performed. There were no cases of craniocervical pseudarthrosis or hardware failure during a mean 26-month follow-up period. The mean American Spinal Injury Association (ASIA) motor score of 50 improved to 79, and the number of patients with useful motor function (ASIA Grade D or E) increased from seven (41%) preoperatively to 13 (76%) postoperatively.

Conclusions. The diagnosis of CCD was frequently delayed, and the delay was associated with an increased likelihood of neurological deterioration. Early diagnosis and spinal stabilization protected against worsening spinal cord injury.

Key Words • **spinal cord injury** • **craniocervical dislocation** • **fracture** • **atlantooccipital joint** • **atlantoaxial joint** • **tetraplegia**

Although acute traumatic osseoligamentous instability at the CCJ is usually fatal, improvement in extricating victims from accident sites and better emergency management techniques have increased the rate of survival in patients being transferred to the hospital.[22,24,26,30,31,33,35,36,47,54,62,63,66,67,79,81,87–89] A delay in diagnosis can have potentially devastating consequences. Unfortunately, an accurate diagnosis is frequently not established at the time of initial evaluation. Causes of missed diagnosis are manifold, and include low clinical suspicion, the presence of multiple life-threatening injuries, and difficulty and inexperience in delineating CCJ relationships on conventional radiography.[1,11,13,14,19,20,34,37,52] Thus, our goal was to evaluate potentially correctable contributory causes of delayed diagnosis of CCD while undertaking a comprehensive evaluation of patients who underwent posterior instrumentation-augmented occipitocervical fusion.

Abbreviations used in this paper: ASIA = American Spinal Injury Association; ATLS = advanced trauma life support; BAI = basion–axial interval; BDI = basion–dens interval; CCD = craniocervical dislocation; CCJ = craniocervical junction; CT = computerized tomography; HMC = Harborview Medical Center; MR = magnetic resonance; MVA = motor vehicle accident; SCI = spinal cord injury.

Clinical Material and Methods

Data Collection

We conducted a retrospective study of all survivors of CCD treated at HMC between 1994 and 2002. Following institutional review board approval, 17 consecutive cases were identified using the HMC trauma registry, University of Washington spine registry, and Northwest Regional

Spinal Cord Injury System. Prospectively collected data from these three registries, all medical records, and all spinal imaging studies were retrospectively reviewed to identify the following variables: 1) the timing of and manner in which the diagnosis of CCD was established; 2) the effect of delayed diagnosis on neurological function, including the timing of and circumstances surrounding any incident of worsened neurological status; 3) the diagnostic reliability of the lateral cervical radiography; 4) the clinical and imaging characteristics that might serve as potential harbingers of craniocervical instability, such as the mechanism of injury, associated injuries, and neurological deficit patterns; 5) surgery- and nonsurgery-related complications; and 6) neurological outcome. A delay in diagnosis was defined as the failure to identify or suspect CCD after completing the ATLS-mediated spinal evaluation,[2] typically based on the acquisition and examination of a lateral plain radiograph and CT scan of the cervical spine.

Patient Population

Table 1 provides a summary of data acquired in the 17 patients. In all cases the patients sustained high-energy injuries in an MVA (11 occupants and three pedestrians) or fall from a height (three patients). Two patients (12%) exhibited normal sensorimotor function of the extremities, and 15 (88%) suffered complete (two) or incomplete (13) SCI. In 10 patients the SCI was severe enough to compromise useful motor function (ASIA[3] Grades A–C). Functional motor activity was maintained in seven patients (ASIA Grade D or E). Although the CCJ complex can be viewed as extending from the occiput to C-2, for illustrative purposes we categorized these distractive injuries as affecting primarily the occipitoatlantal joint, the atlantoaxial joint, or both joints to an equivalent degree (Table 2). Nondistractive C1–2 injuries were excluded. Associated injuries were present in all patients (Table 3).

Evaluation Parameters

On arrival at HMC, patients were evaluated according to standard ATLS protocol.[2] A cross-table lateral conventional radiograph of the spine was obtained as part of the initial trauma evaluation. Head and cervical CT scans (occiput–T3) were obtained in all patients because of the high-energy mechanism of their injuries.[43] Radiographic screening of the CCJ included the use of Harris lines,[45,46] the Wackenheim line,[82,83] and the Powers ratio.[67] Nine patients were seen at other hospitals first and may have been initially treated according to a different algorithm; after transfer to HMC, they were reevaluated as acute trauma patients in the aforementioned manner.

