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Editor
Nancy E. Epstein, MD
Professor of Clinical Neurosurgery, School of Medicine, State U. of NY at Stony Brook

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Original Article

Functional and radiological outcomes following craniovertebral junction surgery

Tushar N. Rathod, Sachin Mohana Khemkar, Tapas Mohanty, Arjit Vashishtha, Arun Kale, Akshay Phupate

Department of Orthopaedics, Seth Gordhandas Sunderdas Medical College and King Edward Memorial Hospital, Mumbai, Maharashtra, India.

E-mail: Tushar N. Rathod - tusharrathod@kem.edu; *Sachin Mohana Khemkar - sachinkhemkar@gmail.com; Tapas Mohanty - tapas.ku.mohanty@gmail.com; Arjit Vashishtha - vashishtharjit96@gmail.com; Arun Kale - kalearun74@gmail.com; Akshay Phupate - akshayphupate2@gmail.com

***Corresponding author:**
Sachin Mohana Khemkar, Department of Orthopaedics, Seth Gordhandas Sunderdas Medical College and King Edward Memorial Hospital, Mumbai, Maharashtra, India.

sachinkhemkar@gmail.com

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ABSTRACT

Background: Craniovertebral junction (CVJ) pathologies include atlantoaxial instability/deformities resulting in myelopathy, respiratory failure, and even death. Here, we describe the indications, preoperative planning, and intra-operative/postoperative complications following surgical management of CVJ anomalies.

Methods: A prospective analysis of 34 patients with CVJ pathology was evaluated between 2015 and 2022. Their various etiologies included atlantoaxial instability, trauma, tuberculosis, Down's syndrome, Morquio syndrome, os odontoideum, and atlantoaxial abnormalities. Clinical outcomes were assessed using the American spinal injury association (ASIA) impairment scale score and Benzel's modified Japanese Orthopedic Association (mJOA) score. Surgical assessments included length of hospital stay, operative time, blood loss, and intraoperative postoperative complications. Radiological parameters included fusion (i.e., implant loosening/implant failure), preoperative/postoperative atlanto-dens interval (ADI), clivus canal angle (CCA), and space available for cord (SAC).

Results: Five patients were managed conservatively, while 29 patients had surgery. Operations included occipitocervical fusion (14 patients), C1–2 fusion (10 patients), C1–2 transarticular screw fixation (four patients), and one patient underwent anterior corpectomy decompression/fusion. Seven patients had vertebral artery anomalies, and 13 patients had atlantoaxial abnormalities. At the final follow-up, atlantoaxial instability (i.e., mean preoperative ADI of 6.6 ± 2.3 mm) was restored to 4.2 ± 0.6 mm, significant cord compression (i.e., with mean SAC of 8.3 ± 2.9 mm) was relieved to 17.2 ± 1.6 mm, and the mean preoperative CCA (i.e., 130.2 ± 15.3) was improved to 143.3 ± 8.3°. There was also a statistically significant improvement in the ASIA scale and mJOA score.

Conclusion: Surgical management of CVJ abnormalities requires expertise and meticulous planning to avoid devastating complications such as wound dehiscence and catastrophic vertebral artery injury.

Keywords: Atlantoaxial instability, Clivus canal angle, Craniovertebral junction, Space available for cord, Vertebral artery abnormalities

INTRODUCTION

Craniovertebral junction (CVJ) pathologies are commonly associated with atlantoaxial instability and deformities resulting in cervical myelopathy, respiratory failure, or even death.[9] Various etiologies of CVJ include trauma, os odontoideum, tuberculosis, rheumatoid arthritis, degeneration, and tumors resulting in atlantoaxial instability (AAI). Radiologically, the atlanto-dens interval (ADI) (i.e., abnormal if more than 3 mm in adults and more than 5 mm in children),

space available for cord (SAC) (i.e., between posterior dens and anterior-posterior atlantal ring), and atlantoaxial dislocation (AAD) (i.e., decrease in the SAC resulting in spinal cord compression with <14 mm predicts paralysis) are defined.[1] Surgical management mainly focuses on correcting sagittal alignment and stabilization.[10] Here, we reviewed the indications, preoperative planning, intraoperative, and postoperative complications in the management of CVJ anomalies at a tertiary apex referral center over a period of 7 years.

