Editorial

Degenerative arthritis of the craniovertebral junction

A review of related publications in the literature suggests that cervical spinal degeneration has more often been identified and surgically treated in the lower cervical spine, less frequently in the upper cervical spine, and only rarely in the craniovertebral junction. Our observations identify that degenerative changes in the craniovertebral junction are relatively frequent and are probably underdiagnosed and undertreated.[1]

Our recent articles relate the term "degeneration" to "instability." "Vertical" spinal instability (VSI) related to weakness of muscles that partake in standing human position and facilitate major motor activities is the nodal point of genesis, propulsion, and propagation of process of degeneration.[2] This understanding is in variance with the existing concept that disc space reduction or "old" age-related loss of water content of the disc form the basis of spinal degeneration. VSI results in telescoping of the spinal segments and initially or primarily leads to listhesis of rostral facet over caudal facet, a process termed as retrolisthesis of facets.[2] All the known so-called "pathological" alterations in spinal degeneration such as buckling of the intervertebral ligaments that include ligamentum flavum and posterior longitudinal ligament, osteophyte formation, and disc space reduction that are ultimately associated with reduction in the spinal canal and intervertebral foraminal height are secondary alterations in the face of VSI. Our studies in the craniovertebral junction and spine conclude that all the "secondary" issues related to spinal instability have a natural protective role and more importantly are reversible following stabilization of the affected spinal segment.[3]

The relevant concept is that more mobile a spinal segment, more likely it is to develop instability and spinal degeneration. Lower cervical spinal segments (C4-5, C5-6, and C6-7) and lower lumbar spinal segments (L4-5 and L5-S1) have been more often associated with degeneration. These "junctional" spinal segments are the more mobile regions of the spine and are consequently more likely to develop instability. Our concept is that it is not neural compression or deformation, but it is spinal instability that is the cause of symptoms.[4] Consequently, there can be symptoms without any radiological evidence of neural compression.[5] Both clinical features and radiological guides direct toward the unstable spinal segments. Identification of instability in segments adjoining those suggested by clinical and radiological guides can be done by manual manipulation of bones during surgery. Ignoring such instability appears to be a cause for the so-called "adjacent segment disease" often encountered in cases operated for degenerative spinal issues.[5,6]

The process of spinal "degeneration" or spinal "instability" is more often chronic or longstanding in nature. Secondary natural alterations are more prominent in cases where the instability is subtle. In cases where the secondary natural processes are prominent, the symptoms are relatively minor or subtle and longstanding but are relentlessly progressive. It appears that once the instability begins, the process of spinal degeneration becomes progressive. However, if exercising and muscle strengthening therapy can restore the effectiveness of the affected muscles, the instability can probably be reversed.

Stability and mobility are hallmarks of the craniovertebral junction. The occipitoatlantal joint is the most stable, and the atlantoaxial joint is the most mobile joint of the body. Degenerative alteration in the occipitoatlantal joint is uncommon, and it is probably related to the strength of the joint and to the limited range of movements. Considering the wide-ranging and circumferential mobility in the atlantoaxial articulation, the probability of complex instability or degeneration is highest when compared to the rest of the spinal segments.

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.jcvjs_142_21	

Submitted: 16-Nov-21 **Accepted:** 22-Nov-21
Published: 11-Dec-21

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

How to cite this article: Goel A. Degenerative arthritis of the craniovertebral junction. J Craniovert Jun Spine 2021;12:323-6.

Probably related to longstanding and subtle nature of instability, a range of natural "protective" measures, and the presence of significant buffer of cerebrospinal fluid in the vicinity, manifestation by clinical symptoms and neurological deficits can be delayed. Pain and chronic spasms of the neck muscles, both natural processes aim to limit the range of the neck movements and prevent or delay the impact of instability. Reduction in the atlantoaxial joint space, buckling of the posterior longitudinal ligament, retroodontoid thickening of ligaments or pseudotumor formation, para-articular osteophyte formation, ossification in the vicinity of the ligaments of the region (apical and transverse), facetal bone destruction or hypertrophy, and several similar events are indicators of the presence of atlantoaxial instability, are secondary in nature, are naturally protective and have the potential of reversal following atlantoaxial stabilization[1] [Figures 1-5].

Chronic atlantoaxial instability can be associated with short neck, short head, short spine, torticollis, dorsal spinal kyphoscoliosis, basilar invagination, Chiari formation, syringomyelia, Klippel-Feil abnormality, platybasia, C2-C3 fusion, assimilation of the atlas, bifid arches of the atlas and axis, and a range of other secondary alteration.[7-16] Subaxial spinal instability is frequently observed in cases with chronic atlantoaxial instability that is identified to be "degenerative" in nature.[17] The presence of disc bulges at the C2-C3 spinal segment is a frequent observation and is more often a secondary alteration to primary atlantoaxial instability.[18] Disc bulges indenting into the spinal cord at the lower cervical spinal segments can be observed. It is critical to evaluate if the changes in the subaxial cervical spine are secondary events related to atlantoaxial instability or are primary in nature.

