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# Atlanto-Axial Instability and its Management

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## INTRODUCTION

The treatment of patients with atlantoaxial instability is a surgical challenge and achieving a successful outcome for these patients is gratifying. The complications of surgery, however, are potentially lethal. Various methods of fixation have been described and used successfully in the treatment of patients with atlantoaxial instability. The techniques of craniovertebral fixation evolved during the 20th century as the anatomy and biomechanics of the craniovertebral region became clearer.

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## SURGICAL ANATOMY

The course of the vertebral artery,extensive venous channels in the region, relationships of C2 ganglion, atlantoaxial joint and the dural tube and the normal and abnormal alignments of the region has to be understood prior to surgery on the basis of all the available radiological information. Our belief is that experience with cadaveric dissections is mandatory for successful conduct of the surgery.

The C1 and C2 vertebrae are called 'atypical vertebrae' and have unusual shape and architecture and a complex and important vertebral artery relationship. Injury to the artery during surgery can lead to catastrophic intra-operative bleeding and compromise to the blood flow can lead to unpredictable neurological deficits, which will depend on the adequacy of blood flow from the contralateral vertebral artery.

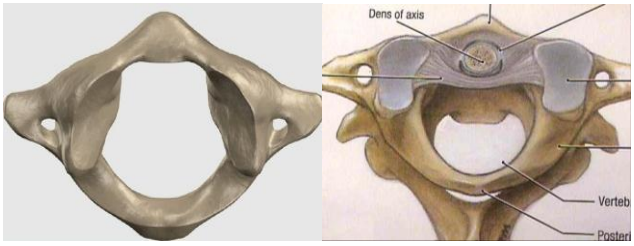



**Fig. 1: Atlas and the Axis**

The vertebral artery adopts a serpentine course in relationship to the craniovertebral region. The artery has multiple loops and an intimate relationship with the atlas and axis bones. We observed a wide variability of the course of the artery in our specimens. The shape, size and location of the vertebral artery groove on the inferior aspect of the superior articular facet of the C2 and over the posterior arch of the atlas had wide variations. The vertebral artery during its entire course is covered with a large plexus of veins. The venous plexuses are the largest in the region lateral to the C1-2 joint.

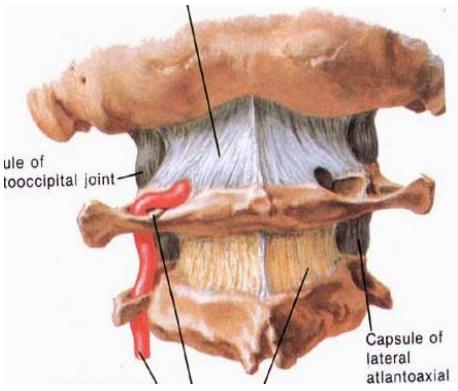



**Fig 2: course of the vertebral artery at the CVJ**

After a relatively linear ascent of the vertebral artery in the foramen transversarium of C6-to C3, the artery makes a loop medially towards an anteriorly placed superior articular facet of the C2 vertebra, making a deep groove on its inferior surface. The extent of medial

extension of the loop varies. The distance of the artery from the midline of the vertebral body of C2 as would be observed during a transoral surgical procedure is on an average 12 mm[3]The vertebral artery loops away from the midline underneath the superior articular facet of the C2.

