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Case Report

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# Problems in Instrumentation of Syndromic Craniovertebral Junction Anomalies – Case Reports

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The aim of this study is to highlight the complications of instrumentation in the setting of syndromic craniovertebral junction (CVJ) anomalies. The records of patients with syndromic CVJ anomalies treated by this author during the period of 2012–2017 were retrospectively reviewed. Patients in whom intraoperative difficulties and complications were encountered were culled out from the database. Complications were divided into (1) technique related, (2) neural injury, (3) vascular injury, (4) instrumentation pull out/breakage, (5) inaccurate screw placement and, (6) where postoperatively, the surgeon felt an alternate surgical technique could have yielded better results. Four patients with either unexpected intraoperative difficulties or complications or in whom the technique could have been refined were identified. There were 2 patients with proatlas segmentation anomalies and 2 with Morquio's-Brailsford disease. The first patient had cage migration which necessitated a second procedure during craniovertebral realignment, the second had partial penetration of the screw into the transverse foramen, the third with bipartite atlas underwent a C1–2 fixation without a horizontal cross-connector and, the fourth had screw pull outs from the subaxial cervical spine intraoperatively during an attempted occipitocervical fusion. In children with syndromic CVJ anomalies, the surgeon should be aware of the high risk of intraoperative difficulties and complications. Potential pitfalls and the ways to avoid these complications are discussed.

**Keywords:** Bifid atlas, C2 translaminar screw, Craniovertebral junction, Dystopic os odontoideum, Morquio's-Brailsford disease, Proatlas segmentation anomaly

## INTRODUCTION

Craniovertebral junction (CVJ) is different from the rest of the spinal column because of its unique anatomical and physiological characteristics and its propensity for a multitude of developmental anomalies. In recent times, surgical management of CVJ pathologies has undergone a sea change because of improvements in the understanding of the biomechanics of the CVJ, improved imaging techniques and the availability of different types of hardware to stabilize the CVJ. However, concurrent to the increased use of hardware, the number of reported and often under-reported complications pertaining to the instrumentation of CVJ has also increased. This is especially so in patients with syndromic anomalies of the CVJ.[1,2] In this report, the author reports 4 patients in whom there were intraoperative difficulties/complications or where postoperative imaging showed inadequate screw placement or where the author in retrospect realized a better technique could have been used to optimize the outcomes.

## CASE REPORTS

This was a retrospective analysis of the author's database of syndromic CVJ anomalies from 2012 to 2017. During the study period, 17 patients with syndromic CVJ anomalies were evaluated by this author. They included: Proatlas segmentation anomalies (n=9), Morquio's-Brailsford disease (n=4), osteogenesis imperfecta (n=2), and Down syndrome (n=2). Of these 17

patients, 6 patients did not undergo surgery. Intraoperative difficulties and postoperative imaging evaluated accuracy of screw placement are discussed in the following 4 case examples.

**1. Case 1**

A 10-year-old male patient presented with progressive difficulty in walking and in using the upper extremities. Examination revealed mild torticollis and spastic quadriparesis. His plain radiographs (not shown) showed evidence of basilar invagination and atlantoaxial dislocation. Magnetic resonance imaging (MRI) showed significant compression at the cervicomedullary junction with intramedullary signal changes (Fig. 1A, B). This section computed tomography (CT) of the CVJ showed evidence of basilar invagination, atlantoaxial dislocation, hypoplastic occipital condyle on the right side with absence of the lateral mass of atlas on the right side (Fig. 2A, B). These radiological features were consistent with Proatlas segmentation anomaly.[3] The patient was planned for reduction of the basilar invagination and atlantoaxial dislocation with instrumentation through an anterior retropharyngeal approach which included the distraction of the CVJ using titanium spacers in the C1–2 joints and placing plates and screws in C1–2 anteriorly in the distracted position. During surgery, with the patient placed in traction, the C1–2 joints were exposed through a conventional anterior retropharyngeal approach, the joints were curetted using osteotome. Trial spacers were placed in facet joints on both sides. On one side, the largest available trial spacer fitted snuggly while on the opposite side even the largest available spacer was not snuggly fitting. It was presumed that the spacer would remain in place once the traction was released. Hence, on one side, the largest available titanium spacer was placed and on the other side, while the spacer was placed in the joint space, it slipped and passed posterior to the facet joint. Under fluoroscopic control, multiple attempts were made to retrieve this spacer. How-

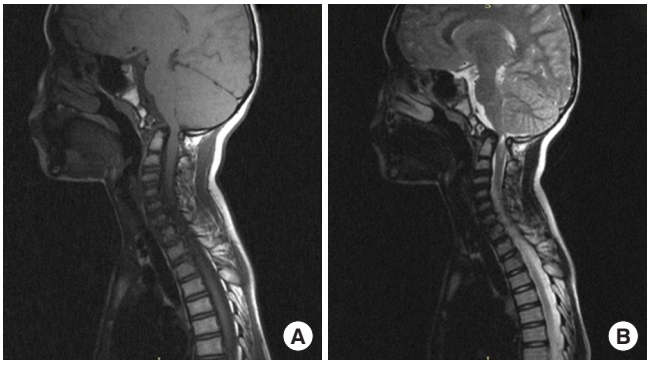


**Fig. 1.** (A, B) Sagittal T1- and T2-weighted sequences showing significant compression of the cord at the craniocervical junction with intramedullary signal changes.

