RADIOLOGICAL EVALUATION OF CRANIOVERTEBRAL JUNCTION ANOMALIES
O. Joji Reddy¹, P. Kavitha², Abdul Gafoor³, Suresh B⁴, Harinath⁵, Padmalatha⁶

HOW TO CITE THIS ARTICLE:

ABSTRACT: INTRODUCTION: Detailed discussions of the CVJ are conspicuously absent in many standard textbooks and chapters addressing the skull or cervical spine, since it lies in between these regions. CVJ anomalies are common in India subcontinent. OBJECTIVES: To outline the normal anatomy and various abnormalities of craniovertebral junction. To evaluate the most common developmental and acquired craniovertebral junction abnormalities. CRANIOMETRY AND DIAGNOSIS: Radiological evaluation of CVJ requires identification of only a few anatomic structures. Over the years multiple lines, planes and angles have been described for assessment of CVJ relationship, initially with radiography and later with polytomography. Two lines have remained particularly useful for evaluation of CVJ relationship with virtually any imaging modality: the chamberlain’s line and weckenheim’s clivus base line. Two angles also continue to be useful: the welcher basal angle and atlanto occipital joint axis angle. PATIENTS AND METHODS: The prospective study of craniovertebral junction anomalies was carried out at Kurnool medical college, Government general hospital Kurnool from NOV 2012 to AUG 2014. The patients are subjected to clinical evaluation and radiological evaluation. OBSERVATIONS AND RESULTS: In our study there is male predominance with male to female ratio of 2:1. Majority of patients are in the age group of 11-40 (73.26%). The commonest symptom seen is weakness of extremities (70%) with associated numbness (50%). On clinical examination pyramidal tract involvement noticed in 70% of cases. Basilar invagination is the most common followed by Atlantoocoipital assimilation (40%) and AAD (30%). CONCLUSION: Computed tomography and magnetic resonance imaging are invaluable adjuncts to the plain radiographs in the evaluation of the craniovertebral junction anomalies. Chamberlain’s line and McGregor line are the most commonly applied craniometric measurements. KEYWORDS: Basilar Invagination, AAD, Condylar hypoplasia, Platybasia.

INTRODUCTION: The cranio vertebral junction or craniocervical junction (CVJ) is a collective term that refers to the occiput (posterior skull base), atlas, axis and supporting ligaments. It encloses the soft tissue structures of the cervico medullary junction (Medulla, spinal cord and lower cranial nerves).[1] With virtual extinction of polytomography in many institutions, detailed evaluation of this region fell to the realm of computed tomography or CT myelography. Now with wide spread availability of MR imaging, the CVJ is seen in virtually every sagital MR imaging study of brain. Dynamic MRI with flexion and extension views can be used to know the instability.[2] Wadia (1960) drew attention to its high incidence in India. The high incidence in India has been emphasized by several authors though the explanations for the same remain speculative. Apart from spinal tumours, tuberculosis and trauma, CVJ anomalies formed the 4th commonest cause of
spinal compression. Tuberculosis involvement of the CVJ is not uncommon in India. Timely detection and therapy would result in complete cure for an otherwise dangerous ailment.[3]

AIMS AND OBJECTIVES OF THE STUDY:
1. To outline the normal anatomy and various abnormalities of craniovertebral junction.
2. To evaluate the most common developmental and acquired craniovertebral junction abnormalities that is often encountered during plain film radiography, CT or MR evaluation.
3. To arrange frequently detected CVJ pathological imaging findings by etiological categories.
4. To stress the importance of CT and MRI in detailed evaluation of CVJ anomalies.
5. Importance of imaging findings in planning the surgical management.

EMBRYOLOGY AND DEVELOPMENT: Development of the cartilaginous cranium and adjacent structures begins during early weeks of intrauterine life. In 2nd week mesodermal cells condense in midline to form notochordal process. In 3rd week notochordal process invaginates between ectoderm and endoderm to form notochord. Dorsal ectoderm thickens to form neural groove which folds, fuses and becomes neural tube. Between 3rd and 5th week: Part of mesoderm which lies on either side of notochord (Paraxial mesoderm) gives rise to somites. Each sclerotome differentiates in to a cranial loosely arranged portion and a caudal compact portion by a fissure known as “fissure of Von Ebner”. Mesenchymal cells of the fissure condense around the notochord to form the intervertebral disc. Notochord disappears at the disc but persist as nucleus pulposus at disc.[4] The first four sclerotomes do not follow this course and fuse to form the occipital bone and posterior margin of foramen magnum. This membranous stage is followed by stages of chondrification and ossification.