MATERIALS AND METHODS

This was a prospective analysis of clinical, radiological, and surgical complications for 34 patients presenting with CVJ pathologies from 2015 to 2022 and is attributed to atlantoaxial instability, trauma, tuberculosis, Down's syndrome, Morquito syndrome, os odontoideum, and atlantoaxial abnormalities [Table 1]. Preoperatively, cervical spine X-rays, computed tomography angiography (CTA), and magnetic resonance (MR) studies were performed [Figure 1]. Clinical outcomes were assessed preoperatively, then at 4 weeks, 3 months, 6 months, and 1 year postoperatively (i.e., using the ASIA score and Benzel's modified Japanese orthopedic association [mJOA] score). Other data studied included length of hospital stay, operative time, blood loss, and intraoperative and postoperative complications. Radiological outcomes were assessed using ADI, clivus canal angle (CCA), and SAC [Figure 2]. Fusion was confirmed by the presence of bony trabecular bridging between C1 and C2 and the occipitocervical junction on computed tomography (CT) scan [Figure 3].

Surgical protocol

Surgical indications were ADI >5 mm for adults and in children with one or more of the following: neurologic involvement, persistent anterior displacement with ADI >4 mm, deformity present >3 months, or recurrence of deformity following 6 weeks of immobilization. Preoperative CT scans also were used to assess the pedicles, while CTA identified vertebral artery anomalies [Figures 4 and 5]. MR scans further documented basilar invagination with/without syrinx, signal changes in the spinal cord consistent with cord compression.

The five patients undergoing conservative management were given custom-fit cervical orthoses. Those patients having CVJ tuberculosis underwent fluoroscopy-guided biopsy and were given antitubercular treatment for 18 months as per our protocol (4 months of intensive phase which included isoniazid, rifampicin, ethambutol, pyrazinamide, and 14 months of continuation phase with isoniazid, rifampicin, and ethambutol). There were 29 patients who underwent surgery: occipitocervical fusion (14 patients), C1–2 fusion (10 patients), C1–2 transarticular screw fixation (four patients), and one anterior corpectomy decompression and fusion. In the immediate postoperative period, all patients were treated with custom-fit cervical orthoses. Routinely, surgical patients received parenteral antibiotics for 5 postoperative days. Wound checks were done on days 3 and 7. Drain removal was done 48 h following surgery. Suture removal was performed on postoperative day 15. The braces were continued till fusion was documented on a CT scan.

Table 1: Demographics of patients included in the study.

Male/female	20/14
Mean age (years)	26.9±16.1
Etiology	
CVJ tuberculosis	11
Traumatic	6
Downs syndrome	1
Atlantoaxial abnormalities	15
Morquio syndrome	1

