

A Comprehensive Study of Craniovertebral Junction Anomalies

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Abstract

Introduction: CVJ anomalies are very common in Neurosurgical practice. The treatment modality & surgical technique, still remains a challenge.

Objectives: To study the various congenital & Acquired CVJ anomalies, their clinical presentation, management and Post surgical functional outcome.

Materials and Methods: About 50 patients of CVJ anomalies who were presented at the “Institute of Neurosurgery, Madras Medical College, Chennai-3 were analysed by their demographic, clinical & radiological parameters, and various surgical procedures.

Results: About 34 patients had congenital anomalies & 16 patients had acquired anomalies.. AAD is the commonest abnormality noted. ADI determines the postsurgical outcome. C1 C2 Stabilisation carries good results.

Conclusions: Bony anomalies are more than soft tissue anomalies. Craniometric assessments are very arbitrary. Congenital anomalies are very common than that of acquired ones.

Keywords: Craniovertebral Junction Anomalies; Atlanto Dental Interval; Atlanto Axial Dislocation.

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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 subject of CVJ is under discussion and evaluation over a century and numbers of classical reviews have attempted to clarify a variety of complex associated issues.

The incidence of different types of CVJ Anomalies varies with demographic environment & ill-defined

genetic factors. CVJ Anomalies are more frequently found in Indian subcontinent than anywhere else in the world. Even in India, these anomalies are more frequently documented from Bihar, Uttar Pradesh, and Rajasthan & Gujarat. The reason for this geographical clustering is more speculative. The CVJ Anomalies can be either due to Bony or Soft tissue Anomalies. They are common in all age groups and almost equal in both sex groups. The anomalies can be due to congenital and acquired causes. There has been a renewed interest in the normal anatomy & pathological lesions of CVJ Anomalies with Dynamic X rays, CT & MRI. The clinical features are often delayed upto 2nd or 3rd decades, since they are subtle and often missed. The surgical management of CVJ Anomalies is complex due to the relative difficulty in accessing the region, critical Neurovascular structures and the intricate Biomechanical issues involved. In spite of various surgical procedures, the commonly done procedures are Foramen Magnum Decompression,

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C1 C2 wiring, C1 lateral mass & C2 pedicle screw fixation and Trans Oral Odontoidectomy. The surgical indications for each approach are still under discussion.

The management protocol varies with each patient and there are several studies conducted on this issue, trying to arrive at a consensus.

Hence an attempt was made to analyse comprehensively on the incidence of CVJ anomalies among various age groups, its various clinical features, different radiological investigations, various etiological diagnosis, different surgical procedures and its final surgical outcome.

Objectives

The Aims & Objectives of this study are as follows;

1. To study the incidence of various Bony & Soft tissue CVJ Anomalies.
2. To study the different clinical features of various CVJ Anomalies.
3. To analyse the various etiological factors contributing to CVJ Anomalies.
4. To study the different radiological Investigations & their correlation with the final surgical outcome.
5. To analyse the various surgical procedures done for CVJ Anomalies & its final surgical outcome.

Materials and Methods

About 50 patients with Craniovertebral Junction Anomalies who were treated in the Institute of Neurology, Rajiv Gandhi Government General Hospital, Chennai during the period of May 2014 to May 2016 have been studied prospectively.

Inclusion criteria for this study are as follows:

1. Bony CVJ Anomalies.
2. Soft tissue Anomalies.
3. All Age groups.

Exclusion criteria for this study are as follows:

1. Incidental Arnold Chiari Malformation.
2. Patients with Irreducible CV Junction Anomalies.
3. Patients who are not willing for surgery.
4. Patients who have lost follow up.

This is a prospective analytical study and all the patients in the study were entered into a proforma.

All the clinical, radiological, surgical treatment & outcome details were entered into the proforma.

Criteria for Detecting the Incidence

All the patients' personal details like name, age, sex were entered into the proforma.

Clinical Criteria for the Analysis of Symptomatology

The patient's symptoms of sensorimotor disturbances, cranial nerve disturbances, features of increased intracranial hypertension, autonomic disturbances, symptoms of vertebrobasilar insufficiency (vertigo, syncope) were analysed.

