# Craniovertebral Junction Injury: Clinical Presentation and Outcomes of Surgical Treatment

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***Background:*** *Craniovertebral junction (CVJ) injury is an uncommon entity of vertebral trauma, which can lead to lethal outcome or permanent neurological morbidity caused by associated spinal cord injury (SCI).*
***Objective:*** *To study clinical presentation and surgical outcomes of patients with CVJ injury.*
***Material and Method:*** *All patients with traumatic CVJ who underwent surgical treatment at Siriraj Hospital from 1997 to 2007 were studied. Various types of operation were selected based on pattern of spinal instability, location and severity of spinal cord compression, as well as underlying spinal disorder. Clinical outcomes were evaluated by the Functional Independence Measure (FIM) score.*
***Results:*** *Twenty-five patients were divided into 2 groups; 18 patients were in the group of acute (early) presentation and 7 in the group of chronic (late) presentation. The most common type of injury was C1-2 subluxation followed by odontoid fracture. Four patients in the acute group had associated traumatic brain injury (TBI). Posterior cervical fixation using various types of instrumentation was done in all patients. Good outcome (the FIM score of 4 or more) were found in 22 patients. On the contrary, poor outcome (the FIM score of 1 or complete dependence) was found in the remaining 3 patients; all of them were in the acute group. Two of them had associated TBI and the other was a patient with Klippel-Feil syndrome manifested by quadriplegia due to severe SCI.*
***Conclusion:*** *The surgical treatment of CVJ injury renders satisfactory result in most patients. Factors which may influence the outcomes included associated TBI and severity of SCI.*

***Keywords:*** *Craniovertebral junction injury, Cervical spine injury, Spinal cord injury*

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The craniovertebral junction (CVJ) is a unique spinal region. It presents a challenge to surgeons to operate on, due to its bony and articular complexity, biomechanics, as well as adjacent neural and vascular structures. Common causes of pathological condition in this region include trauma, rheumatoid arthritis, infection and neoplasm. Results of these processes are spinal stenosis, neural compression, instability and deformity. Traumatic CVJ has a life-threatening consequence. Clinical presentation of traumatic CVJ is varied, usually fatal[1,2]. Improvement in prehospital management increases number of surviving patients who are transferred to hospitals[3]. Patients may present with neck pain, various degree of neurological deficits, lower cranial nerve dysfunction, or instability of the craniovertebral area. Standard guideline of trauma care for preventing further deterioration and accurate diagnosis are established. Appropriate management in these patients is crucial for their recovery and quality of life. In the present article, we present a retrospectively study of particular patients with CVJ injury, who underwent surgical treatment at Siriraj Hospital during 10-year period.

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## Material and Method

### *Data collection*

After approval of the study from the institutional review board, we retrospectively collected data of all patients who were diagnosed CVJ injury and treated by surgery at Siriraj Hospital between 1997 and 2007. Twenty-five patients were identified from the database. All medical records and radiographic studies were reviewed to collect demographic characteristic, clinical presentation, classification of injury, surgical

management, and clinical outcome.

***Patient population***

The patients were divided into two groups, including the group with acute (early) and chronic (late or delayed) presentation. Those, who were treated immediately after their injuries, were in the acute group. The chronic group consisted of patients with undiagnosed CVJ injury and patients who came to medical attention lately.

***Neuroimaging evaluation***

All patients were evaluated with plain radiographs of the cervical spine, including lateral-cross table, anteroposterior, and open mouth views. Cervical spine computerized tomography (CT) scan was done for better visualization of bony structures, and for definite classification of fracture/dislocation. Magnetic resonance imaging (MRI) of the cervical spine was performed when patients were found to have neurological deficit, or was done to identify associated ligamentous injury.

***Surgical management***

Surgical options in all patients were selected by the senior neurosurgeons. Decompression technique was chosen from compression site and surgeon's preference. Regarding fixation technique and instrumentation, contoured rods with wiring were used in the early period of this study. Later, we used more rigid system, screw and rod fixation, for stabilization. All patients were followed at the outpatient department for clinical and radiological outcomes. Postoperative radiograph was done periodically to determine fusion and alignment. They were instructed to wear the appropriate cervical orthosis, until bony fusion was evident on postoperative radiograph. The Functional Independence Measure (FIM) score was utilized to evaluate the results.

