Original article

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Comprehensive assessment of surgical, clinical, and radiological outcomes in craniovertebral junction anomalies with basilar invagination and atlantoaxial dislocation: An initial experience of 5 cases

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Objective: The craniovertebral junction (CVJ) plays a pivotal role in stabilizing and facilitating movement within the craniospinal axis. This study aimed to evaluate the clinical, radiological characteristics and surgical outcomes in patients with CVJ anomalies associated with basilar invagination and atlantoaxial dislocation.

Materials and methods: A retrospective analysis of five patients with CVJ anomalies, who underwent surgical management at Sardar Vallabhbhai Patel (SVP) Hospital, Ahmedabad, Gujarat, India. They were analyzed for clinical characteristics, radiological parameters and various surgical procedures. Patient's clinical and radiological status was assessed pre- and postoperatively at discharge and at 6 months of follow-up. Nurick grading system and Modified Japanese Orthopedic Association (mJOA) score was used. Radiological assessment was done by atlantodental interval (ADI), craniobasal angle, and craniometric lines.

Results: Most patients presented with neck pain, followed by motor weakness as the second most common symptom, while sensory deficits were the least frequent. Congenital atlantoaxial dislocation was the most prevalent CVJ anomaly observed. Clinically, significant postoperative improvements were observed in both Nurick grade and Modified Japanese Orthopedic Association (mJOA) score. Radiological findings showed a reduction in the atlanto-dens interval (ADI), a less acute clivus-canal angle, and downward movement of the odontoid process in the postoperative period. All cases had favorable postoperative outcomes, with no mortality reported at the one-year follow-up, and the condition of all patients stabilized.

Conclusion: Favorable outcomes were achieved through posterior fixation without anterior exposure in selected cases. The key to achieving excellent clinical and radiological outcomes with minimal complications lay in a thorough preoperative evaluation, timely surgical intervention, and the selection of an individualized surgical technique.

Keywords: *craniovertebral junction; basilar invagination; atlantoaxial dislocation; posterior fixation; surgical clinical; outcomes; case series*

Introduction

The craniovertebral junction (CVJ) plays a critical role in maintaining both mobility and biomechanical stability. Common developmental anomalies in this region include irreducible atlantoaxial dislocation (AAD) and basilar invagination (BI), which are frequently seen alongside occipital fusion of the C1 arch. Basilar invagination refers to the congenital upward shift of the upper cervical spine, particularly the odontoid process, into the foramen magnum, and it is often associated with AAD [1].

The aim of treatment is to stabilize the craniovertebral junction, correct any deformities or misalignments, and relieve pressure on the neural structures. The purpose of using instrumentation is to achieve immediate stability, enhance fusion outcomes, minimize postoperative external immobilization, and shorten rehabilitation periods. Common complications that may arise include dural tears, CSF leakage, infections at the surgical site, nerve or cord damage, vertebral artery damage, and failure of fusion at the bone or screw interface [2].

The progress made in neuroradiology and surgical techniques has led to improved safety, increased fusion success, fewer complications, and better clinical outcomes [3, 4]. Nevertheless, the posterior approach, including occipitocervical fusion or C1-2 fusion with or without bony decompression — remain a complex procedure due to the intricate bony and neurovascular anatomy at the CVJ [5, 6].

This case report series presents our institutional experience with posterior surgical approaches in five patients diagnosed with congenital BI and AAD.

This study sought to analyze the clinical characteristics, Nurick grading, Modified Japanese Orthopedic Association (mJOA) score and the outcomes of patients with craniovertebral instability who underwent posterior surgical treatment, either with or without decompression. It also aimed to assess the radiological results, including the measurement of craniometric and craniobasal angles, atlanto-dental interval (ADI), and bony fusion.

Material and methods

Study participants

A retrospective review was conducted on 5 patients with developmental craniovertebral junction anomalies operated on at the Neurosurgery Department SVP Hospital, Ahmedabad.

Inclusion criteria

Patients with developmental CVJ anomalies managed surgically through a posterior approach either occipitocervical or atlantoaxial fixation and fusion who had developmental irreducible BI with AAD with minimum 6 months' follow-up were included in our study.

Exclusion criteria

The following conditions led to exclusion from the study:

1. Traumatic AADs with fractures of the C1 or C2 arches or facet joints,
2. Polytrauma involving other areas of the cervical spine.
3. Rheumatoid arthritis.

Preoperative Assessment

Cases followed our inclusion criteria and they were analyzed for age, sex, clinical characteristics, radiological diagnosis, and treatment given. Patient's clinical status was assessed pre- and postoperatively (at 6 months) by the Nurick grading system and Modified Japanese Orthopedic Association (mJOA) score.