Clinical Criteria for the Diagnosis

After complete clinical examination, the sensory level, the motor level and the reflex level was found and the diagnosis of CVJ anomalies were attained. The other associated clinical features like Head tilt, short neck, webbed neck, Downbeat Nystagmus, Cranial nerve deficits, features of congenital syndromes like Down syndrome, Morquio syndrome, Klippel - Feil syndrome were taken into account.

Radological Criteria for the Diagnosis

X ray cervical spine:

- a. Occipitalisation of Atlas due to the assimilation of Atlas to the Occiput.
- b. Defect in the arches of Atlas.
- c. Atlanto Dental Interval (ADI): It is the interval between posterior surface of Anterior Arch and the anterior surface of Atlas. In adults, ADI >3mm & in children, ADI > 4mm was taken as positive for Atlanto Axial Subluxation.
- d. Block Vertebrae - Congenital fusion of vertebral bodies.

Dynamic Xrays CVJ:

Both Flexion & Extension Lateral views of CVJ were viewed.

The increased ADI on Flexion view implies irreducible Atlanto Axial Subluxation.

Craniometric Assessment:

Various anatomical lines were studied from X Ray CVJ, CT scan CVJ and MRI CVJ.

1. *Mc Rae's Line:* It is the foramen magnum line connecting the Basion and Opisthion.
2. *Mc Gregor Line:* It is the line connecting the posterior surface of palate to the Occiput
3. *Chamberlain Line:* It is the line connecting the posterior surface of palate to the inferior surface of the Occiput.
4. *Wackenheim's Clivus Canal Line:* It is the tangential line drawn along the clivus & by extrapolating it downwards. Normally the Odontoid should lie below or should not exceed 2.5mm above this line.
5. *Welcher's Basal Angle:* It is the Angle between the nasion tuberculum line & tuberculum Basion line. The angle above 130 degrees was taken as positive for Basilar Invagination.
6. *Powers Ratio :* It is the ratio between the lines connecting Basion to anterior edge of posterior arch of Atlas and the line connecting the Opisthion to the posterior surface of Anterior arch of Atlas. Normal: 0.77. Abnormal values (>0.77) implies atlanto occipital Dislocation.

CT scans CV Junction:

Various Bony anomalies like defects in the arches of Atlas, Assimilation of Atlas, Block Vertebrae, Basilar Invagination, Atlanto Occipital Dislocation, Odontoid fracture were studied.

MRI CV Junction:

Apart from Craniometric assessment, Atlanto axial subluxation, tonsillar herniation, cerebellar descent, syringomyelia, intrinsic cord changes, prevertebral soft tissue collections were studied.

Criteria for Etiological Diagnosis

Based on the history & thorough clinical examination, the CVJ Anomalies were broadly categorized into Congenital and Acquired.

Among the congenital Bony CVJ Anomalies, congenital Atlanto Axial subluxation, Basilar Invagination, Platybasia, Occipitalisation of Atlas, Defect in arches of Atlas were studied.

Among the Congenital Soft tissue CVJ Anomalies, Arnold Chiari malformations, Syringomyelia were studied. The associated features of congenital

syndromes like Down syndrome, Morquio syndrome & Klippel Feil Syndrome were studied.

The Acquired CVJ Anomalies were studied under traumatic causes, Infective causes – Grisel Syndrome following Pharyngeal Infection, Inflammatory causes – following Tuberculosis, Rheumatoid arthritis, Neoplastic causes – following Foramen Magnum meningiomas, Neurofibromas.

Criteria for Studying the Surgical Treatment

The patients who underwent the following surgeries were studied.

1. Foramen magnum Decompression.
2. C1 C2 Wiring using Stainless Steel or Titanium wires.
3. C1 Lateral mass & C2 pedicle screw fixation.
4. Transoral Odontoidectomy
5. Occipito Cervical Fusion.

Criteria for Studying the Outcome;

The patients were followed up at 1 month, 3 months, and 6 months. Thorough clinical examination was made and the outcome was measured based upon the improvement/deterioration/static neurological status by assessing the motor power by MRC Grade. Post operative Imaging was done to assess the optimal reduction and restoration of Craniovertebral junction.

