



Original Article

# Foramen magnum decompression without bone removal: C1–C2 posterior fixation for Chiari with congenital atlantoaxial dislocation/basilar invagination

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## ABSTRACT

**Background:** Atlantoaxial dislocation (AAD) and basilar invagination (BI) may coexist with Chiari malformations (CM) and a small posterior fossa volume. These are typically treated with craniocervical junction fusion and foramen magnum decompression (FMD). Here, we evaluated whether C1–C2 posterior reduction and fixation (which possibly opens up the ventral foramen magnum) would effectively treat AAD and CM without additionally performing FMD.

**Methods:** This is a retrospective analysis of 38 patients with BI, AAD, and CM who underwent C1–C2 posterior reduction and fusion without FMD. Baseline and follow-up clinical, demographic, and radiological data were evaluated.

**Results:** The vast majority of patients (91.9%) improved both clinically and radiographically following C1–C2 fixation alone; none later required direct FMD. Notably, AAD was irreducible in 25 (65.8%) patients. Preoperatively, syringomyelia was present in 28 (73.7%) patients and showed resolution. In 3 (8.1%) patients, resolution of syrinxes did not translate into clinical improvement. Of interest, 5 patients who sustained inadvertent dural lacerations exhibited transient postoperative neurological worsening.

**Conclusions:** Posterior C1–C2 distraction and fusion alone effectively treated AAD, BI, accompanied by CM. The procedure sufficiently distracted the dens, reversed dural tenting, and restored the posterior fossa volume while relieving ventral brainstem compression making FMD unnecessary. Surgeons should, however, be aware that inadvertent dural lacerations might contribute to unwanted neurological deterioration.

**Keywords:** Atlantoaxial dislocation, basilar invagination, C1–C2 distraction, chiari, foramen magnum decompression, syrinx, ventral decompression

## INTRODUCTION

Chiari malformations (CM), associated with a reduced volume of the posterior fossa, are typically treated with foramen magnum decompression (FMD) (i.e., to enlarge the posterior fossa volume).<sup>[4,9,10]</sup> When

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CM are combined with atlantoaxial dislocation (AAD) and basilar invagination (BI), the posterior fossa volume is even further reduced; the instability requires C1–C2 distraction, realignment, and arthrodesis.<sup>[1,5-7,10]</sup> The fixation *per se* is likely to relieve anterior cervicomedullary compression and increase the overall posterior fossa volume.

In this study, we, therefore, evaluated whether C1–C2 distraction and arthrodesis obviated the need for additional FMD.

## MATERIALS AND METHODS

### Patient data

In this retrospective analysis (2015–2017), 38 patients with AAD/BI and CM underwent C1–C2 fixation without FMD (neither bony nor dural decompression). Patients averaged 40.5 years of age and demonstrated a mean modified Japanese Orthopaedic Association (JOA) score at the time of presentation of 11.3 [Table 1]. They all underwent dynamic plain radiographs showing a mean preoperative atlantodental interval of 6.8 mm.

### Computed tomography (CT) and CT angiography (CTA): Preoperative findings

Preoperative CT studies were utilized to evaluate the craniovertebral junction, while CTA additionally documented any anomalous vertebral arteries [Table 2].

### Magnetic resonance imaging (MRI) findings

MRI confirmed the diagnosis of CM, syringomyelia, and the degree of cervicomedullary compression. For those with an intact C1 arch, the extent of tonsillar descent of at least 5 mm below the FM while for those with assimilated C1 arches, any descent of the tonsils below the posterior arch of C1 was considered CM.

### Perioperative and operative considerations

About 48-h before surgery, cervical traction was applied. The AAD was reducible in 13 (34.2%) patients but was irreducible in 25 (65.8%) who underwent intraoperative reduction.

All patients then had C1–C2 posterior reduction and fusion.<sup>[5-7]</sup> Screw and spacer positions were confirmed with the O-arm. Postoperatively, the patients wore a Philadelphia collar without an extension brace for 3 months.

### Clinical and MR/CT follow-up

At 6 postoperative months, the thin slice CT documented bony fusion, while the MRI scans documented the extent of cephalad cerebellar tonsil migration, the degree of cerebrospinal fluid (CSF) flow across foramen magnum, and whether there was the resolution of the syrinxes.

### Statistical analysis

Statistical Package for the Social Sciences version 21 (IBM Corp., New York, USA) was used for the data analysis. The mean preoperative and follow-up modified JOA score were compared using the paired *t*-test.  $P < 0.05$  was considered significant.