Nurick grades 0, I, and II have been considered as good and III to V as poor Nurick grade. Various craniometric lines (McRae, McGregor, Wackenheim Line, Chamberlain, Modified Ranawat Line and clivus canal line), craniobasal angle (clivus canal angle), and ADI were recorded. Standard definition exists for all the parameters mentioned.

The atlantodental interval was measured to evaluate the horizontal dislocation of C1 over C2 or C2 over assimilated C1.

The following measurements were used to assess the BI

Chamberlain Line (CL): The normal range was considered as dens being about 2.3 - 2.6 mm below this line.

Wackenheim Line: The normal value was considered being about 0.9 –2.2 mm below this line.

McRae Line (ML): The normal value was considered being about 5.8±1.6 mm below this line.

Modified Ranawat Line: From the midpoint of the base of C2, a line was drawn to meet the line drawn from the center of the anterior arch of C1 to the center of the posterior arch. The distance between the 2 lines along the long axis of C2 was measured (normal value is about 17 +-2.6 mm; a value less than this indicates BI).

The realignment was measured using the clivus canal angle (normal value ~ 150°).

CT angiogram was performed in patients to explore the size and course of the vertebral arteries between C1 and C2 segments.

Associated Chiari malformation, syringomyelia, or other intramedullary signal changes were also evaluated. Associated syringomyelia was found in 3 patients.

Postoperatively at 1 year follow-up, dynamic computerized tomography (CT) scan CVJ region was done in each case and we reassessed the various craniometric lines, ADI, craniobasal angles, and bone fusion between C1 and C2.

The posterior margin of the foramen magnum was removed in all cases, making it impossible to directly draw the cranial line (CL) and mandibular line (ML) on the postoperative images. To circumvent this, a technique based on the approach by Jian et al. was applied. A reference line was drawn from the posterior edge of the hard palate to the anterior edge of the foramen magnum. Then the angles between this reference line and CL (hard palate to posterior foramen magnum) and ML (anterior to posterior foramen magnum) were measured. On the postoperative CT scans, the reference line was drawn first, and then the CL and ML were reconstructed by transferring the measured angles.

Operative technique

All cases underwent posterior fixation and decompression. Out of 5 cases of posterior fixation, one case underwent C1 lateral mass and C2 pars screw fixation for reducible atlantoaxial dislocation (AAD) (see ***Fig. 1, Fig. 2***). O-C2 fixation (***Fig. 3, Fig. 4***) was performed in one case of C1-C2 subluxation was associated with C1 defects (assimilation, absent posterior arch) and abnormal C1-C2 joint anatomy (vertically oriented or deformed C1-C2 joint). O-C2-C3-C4 fixation was performed in three cases.

Intraoperatively, vertebral artery rupture leading to bleeding, had occurred in one case during C2 screw placement. Postoperatively, a wound infection was noticed in the same case. Both complications were managed conservatively and the patient was subsequently discharged.

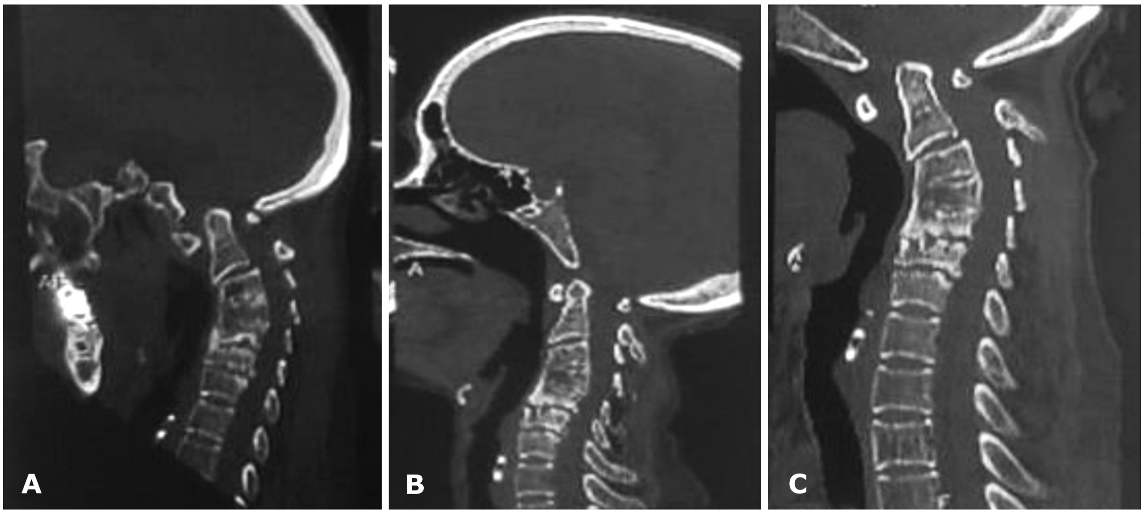
Statistical analysis

Descriptive statistics were used to summarize demographic data, clinical characteristics, radiological parameters, and outcome measures. Continuous variables such as age, ADI (atlanto-dental interval), clivus canal angle, and mJOA scores were expressed as mean ± standard deviation (SD) or median where appropriate. Categorical variables such as Nurick grades and presence of associated anomalies were reported as frequencies and percentages.

