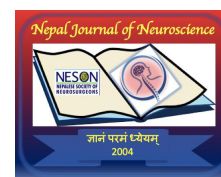


# Clinico Radiological Profile And Surgical Outcome Of Congenital Craniovertebral Junction Anomalies Surgery: An Institutional Experience

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## Abstract

**Introduction:** Congenital Craniovertebral junction (CVJ) anomaly has always proved a surgical challenge due to the complex anatomical location, unique embryological development, high mobility of the joints and its intricate relationship to vital neuromuscular structures. Patients usually presents with myelopathy, neck pain, and functional disability, such as difficulty swallowing and a associated neck deformity. Such patients requires early surgical treatment to prevent irreversible neurological deficit. But usually such patient presents late and sometimes respond late to surgical management offered to them. Due to limited literature available regarding surgical outcomes in patients with pre- operative poor neurological grade. Our study is thus an effort to determine the impact of surgery in neurological outcomes in such patients. Our objective was to review the clinico-radiological profile and surgical outcome of congenital craniovertebral junction (CVJ) surgery and also to determine the impact of surgery on neurological outcomes in patients with extensive neurological deficits

**Materials & Methods:** In our study, 32 patients of Congenital CVJ anomalies who underwent surgeries were retrospectively analysed for Demographic data, duration of symptoms, clinical presentation, neuroimaging, type of surgery, complications and post surgical outcome.

**Results:** Clinical and radiological datas of 28 patients were retrospectively analysed and the patients were followed up for 6 months post operatively. Pre operative Nuricks grade were compared with follow up Nurick grade for any improvement in functional outcome. Other independent variables such as age, sex, time of presentation, clinical features were analysed for any correlation with final functional outcome in such patients.

**Conclusions:** Early surgical intervention has better outcomes in congenital CVJ anomalies patients. However some patients with poor pre operative neurological grade also showed promising functional outcome thus emphasising the role of surgery in such patients.

**Key words:** Craniovertebral junction, Spine surgery, myelopathy, vertebral artery

## Introduction

Craniovertebral junction is a complex anatomical area due to its relation to lower medulla, vertebral artery and lower


cranial nerves. CVJ is one of the most mobile joints in our body. But this mobility comes at the cost of instability Due to its unique embryological development and high mobility. It is the site of multiple congenital anomaly<sup>1,2</sup> Surgery always poses a great challenge in this area due to its close proximity to critical anatomical structures<sup>3,4</sup>. Surgeries are mainly Anterior and posterior approach combined or Posterior approach alone<sup>5</sup>. Good neurological outcome following surgery is always attributed to early intervention and several literature suggest that patient presenting late with higher grades of myelopathy is the sole reason for poor surgical outcome<sup>6,7,8</sup>. Here in our study we tried to determine the outcome of surgery in patients with higher grades of myelopathy presenting to our institute

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Fig 1: CT scan and MRI images of CVJ anomaly

## Methods

### Aims and Objective:

1) To review the clinico-radiological profile and surgical outcome of congenital craniovertebral junction (CVJ) anomalies operated at our institute

2) To assess the impact of surgery on neurological outcome in patients with extensive neurological deficits.

We retrospectively analysed 32 cases of congenital CVJ anomalies operated at our institute in between February 2021 and January 2023 with a minimum of 2 years of followup. All Neuroimaging studies reviewed for bony and neurovascular details. Detailed history, duration of symptoms, neurological examination and Nurick scoring done. Diagnosis was made with static and dynamic X ray CVJ, static and dynamic Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) studies. Vertebral artery CT angiogram was done in all patients for pre-operative planning. Outcome was assessed by comparing pre and post operative Modified Nurick grade.

Table 1: Modified Nurick Classification

Modified Nurick Classification (24)	
<b>Grade 0</b>	No root or cord symptoms
<b>Grade I</b>	Root signs or symptoms. No evidence of cord involvement.
<b>Grade II</b>	Signs of cord involvement. Normal gait.
<b>Grade III</b>	Mild gait abnormality. Able to be employed.
<b>Grade IV</b>	Gait abnormality prevents employment.
<b>Grade V</b>	Able to ambulate only with assistance.
<b>Grade VI</b>	Chair bound or bedridden.

