ORIGINAL RESEARCH PAPER

Radiodiagnosis

KEY WORDS: Congenital, Craniovertebral Junction, Plain Radiograph, Computed Tomography

CONGENITAL CRANIOVERTEBRAL JUNCTION ANOMALIES ON X-RAY AND ITS CORRELATION WITH MULTIDETECTOR COMPUTED TOMOGRAPHY

Dr Rajeev Singh Resident, Department Of Radio-Diagnosis, Gajra Raja Medical College, Gwalior, Madhya Pradesh.

Dr Pankaj Yadav Professor, Department Of Radio-Diagnosis, Gajra Raja Medical College, Gwalior, Madhya Pradesh.

Dr Ratnesh Jain* Assistant Professor, Department Of Radio-Diagnosis, Gajra Raja Medical College, Gwalior, Madhya Pradesh *Corresponding Author

ABSTRACT

OBJECTIVES: To outline the normal anatomy and various abnormalities of craniovertebral junction. To evaluate the most common developmental craniovertebral junction abnormalities. **MATERIAL & METHODS:** A prospective study carried out at the Department of Radiology GR Medical College JAH Hospital Gwalior. In our study 26 nonconsecutive patients with all age groups with clinical suspicion of congenital craniovertebral junction anomalies referred from the neurosurgery department for diagnosis and evaluation would be subjected to Computed tomography & x-ray of head & neck from February 2019 to July 2020 were studied. **RESULTS:** In our study of 26 patients, male to female ratio was 3.3:1(20males and 6females) . Congenital CVJ anomalies were seen in 26 cases. Anomalies seen were either singly or in combination. The most common anomaly was basilar invagination (BI) seen in 73% of cases. **CONCLUSION**-Plain radiographs form the initial modality of investigation in evaluating a case of congenital craniovertebral junction anomaly. Computed tomography are invaluable adjuncts to the plain radiographs in the evaluation of the congenital craniovertebral junction anomalies.

INTRODUCTION

Craniovertebral Junction, being the transit zone between cranium and spine, is the most complex and dynamic region of the cervical spine. It has complex bony anatomy and intricate tissues and major neurovascular structures. The craniovertebral junction (or craniocervical) (CVJ) consists of the occiput (posterior skull base), foramen magnum, clivus, atlas, axis, ligaments of atlantooccipital and atlantoaxial articulations.[1,2] It encloses the soft tissue structures of the cervicomedullary junction (medulla, spinal cord, and lower cranial nerves). CVJ may be congenital, developmental, or due to malformation secondary to any acquired disease process. These anomalies can lead to neural and vascular compromise, obstructive hydrocephalus, and cerebrospinal fluid dynamics.[3]

Conventionally, an X-ray of the skull with the cervical spine was the imaging modality used for the assessment of basilar impression. CVJ can now be visualized much better using modern-day imaging modalities including computed tomography (CT) which offers a three-dimensional visualization of this region with relatively complex anatomy. CT scan can provide good spatial resolution combined with speed and ability to perform high-quality multiplanar imaging. It provides details of the bony anatomy which is superior to that of plain X-ray. CT is a reliable diagnostic modality for the accurate assessment of the classical lines and angles, transverse and anteroposterior (AP) diameters of the foramen magnum and spinal canal. The skull baselines namely Chamberlain's, McGregor's, and McRae's lines are the standard reference measurements used for the evaluation of basilar impression, in defining the anatomy of the CVJ, in pre and post-operative assessment and follow up of any CVJ pathology.[4,5]

The aim of the current study was to review craniovertebral junction complex anatomy, normal variants, congenital, and acquired abnormalities. Conventionally, an X-ray of the skull with the cervical spine was the imaging modality used for the assessment of CVJ anomalies, Due to advances in computed tomography complex CVJ anatomy is well understood.

AIMS AND OBJECTIVES

The Aims & Objectives of this study are as follows;

1. To study the incidence of various congenital CVJ Anomalies.
2. To outline normal anatomy of the craniovertebral junction (CVJ).
3. To study the most common developmental CVJ abnormalities.
4. To arrange frequently detected CVJ pathologic imaging findings.