Results

Neck pain was the most common presenting symptom in our study. Other frequent manifestations were motor weakness, sensory disturbances, and restricted neck movement. Additional clinical findings are given in ***Table 1***. The majority of cases presented with symptoms duration longer than 12 months.

This article contains some figures that are displayed in color online but in black and white in the print edition.



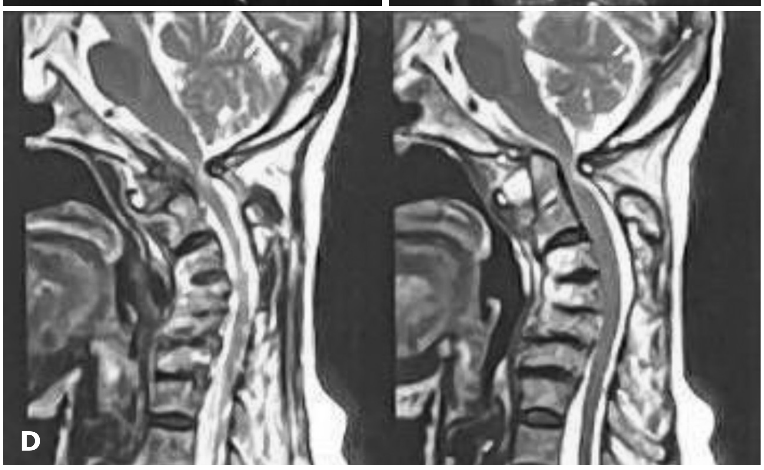


Fig. 1. Preoperative non-contrast computed tomography (CT) of craniovertebral junction (CVJ).

A - sagittal view showing atlantoaxial dislocation (AAD) in flexion

B - sagittal view showing in neutral position

C - reduction of AAD in extension

D - T2-weighted (T2W) magnetic resonance imaging (MRI) of CVJ in sagittal view

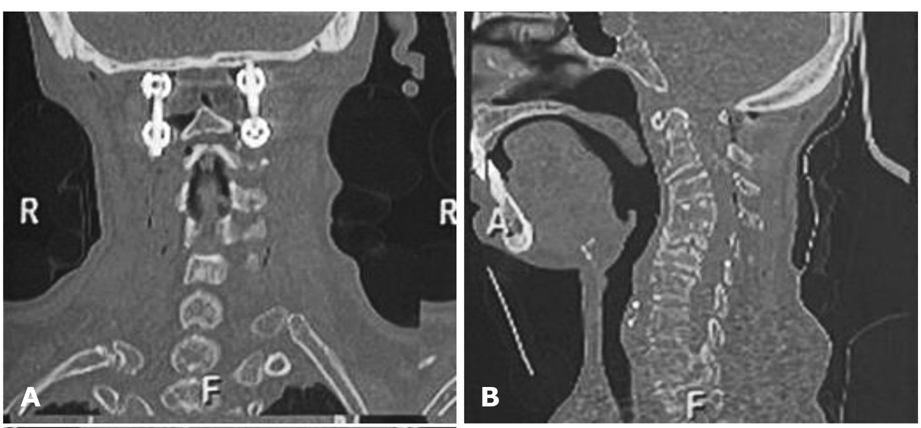


Fig. 2. Postoperative:
A – Non-contrast computed tomography (CT) showing bilateral C1 lateral mass and C2 pars screw fixation in coronal view

B – Non-contrast computed tomography (CT) of CVJ showing reduction of atlantoaxial dislocation (AAD) and widened spinal canal

C – Non-contrast computed tomography (CT) showing bilateral C1 lateral mass and C2 pars screw fixation in saggital view