## RESULTS

After C1–C2 fusion, satisfactory reduction and realignment were achieved in all cases [Figures 1-3]. The mean descent of tonsils in patients with assimilated versus intact C1 arches was, respectively, 8 mm and 5.2 mm below the FM. There was no posterior CSF space seen in any of the patients. Although only 7 patients had symptoms attributable to syringomyelia, 28 (73.7%) showed syrinx formation in addition to CM.

### Immediate postoperative complications

Six patients exhibited transient neurological worsening on postoperative days 2–3 [Table 3] [Figure 4]. Notably, 5 among them had an inadvertent dural laceration and CSF leak.

**Table 1:** Clinical manifestations of patients ( $n=38$ ).

Clinical features	n (%)
Spastic quadriparesis	36 (94.7)
Neck pain	14 (36.8)
Posterior column involvement	29 (76.3)
Hoarseness of voice	8 (21.1)
Dysphagia/nasal regurgitation	9 (23.7)
Bladder/bowel complaints	11 (28.9)

**Table 2:** Radiological findings of patients ( $n=38$ ).

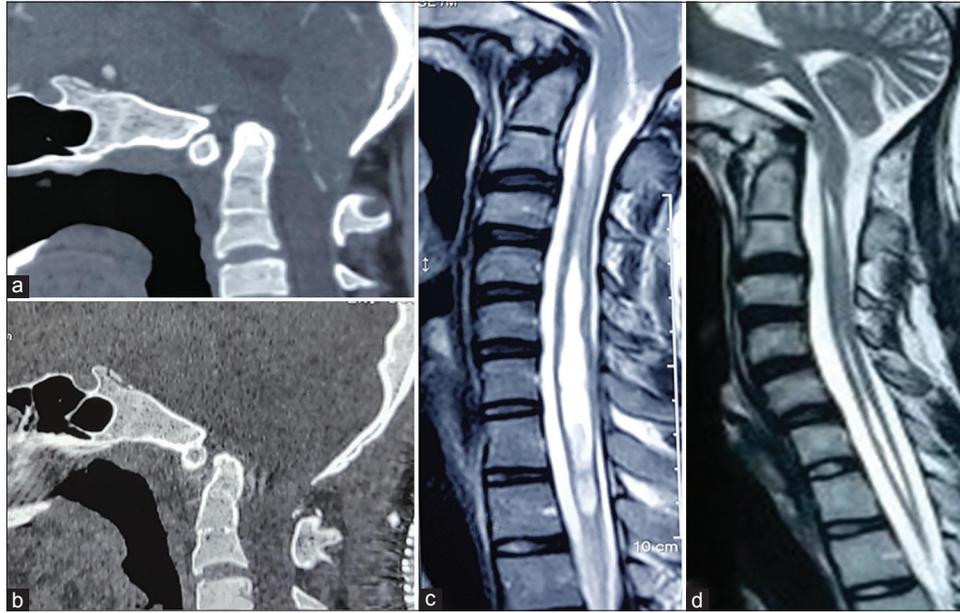
Parameter	Assimilated C1 arch	Intact C1 arch
Mean tonsillar descent	8 mm	5.2 mm
Irreducible AAD* ( $n=25$ )	25 (65.8%)	0
Reducible AAD ( $n=3$ )	7 (18.4%)	6 (15.8%)
Vertebral artery anomaly	11 (28.9%)	0

