Facetal alignment: Basis of an alternative Goel’s classification of basilar invagination

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Basilar invagination is a complex subject. Understanding of the pathogenic issues is crucial to evaluate and treat this problem. Whilst a successful treatment can lead to quick neurological recovery and an excellent post-operative and long term outcome, complications can be devastating and life threatening.

We recently proposed an alternative concept in the management of basilar invagination and analyzed the clinical course and management issues in approximately 1000 surgically treated cases. Our concept in management of basilar invagination is based on the understanding that atlantoaxial dislocation or instability is the primary cause of all types of basilar invagination, irrespective of whether it is demonstrated or not on dynamic imaging.[1,2] Long-standing instability results in a variety of structural alterations in the soft tissues and bones that ultimately culminates in basilar invagination. Short neck, low hairline, web-shaped neck muscles, torticollis, reduction in the range of neck movements, and several such physical variations have been described as hallmarks of basilar invagination. A number of bone fusion deformities and platybasia have also been recorded. Neck pain, muscle spasms, and restriction of neck movements are frequently noted and suggest instability of the region. The instability of the region is manifested by alterations in alignment of the facets.

The musculoskeletal and neural alterations observed in cases with basilar invagination will depend on the severity or type of atlantoaxial dislocation. In more acute form of basilar invagination, the facetal dislocation is usually of Type A [Figure 1].[3] In this type of basilar invagination, the odontoid process migrates superiorly and posteriorly and indents into the brainstem and there is resultant neural compromise relatively early in the clinical course. Such form of dislocation is usually seen in younger patients, the clinical symptoms are relatively acute and severe. The structural bone and soft tissue including neural malformations are not so prominent in this group of patients. There is a progressive slippage of the facet of atlas over the axis secondary to this malalignment, a process similar to spondylolisthesis in the lumbosacral spine.[4]

The more chronic form of basilar invagination is associated with Type B or Type C atlantoaxial facetal dislocation. In Type B atlantoaxial facetal dislocation, the facet of atlas is dislocated posterior to the facet of axis in lateral profile or sagittal imaging [Figure 2]. In Type C atlantoaxial dislocation, the facets of atlas and axis are in alignment and the dislocation or instability is identified only during surgery [Figure 3]. We labeled such dislocation as “central” facetal dislocation. In Types B and C dislocation, the alignment of the odontoid process with the anterior arch of atlas is not remarkably affected and the atlantodental interval remains normal. In both these forms of dislocation the atlantodental interval is not increased and the neural compromise is not early in clinical course. The symptoms in such cases are subtle and long-standing. However, the longstanding nature of instability results in marked structural alterations in the bone and soft tissue structures. Chiari malformation and syringomyelia are components of such secondary alterations.

**CLINICAL FEATURES**

Trauma of varying severity was a noteworthy precipitating factor in cases of basilar invagination with Type A atlantoaxial facetal dislocation.[5] Trauma seldom plays any major role in precipitating...
The pyramidal symptoms formed a dominant component in basilar invagination associated with Type A atlantoaxial facetal dislocation. Kinesthetic sensations were affected in 55% cases. Spinothalamic dysfunction was less frequent (36%). Neck pain as a major presenting symptom was in 80% cases. Torticollis was present in 45% cases. The analysis of radiological and clinical features in cases with basilar invagination having Type A atlantoaxial facetal dislocation suggests that the symptoms and signs were a result of brainstem compression by the odontoid process. The presentation was relatively acute in such cases. Symptoms in patients having Types B and C atlantoaxial facetal dislocation were longstanding and slow progressive. In such cases the onset of symptoms and their evolution were insidious.

**Associated clinical features**

Mere inspection of the patients with basilar invagination was of diagnostic value in the majority of cases, more particularly in cases having Type B and Type C atlantoaxial dislocation. Short neck and torticollis were more frequently encountered in cases with Type B and Type C dislocation.
Goel’s Clinical grading system
Depending on the extent of neurological disability, the patients were divided into five clinical grades. Grade 1: Independent and normally functioning, Grade 2: Walks on own, but needs minimal support/help to carry out routine household activities, Grade 3: Walks with minimal support and requires help to carry out household activities, Grade 4: Walks with heavy support and unable to carry out household activities, Grade 5: Unable
Goel: Goel's classification of basilar invagination

Radiological criteria

Chamberlain's line: Basilar invagination was diagnosed when the tip of the odontoid process was at least 2 mm above the Chamberlain's line. Measurement of Chamberlain's line on lateral sagittal reconstruction pictures of CT scan and sagittal MRI were seen to be reliable and accurate. The analysis of basilar invagination on the basis of Chamberlain's line suggested that the basilar invagination is much more severe in cases with Type B and C atlantoaxial facetal dislocation than in Type A atlantoaxial dislocation.