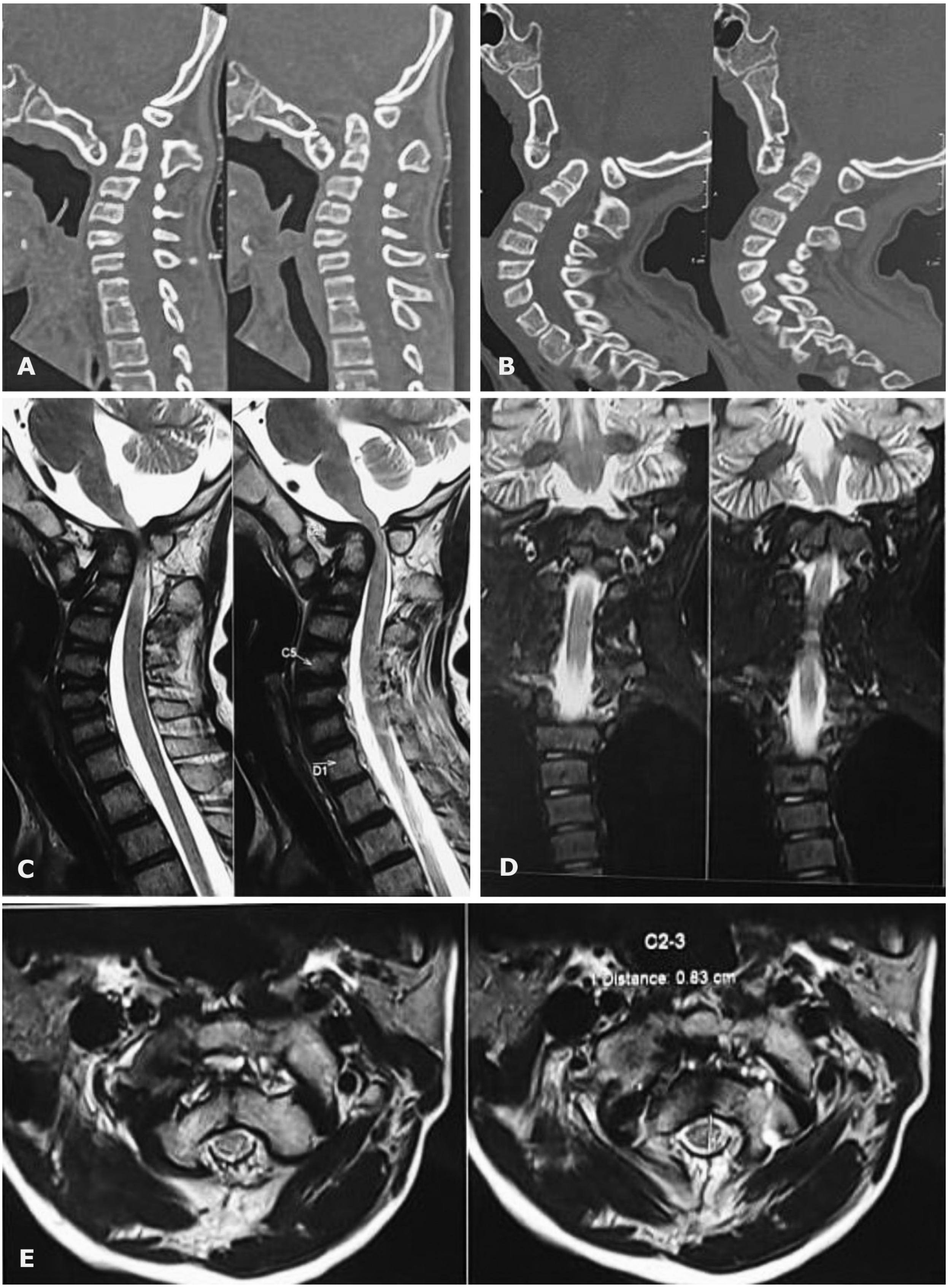


Fig. 3. Preoperative non-contrast computed tomography (CT) of craniovertebral junction (CVJ).
A - sagittal view showing in flexion position
B - sagittal view showing in extension position
C,D,E - T2-weighted (T2W) magnetic resonance imaging (MRI) CVJ sagittal view and axial cuts, showing cord compression with hyperintense changes in the cord at the CVJ region.

Fig. 4. Postoperative computed tomography (CT) of craniovertebral junction (CVJ).
A - Coronal view showing occipital plate and C2 pars screw
B - Sagittal view showing occipital plate and C2 pars screw

Table 1. Clinical findings and duration of symptoms

Clinical findings		Number
Neck pain		4
Restricted neck movement		2
Motor weakness	Quadriparesis	0
	Hemiparesis	4
Sensory disturbances	Posterior column involvement	3
	Spinothalamic tract involvement	2
Cerebellovestibular disturbances		0
Lower cranial nerve palsy		0
Sphincter disturbances		0
Respiratory involvements		0
Neck tilt, low hair line, short neck		4
Thenar/hypothenar muscle wasting		1
Duration of symptoms (in months)	Up to 6	1
	7–12	1
	13–24	2
	≥ 25	1

Congenital AAD was the most common congenital CVJ anomaly. Other common congenital CVJ anomalies are given in ***Table 2***.

In preoperative period, all patients were classified as having poor Nurick grades and low mJOA scores. In the postoperative period, all patients had improved in Nurick and m-JOA grading as compared with preoperative scores. (***Table 3***).