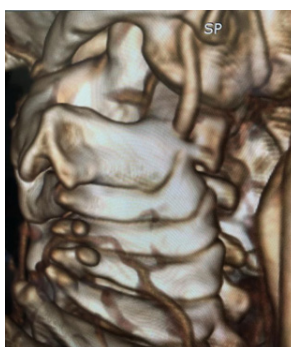


Fig 2: CT angiogram showing anomalous left vertebral artery

### Inclusion Criteria:

1) All congenital CVJ anomaly cases operated at our institute

### Exclusion Criteria:

1) Patients with congenital CVJ anomaly operated elsewhere

2) CVJ anomaly due to trauma, infection, tumour

## Results

Analysing the data of 32 patients (M= 20, F-12) median age of presentation was 21 years. Patients mostly presented after 2 years of initial presentation of symptoms.

CVJ anomalies according to age group

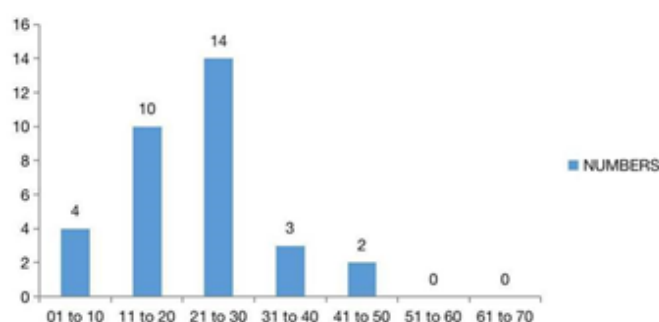


Fig 3: Age wise distribution of CVJ anomalies

Duration of Symptoms

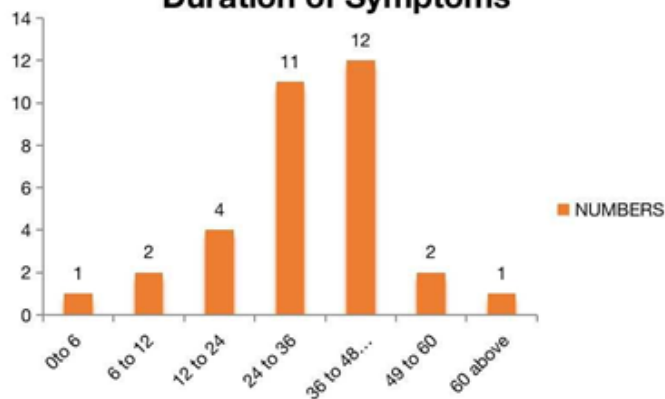


Fig 4: Duration of symptoms in CVJ anomalies

Neck pain (77.7%), Tingling numbness (74.1%) and gait impairment (66.7%) were most common presenting symptoms.

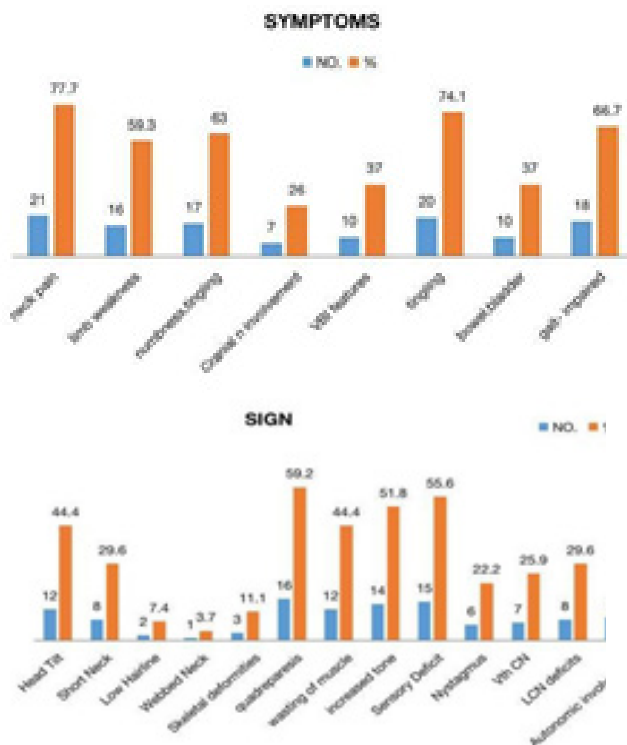


Fig 5: Signs and symptoms of patients with CVJ anomalies

Most common congenital CVJ anomaly detected was Atlantoaxial Dislocation in 75% cases. Two third of AAD were reducible type.