Provisional Craniocervical Stabilization

Once CCD was identified or suspected, we secured provisional stabilization by taping the patient's head to adjacent sandbags on a backboard and, if tolerated, placing the patient in the reverse Trendelenberg position to counteract distractive forces until a halo vest assembly could be applied. Manual reduction and halo vest application under fluoroscopic guidance were used to achieve emergent closed reduction, when clinically appropriate (Fig. 1). Cervical spine MR imaging was then performed to determine the extent of the ligamentous disruption and SCI. Patients in whom there was MR imaging evidence of craniocervical osseoligamentous injury and grossly preserved alignment (≤ 2 mm displacement) and whose craniocervical stability remained uncertain underwent trac-

TABLE 1

*Summary of initial data obtained in 17 patients surviving a high-impact craniocervical injury**

Case No.	Age (yrs), Sex	Injury Mech	Ejected from Vehicle	Death at Scene†	GCS Score	Revised Trauma Score	LOC	CHI	Time to Diagnosis (days)	FU (mos)	ASIA Grade (motor score)
1	12, M	MVA	yes	yes	7	6	yes	yes	1	12	E (100)
2	17, M	MVA	no	yes	9	10	no	yes	0	14	D (82)
3	17, F	MVA	yes	no	15	12	yes	yes	1	7	C (6)
4	26, M	MVA	no	no	2	6	yes	yes	3	114	A (3)
5	27, F	MVA	yes	yes	6	5	yes	yes	1	4	C (18)
6	39, F	MVA	no	yes	13	12	yes	yes	2	45	D (94)
7	45, M	MVA	yes	yes	3	5	yes	yes	1	85	D (89)
8	49, F	MVA	yes	yes	3	4	yes	yes	1	26	C (4)
9	67, F	MVA	yes	no	8	10	yes	yes	1	40	C (56)
10	23, F	MVA	no	NA	15	12	yes	no	1	12	D (89)
11	22, M	MVA	no	NA	3	7	no	yes	2	33	C (26)
12	35, F	ped MV	NA	NA	3	3	yes	yes	0	16	A (0)
13	43, M	ped MV	NA	NA	3	5	yes	yes	1	10	C (48)
14	48, F	ped MV	NA	NA	3	6	yes	yes	0	82	C (10)
15	17, F	fall	NA	NA	8	12	yes	no	15	12	C (32)
16	33, M	fall	NA	NA	15	12	no	no	0	16	E (94)
17	84, F	fall	NA	NA	15	12	no	no	1	6	E (100)
mean	35 (10 F, 7 M)		NA	NA	8	9	13 yes, 4 no	13 yes, 4 no	2	26	(50)

* CHI = closed head injury; FU = follow up; GCS = Glascow Coma Scale; LOC = loss of consciousness; Mech = mechanism; NA = not applicable; ped MV = pedestrian struck by motor vehicle.

† In this column, "yes" indicates that there were other occupants who died at the scene; "no," that there were other occupants but none died; and "NA," that the driver was the only occupant.

TABLE 2

*Summary of craniocervical injury patterns in 17 patients with CCD**

Injury Pattern	No. of Patients (%)
primarily occiput–C1 distraction	7 (41)
occiput–C1 & C1–2 distraction	6 (35)
primarily C1–2 distraction	4 (23)
associated increased ADI	5 (29)

* ADI = atlas–dens interval.

tion tests performed by a spine surgeon who used markers or reference points of known dimensions to evaluate the extent of provocative distraction (Fig. 2).

Operative Technique

Definitive management of CCD involving rigid occipitocervical instrumentation–augmented arthrodesis was performed as soon as the physiological condition of patients permitted (Fig. 3). Surgical management was performed in the following sequence. We inserted a fiberoptic tube into the patient who was awake and in the supine position; baseline somatosensory evoked potentials were measured. In the prone position the patient was placed on a spinal surgery operating table (Jackson Table; OSI, Union City, CA) to which a rigid halo ring was attached. Final closed reduction was established under fluoroscopic guidance. Posterior decompression was conducted as needed; rigid instrumentation titanium craniocervical plates or rod/plate devices (Synthes Spine, Paoli, PA) were used to fixate the affected segments. Arthrodesis was undertaken by implanted bone graft secured to the occiput and cervical spine as previously described.[85] Either a tricortical posterior iliac crest autologous graft or structural distal femoral allograft was used, the choice being based primarily on the surgeon's preference. Occasionally, the perceived need to expedite the procedure and minimize operative time, dissection, and blood loss due to the patient's physiological status also influenced the decision to use allograft. Postoperative immobilization was achieved with a brace or halo vest based on the surgeon's degree of confidence in the strength of their fixation. Postoperative multiplanar reformatted CT scans were obtained to assess alignment and the adequacy of hardware placement. At 3 months postoperatively, upright flexion–extension lateral radiography was used to evaluate healing in all patients without external immobilization. Clinical follow-up data were obtained using the Northwest Regional Spinal Cord Injury System.

Results

Diagnosis of CCD

In four (23%) of 17 patients a diagnosis of CCD was established during the initial trauma evaluation. In two patients injuries were detected on the initial lateral cervical radiographs. In the other two patients diagnosis was made based on the cervical CT evidence. Neurological function did not deteriorate in any of these four patients before they underwent operative stabilization.