MATERIAL AND METHODS

A prospective study carried out at the Department of Radiology GR Medical College JAH Hospital Gwalior. In our study nonconsecutive patients with all age groups with clinical suspicion of congenital craniovertebral junction anomalies referred from the neurosurgery department for diagnosis and evaluation would be subjected to Computed tomography & x-ray of head & neck from February 2019 to July 2020 were studied. Inclusion Criteria: Patients with congenital craniovertebral junction anomalies irrespective of age. Exclusion criteria: Not providing consent, Acquired craniovertebral junction anomalies & Patients with Chiari malformation and associated soft tissue lesions.

TECHNIQUE OF XRAY & CT SCAN examination

The x-ray was taken by 625 milliamperes, 3phase(440 voltage) Allenger medical system machine for analyzing x-ray, a spherical marker of known dimensions was kept in the field for comparison above the thyroid cartilage in the midline.

For taking an x-ray, the patient was positioned in a supine position with the patient's shoulder pulled down with the neck lying against a vertical cassette holder. The mid coronal plane (The plane that passes through the mastoid tips) was in the midline of the cassette. The patient was asked to elevate the chin to prevent the superimposition of the upper cervical spine by the mandible. The central X-ray was perpendicular to the cassette and was directed horizontally to C-4 (level of the upper margin of thyroid cartilage).

CT Scan Measurements –

CT images of the craniovertebral junction on 128 Slice CT Siemens Somatom-AS using a rotation time of 600 msec, tube voltage of 120 kV, and tube current of 120–250 mA. The images were reconstructed into 1mm-thick slices with space between slices of 0.3 mm. All scans were analyzed on a present bone window setting: length 300 HU and width 2500 HU. The distance between the tip of the odontoid process and the skull

baselines was measured on the sagittal image of the CT scan with the coronal section centered on the dens.[6]

This study also compares dimensions as measured on X-ray and CT scan to assess whether X-ray is a reliable diagnostic tool to evaluate the CVJ in an emergency setting.

OBSERVATIONS & RESULTS

TABLE 1. Age Distribution of patients studied:-

Age in Years	Number of Patients	Percentage %
1-10	2	7.6%
11-20	10	38.4%
21-30	6	23%
31-40	4	15.3%
41-50	3	11.5%
>50	1	3.8%
TOTAL	26	100.0%

TABLE 2. Gender Distribution of patients studied:-

Gender	Number of patients	Percentage %
Male	20	77%
Female	6	23%
TOTAL	26	100%

Table No. : 3 Distribution of X-ray and CT finding in Congenital CVJ anomalies (n= 26)

Developmental anomalies	No. of patients In CT		No. of patients In X-ray	
	No. of cases	(%)	No. of cases	(%)
Basilar invagination (BI)	19	73%	9	34.2%
Atlanto axial dislocation (AAD)	17	65%	9	34.2%
Atlanto occipital assimilation (AOA)	16	61%	6	23%
Platybasia (PLB)	2	7.6%	2	7.6%
Osodontoideum (OO)	4	15%	2	7.6%
Ossiculum terminale (OT)	2	7.6%	1	3.8%
Odontoid hypoplasia(OH)	3	11.5%	0	0%
C2-C3 Block Vertebra (BV)	11	42%	7	27%
Rachischisis	2	7.6%	0	0%
Hypoplastic atlas	2	7.6%	0	0%
Hypoplastic clivus	1	3.8%	0	0%

Table 4: Combinations of Congenital CVJ Anomalies (n=26)

Combination	Number	Percentage
BI +AOA	13	50%
BI+AAD	11	42.3%
AAD+AOA	11	42.3%
BI+BV	10	38.4%
BI+AOA+AAD	9	34.6%
OO+AAD	3	11.5%
OH+AAD	3	11.5%
BI+PLB	2	7.6%

Table 5: Associated subaxial anomalies with Congenital CVJ (n=26)

Associated anomaly	Number	Percentage
Scoliosis	4	15.3%
Hemivertebra	2	7.6%
Multiple vertebral anomalies	1	3.8z%
Limbus vertebra	1	3.8%
Bifid spinous process of C4&C5	1	3.8%
Hypoplastic spinous process of C4 & C5	1	3.8%

