Cranio-vertebral junction anomalies with Atlanto-axial instability: Management and complications- Our experience of 63 cases at a tertiary care center in India

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ABSTRACT

IntroductionManagement of complex cranio-vertebral junction anomalies with atlanto-axial instability require extensive pre-operative work up with various intra-operative (and/or preoperative)manouvres and bony fusion procedures and post-operative rehabilitation and support. This study focuses on various management strategies in terms of its outcome and complications. **Materials and methodology:**This clinical analytical study shares our experience of 63 cases of cv junction anomalies with atlanto-axial instability operated at tertiary care center in india. 63 patients operated during the period spanning from June-2019 to November-2021 who had atlanto axial instability (AAD WITH/WITHOUT BI). Patients were assessed pre-operatively and post-operatively up to 3 months objectively and subjectively in terms of outcome and post op radiology was also done. **Results:**Out of 63 atlanto-axial operated patients, 42 improved, 3 had stable disease and 12 deteriorated clinically some of whom required also secondary procedures for it. Out of 63 ,8 patients had died,4 of them were pediatric and 4 were adults. Various procedure related complications occurred in 12 patients. **Conclusion:** Thus, with proper pre-operative diagnosis of type of congenital anomaly combined with appropriate surgical plan can give impacting results in these pathologies with minimal complications.

Key words: cranio-vertebral junction anomalies, atlanto-axial instability, DCER, C1-C2 fusion

INTRODUCTION

Cranio-vertebral junction is anatomically very complex region and protects the critical structure of cervico-medullary junction and is a seat of many pathologies. This clinical analytical stdy focuses on management in terms of outcome and complications of cranio-vertebral junction anomalies which are mostly congenital/developmental in nature and have inherent atlanto-axial instability and shares the experience of 63 cases operated at a high-volume tertiary care center in India.

AIM & OBJECTIVE

To audit various operative techniques for cranio-vertebral junction anomalies with atlanto-axial instability for their outcome and complications in terms of morbidity and mortality.

METHODOLOGY

Type: clinical analytical study- case series

Patients operated at a tertiary care center at Ahmedabad, Gujarat, India from June 2019 to November 2021 with at least 3 months post op follow up.

Inclusion criteria:

1. Age above 2 years
2. Progressive neurological deficit attributable to atlanto-axial instability due to cranio-vertebral junctionanomalies which may be congenital or developmental in origin
3. Radiological imaging suggestive of atlanto-axial instability with compressive myelopathy with minimal neurological impairment.

Exclusion criteria:

1. Age below 2 years with cranio-vertebral junction developmental anomalies
2. Progressive neurological deficit not attributable to cranio-vertebral junction anomalies with radiology suggestive of presence of congenital/developmental cranio-vertebral junction anomalies without compressive myelopathy.
3. Cranio-vertebral anomalies with compressive myelopathy and/or progressive deficit secondary to some etiology i.e.,trauma, infective cause or auto-immune/systemic diseases.
4. cranio-vertebral anomalies without any definitive radiological/objective evidence of atlanto-axial instability i.e., isolated chiari-1 malformations

Atlanto-axial instability/ Atlanto-axial dislocation (AAD) is defined as atlanto-dental interval more than 3 mm in adults and in pediatric patients of atlanto-dental interval up to 5 mm up to 8 years.
It may be associated with basilar invagination in many cases.
All patients were assessed clinically as well as radiologically by doing MRI cranio-vertebral junction with whole spine screening with/without brain screening and CT- cranio-vertebral junction dynamic (flexion,extension,neutral)study pre-operatively and post operatively at 3 months.in addition to this,CT angiography of neck was done to check for vertebral artery anomaly.
Results were analyzed prospectively in terms of various complications and long-term morbidity and mortality rate.

Management strategies used:

For treatment of these cases, there are three components to consider.

1.Reduction:

a. Skeletal traction[1]: may be given pre-operatively or intra-operative and force of traction according to age and weight of the patient with maximum up to 12-15 pounds in adults.

b. DCER (Distraction, Compression, Extension, Reduction)[2]:

DCER stands for distraction, compression, extension, and reduction. This is a surgical technique to reduce, realign and correct (even very severe) basilar invagination (BI), atlanto-axial dislocation (AAD) with a posterior only, single staged approach. This involves motion in 2-axis using the lever principle. This is a technique which was pioneered by PS Chandra et al[2].
DCER is currently indicated for AAD with BI in developmental anomalies with atlanto occipital assimilation. It involves intraoperative reduction of anomalies to anatomical position with occipito-cervical fusion with fixation and/or spacer insertion in cases of severe basilar invagination to maintain reduced position.