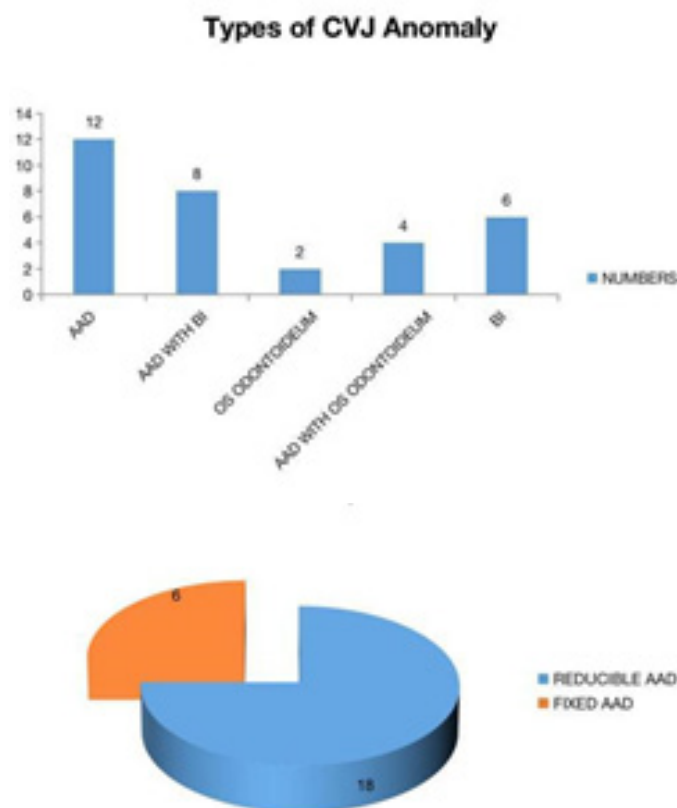


Fig 6: Different types of CVJ anomalies

We operated 5 cases by Anterior decompression followed by posterior stabilisation whereas rest 27 cases were operated by posterior approach only.

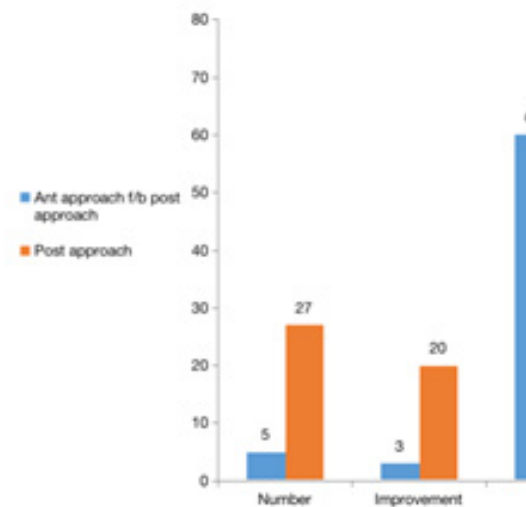


Fig 7: Different approaches of CVJ anomaly surgery

Post operatively occipital neuralgia was the most common complain among patients. We had 2 cases each of post operative CSF leak and new neurological deficit which were managed conservatively and patient improved subsequently.<sup>1</sup> case of implant failure which required revision surgery. Intraoperatively vertebral artery injury was detected in 1 case which was managed during the surgery and later patient developed pseudoaneurysm and underwent coil embolization.

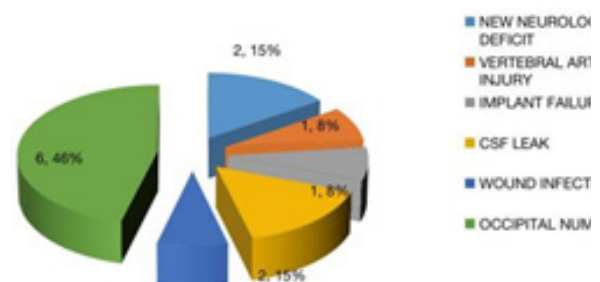


Fig 8: Complications following CVJ Anomaly surgery



71.5% of our patients showed improvement in neurological deficit post operatively while 21% patients remained same. Improvement in pre operative myelopathy grade was observed in 85% cases in mild pre operative myelopathy score (Nurick 0-3) while 61% patients with Nurick score (4-6) showed improvement. No mortality was seen in our study.

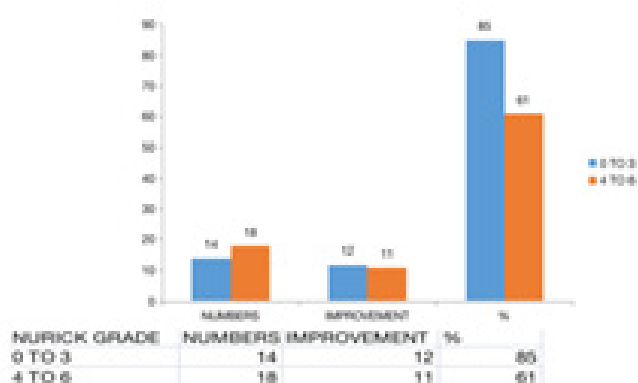
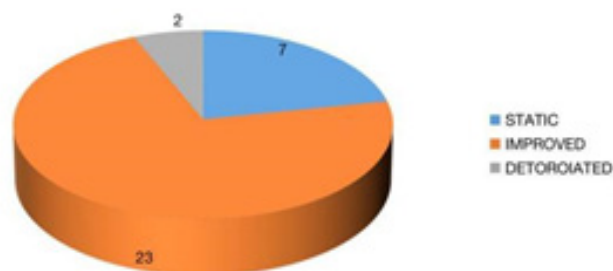


