Editorial

Is central atlantoaxial instability the nodal point of pathogenesis of "idiopathic" dorsal spinal kyphoscoliosis?

"Idiopathic" dorsal spinal kyphoscoliosis (SKS) is a relatively rare clinical observation and a formidable surgical management issue. The patients usually present with spinal bony deformity or SKS as a major symptom. Other more recent-onset symptoms that force them to seek early medical treatment include breathing disturbance, sleep apnea, breathlessness on exertion, and spasticity in the limbs. The general opinion regarding the pathology of SKS is that the deformity is of a "fixed" or a "stable" nature. Abnormal bone curvatures lead to neural and lung compressive symptoms. Apart from the correction of deformity for cosmetic purposes, the aim of the surgical endeavor is to relieve the spinal cord and the lungs of compression. Some anterior, posterior, combined anterior and posterior, and lateral decompressive bone resection procedure is followed by attempts toward the correction of the deformity using multi-level screws and rod construct. Despite the number of recommended treatment forms, it may only be correct to state that no gold standard or universally accepted technique or philosophy has been identified. The controversy of treatment of SKS in the presence of syringomyelia and Chiari formation is still rife and no definite treatment paradigm or sequence of treatment has received general approval.[1]

We have analyzed the subject of craniovertebral junction instability over several years. It appears that atlantoaxial instability is an under-recognized and under-treated clinical entity. For several decades, the only parameter to diagnose atlantoaxial instability has been by evaluating the abnormal alteration of atlantodental interval on dynamic imaging. Indentation of the dural tube and neural structures opposite the odontoid process are considered to be valuable indicators that suggest abnormality of the craniovertebral junction. We recently identified that there can be atlantoaxial instability even in the absence of abnormal alteration of atlantodental interval and even when there is no dural or neural indentation by the odontoid process. We proposed an alternative classification of atlantoaxial instability on the basis of evaluation of facets of atlas and axis on lateral profile imaging in the neutral head position.[2] Type 1 atlantoaxial facetal instability was when the facet of atlas was dislocated anterior to the facet of axis. Type 2 atlantoaxial facetal instability was when the facet of atlas was dislocated posterior to the facet of axis. Type 3 atlantoaxial facetal instability was when the facets of atlas and axis were in alignment and atlantoaxial instability was identified only on direct physical manipulation of bones during the surgery. Types 2 and 3 atlantoaxial instability is labeled as central or axial as there may not be any abnormal alteration of atlantodental interval, and there may not be any dural or neural indentation in the region of the odontoid process.[3] High degree of clinical suspicion that is based on telltale radiological evidence is mandatory to be able to diagnose the central atlantoaxial instability.

Understanding of the concept of central or axial atlantoaxial instability has wide range of clinical implications. In general, while Type 1 atlantoaxial instability is associated with relatively acute clinical manifestations, central atlantoaxial instability is more often associated with chronic or long-standing clinical symptomatic progression. The natural reparative or protective maneuvering is significantly more evident in cases with central atlantoaxial instability. A range of musculoskeletal and neural alterations that are usually considered to be either embryological disorders or anomalies are actually natures endeavors that are directed on neural protection in the event of subtle, manifest or potential atlantoaxial instability.

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.JCVJS_18_19	

How to cite this article: Goel A. Is central atlantoaxial instability the nodal point of pathogenesis of "idiopathic" dorsal spinal kyphoscoliosis?. J Craniovert Jun Spine 2019;10:1-2.

The short neck, torticollis, and related bone fusions and deformities have been conventionally identified to be fixed or stable spinal abnormalities. In 2009, we identified that these musculoskeletal alterations that are grouped under the cohort of basilar invagination complex are protective secondary responses that have origin related to primary atlantoaxial instability.[4] More importantly, it was identified that atlantoaxial stabilization can result in reversal of the alterations as early as in the immediate postoperative phase. Chiari formation and syringomyelia are neural alterations in the event of atlantoaxial instability. Atlantoaxial stabilization can result in reversal of both tonsillar herniation and reduction in the size of syrinx and symptoms related to these clinical conditions in the "immediate" postoperative period.[5,6]

It was observed that in situations with chronic atlantoaxial instability in addition to the short neck, there is short head that is observed by reduction in the size of the clivus and flattening of its inclination that results in platybasia.[7] In addition, the length of entire spine is reduced or there is a short spine. The neck and cranial compartment, more evidently the posterior cranial fossa is vertically reduced in length but is transversely increased in their dimensions. The neural structures are thinned out both in the cranial and in the spinal compartments. With the increased bony canal dimensions and decreased neural volume, the cerebrospinal fluid spaces are increased either inside the spinal cord (syringomyelia) or outside the spinal cord (external syringomyelia) and inside the brainstem (syringobulbia) or outside the brainstem (external syringobulbia).[8-10] The general purpose of the secondary alterations is to make the traverse of the spinal cord lax and to float it away from the odontoid process.

We have observed that SKS can be a result of reduction in the vertical length of the spinal column. Tell-tale evidence of the presence of Chiari formation, syringomyelia, and bone abnormalities in the craniovertebral junction can be further pointers toward the basic pathology of atlantoaxial instability.[1] Like other musculoskeletal and neural alterations, atlantoaxial fixation can result in immediate postoperative improvement is SKS and relief from all symptoms that are attributed to cord and lung compression.[1]

Atul Goel

Department of Neurosurgery, Seth G.S. Medical College and K.E.M Hospital, Mumbai, Maharashtra, India

Address for correspondence: Prof. Atul Goel, Department of Neurosurgery, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai - 400 012, Maharashtra, India.
E-mail: atulgoel62@hotmail.com

REFERENCES

1. Goel A, Vutha R, Shah A, Dharurkar P, Jadhav N, Jadhav D, *et al.* Spinal kyphoscoliosis associated with chiari formation and syringomyelia 'recovery' following atlantoaxial fixation: A preliminary report and early results based on experience with 11 surgically treated cases. World Neurosurg 2019. pii: S1878-8750 (19) 30324-9.
2. Goel A. Goel's classification of atlantoaxial "facetal" dislocation. J Craniovertebr Junction Spine 2014;5:3-8.
3. Goel A. Central or axial atlantoaxial instability: Expanding understanding of craniovertebral junction. J Craniovertebr Junction Spine 2016;7:1-3.
4. Goel A, Shah A. Reversal of longstanding musculoskeletal changes in basilar invagination after surgical decompression and stabilization. J Neurosurg Spine 2009;10:220-7.
5. Goel A. Is atlantoaxial instability the cause of chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine 2015;22:116-27.
6. Goel A. Is chiari malformation nature's protective "air-bag"? Is its presence diagnostic of atlantoaxial instability? J Craniovertebr Junction Spine 2014;5:107-9.
7. Goel A. Short neck, short head, short spine, and short body height-hallmarks of basilar invagination. J Craniovertebr Junction Spine 2017;8:165-7.
8. Goel A, Jain S, Shah A. Radiological evaluation of 510 cases of basilar invagination with evidence of atlantoaxial instability (Group A basilar invagination). World Neurosurg 2018;110:533-43.
9. Goel A, Nadkarni T, Shah A, Sathe P, Patil M. Radiologic evaluation of basilar invagination without obvious atlantoaxial instability (Group B basilar invagination): Analysis based on a study of 75 patients. World Neurosurg 2016;95:375-82.
10. Goel A, Sathe P, Shah A. Atlantoaxial fixation for basilar invagination without obvious atlantoaxial instability (Group B – basilar invagination): Outcome analysis of 63 surgically treated cases. World Neurosurg 2016;95:375-82.