Short neck, short head, short spine, and short body height – Hallmarks of basilar invagination

Group A basilar invagination is characterized by an increase in atlantodental or clivodental distance that are manifest radiological evidences of atlantoaxial instability. The odontoid process migrates rostrally and indents into the cervicomedullary cord.[1] As von Torklus and Gehle described in 1962, the radiological scene appears as if the spine has migrated into the cranial cavity.[2] The clinical presentation is in the form of progressive quadriparesis over a period of time. The patients are usually young adolescents and have short neck and torticollis since childhood. In Group B basilar invagination, the entire craniovertebral junction complex is migrated rostrally.[1] The basilar invagination is significantly more marked but the odontoid process does not transgress the Wackenheim's clivus line, and the atlantodental interval is not abnormally altered. The symptoms are longstanding and progressive, and the affected patient age group is "adults." The musculoskeletal affections such as the short neck, torticollis, and Klippel-Feil abnormalities are remarkably more pronounced.

The general understanding has been that development of short neck is a result of embryogenic dysgenesis, wherein the formation of the vertebral spinal column is incomplete and inappropriate. Indentation of the odontoid process into the cervicomedullary cord is considered to be a direct physical consequence of shortening of the neck size. We earlier speculated that short neck is not a pathological or abnormal event but is a protective natural response and is a direct consequence of long-standing atlantoaxial instability.[3,4] While an acute atlantoaxial instability results in neck pain, muscle spasm and stiffness that result in restriction of movements, chronic, or long-standing atlantoaxial dislocation results in shortening of the neck.

Short neck is usually associated with torticollis. Although spasm of the muscles of nape of the neck is not as prominent as in acute atlantoaxial instability related to trauma, it is remarkable and present. Similarly, neck pain is not as severe as in an acute situation but is frequently associated. The overall effect of shortening of the neck is restriction of flexion movements and exaggeration of extension movement of the neck.

The presence of short neck is usually noticed early in childhood and even during infancy. The exact time of development of atlantoaxial instability during the physical growth of the person cannot be pointed but is speculated to be during late fetal life or early infancy, during the time when structural development, growth, and organogenesis is still in progress. Injury to the neck during fetal delivery, poor practices of delivery of a fetus by forceps, and protein-calorie malnutrition may be the incriminating factors that result in atlantoaxial instability and its related consequences. Disproportionately high incidence in relatively poor population of the world lends credence to such hypothesis.

In general, basilar invagination was considered to be a "fixed" atlantoaxial instability.[5] In the year 2004, for the first time in literature, we proposed that atlantoaxial instability in Group A basilar invagination is not only not fixed, but it is pathologically mobile and more importantly, it can be reduced by direct surgical manipulation of the facets.[1] There is Type A atlantoaxial facet instability in such cases, wherein the facet of atlas is dislocated anterior to the facet of the axis.[6] The alignment of the facets of atlas and axis mimicked vertebral bodies of lumbosacral bones in cases with spondylolisthesis. The proposed concept speculated that Group A basilar invagination is a result of "listhesis" of a facet of atlas over the facet of the axis as a result of the instability of the region.[7] The treatment that was essentially focused on decompression of bone now shifted to stabilization and fixation and aimed at

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craniovertebral junction realignment.[1] The technique involved opening of the atlantoaxial joint, denuding of the articular cartilage, stuffing of bone graft within the articular cavity, and manipulation of facets that would result in their realignment.[1,8,9]

Short neck is associated with a host of musculoskeletal abnormalities of the neck and skull. Torticollis is a frequent association, wherein the odontoid process is eccentrically poised. Abnormal bone fusions of the cervical spine include assimilation of atlas, C2-3 fusion, and Klippel-Feil abnormalities.

We recently identified that short neck is associated with simultaneous and proportionate decrease in vertical height of the skull in general and posterior cranial fossa in particular.[10] Shortening of clivus length and its rostral migration results in platybasia. We labeled shortening of clivus size as "shortening of the head."[10] Shortening of the head is generally associated with chronic or long-standing atlantoaxial instability. More frequently, Type B and Type C facet malalignment is associated with chronic atlantoaxial instability, usually identified in Group B basilar invagination. Long-standing "subtle but present and relentlessly persistent" instability provides an opportunity for natural "protective" process to mature.