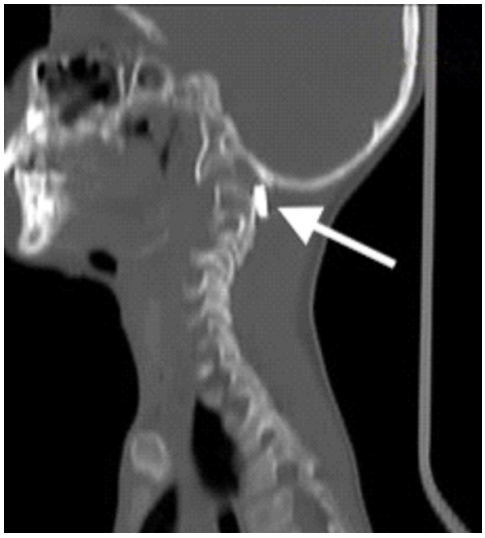

**Fig. 3.** Sagittal computed tomography showing titanium spacer displaced posterior to the facet joint (arrow).

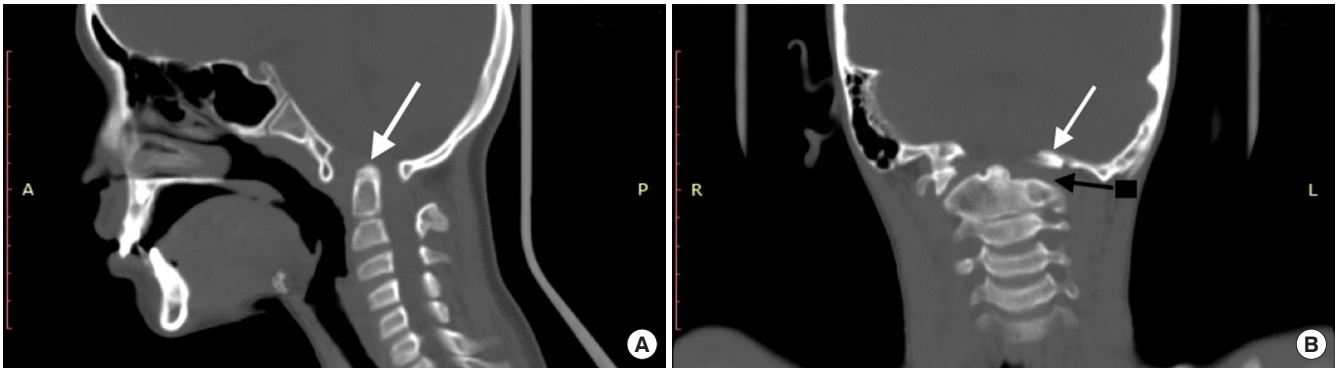


**Fig. 2.** (A) Sagittal computed tomography (CT) showing basilar invagination (white arrow) with atlantoaxial dislocation. (B) Coronal CT showing hypoplastic occipital condyle (white arrow) and aplasia of the lateral mass of atlas on one side (black arrow).


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ever, this could not be done and hence the spacer on the opposite side was removed and the wound was closed. A CT scan done immediately postoperatively showed the spacer just behind the facet joint (Fig. 3). Patient's neurological status did not worsen postoperatively. The very next day, the patient was once again placed in traction and prone position. A midline incision was made from the external occipital protuberance to the mid cervical region and on subperiosteal exposure the spacer was found in the C1–2 interspace was removed. This was followed by an occipitocervical fusion after radiological confirmation of reduction of basilar invagination and atlantoaxial dislocation. The hardware fusion was supplemented by autologous rib grafts. Postoperatively, CT showed reduction of basilar invagination and AAD (Fig. 4A, B). The patient was ambulant by the fifth postoperative day and was discharged without sequelae. The parents of this patient gave consent to publish this material.

This case illustrates that in patients with developmental anomalies, a careful study of the imaging should be done and the surgeon should remember that the facet joint space will increase under traction, especially, under anesthesia with muscle relaxants. The following lesson was learned: after finding that the largest available spacer was not fitting snuggly in the facet joint, instead of titanium spacer, an appropriately sized autologous iliac crest bone graft with supplemental instrumentation during the anterior retropharyngeal approach should have achieved the surgical goals. This could have also avoided a second surgery.