CVJ: Craniovertebral junction

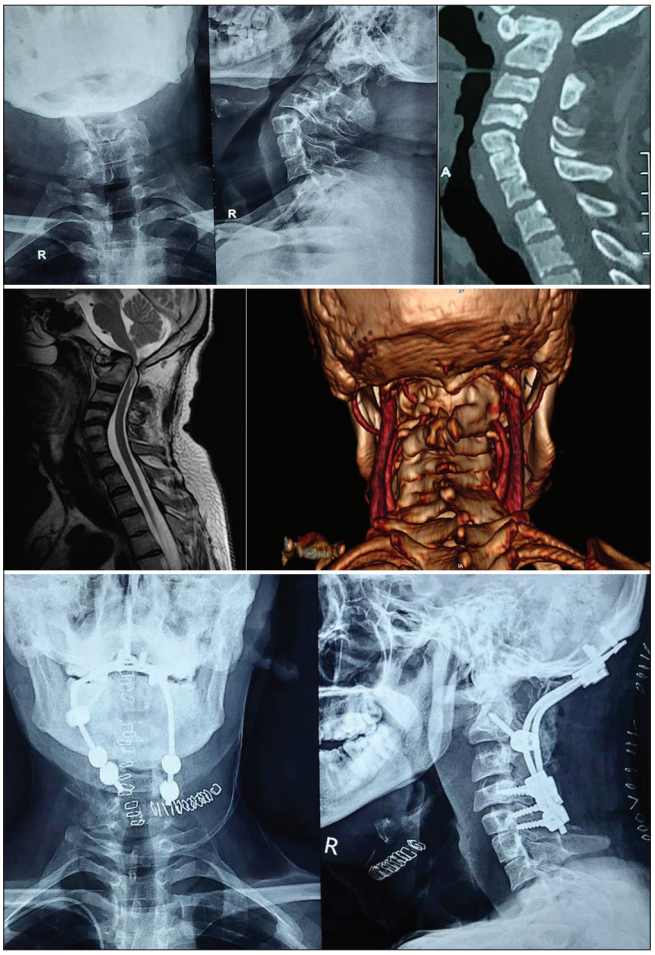


Figure 1: Preoperative X-ray, computed tomography scan with angiography and magnetic resonance imaging with postoperative X-ray.

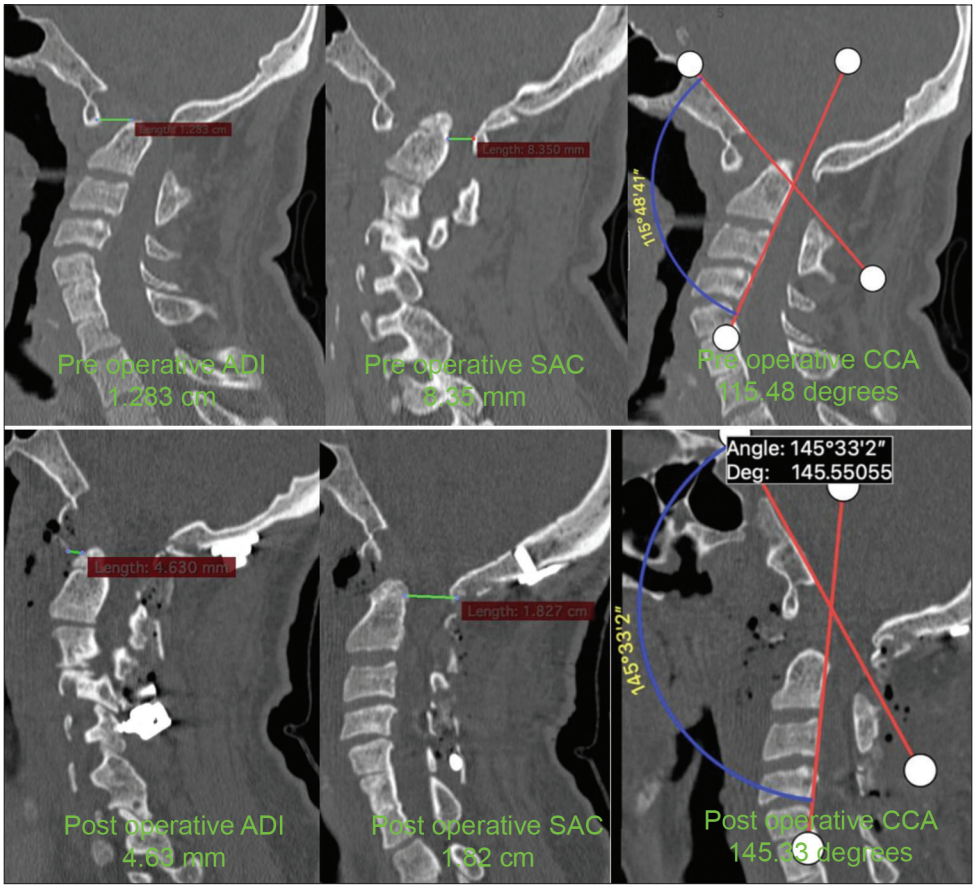


Figure 2: Pre and postoperative radiological parameters atlanto-dens interval (ADI), space available for cord (SAC), and clivus canal angle, respectively.
Green line- ADI and SAC, Brown bar - indicates measurement of ADI and SAC, Red lines with white dots- CCA, Blue arc- angle measurement in degrees

