



## Case Report

# Dysplasia and anomalies of atlas result in pediatric torticollis: A case report and literature review

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## ABSTRACT

**Background:** Often, the cause of bony torticollis is difficult to determine, especially in cases of multiple craniovertebral junction anomalies.

**Case Description:** We report a rare case of a dysplastic C1 vertebra (assimilation to the right occiput and C2, a nonseparated left odontoid, and discontinuity in both anterior and posterior arches of the atlas) in a 6-year-old child with progressive torticollis. Notably, the mechanism of torticollis was not a rotatory subluxation of C1-C2, but differential growth between C1-C2. The child underwent a successful C1-C2 Goel and Harms fusion with reduction/correction of the torticollis.

**Conclusion:** Torticollis caused by differential growth between the C1 and C2 vertebrae resulting in a nonrotatory subluxation/torticollis in a 6-year-old child, was successfully managed with a C1-C2 Goel and Harms fusion.

**Keywords:** Dysplasia, Occipitalization, Torticollis

## INTRODUCTION

Torticollis may result from a congenital craniovertebral junction (CVJ) anomaly. Here, in a 6-year-old child with progressive torticollis, we recognized a dysplastic C1 vertebra showing assimilation to the right occiput and C2, accompanied by a nonseparated left odontoid, and discontinuity in both anterior and posterior arches of the atlas. This was successfully managed with a C1-C2 Goel and Harms fusion.

## CASE DESCRIPTION

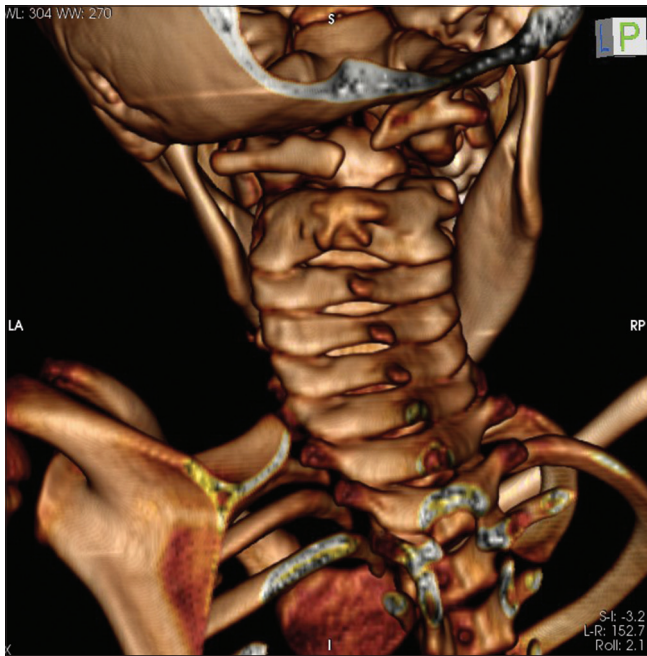
A 6-year-old female presented with a 3-year history of a progressive torticollis toward the right side. It was associated with facial asymmetry including a smaller right eye [Figure 1]. There was no history of any prior untoward events during the pregnancy. The patient had no neurological deficits. The radiographs revealed evidence of fusion between the C2 and C3 vertebral bodies with a dysplastic C1, while the CT scan confirmed an absent C1 spinous process and adjacent lamina (posterior arch deficiency) [Figure 2]. The CT scan showed additional bony anomalies as explained in [Table 1] and [Figures 3 and 4]. MRI showed no evidence of cord compression [Figure 5].

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**Figure 1:** Clinical photograph showing right-sided torticollis with features of facial asymmetry including smaller eye size.

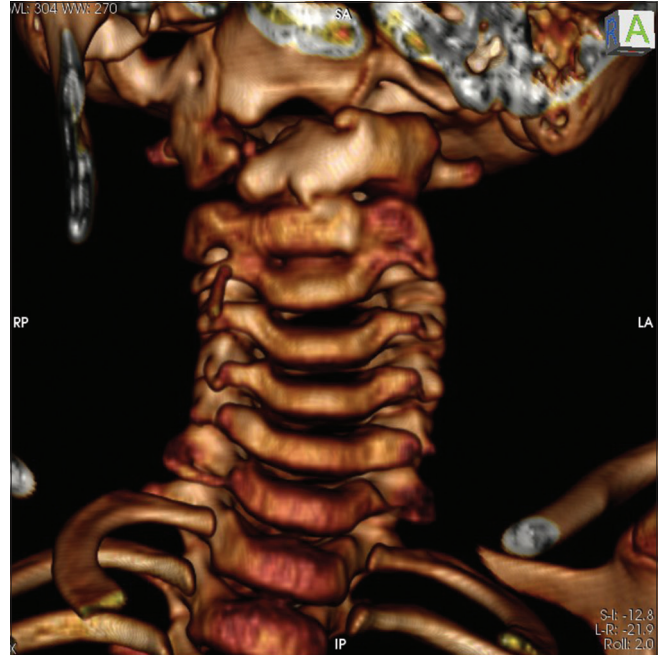


**Figure 2:** 3D CT scan showing absent C1 spinous process with adjacent lamina (posterior arch deficiency) and complete fusion of posterior elements of C2 and C3.

