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Radiological Evaluation of Craniovertebral Junction Anomalies

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Abstract: ***Introduction: The craniovertebral junction (CVJ), a complex area where the skull meets the spine, is critical for head movement and brainstem protection. Anomalies in this region can cause neurological complications. This thesis investigates the role of radiological evaluation in diagnosing CVJ anomalies. The thesis reviews the normal CVJ anatomy and various imaging techniques used for assessment, including plain radiography, computed tomography (CT), and magnetic resonance imaging (MRI). It then explores the spectrum of congenital and acquired CVJ anomalies, detailing their radiological presentations and classifying them into appropriate etiological categories to aid management. Methodology: The research component of the thesis involves analyzing patient data, evaluating different imaging modalities, or investigating the effectiveness of specific radiological craniometric measurements in diagnosing CVJ anomalies. Conclusion: The thesis concludes by emphasizing the importance of accurate radiological evaluation in early diagnosis and management of CVJ anomalies, potentially improving patient outcomes.***

Keywords: CVJ Malformations (focuses on anomalies specifically), Diagnostic imaging, Craniometric measurements, Neurological complications, Early Diagnosis

1. Introduction

The cranio cervical junction (CVJ), where the skull meets the spine, is a critical area for both movement and protecting vital structures in the brain. Abnormalities in this region can cause serious neurological problems, particularly in populations like those in the Indian subcontinent. These problems can range from fluid buildup in the brain (hydrocephalus) to nerve and artery compression, even spinal cord injuries. Therefore, it's crucial to consider CVJ anomalies in patients with these symptoms. (1).

Thankfully, various imaging techniques such as X - rays, CT scans, and MRIs can clearly visualize the CVJ. These scans can even provide dynamic information when performed with specific movements. Normally, the CVJ is somewhat visible in standard brain scans. On X - rays, the CVJ typically extends from the level between the second and third vertebrae in the neck to a line drawn between a bony bump on the skull (internal occipital protuberance) and a specific point on the sphenoid bone (dorsum sellae).

The CVJ itself is a complex area encompassing the occipital bone, a bony projection at the skull base (clivus), the large opening at the base of the skull (foramen magnum), and the top two cervical vertebrae. It also includes the joints between these bones, the ligaments connecting them, and the soft tissues like the brainstem (medulla), spinal cord, cerebellum, and lower cranial nerves. The primary ligaments responsible for stability are the alar ligaments and the transverse ligament. (2).

Abnormalities of the CVJ can compress the brainstem and spinal cord, cranial nerves, or the vertebral artery. This compression can lead to various symptoms like neck pain and weakness in the limbs. These abnormalities can be present at birth (congenital) or develop later in life due to trauma, infections, or inflammatory conditions.

The CVJ's unique anatomy and function set it apart from both the skull and the cervical spine. It contains muscles, ligaments, membranes, and bones connected by joints. This area also cradles the spinal cord, several cranial nerves, and vital blood vessels supplying the brain and spinal cord. Damage to the CVJ can have severe consequences, even leading to death. The CVJ faces a demanding challenge: it must allow for a wide range of motion while simultaneously protecting these critical structures essential for survival.

The craniocervical junction (CVJ) can be a complex area for medical residents to understand. By having more in - depth discussions and evaluations of CVJ pathologies, residents can improve their knowledge and decision - making skills. This will ultimately lead to better patient care and outcomes.

2. Literature Survey

2.1 Embryology (3, 4, 5)

The embryology of the craniocervical junction (CVJ) is a fascinating and intricate process that lays the foundation for its complex structure and function. Here's a breakdown of the key players:

Building Blocks:

- **Somites:** These are segmented blocks of tissue that appear along the developing embryo's back. Specific somites

contribute to the formation of the vertebrae and associated structures.
- **Sclerotomes:** Each somite gives rise to a sclerotome, a mesenchymal structure that eventually forms cartilage and bone.

Key Contributors:
- **Occipital Sclerotomes (Somites 1 - 4):** These contribute to the formation of the occipital bone at the base of the skull, including the clivus and the occipital condyles (bony bumps that connect the skull to the spine).
- **Proatlas:** This transient structure derived from the fourth occipital sclerotome eventually forms the anterior arch of the atlas (the first cervical vertebra).
- **First and Second Cervical Sclerotomes (Somites 5 - 6):** These contribute to the formation of the remaining parts of the atlas and axis (the second cervical vertebra).