TABLE 3

*Summary of associated injuries**

Associated Injury	No. of Patients (%)
overall	
CHI w/ intracranial hemorrhage	13 (76)
craniofacial injury	12 (71)
musculoskeletal injury (nonspine)	11 (65)
thoracic injury	9 (53)
spinal fracture†	7 (41)
abdominal injury	6 (35)
associated spinal injury	
occipital condyle alar ligament avulsion	7 (41)
C-2 fracture	7 (41)
odontoid tip alar or apical ligament avulsion	6 (35)
C-1 fracture	4 (23)
thoracic fracture	2 (12)
C-1 transverse ligament avulsion	1 (6)
C3–7 fracture	1 (6)
lumbar fracture	1 (6)
cranial nerve injury	
V	2 (12)
VI	2 (12)
VII	2 (12)
XII	2 (12)
Horner syndrome	1 (6)

* The sum of percentages may exceed 100 because patients may belong to more than one category.

† The spinal fracture row does not include avulsion fractures of the upper cervical spine.

In 13 (76%) of 17 patients the diagnosis of CCD was delayed by a mean of 2 days (range 1–15 days). Five (38%) of these 13 patients suffered profound neurological deterioration before CCD was clinically recognized.

The specific abnormal imaging characteristics that heralded the diagnosis of CCD and the clinical circumstances that led to a focused evaluation of the CCJ are summarized in Table 4.

Operative Treatment

Surgical stabilization was undertaken in all 17 patients (Table 5). The mean follow-up duration was 26 months (range 4–114 months).

Surgery-Related Complications

One patient experienced acute worsening of his sensorimotor function postoperatively. His CCJ had been stabilized in an extended and 50% anteriorly subluxed position (Fig. 4). Hydrocephalus and a cerebellar infarction were diagnosed, presumably due to vertebrobasilar arterial insufficiency,[8,57] and the patient underwent emergency occipitocervical realignment and stabilization, posterior fossa decompression, and placement of a ventriculoperitoneal shunt. He was the only patient who required early return to the operating room after craniocervical stabilization. At the 16-month follow-up examination, the tetraplegia had completely resolved; a mildly broad-based gait was the only sign of neurological dysfunction. Another patient with an ASIA Grade C SCI experienced temporary worsening when, after being positioned prone for occipitocervical arthrodesis, a decrease in baseline somatosensory evoked potentials necessitated that the procedure be abort-

FIG. 1. Case 12. Lateral cervical radiographs documenting widely displaced Stage 3 CCD, obtained after injury *(left)*, after provisional closed reduction with a halo vest *(center)*, and internal fixation *(right)* in a patient who was struck by a motor vehicle. Even after open reduction, the occipitoatlantal articulation remained distracted and anteriorly translated *(black lines)*.

ed before the incision could be made. This patient's ASIA motor score decreased from 16 to 9, but it returned to its baseline value within 24 hours. Posterior stabilization was performed without event 2 days later.

There were no cases of occipitocervical pseudarthroses or hardware failure; however, in the patient who required occiput–T2 fusion for multilevel cervicothoracic injuries we observed a T1–2 pseudarthrosis and loosening of the caudal hardware, requiring revision of the posterior instrumentation 1 year postoperatively.

A postoperative superficial wound infection, treated with antibiotic agents and local wound care, occurred in one patient (6%). In one patient (6%), who had undergone repair of a traumatic dural tear, cerebrospinal fluid leaked from the surgical incision. This was treated successfully by insertion of a lumbar subarachnoid drain.

Neurological Outcome

The mean ASIA motor score improved from 50 preoperatively to 79 at the final follow-up examination (Table 6). Of 13 patients with incomplete SCI, impairment in 11 (85%) improved by at least one ASIA grade. Neither of the two patients who presented without SCI worsened neurologically. Of the two patients with complete SCI, improvement occurred in one such that the high cervical level deficit improved to a T-5 level, with corresponding improvement in ASIA motor score from 16 to 50. The second patient, who presented with wide craniocervical displacement and a complete SCI above the C-5 level (Fig. 1) remained dependent on the assistance of a ventilator; her ASIA motor score was 0 at the 16-month follow-

FIG. 2. Case 6. Lateral *(left)* and traction *(right)* cervical radiographs documenting mildly displaced Stage 2 CCD; the BDI is within 2 mm of normal, and only mild, unilateral right-sided loss of occipitoatlantal congruence was observed on MR imaging (not shown). The traction radiograph was used to distinguish this as a highly unstable Stage 2 injury *(black lines)* requiring internal fixation rather than a Stage 1 injury in which closed treatment would have sufficed.

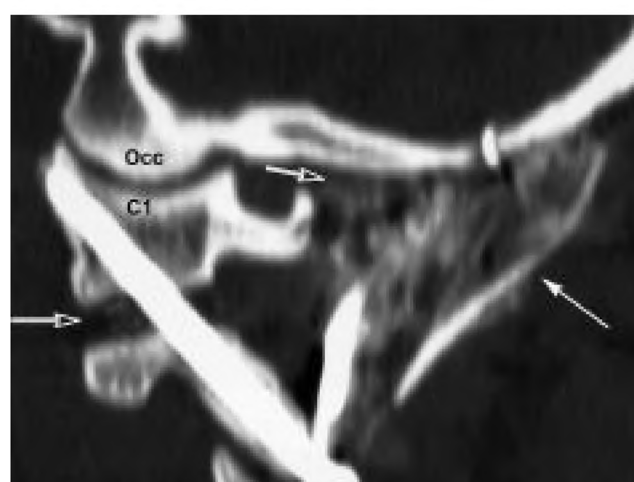

FIG. 3. Operative technique. Postoperative sagittal CT reformation illustrating congruent reduction of occipitoatlantal and atlantoaxial joints and arthrodesis in a 17-year-old patient who underwent placement of segmental occipitocervical instrumentation: C1–2 transarticular screw fixation, placement of structural bone graft material with suboccipital and C2–sublaminar fixation *(solid arrow)* and placement of autologous cancellous bone grafting material in the decorticated C1–2 joints and the CCJ *(open arrows)*. Occ = occiput.