Table 6 : Congenital CVJ Anomalies(Correlation between X-ray & CT) - An evaluation

	Sensitivity	Specificity	PPV	NPV	Accuracy
Basilar Invagination	50.00	100.00	100.00	75.00	80.00
Atlanto axial dislocation	50.00	100.00	100.00	78.00	82.00
Atlantooccipital dislocation	31.20	100.00	100.00	75.60	78.00
Blocked Vertebra	63.60	100.00	100.00	90.70	92.00
Platybasia	100.00	100.00	100.00	100.00	100.00
OsOdontoideum	50.00	100.00	100.00	95.80	96.00
Ossiculum Terminale	50.00	100.00	100.00	98.00	98.00
Odontoid hypoplasia	0.00	100.00	0.00	94.00	94.00
Hypoplastic Clivus	0.00	100.00	0.00	98.00	98.00
Rachischisis	0.00	100.00	0.00	96.00	96.00
Hypoplastic atlas	0.00	100.00	0.00	96.00	96.00
Other associated subaxial anomalies	0.00	100.00	0.00	86.00	86.00

IMAGES OF CONGENITAL CVJ ABNORMALITIES

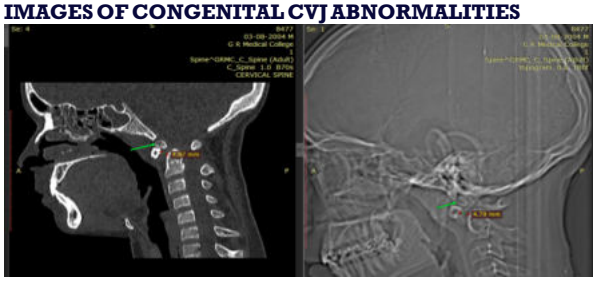

Figure1- OSODONTOIDEUM - Sagittal CT image showing rounded bony fragment lying above and anterior to the base of dens. Dens is hypoplastic, smooth and well corticated(arrow) and anterior arch is hypertrophied and rounded differentiating the condition from fracture.

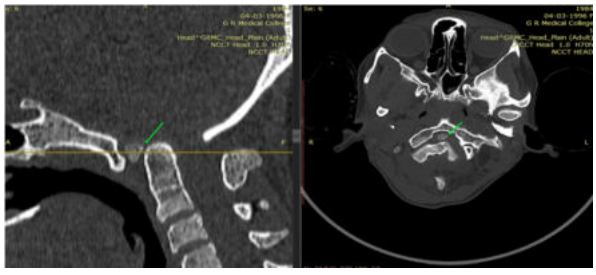

Figure2- PERSISTENT OS TERMINALE: Sagittal & axial CT image showing The terminal ossicle is seen separate from dens due to failure of fusion. It may be confused with a type 1 odontoid fracture. The odontoid process is usually normal in height.

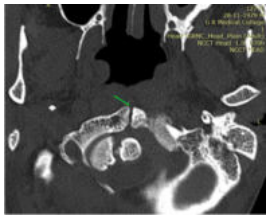

Figure3- ANTERIOR ARCH RACHISCHISIS : Axial CT scan clearly demonstrates the anterior arch rachischisis (arrow)

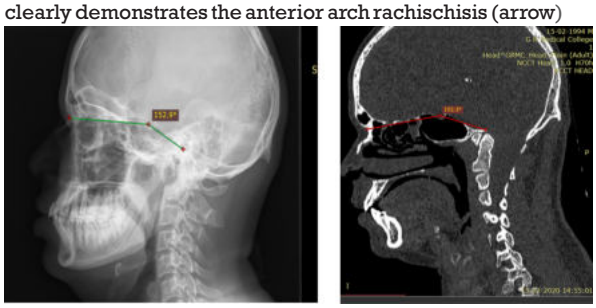

Figure 4-PLATYBASIA - Cervical spine X-ray & CT scan sagittal view show increased basal angle.