The Big Picture:
- The occipital bone, clivus, and occipital condyles develop from cartilage models that are later replaced by bone through a process called enchondral ossification.
- The atlas and axis form through a combination of enchondral ossification and intramembranous ossification (bone formation directly from mesenchymal tissue).
- The ligaments that stabilize the CVJ also arise from specific mesenchymal tissues surrounding the developing vertebrae.

2.2 Anatomy

The craniocervical junction (CVJ) is a critical anatomical region where the skull meets the spine. This complex area plays a vital role in both stability and movement, allowing for a wide range of head motions while protecting the brainstem, spinal cord, and other vital structures. Here's a breakdown of its key anatomical components:

Bony Elements:
- **Occipital Bone:** Forms the posterior base of the skull, including the clivus (a bony projection) and the occipital condyles (rounded knobs that articulate with the atlas).
- **Atlas (C1):** The first cervical vertebra, unique in its ring - like shape. It lacks a vertebral body and instead has a large opening (foramen magnum) that allows the brainstem to pass through.
- **Axis (C2):** The second cervical vertebra, notable for its prominent dens (odontoid process) that projects upwards and articulates with the atlas.

Ligaments: (6)
- **Cruciate Ligament:** Located within the atlas, with the transverse ligament forming the anterior part and the alar ligaments extending posteriorly. It stabilizes the dens and prevents excessive movement of the head on the axis.
- **Tectorial Membrane:** A thin membrane stretched across the dens, providing additional support and housing the alar ligaments.
- **Occipitoatlantal and Atlantoaxial Ligaments:** These ligaments connect the occiput to the atlas and the atlas to the axis, respectively, providing additional stability and limiting excessive motion.

Soft Tissues:
- **Medulla Oblongata:** The lower portion of the brainstem, responsible for essential functions like breathing, heart rate, and swallowing. It passes through the foramen magnum in the occiput.
- **Spinal Cord:** The continuation of the brainstem that descends through the spinal canal within the vertebrae.
- **Lower Cranial Nerves:** Several cranial nerves (glossopharyngeal, vagus, accessory, and hypoglossal) originate from the brainstem and pass through the CVJ, innervating structures like the tongue, throat, and muscles.
- **Vertebral Arteries:** These arteries supply blood to the brainstem and cerebellum, entering the skull through the foramen magnum.

Functional Significance:
- The unique articulation between the occiput, atlas, and axis allows for a wide range of head movements, including flexion, extension, rotation, and lateral bending.
- The strong ligaments and bony structures provide stability and prevent excessive movement that could damage the brainstem, spinal cord, and nerves.
- The CVJ protects these vital structures from injury due to impacts or sudden movements.