Apart from musculoskeletal alterations, we identified that in the face of atlantoaxial instability, there is extensive neural reformation that is aimed at protection from cord injury due to the indentation of the odontoid process into the cervicomedullary cord and repeated "micro-injuries" related to instability. Chiari formation, wherein the cerebellar tonsils herniate into the spine, appears to provide a soft cushion to the cord in the event of manifest or potential atlantoaxial instability and prevents direct pinching of critical neural structures between bones.[11,12]

It was identified that in the presence of vertical reduction of the head and neck size, there is an anteroposterior increase in both the spinal canal and the posterior cranial fossa dimensions. The entire spinal column is increased in its transverse dimension but is reduced in its vertical height. The gross result is reduction in spinal height or "short spine."[10]

The neural structures, both in the posterior cranial fossa and in the spinal canal are reduced in their girth or are "atrophic" in their nature.[4,10] Atrophy of the cerebellum and brainstem in the skull and spinal cord in the spine in the presence of an increase in the anteroposterior and transverse dimensions of the bony housing results in a space that is occupied by cerebrospinal fluid (CSF). CSF can be present inside the spinal cord (syringomyelia), outside the spinal cord (external syringomyelia), or both inside and outside the spinal cord. In the similar fashion, increase in the amount of CSF can be present within the cerebellum and manifested as cerebellar atrophy, within the brainstem (syringobulbia) or outside the cerebellum and around the brainstem (external syringobulbia).

Although the neural structures are "atrophic" and filled with or surrounded by CSF, the vertical length of the neural column is not decreased or may even be a shade increased when compared to normal. The presence of normal or longer length of neural structures within a shorter spinal column that is filled in with an excessive quantity of CSF allows the neural structures to drift away from the indenting odontoid process and float as freely as possible. Stretch injury to the neural structures from the indenting odontoid process is thus minimized.

Reversibility of musculoskeletal and neural formations after atlantoaxial fixation, sometimes in the immediate postoperative period, lends support to the speculations. The clinical recovery in symptoms can be "magical."

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REFERENCES

1. Goel A. Treatment of basilar invagination by atlantoaxial joint distraction and direct lateral mass fixation. J Neurosurg Spine 2004;1:281-6.
2. Von Torklus D, Gehle W. The Upper Cervical Spine: Regional Anatomy, Pathology, and Traumatology. A Systematic Radiological Atlas and Textbook. New York: Grune & Stratton; 1972. p. 1-98.
3. Goel A, Shah A. Reversal of longstanding musculoskeletal changes in basilar invagination after surgical decompression and stabilization. J Neurosurg Spine 2009;10:220-7.
4. Goel A, Nadkarni T, Shah A, Sathe P, Patil M. Radiological evaluation of basilar invagination without obvious atlantoaxial instability (Group B-basilar invagination): An analysis based on a study of 75 patients. World Neurosurg 2016;95:375-82.
5. Goel A, Bhatjiwale M, Desai K. Basilar invagination: A study based on 190 surgically treated patients. J Neurosurg 1998;88:962-8.
6. Goel A. Goel's classification of atlantoaxial "facetal" dislocation. J Craniovertebr Junction Spine 2014;5:3-8.
7. Kothari M, Goel A. Transatlantic odonto-occipital listhesis: The so-called basilar invagination. Neurol India 2007;55:6-7.
8. Goel A, Laheri VK. Plate and screw fixation for atlanto-axial dislocation. (Technical report). Acta Neurochir (Wien) 1994;129:47-53.

9. Goel A, Desai K, Muzumdar D. Atlantoaxial fixation using plate and screw method: A report of 160 treated patients. Neurosurgery 2002;51:1351-7.
10. Goel A, Jain S, Shah A. Radiological evaluation of 510 cases of basilar invagination with evidence of atlantoaxial instability (Group A). World Neurosurg. [In press].
11. 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.
12. 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.