**Results**

There were 25 patients, including 21 males and 4 females. Patients' age ranged from 2 to 80 years. Eighteen patients (72%) were transferred to hospitals immediately after the accident, so they were in the acute group. The remaining 7 (28%) were in the chronic group. The major cause of the injury was motor vehicle accident, accounting for 15 of 25 (60%). The remaining 10 (40%) had CVJ injury due to falling from height. In the acute group, 8 of 18 patients (44%) presented with only neck pain without neurological deficit. Four patients in the acute group had associated traumatic brain injury (TBI). In the chronic group, two of seven patients presented with only neck pain. Five patients had spinal cord injury (SCI) with neurological deficit.

Of all patients, the most common type of CVJ injury was atlanto-axial dislocation found in 13 patients, followed by odontoid fracture in 10 patients. Also, there was one patient with atlas fracture and one with hangman's fracture. Three patients had pre-existing congenital spinal disorders, including one with Chiari I malformation, one with achondroplasia, and one with Klippel-Feil syndrome.

Regarding surgical treatment, 4 of 25 patients required spinal decompression with posterior fixation. A 33-year-old female with odontoid fracture after falling, was found to have Chiari I malformation. She underwent foramen magnum decompression, and C1-2 posterior sublaminar wiring. One patient with history of falling from height had os odontoideum which required transoral decompression to remove the ventral pannus. It was followed by C1-2 fixation using transarticular screws. Posterior decompression was done in three patients. A 3 years old girl with achondroplasia, who sustained C1 fracture with atlanto-axial dislocation, underwent posterior foramen magnum decompression with C1-2 fixation. Twenty patients underwent posterior fusion without spinal decompression. Occipito-cervical fixation using contoured rods and wiring were used in 9 patients. C1-2 sublaminar wiring was performed in 6 patients. Seven patients underwent C1-2 transarticular screw fixation, and 3 patients underwent C1-2 lateral mass screw and rod fixation.

Three patients had poor outcome with the FIM score of 1 (complete dependence with total assistance). Two of them also suffered from associated TBI. Twenty-two patients (88%) showed satisfactory outcome with the FIM score of 4 (function with minimal assistance) or better. All of 8 patients in the acute group presenting with neck pain without neurological deficit had excellent outcome with the FIM score of 7 (complete independence). Demographic characteristics, surgical treatments and outcomes are presented in Table 1.

Of all cases, no evidence of instrumentation failure was found on postoperative radiographic studies.

**Discussion**

Although craniovertebral injury is relatively uncommon spinal injury, it typically results in SCI or spinal instability. High index of suspicion in diagnosis and prompt treatment are crucial to prevent further

**Table 1.** Demographic characteristics, surgical treatments and outcomes

| Gender/age (years) | Mechanism of injury | Timing | Clinical presentation | Diagnosis | Underlying spinal disorder | Surgical treatment | FIM score |
|---|---|---|---|---|---|---|---|
| M 13 | MVA | Acute | SCI, TBI | C1-2 subluxation | - | OC fusion | 5 |
| M 38 | Falling | Chronic | SCI | C1-2 subluxation | - | OC fusion | 4 |
| F 33 | Falling | Acute | SCI, mild | Odontoid fracture | Chiari I malformation | Foramen magnum decompression, C1-2 wiring | 6 |
| M 51 | MVA | Chronic | Neck pain | C1-2 subluxation | - | OC fusion | 7 |
| M 63 | MVA | Acute | Neck pain | C1-2 subluxation | - | C1-2 sublaminar wiring | 7 |
| F 28 | Falling | Acute | Neck pain | Odontoid fracture | - | OC fusion | 7 |
| M 52 | Falling | Chronic | SCI, mild | Os odontoideum, C1-2 subluxation | - | Transoral decompression, C1-2 transarticular screw fixation | 6 |
| M 24 | MVA | Acute | Neck pain | Odontoid fracture | - | C1-2 sublaminar wiring | 7 |
| M 27 | MVA | Acute | Neck pain | Hangman's fracture | - | OC fusion | 7 |
| M 52 | Falling | Chronic | SCI, mild | Os odontoideum, C1-2 subluxation | - | C1 laminectomy C1-2 lateral mass screw fixation | 6 |
| M 42 | MVA | Acute | Neck pain | Odontoid fracture | - | C1-2 lateral mass screw fixation | 7 |
| M 41 | MVA | Acute | Neck pain | C1 fracture | - | OC fusion | 7 |
| M 20 | MVA | Acute | Neck pain | Odontoid fracture | - | C1-2 transarticular screw fixation | 7 |
| M 29 | MVA | Acute | SCI | C1-2 subluxation | - | C1-2 sublaminar wiring | 7 |
| F 3 | Falling | Acute | SCI | C1 fracture, C1-2 subluxation | Achondroplasia | Foramen magnum decompression, C1-2 transarticular screw fixation | 6 |
| F 59 | MVA | Acute | SCI | Odontoid fracture | - | C1-2 transarticular screw fixation | 6 |
| M 79 | MVA | Acute | SCI | Odontoid fracture | - | Posterior decompression, C1-2 lateral mass screw fixation | 5 |
| M 53 | Falling | Acute | SCI | C1-2 subluxation | Klippel-Feil syndrome | OC fusion | 1 |
| M 69 | MVA | Acute | SCI, TBI | Odontoid fracture | - | OC fusion | 1 |
| M 33 | Falling | Acute | SCI, TBI | Odontoid fracture | - | OC fusion | 1 |
| M 41 | Falling | Acute | SCI, TBI | C1-2 subluxation | - | C1-2 sublaminar wiring | 6 |
| M 26 | MVA | Acute | Neck pain | Odontoid fracture | - | C1-2 transarticular screw fixation | 7 |
| M 28 | MVA | Chronic | Neck pain, torticollis | C1-2 subluxation | - | C1-2 transarticular screw fixation | 7 |
| M 2 | MVA | Chronic | SCI | C1-2 subluxation | - | C1-2 sublaminar wiring | 4 |
| M 80 | Falling | Chronic | SCI | C1-2 subluxation | - | C1-2 transarticular screw fixation | 5 |