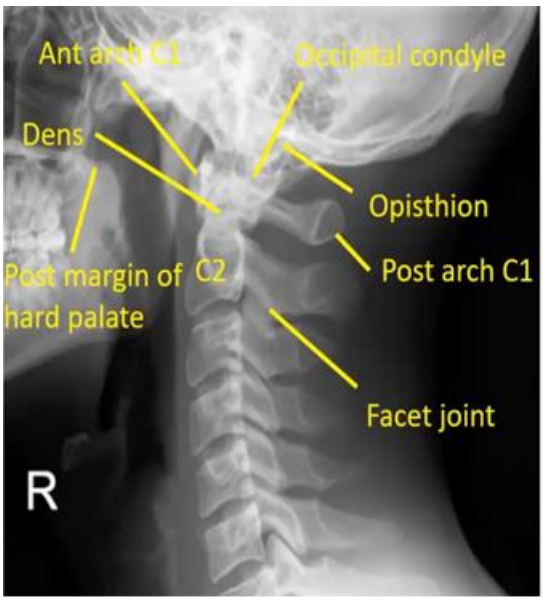


Figure 1: X - Ray Anatomy of Craniovertebral Junction

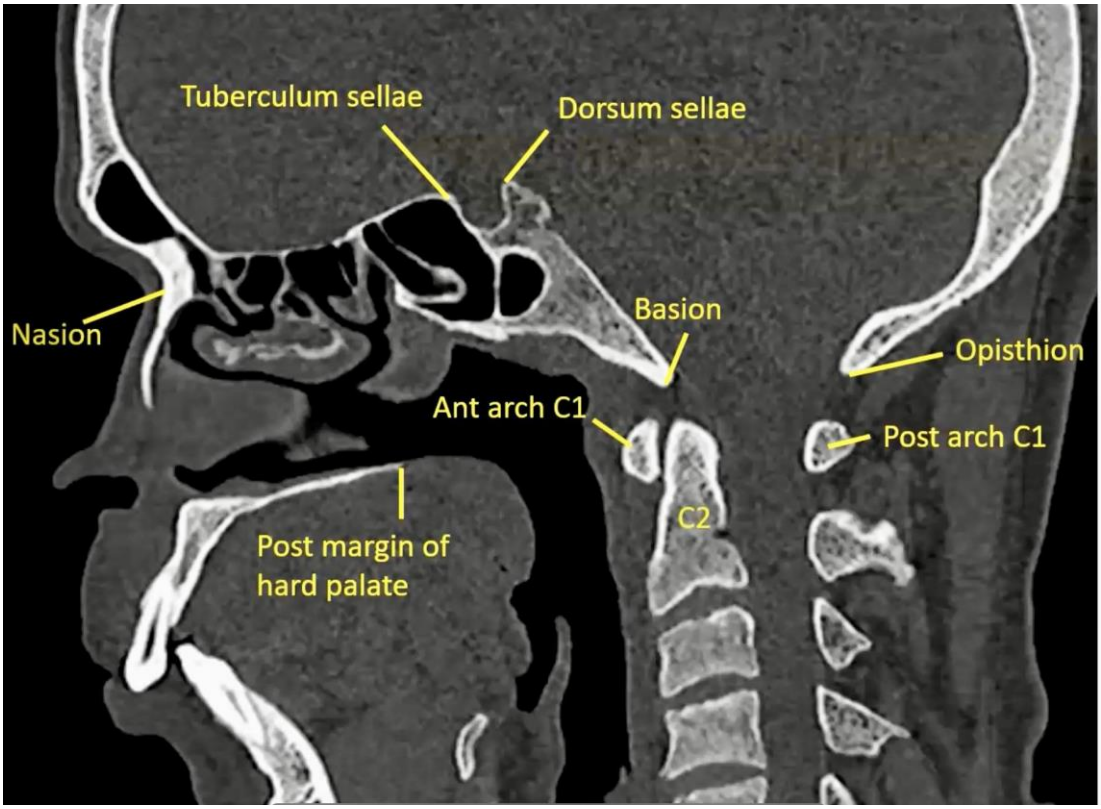


Figure 2: CT Anatomy of Craniovertebral Junction

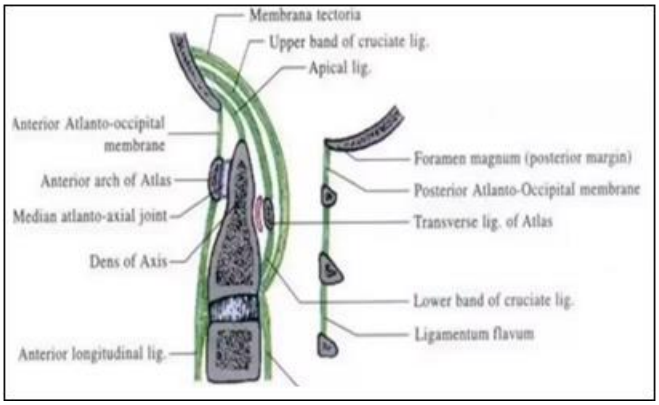


Figure 3: Understanding the Ligaments Supporting the Craniovertebral Junction

2.3 Craniometric Measurements

Table 1: Essential Craniometry To Evaluate Cvj Anomalies

	Normal Range	Remarks
Chamberlain line (Palato Occipital line)	Dens apex < 3 mm above this line, anterior arch of C1 typically lies below	Diagnosis of basilar invagination. (Posterior rim of foramen magnum shows great anatomic variability and also it may be difficult to radiologically pinpoint opisthion)
McGregor line (Palato Suboccipital line)	Dens apex < 7 mm above this line, anterior arch of C1 typically lies below	Diagnosis of basilar invagination
McRae line (Foramen magnum line)	Tip of dens below this line	Assess the decrease in content injury
Wackenheim line (craniovertebral or clivus - canal angle)	Line falls tangent to, or intersects, the posterior one - third of the odontoid	Assessment of CVJ traumatic injuries
Welcher basal angle	125 - 143	Assessment of platybasia