In the postoperative period, there was a significant shortening of distance of the odontoid process above the defined craniometric lines. Radiologically, a significant decrease in the ADI was observed postoperatively. Preoperatively, clivus canal angle was more acute as compare to the postoperative period (***Table 4***). Both, the decrease in ADI and the increase in clivus canal angle, suggest a reduction in the ventral compression of the cord postoperatively.

Various radiological craniometric lines were calculated. Some of examples are given in the image below (***Fig. 5, Fig. 6***).

Table 2. Distribution of congenital anomaly

Congenital anomaly	Number
Atlantoaxial dislocation	5
Basilar invagination	4
Chiari malformation	2
Assimilation of atlas	4
Unilateral deficient posterior arch of atlas	0
Block vertebrae	1
Klippel–Feil syndrome	0
Platybasia	0

Table 3. Associated radiological anomalies, surgery performed, clinical outcomes

No	Age	Sex	Radiology	Surgery	Nurick grading		m-JOA grading	
					pre-op	post-op	pre-op	post-op
1	29	F	C1A,Syr	Oc- C2-3-4	4	1	14	16
2	57	M	C1A	Oc-C2-3-4	3	0	15	17
3	52	F	C1A,Syr	Oc-C2-3-4	4	2	11	10
4	11	M	C1A	Oc-C2	4	2	10	14
5	61	F	C1A	C1-C2	4	1	15	18

Notes. M=male; F=female; C1A=C1 arch assimilated with occiput; C1NA=C1 arch not assimilated with occiput; Ch=Chiari malformation; Syr=syringomyelia; Oc-C2= occipito-C2 distractive compressive reduction performed; C1/C2=C1 and C2 distractive compressive reduction performed; preop=preoperative; postop=postoperative, Syr=syringomyelia

Table 4. Pre- and postoperative radiological findings

Patient No.	Age,Y	Sex	ADI, mm		CL (Normal = 2.3±2.6 mm)		ML (Normal = 5.8±1.6 mm)		WL (Normal = 0.9±2.2 mm)		CCA (Normal.150°)		RL (Normal = 17±2.6 mm)	
			pre-op	post-op	pre-op	post-op	pre-op	post-op	pre-op	post-op	pre-op	post-op	pre-op	post-op
1	29	F	3.9	2.2	-8.9	3.3	-3.2	4.8	26.4	1	115	155	7	17
2	57	M	2.1	1.8	-7.1	3.8	-3.58	4.5	22.4	1.2	106	130	3	19
3	52	F	3.1	1.4	-9.8	0.2	-4.5	1.2	25.4	1.1	141	135	10	23
4	11	M	5	1	-12.9	3.6	-15.3	1.1	-11.3	1.5	138	156	3.5	23
5	61	F	4	0	-6.1	2.2	-7.4	1.9	-6.5	-1.2	116	134	9	20

Notes. ADI, atlantodental interval; CL, Chamberlain line; ML, McRae line; WL, Wackenheim line; CCA, clivus canal angle; RL, modified Ranawat line; M, male; F, female; preop, preoperative; postop, postoperative.

Negative value indicates that the dens is placed above this line.