Fig 9: Surgical Outcomes

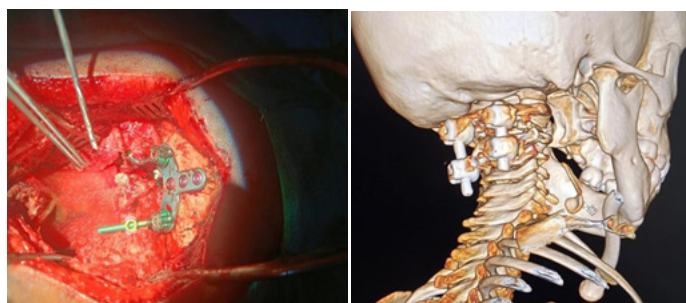


Fig 10a&b: Intraoperative image and post operative 3D reconstruction of CVJ surgery

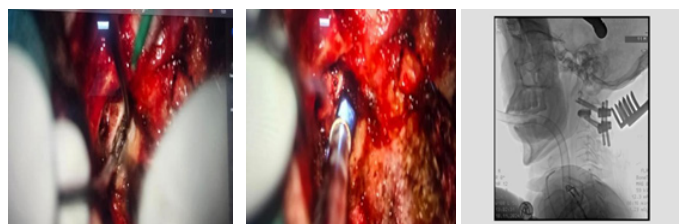


Fig 11a,b,c : Intraoperative C1-C2 joint dissection and post operative image of C1-C2 fixation with joint spacer

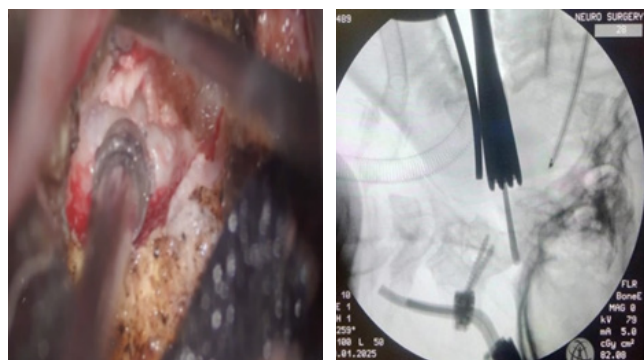


Fig 12 a &b: Intraoperative image of transoral odontoidectomy and C-arm image to confirm complete posterior cortex drilling of odontoid process

## Discussion

Congenital CVJ anomalies are more common in young adults with equal sex distribution<sup>9,10</sup>. We found congenital AAD as the most common anomaly in our patients like other studies<sup>10,11</sup>. Neck pain and quadriparesis were the most common signs and symptoms in patients with congenital CVJ anomaly<sup>12</sup>. Post surgery number of patients improved in our cases were similar to other studies<sup>7,13,14</sup>. Complications in the form of instrumentation, wound infection, CSF leak, vertebral artery injury are well known following CVJ surgery<sup>15,16</sup>. Vertebral artery injury is the most dreaded complication during CVJ surgery accounting to 2.4% in different literatures<sup>17</sup> we had 1 patient of vertebral artery injury in our study. Mortality in CVJ surgery is rare in larger series it is reported around 3%<sup>17</sup> we did not have any mortality in our series. Pre operative poor myelopathy grade is one of the most important determinants of outcome following CVJ surgery however we found that even patients with poor pre operative myelopathy grade shows considerable improvement post operatively<sup>18</sup>.

## Conclusion

Early surgical intervention in patients with congenital craniovertebral anomaly helps with better functional outcome in majority. But surgical intervention even in patients with delayed presentation and higher grades of myelopathy is also associated with good functional outcome in some and prevents further neurological deterioration in others. Reversal of neurologic deficits after surgery in patients with poor pre-operative neurologic grade needs further studies.

## References

- 1) Offiah CE, Day E. The craniocervical junction: embryology, anatomy, biomechanics and imaging in blunt trauma. Insights Imaging. 2017 Feb;8(1):29-47. doi: 10.1007/s13244-016-0530-5. Epub 2016 Nov 4. PMID: 27815845; PMCID: PMC5265194.

