


Craniovertebral Junction Anomalies: Changing Paradigms, Shifting Perceptions: Where Are We and Where Are We Going?



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Craniovertebral junction especially with bony anomalies like basilar invagination (BI) and atlantoaxial dislocation (AAD) have often been considered as the last bastion of the spine surgeons. Some tread with caution, other decide to veer clearly away from it. Perhaps the single most factor contributing towards this thinking has been the inability for the physicians to look at this pathology from a different window. The treatment paradigm has always been directed towards (1) classifying these pathologies as reducible or irreducible, based on flexion/extension digital X-rays; (2) treating irreducible pathologies with transoral excision of the dens followed by posterior fixation; and (3) treating reducible pathologies with wiring techniques.

For several decades, these techniques became so standard that it was felt that no other treatment was possible for these complex pathologies. Let's explore how these myths concepts were systematically exploded.

Irreducible AAD & BI Cannot Be Reduced: The concept that AAD and BI cannot be reduced is fast disappearing. Following the pioneering work of Goel et al.,^{1,2} it is now clear that there is nothing like 'irreducible' as long as there is no bone fusion (either ventral or dorsal). It is thus, best to reduce and realign the deformity intraoperatively. C1-2 fusion has been also demonstrated to be the best possible option to achieve a stable, short segment fixation. World over, this technique is now accepted. The surgical technique should of course be done with a certain degree of caution as if not careful, can lead to uncomfortable blood loss. It is also mostly advised in cases where the C1 is not fused with occiput (see below).

Distraction is the Only Movement That May Be Achieved With Spacers; No Other Motion Is Possible: Goel et al.² have shown that distraction is eminently possible which can correct BI. However, it was often seen that there were other challenges to overcome. Some of them include: (1) both BI and AAD resulting in severe superior and posterior tilt of the dens causing severe cord compression, (2) C1-2 joints becoming completely vertical, (3) anomalous vertebral artery positioned directly over the joint. All the above were situations, where the C1 arch was generally fused with the occiput. These challenges were addressed by the author with the development of the technique of distraction, compression, extension and reduction (DCER) along with all its modifications (joint remodeling, extra-articular distraction, and vertebral artery mobilization). The author also described the concept of pseudo-joints and how they may be used for reduction and realignment. We also introduced the concept of 3

axis reduction, where C2 could be aligned through reduction in multiple axis of motion.³⁻⁸

The Only Treatment for Chiari Malformation Is Foramen Magnum Decompression With or Without Duroplasty: Behari et al.⁹ in his impressive review of literature has shown significant improvement (upto 70%) may be achieved both with foramen magnum with or without duroplasty.¹⁰ He also stressed the low mortality of this procedure (<1%). This is supported ample objective imaging evidence. However, Behari et al.⁹ concluded from his extensive review that fixation is necessary in cases of Chiari associated with AAD and BI. Goel et al.^{11,12} recently have suggested that C1–2 fixations should be performed for all cases of Chiari malformations. He proposed that tonsillar herniation is secondary (and a protective and compensatory mechanism) to the subtle or gross instability which happens in all cases of Chiari malformations. Thus, an instrumented fixation without foramen magnum decompression is very effective for all Chiari malformations. While Dr. Goel is quite positive about this method of treatment, it is yet to gain universal acceptance as a standard of mode of treatment for Chiari. Well-designed randomized double blinded studies would be necessary to establish the role of such treatment modality. But the fact remains the actual cause of this ambiguous pathology remains elusive.

Is Occipital Purchase Justified: Goel¹³ has been skeptical about occipital fixation. This fact was also supported well by Sanjay Behari⁹ in his article who mentioned that occipital fixation is known to lead greater incidence of biomechanical instability (thin bone, longer lever of fixation).¹⁴ Behari also pointed out that occipital cervical fusion usually excludes the C1 arch. However, he also cautioned that in the presence of significant bleeding from paravertebral venous plexus; a very high BI, condylar hypoplasia and occipitalised atlas, where the occipital condyle and lateral mass of atlas are fused on either side, gross C1–2 rotation or vertical C1–2 joints with unilateral C1 or C2 facet hypoplasia, as well as in the presence of subaxial scoliosis, where insertion of C1–2 screws may endanger the neuraxis or the ipsilateral vertebral artery. Chandra et al.³⁻⁸ in his earlier articles has used occipital fixation in all cases of his described technique DCER both for reason mentioned above and also for the advantage of maintaining a long lever arm, which is one of the fundamental bases of DCER. He also showed that in vertical joints, posterior along fixation may be done with the technique of extra-articular distraction.

The Dens Always Dislocates Backward: Another fundamental concept of AAD has been that the dens always dislocates posteriorly. Goel¹³ has shown that it is not the dens that dislocates but the joints. When the C1 joint is dislocated posteriorly, AAD is produced (type I). When the C1 joint is dislocated forwards over C2 (type II), there may be no AAD, but the joint is still unstable, hence the spine has to be fixed. He added a third interesting category of type III stating that even in cases where there is no joint dislocation, but the patient having Chiari and 'loose joints' are observed at surgery, this was enough to diagnose bony instability and the patient should undergo bony fusion. What is interesting is that both type II and III examples shown by Dr. Goel had severe platybasia which was not commented upon. This thinking is of course a shift of paradigm and more studies will be required to justify this hypothesis.

Can the Concept C1–2 Instability Be Extended to Subaxial Cervical Spine: Goel et al.¹³ finally end their article stating that ossified posterior longitudinal ligament and even Hiramaya's disease occurs because of subaxial joint instability and a multiple long segment joint screw fixation without laminectomy is enough to treat the pathology.

Finally, I will conclude by saying that "Change is the only thing that is constant." The past decade has seen a shift in the paradigm of treatment of craniovertebral junction. Some changes like all changes have evoked intense criticism and some appreciation. But nevertheless, one cannot ignore them.

Only time will tell.

I wish the reader a happy reading of all these articles.

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Title: Rest
 Artist: Pablo Picasso
 Year: 1932

It was painted in a more surrealist and postimpressionist style than his more famous Cubist paintings, both in the use of colour and form. In *The Rest*, Picasso depicts a woman sleeping peacefully on her arms. The painting contains zones of colour with deep curved outlines. In one hand, there is a casual extension of her hand, making the limb placement highly personal as a lover's view. The woman in this painting is Marie-Thérèse Walter, one of Picasso's first mistresses. She is the subject of many of Picasso's works including slight variations of this one, also called *Le Repos*.

More information: <http://www.pablocicasso.net/rest/>

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