TABLE 4

Modality used for establishing the correct diagnosis

Variable	No. of Patients (%)
diagnostic study	
initial lat cervical radiograph	2 (12)
cervical CT scan	6 (35)
FU lat cervical radiograph	5 (29)
cervical spine MRI	3 (18)
brain MRI	1 (6)
stimulus for diagnosis	
neurological deterioration	5 (29)
transferred to HMC—repeated ATLS evaluation	5 (29)
initial ATLS evaluation	4 (24)
unanticipated finding on FU radiograph	2 (12)
unexplained, nonprogressive neurological deficit	1 (6)

up visit. In the other 16 patients ASIA motor score–based function improved at follow up.

Cranial Nerve Injury

In four patients (23%) preoperative cranial nerve injuries were documented (Table 3). Each of these patients sustained head injuries and suffered intracranial hemorrhage. In three patients (17%) more than one cranial nerve was involved. The trigeminal, abducent, facial, and hypoglossal nerves[76] were each affected in two patients (12%). In one patient (6%) Horner syndrome was diagnosed. Four patients (23%) had dysphagia, an absent gag reflex, and impaired vocal cord function, which could potentially have resulted from cranial nerve dysfunction. In each of these patients a tracheostomy tube had remained in place for prolonged periods.

Imaging Results

After surgery the mean BDI improved from 17 to 11 mm, and the mean BAI decreased from 10 to 8 mm (Table 7). Whereas an abnormal BDI was present preoperatively in 16 patients, it had normalized postoperatively in all but two patients. One of these patients presented with severe distraction that could not be completely reduced, resulting in a postoperative BDI of 18 mm (Fig. 1).

Additional spinal fractures, particularly ligamentous avulsions and associated fractures of the upper cervical spine, were commonly identified (Table 3 and Fig. 5).

Discussion

Injuries to the CCJ constitute almost one fourth of all cervical injuries.[11] In autopsy studies investigators have ascribed up to 90% of traumatic fatalities to upper cervical injuries.[1,13,19,52] Based on their prospective morgue study, Bucholz and Burkhead[13] estimated a survival chance of 0.65 to 1% for patients with CCD. In the 25 years since publication of these preliminary studies, however, the emergency management of trauma victims has been dramatically improved. Accordingly, multiple case reports have documented survival after CCD.[16,22,24,26,30,31,33,35,36,47,53,54,62,63,66,67,74,79,81,87–89] The burden of appropriately anticipating, identifying, and treating the survivors of this life-threatening injury has thus shifted to the emergency department, trauma, and spine surgery teams.

Delay in Diagnosis

The previously recognized difficulties in prompt diagnosis of craniocervical instability were highlighted in this series.[11,14,20,34,37,63,69] The highly unstable nature of these lesions was either entirely unrecognized or underappreciated in more than 75% of our patients. This problem is often attributed to misleading clinical and imaging features but may be influenced by the absence of a systematic approach to imaging evaluation of the CCJ. Most of our patients suffered cognitive impairment due to loss of consciousness (13 patients [76%]) or a closed head injury with intracranial hemorrhage (13 patients [76%]), which contributed to diagnostic difficulties. Because lateral cer-

TABLE 5

*Summary of surgery-related data stratified by hardware specifications**

Case No.	Levels Fused	Implant	C1–2 Transarticular Screws	Structural Bone Graft	C-1 Laminectomy
5	Oc–C4	HOC plate	unilat	autograft	no
2	Oc–C3	HOC plate	bilat	allograft	no
3	Oc–C3	HOC plate	bilat	allograft	yes
7	Oc–C2	HOC plate	bilat	autograft	no
12	Oc–C3	HOC plate	bilat	allograft	no
13	Oc–C2	HOC plate	unilat	allograft	no
14	Oc–C2	HOC plate	bilat	autograft	no
15	Oc–C3	HOC plate	bilat	autograft	no
16	Oc–C2	HOC plate	bilat	autograft	yes
8	Oc–C2	titanium plate	bilat	autograft	no
10	Oc–C2	titanium plate	bilat	autograft	yes
17	Oc–C3	titanium plate	unilat	autograft	no
4	Oc–C4	titanium plate	NA	autograft	yes
11	Oc–C2	titanium plate	NA	autograft	no
9	Oc–T2	Cervifix rod/plate	bilat	allograft	no
1	Oc–C2	Cervifix rod/plate	bilat	allograft	no
6	Oc–C2	Cervifix rod	bilat	allograft	no

* The Harborview Occipito-Cervical (HOC) plate, titanium plate, Cervifix rod/plate, and Cervifix rod were all obtained from Synthes Spine.