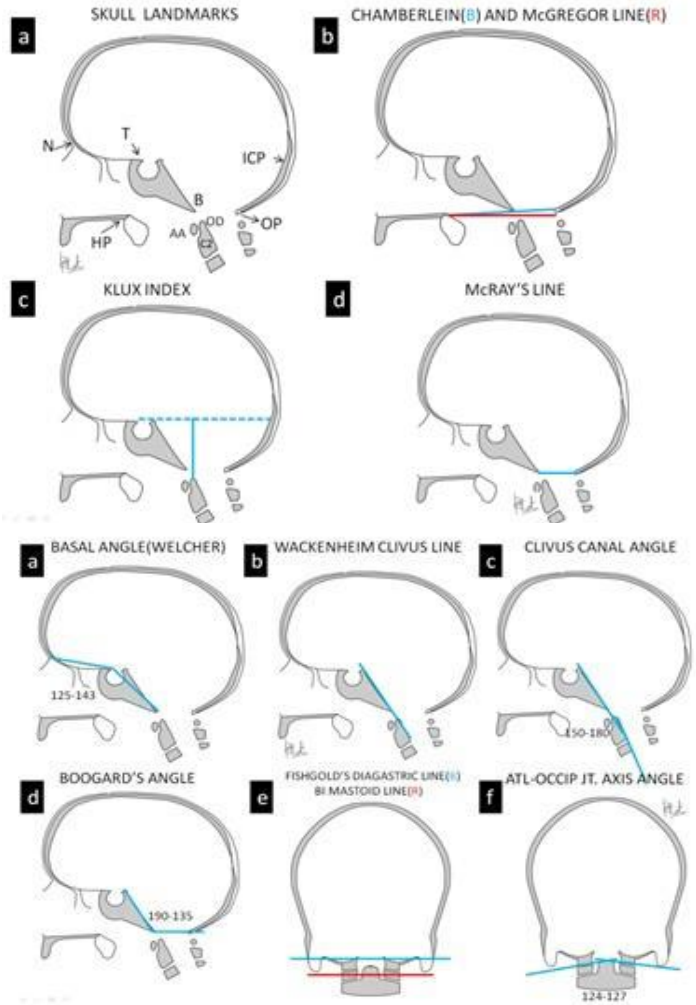


Figure 4: Essential Craniometry to Evaluate CVJ Anomalies

2.4 Classification of CVJ Anomalies

Table 2: Etiological Classification of Anomalies

Common pathological conditions of CVJ			
Category	Specific entity	Presentation - features	Additional significance
Congenital anomaly	Rachischisis	Incidental	Associated anomalies
	Clival hypoplasia	Incidental	
	Pro - atlas anomaly	Occipitalization of C1, Condylar defects	Limitation of movement
			Susceptible for trauma
	Atlanto - axial instability		Unstable
	Fusion - segmentation defect		Scoliosis
	Klippel Feil anomaly	Syndromic, Multisystem	Short spine, Anomalies
Syndromic	Morquio Syndrome	Congenital entities with variable presentation	Subluxation, spinal compression.
	Spondylo - epiphyseal dysplasia,		Late manifestation of symptoms
	Cleido cranial dysplasia		
	Down's Syndrome		A - A Subluxation
Trauma	Non - displaced Fracture	Minor	Stable
	Displaced Injury/dislocation	High velocity injury	Unstable, Neural injury
	Rotatory subluxation	Pediatric, Trauma, inflammatory	Self limiting
Inflammatory	Septic	Diabetic, acute symptoms	Epidural abscess
	Tuberculosis	Insidious onset, subluxation	Subluxation, epidural abscess
	Rheumatoid	Systemic, Erosions	Subluxation
	Eosinophilic granuloma	lytic lesions	Healing - recovery
Neoplastic	Primary - Chordoma, Sarcoma, GCT	Expansion, osteolysis /sclerosis, new - bone formation	Needs MRI, PET
	Secondary	Breast, Thyroid, Lung	
Misc	Paget's disease	Skull softening, BI,	Deformity
	Osteogenesis imperfecta	deformity	Cord compression
	Osteomalacia, Hyperparathyroidism		