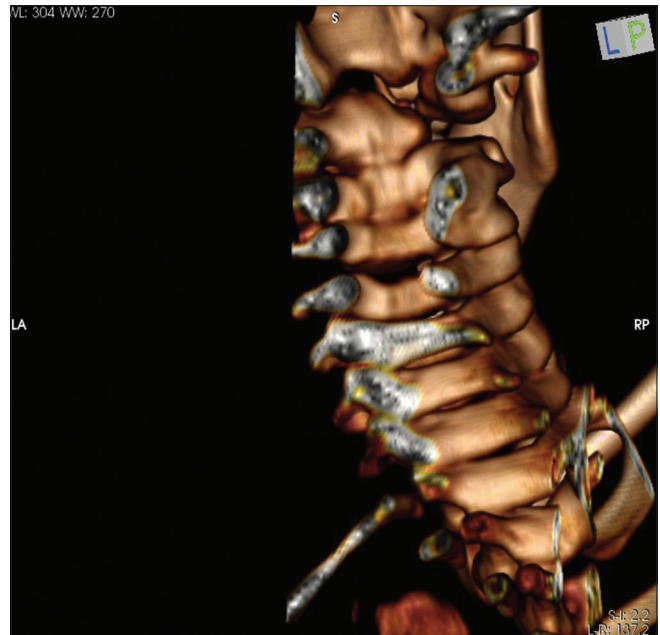
### Surgical management

The progressive torticollis was treated with a C1 and C2 Goel and Harm fusion. During the surgery, on the right side, the interfacet space between C1 and C2 was large in contrast to the left side. Articular cartilage was debrided and cancellous bone grafting (e.g., harvested from the posterior iliac crest) was performed.

Similar steps were followed on the left side. Next, C1 lateral mass and C2 pedicle screw were inserted with increased



**Figure 3:** 3D CT scan showing occipitalization of the right side of lateral mass of C1, discontinuity in anterior arch of C1, left side lateral mass of C1 with residual anterior arch of C1 showed fusion with body of C2 anteriorly with minimal space between left lateral mass of C1 and C2 in contrast to the right side.



**Figure 4:** CT scan showing subtraction view showing odontoid nonseparation and its assimilation with anterior arch of atlas.

attention to the dysplastic C1 and C2 anatomy. The screw trajectory on the right side was kept high in sagittal plane to adjust for occipitalization of the right lateral mass of the

**Table 1:** Anomalies as evident on CT scan.

S. No.	Anomalies present
1.	Occipitalization of the right side of atlas
2.	Discontinuity in the anterior arch of C1
3.	Fusion of the left lateral mass of C1 (with its residual anterior arch) with body of C2 anteriorly
4.	Reduced interfacet distance on the left side between C1 and C2
5.	Assimilation of odontoid with remnant of anterior arch of atlas
6.	Complete fusion between posterior elements of C2 and C3

atlas. As there was fusion between the C1 and C2 bodies, we passed larger bilateral pedicle screws into the C2 vertebral body [Figure 6].

Intraoperative reduction/compression was performed on the right side between the C1 and C2 screws to correct the torticollis [Figure 7]. Two years later, the patient remains asymptomatic [Figure 8].

## DISCUSSION

It is extremely rare to find dysplastic C1 vertebrae causing torticollis without rotatory subluxation of C1 and C2. A report by Geipel<sup>[3]</sup> stated that incidence of defects in posterior arch of atlas was about 4%, and defects of the anterior arch of the atlas were 0.1%.