The Atlantodental Interval (ADI 3-5mm, >5mm) was correlated with the final outcome of the patients and the statistical significance was found. The Craniometric lines were correlated with the final outcome and the statistical significance was found out.

The statistical significance was found in the patients, who underwent different surgical procedures by correlating with the outcome.

The Statistical Analysis was done by Chi-Square test by using Graph Pad Stat Software.

Observation and Results

About 50 cases of Craniovertebral junction Anomalies treated during the period of May 2014 to May 2016 were studied.

Age Distribution

About 14 patients were below 10 years, 13 patients were seen from 11 to 20 years, 9 patients

were seen from 21 to 30 years, 10 patients from 31 to 40 years, 3 patients from 41 to 50 years and 1 patient from 51 to 60 years. (Fig. 1).

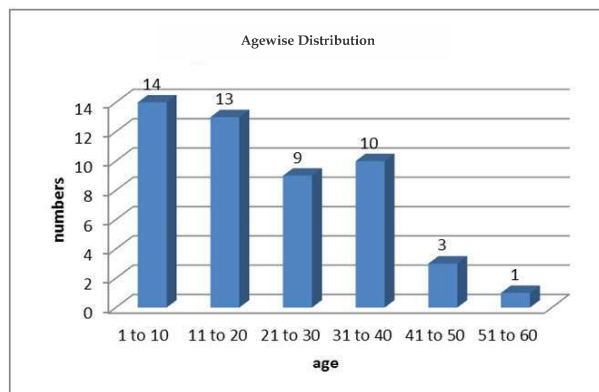


Fig. 1:

Sex Distribution

Out of 50 patients, about 27 patients were males and 23 patients were females.

Distribution of Age According to Gender

Out of 14 patients under 10 years, 8 were males & 6 were females. Under 11 to 20 years of age, about 8 patients were males & 5 patients were females, out of 13 patients. About 5 patients were males & 4 patients were under 21 to 30 years category, out of 9 patients.

Out of 10 patients under 31 to 40 years category, about 6 were males & 4 were females. All the three affected patients were female's under 41 to 50 years category and the only affected patient is a male under 51 to 60 years category. (Fig. 2).

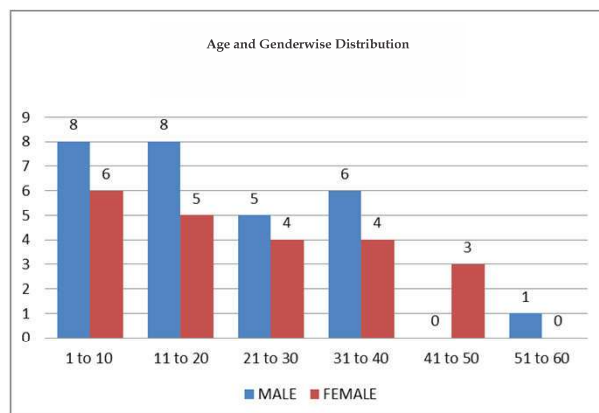


Fig. 2:

Distribution of Symptoms

About 48 patients had presented with motor weakness in the form of ascending type of spastic

quadriparesis, about 44 patients presented with sensory disturbances involving the anterolateral spinothalamic tract & Posterior column, 43 patients presented with neck pain (22 had localised Bony pain & 9 had Suboccipital Headache), about 8 patients had recent significant Head & Neck Trauma, about 38 patients presented with Autonomic disturbances (Isolated Bladder involvement in 22 patients, combined Bladder & Bowel in 38 and patients, erectile dysfunction in 4 patients) and about 6 patients presented with features of Vertebrobasilar insufficiency in the form of vertigo.

Distribution of Clinical Signs

Out of 50 patients, about 16 had Head tilt, 18 had short Neck, 11 had Low hairline, 7 had Webbed neck and increased Height Neck ratio in about 29 patients.

Skeletal deformities were found in 4 patients (3 had kyphosis, 1 had scoliosis), Spastic Quadriparesis in 48 patients, sensory disturbances in 44 patients, Downbeat Nystagmus in 5 patients, Facial Sensory loss in about 4 patients, Lower Cranial nerve deficit in about 6 patients, and Autonomic disturbances in 38 patients. (Fig. 3.)