3. Methodology and Approach

3.1 Inclusion Criteria: About 74 patients reffered to the department of radiodiagnosis from other IPD and OPD department of hospital was considered for study. Written consent was taken from the participants prior to enrollment in the study after explaining the details of the research project. A pre structured proforma was used to collect the relevant clinical data. The patient was subjected to MRI and/or CT and /or XRAY. Descriptive data was collected and studied accordingly.

3.2 Exclusion Criteria: Patients reluctant to participate in the study and claustrophobic patients were excluded.

3.3 Radiological Criteria for the Diagnosis

Imaging analysis focused on the integrity and alignment of the occiput, atlas, axis, and other relevant vertebrae. Dynamic X - rays of the CVJ were analysed in flexion and extension to evaluate atlanto - axial sublaxation Craniometric measurements like the basion - opsithion distance and atlanto - dens interval were assessed using established protocols. MRI scans were evaluated for ligamentous structures, brainstem, spinal cord, and nerve root involvement. The prevalence of different types of CVJ anomalies was determined based on radiological findings.

3.4 Table 3. Performa Used to Evaluate CVJ Anomalies

Serial Number		
Age		
Clinical Complaints		
Chamberlein Line		
Wackenheim's Line		
Basilar Invagination		
Boogard's Angle		
Welcher Basal Angle		
Platybasia		
Aad/Aas		
Foramen Magnum Stenosis		
Posterior Atlanto - Dental Interval		
Clivus		
Occipital Condyle		
Atlas		
Axis		
Brain Stem Herniation		
Spinal Cord Changes		
Syrinx		

4. Observation and Results

This study investigated craniovertebral junction (CVJ) anomalies in 74 patients. Here's a concise breakdown of the findings:

4.1 Age Distribution: The average age was 39.7 years, with a wide range (1 - 85 years). The most frequent age group was 41 - 50 years (24.3%). In contrast, the study by **Dhadve R et al** (1). found that the most common age group for CVJ disorders was 21 - 40 years, accounting for 30.64% (19 patients), followed by those over 60 years, at 27.41% (17 patients).

4.2 Gender Distribution: Males were more predominant (56.8%) than females (43.2%)

4.3 Craniometric Assessment

4.3.1 Basilar Invagination: Measured by **Chamberlain's and Wackenheims lines,** 24% of patients showed signs of basilar invagination (dens above the line). In comparison, **Singh N et al** (2). reported a higher prevalence of basilar invagination (54.39%), emphasizing its association with atlantoaxial instability, noted in 26 out of 31 patients with basilar invagination in their study,

4.3.2 Platybasia: Measured by **Boogard's and Welcher angles,** only 2.7% of patients had platybasia (increased angle)

4.4 Additional MRI Findings:

a) Tonsillar/brainstem herniation: 6.8%.
b) Spinal cord changes: 28.4%
c) Syrinx

4.5 Classification of Cases on the Basis of Etiology

Table 4: Etiological Classification

Diagnosis	Frequency	Percent
Acquired	39	52.7
Developmental	23	31.1
Acquired, developmental	12	16.2
Total	74	100

Acquired	Developmental
Trauma 14	
Tumour 6	
Infections 8	
Degenerative changes 12	
Basilar invagination.13	
Atlanto - axial sublaxation 18	

Atlanto - axial sublaxation was the most common finding under acquired conditions in the study followed by trauma, basilar invagination and degenerative changes. Almost all the acquired infectious conditions were found to be of tubercular etiology.