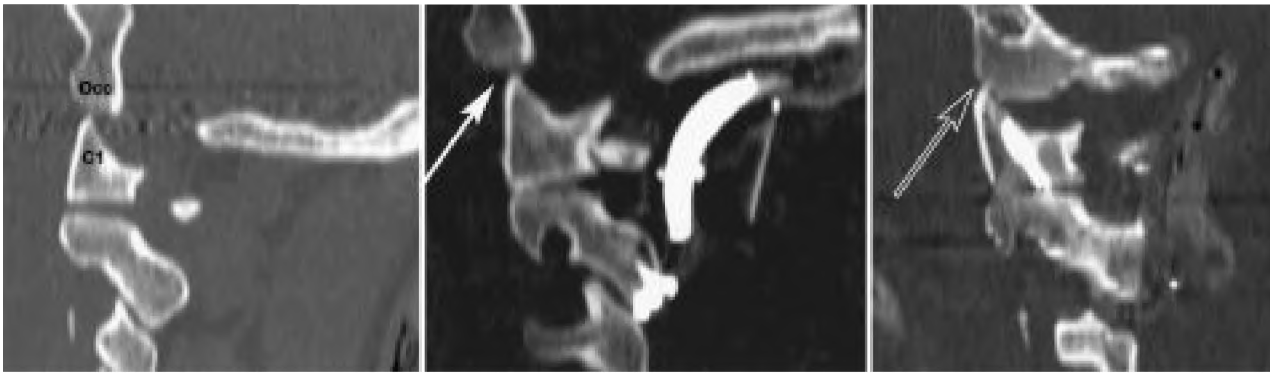

FIG. 4. Case 16. Sagittal CT reformations revealing malreduction and postoperative neurological deterioration after injury *(left)*, initial posterior craniocervical fixation *(center)*, and emergency revision craniocervical fixation *(right)* in a patient in whom CCD was caused by a 30-ft fall. The patient suffered neurological decline after internal fixation, and his postoperative imaging studies showed that the CCJ had been stabilized in an extended and anteriorly displaced position *(solid arrow)*. After repeated stabilization and improved column alignment *(open arrow)*, the patient's ASIA motor score–reflected status eventually normalized.

vical spine radiography remains key in the initial imaging evaluation of a patient with suspected cervical spine trauma,[2,61,72] the most alarming finding of our study was how infrequently craniocervical instability was suspected on standard radiographs. This finding may be related to the more subtle and diagnostically challenging injuries that occur in survivors of CCD. Based on the initial lateral radiograph, the injury was identified or suspected in only two patients. The radiographic findings were interpreted as normal for craniocervical injury in 14 patients and were declared inadequate to allow for craniocervical assessment in one patient. A retrospective review of these x-ray films, however, showed both the BDI and BAI[45,46] to be within normal parameters in only one patient.

Even a short delay in diagnosis and treatment was associated with a considerable risk of severe adverse, life-threatening consequences. Five (38%) of 13 undetected injuries were recognized when neurological deterioration was noted.[58,80] In two (15%) of the 13 patients in whom the diagnosis was delayed, the cervical spine injury had been identified without appreciating the magnitude of associated craniocervical instability (Fig. 5). Despite cervical collar immobilization and continued total-spine precautions, one patient suffered profound deterioration in spinal cord function, which highlights the inadequacy of standard brace therapy in stabilizing CCD.[10]

As with any joint dislocation, CCD may initially appear to be partially reduced. This compounds the diagnostic challenge in interpreting standard cervical radiographs because visualization of the CCJ is impeded by overlying structures and parallax.[86] This difficulty in appreciating the articular congruence at the CCJ has given rise to methods of indirect assessment using a multitude of radiographic lines,[45,46,56,67,82,83,86] most of which are limited by their complexity, lack of interobserver reliability, or directional limitations.[67] We have relied mainly on the BDI and BAI.[45,46] Despite the low diagnostic success in this series, the finding that these relationships were within normal limits in only one patient corroborates their importance.

Other pitfalls in the radiographic diagnosis of craniocervical instability can be attributed to associated fractures of the upper cervical spine (Table 3 and Fig. 5). Once a separate cervical injury had been identified, the attention given to further evaluation of the CCJ may have been less urgent or thorough. Although the ligament-related characteristics of CCD have not been completely defined, the tectorial and alar ligaments appear to be important stabilizers, and additional stabilization is provided by the apical and transverse ligaments.[84] Accordingly, alar ligament avulsions have been reported in 30 to 50% of patients with craniocervical instability.[5,6,13,52] Transverse ligament injuries are thought to occur in 36%,[32,77] some of which manifest as osseous avulsions, and there is no documentation of the incidence of apical ligament avulsion.[77] In the present study avulsion fractures were commonplace, occurring in nine patients (53%), with more than one fracture type seen in six patients (35%). The frequency with which avulsion fractures were identified (Table 3) suggests that their presence should heighten the suspicion for CCD and indicate the need for early MR imaging follow-up investigation of ligamentous integrity.