\*AAD: Atlantoaxial dislocation

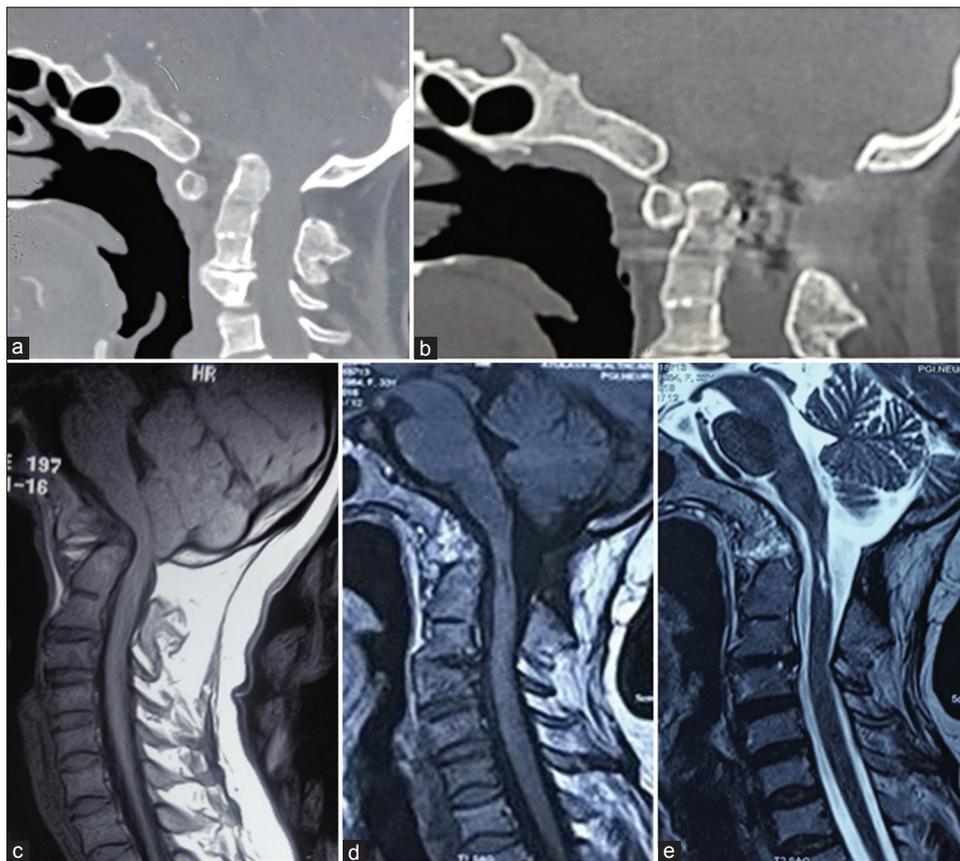
**Table 3:** Immediate postoperative complications ( $n=6$ ).

Clinical manifestations	n
Respiratory distress	6
Ataxia	6
Cerebellar infarct and hydrocephalus*	1
Cerebellar ischemia <sup>†</sup>	1
Vertebral artery injury	1
CSF leak	5

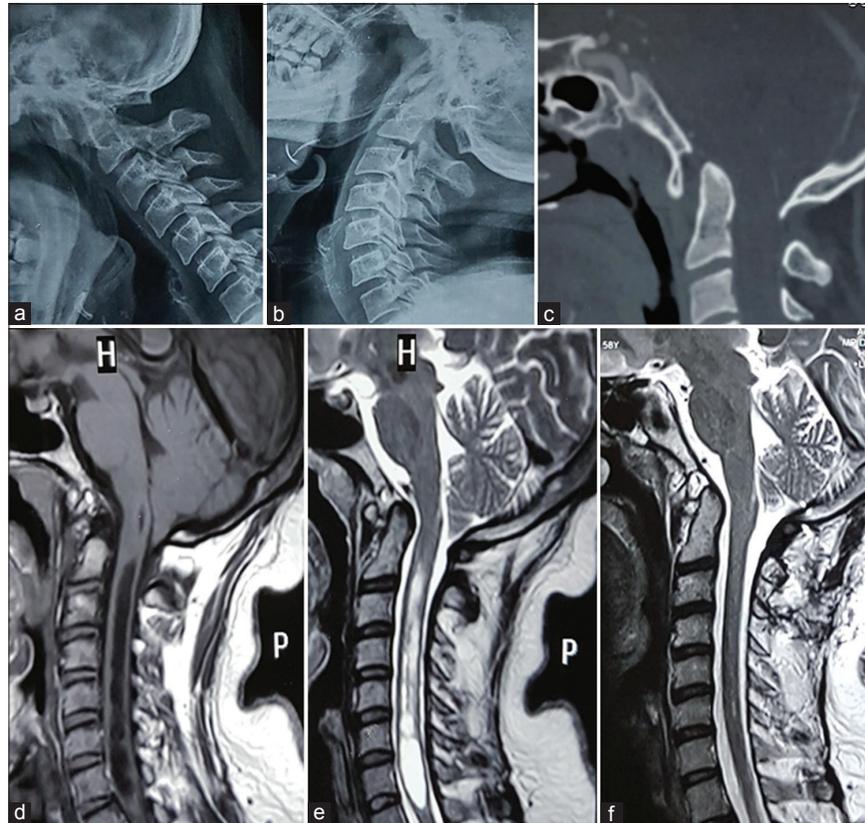
\*Required ventriculoperitoneal shunt, <sup>†</sup>Expired at 3 months due to chest infection