2. Decompression[1]:

according to site of compression over cervico-medullary junction, it may be ventral or posterior decompression.

a. posterior decompression:

it is done by foramen magnum bony decompression and indicated in cases of dorsal compression over cervico-medullary junction as a stand-alone procedure or combined with posterior cervical fusion procedures.

b. ventral decompression[1]:

when there is ventral compression at cervicomedullary junction by odontoid process or any bony part of cranio-vertebral junction due to congenital anomalies, they are removed by ventral procedure done via transoral transpharyngeal route, transcervical extrapharyngeal or endoscopic routes. We have used transoral transpharyngeal route. These procedures are done as part of two stage procedures combined with posterior cervical fusion procedures.
Indicated in cases of severe ventral compression in operated cases of posterior cervical fusion procedures with persistent ventral compression.

3. Fusion procedures[3].

In Almost all cases of atlanto-axial instability, posterior cervical fusion procedures are carried out to maintain reduced (anatomical) state at cranio-vertebral junction and to prevent future chances of instability. They are generally combination of instrumented fixation with bony fusion procedures.
Various posterior cervical fusion procedures are:

a. Wiring techniques:

atlanto-axial fusion carried out by interwining malleable titanium wires between C1-C2 vertebrae with bone graft placed between them. Various methods are, gallie's fusion, sonntag's modification and brook's and jenkins technique.

b. C1-C2 fusion:

this fusion technique popularized by Goel et al[8]. In this procedure, lateral C1-C2 joints are fused by bone graft with/without metal spacers followed by instrumented fixation of C1 and C2. Indicated in cases of atlas vertebrae with good lateral masses and difficult in cases of patients with atlanto-occipital assimilation.

c. Occipito-cervical fusion:

indicated for cases with atlanto-occipital assimilation or very severe BI cases. Associated with DCER, this procedure provides a good treatment strategy for severe bi or irreducible type of BI cases. In these cases, instrumented fusion extends fro occipital bone to various levels of cervical vertebrae.

RESULTS

There were total 63 patients operated for cranio-vertebral junction anomalies with atlanto-axial instability during the period spanning from June-2019 to November-2021 and results were analyzed prospectively.

1. Clinical Outcome:

Outcome	**No .of patients(%)**
Improved	42(66.66%)
Deteriorated	8(12.69%)
Stabilized	12(19.04%)
Lost to follow up	1(1.58%)

2. Radiological Outcome:

In addition to, subjective and objective clinical outcome, radiological outcome has also been measured in terms of, anterior atlanto-axial(dental)(ADI) distance and basilar invagination above chamberlein's line (palato-occipital line) (BI). For successful reduction and fixation, we have taken atlanto-dental interval should be 3mm or less than that and basilar invagination of 5mm or less than that.

Status	AAD (mean)	BI (mean)
Pre -op	5.079mm	9.65mm
Post op	2.877mm	4.385mm
Successful reduction (no. of patients (%))	51(89.47%)	45 (78.94%)

1 patient died in post op period in whom post op imaging could not be achieved.

3.mortality:

Mortality	No. of patients
Immediate post op/intra-op	1
Post operative	3
Follow up period(upto 3 months)	2
Extended follow up(after 3 months)	2

Out of 63 patients operated, mortality happened in 8 patients which is 13% of all patients.

Out of 8 patients died, 4 were pediatric and 4 were adult patients.

4. complications:

out of 63 patients operated, 12 patients had procedure related complications.

Complication	**NO. of patients**	**No. of pts. Required re-exploration/surgery**
Spacer dislocation	3	3
Loosened occipital/lateral mass screws	2	2
Vertebral artery injury	2	1
Deep seated infection	2	2
Post op CSF LEAK	3	0

5.Type of Surgery:

Type of surgery	**No. of patients**
Occipitocervical fusion with DCER	41
C1-C2 FUSION	17
Primary ventral decompression (trans-oral approach)	2
Secondary ventral decompression (trans-oral approach)	2
Only foramen magnum decompression	1
Syringe-subarachnoid shunt (with FMD)	1

DISCUSSION

Treatment of cv junction has changed from troublesome traction and immobilization in a brace to intra-op reduction and posterior fusion with titanium implants and endoscopic transnasal/transoral decompression at cv junction.

Pioneers among treatment of cv junction anomalies are Menezes et al, klekamp et al, goel et al, PS Chandra et al etc.

Menezes[4,5] in the United States have provided insights regarding factors causing cv junction instabilities and management of various anomalies. His largest contribution is in role of ventral approaches in ventral compressive myelopathy[6].

Klekamp et al[7] from Germany has provided largest case series in Europe with support toward posterior approaches. He insisted on foramen magnum decompression for only BI patients without ventral compression and to do cranio-vertebral realignment and posterior fusion for reducible lesions with moderate ventral compression and reserved ventral decompression for severe ventral compression only.