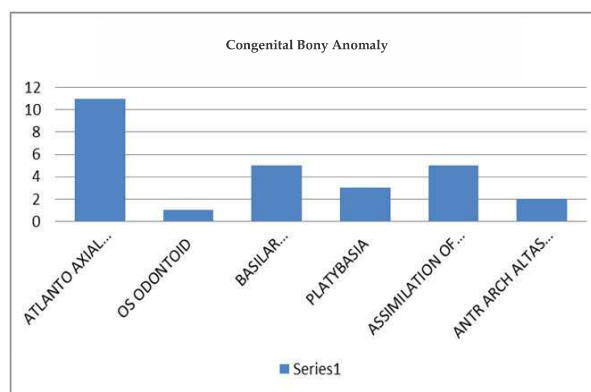


Fig. 3:

Distribution of Clinical Signs

On examining the spinomotor system of these patients, the motor power was examined under MRC Grade. About 4 patients had Quadriplegia, one patient presented with Grade 1 power, about 24 patients had presented with Grade 2 power, about 15 patients had presented with Grade 3 power and 6 patients had presented with Grade 4 power.

Distribution of Etiological Diagnosis

Out of 50 patients, 34 patients had congenital Anomalies and 16 patients had Acquired Anomalies.

Among the 34 congenital Anomalies, 22 had Bony Anomalies & 12 had soft tissue Anomalies. Among the 22 patients, 11 had Atlantoaxial subluxation, 5 had Basilar Invagination, 3 had Platybasia, 5 had Assimilation of Atlas, 2 had defect in Anterior arch of Atlas, 1 had Os Odontoid. One patient had features of Down syndrome with Atlantoaxial subluxation.

Out of 12 patients with soft tissue CVJ Anomalies, 12 had Arnold Chiari malformation.

Among the Acquired CVJ Anomalies (16 patients), about 8 had developed the Anomaly following trauma, 2 had developed following Pharyngeal infection (Grisel Syndrome), 3 had developed following inflammatory causes (Tuberculosis in 2 patients & Rheumatoid Arthritis in 1 patient), 3 had developed this Anomaly following tumors (2 had Foramen magnum meningioma & 1 had Neurofibroma).

Tuberculosis CVJ Anomaly was instituted with Anti Tuberculous Therapy. (Fig. 4 and 5.)

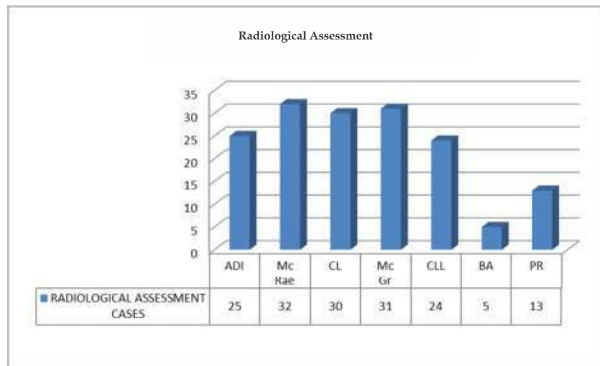


Fig. 4:

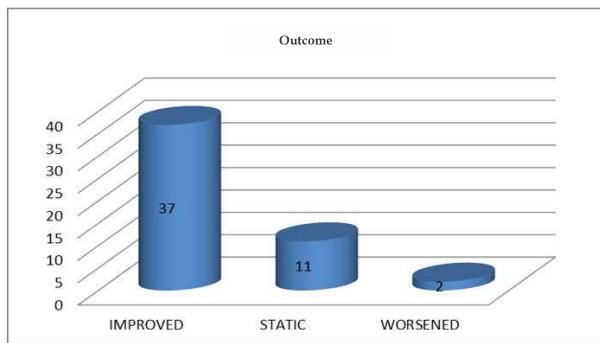


Fig. 5:

Distribution of Radiological Abnormalities

The Atlanto Dental Interval (ADI 3-5 mm in 13 pts, >5mm in 12 pts.) was increased in about 25 patients.