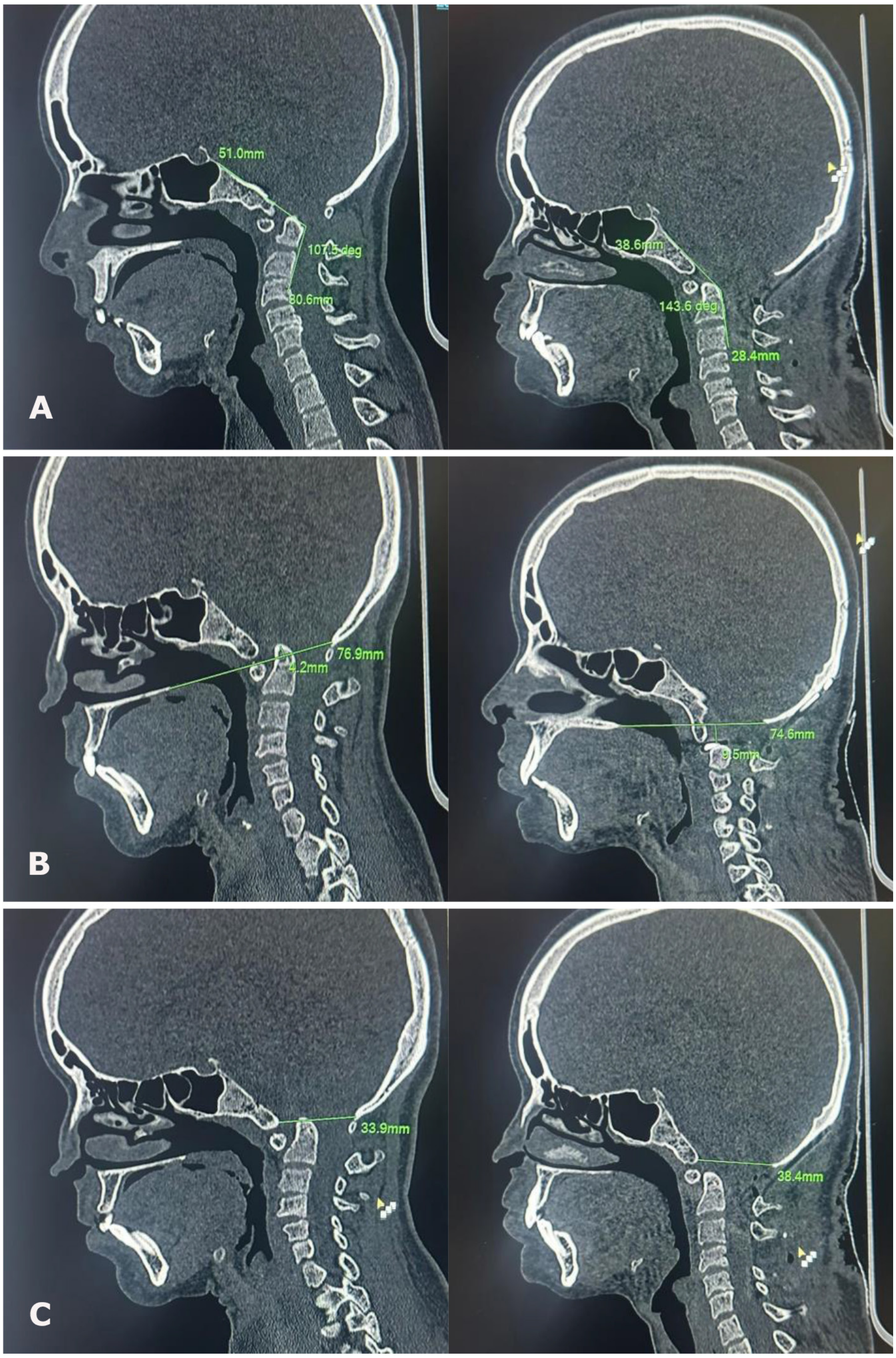


Fig. 5. A - pre-op and post op clivus canal angle
B - pre and post op chamberlain line
C - pre and post op McRae line