Distance between odontoid tip to the pontomedullary junction: The distance of the tip of the odontoid from the pontomedullary junction, as observed on MRI is a useful index to define the status of the bones at the craniovertebral junction. The distance was markedly reduced in cases with Type B and C atlantoaxial dislocation while it was relatively large in cases with Type A atlantoaxial dislocation.

Wackenheim's clival line: Wackenheim's clival line is a line drawn along the clivus. The tip of the odontoid process was significantly superior to the Wackenheim's clival line in patients with Type A atlantoaxial dislocation. In cases with Type B and
Type C atlantoaxial dislocation, the relationship of the tip of the odontoid process and the lower end of the clivus and the atlanto-dental and clivo-dental interval remained relatively normal. In a majority of these cases, the tip of the odontoid process remained below the Wackenheim’s clival line and McRae’s line of foramen magnum. The basilar invagination thus resulted from the rostral positioning of the plane of the foramen magnum in relation to the brainstem [Figure 1].

**Platybasia:** A line is drawn along the anterior skull base. The angle of this line to the clivus is referred to as the basal angle. Reduction of the basal angle is referred to as platybasia.

**Omega angle:** Although not frequently used, the Omega angle or the angulation of the odontoid process from the vertical as described by Klaus was found to be a useful guide. Goel described a modified omega angle as the measurement of the angle from the vertical as determined by the flexion and angulation of the odontoid and the cervical spine during movement of the neck. A line was drawn traversing through the center of the base of the axis parallel to the line of the hard palate. The line of the hard palate was unaffected by the relative movement of the head and the cervical spine due to movement of the neck in these “fixed” craniovertebral anomalies. Facial hypoplasia or hard palate abnormality was not seen in any case in this series and did not affect the measurements. The Omega angle depicted the direction of displacement of the odontoid process. The Omega angle was severely reduced in patients with Type A atlantoaxial dislocation while it was much larger in patients having Type B and Type C atlantoaxial dislocation. The reduction in the Omega angle depicted that in patients with Type A dislocation the odontoid process had tilted towards the horizontal and was posteriorly angulated while it was near vertical and superiorly migrated in patients having Type B and Type C atlantoaxial dislocation.

**Brainstem girth:** The effective brainstem girth measured on MRI was a useful additional parameter. Whilst the brainstem girth was markedly reduced in patients having Type A atlantoaxial dislocation, the girth was only marginally affected or unaffected in cases having Type B and Type C atlantoaxial dislocation indicating thereby that there was no direct brainstem compression as a result of the odontoid process in the latter group.

**Occipitalization of the atlas:** Occipitalization of the atlas associated with basilar invagination was noted first by Rakitansky [cited by Grawitz 1880] and has since been referred to frequently. Many authors have regarded assimilation as a characteristic feature of basilar invagination. The assimilation of atlas can be partial or incomplete. Occipitalization of atlas was identified to be an indicator of long-standing instability of the region and was identified in all types of basilar invagination.

**Neck size:** Measurement of craniovertebral height can be done using a modification of Klaus’s posterior fossa height index. The cervical height was measured from the tip of the odontoid process to the mid-point of the base of the C7 vertebral body. The parameter of direct physical measurement of the neck length from inion to the tip of the C-7 spinous process can be useful. The reduction in the neck length was more marked in patients having Type B and C atlantoaxial facetal dislocation. **Cervical lordosis** was evaluated with a modification of the Klaus omega angle and a modified omega angle. (2)

### SURGICAL MANAGEMENT

**Atlantoaxial fixation for all cases of Basilar Invagination**

The conventional form of treatment of basilar invagination with Type A atlantoaxial dislocation is a transoral decompression that is followed by posterior occipitocervical fixation. However, the long-term clinical outcome following the twin operation of transoral decompression followed by posterior stabilization was seen to be inferior to the clinical outcome following surgery that involves atlantoaxial fixation and craniovertebral realignment without any bone, dural or neural decompression. An attempt can be made to reduce basilar invagination by performing occipitocervical fixation following institution of cervical traction. However, all our cases treated in this manner subsequently needed transoral decompression as firm fixation, reduction of the basilar invagination and of atlantoaxial dislocation could not be sustained by the implant. The technique of craniovertebral realignment by wide removal of atlantoaxial joint capsule and articular cartilage by drilling and subsequent distraction of the joint by manual manipulation provided a unique opportunity to achieve fixation and to obtain reduction of the basilar invagination and of atlantoaxial dislocation.