Abnormal Mc Rae’s line was found in about

32 patients, Chamberlain line in 30 patients, Mc Gregor’s line in 31 patients, Wackenheimer Clivus Canal Line in 24 patients.

Abnormal Welcher’s Basal Angle >130 degrees was found in about 5 patients and the increased Power’s ratio > 0.77 mm was seen in about 13 patients. (Fig. 6)

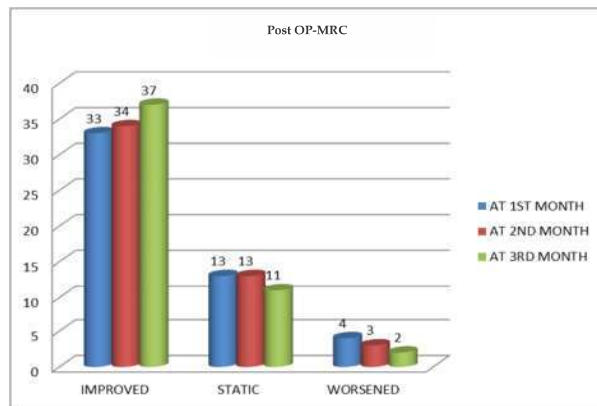


Fig. 6:

Distribution of Various Surgical Treatments Done:

Out of 50 patients, about 14 patients with Atlantoaxial subluxation had undergone C1 lateral mass & C2 pedicle screw fixation. About 14 patients underwent Foramen magnum decompression for 12 patients with Arnold Chiari malformation & 2 patients with Foramen magnum tumors.

About 6 patients underwent C1 C2 wiring for Atlantoaxial Dislocation.

About 7 patients underwent Transoral Odontoidectomy (for 2 patients with Grisel syndrome, 1 with Os Odontoid, 4 patients with Basilar Invagination).

About 9 patients underwent Occipitocervical Fusion using contoured steel rod for 5 patients with Basilar Invagination & 3 patients with Platybasia, 1 with AAD.

Distribution of Surgical Outcome

The Neurological outcome was assessed in these 50 patients by thorough clinical examination of motor power under MRC Grade.

About 33 patients got improved, 13 patients had residual deficit as that of pre operative status and 4 patients got deteriorated after surgery, at 1 month follow up.

About 34 patients had improved and 13 remained static, 3 patients got worsened, at the end of 3 months follow up.

About 37 patients had improved at 6 months and 11 patients remained static, 2 patients got deteriorated at 6 months follow up.

All the patients had optimal reduction in the post operative imaging.

Among the 13 patients with ADI 3-5mm, about 10 patients and among the 12 patients

with ADI > 5mm, about 6 patients have showed significant postoperative improvement. On

Statistical analysis by Chi-square test, the P value is 0.001 (p value<0.05) and hence it is statistically significant. (Fig. 7).

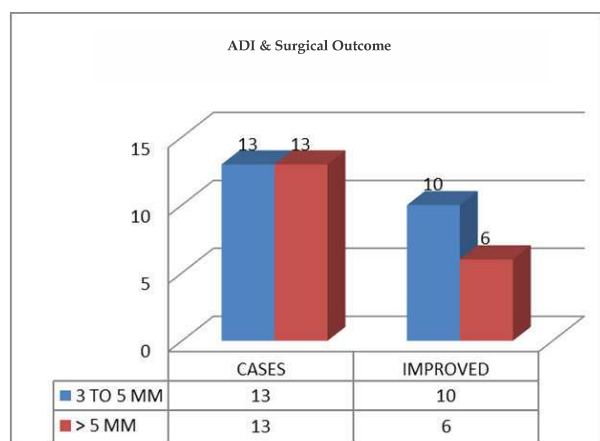


Fig. 7:

Among the 32 patients with abnormal McRae’s line, 9 patients have improved, 8 patients have improved out of 30 patients with abnormal Chamberlain’s line, 8 patients have improved among the 31 patients with abnormal McGregor’s line and 7 patients have shown improvement among the 24 patients with Wackenheim’s Clivus Canal line, post operatively. (Fig. 8).