- 2) Tubbs RS, et al. Embryology and anatomy of the craniovertebral junction. *Handbook of Clinical Neurology*, Vol. 100, 2011, Elsevier.
- 3) Ropper AE. From Anatomic to Genetic Understanding of Developmental Craniovertebral Junction Abnormalities. *Neurospine*. 2020 Dec;17(4):859-861. doi: 10.14245/ns.2040548.274. Epub 2020 Dec 31. PMID: 33401863; PMCID: PMC7788420.
- 4) Chandra PS, et al. Complications of craniovertebral junction surgery: A 25-year experience. *Neurosurgical Review*, 42(4): 925–934, 2019 [PMID: 30460581]
- 5) Hong JT, Kim IS, Lee HJ, Park JH, Hur JW, Lee JB, Lee JJ, Lee SH. Evaluation and Surgical Planning for Craniovertebral Junction Deformity. *Neurospine*. 2020 Sep;17(3):554-567. doi: 10.14245/ns.2040510.255. Epub 2020 Sep 30. PMID: 33022160; PMCID: PMC7538356.
- 6) Goel A, Shah A. Progressive myelopathy despite apparent neural decompression following craniovertebral junction surgery: Is instability the cause? *Journal of Craniovertebral Junction and Spine*, 6(2): 65–70. 2015 [PMID: 26435794]
- 7) Chibbaro S, et al. Management of craniovertebral junction anomalies: review of 137 cases. *European Spine Journal*, 19(1): 20–28. 2010 [PMID: 19760208]
- 8) Sai Kiran NA, Vidyasagar K, Sivaraju L, Raj V, Aryan S, Thakar S, Mohan D, Hegde AS. Outcome of Surgery for Congenital Craniovertebral Junction Anomalies with Atlantoaxial Dislocation/Basilar Invagination: A Retrospective Study of 94 Patients. *World Neurosurg*. 2021 Feb;146:e313-e322. doi: 10.1016/j.wneu.2020.10.082. Epub 2020 Oct 20. PMID: 33096283.
- 9) Rao G, Das D, Rao S, et al. Congenital anomalies of the craniovertebral junction: A retrospective analysis of 40 cases. *Journal of Neurosurgery: Spine*, 7(6): 613–621. 2007 [PMID: 18074681]
- 10) Menezes AH. Craniovertebral junction anomalies: Diagnosis and management. *Seminars in Pediatric Neurology*, 15(3): 141–149. 2008 [PMID: 18760215]
- 11) Goel A, Shah A. Atlantoaxial instability: The most common cause of craniovertebral junction anomalies. *Journal of Craniovertebral Junction and Spine*, 1(1): 1–2. 2010 [PMID: 21886905]
- 12) Shankar D R, S Balamurugan, A Comprehensive study of Craniovertebral Junction Anomalies. *Int J of Neurol Neurosurg*, 2019; 11(1):78-86
- 13) Goel A, Shah A. Progressive myelopathy despite apparent neural decompression following craniovertebral junction surgery: Is instability the cause? *Journal of Craniovertebral Junction and Spine*, 6(2): 65–70. 2015 [PMID: 26435794]
- 14) Salunke P, et al. Outcome of posterior C1-C2 fixation in congenital atlantoaxial dislocation presenting with high-grade myelopathy. *World Neurosurgery*, 95: 406–412. 2016 [PMID: 26774436]
- 15) Salunke P, et al. Complications in posterior surgery for congenital atlantoaxial dislocation: Avoidable or not?
- 16) Visocchi M, Mattogno PP, Signorelli F, Zhong J, Iacopino G, Barbagallo G. Complications in Craniovertebral Junction Instrumentation: Hardware Removal Can Be Associated with Long-Lasting Stability. Personal Experience. *Acta Neurochir Suppl*. 2017;124:187-194. doi: 10.1007/978-3-319-39546-3\_29. PMID: 28120073.
- 17) Song GC, Cho KS, Yoo DS, Huh PW, Lee SB. Surgical treatment of craniovertebral junction instability : clinical outcomes and effectiveness in personal experience. *J Korean Neurosurg Soc*. 2010 Jul;48(1):37-45. doi: 10.3340/jkns.2010.48.1.37. Epub 2010 Jul 31. PMID: 20717510; PMCID: PMC2916146.
- 18) Thakar S, Mohan D, Hegde AS. Outcome of Surgery for Congenital Craniovertebral Junction Anomalies with Atlantoaxial Dislocation/Basilar Invagination: A Retrospective