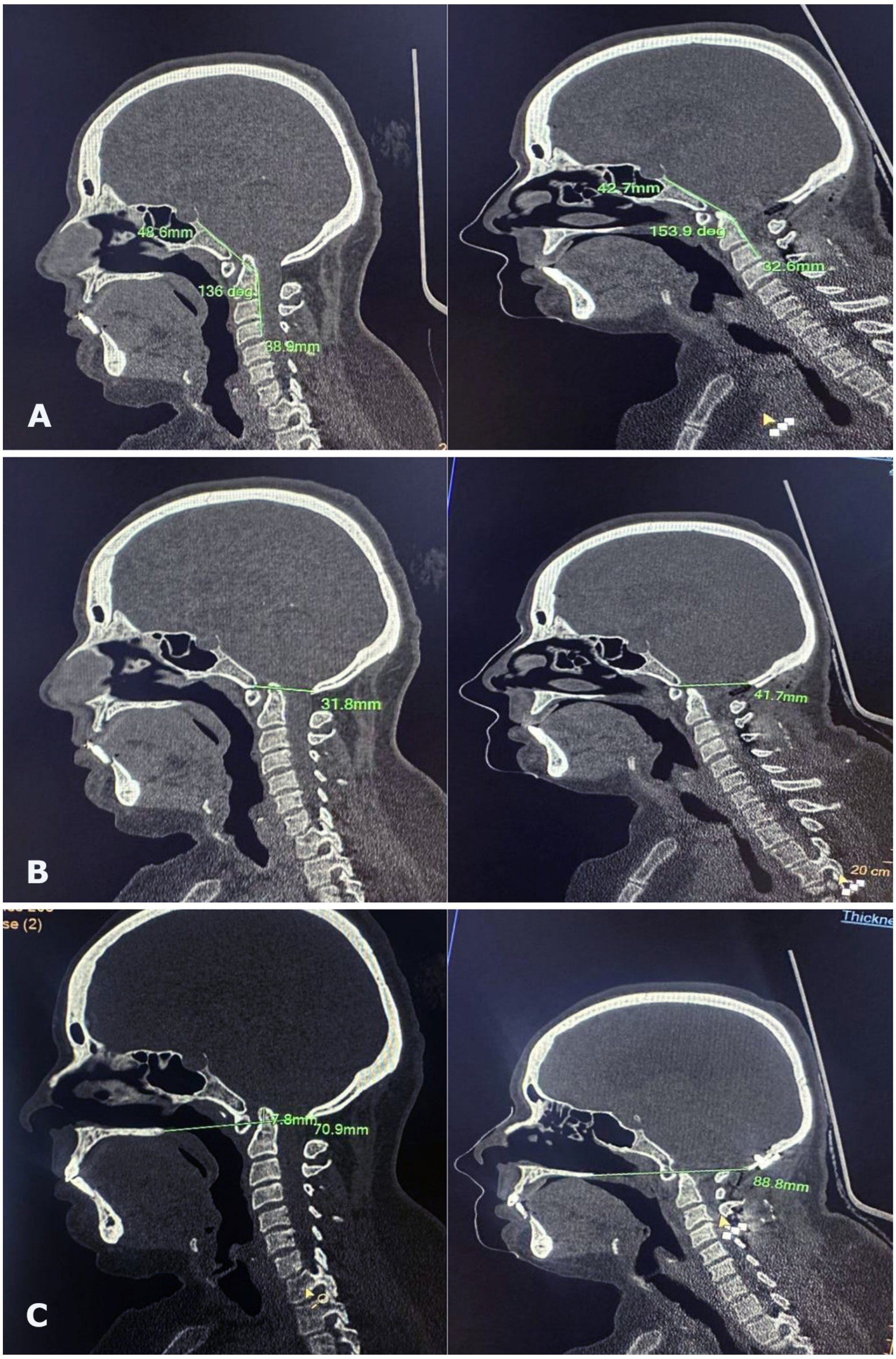


Fig. 6. A - pre and post op clivus canal angle
B - pre and post op McRae line
C - pre and post op Chamberlain line

Discussion

The main objectives of surgery in CVJ anomalies are to alleviate compression and stabilize the CVJ. In most cases, both of these goals can be effectively achieved through a single posterior midline approach. While posterior approaches have gained popularity due to lower morbidity and improved instrumentation, ventral decompression techniques remain crucial in managing rigid anatomical constraints and severe ventral cord compression.

Previously, semi-rigid wire and loop fixation devices were used for stabilization [7]. However, recent studies have shown that rigid screw fixation provides superior results [8, 9].

Atlantoaxial dislocation (AAD) and basilar invagination (BI) are complex conditions that lead to progressive cervicomedullary compression and neurological deficits. The primary goal of treatment is to alleviate compression, provide adequate stability, and correct any deformities. These congenital anomalies typically cannot be reduced using cervical traction with dynamic X-rays. Most cases are associated with atlas assimilation to the occiput. Notably, most of our patients had irreducible AAD and BI. Traditionally, BI with AAD was managed through an anterior transoral approach to remove the odontoid process, followed by posterior instrumented fixation.

The common clinical presentations in our study were neck pain, motor weakness, sensory disturbances, restricted neck motion. A study done by Goel has similar presentations except lower number of cases presenting with cerebellovestibular disturbances and respiratory involvement [10]. The differences may be due to less number of cases in our study.

Goel compared the ADI preoperatively and postoperatively and achieved a satisfactory decrease in ADI in more than 80% of the patients [10]. In our study, we also attained a satisfactory decrease in ADI in all cases.

After fixation, the bone fusion rate of CVJ anomalies has been reported to be highly successful (75–100%) as described by previous studies regardless of fixation material used and underlying pathologies [11, 12]. In our study, 6 months after surgery solid bone fusion was achieved in all cases, this is comparable to other studies. Excellent results have been reported by Grob et al (100%) and Wertheim et al (100%) [13, 14].

Complications such as dural tear was noted in one case. Intraoperative verterbral artery rupture, wound infection had occurred in one case in our study.

Distraction, compression, extension, and reduction. Chandra et al [15, 16] described an innovative technique for reducing BI with AAD via a single-stage posterior approach. This approach avoids the need for an anterior transoral procedure and involves both the use of an interarticular spacer to distract the C1-C2 joints and the application of horizontal occipito-cervical manipulation to address the CVJ deformity secondary to AAD. The technique is especially suited for cases where C1 is fused with the occiput.