Table 5: Distinct Patterns of Injury in Trauma

Trauma	N	%
Odontoid fracture	7	50%
Rotatory sublaxation	4	29%
Fractures involving other bones at CVJ	3	21%
Total	14	100%

Table 6: Distinct Patterns of Tumour at CVJ

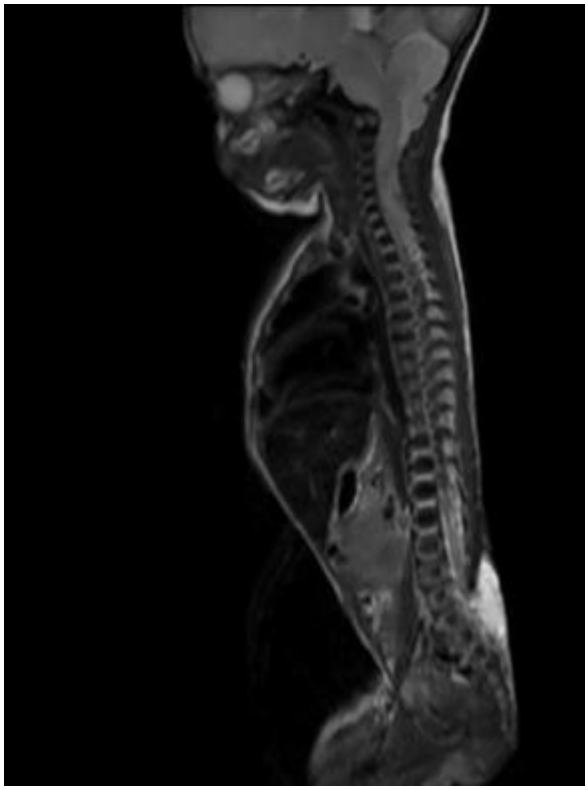
Tumour	N	%
Indeterminate	1	16%
Chordoma	1	16%
Meningoma	2	33%
Metastatic	2	33%
Total	6	100%

Table 7: Patterns of Developmental Conditions at CVJ

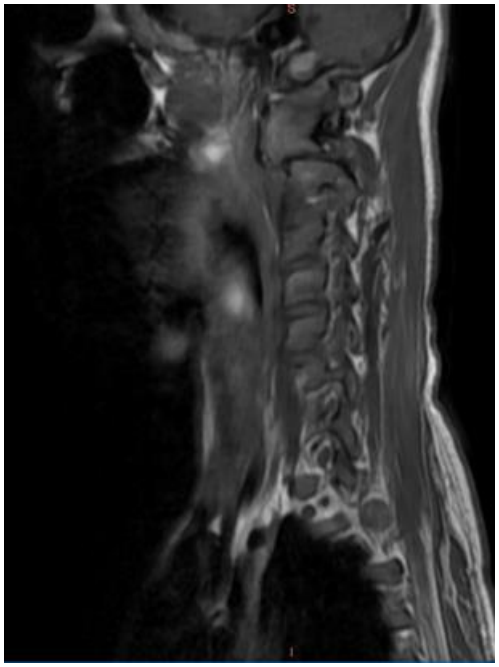
Developmental	N	%
Arnold Chiari Malformation	6	13%
Os odontoideum	5	11%
Platybasia	2	4%
Occipitalisation of atlas	26	58%
Mucopolysaccharoidosis	1	2%
Klippel fiel syndrome	2	4%
Aplasia/hypoplasia of posterior arch of atlas	3	7%
Total	45	100%

Comparatively, **Dhadve R et al** (1). reported findings on degenerative changes and various etiologies affecting the CVJ, including infective conditions such as tuberculosis (TB) in four patients and inflammatory pathology like rheumatoid arthritis (RA) in three patients. Among neoplasms, they identified one case each of meningioma, chordoma involving the axis and C3 vertebra, and a nerve sheath tumor at the C2 vertebral level. They also noted syndromic associations in two patients, including Klippel - Feil syndrome and Down syndrome.