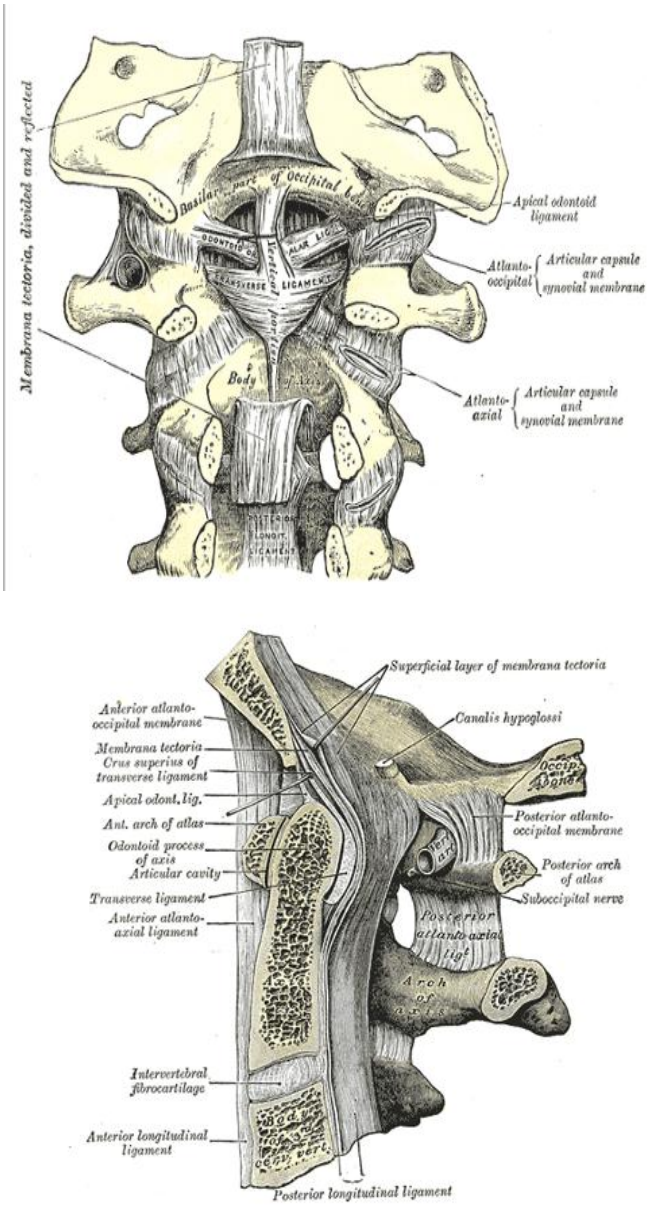



**Fig 3: CVJ ligamentous anatomy**

The dens or the odontoid process is flanked by two large, superior facets, extending laterally on to the adjoining pars-interarticularis and articulating with the inferior atlantal facets. Superior facet of C2 vertebra differs from the facets of all other vertebrae in two important characters, which make this region prone to vertebral artery injury during screw fixation. First is that the superior facet of C2 is present in proximity to the body when compared to other facets which are located in proximity to the lamina. The second is that the vertebral artery foramen is present partially or completely in the inferior aspect of the superior facet of C2, while in other cervical vertebrae, vertebral artery foramen is located entirely in relationship with the transverse process. Unlike superior facets of all other vertebrae, they do not form a pillar with the inferior facets, being considerably anterior to these. The pedicle of the C2 vertebra is relatively small. The course of the vertebral artery in relationship to the inferior aspect of the superior articular facet of the C2 makes its susceptible to injury during transarticular and inter-articular screw implantation techniques. It was observed that the screw implantation in the superior facet of the C2 vertebra has to be sharply medial and directed towards the anterior tubercle of the C1 for trans-articular fixation and towards the vertebral body of C2 for interarticular fixation. As discussed in our previous paper on this subject, the pars-interarticularis can be divided into nine quadrants[4]The superior and medial compartment can be used for inter-articular technique of screw implantation. The average distance of the artery from the ganglion was 7.5 mm. It suggests that the dissection around the lateral end of the ganglion should be carefully done and under vision.

The inferior facet of the atlas is almost circular in most of the vertebrae without any significant difference in the mean anteroposterior and transverse (15 mm) dimensions. The thickness of the inferior facet under the posterior arch of the atlas is on an average 3.5 mm. The thickness of the posterior arch of the atlas separating the vertebral artery groove from the inferior facet is about 3.5 mm.

## HISTORY OF SURGICAL INTERVENTIONS

Attempts at surgical stabilization of C1 and C2 from a posterior approach date to 1910, when Mixter and Osgood described using heavy silk thread to wire the spinous processes of C1 and C2 together.[1]

The use of posterior cervical wiring of the lamina of C1 and C2 dates to 1939 in a report by Gallie.[2] Brooks and Jenkins offered an alternative method of posterior C1-C2 laminar wiring in 1978[3]Dickman and Sonntag, et al further modified the posterior wiring technique in 1991[4]We will further explore each of these techniques in this review article.

In the 1980's interlaminar clamps[5]were popularized as an alternative method of posterior C1-C2 fixation.

Posterior screw fixation utilizing C1-C2 transarticular screws and C1 lateral mass screws with C2 pars screws were the final two alternative methods of posterior C1-C2 fixation. The C1 lateral mass screw with C2 pedicle screw construct was initially created with plates and screws by Goel et al in the 1980's.[6],[7] The method has recently gained popularity and a variety of instrumentation is now available for application with this technique[26]

Transoral instrumentation has also been described for stabilization of unstable craniovertebral region, but has received only limited support.

## CLINICAL FEATURES

The presentation of atlantoaxial dislocation may range from minor axial neck pain to death. Approximately 50% of patients present with neck pain and/or neck movement restriction, 70% with weakness and/or numbness, and 90% with pyramidal signs.[33], [36], [37] Other preoperative clinical presentations include sphincter disturbances, lower cranial nerve dysfunction, and respiratory distress. Other serious sequelae include myelopathy, respiratory failure, vertebral artery dissection, neurologic compromise, and rarely quadriplegia or death if left untreated.[38], [39], [40] The differential diagnosis of atlantoaxial dislocation includes torticollis, atlantoaxial rotary fixation, and odontoid fractures without atlantoaxial dislocation.