On the basis of alignment of the facets of atlas and axis on lateral profile imaging and direct observation of instability by manual manipulation of bones of the region, we classified

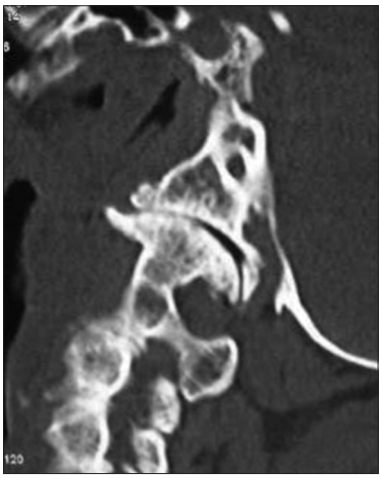

Figure 1: Sagittal cut of computed tomography scan showing the C1-C2 articulation. There is a reduction in the C1-C2 joint space and the presence of para-articular osteophytes

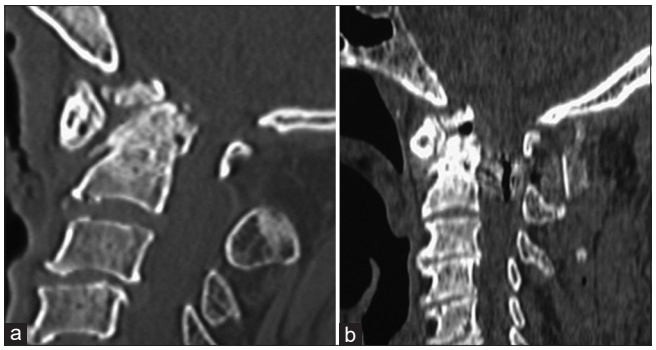

Figure 3: (a) Preoperative sagittal computed tomography image showing atlantoaxial instability and the craniovertebral junction degenerative arthritis. Note the calcification in the apical and transverse ligaments. (b) Postoperative computed tomography image showing the fixation

Figure 2: Coronal computed tomography image showing reduction in the C1-C2 joint space and degenerative changes in the facets of C1 and C2

Figure 4: Sagittal computed tomography image showing basilar invagination. There is ossification of the posterior longitudinal ligament at the C2-C3 level and ossification in the region of the apical ligament

Figure 5: (a) T2-weighted sagittal magnetic resonance imaging showing retroodontoid thickening of the ligaments or pseudotumor formation. (b) Sagittal computed tomography scan showing craniovertebral junction degenerative arthritis. (c) Sagittal computed tomography scan with cut through the facets showing reduction in joint space with facet arthritis

atlantoaxial instability into three types.[19] Type 1 atlantoaxial instability is when the facet of atlas is dislocated anterior to the facet of axis. In such cases, atlantodental interval is increased, and there is neural and dural compression by the odontoid process. In such cases, atlantoaxial instability is more often of acute or relatively acute nature, symptoms are more pronounced and secondary natural alterations are not frequent or prominent. Type 2 atlantoaxial instability is when the facet of atlas is dislocated posterior to the facet of axis. Type 3 atlantoaxial instability is when the facets are in alignment. In both Types 2 and 3 atlantoaxial instability, atlantodental interval may not be abnormally altered, and there might not be any neural or dural compression by the odontoid process. Such types of atlantoaxial instability are labeled as "central" or "axial" atlantoaxial instability (CAAD). CAAD is identified by telltale clinical and radiological evidence and by the identification of musculoskeletal and neural alterations and is confirmed by direct manipulation of bones during surgery.[20] Degenerative alterations in the craniovertebral junction are usually associated with CAAD. Symptoms are "longstanding" and secondary alterations in the craniovertebral junction and subaxial cervical spine can be prominent. Compression of the dural or neural tube is not the hallmark. Identification of the presence of atlantoaxial instability and its appropriate treatment can result in dramatic clinical recovery.

For the sake of understanding of the participant and for formulating a surgical strategy, we divided the craniovertebral junction degeneration into three types.

Type 1 atlantoaxial degeneration: the instability in such cases is essentially limited to the atlantoaxial joint. There can be telltale evidence of instability in the form of reduction in the joint space, osteophyte formation, retroodontoid "pseudotumor, buckling of the atlantoaxial ligaments, osteophyte formation in the region, facetal destruction or hypertrophy, evidence of pseudo-fusion of the atlantoaxial facetal articulation, and similar such alterations [Figures 1-5]. The affection of subaxial spinal segments is not prominent. Surgical treatment in such cases is atlantoaxial fixation with the aim to achieve atlantoaxial arthrodesis.