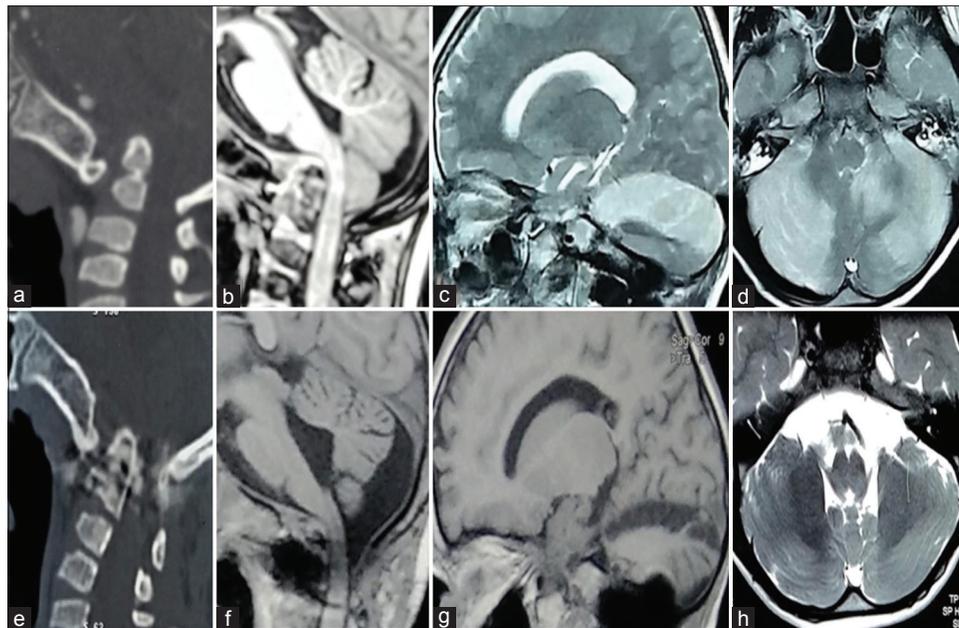
**Figure 1:** (a) Preoperative midsagittal computed tomography (CT); basilar invagination and atlantoaxial dislocation (AAD) with segmentation defects. (b) Postoperative CT; reduction of deformity with intact foramen magnum margin. (c) Preoperative T2-weighted sagittal magnetic resonance imaging; Chiari malformations and syringomyelia. (d) Follow-up imaging (6-months) shows significant resolution of the syrinx.



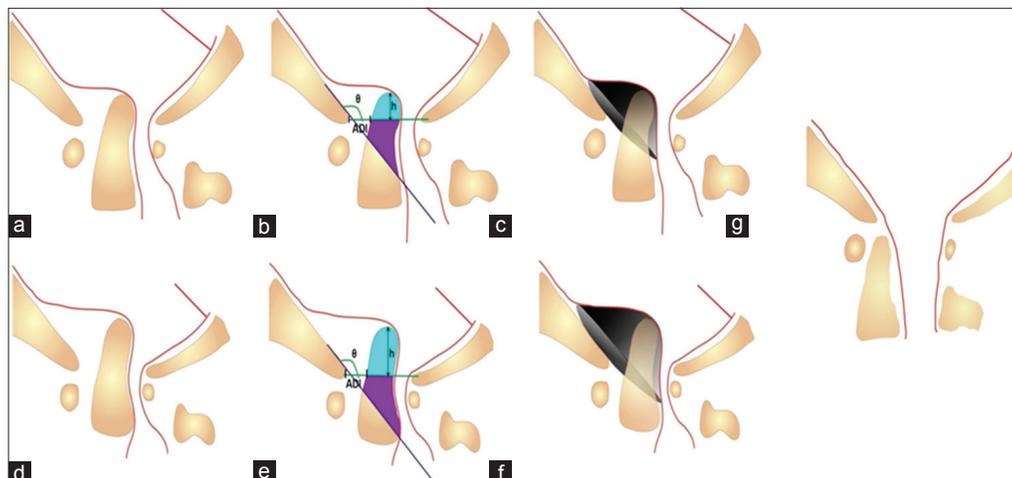
**Figure 2:** (a) Computed tomography (CT) shows basilar invagination and atlantoaxial dislocation with segmentation defects. (b) Postoperative CT after distraction and realignment of dens. (c) T1-weighted magnetic resonance imaging (MRI) shows Chiari with crowding of posterior fossa structures. (d and e) Postoperative MRI; T1-weighted (d) and T2-weighted (e) show ascent of tonsils and the opening of cerebrospinal fluid space.