Anatomical division of CVJ in to axial and flanking components has close analogy in the embryology of this region. The constituents of central pillar are all derived from axial portion of the occipital and upper two cervical sclerotome whereas the ring structures all come from lateral portion of these same sclerotome.[5]
Apical ligament and alar ligament develop from proatlas.\[^6\] Atlas forms from proatlas and first spinal sclerotome. Tip or apex of dens arises from centrum of proatlas, body of odontoid from first spinal sclerotome, axis body, facets and posterior arch of axis from second spinal sclerotome. At birth, the odontoid base is separate from body of axis by a cartilage which persists until the age of 8, later the centre gets ossified or may remain separate as osodontoidium. At birth, the tip of the dens is represented by a separate ossification centre, ossiculum terminale, which is usually seen at 3yrs of age, but fuses with the remainder by age 12.\[^7\] If it fails to fuse with dens, it is called ossiculum terminale persistens. Developmentally, this condition is the result of failure of proatlas and dens to fuse. Conaradi’s syndrome and Morquios syndrome are often associated with ossiculum terminale.\[^8\]

**OSSIFICATION CENTRES:**

**OCCIPUT AND BASIOCCIPUT:**
- Two occipital squamous portions – 2 Centre’s.
- Basiocciput (Clivus) - 1 Centre.
- 2 jugular tubercles - 2 Centre’s.
- 2 occipital condyles – 2 Centre’s.

**ATLAS:** Ossifies from 3 Centre’s.\[^9\]
- 2 Centre’s, 1 for each half of posterior arch with lateral mass appears at 7 to 9 weeks and unites at 3-4yrs. For anterior arch, a separate centre appears at 1 to 2yrs, unites with lateral mass at 6-8yrs.
**AXIS:** Ossifies from 5 primary and 2 secondary Centre’s.[9]

- 2 neural arches – 2 Centre’s appear at 7-8wks.
- Body of axis – 1 Centre appears at 4-5 months.
- Body of dens – 2 Centre’s appear at 6-7 months.

These join to form a conical mass after birth.

At birth, the axis is in four pieces which unite between 3 and 6 yrs. The summit of the odontoid is formed by a wedge shaped piece of cartilage which fills the deep cleft of conical mass. A Centre appears in this cartilage about the second year and it unites with the main mass of the odontoid process by 12yr. The base of the process is separated from the body of axis by a cartilaginous disc. Its circumference ossifies but the Centre remains cartilaginous until advanced age.

**ANATOMY OF CRANIOVERTEBRAL JUNCTION:** The craniovertebral junction (CVJ) is a collective term that refers to occiput, atlas, axis and supporting ligaments. It is transition zone between a mobile cranium and a relatively rigid spinal column. It encloses the soft tissue structures of the cervicomedullary junction (Medulla, spinal cord and lower cranial nerves).

**ATLAS:**

**AXIS:**

![Figure 2](image)

![Figure 3](image)
Ligamentous anatomy of CVJ: Principal stabilizing ligaments of C1 are,
  a. Transverse atlantal ligament.
  b. Alar ligament.

Secondary stabilizing ligaments of CVJ are more elastic and weaker than the primary ligaments;
  a. Apical ligament.
  b. Anterior and posterior atlanto occipital membranes.
  c. Tectorial membrane.
  d. Ligamentum flavum.
  e. ALL & PLL.
  f. Capsular ligaments.

Kinetic Anatomy of CVJ: CVJ units are unique with respect to rest of the spine in that they do not bear weight through discs, but rather through synovial joints.

Atlanto-occipital joints: The joint is biaxial having movements only around the transverse and AP axis. Flexion – 10 degrees, extension -25 degrees, lateral bending – 8 degrees and no rotation

Atlantoaxial Joint: Rotation of atlas occurs around the odontoid process like a wheel around an axle and its axis passes centrally through the annulus osteofibrosis. Flexion – 5 degrees, extension - 10 degrees, lateral bending -0 degrees, axial rotation – 45 degrees.

The ratio of extension to flexion is approximately 2: 1 and this ratio maintains itself at both O-A & A-A joints. Total rotation of entire cervical spine is up to 90 degrees and approximately half occurs at the A-A joint. Thus the anatomical structures that provide stability of the O-A joint include – Cup shaped configuration of the joint & Anterior and posterior A-O...
membranes. Additional stability is provided by ligamentous connections between the occiput and the axis that include the tectorial membrane, alar and apical ligaments.\[14\]

**Neural structures related to CVJ:** Caudal portion of brain stem;
- Cerebellum (Tonsils, Biventral lobules and lower part of vermis;
- Fourth ventricle.
- Rostral part of spinal cord.
- Lower four cranial nerves and upper cervical nerves. (C1, C2, C3)

**Arterial anatomy of CVJ:** The major arteries related to CVJ are;
- Vertebral.
- PICA.
- Meningeal branches of vertebral, external and internal carotid arteries.