DISCUSSION

A prospective study carried out at the Department of Radiology GR Medical College JAH Hospital Gwalior. In our study nonconsecutive patients with all age groups with clinical suspicion of congenital craniovertebral junction anomalies referred from the neurosurgery department for diagnosis and evaluation would be subjected to Computed tomography & x-ray of head & neck from February 2019 to July 2020 were studied. On completion of the study, analysis of the obtained radiological data was done. They were divided into six groups according to age in a decade. Side and type of anomaly, clinical presentation, other associated malformations were recorded.

AGE & SEXWISE DISTRIBUTION OF THE PATIENTS

In our study of 26 patients, male to female ratio was 3.3:1 (20males and 6 females) (Table no-2) and the most common age group was 11-30 years which correlated well with the study by Jawalkar et al and Sankhe and Kumar[10,16].

Table7 : Sex Distribution of CVA- comparative study

Name of Study	Shukla et al	Erbengi et al	Present Study
No of patients	28	56	26
Male	23	40	20
Female	05	16	06
M:F Ratio	4.6:1	2.5:1	3.3:1

EVALUATION OF CONGENITAL ANOMALIES OF CV JUNCTION.

A retrospective cross-sectional study was conducted at NKP Salve Institute of Medical Sciences and Research Center and Lata Mangeshkar Hospital, Nagpur[17]. A total of 26 patients with bony congenital malformations age eight years and above were included in this study over a period of eight years. The most common congenital anomaly observed in this study was BI in15 cases. Other studies reported the commonest anomaly as BI (48%) by Mwang'ombe and Kirongo at Kenyatta national hospital, Nairobi, East Afr[18].

The most common congenital anomaly observed in the present study was Basilar invagination in 19 cases. Other studies reported the commonest anomaly as BI (73%). BI is due to basioccipital dysgenesis in which the vertebral column remains high and is seen above the margins of the foramen magnum.

As the basilar part of occipital bone and margins of the foramen magnum are less developed, the odontoid process and arch of atlas invaginate resulting in basilar invagination. In 13 cases (50%) BI was associated with atlantooccipital assimilation. Another study also reported a similar combination in 14.5% of cases.

The second most common anomaly observed in the present study was atlantoaxial dislocation in 17cases (65%). The third most common anomaly observed in the present study was atlantooccipital assimilation in 16 cases (61%) which coincided with the study conducted by N.J.M. Mwang'ombe[18].

The most common type of lesions in the study conducted by N.J.M. Mwang'ombe, were basilar invagination (48%) followed by atlantoaxial dislocation (28%) and occipitalization of the atlas (28%).

Other atlas anomalies included two cases of defect in the arch of atlas, one case of the hypoplastic posterior arch, absence of lateral mass in one case in our study.

PLATYBASIA

We observed platybasia in two cases which is the same as a study done by Deepali & Chetna at NKP Salve Institute of Medical Sciences and Research Centre and Lata Mangeshkar Hospital, Nagpur[17]. Hypoplastic clivus was seen in one case in the present study.

OS ODONTOIDEUM

In our study, os odontoideum was found in four patients with a male to female ratio of 3:1 (3males:1 female) which correlated with a study done by Dai et al. and Spiering and Braakman [20,21] where male to female ratio was almost 3:1. This odontoid process may remain separate from the body of the axis partially or completely.

OS TERMINALE

In two cases Os terminale was seen. The ossiculum is the separated apical portion of dens, which is derived from the proatlas centrum. The detachment is due to the failure of upper dental synchondrosis.

Table8: Comparison of Findings in developmental anomalies on CT

Findings	Erbengi & Oge[8]	A.H. Menezes[9]	Present Study
AAD	20%	44%	65%
BI	65.2%	94%	73%
AOA	56%	92%	61%

CONCLUSION

- Congenital CVJ anomalies were seen in 26 cases either singly or in combination. The most common anomaly was basilar invagination (BI) seen in 73% of cases. The most common combination of developmental anomalies found was 50 % for BI + AOA followed by 42.3% for BI+ AAD & AAD +AOA BI was seen in combination with Atlanto-occipital assimilation, atlantoaxial assimilation, platybasia & blocked vertebrae. In six cases additional anomalies of other vertebrae were present.
- Plain radiographs form the initial modality of investigation in evaluating a case of Craniovertebral junction anomaly. Computed tomography are invaluable adjuncts to the plain radiographs in the evaluation of the craniovertebral junction anomalies. 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.