C = cervical; FIM = Functional Independence Measure; MVA = motor vehicle accident; OC = occipitocervical; SCI = spinal cord injury; TBI = traumatic brain injury
FIM score description as the follows; Independence: 7 = complete independen ce; 6 = modified independence (patient requires use of a device, but no physical assistance). Modified dependence: 5 = supervision or setup; 4 = minimal contact assistance (patient can perform 75% or more of task); 3 = moderate assistance (patient can perform 50% to 74% of task), Complete dependence: 2 = maximal assistance (patient can perform 25% to 49% of task); 1 = total assistance (patient can perform less than 25% of the task or requires more than one person to assist)

injury and maximize treatment outcomes. In the present study, 8 of 18 patients (44%) who arrived hospitals right after the injury, presented with only neck pain without neurological deficit. The diagnosis of CVJ injury was established based on findings on radiographic investigation, as per advanced trauma guideline. Accordingly, unstable craniovertebral junctions must be surgically stabilized, and protected against SCI[4]. All of eight patients had excellent outcome with the FIM score of 7.

Among 4 patients with associated TBI, 2 of them (50%) had poor outcome with the FIM score of 1. Horn et al reported 22 patients with occipitoatlantal dislocation who were treated and survived. Of them, 10 presented with TBI, their status was improved in six and remained unchanged in four[4]. In the present study, 3 of 15 patients presenting with SCI had poor outcome with the FIM score of 1. Two of the three patients had associated TBI, which might influence the outcome. The other patient with Klippel-Feil syndrome and C1-2 subluxation after falling from height sustained severe SCI. The patient's neurological function did not improve after treatment. Horn et al reported similar results. They concluded that the best predictor of outcome is the severity of neurological injuries at the presentation[4]. Severe TBI associated with brainstem dysfunction and complete cervical SCI lead to poor outcome. In contrast, neurological status of patients with incomplete SCI or less severe TBI can be improved after surgical stabilization.

C1-2 subluxation was the major type of CVJ injury in the present study. Two patients with os odontoideum on had history of falling for a long time before the presentation. One of them had associated ventral pannus compressing the cervicomedullary junction. Transoral decompression followed by dorsal atlanto-axial arthrodesis was performed as previously described by Menezes[5].

Several fixation techniques and instrumentations were used to stabilize CVJ instability. In the early period of this study, a less rigid contour rod and wiring technique were used in all cases. It necessitated a prolonged use of rigid cervical orthosis[6]. Since 2005, we have utilized more rigid fixation techniques; occipitocervical, C1-2 transarticular, and lateral mass screw and rod fixation. These fixation techniques are superior biomechanically, thus providing immediate stabilization with much less pseudoarthrosis rate[6-10]. We did not found postoperative instrumentation failure in the present study.

**Conclusion**

CVJ injury is a serious and life-threatening condition. Awareness of this uncommon injury leads to timely diagnosis. With appropriate surgical management, satisfactory results are usually achieved. Factors which may determine the outcomes include associated TBI and severity of SCI at the presentation.

**What is already known from this topic?**

Patients with suspicious of CVJ injury should be managed and transferred carefully before arriving hospital. Early diagnosis and appropriate treatment are crucial for preventing additional neurological morbidity.