The tonsillomedullary PICA segment, which forms the caudal loop related to the lower part of the tonsil, is most intimately related to foramen magnum.\[15\]

**DIAGNOSIS OF CRANIOVERTEBRAL JUNCTION ANOMALIES:** Diagnosis can be divided into;

A. **Clinical:**
   i. General physical examination.
   ii. Neurological.

B. **Radiological:**
   i. Plain X-ray studies.
   ii. Conventional tomography.
   iii. Myelography.
   iv. CT scan.
   v. MRI.
   vi. Angiography.

**CLINICAL EXAMINATION: General Physical Examination:** An abnormal general appearance more often involving the neck is seen in patients with congenital abnormalities of craniovertebral junction. Most common finding is atlantooccipital fusion which has high incidence in patients with klippel – feil syndrome.

**Clinical:**
1. Short neck definition: sinh’s criteria:
   - Height/ neck index can be calculated as follows:
     - Height in cms/ neck in cms $\times 100$.
     - When it is greater than 13 it is considered as short neck.
2. Low hair line _ hair line at the level of spine of 5th cervical vertebra.
3. Malformations of face and mouth: Hemifacial atrophy, Cleft lip & palate, High arched palate, Micrognathia, Microtia, Low set ears, Hearing loss, Lateral rectus palsy, Duane`s contracture.
7. Sprengel shoulder.
8. Accessory nipple

**CLINICAL FEATURES OF CRANIO–VERTEBRAL ANOMALIES:** Compromise of cervicomedullary junction results in a multiplicity of signs and symptoms which may be indicated by dysfunction of the brainstem, cerebellum, cervical spinal cord, cranial nerves, cervical roots etc. The symptoms are the direct result of:
   a. Direct compression of the neural tissue by bone or soft tissue.
   b. Compromise of their blood supply or.
   c. Combination of both.

**Age:** The age of presentation can vary between first to the fourth decade.

**Precipitating Factors:** In majority, the disease manifests spontaneously. Nonetheless, precipitating factor may be so trivial especially in atlantoaxial dislocation. Carrying heavy loads on the head or back is a common practice in the tropics and is thought to be a predisposing factor in patients of AAD. Basilar invagination is significantly common in India. Even in India, there is a disproportionately high incidence in north-western belt of country. No genetic factor has been identified that could explain the discrepancy in incidence. Degeneration of spine in general and cranio vertebral junction in particular can lead to instability and subsequently to basilar invagination.

**Clinical Presentation:** The clinical manifestations can be grouped as under;
- **Cervical Manifestations:** pain, stiffness and varying degree of restriction of neck movements are frequent. Torticollis is common.
- **Transient Attacks (Vascular Symptoms):** These symptoms may be manifested as unconsciousness, vertigo, intermittent periods of altered sensorium, confusion, transient paralysis, transient loss of visual fields, transient paraesthesias etc.[16]
- **Myelopathic Features:** Motor deficits include monoparesis, hemiparesis, paraparesis and quadriplegia.[17] Posterior column signs affecting all the four limbs to an equal extent in the form of loss of vibration and position sense are next in frequency.[18]
- **Brain Stem Dysfunction:** Brainstem dysfunction manifests as internuclear ophthalmoplegia, dysphagia, respiratory abnormalities and sleep apnoea.[19] Down beat nystagmus can be present and may be indicative of an ACM.[20]
- **Cerebellar Signs:** They are usually seen in association with Chiari malformation or in basilar invagination or occipitalization.[20]
- **Cranial Nerve Involvement:** In klippel – Fiel syndrome, as many as 30 % of patients can have hearing impairment.[21,22]
**Other Features:** localized wasting affecting the shoulder girdle and the upper limbs with or without fasciculations, may be present.