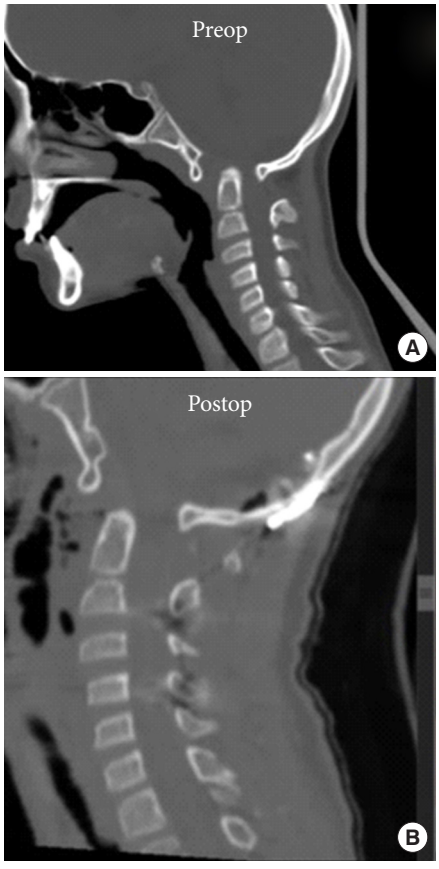


Fig. 4. Sagittal computed tomography preoperative (A) and postoperative (B) showing reduction of the basilar invagination and atlantoaxial dislocation.

**2. Case 2**

This was a 14-year-old male patient who presented with difficulty in walking and weakness of all 4 extremities following a trivial fall. Examination revealed spastic quadriparesis. His dynamic cervical radiographs showed evidence of atlantoaxial dislocation (Fig. 5). MRI showed significant compression of the cord at the cervicomedullary junction with intramedullary signal changes. His CT showed evidence of os avis or dystopic os

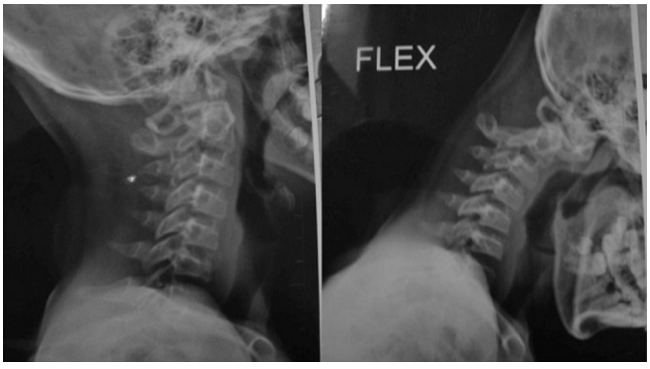


Fig. 5. Dynamic radiographs showing atlantoaxial dislocation.

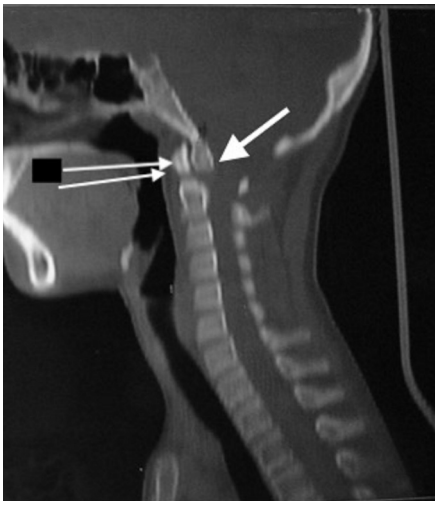

Fig. 6. Sagittal computed tomography; single white arrow points to the dystopic os odontoideum, double white arrows point to the anterior arch of atlas lying over the body of C2.

odontoideum[4] with the anterior arch of atlas directly over the body of C2 (Fig. 6). This patient underwent a classical C1 lateral mass – C2 pars screw fixation and improved with the same. His postoperative radiographs showed good screw position. However, as is customary for this author, postoperative CT evaluation showed a grade II C1 lateral mass screw violation of the transverse foramen on one side (Fig. 7) according to Bransford et al.[5] classification. As grade II violation is not considered severe enough to warrant revision surgery and as the patient had improved with no sign of vascular injury, further investigations were not done. The patient was followed for 2 years and he had resumed normal activities and had started attending school. The parents of this patient gave consent to publish this material.

The reason for lateral violation of C1 lateral mass screw is as follows: the basic physiological movement in the C1–2 region is rotation and this movement is likely to be exaggerated when there is instability. Therefore, while drilling and tapping the C1 lateral mass, it is imperative for the assistant surgeon to exert counter pressure on the opposite side C1 lateral mass. Failure to perform this simple maneuver will result in lateral rotation of the atlas with consequent screw violation, especially, laterally.