REFERENCES

1. Gray's anatomy 37th Ed Philadelphia, Pa: Lea & Febiger, 1989.
2. Smoker WRK Craniovertebral junction: normal anatomy, craniometry, and congenital anomalies. Radiographic 1994; 14:2555-7.
3. Von Torklus D, Gehle W. The upper cervical spine: Regional Anatomy, Pathology and traumatology. A systematic Radiology atlas and textbook, New York: Grune and Stratton; 1972. P1-98.
4. Kwong Y, Rao N, Latief K. Craniometric measurements in the assessment of craniovertebral settling: Are they still relevant in the age of cross-sectional imaging? AJR Am J Roentgenol 2011;196:W421-5.
5. Cronin CG, Lohan DG, Mhuircheartigh JN, Meehan CP, Murphy J, Roche C. CT evaluation of chamberlain's, McGregor's, and McRae's skull-base lines. Clin Radiol 2009;64:64-9.
6. X-ray and Computed Tomography Scan-Based Morphometric Analysis of Skull Baselines in Indian Population by Nandan Amrit Marathe, Aditya A. Dahapute, Jigar Rajesh Desai, Kiran P. Dhole1 , Shubhanshu Bhaladhare1 ,

Swapneel Shah
7. Batista UC, Joaquim AF, Fernandes YB, Mathias RN, Ghizoni E, Tedeschi H. Computed tomography evaluation of the normal craniocervical junction craniometry in 100 asymptomatic patients. Neurosurg Focus 2015;38:E5.
8. Erbengi A, Oge HK. Congenital malformations of the Craniovertebral junction: classification and surgical treatment. Acta Neurochir (Wien). 1994; 127(3-4):180- 5.
9. Menezes AH, Ryken TC. Craniovertebral abnormalities in Down's syndrome. Pediatr Neurosurg. 1992; 18(1):24-33.
10. Sankhe SS, Kumar SK. Neuroimaging in craniovertebral anomalies as seen in the tropics. Neuroimaging Clin N Am 2011;21:879-95.
11. Kale SS, Ailawadhi P, Yerramneni VK, Chandra PS, Kumar R, Sharma BS, et al. Pediatric bony craniovertebral junction abnormalities: Institutional experience of 10 years. J Pediatr Neurosci 2011;6:S91-5.
12. Kumar R, Kalra KS. Management concerns of pediatric congenital atlantoaxial dislocation in the developing milieu. Pan Arab J Neurosurg 2007;11:28-37.
13. Bharucha EP, Dastur HM. Craniovertebral anomalies (A report of 40 cases). Brain 1964;87:469-80.
14. Bhagwati SN. Congenital atlantoaxial dislocation. A study of 99 cases. Neurol India 1990;38:317-24.
15. Chopra JS, Sawhney IM, Kak VK, Khosla VK. Craniovertebral anomalies: A study of 82 cases. Br J Neurosurg 1988;2:455-64.
16. Jawalkar S, Chopra JS, Kak VK, Rao JP, Gulati DR. Craniovertebral anomalies in north-western India. Neurol India 1983;31:15-26.
17. Evaluation of congenital anomalies of craniovertebral junction by computed tomography and its embryological basis Deepali Onkar1*, Chetana Ratnaparkhi2 , Prashant Onkar3
18. Mwang'ombe NJM, Kirongo GK. Craniovertebral junction anomalies seen at kenyatta national hospital, Nairobi. East Afr Med J 2000;77(3):162-4.
19. Goel A. Basilar invagination, Chiari malformation, syringomyelia: A review. Neurol India 2009;57(3):235-46.
20. Dai L, Yuan W, Ni B, Jia L. Os odontoideum: Etiology, diagnosis, and management. Surg Neurol 2000;53:106-8.
21. Spierings EL, Braakman R. The management of os odontoideum. Analysis of 37 cases. J Bone Joint Surg Br 1982;64:422-8.