**CRANIOMETRY:**

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Definition</th>
<th>Normal measurements</th>
<th>Implication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Chamberlains` line (palato occipital line)</td>
<td>Joins posterior part of hard palate to opisthion</td>
<td>Odontoid process should not project above this line &gt; 3 mm</td>
<td>Tip of dens &gt;6.6 mm above this line strongly indicates basilar invagination</td>
</tr>
<tr>
<td>2. McGregor`s line (basal line)</td>
<td>Hard palate to lowest point of occipital bone[23]</td>
<td>Tip of dens should not exceed 4.5 mm above the line</td>
<td>Tip of dens &gt; 4.5 mm above this line indicates basilar invagination</td>
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<tr>
<td>3. McRae`s line (foramen magnum line)</td>
<td>Joins anterior and posterior ends of foramen magnum</td>
<td>Tip of dens should not exceed this line.</td>
<td>When effective sagital canal diameter is less than 20 mm neurological symptoms occur</td>
</tr>
<tr>
<td>4. Wackenheim`s line (clivus canal line)</td>
<td>Line drawn the clivus in to cervical canal</td>
<td>Odontoid tip is ventral and tangential to this line.</td>
<td>Odontoid process transects the line in basilar invagination or forward position of skull</td>
</tr>
<tr>
<td>5. Boogard`s line</td>
<td>Joins nasion to opisthion</td>
<td>Basion below this line</td>
<td>Altered in basilar impression</td>
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<tr>
<td>6. Ranawat line/criterion</td>
<td>Distance between the line joining centre of anterior &amp; posterior arch of C1 to the line along the axis of odontoid from the centre of pedicle of C2.</td>
<td>Normal average distance in men is 17mm and in women 15mm</td>
<td>A decrease in this distance indicates cephalad migration of C2.</td>
</tr>
<tr>
<td>7. Klau`s index</td>
<td>Distance between tip of dens and tuberculum – cruciate line</td>
<td>40 – 41 mm</td>
<td>&lt; 30 mm seen in basilar invagination</td>
</tr>
<tr>
<td>8. Spinolamellar line</td>
<td>Line drawn from inter occipital ridge above and down along the fused spinous process C2 and C3.</td>
<td>Should intersect posterior arch of atlas</td>
<td>If atlas is fused, posterior arch is anterior to the line, posterior compression of spinal cord may occur.</td>
</tr>
<tr>
<td>9. Power`s ratio</td>
<td>Calculated by dividing the distance between the tip of basion to</td>
<td>Normal power`s ratio &lt; 1.</td>
<td>Used in detection of atlanto occipital dissociation[25]</td>
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<tr>
<td></td>
<td>Description</td>
<td>Normal Range</td>
<td>Abnormality</td>
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<td>----------------------------------------------------------------------------</td>
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<tr>
<td>10. BAI</td>
<td>Distance between basion and rostral extension of posterior cortical margin of body of axis.</td>
<td>Should be &lt; 12mm</td>
<td>Increased in Atlanto occipital dissociation[25]</td>
</tr>
<tr>
<td>11. BDI</td>
<td>Distance from the most inferior portion of the basion to the closest point of the superior aspect of dens</td>
<td>Should be &lt; 12mm</td>
<td>Increased in Atlanto occipital dissociation[25]</td>
</tr>
<tr>
<td>12. ADI</td>
<td>Distance between posterior aspect of anterior arch of C1 to the most anterior aspect of dens</td>
<td>Normally &lt;3mm in men and &lt; 2.5 mm in women</td>
<td>Increased in Atlanto axial dissociation[26]</td>
</tr>
<tr>
<td>13. PADI</td>
<td>Distance between posterior surface of odontoid and anterior margin of posterior ring of C1.</td>
<td>Normal valve at C1 – 17 – 29mm</td>
<td>&lt;14mm predicts cord compression [25]</td>
</tr>
<tr>
<td>14. AOI</td>
<td>Distance between the articular surfaces of the occipital condyles and the lateral mass of C1.</td>
<td>Normally &lt;5mm in children and &lt; 2mm in adults</td>
<td>Increased in Atlanto occipital dissociation[27,28]</td>
</tr>
<tr>
<td>15. Fishgold’s digastrics line</td>
<td>Joins the fossae for digastic muscles on undersurface of skull just medial to mastoid process</td>
<td>Dens tip should not project above this line, central axis of dens should be perpendicular to the line</td>
<td>Corresponds to the Mc Rae`s line on lateral view, may be oblique in U/L Condylar hypoplasia, oblique odontoid suggests paramedian abnormality [29]</td>
</tr>
<tr>
<td>16. Fishgold’s bimastoid line</td>
<td>Line connecting tips of mastoid process</td>
<td>Runs across atlantooccipital joints, line is 10 mm below digastrics line</td>
<td>Odontoid tip &gt; 10 mm above the line indicates basilar invagination[29]</td>
</tr>
<tr>
<td>17. Welcher basal angle</td>
<td>Angle between nasion – tuberculum line to</td>
<td>Should be &lt; 132 degrees</td>
<td>&gt;140 degrees indicates platybasia[30, 31]</td>
</tr>
<tr>
<td>18. Clivus canal angle</td>
<td>Between clivus and posterior axial line</td>
<td>Flexion -150 degrees Extension – 180 degrees</td>
<td>Ventral spinal cord compression occurs when the angle &lt; 150 degrees(^{[31]})</td>
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<tr>
<td>19. Atlanto occipital joint axis angle</td>
<td>Angle of axis of atlanto occipital joints</td>
<td>124 – 127 degrees. Should be measured in plane of dens on tomography</td>
<td>Angle is wider in condylar hypoplasia(^{[30]})</td>
</tr>
<tr>
<td>20. Boogard’s angle</td>
<td>A line is drawn from basion to opisthion and another line along the plane of clivus to the basion, intersecting the first line</td>
<td>The normal valve is 126 +/- 6 degrees.</td>
<td>In platybasia it exceeds 136 degrees</td>
</tr>
<tr>
<td>21. Bulls angle</td>
<td>Posterior angle between first line from posterior tip of hard palate to posterior margin of foramen magnum to the second line b/w anterior and posterior tubercles</td>
<td>&lt; 10 degrees – normal 10 – 13 degrees - tendency to BI</td>
<td>&gt; 13 degrees indicates basilar invagination</td>
</tr>
</tbody>
</table>
**Welcher Basal Angle:** angle subtended by the junction of nasion - tuberculum to basion tuberculum tangents

**Clivus Canal Angle:** Angle formed between Wackenheims line and posterior vertbral body line. Atlanto occipital joint axis angle: Angle formed by the junction of lines drawn through atlantooccipital joints.
CLASSIFICATION OF ANOMALIES OF CVJ: The anomalies in the CVJ may be;
   a. Skeletal.
   b. Neural.
   c. Combination of both.