Neurological Deficits

Patients in whom SCI has been caused by upper cervical trauma may present with neurological deficits ranging from complete tetraplegia with head and neck involvement to atypical incomplete injuries, such as Bell cruciate paralysis or other variants of cervicomedullary syndromes.[9,25,55] Only two of our patients presented without SCI. The most common neurological deficit was motor dysfunction that 1) was more severe in the upper than in the lower extremities; 2) was asymmetrical, affecting one side more than the contralateral side; and 3) involved the muscles innervated by C-5, the most cranially examinable cervical motor root.

Injury Classification

Traynelis and colleagues[79] have identified three occipitocervical injury patterns that are based on the direction of displacement. Injury classification based on directional criteria, however, may be misleading because the position

TABLE 6

Summary of neurological outcome stratified by ASIA grade category

	Preop			Postop			
Case No.	ASIA Grade	Spinal Level*	ASIA Motor Score	ASIA Grade	Spinal Level*	ASIA Motor Score	Change in ASIA Motor Score
1	E	NA	100	E	NA	100	0
2	D	C-5	82	E	NA	100	18
3	C	C-5	6	D	C-5	82	76
4	A	C-5	3	A	T-5	50	47
5	C	C-5	18	D	C-5	83	65
6	D	C-5	94	E	NA	100	6
7	D	C-5	89	E	NA	100	11
8	C	C-5	4	C	C-5	39	35
9	C	C-5	56	E	C-5	100	44
10	D	C-5	89	E	NA	98	9
11	C	C-5	26	D	L-1	85	59
12	A	C-5	0	A	C-5	0	0
13	C	C-5	48	D	C-5	70	12
14	C	C-5	10	D	C-5	95	85
15	C	C-5	32	C	C-5	46	14
16	D	C-5	94	E	NA	100	6
17	E	NA	100	E	NA	100	0
mean			50			79	29

* Here C-5 reflects the highest possible examinable motor level.

of the head in relation to the spine is arbitrary given the total ligamentous disruption and severe instability fundamentally caused by these injuries.[51] Furthermore, this injury classification does not convey the severity of the injury nor does it address spontaneously repositioned injuries or rotatory injuries such as unilateral alar ligament disruptions.[5,27–29] The classification proposed by Traynelis and colleagues was of little use in the present series, in which all CCDs were a combination of distraction and anterior subluxation. Based on our clinical experience, we propose a three-stage classification system with therapeutic implications (Table 8).[15] A Stage 1 injury is defined as a stable minimally or nondisplaced craniocervical injury in which there is sufficient preservation of ligamentous integrity to allow for nonoperative treatment; such injury types include unilateral alar ligament avulsion or a partial ligamentous injury (or sprain), any of which would be documented on MR imaging.[5,23,44] A Stage 2 injury represents a partially or completely spontaneously reduced bilateral CCD involving minimal displacement (BAI/BDI $\leq$ 2 mm beyond the upper limit of normal) in which a positive traction test confirms a complete loss of craniocervical ligamentous integrity, requiring internal fixation.[7,12,19,71] A Stage 3 injury denotes a highly unstable injury defined by gross craniocervical malalignment (BAI/BDI $>$ 2 mm beyond acceptable limits), also requiring internal fixation. Although these represent a spectrum of craniocervical injury severity, we reserve the term "craniocervical dissociation" for Stage 2 and 3 injuries, where ligamentous instability is complete. According to this classification system, four Stage 2 and 13 Stage 3 injuries were documented in survivors of CCD at our institution during an 8-year period.

Surgical Stabilization

A delay in surgical stabilization was associated with a greater neurological risk than early stabilization. In five patients (29%) neurological deterioration occurred between the time of injury and stabilization. Although fracture displacement was reduced and stabilized by immediate application of a halo vest or by postural changes, these methods were entirely provisional because external immobilizaton does not appear to effectively stabilize Stages 2 and 3 craniocervical injuries. Although we found that halo vest immobilization was the better alternative

TABLE 7

Comparison of pre- and postoperative BDI and BAI measurements

	Measurement (mm)			
	Preop		Postop	
Case No.	BDI	BAI	BDI	BAI
1	15	16	8	2
2*	16	14	10	10
3	16	18	11	8
4	22	15	11	8
5	21	16	11	9
6	14	11	9	8
7	15	15	9	10
8	18	10	10	6
9	10	11	12	8
10	15	9	10	6
11	18	15	14	10
12*	26	14	18	10
13	16	13	12	10
14*	17	14	10	4
15	15	14	12	8
16*	13	12	9	11
17	17	13	9	6
average	17	14	11	8
no. abnormal (%)†	16 (94)	12 (71)	2 (12)	0

* Diagnosis was not delayed in these cases.

† Intervals greater than 12 mm represent abnormal BDIs and BAIs.