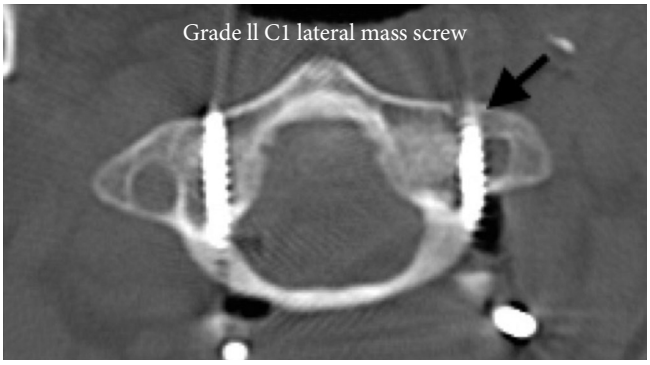


**Fig. 7.** Axial computed tomography showing Bransford grade II violation of the C1 lateral mass screw on the left side (black arrow).

### 3. Case 3

This patient was a 12-year-old male who presented with difficulty in walking. He had a history of spinal deformity since birth and short stature. On examination, this patient had all the features of Morquio's- Brailsford disease – short stature, short

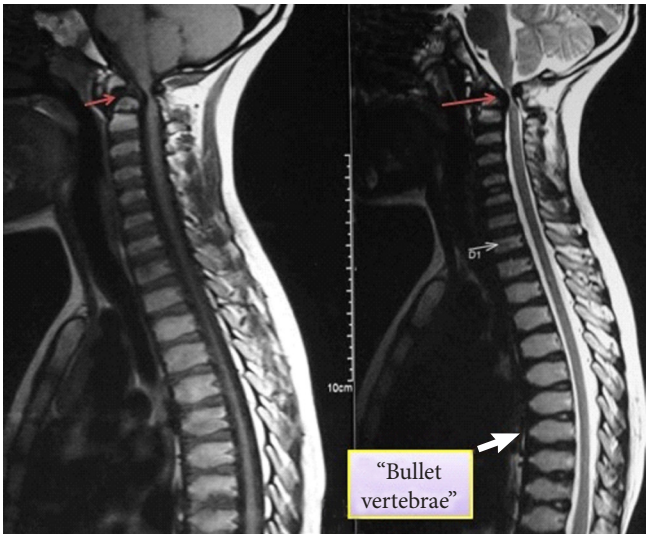


**Fig. 8.** Sagittal T1 and T2-weighted magnetic resonance imaging sequences showing significant compression of the cervicomedullary junction (red arrows) and bullet-shaped vertebrae (white arrow).

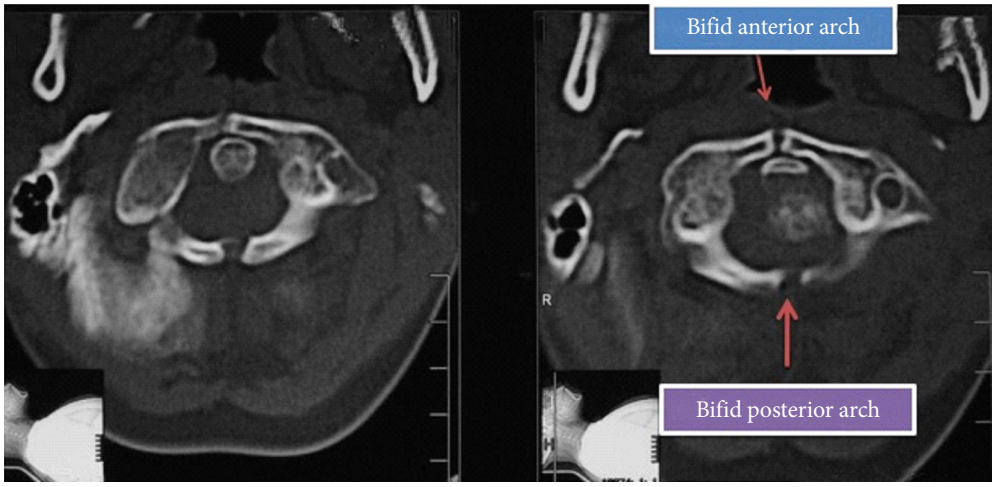


**Fig. 9.** Axial computed tomography showing bifid anterior and posterior arches of atlas.


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neck, pectus carinatum, corneal clouding, etc. His plain radiographs showed evidence of "bullet-shaped" vertebrae which is considered to be classical of this disease (not shown). His MRI of the spine showed evidence of odontoid hypoplasia, atlantoaxial dislocation with severe compression at the cervicomedullary junction and anterior beaking of the thoracic vertebrae ("bullet-shaped vertebrae") (Fig. 8). His CT showed bifid anterior and posterior arch of atlas with odontoid hypoplasia (Fig. 9). This patient underwent a routine C1 lateral mass and C2 pars screw fixation and improved with the same. His postoperative radiographs showed the implants in situ (Fig. 10). The postoperative CT scans showed accurate screw placements (Fig. 11A, B). The patient was followed for 2 years and was found to be walking without support and the implants were *in situ*. The parents of this patient gave consent to publish this material.