The common skeletal craniovertebral anomalies can be classified as follows;\textsuperscript{[32]}
   1. Basilar invagination and basilar impression.
   2. Malformations of the occipital bone.
   3. Malformation of atlas.
   5. Atlantooccipital dislocation.

The neural anomalies at the CVJ are;
   1. Arnold chiari malformation.
   2. Syringohydromyelia.
   3. A combination of above two.

Basilar invagination is used for the primary form of basilar impression consisting of a distinct developmental defect of the chondrocranium which is often associated with other bony anomalies such as basiocciput hypoplasia, occipital condyle hypoplasia, various atlanto occipital assimilations. There is an increased prevalence of neural dysgenesis associated with basilar invagination such as chiari malformation or syringohydromyelia.\textsuperscript{[30,32,33]}

Basilar Impression (Secondary Basilar Invagination): Basilar impression refers to secondary/ acquired forms of basilar invagination and is due to softening of bone and is seen in conditions such as rickets, hyperparathyroidism, osteogenesis imperfecta, pagets disease, etc.,

Condylar Tertius: ossified remnant may be present at the distal end of clivus, called as third occipital condyle or condylar tertius. Although typically single, multiple supranumerary ossicles may be present.\textsuperscript{[34]}

Condylar Hypoplasia: In condylar hypoplasia the occipital condyles are underdeveloped and have flattened appearance leading to basilar invagination and widening of atlanto occipital joint axis angle.\textsuperscript{[35]}

Basiocciput hypoplasia: It results in shortening of clivus and violation of chamberlain line and is virtually always associated with basilar invagination. The clivus canal angle is typically decreased and there is bowstring deformity of the cervicomedullary junction.\textsuperscript{[36,30]}

ANOMALIES OF ATLAS: With the exception of various atlantooccipital assimilation various atlas anomalies when isolated anomalies produce no abnormal CVJ relationships.
Osodoontoideum: This term is first introduced by Giacomini in 1886, refers to an independent osseous structure lying cephalad to the axis body in the location of odontoid process. The anterior arch of the atlas is rounded and hypertrophic but the posterior arch is hypoplastic.

Occasionally, the differentiation between an osodoontoideum and a type 2 odontoid fracture on a lateral radiograph may be problematic.

Persistent Ossiculum Terminale: Also called Bergman ossicle, results from failure of fusion of terminal ossicle to the remainder of odontoid process. The fusion typically is accomplished by 12 yrs of age.

KLIPPEL FEIL SYNDROME: Classical triad seen in < 50 % of cases: Low posterior hair line, Short neck, limited range of neck movements. The most consistent finding is limitation of neck movement.[37]

ATLANTOAXIAL DISLOCATION: Basic defect may be an abnormal odontoid, atlanto-occipital fusion or laxity of the transverse atlantal ligament leading ultimately to narrowing of spinal canal and narrowing of spinal canal and impingement on the neural elements. Greenberg’s classification: All cases regardless of etiology are classified as those caused by
1. Incompetence of odontoid process: congenital, traumatic, Infections, tumour.
2. Incompetence of transverse atlantal ligament (TAL): congenital (idiopathic mongolism), traumatic, inflammation (Pharynx and nasopharynx infections, tuberculosis. RA, Ankylosing spondylitis). A hook like appearance of odontoid process occurs secondary to the cruciate ligament pannus eroding in to the odontoid process in Rheumatoid arthritis.[38]

PATIENTS AND METHODS: The prospective study of craniovertebral junction anomalies was carried out at Kurnool medical college, Government general hospital Kurnool from Nov 2012 to Aug 2014. The patients are subjected to the following;
  a. Clinical evaluation.
  b. Radiological evaluation.

The patients taken in to study include patients with specific symptoms related to CVJ and patients with nonspecific symptoms having CVJ anomalies. Both group of patients having CVJ anomalies are evaluated using a predetermined proforma and they were subjected to radiological investigation. The proforma is designed keeping in mind the diversity of clinical manifestations of CVJ.

The following radiological investigations were done.

Plain radiography: All patients are subjected to plain radiographs of CVJ flexion, extension views on lateral projection to r/o atlantoaxial subluxation/occipitalisation of atlas and open mouth view are also obtained.