Figure 3: Bony fusion at the occipitocervical junction.

Statistical analysis

Preoperative values of the visual analog scale score, mJOA, ADI, SAC, and CCA were compared with postoperative values using the Mann–Whitney *U*-test, Wilcoxon-matched paired *t*-test, and Z-test [Table 2].

RESULTS

Surgical parameters were assessed using a mean operative time of 155.09 min, the mean intraoperative blood loss of 679.6 mL, and the length of stay in the hospital range from 15 to 30 days [Table 3]. There was a significant improvement in the ASIA scale postoperatively [Graph 1]. The intensity of pain was assessed using a visual analog score which significantly improved from a mean preoperative value of 5.65 ± 1.03 to a final follow-up value of 1.3 ± 0.47. Benzel's

Figure 4: Anomalous course of the left vertebral artery.

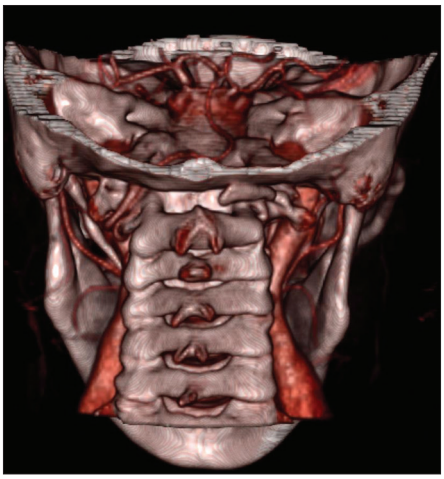


Figure 5: Hypoplastic right vertebral artery.

mJOA score improved from a mean preoperative value of 12.14 ± 2.6 to a postoperative mean value of 16.03 ± 0.7 and was statistically significant [Graph 2].

CVJ measurements

At the final follow-up, atlantoaxial instability was restored to 4.2 ± 0.6 mm, the mean SAC was 17.2 ± 1.6 mm, and the mean CCA was 143.3 ± 8.3°. Furthermore, fusion was achieved in 31 patients (91.17%).

Complications

Complications were seen in 3 patients (8.82%). One patient had an intraoperative massive bleed leading to death. Another patient died in the immediate postoperative period due to brainstem dysfunction. The third patient had a wound infection which led to a progressive neurological deficit requiring debridement and intravenous antibiotics.

Table 2: Functional and radiological outcomes.

Parameters	Preoperative	Final Follow-up	*P*-value
VAS score	5.65±1.03	1.3±0.47	0.001
Benzel's mJOA score	13.39±2.61	16.02±1.8	0.003
Atlanto-dens interval (mm)	6.6±2.3	4.2±0.6	0.216
Space available for cord (mm)	8.3±2.9	17.2±1.6	0.159
Clivus canal angle (degrees)	130.2±15.3	143.3±8.3	0.291

mJOA: Modified Japanese Orthopedic Association, VAS: Visual analog scale

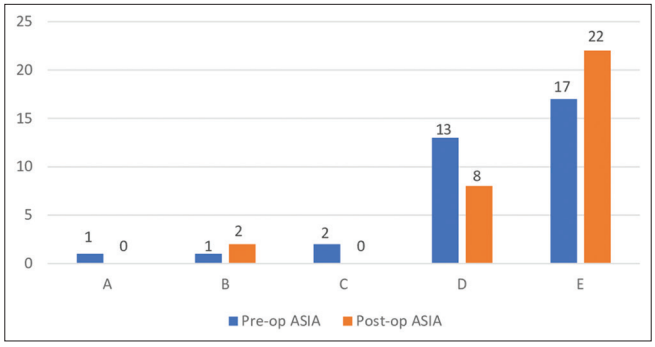
Table 3: Surgical parameters.