**Figure 3:** (a and b) Preoperative dynamic radiographs show reducible atlantoaxial dislocation (AAD). (c) Midsagittal computed tomography demonstrates basilar invagination and AAD; C1 is assimilated. (d and e) T1 (d) and T2 (e) weighted magnetic resonance imaging (MRI); Chiari with crowded posterior fossa and syrinx. (f) Follow-up MRI (12 months) shows ascent of tonsils and striking resolution of the syrinx.



**Figure 4:** Postoperative course with the inadvertent dural opening. Preoperative computed tomography (CT) (a) depicts BI with atlantoaxial dislocation; T1-weighted magnetic resonance imaging (MRI) (b) shows Chiari. (c and d) MRI show acute cerebellar infarcts in the postoperative period. (e) Postoperative CT shows a reduction of the deformity. (f-h) On follow-up, there is the ascent of tonsils with the good restoration of CSF space (f) and resolving infarcts (g-h).



**Figure 5:** Dural indentation in basilar invagination (BI) with Chiari. (a-c), moderate and (d-f), severe dislocation. A portion of dens that lies above foramen magnum (FM) and that violates Wackenheim's line is marked in different colors (b and e). This causes ventral compression. However, the actual volume of posterior fossa (PF) (gray cone) that is reduced is far more than the volume of dens alone (c and f). C1–C2 distraction and reduction not only relieves ventral compression but also restores PF vol. PF volume is given by Boogaard's angle ( $\theta$ ), atlantodental interval and height of dens (vertical dislocation) that invaginates above FM (h). (g) Normal anatomy.

### Long-term outcome

Fusion was documented in 37 of 38 patients from 11 to 40 months postoperatively; of interest, none required additional FMD. The mean improved modified JOA score of 13.6 ( $P < 0.01$ ) was seen in 34 patients (91.9%) who showed neurological improvement. Their MRI studies also documented ascent of the tonsils with the restoration of CSF space and resolution of syrinxes [Figures 1-3]. Of interest, for 3 (8.1%) of patients, despite radiological re-establishment of CSF flow and syrinx resolution, there was no correlation with clinical improvement. This was possibly due to pre-existing irreversible damage to the spinal tracts.

### DISCUSSION

AAD/BI with CM was successfully managed in this series utilizing posterior C1–C2 reduction and fusion without FMD.<sup>[1,10]</sup> Wang *et al.* successfully performed posterior C1–C2 fusions with bony FMD without duraplasty; they demonstrated adequate decompression and a 93% incidence of syrinx resolution.<sup>[10]</sup> Goel *et al.*, however, proposed treating CM with or without AAD, utilizing C1–C2 fusion alone.<sup>[2,3]</sup> He also proposed that CM are a result of underlying instability whereas we suggest that distraction, reduction, and C1–C2 arthrodesis reverse ventral dural kinking and restores posterior fossa volume thereby addressing Chiari. Notably, in our series, all 38 patients underwent C1–C2 short segment fusion (irrespective of segmentation anomaly), achieved adequate decompression and improved without FMD.<sup>[8]</sup> In fact, dural opening may be dangerous as noted in our study. Patients with inadvertent dural lacerations developed transient neurological worsening possibly due to a ball valve mechanism and change in CSF dynamics.

### Mathematical formula to estimate the reduction in posterior fossa volume secondary to encroachment by dens in AAD/BI and the rationale of surgery

The absolute volume of conical dens that violates the Wackenheim's clivus canal line is given by the formula  $\pi d^2 [VD/12 + \tan(180-\theta) \cdot d/8]$ ,  $d$  being the diameter of dens at the level of FM and  $VD$ , the height of dens that invaginates above FM. The effective reduction in posterior fossa volume is far greater than the absolute volume of dens and is approximately  $\pi [VD \cdot \cos(180-\theta) + ADI \cdot \sin(180-\theta)]^3 / 3 \cdot \tan(180-\theta)$  where  $\theta$  is the Boogaard's angle and  $ADI$  is the atlantodental interval [Figure 5]. Our technique of C1–C2 distraction and realignment relieved ventral compression and restored posterior fossa volume obviating the need for a transoral surgery as well as FMD.

### CONCLUSIONS

C1–C2 fusion without FMD adequately addressed AAD, BI with CM in 38 patients in this series. Distraction, reduction, and C1–C2 arthrodesis increased the posterior fossa volume resulting in adequate ventral cervicomedullary junction decompression without the need for additional FMD.

### Compliance with ethical standards

The procedures performed were in accordance with the ethical standards of the Institutional Ethics Committee and with the 1975 Helsinki declaration and its later amendments.

### Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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