Computerized tomography: MDCT 16 Slice belonging to GE Company was used. Volume scans were performed from the craniovertebral junction down to the level of C7 vertebral body employing 0.625 mm sections. All patients were subjected to CT. Sagital and coronal reformations
were done. 3 D reformation was done in most of the patients which helped in easily understanding the bony CVJ anomalies.\textsuperscript{(23)}

**MRI:** MRI was performed in most of the patients using 0.35 T MRI belonging to XGY opera company. The following sequences were done

- T1W – Sagittal plane, axial.
- T2W- Sagittal plane, axial, coronal.
- GRE – sagittal & coronal plane if required for osseous details. Post-operative radiographs were taken where ever possible.

**CASE 1:**

![X ray C Spine showing AAD and Basilar invagination](image)

![Plain CT sagital and axial sections in bone window shows AAD with Dystopic Osodontoideum with Basilar Invagination and cleft of posterior arch of atlas (Arrow)](image)
CASE 2:

T1 & IRFSE sagital images showing AAD (White arrow) and compression of spinal cord by dens at foramen magnum causing edema (Black arrow)

X ray C spine in flexion view showing basilar invagination (black arrow), basiocciput hypoplasia (White arrow)

Violation of chamberlains line but wackenheims clival line and McRaes line are normal

Klau’s posterior fossa height index very much reduced and there is platybasia
Plain CT sagittal images in bone window showing Basiocciput hypoplasia (Curved arrow), Basilar invagination, Atlanto occipital assimilation (Black arrow), Klippel feil syndrome

Plain CT coronal image in bone window showing condylar hypoplasia with atlantooccipital assimilation, butterfly vertebra (Black Arrow)

MRI T2 weighted images in sagital and coronal section showing Basiocciput hypoplasia, Basilar invagination, Atlanto occipital assimilation, condylar hypoplasia and multiple butterfly vertebra
CASE 3:

Sagital T1 weighted MRI image showing elongated peg like cerebellar tonsil extending below foramen magnum with posteriorly angled odontoid

Plain X rays C spine in flexion and extension showing hypoplastic posterior arch

Sagital CT section in bone window showing posteriorly angled C2. Axial CT section in bone window showing cleft in posterior arch.
CASE 4:

T1 and T2 sagital images showing Basilar invagation of (Curved arrow), Atlantoaxial subluxation, Atlantooccipital assimilation, Spinal cord compression and edema at foramen magnum (Black arrow)

CASE 5:

X ray C spine lateral view in flexion and extension showing increased AAD (Curved arrow), Basilar invagination(Black arrow) and bone graft with suture material (White arrow)
OBSERVATIONS AND RESULTS: This prospective study was carried out from Nov 2012 to August 2014 i.e., a period of 21 months. 30 cases of CVJ anomalies admitted during that period in Kurnool medical college, Government General hospital were studied in relation to clinical presentation, radiography, CT scans, MRI and postoperative findings (wherever possible). The summary of the study is given below;

<table>
<thead>
<tr>
<th>Sex</th>
<th>No. of patients</th>
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<tbody>
<tr>
<td>Male</td>
<td>20</td>
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<tr>
<td>Female</td>
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<td>Total</td>
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<table>
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<tr>
<th>Age</th>
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<td>16.65</td>
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<td>11-20</td>
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<td>21-30</td>
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<tr>
<td>TOTAL</td>
<td>30</td>
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</table>

Table 1: Sex Incidence

Table 2: Age Distribution
Graph 1: Clinical features in the study group (n=30)

Graph 2: Clinical examination findings in the study group (n=30)

Graph 3: The distribution of CVJ anomalies in the study group (Values shown are in %)
Basilar invagination is the most common anomaly noticed in our study. (50%) Basilar invagination is the most common followed by Atlantooccipital assimilation (40%) and AAD (30%). Arnold Chiari malformation is the most common anomaly noticed (30%) of soft tissue anomaly. Osodontoideum (20%) and Block vertebrae (17%) are the next most common anomaly noticed. split atlas was noticed in two cases. Condylar hypoplasia and basioociput hypoplasia noticed in 13.2% of the cases. Normal anatomical variant ponticulusposticus noticed in one case. Posterior arch anomaly noticed in 14% of cases. Spinal cord compression noticed in 43% of cases, the location being foramen magnum in 90% of cases.

<table>
<thead>
<tr>
<th>Soft tissue anomaly</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACM</td>
<td>8</td>
<td>88.8</td>
</tr>
<tr>
<td>Syrinx</td>
<td>7</td>
<td>77.7</td>
</tr>
<tr>
<td>Dandy walker variant</td>
<td>1</td>
<td>11.1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>9</strong></td>
<td></td>
</tr>
</tbody>
</table>

Table 3: Various Types Of Bony CVJ Anomalies Seen At Kurnool Medical College

The most common soft tissue anomaly in our study is ACM type I in 88.8 % of cases. One case of Dandy walker variant is seen.