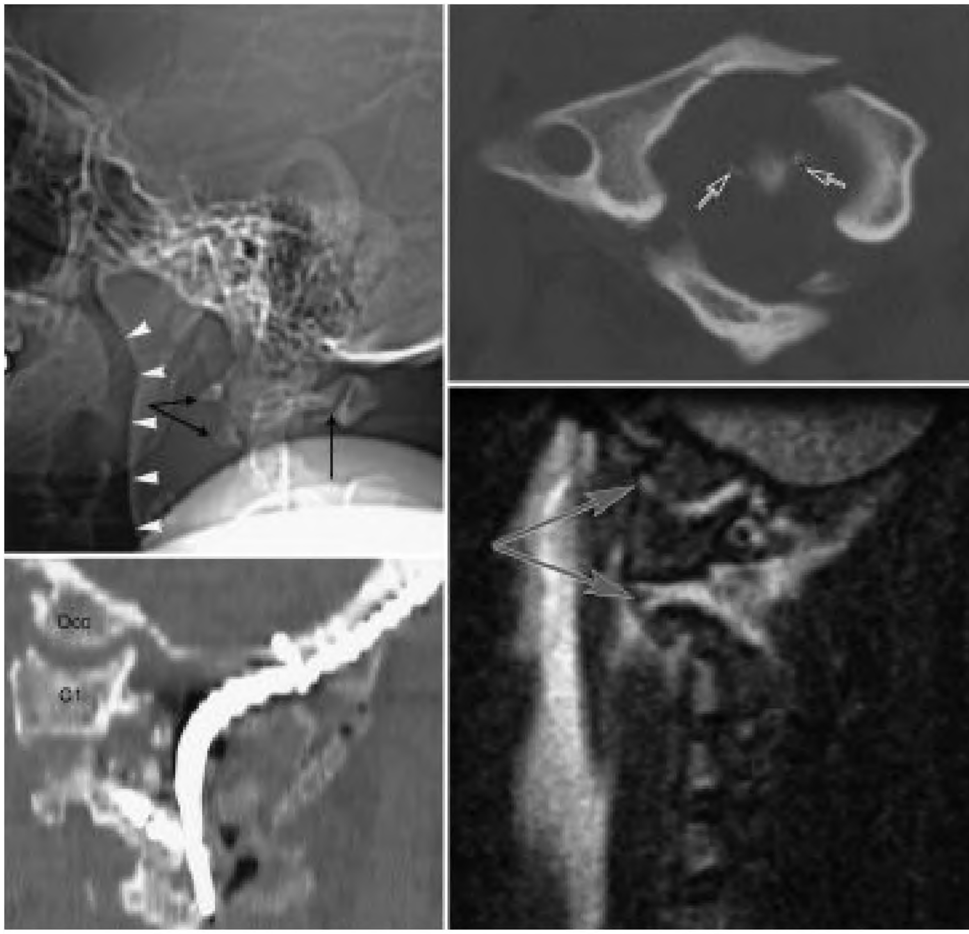

FIG. 5. Case 3. Imaging studies documenting neurological decline attendant on a delayed diagnosis: lateral x-ray study (*upper left*), axial CT (*upper right*), sagittal CT reconstruction (*lower left*), and MR image (*lower right*) obtained in a patient involved in an MVA who presented with neck pain and primary right arm and leg weakness. The C-1 fracture (*black arrows [upper left]*) was erroneously identified as a Jefferson-type fracture. Widening of the ADI and the significance of bilateral alar ligament avulsions off the odontoid tip (*open arrows [upper right]*) were underappreciated. Note the anterior soft-tissue swelling (*white arrowheads [upper left]*). During transfer to the MR imaging unit for additional evaluation, she suffered cardiorespiratory arrest and lost all but trace movement in her extremities despite cervical collar immobilization. Craniocervical dislocation involving the occipitoatlantal and atlantoaxial articulations was confirmed on MR imaging (*gray arrows [lower right]*). The patient underwent emergency decompression and stabilization (*lower left*) and injury status improved to an ASIA Grade D and motor function to a score of 82.

pending internal fixation, it occasionally accentuated the distractive deformity (Fig. 6). More definitive stabilization options include the placement of occipitocervical bone grafts and wire-based hardware;[18,39,42,60,64,79] posterior fixation involving contoured structural rods with suboccipital and sublaminar wires;[17,49,68] and posterior segmental instrumentation.[4,41,59,70,73,75,78,85] Transarticular screws implanted across the occipitoatlantal joint[38,40] and anterior fixation via an extrapharyngeal approach[21] have also been advocated. In our experience, patients with CCD benefit from rigid posterior segmental stabilization in which the hardware extends from the occiput to at least C-2, preferably involving transarticular C1–2 screw fixation.[4,48,50,65,73] Because the primary craniocervical ligamentous stabilizers extend from the occiput to C-2 and essentially bypass C-1, as a general rule we extend the hardware from the occiput to at least C-2 in all cases of CCD, even if the distraction occurs primarily at the occipitoatlantal joint. These rigid constructs maintained the reduction without a single instance of craniocervical pseudarthrosis.

TABLE 8
Proposed classification for craniocervical injuries

Stage	Description of Injury
1	MRI evidence of injury to craniocervical osseoligamentous stabilizers; craniocervical alignment w/i 2 mm of normal; distraction of ≤2 mm on provocative traction radiography.
2*	MRI evidence of injury to craniocervical osseoligamentous stabilizers; craniocervical alignment w/i 2 mm of normal; distraction of >2 mm on provocative traction radiography.
3*	craniocervical malalignment of >2 mm on static radiography.