Goel et al[8] revolutionized the treatment of AAD with BI with their landmark paper of C1-C2 lateral mass screw fixation. Goel et al put stress on C1-C2 joint to be of center of origin of all kind of instability and by fixing this joint in reduced condition would reverse all the changes of compressive myelopathy.

Despite highly successful technique, goels's C1-C2 lateral mass fixation technique has limitations in cases like atlanto-occipital assimilation, anomalous vertebral artery origin,difficulty in achieving

cranio-vertebral realignment in cases of severe AAD with BI and severe ventral compression. All above limitations lead to development of DCER technique by P. Sarat Chandra.

P. Sarat Chandra et al[2] described various types of atlanto-axial joint morphology and angulations and described occipito-cervical fusion procedures by joint modification and joint preparation and putting a metallic spacer or bone graft in joints, thus providing reduction and posterior fusion. This technique is very useful in severe atlanto-axial instability.

With advancement in instrumentation and radiology, there is rise of interest in decompression and fixation through microscopic and endoscopic techniques via anterior approach.

Our experience:

In our case series, out of 63 patietns, clinical improvement was seen in 42(66.66%) patientswhereas 12(12.70%) patients had stabilized disease progress, constituting overall 79.36% successful outcome in terms of halting clinical deterioration, where as in P. Sarat Chandra et al[2] case series, overall improvement rate is 93.2% (138 out of 148) had clinical improvement and in goel at al[8]is approximately 98% (157 out of 160).

Overall complication in our study group (AAD group) is around 13%, which is equivalent to P.S Chandra et al[2] (14%).

The difference in results of our series might be due to small number of cases as compared to other studies.

Spacer, Traction and Bone Graft:

We have applied traction intra-op in 38 patients and pre-op traction in 10 patients. When results are compared, there was more reduction in AAD and BI in intra-op traction group versus pre-op traction group. Moreover, patients with pre op traction either required secondary ventral decompression (5 out of 10) or deteriorated in outcome measurement. This may be because of, these group of patients had severe atlanto-axial instability with severe compressive myelopathy.

We have used metallic spacer in 29 out of 58 patients, out of which 25 wereinserted in DCER patients and 4 in C1-C2 fixation patients. In our experience,we found spacer insertion useful to maintain reduction of BI, especially in DCERgroup of patients.

Out of 58 patients, we have used iliac crest graft in 2 patients and rib graft in only 1 patient. In the rest 55 patients, we acquired autologous bone graft from same incision through partial spino-laminectomy and bony FMD. Thus, by avoiding additional incision, we provide much of pain relief and less morbidity to this patient.

Trans-Oral (Ventral Decompression)

10 patients were operated with trans-oral decompression, 8 of whichfor secondary ventral decompression after posterior fusion for persistent ventral compression, 1patient had clival hypertrophy which immediately died in post op period and 1 patient had clival/pro-atlas segmentation defect whom we have not counted in result analysis, both of them underwent primary trans-oral decompression.

No post op CSF leak or meningitis occurred in these patients. 1 patient had intra-op Dural leak for which tissue adhesive glue was used and post op lumbar drain was inserted.

2 patients had velopharyngeal insufficiency and 1 patient had dysphagia, all of which improved gradually without any intervention or prolonged enteral nutrition.

Out of 7 patients alive after trans-oral decompression, all of them have subjective as well as objective improvement and radiological decompression achieved.

In trans oral group, out of 10, 2 patients died in immediate post operative period and 1 patient died after extended period due to aspiration pneumonitis. In rest of 7 patients, there is overall subjective and objective improvement,as is scenario in case with Menezes et al[6].

Overall, from our study, we concluded that, for patients with atlanto-axial instability, DCER and C1-C2 posterior fusion techniques are equivalent in terms of clinical outcome. One of which is used for isolated C1 vertebrae with good lateral masses(C1-C2 fusion) and other is for atlanto--occipital assimilation with severe AAD (with almost vertical C1-C2 joints)(DCER). Despite efficiency of these procedures, some patients require secondary ventral decompression procedures

which are technically complex and moribund procedures, but if successfully executed, it can give lasting results combined with posterior fusion procedures.

LIMITATIONS

Main limitation of our study is, small number of subjects with heterogenous group of cases, absence of control arm group and short term follow up. For, more tangible conclusions, we need to conduct study with large no. of cases with control groups if possible.

CONCLUSION

Cranio-vertebral junction anomalies are very complex anomalies, which require proper understanding of pathology and pre-operative planning as well as adequate resources and instrumentation and most importantly, expertise in these types of cases. Decision making in this type of cases should be on a case-by-case basis and should not be rigid to any strict criteria. Thus, Cranio-Vertebral junction anomalies should be managed at a high-volume center with expertise in these cases with well-established pre-op planning and post op rehabilitation program and adequate care to improve quality of life for these patients with minimal complications and mortality.

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