**DISCUSSION:** The subject of craniovertebral junction anomalies is under discussion and evaluation for over a century. A number of classical reviews have attempted to clarify a variety of complex associated issues. Despite the volumes of publications on the subject, it appears that the last word has not yet been said. Craniovertebral anomalies are more frequently found in the Indian subcontinent than anywhere else in the world. Even in India, these anomalies are more frequently documented form Uttar Pradesh, Bihar, Rajasthan and parts of Gujarat. The reasons for this geographical clustering are speculative. Over the years, there have been a relatively large number of significant and authoritative publications from India on this subject.

The surgical management of congenital craniovertebral anomalies is complex due to the relative difficulty in accessing the region, critical relationships of neurovascular structures and the intricate biomechanical issues involved. Basilar invagination forms a prominent component of the craniovertebral anomalies. Chiari malformation and syringomyelia are common associates of basilar invagination and are the soft tissue components of the dysgenesis. Plain radiological and tomographic parameters have been principally used to diagnose basilar invagination for many years. There has been a renewed interest in the normal anatomy and pathologic lesionsof the craniovertebral junction with the development of imaging by high-resolution computer tomography (CT) scan and magnetic resonance imaging (MRI). Improved imaging has provided an opportunity to clearly observe the bony abnormality and the distorted neural and vascular relationships. Dynamic MRI and CT scan have helped in the evaluation of the pathology of basilarinvagination, in the assessment of the biomechanics of the joints and in the formulation of a rational surgical strategy. Despite the clarity of imaging, controversy regarding the management of basilar invagination continues. Even the natural history has not been clearly elucidated in the literature.
The incidence of craniovertebral junction anomalies appears to be frequent in our area. This prompted us to carry out this prospective study comprising 30 cases admitted in the various departments mostly in the neurosurgery department over a period of 26 months from Jun 2012 to Aug 2014. Observations relevant to this study has been made and a comparative evaluation and discussion is done.

In our study there is male predominance with male to female ratio of 2:1 which coincided with the study conducted by N.J.M. Mwang’ombe. The youngest patient in our study was 1 year old whereas the oldest patient was 60. Majority of patients are in the age group of 11-40 (73.26%) mostly in the 30-40 age group (26.64%) followed by 20-30 (23.31%) & 11-20 (23.31%). There was only two patients below 10 months and only one patient above 60 yrs. In studies conducted by N.J.M. Mwang’ombe the highest peak of incidence was in third decade, as in our study. Somewhat high incidence of detection of cases below 1yr in this study is due to use of MDCT which helped in accurate assessment. In our study predisposing factor was present in 25% of cases. H/O trauma in the form of carrying heavy loads noted in 7 cases. Minor trauma history noted in 75% of cases of basilar invagination. H/o rheumatoid arthritis present in one case.

The commonest symptom seen is weakness of extremities (70%) with associated numbness (50%). Other symptoms were headache (40%) and neck pain (40%). On clinical examination pyramidal tract involvement noticed in 70% of cases. Low hair line (25%) and short neck (25%) are the other findings on clinical examination. Hand muscle wasting noticed in 15% of cases. Other common neurological findings were nystagmus, sensory loss, cerebellar ataxia. Respiratory or bladder involvement not seen in any of the cases in my study. Motor symptoms are the predominant manifestations even in studies conducted by N.J.M. Mwang’ombe followed by sensory symptoms. ACM patients usually presented with cerebellar dysfunction signs such as down beat nystagmus, ataxia e.t.c. They were seen in all 30 patients which formed the criteria for our study. Basilar invagination is the most common anomaly noticed in our study (50%). Basilar invagination is the most common followed by Atlantococipital assimilation (40%) and AAD (30%). The most common type of lesions in the study conducted by by N.J.M. Mwang’ombe were basilar invagination (48%) followed by atlanto-axial dislocation (28%) and occipitisation of the atlas (28%).

Arnold Chiari malformation is the most common anomaly noticed (85%) of soft tissue anomaly. Osodontoideum (20%) and Block vertebrae (17%) are the next most common anomaly noticed. split atlas was noticed in two cases. Condylar hypoplasia and basiociput hypoplasia noticed in 13.2 % of the cases. Normal anatomical variant ponticulusposticus noticed in one case. Posterior arch anomaly noticed in 14% of cases. Spinal cord compression noticed in 46% of cases, the location being foramen magnum in 90% of cases.