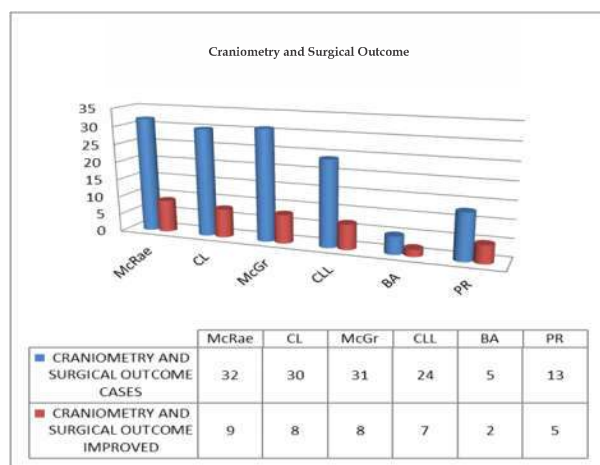


Fig. 8:

Statistics

Craniometry & Outcome

Chi-squared Test for Independence

Chi-square: 1.106

Degrees of Freedom: 5

Table size: 6 rows, 2 columns.

The p value is 0.9536.

The row and column variables are not significantly associated.

Chi-square calculations are only valid when all expected

Values are greater than 1.0 and at least 80% of the expected

Values are greater than 5.

Chi-Squared Test for Trend.

Chi-squared for trend = 0.5674 (1 degree of freedom)

The p value is 0.4513.

There is not a significant linear trend among the ordered categories

defining the rows and the proportion of subjects in the left column.

Summary of Data

Table 1:

Row	Total	Percent
1	32	23.70%
2	30	22.22%
3	31	22.96%
4	24	17.78%
5	5	3.70%
6	13	9.63%
Total	135	100.00%

Table 2:

Column	Total	Percent
Not Imp	96	71.11%
Improved	39	28.89%
Total	135	100.00%

On statistical Analysis of the Craniometric assessment and the surgical outcome, the p value is 0.4513 (p value > 0.05) and hence it is not statistically significant.

The outcome was assessed in each surgical procedure. About 12 patients have showed significant post operative improvement in 14 patients who have undergone C1 Lateral mass & C2 pedicle screw fixation. About 11 patients

have improved among the 14 patients, who have undergone Foramen Magnum Decompression.

About 4 patients have improved among the 6 patients who have undergone C1C2 Wiring. About 3 patients have shown significant post operative outcome in the 7 patients, who have undergone TransOral Odontoidectomy and about 7 patients have improved in the 9 patients, who have undergone OccipitoCervical Fusion. (Fig. 9)

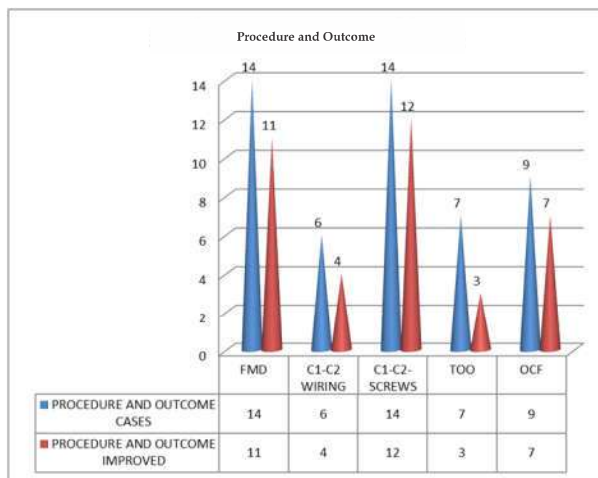


Fig. 9:

Statistics

Surgical Procedures & the Outcome

Chi-squared Test for Independence

Chi-square: 4.914

Degrees of Freedom: 4

Table size: 5 rows, 2 columns.

The p value is 0.2963.

The row and column variables are not significantly associated.

Chi-Squared Test for Trend.

Chi-squared for trend = 0.2753 (1 degree of freedom)

The p value is 0.5998.

There is not a significant linear trend among the ordered categories

defining the rows and the proportion of subjects in the left column.

Summary of Data

Table 3:

Row	Total	Percent
1	14	28.00%
2	6	12.00%
3	14	28.00%
4	7	14.00%
5	9	18.00%
Total	50	100.00%

Column	Total	Percent
Not Imp	13	26.00%
Improved	37	74.00%
Total	50	100.00%

On Statistical Analysis of the different surgical procedures and the outcome, the p value is 0.5998 (p value > 0.05) and hence it is not statistically significant.