However, a retrospective analysis of this case showed that we had fused the C2 to an essentially bipartite atlas, the two halves of which were separate. This was not a biomechanically ideal situation. In this particular case, a horizontal cross-connector should have been placed between the 2 sides or an occipitocervical fusion should have been done. However, a revision surgery was not done as the patient had improved considerably and the parents were not willing for revision surgery in view of the good postoperative neurological status of the child.

**4. Case 4**

This 6-year-old male patient presented with a history of spinal deformity since birth, short stature and difficulty in walking. On examination, he had classical features of Morquio's disease which included short stature, extremely short neck, pectus carinatum, significantly kyphotic thoracic spine and limb deformities. His plain radiographs showed evidence of Morquio's disease in the form of "bullet-shaped vertebrae" (Fig. 12). His CT scan showed odontoid hypoplasia and both anterior and posterior arches of atlas were bifid (Fig. 13). A 3-dimensional (3D) reconstruction showed in addition to the above features, a thick bifid C2 spinous process with a robust C2 lamina (Fig. 14). His MRI showed significant compression at the cervicomedullary junction with intramedullary signal changes with evidence of kyphosis at both the cervicothoracic and lumbosacral junctions (Fig. 15). This patient was planned for Transarticular C1–2 fixation under intraoperative neuromonitoring. In view

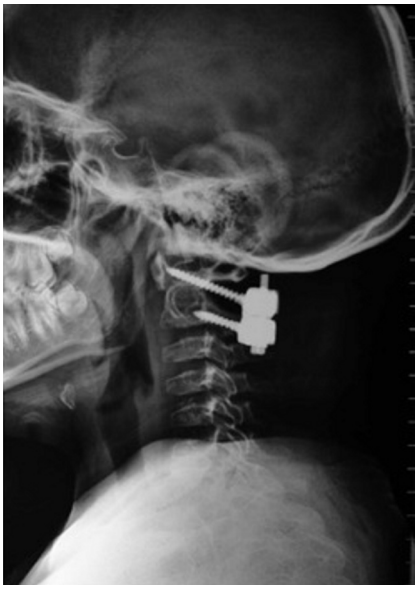

**Fig. 10.** Postoperative radiograph showing the implants *in situ*.

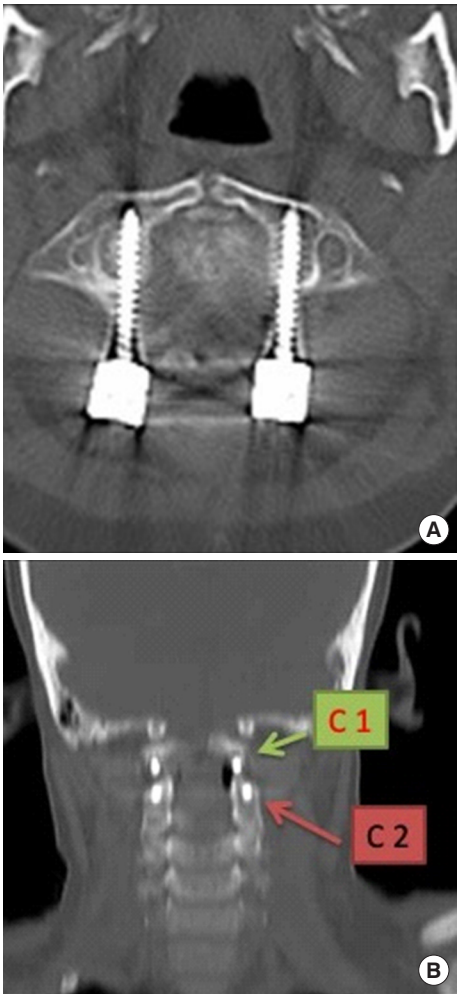


**Fig. 11.** (A) Postoperative axial computed tomography (CT) showing accurate placement of the C1 lateral mass screws. (B) Postoperative coronal CT showing accurate placement of C1 and C2 screws.

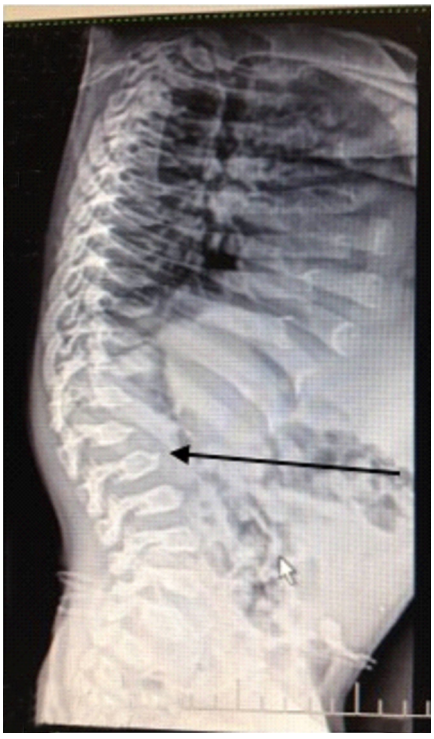

Fig. 12. Plain radiograph showing bullet-shaped vertebra (black arrow).