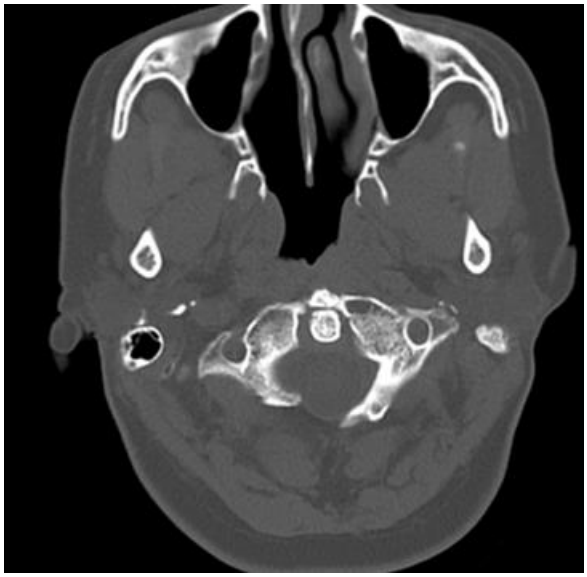
5. Cases



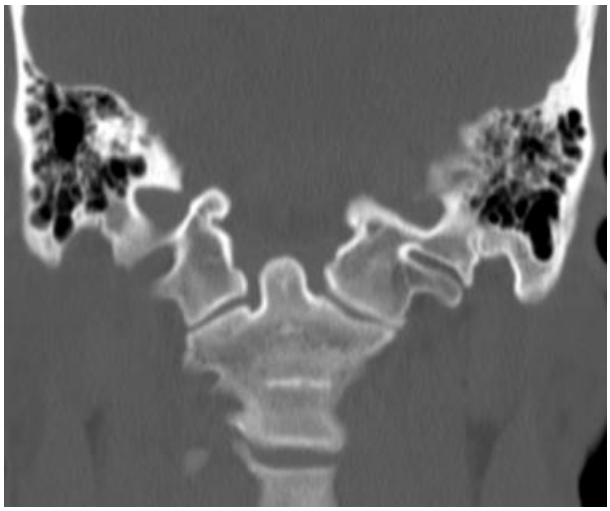
Case 1: Tonsillar Herniation with Myelomeningocele Arnold Chiari Malformation Type 2



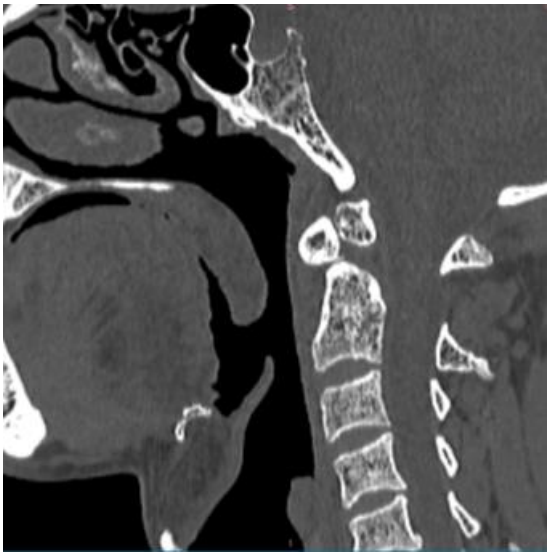
Case 2: Fusion of right lateral mass of atlas with occiput with mild anterior rotatory sublaxation of atlanto - axial joint



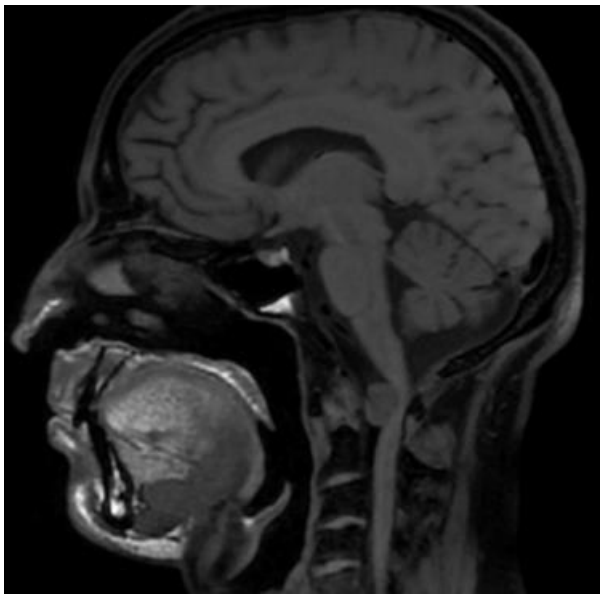
Case 3: Hypoplastic right posterior arch of atlas