* Represents an injury defined as CCD.

FIG. 6. Pitfalls of closed provisional stabilization of CCD. *Left* and *Center*: Lateral cervical radiographs showing a Stage 3 CCD before *(left)* and after *(center)* application of a halo vest demonstrate how application of a halo vest may result in worsened craniocervical distraction. The radiograph on the *left* also illustrates the appropriate use of Harris lines, with an abnormal BDI of 15 mm and a normal BAI of 9 mm. The BDI is measured from the basion to the tip of the dens and is 12 mm or smaller in most patients; the BAI is measured from the basion to the posterior axial line, a line that extends along the posterior cortex of the body of the axis (but not necessarily of the dens, which might be angled with respect to the axis). The BAI ranges from −6 to 12 mm in most patients. Any BDI or BAI measurement outside the normal range requires further imaging studies to evaluate for the presence of CCD. *Right:* Schematic illustration of Harris lines.

Pattern of Associated Injuries and Injury Mechanisms

We attempted to identify variables that might serve as warning signs of craniocervical instability. All patients were involved in high-energy injuries affecting an average of three organ systems (Table 3). Of nine patients involved in motor vehicle collisions in which there was more than one person, at least one death occurred at the scene in six cases (67%). Facial or skull injuries were present in 12 patients (71%). On initial evaluation neurological deficits were identified in 15 patients (88%); these were most commonly (76%) ASIA Grade C or D deficits with asymmetrical motor deficits involving the C-5 level. The polytraumatized patient with intracranial hemorrhage and obvious injury to the face or skull is at risk particularly when neurological dysfunction involves high cervical levels and exhibits side-to-side asymmetry.

Measures Taken to Reduce Missed Injuries

Because of the potentially devastating consequences of failing to diagnose and treat CCD injuries promptly, several measures have been instituted at our center to recognize them as early as possible. These measures have focused on educating healthcare personnel at all levels about the importance of careful imaging evaluation of the CCJ in all trauma patients, the identification of clinical clues (as described in the previous section) that suggest the presence of a high cervical cord injury, and the vigilance required—even once the diagnosis has been made—to avoid secondary neurological deterioration in patients with this severe instability pattern. A fundamental aspect of this approach is to educate all participants who are involved in the diagnosis and treatment of trauma patients about these issues to establish as many layers of redundancy as possible. This approach allows, for example, for the screening lateral cervical radiographs and cervical CT scans to be scrutinized by the radiology, emergency department, general surgery, orthopedic, and neurosurgery teams, increasing the likelihood that one of these clinicians might identify injuries that may have been less obvious to others. Both the rarity of these injuries and the frequent turnover of personnel that is typical at an academic trauma institution require that the educational process be repeated frequently, through various educational conferences, quality-improvement forums, and continual individual instruction, to maintain the appropriate level of vigilance and prevent complacency during the relatively long interval between cases. Once a CCD is diagnosed, our protocol for first provisional and then definitive treatment (see *Clinical Material and Methods*) should be implemented.

Summary of Points

A high index of suspicion is important for prompt diagnosis and stabilization of craniocervical instability. Of the 17 survivors of CCD in our series, a delay in diagnosis either at our institution or the transferring hospital was demonstrated in 13 cases. Based on the findings in our study, a disciplined, systematic approach to the evaluation of screening cervical radiographs and adjunctive imaging studies offers the highest potential for identifying most injuries. In addition to its therapeutic implications, a classification system that includes a category for partially reduced yet highly unstable injuries may heighten awareness that even a life-threatening injury such as CCD may present with a misleadingly subtle imaging appearance. Atypical patterns of neurological dysfunction, particularly with asymmetrical proximal upper-extremity involvement, should raise suspicions about cervicomedullary syndromes. Additional research efforts are needed to establish stability criteria and assessment protocols for the CCJ. Within the limitations of a retrospective and purely observational study in which a control group could not obviously be established, we found that delay in diagnosis places the patient at risk for neurological worsening. If a timely diagnosis is made and internal occipitocervical fixation performed, even patients with severe neurological deficits may experience significant functional improvement.

Conclusions

Despite abnormalities in the BDI and/or BAI in all but one patient in our series, CCD was rarely identified on initial lateral cervical radiography, and it frequently remained undiagnosed after completion of the trauma evaluation. Even short delays in diagnosis of CCD may have severe adverse neurological consequences. Because the severity of craniocervical instability is more relevant than the arbitrary direction of displacement, classification systems should be developed accordingly. An occipitocervical fusion procedure is neuroprotective, provides the stable environment necessary for neurological recovery, and should be performed as early as safely possible in these polytraumatized patients.

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Address reprint requests to: Carlo Bellabarba, M.D., Department of Orthopaedics and Sports Medicine, Harborview Medical Center, 325 Ninth Avenue, Box 359798, Seattle, Washington 98104. email: cbella@u.washington.edu.