**Surgical Technique of atlantoaxial fixation and craniovertebral realignment**

The primary aim of surgery is atlantoaxial fixation and craniovertebral stabilization in all cases having basilar invagination. The procedure of stabilization by our technique by itself will result in craniovertebral realignment, more particularly in cases with Type A atlantoaxial dislocation. The steps of introduction of bone graft within the joint are mandatory in all cases. Introduction of spacers within the joint cavity is necessary when it appears that the spacers will provide additional stability to the region. Essentially, spacers are placed to provide enhanced stability to the region rather than being aimed at reducing the basilar invagination.

The exposure of the atlantoaxial joint in cases with basilar invagination is significantly more difficult and technically challenging when compared to a normally aligned atlantoaxial joint encountered during the treatment of post-traumatic instability. The joint is rostral in location and the microscope needs to be appropriately angled. The difficulty in exposure is more in cases having Type B and Type C dislocation. In all cases of basilar invagination, the atlantoaxial facet joints are widely exposed on both sides after sectioning of the large C2 ganglion. The joint capsule is excised and the articular cartilage is widely removed using microdrill. The joint on both sides are distracted using an osteotome. The flat edge of the
osteotome is introduced into the joint and it is then turned vertical to effect distraction. The status of the dislocation and of basilar invagination is evaluated by intraoperative radiographic control. Corticocancellous bone graft harvested from the iliac crest is stuffed into the joint in small pieces. Specially designed Titanium spacers are used in selected cases as strut graft and impacted into the joints to provide additional distraction and stability. Subsequent fixation of the joint with the help of interfacet monoaxial (or polyaxial) screws and a metal plate (or rod) provided a biomechanically firm fixation and sustained distraction. Holes in the titanium metal spacer provided space for bone fusion. The fixation was seen to be strong enough to sustain the vertical, transverse and rotatory strains of the most mobile region of the spine. Post-operatively the traction is discontinued and the patient is placed in a four-post hard cervical collar for 3 months and all his physical activities involving the neck are restrained during the period.

**Reversibility of musculoskeletal changes following surgery**[11]

A number of bone and soft tissue anomalies are associated with basilar invagination. These include short neck, torticollis, platybasia, cervical vertebral body fusion (Klippel — Feil abnormality) including assimilation of atlas, spondylotic spinal changes and restriction of neck movements. A number of these abnormalities were seen to be reversible following decompression and stabilization of the region. Our recent studies suggested that even neural abnormalities like Chiari malformation and syringomyelia are also reversible following surgical procedures that involve stabilization of the atlantoaxial joint. Considering that several physical features associated with this group of basilar invagination are reversible, it appears that the pathogenesis in such cases may be more due to mechanical factors rather than congenital causes or embryological dysgenesis. The common teaching on the subject is that the short neck and torticollis are a result of embryological dysgenesis and effectively result in indentation of the odontoid process into the cervicomедullary cord. However, it appears that basilar invagination is a result of atlantoaxial instability that is the primary event and all the physical alterations and bony abnormalities, including the short neck and torticollis are secondary natural protective responses that aim to reduce the stretch of the cord over the indenting odontoid process. Chiari malformation is Nature’s “air-bag” that protects the critical neural structures from getting pinched between bones. Pain, restriction of neck movements and hyperlordosis of the most mobile region of the spine are secondary natural protective responses that aim to reduce the stretch of the cord over the indenting odontoid process. Reduction of the disc spaces, osteophytes formation, incomplete and complete cervical fusions, and alterations in the craniospinal and cervical angulations appear to be directly related to the reduction in neck length.[17-20] The reduction in the disc-space height and fusions are more prominently seen in the upper cervical vertebrae, like assimilation of atlas and C2-3 vertebral fusions. It appears that cervical fusions and assimilation of the atlas may be related to long-standing and progressive reduction in the disc-space height.

**Occipital fixation and Foramen magnum decompression:** As our experience in the subject is growing, it appears that atlantoaxial stabilization is mandatory and necessary in all cases with basilar invagination. Inclusion of the occipital bone in the fusion construct is not only unnecessary, it negatively affects strength of the implant and severely restricts the neck mobility. Foramen magnum decompression is not necessary and has negative effects in the long run.

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