

## Is Chiari a “formation” or a “malformation?”

*Look deep into nature – you will understand everything  
better – Einstein*

*If you can't explain it simply, you don't understand it well  
enough. – Einstein*

*Nature has its own incorrigible ways,*

*Unseen and protective,*

*Behind a cloak of invisibility!*

*Unveil the cape,*


*Therein lays elucidation of all mystery!*

In the year 2004, we discussed that a host of soft tissue and bone malformations of the craniocervical junction that includes short neck, torticollis, Klippel–Feil abnormality, assimilation of atlas, C2–C3 fusion, and platybasia are a protective natural response to long-standing atlantoaxial instability.<sup>[1,2]</sup> Recently, we introduced the term “short head” and identified that the short head (meaning thereby-short clivus and short vertical height of posterior cranial fossa) and short neck are both simultaneous and proportional.<sup>[3,4]</sup> It is now realized that the entire spine length is vertically shortened, and the spinal canal is transversely widened. Essentially, the responsive bone and soft tissue alterations are an attempt by the Nature to reduce the length of the spinal and cranial canal or neural highway designed for the traverse of brainstem and spinal cord. Accordingly, the clivus length/posterior cranial fossa height and neck and spine height are reduced by a variety of bone fusions. We have discussed earlier, the open-close door pattern of movements of bifid posterior arch of atlas on flexion and extension of neck and its role as a natural decompressive laminectomy. The bifid arch of atlas also assists in widening the spinal canal at the level of atlas.<sup>[5]</sup> Os-odontoideum appears to be a self-bone section designed to avoid injury to the craniocervical cord on neck flexion.

Neural changes include reduction in thickness or atrophy of the spinal cord and brainstem-cerebellum. In addition, there occurs an increase in the length of the neural structures. The angulations of the brainstem and spinal cord change. The net effect is that the longer-thinner brainstem-spinal cord traverses a shorter distance, a process that minimizes the stretch of the indenting odontoid process.

This miracle of nature can be explained by simple physics. It is easier to snap a taut tense rope than a loose one. In a taut cable, the added tension adds stress, which brings the cable closer to breaking point on the application of a small force. In case, the rope is loose the momentum of the same force will get dissipated throughout the cable and will be unable to have enough impact to snap it. The bone fusions provide a stress-free traverse to the cord thus making it “loose” in the spinal canal and able to withstand a lot more injury; then, it would have been able to if it had been taut and draped over the unfused vertebral bodies.

The spinal canal increases in its anteroposterior dimension, despite the reduction of thickness of neural structures. The vertical height of the posterior fossa is reduced, but the anteroposterior dimension increases. The effects of bone fusions and soft tissue alterations make the neck prefer an extension (or hyper-extension) posture, and there is reduction in neck flexion. Platybasia seems to be a process that lifts brainstem superiorly along with the spinal cord. Both the neck and clivus manoeuvres are designed to reduce the contact of the junction of brainstem and spinal cord with the tip of the odontoid process. The divine games of the nature that come into play simultaneously to thwart off the potential or manifest dangers related to atlantoaxial instability can only be admired with awe and respect. On the basis of this understanding, it is proposed that these musculoskeletal formations should be labeled as “craniovertebral alterations” and not “craniovertebral anomalies.” An anomaly is a change or a deviation from the norm it is a peculiarity that does not fit in. An alteration is a

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change that is performed to make a fitting or appearance of an object better. If all the changes seen in the craniovertebral junction are just defending mechanisms brought in by nature to provide sanctity to the neural structures, then wouldn't we be better off calling them "alterations" rather than "anomalies?"

Neural alterations occur that have the same function of protecting the cord from dangers of odontoid process indentation. The tonsillar part of the cerebellum herniates into the spinal canal to provide a protective cushion for the craniocervical cord in an effort to protect the neural structures from getting pinched between the bones.<sup>[6,7]</sup> Simultaneously, "water" or "neuroaqua" delivers its motherly protective properties by increasing its presence inside (syringomyelia) and/or outside the spinal cord (external syrinx) and inside (syringobulbia) and/or outside the brainstem and cerebellum (external syringobulbia).<sup>[3,4,8]</sup> Such excessive presence of water has the function of making the neural structures float within an excessive pool of cerebrospinal fluid such that trauma to the neural structures at the tip of the odontoid process is minimized. Simultaneously, there is a reduction in water content within the neural axis that makes spinal cord and brainstem thinner (or atrophied) than its normal girth. The overall impact of the neural alterations is to delay or stall the symptoms related to neural compression. Observation of association of structural malformations with minimum or only marginal clinical symptoms displays the complex and protective orchestra of the creator. To respect the magnificence, it is only appropriate that the term Chiari "malformation" be changed to Chiari "formation." This will only help in relaying the fact that tonsillar herniation is not an error by the nature but is a divine intervention. Profound poetry should not be undermined by pedestrian prose. It

seems that the only anomaly in the craniovertebral junction is atlantoaxial instability. Rest of everything that happens is a response that is protective.

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