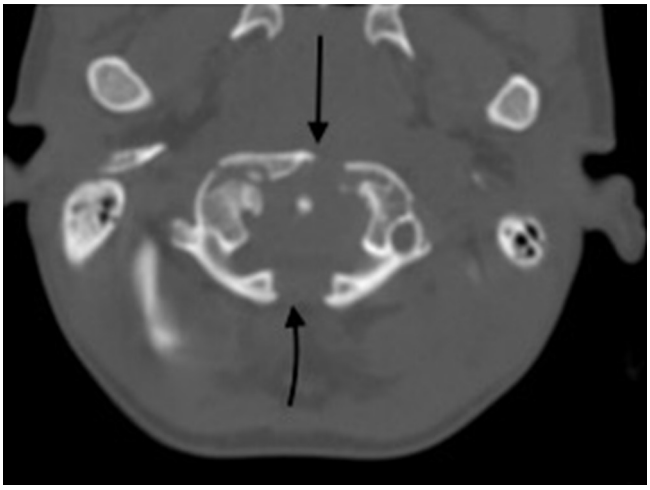

Fig. 13. Axial computed tomography showing bifid anterior arch (straight black arrow) and bifid posterior arch (curved black arrow).

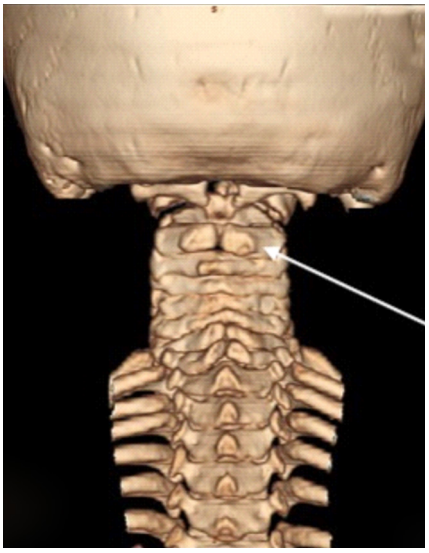

Fig. 14. Three-dimensional computed tomography showing robust C2 spinous processes and lamina. Arrow indicates C2 process.

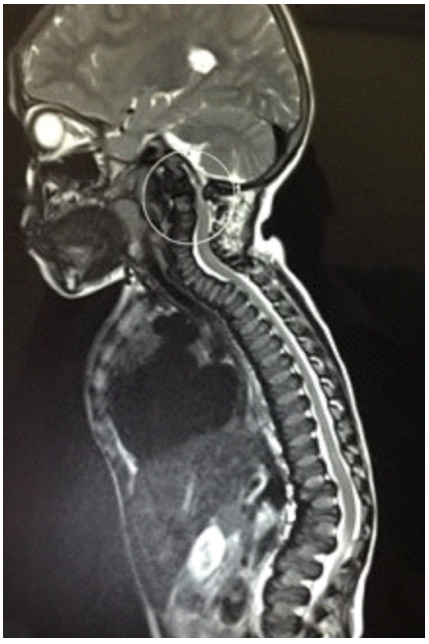

Fig. 15. Sagittal T2-weighted magnetic resonance imaging sequence showing significant compression of the cord at the cervicomedullary junction (circle) with associated cervicothoracic and thoracolumbar kyphosis.

of the extremely short neck and grossly aberrant anatomy of the CVJ, the alternate surgical options for this patient were to do a classical C1 lateral mass – C2 pars screw fixation or an occipitocervical fusion extending to the subaxial cervical spine. Preoperatively, all the potential screw insertion locations were measured in the CT to assess their ability to take a 3.5-mm screw. However, intraoperatively, transarticular C–2 fixation could not be done because the necessary angulation for transarticular screw could not be obtained because of both the extremely short neck and the exaggerated thoracic kyphosis and pectus carinatum. A classical Goel's fusion could not be done


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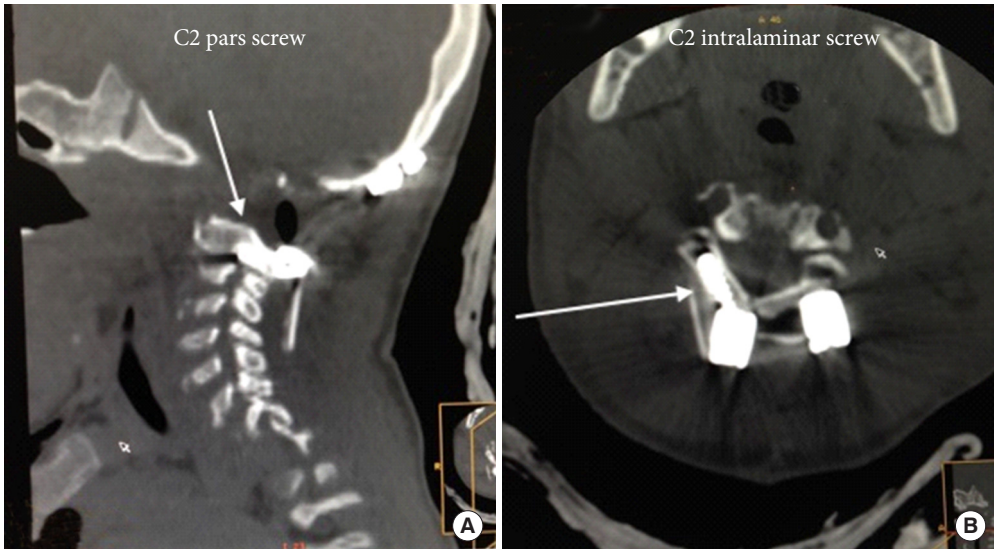