Surgical parameters	Mean+SD
Operative time (minutes)	155.09±48.21
Blood loss (mL)	679.6+399.3
Length of stay in hospital (days)	20.85±10.61

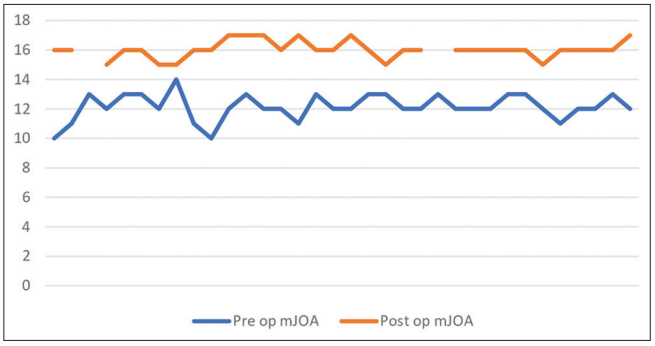
SD: Standard deviation

DISCUSSION

Management of CVJ abnormalities should be based on the type of instability, the integrity of posterior cervical elements, individual anatomic variation, and the surgeon's familiarity with the techniques. 7 patients with significant cervicomedullary compression and persistent AAI required posterior decompression/fusion. Shukla *et al.* reported 24 cases of CVJ tuberculosis: 5 patients underwent transoral biopsy only, 9 patients had transoral decompressions/posterior fusions, and 6 patients underwent only posterior fusion.[6] Salunke *et al.* described utilizing anterior releases



Graph 1: Preoperative and postoperative American spinal injury association (ASIA) impairment scale.



Graph 2: Preoperative and postopertaive modified Japanese orthopedic association (mJOA) score.

Table 4: Vertebral artery abnormalities.

Vertebral artery abnormalities	Number of patients
Hypoplastic right vertebral artery	3
Hypoplastic left vertebral artery	1
Left internal carotid artery retropharyngeal anomalous course	1
Extracranial sigmoid sinus course	1
Hypoplastic left common carotid artery	1

in patients with irreducible AAD, while 2 patients with atlanto-occipital assimilation and 1 patient with os odontoideum underwent anterior release with posterior instrumentation and fusion.[5] Srivastava *et al.* described the management of irreducible atlantoaxial dislocation with basilar invagination utilizing a single stage anterior release and posterior instrumented fusion in 19 patients.[7]

We had 1 patient with Down's syndrome and os odontoideum with AAD managed by Goel/Harms fixation.[3] Pueschel and Scola in their study of 404 patients with Down's syndrome found that 59 patients (14.6%) had AAI and 53 were asymptomatic, while 6 patients with symptomatic required surgery.[4] In our study, 1 patient with Morquio syndrome with a hypoplastic dens was managed with C1–2 transarticular screw fixation. Stevens *et al.* evaluated 13 patients with Morquio syndrome and atlantoaxial subluxation and found that odontoid dysplasia was present in all cases along with severe spinal cord compression requiring posterior occipitocervical fusion.[8] Vertebral artery anomalies at the CVJ can lead to catastrophic complications. Byun *et al.* in their study demonstrated right vertebral artery dominance in 4 patients (6.7%) and left vertebral dominance in 17 patients (28.3%) with AAI.[2] We had 7 vertebral artery anomalies and one fatal complication occurred due to massive bleeding on the dominant left vertebral artery [Table 4].

CONCLUSION

Surgical management of CVJ abnormalities requires expertise at a tertiary center, with a mandatory CTA, and careful operative planning to avoid devastating complications.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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