Discussion

The CV Junction Anomalies are more common in young adults (28%). The incidence is almost equal in both sexes, with slight male (54%) predominance.

The majority of CVJ Anomalies are of congenital variety (68%). Among the congenital Anomalies, the Bony anomalies (44%) are common than the soft tissue Anomalies (24%). The Alanto axial Dislocation (22%) is the most common congenital Bony Anomaly and the Arnold Chiari malformation (24%) is the most common soft tissue Anomaly. Among the Acquired CV Junction Bony Anomalies, trauma (16%) is the most common etiological factor.

These results are well comparable to the studies done worldwide, as described in the literature.

Inspite of the varied clinical presentations, motor weakness (96%), followed by sensory disturbances (80%) are the most common clinical features. Pre operative MRC Grading of motor power implies the post operative prognostic outcome.

Dynamic X ray cervical spine, Craniometric Assessment in X ray cervical spine, CT scan & MRI cervical spine are the important tool to diagnose and to tailor the surgical management. About 50% of patients had increased Atlantodental Interval. The ADI in the preoperative imaging is well correlated with the final surgical outcome and showed statistical significance.

The Craniometric assessment showed abnormal Mc Rae’s line in about 64%, Chamberlain line in 60%, McGregor’s line in 62%, Wackenheimer Clivus Canal Line in 48% of patients.

The increased Basal Angle was seen in about 10% and about 26% showed abnormal Power's Ratio.

Even though, the different surgeries are indicated, C1 Lateral Mass & C2 pedicle screw fixation (28%) is commonly done for Bony Anomalies and Foramen Magnum Decompression (24%) is commonly done for Soft tissue Anomalies.

About 77% of patients with abnormal ADI 3-5mm, showed significant post operative Outcome and 50% of patients with abnormal ADI >5mm have improved postoperatively. The ADI is statistically significant and well correlated with the post operative outcome. It is the most important and reliable pre operative marker in predicting the prognostic outcome.

About 28% of patients had improved postoperatively with abnormal McRae's lines, 26% of patients had improved with abnormal Chamberlain lines, 25% of patients have improved with abnormal McGregor lines.

About 29% of patients had improved with abnormal Wackenheim Clivus canal line. About 40% of patients had improved with increased Basal Angle and about 39% of patients had improved with increased Power's ratio.

The Craniometric lines are not well correlated with the final surgical outcome and it is not statistically significant.

The Craniometric lines are very arbitrary and poorly correlated with the post operative prognostic outcome.

In spite of different surgical procedures, C1 Lateral mass & C2 Pedicle screw fixation is gaining popularity nowadays. About 86% of patients had improved after this surgery and the improvement is very less (43%) in the TransOral Odontoidectomy.

About 79% of patients had shown improvement following Foramen magnum Decompression and about 67% of patients had improved following C1C2 Wiring.

About 78% of patients had improved following Occipitocervical Fusion using contoured rod. In spite of different surgical techniques described, no particular technique is better correlated with the post operative outcome and it is statistically insignificant. Hence, the each surgical procedure may be indicated depending on the case. This is comparable to the studies done worldwide, where there are no definite indications for each surgical technique and no consensus is described in the literature.

Many patients have improved neurologically

(54%), after surgery at the end of 6 months follow up.

Since the sample size is very small, the study with large sample size is needed to obtain still better results.

Conclusion

- 1) Among all the Craniovertebral Junction Anomalies, Bony Anomalies are more common than soft tissue Anomalies. It is more common in young adults. It is almost equal in both the sexes with slight male predominance.
- 2) Congenital CVJ Anomalies are more frequent than Acquired Anomalies.
- 3) Atlanto Dental Interval is the single important & reliable marker to assess the prognostic outcome.
- 4) Craniometric assessment is very arbitrary and not correlated with the final surgical outcome.
- 5) In spite of different surgical procedures, each surgical technique is tailored according to the patient. Early surgery carries good prognostic outcome especially in patients with better motor power.

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