**Fig. 16.** (A) Sagittal computed tomography (CT) showing C 2 pars screw (arrow). (B) Sagittal CT showing C2 intralaminar screw (arrow).

because of the significant blood loss in this child while dissecting the C2 ganglion. Hence, an occipitocervical fusion extending up to the subaxial cervical spine was attempted. However, while tightening the rods, all the subaxial cervical lateral mass screws pulled out. Eventually, on the basis of the preoperative CT evidence of a robust bifid C2 spinous process with a robust lamina, a C2 translaminar screw was placed on the right side and a pars screw was placed on the left side.[6] Postoperative CT scan showed both the left side C2 pars screw and the right side C2 translaminar screw to be *in situ* (Fig. 16A, B). This patient improved neurologically and has been followed for more than 3 years without any worsening of the spinal alignment. The parents of this patient gave consent to publish this material.

This case shows that a surgeon planning to operate on a patient with syndromic CVJ anomaly such as severe case of Morquio's should have multiple surgical options available and a careful study of the imaging studies is required to change the strategy intraoperatively, if the need for the same arises.

## DISCUSSION

CVJ is a complex area, embryologically, anatomically and biomechanically. The complexity of this region is increased by the high propensity of this region for congenital anomalies. The problems become manifold when these developmental anomalies occur in the pediatric population. The pediatric spine continues to ossify until the ages of 8–9 years.[6] Until this age, it has increased mobility when compared to adults. Below 9 years of age, the spine is immature, between 9 and 12 years, it has an intermediate configuration, between 14 and 16 years, the spine acquires an adult like configuration. More importantly, the non-osseous components of the spine do not resemble those in adults until adolescence.[6]

There are special issues involved in instrumentation of the pediatric spine. These include: (1) diminutive osseous structures, (2) incompletely ossified structures, (3) lax ligamentous structures, (4) the presence of syndromic anomalies, (5) different injury patterns and, (6) Increased complications associated with external immobilization.[7,8] The goals of CVJ fusions are to immobilize the unstable joints, to relieve the neural compression, to resist forces applied to that joint motion, to recreate sagittal and coronal balance and, to create a stable biomechanical environment for osseous integration to take place.[7]

Earlier techniques of wire/cable or contoured rod fixation supplemented with autologous bone grafts were associated with unacceptable failure rates and perioperative morbidity.[8] However, in the past two decades CVJ fusions have undergone significant changes due to improved understanding of the biomechanics of this region, improved imaging techniques coupled with increased availability of an array of armamentarium for instrumentation of this region. While these have led to improved neurological and radiological outcomes, this, however, comes with a price viz; complications either due to technical reasons or due to instrumentation failure. As expected, complications are often reported in the surgical literature. However, complications provide an opportunity to learn from our mistakes so as

to prevent such mistakes from happening in the future.

Studies have shown that the outcomes in patients with syndromic CVJ anomalies are less when compared to those without these anomalies.[1] In a recent review of complications associated with craniocervical fusion surgery, Lall et al.[9] showed an instrumentation failure rate of around 7% with occipitocervical fusions, 6% with atlantoaxial fusion and vertebral artery injury rates varying between 1% and 4%.

**1. Lessons Learned From Case 1**

CVJ is a notoriously susceptible region for syndromic CVJ anomalies. These anomalies often include the coexistence of basilar invagination and atlantoaxial dislocation. One of the under-recognized cause of syndromic CVJ anomalies is Proatlas segmentation anomaly.[3,4,10] Patient number 1 in this series had the following anomalies: hypoplasia of the occipital condyle on the left side, aplasia of the lateral mass of atlas on the left side, basilar invagination and atlantoaxial dislocation. In this patient, the articulation between the hypoplastic occipital condyle and the C2 was wide and abnormally angled. Therefore, placement of even the largest available titanium spacer was not sufficient to distract the CVJ to reduce the odontoid invagination. The combination of a wide joint which was accentuated by the intraoperative traction along with the abnormal angulation of the joint led to the migration of the spacer posteriorly necessitating a second posterior approach. The alternative techniques that could have prevented this complication is as follows: during the initially planned anterior retropharyngeal approach to CVJ, titanium spacer should have been placed on one side where the spacer was snuggly fitting within the joint and on the other side, instead of the titanium spacer, an appropriately sized autologous iliac crest graft followed by anterior plating between C1 and C2 should have reduced the invagination thereby avoiding the complication of cage migration and the need for a second surgery.

