# Craniovertebral Junction Anomaly

- Rare tandem anomalies of the vertebral artery at the craniovertebral junction - What the mind does not know the eyes do not see!!
- A Case Report of an Adverse Outcome: Development of a Dural Arteriovenous Fistula Following Foramen Magnum Decompression for Chiari Malformation
- Surgical Treatment of Basilar Invagination without Evident Atlantoaxial Instability (Type B) - A Systematic Review
- Common origin of the right vertebral artery and the right costocervical trunk diagnosed by magnetic resonance angiography
- Occipitocervical fusion and serious airway adverse events: A systematic review
- Morphometric analysis of the lateral mass of atlas and its clinical significance in craniovertebral junction surgeries
- SEMANTICS AND DYNAMICS OF HEADACHE IN PATIENTS WITH CHIARI MALFORMATION TYPE I AFTER DECOMPRESSION SURGERY: EXPERIENCE FROM AZERBAIJAN
- Mapping, classification, and surgical strategy for vertebral artery variation in posterior atlantoaxial joint release, distraction, and fusion surgery for basilar invagination and atlantoaxial instability

Anomalies of the craniovertebral junction can result from congenital, developmental, or traumatic causes. These anomalies can affect the stability, movement, and neurological function in the region and may require surgical intervention, especially if they lead to brainstem or spinal cord compression.

## Pediatric Craniovertebral Junction Anomaly

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## Common Anomalies of the Occipitocervical Articulation

**Basilar Invagination**

**Craniovertebral Instability**

### Other

**Atlanto-occipital Assimilation**

**Chiari Malformation**

**Klippel-Feil Syndrome**

**Platybasia**

Congenital craniovertebral junction deformities can be associated with an anomalous vertebral artery (VA). At times, the artery crosses the joint posteriorly (i.e., persistent first intersegmental artery) and is at risk during posterior approach.

Karthigeyan et al., report a new variant, wherein the bilateral VA coursed medially after exiting the C3 transverse foramina to lie beneath C2 pars interarticularis and enter the foramen magnum (without passing through C2 transverse foramen anywhere along its course). This is possibly a result of bilateral persistent second intersegmental arteries. It is pertinent to recognize this unusual variant to avoid VA injury, especially while inserting C2 pars/transarticular screw [1)].

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A retrospective study was performed on total of 38 patients who met the inclusion criteria for analysis. These patients were contacted through telephone and letters and their clinical examination and radiological investigations were performed at follow up visit. The preoperative, the postoperative and follow-up clinical evaluation of the patients were done by the Nurick grading system.

The age ranged from 4 to 60 years with average being 20.5 years. There were 31 males and 7 females with ratio of male to female being 4.43:1. 13 cases were of fixed atlanto-axial dislocation (AAD) 17 were of mobile AAD, 6 were traumatic and 2 patients had post-infective AAD. Majority of these patients (n=29, 76.31%) presented with neck pain followed by cerebello-vestibular disturbances (n=27, 71.1%). Sphincter disturbances were observed in nine patients. Significant increase in cranio-vertebral angle was observed in postoperative period in all the patients. Initially, 84% of the patients were in poor Nurick grade, which were reduced to 28% after the surgical intervention. 100% bony fusion was attained in patients who underwent rigid fixation technique and 80% in semi rigid fixation technique.

The key to successful management of craniovertebral junction disease is individualized selection of judicious surgical intervention from various available techniques [2)].

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Manifestation of congenital anomalies of cranio-vertebral junction (CVJ) in the later half of life is unusual and intriguing. Coexisting cervical spondylotic changes with multilevel compression, poorer bone quality as well as less smooth post-surgical recuperation make management of elderly Congenital Atlantoaxial Dislocation/ Basilar Invagination (CAAD/BI) challenging.

Clinico-radiological data of 20 patients of CAAD/BI (with markers of congenital anomalies) presenting after 50 years of age, the challenges faced and outcomes after C1-C2 fusion have been analysed.

Three distinct groups were identified. Seven patients with Os-odontoideum had reducible AAD (Type I). Seven patients had assimilated C1, C2-3 fusion and deformed C1-2 joints with irreducible AAD/BI (Type II). In type III, 4 patients had similar segmentation defects but with compression at both cervico-medullary junction and subaxial spine, although clinical localisation pointed to the CVJ. Spastic quadriparesis was the commonest presentation. All underwent C1-2 fusion alone. There was significant improvement in 18, including those with compression at additional level. Bony fusion was documented in all patients followed up beyond one year.

Congenital CVJ anomalies may present in later half of life, though attempts at reasoning remain

speculative. These patients improve after multiplanar realignment and C1-2 fusion. Careful clinico-radiological evaluation is required in those with additional subaxial compression. Bone quality in elderly is not a deterrent for instrumentation. Fusion eventually occurs in most [3].

## Treatment

Craniovertebral junction surgery.

## Case series

Craniovertebral Junction Anomaly Case Series.

1)
Karthigeyan M, Salunke P, Kataria MS. Bilateral persistent 'second' intersegmental vertebral arteries: illustrated with a case. Acta Neurochir (Wien). 2018 Jun 23. doi: 10.1007/s00701-018-3601-1. [Epub ahead of print] PubMed PMID: 29934840.
2)
Faheem M, Jaiswal M, Ojha BK, Chandra A, Singh SK, Srivastava C. Clinico-Radiological Outcome Analysis in Cranio-Vertebral Junction Diseases: An Institutional Experience of 38 Patients in a Tertiary Care Centre. World Neurosurg. 2018 Jun 21. pii: S1878-8750(18)31315-9. doi: 10.1016/j.wneu.2018.06.099. [Epub ahead of print] PubMed PMID: 29936209.
3)
Salunke P, Karthigeyan M, Sunil N, Rangan V. 'Congenital anomalies of craniovertebral junction presenting after 50 years of age': An oxymoron or An unusual variation? Clin Neurol Neurosurg. 2017 Dec 21;165:15-20. doi: 10.1016/j.clineuro.2017.12.015. [Epub ahead of print] PubMed PMID: 29289915.