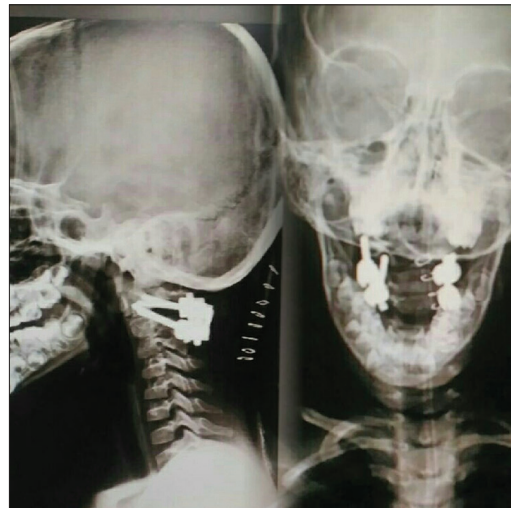
Cave had classified atlantoaxial congenital fusion into three types: (1) fusion of a separated odontoid process with the anterior atlantal arch; (2) complete (bilateral) fusion of atlas and axis; and (3) incomplete (unilateral) fusion, with or without some degree of assimilation.<sup>[1]</sup> The present case was fitting in to subclass of 3 of Cave classification with regard to atlantoaxial congenital fusion in addition to other congenital anomalies. It is extremely rare to find fusion of a nonseparated odontoid process with the anterior arch of the atlas.

Previously published literature has reports about bipartite atlas. However, in our case, atlas cannot be called bipartite, as defect on anterior side was not in midline and there was unilateral assimilation of atlas with axis and occiput. As C2-C3 had fused from both anterior and posterior side symmetrically, it did not have any effect on torticollis caused at CVJ.

Occipitalization of the atlas occurs as a result of the failure of segmentation between the fourth occipital sclerotome and the first cervical sclerotome.<sup>[4]</sup> Hemiplasia or hypoplasia of the atlas can cause a marked torticollis, which if untreated, constitutes an unacceptable deformity. Dubousset described



**Figure 5:** T2 Sagittal image sequence of MRI showing no evidence of cord compression.

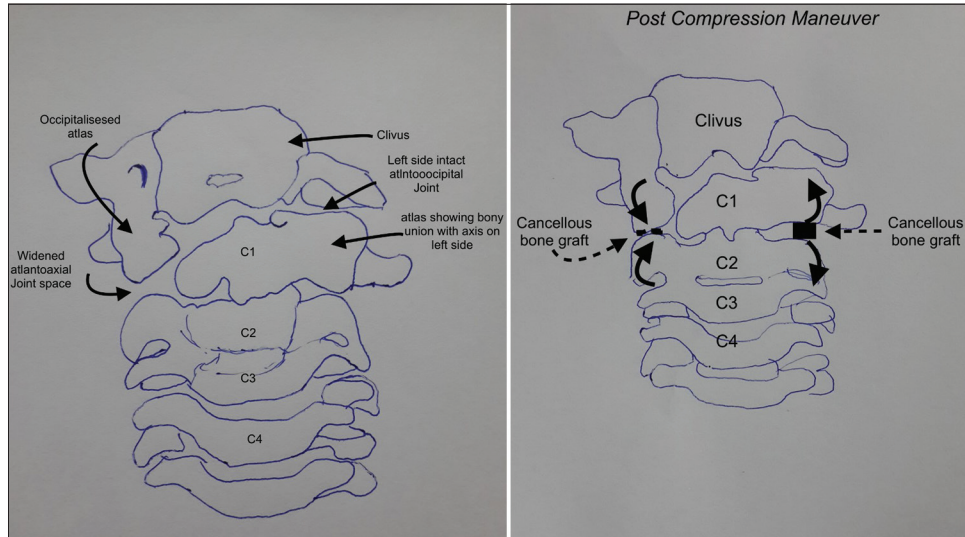


**Figure 6:** Immediate postoperative lateral and anteroposterior X-ray showing C1 C2 fixation by Goel and Harm technique.

three forms of the anomaly.<sup>[2]</sup> Type I is characterized by an isolated hemiatlas; Type II, by partial or complete aplasia of one hemiatlas with congenital fusions in the cervical spine; and Type III, by partial or complete occipitoatlantal fusion and symmetrical or asymmetrical hemiatlas aplasia with or without anomalies of the odontoid or other cervical vertebrae.

The distinguishing feature in our case was the torticollis caused by differential growth between C1 and C2. (minimal growth due to fusion of C1 and C2 on the left half and continuous growth on the right side between occipitalized C1 lateral mass and C2). Our patient underwent a C1-C2 fusion using the Goel and Harm technique to reduce the torticollis and achieve stability [Figure 6].





**Figure 7:** Schematic representation of intraoperative reduction maneuver.



**Figure 8:** Clinical photograph on postoperative day 2 showing correction of torticollis and maintained in cervicothoracic orthosis.

## CONCLUSION

Torticollis caused by anomalous/differential growth of C1-C2 vertebrae resulting in a nonrotatory subluxation is exceedingly rare. Here, a 6-year-old female with a dysplastic C1 and multiple other congenital anomalies required a C1-C2 Goel and Harm fusion to reduce the torticollis.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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## Conflicts of interest

There are no conflicts of interest.

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