In subgroup of patients having basilar invagination where there was clear radiological evidence of instability of the region that was manifested by distancing of the odontoid process from the anterior arch of the atlas. Considering this current evaluation basilar invagination can be into two groups based on parameters that determine an alternative treatment strategy. In Group A basilar invagination there was a ‘fixed’ atlantoaxial dislocation and the tip of the odontoid process ‘invaginated’ into the foramen magnum and was above the Chamberlain line, McRae line.
of foramen magnum and Wackenheim’s clivalline. The definition of basilar invagination of prolapsed of the cervical spine into the base of the skull, as suggested by von Torklus, was suitable for this group of patients. Group B basilar invagination was where the odontoid process and clivus remained anatomically aligned despite the presence of basilar invagination and other associated anomalies. In this group, the tip of the odontoid process was above Chamberlain’s line but below McRae’s and Wackenheim’s lines. The radiological findings suggested that the odontoid process in Group A patients resulted in direct compression of the brainstem. Essentially, in Group A basilar invagination, there was an element of instability of the region that was manifested by the tip of the odontoid process distancing itself from the anterior arch of the atlas or the lower end of the clivus. Group A basilar invagination forms a larger subgroup of patients that are encountered in Indian subcontinent. The above observations are similar to those made by Goel A.

The analysis of basilar invagination into two groups on the basis of Chamberlain’s line suggested that the basilar invagination is much more severe in Group B than in Group A. The distance from the tip of the odontoid to the pontomedullary junction, as observed on MRI, was seen to be a useful index to define the reduction of the posterior cranial fossa bone size. The distance was markedly reduced in Group B patients while it was relatively large in Group A patients.

Out of 14 cases of basilar invagination, 10 cases (71%) belonged to Group A patients and 4 cases (29%) belonged due to Group B. Among secondary causes one case of basilar invagination was due to flurosis and other case was due to rheumatoid arthritis. Osodontoideum was noticed in 50% cases of Group A patients. Among the cases of Osodontoideum, 90% are of dystopic variety associated with AAD and Basilar invagination. One case of orthotopic Osodontoideum was noticed with posterior fixation, which was operated previously for basilar invagination. Although basilar invagination is the most common observation in this study, the frequency is less when compared to others studies and also the frequency of ACM is more in this study. The reason for this being that the cases of basilar invagination and AAD that needed anterior transoral approach for surgical correction were not done in our institution due to lack of instruments and cases of ACM were operated more by foramen magnum decompression.

Out of 30 patients, 40% of patients had atlantooccipital assimilation. Both Group A and Group B patients have atlantooccipital assimilation. 100% of Group B patients had atlantooccipital assimilation. Other occipital bone anomalies such as condylar hypoplasia and basioccipital hypoplasiawere noticed in 100% of cases of Group B Basilar invagination. Platybasia was observed in 13% of cases. Platybasia was noticed both in Group A and Group B but a higher association was found with Group B.

Posterior cranial fossa volume or the Klaus’ height index measured on the MRI was seen to be much more accurate than the conventional measurements based on plain X-rays. The tentorium could be clearly identified on MRI and the distance of the tip of the odontoid from the line of the tentorium indicated the height of the posterior cranial fossa. On the basis of Klaus’ index, the posterior fossa height was found to be markedly reduced in Group B patients while it was only moderately affected in Group A patients.
The effective brainstem girth measured on MRI was a useful additional parameter. Whilst the brainstem girth was markedly reduced in Group A patients, the girth was only marginally affected or unaffected in Group B patients indicating thereby that there was no direct brainstem compression as a result of the odontoid process in the latter group. The anterior concavity of the brainstem was smooth in curvature in Group B patients while it was acute in Group A patients, the angle being formed by the tip of the odontoid process. In Group A, the brainstem distortion was directly a result of indentation of the odontoid process.\[40\]

Among the soft tissue anomaly, ACMis observed in 80 % of cases. Dandy walker variant due to hypoplastic inferior cerebellar vermis is noticed in one case.

The standard and most accepted form of treatment of Group A basilar invagination is a transoral decompression. The majority of the authors recommend a posterior occipitocervical fixation following the anterior decompression.\[41\] The new technique of craniovertebral realignment by wide removal of atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provided a unique opportunity to obtain reduction of the basilar invagination and of atlantoaxial dislocation in Group A.\[40\] Patients in Group B benefited by foramen magnum bony decompression.\[33,41\] The procedure resulted in amelioration of symptoms and at least an arrest in the progression of the disability.

CONCLUSIONS: Plain radiographs form the initial modality of investigation in evaluating a case of Craniovertebral junction anomaly. Computed tomography and magnetic resonance imaging are invaluable adjuncts to the plain radiographs in the evaluation of the craniovertebral junction anomalies. Chamberlain’s line and McGregor line are the most commonly applied craniometric measurements. MRI is more sensitive in detecting soft tissue anomalies whereas CT is more sensitive in detecting the bony CVJ anomalies.

Abnormal kinetodynamics which develop at the site due to bony anomalies predispose to instability and subsequent neurological deficit. So early diagnosis helps in the appropriate management of patient without resulting in a state of irreversible neurological damage. Not all patients with the bony CVJ anomalies develop the neurological deficit.

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