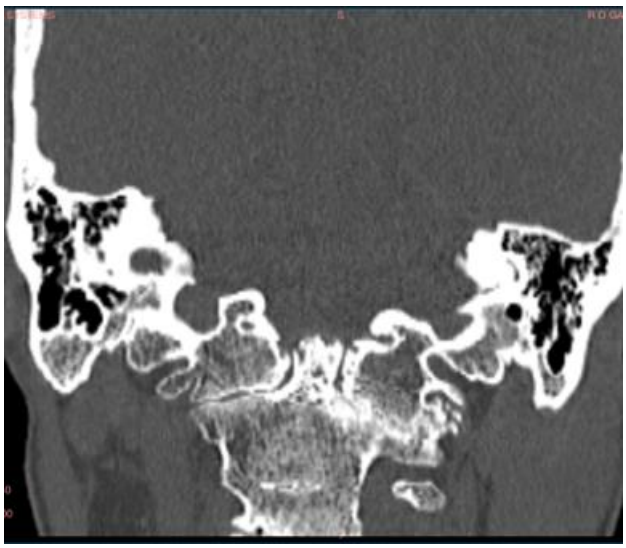
Case 4: Complete assimilation of atlas to basiocciput



Case 5: Os Odontoideum



Case 6: Metastatic lesion from primary spindle cell carcinoma in pre - sacral region



Case 7: Arthopathy and degenerative changes and atlanto - axial joint

6. Conclusion

The craniovertebral junction (CVJ) is a complex anatomical region encompassing critical structures like the occipital bone, atlas, axis, and associated neural elements. Disorders affecting the CVJ can arise from congenital anomalies, developmental abnormalities, or acquired conditions, each presenting unique diagnostic and therapeutic challenges. Our study of 74 cases highlights the diversity and prevalence of various CVJ disorders within our patient population. The age distribution in our study revealed a peak incidence of CVJ disorders in patients aged 41 to 50 years, contrasting with findings from other studies that report higher prevalence in younger age groups. This variation underscores the influence of demographic factors and regional differences on disease prevalence and presentation. A notable male predominance was observed among patients with CVJ disorders in our study, consistent with other literature, suggesting potential gender - specific differences in disease susceptibility or diagnostic patterns. Examination of anatomical structures such as the occipital condyles, clivus, atlas, and axis revealed predominantly normal findings, with occasional anomalies like basilar invagination, platybasia, and developmental abnormalities. These variations emphasize the importance of precise radiological assessment and anatomical knowledge in diagnosing and managing CVJ disorders effectively. Acquired conditions emerged as the leading cause of CVJ disorders in our study, reflecting the impact of trauma, degenerative changes, and inflammatory processes. This contrasts with studies highlighting developmental anomalies as predominant, indicating regional and demographic variability in disease etiology. The association of basilar invagination with atlantoaxial instability underscores the need for comprehensive evaluation and management strategies tailored to individual patient profiles. Similarly, the identification of rare tumors and syndromic associations at the CVJ highlights the diverse pathology encountered in clinical practice. Accurate classification and diagnosis of CVJ disorders are essential for guiding therapeutic interventions, which may include surgical correction, conservative management, or multidisciplinary approaches depending on the nature and severity of the condition. In conclusion, our study provides comprehensive insights into the epidemiology, clinical characteristics, and management considerations of CVJ disorders. Continued research and collaborative efforts are crucial for advancing diagnostic techniques and therapeutic strategies to improve outcomes for patients affected by these complex conditions.

7. Future Scope

Advanced Imaging Techniques:

- The expanding role of advanced imaging techniques like **diffusion tensor imaging (DTI)** and **functional MRI (fMRI)** in evaluating CVJ anomalies' impact on neural structures and blood flow.
- The potential of **artificial intelligence (AI) and machine learning** for automated analysis of CVJ images, aiding in faster and more accurate diagnosis.

Long - Term Management:
The importance of long - term radiological follow - up for patients with CVJ anomalies, including monitoring for potential complications like Chiari malformation progression.

Genetic Considerations:
The growing understanding of the genetic basis of CVJ anomalies and the potential for future genetic testing to guide treatment decisions.

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