Non-reducible basilar invagination is classified into three types based on the sagittal inclination (SI) angle, which is the angle between a line parallel to the axis of the odontoid process (tangential to its posterior border) and a line parallel to the C1-C2 joint surface. The normal range of the SI angle is 87.15 ± 5.65°. The classification is as follows: type I for SI angles <100°, Type II for 100–160°, and Type III for >160°. Chandra et al. advised particular surgical interventions for each type, and further details can be found in their manuscript.:

Type I: DCER

Type II: Joint remodeling+DCER

Type III: Extra-articular distraction+DCER

Two main approaches have emerged for the management of non-reducible BI and AAD:

1. Posterior decompression and reduction with occipitocervical fixation.

2. The DCER procedure that includes the application of a C1–C2 facet joint spacer.

Both recent methods resulted in complete fusion. The techniques including the C1–C2 joint spacer and vertical reduction yielded significantly better outcomes in terms of index correction and reduction rates. Although it is difficult to compare major complications in detail due to the limited number of cases, vertebral artery injury was the most serious complication reported in both techniques, with a slightly higher incidence in DCER procedures. In accordance with current literature and the findings from Chandra et al., we successfully applied the DCER technique for the first time at our institution to treat individuals with BI and AAD.

Ventral decompression techniques in irreducible BI and AAD:

When dealing with fixed deformities and non-reducible basilar invagination (BI) or atlantoaxial dislocation (AAD), direct anterior decompression is often required to relieve pressure on the brainstem and upper cervical spinal cord. In such cases, the transoral odontoidectomy, as described and popularized by Alan Crockard and colleagues, remains the gold standard for direct ventral decompression [17–19].

Crockard's work laid the foundation for modern transoral approaches, particularly in patients with significant ventral compression due to an upwardly displaced odontoid process. His studies demonstrated that transoral odontoidectomy allows for safe and effective removal of the odontoid process through an intraoral route, thereby relieving ventral pressure without the need for extensive brain retraction or dural manipulation [17]. This approach is especially beneficial in cases where the sagittal inclination (SI) angle exceeds 160° (Type III deformity), making posterior reduction inadequate or impossible.

Combined anterior-posterior approach: Crockard's legacy. One of the most important contributions of Alan Crockard to CVJ surgery was his advocacy for a combined anterior-posterior approach — particularly in complex cases involving irreducible BI and AAD. He emphasized that while anterior decompression addresses the immediate threat to neural structures, posterior stabilization provides the biomechanical support necessary for durable outcomes.

In his seminal work, Crockard described a two-stage procedure:

1. Anterior transoral odontoidectomy to remove the offending bony structure causing ventral compression

2. Posterior occipitocervical or C1–C2 fixation to stabilize the CVJ and maintain alignment post-decompression

This combined strategy has since become the benchmark for managing severe, non-reducible CVJ pathologies, especially in the context of congenital anomalies, rheumatoid arthritis, and post-traumatic instability.

Comparison with Posterior Approaches:

While anterior decompression remains the gold standard in certain scenarios, posterior-only approaches have shown promising results in carefully selected patients. As highlighted by Goel and Chandra et al., patients with reducible or semi-reducible BI and AAD can achieve satisfactory outcomes using posterior fixation alone, avoiding the morbidity associated with transoral surgery [10, 15, 16].

In our series, we observed satisfactory clinical and radiological outcomes following posterior decompression and fixation without anterior exposure. Our findings align with those of Goel and Chandra et al., who have demonstrated that carefully selected patients with reducible or semi-reducible BI and AAD can benefit from posterior-only strategies [10, 15, 16]. Specifically, we noted a consistent reduction in the atlantodental interval (ADI) and improvement in clivus-canal angles postoperatively, indicating effective reduction and decompression of the cervicomedullary junction.

As highlighted by Crockard and others, ventral compression in the presence of rigid anatomical constraints or severe fixed deformities often necessitates anterior decompression to avoid catastrophic complications such as tetraplegia or respiratory compromise [17, 18]. Therefore, preoperative imaging—particularly dynamic MRI and CT angiography—is essential to assess reducibility, vertebral artery course, and the degree of ventral cord compression.

Although, the anterior transoral approach remains the gold standard for definitive ventral decompression in non-reducible BI with AAD, our experience suggests that selected patients with certain morphological characteristics may benefit from a posterior-only strategy. Meticulous patient selection, accurate radiological assessment, and intraoperative neuromonitoring are imperative to ensure safe and effective outcomes.

Future studies with larger cohorts and comparative analyses between anterior and posterior approaches are warranted to further refine surgical indications and optimize patient care.

Conclusion

We noted satisfactory clinical improvement and vertical reduction. Congenital CVJ anomalies were the most common lesions found. The majority of patients presented with neck pain, motor weakness, and sensory deficits. Favorable outcomes, both clinically and radiologically, were seen in most cases. Proper preoperative evaluation and careful selection of individualized surgical techniques are crucial for minimizing complications. Our case series supports the use only posterior fixation without anterior exposure in patients with reducible or semi-reducible anomalies. The findings of this study may be subject to revision in the future with a more streamlined, single-stage approach, given the small sample size and potential selection bias.

Disclosure

Conflict of interest

The authors declare no conflicts of interest.

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The study was not sponsored.

Informed consent

Informed and voluntary written consent to participate in the study and publication of data was obtained from all patients.

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