**2. Lessons Learned From Case 2**

Patient number 2 also had a form of proatlas segmentation anomaly. In this patient, the C1 lateral mass screw had partially entered into the transverse foramen on one side, however, without any vascular injury. In retrospect, this complication may be due to 2 reasons: (1) The basic movement at the atlantoaxial joint is rotation which is likely to be aggravated when there is an atlantoaxial dislocation with lax ligamentous structures; therefore, while drilling and tapping the C1 lateral mass, it is imperative that counter pressure should be applied on the opposite side to avoid rotation of the atlas which will lead to the drill/tap being inadvertently directed laterally into the transverse foramen; (2) As shown by Jea et al.,[11] in children the entry point of the C1 lateral mass screw should be different from the adult. In adults, the centre of the C1 lateral mass corresponds to the junction of the C1 posterior arch and atlas whereas in children, this junction is off-set laterally by 2–3 mm so that in children if the same entry point used for adults is used, it will lead to lateral violation as happened in this patient. Instead, the entry point should be just medial to the junction of the posterior arch and lateral mass of C1. An effective way of identifying this entry point would be palpate the medial border of the C1 lateral mass and then proceed accordingly.

**3. Lessons Learned From Case 3**

This patient was a classical patient with Morquio's-Brailsford disease. The following features are common in this syndromic entity: odontoid hypoplasia and atlantoaxial dislocation. These patients have a high mortality rate often due to respiratory failure due to cervicomedullary compression.[12] A recent study by Solanki et al.[13] has shown that children with this entity have narrower cervical spinal canal at C1–2 region with an inverted funnel-shape and their canal diameters were smaller than age-matched controls. These patients require fixation to address the instability. In case 3, this was done by using a classical C1 lateral mass- C2 pars screw fixation. Even though, the patient improved significantly and remained so during follow-up, on retrospective analysis, this author is of the opinion, a horizontal cross-connector between the 2 sides would have been a biomechanically stable construct because the patient had a combined bifid anterior and posterior arch of atlas, a bipartite atlas. However, this was not thought of prior to the surgical procedure because of the rarity of this disease entity and unfamiliarity of the surgeon with this condition at that time.

**4. Lesson From Case 4**

This patient had a very severe form of Morquio's-Brailsford disease with short stature, extremely short neck, severe pectus carinatum, thoracolumbar kyphosis, limb abnormalities, etc. Radiologically, this patient also had odontoid hypoplasia, atlantoaxial dislocation, bifid anterior and posterior arches of atlas. In addition, the 3D reconstruction of the spine showed a robust C2 spinous process with a thick C2 lamina. In this patient, in view of the previous experience with similar cases of Morquio's disease, the following surgical options were preplanned: Option 1: transarticular C1–2 fixation with a horizontal cross-connec-

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tor between the 2 transarticular screws, 2. Option 2: a classical Goel's fusion with horizontal cross connector between the 2 sides and 3. An occipitocervical fusion extending up to the subaxial cervical spine. However, intraoperatively, all the 3 options failed. Option 1, transarticular screw fixation could not be done because of the extremely short neck and an exaggerated thoracic kyphosis; Option 2 could not be done as there was profuse bleeding from the C2 venous plexus probably because prone position hand increased the intrathoracic pressure because of the presence of pectus carinatum. As the child could not withstand significant blood loss, the third option of occipitocervical fusion was attempted. However, during tightening of the rods, the subaxial lateral mass screws pulled out in spite of achieving a bicortical purchase. Hence, C2 translaminar screw was done on one side and C2 pars screw was done on the other side. A recent study by Geck et al.[14] has shown that in children between 2 and 6 years, the C2 pars has a width of only 2.8 mm on average and hence is not generally suitable to accept a 3.5-mm screw. However, in almost all patients, the width of the C2 lamina was found to be around 4.5 mm on average and hence can safely accept a C2 translaminar screw.[14] The lesson learned from this case was that in young children apart from having multiple surgical options, C2 lamina can be a reliable screw insertion point. This was confirmed by another study by Ferri-de-Barros et al.[15]

## CONCLUSION

CVJ fusions in children, generally, is a safe procedure. However, in children with syndromic CVJ anomalies, the distorted anatomy proves to be a challenge to the surgeon performing fusions in this region. Complications can be avoided by adopting the following: a careful and meticulous study of the preoperative imaging studies, having multiple surgical options so that if one option fails, the other option(s) can be used, being familiar with the anatomical constraints of instrumentation in the paediatric age group.

## CONFLICT OF INTEREST

The authors have nothing to disclose.

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