**What this study adds?**

CVJ injury can occur in patients presenting with only neck pain without clinical evidence of SCI. This condition must be recognized in patients undergoing high-velocity motor vehicle accident, including patients with impaired consciousness caused by TBI. With prompt diagnosis and proper surgical treatment, most of patients with CVJ injury have neurological recovery to a satisfactory level.

**Potential conflicts of interest**

None.

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## *การบาดเจ็บบริเวณรอยต่อระหว่างกะโหลกศีรษะและกระดูกสันหลัง: ลักษณะทางคลินิกและผลการรักษาโดยการผ่าตัด*

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***ภูมิหลัง:*** *การบาดเจ็บบริเวณรอยต่อระหว่างกะโหลกศีรษะและกระดูกสันหลังเป็นการบาดเจ็บของกระดูกสันหลังที่พบได้ไม่บ่อย ซึ่งสามารถทำให้ถึงแก่ชีวิตหรือเกิดความพิการทางระบบประสาทแบบถาวรได้*

***วัตถุประสงค์:*** *เพื่อศึกษาลักษณะทางคลินิกและผลการรักษาโดยการผ่าตัดในผู้ป่วยที่มีการบาดเจ็บ บริเวณรอยต่อระหว่างกะโหลกศีรษะและกระดูกสันหลัง*

***วัสดุและวิธีการ:*** *ทำการศึกษาในผู้ป่วยทุกรายที่มีการบาดเจ็บบริเวณรอยต่อระหว่างกะโหลกศีรษะและกระดูกสันหลัง ซึ่งได้รับการรักษาโดยการผ่าตัดที่โรงพยาบาลศิริราชตั้งแต่ พ.ศ. 2540 ถึง พ.ศ. 2550 การเลือกวิธีการผ่าตัด ซึ่งมีหลายวิธีขึ้นอยู่กับรูปแบบความไม่มั่นคงของกระดูกสันหลังตำแหน่งและระดับความรุนแรงของภาวะไขสันหลังถูกกดทับรวมทั้งความผิดปกติของกระดูกสันหลังที่มีอยู่เดิม การวัดผลการรักษาอาศัยคะแนน Functional Independence Measure (FIM)*

***ผลการศึกษา:*** *ผู้ป่วยทั้งหมด 25 รายแบ่งเป็น 2 กลุ่ม ได้แก่ กลุ่มที่มาโรงพยาบาลในระยะเฉียบพลัน 18 ราย และกลุ่มที่มาโรงพยาบาลในระยะเรื้อรัง 7 ราย ชนิดของการบาดเจ็บที่พบบ่อยที่สุดคือการเคลื่อนของกระดูกสันหลัง ระดับคอที่ 1 และ 2 การบาดเจ็บที่พบรองลงมาได้แก่ การหักของ odontoid process ของกระดูกสันหลังระดับคอที่ 2 ผู้ป่วย 4 รายในกลุ่มที่มาโรงพยาบาลในระยะเฉียบพลันมีการบาดเจ็บของสมองร่วมด้วย ผู้ป่วยทุกรายได้รับการผ่าตัด ทางด้านหลังเพื่อยึดตรึงกระดูกสันหลังโดยใช้อุปกรณ์ยึดตรึงกระดูกสันหลังหลายชนิดผู้ป่วย 22 ราย มีผลการรักษาที่ดีโดยมีคะแนน FIM มากกว่าหรือเท่ากับ 4 ในทางตรงข้ามผู้ป่วยที่เหลือ 3 ราย มีผลการรักษาที่ไม่ดีโดยมีคะแนน FIM เท่ากับ 1 หรือช่วยเหลือตนเองไม่ได้เลยโดยทั้ง 3 ราย ดังกล่าวอยู่ในกลุ่มที่มาโรงพยาบาลในระยะเฉียบพลันผู้ป่วย 2 ใน 3 รายมีการบาดเจ็บของสมองร่วมด้วยและอีก 1 รายที่มีความผิดปกติของกระสันหลังแบบกลุ่มอาการ Klippel-Feil มาโรงพยาบาลด้วยอัมพาตของแขนและขาทั้งสองข้างซึ่งมีสาเหตุมาจากการบาดเจ็บของไขสันหลังอย่างรุนแรง*

***สรุป:*** *การผ่าตัดรักษาการบาดเจ็บบริเวณรอยต่อระหว่างกะโหลกศีรษะและกระดูกสันหลังให้ผลการรักษา เป็นที่น่าพอใจในผู้ป่วยส่วนใหญ่ ปัจจัยที่อาจมีผลต่อผลการรักษาได้แก่ การบาดเจ็บของสมองร่วมด้วยและความรุนแรงของการบาดเจ็บของไขสันหลัง*

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