Type 2 atlantoaxial degeneration: in such cases, in addition to atlantoaxial instability and its consequences, there is secondary affection of subaxial spinal segments. More often C2-C3 disc bulges and evidence of adjoining spinal cord compression. The surgical treatment in such cases is atlantoaxial stabilization. Inclusion of the subaxial cervical spine in the fixation construct is not necessary.

Type 3 atlantoaxial degeneration: In patients with Type 3 atlantoaxial degeneration, there is multisegmental subaxial spinal degeneration. The patients are generally disabled and might have evidence of severe myelopathy. The surgical treatment in such cases is atlantoaxial and multisegmental subaxial spinal fixation. More recently, we identified a modified technique of atlantoaxial fixation that included the section of muscles attached to C2 spinous process and trans-articular C2-C3 fixation. The technique retained the rotatory movements executed by muscles attached to the transverse process of atlas and obliterated flexion-extension movements executed by the muscles attached to the C2-spinous process. We have advocated that all these patients need "only-fixation" of the affected spinal segments and despite radiological evidence of spinal cord compression, "decompression" by the resection of bone and soft tissues can have negative consequences.

Atul Goel[1,2,3]

[1]Department of Neurosurgery, Lilavati Hospital and Research Center, [2]Department of Neurosurgery, R. N Cooper Hospital and Medical College, [3]Department of Neurosurgery, Bombay Hospital Institute of Medical Sciences, Mumbai, Maharashtra, India

Address for correspondence: Dr. Atul Goel,
Department of Neurosurgery, Lilavati Hospital and Research Center,
Bandra, Mumbai, Maharashtra, India.
E-mail: atulgoel62@hotmail.com

REFERENCES

1. Goel A, Shah A, Gupta SR. Craniovertebral instability due to degenerative osteoarthritis of the atlantoaxial joints: Analysis of the management of 108 cases. J Neurosurg Spine 2010;12:592-601.
2. Goel A. Vertical facetal instability: Is it the point of genesis of spinal spondylotic disease? J Craniovertebr Junction Spine 2015;6:47-8.
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. Not neural deformation or compression but instability is the cause of symptoms in degenerative spinal disease. J Craniovertebr Junction Spine 2014;5:141-2.
5. Goel A. Beyond radiological imaging: Direct observation and manual physical evaluation of spinal instability. J Craniovertebr Junction Spine 2017;8:88-90.
6. Goel A, Dandpat S, Shah A, Rai S, Vutha R. Muscle weakness-related spinal instability is the cause of cervical spinal degeneration and spinal stabilization is the treatment: An experience with 215 cases surgically treated over 7 years. World Neurosurg 2020;140:614-21.
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 2017;99:164-70.
11. Goel A, Patil A, Shah A, Dandpat S, Rai S, Ranjan S. Os Odontoideum: Analysis of 190 surgically treated cases. World Neurosurg 2020;134:e512-23.
12. Goel A, Vutha R, Shah A, Dharurkar P, Jadhav N, Jadhav D. 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;125:e937-46.
13. 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.
14. Shah A, Sathe P, Patil M, Goel A. Treatment of "idiopathic" syrinx by atlantoaxial fixation: Report of an experience with nine cases. J Craniovertebr Junction Spine 2017;8:15-21.
15. Goel A, Nadkarni T, Shah A, Ramdasi R, Patni N. Bifid anterior and posterior arches of atlas: Surgical implication and analysis of 70 cases. Neurosurgery 2015;77:296-305.
16. Goel A, Darji H, Shah A, Prasad A, Hawaldar A. Retro-odontoid and retro-C2 body pseudotumor, pannus, and/or cyst. A study based on analysis of 63 cases. World Neurosurg 2021;151:e170-7.
17. Goel A. Role of subaxial spinal and atlantoaxial instability in multisegmental cervical spondylotic myelopathy. Acta Neurochir Suppl 2019;125:71-8.
18. Goel A. C2-3 fusion, C3-4 cord compression and C1-2 posterior facetal instability: An evaluation of treatment strategy based on four surgically treated cases. Asian Spine J 2016;10:430-5.
19. Goel A. Goel's classification of atlantoaxial 'facetal' dislocation. J Craniovertebr Junction Spine 2014;5:15-9.
20. Goel A. A review of a new clinical entity of 'central atlantoaxial instability': Expanding horizons of craniovertebral junction surgery. Neurospine 2019;16:186-94.