Most cases of atlantoaxial dislocation appear in the adolescent population and should be considered in a child with inability or unwillingness to turn their head when history and physical examination are inconsistent with torticollis.[41] Typically, congenital atlantoaxial dislocation presents in childhood as chronically progressive spinal canal compression and associated with neurologic and respiratory symptoms.[42] Less commonly, a patient with a congenital yet asymptomatic unstable atlantoaxial joint may have an acute presentation after minor acute trauma. In one study, 9 of 15 patients who presented after acute trauma with congenital atlantoaxial dislocation presented with quadriplegia.[41] The symptoms may also present as relapsing and remitting episodes due to the motion of day-to-day life.[26], [38], [42]

Atlantoaxial dislocation presenting in adulthood has historically been the result of rheumatoid arthritis, with some patients experiencing few symptoms and others experiencing gross instability and neurologic compromise. Clinical findings can be confounded by the severity of systemic involvement. Radiographic changes are common, and the prevalence of neurologic injury is relatively low.[30] More recent series have found a decreased prevalence of atlantoaxial dislocation among patients with rheumatoid arthritis, especially with various immunoregulatory medications showing promising outcomes preventing cervical spine lesions in patients with rheumatoid arthritis.[43], [44] One study that investigated a total of 904 patients with a diagnosis of atlantoaxial dislocation over a period of 12 years showed that only 36 patients (4%) had rheumatoid arthritis.[25] Regardless of the etiology, the clinical consequences of atlantoaxial dislocation have clear potential for neurologic compromise. Therefore, a careful history is critical to identify symptoms of cervical disease, and physical and neurologic examinations should be performed in all at-risk patients.

## CAUSES

### *Traumatic Causes*

A purely traumatic atlantoaxial dislocation in the absence of another predisposing risk factor is extremely rare. A literature review by Venkatesan et al in 2012 found only 12 adult case reports.[8] Traumatic atlantoaxial dislocation is due to forced displacement of the neck resulting in disruption of the transverse ligament. Rarely, injury of the transverse ligament can also involve simultaneous disruption of the alar and apical ligaments. Such injuries may be seen in head injuries, which may occur during tackling in football or rugby. In these ligamentous dislocations, the atlas will lose articulation with the dens, and the anterior atlantal arch may translate completely superiorly and posteriorly with significant damage to the ligaments[9]

Traumatic osseous injuries may also result in atlantoaxial instability. Type II odontoid fractures occur at the base between the transverse ligament and body of C2 body.[10], [11], [12] Type II fractures are the most common odontoid fracture and the only type normally associated with atlantoaxial dislocation.[10] Traditionally believed to be a congenital anomaly, os odontoideum may in fact be caused by an early traumatic injury in which the odontoid is separated completely from the axis and then heals to resemble a separate ossicle. The resulting condition predisposes patients to dislocation.[13], [14]

### *Congenital Causes*

Certain congenital conditions are associated with craniocervical region abnormalities that predispose these populations to developing atlantoaxial dislocation.[15] One particularly well-studied condition is Down syndrome. Down syndrome (trisomy 21) is the most common inherited chromosomal disorder.[16]Predisposing sequela include hypermobility and instability caused by ligamentous laxity and osseous abnormalities.[17], [18] resulting in an increased incidence of atlantoaxial dislocation (15 to 20%).[19] Ligamentous laxity may be due to an inflammatory process or to an intrinsic defect in collagen fibers that form ligaments.[20] Alternatively, chronic environmental trauma seen in these population superimposed on congenitally weakened ligaments can lead to spondylitis with subsequent atlantoaxial dislocation.[18]

Skeletal dysplasias are a heterogeneous group of disorders with resulting abnormal cartilage and bone formation, growth, and remodeling. Craniocervical junction abnormalities, atlantoaxial dislocation, and kyphoscoliotic deformities are among the common spinal problems that are found in certain skeletal dysplasias.[16] Examples of skeletal dysplasias that have been linked with atlantoaxial dislocation include Goldenhar syndrome, spondyloepiphyseal dysplasia, and Morquio syndrome.[16]

Goldenhar syndrome is a clinically heterogeneous disorder characterized by spinal defects, hemifacial microsomia, and epibulbar dermoid appendages. An increased frequency of hypoplasia of the dens with atlantoaxial instability has been reported in children with Goldenhar syndrome.[16] In one series of eight children with Goldenhar syndrome, three had atlantoaxial instability greater than 5 mm with upward migration of the odontoid process. Two of the patients had atlantoaxial instability greater than 7 mm and required surgical treatment.[21]

Spondyloepiphyseal dysplasia encompasses several disorders characterized by abnormal growth of the spinal vertebrae and epiphysis. Typically, individuals with

spondyloepiphyseal dysplasia have short-trunk dwarfism, with short proximal and middle limbs but normal-sized hands and feet.[16] Atlantoaxial instability associated with hypoplasia of the dens or ligamentous laxity is the most common spinal manifestation of congenital spondyloepiphyseal dysplasia in children. An increased incidence, as high as 35% of cervical myelopathy in children with congenital spondyloepiphyseal dysplasia, may be attributable to atlantoaxial dislocation.[22]

Mucopolysaccharidosis type IV (Morquio syndrome) is an autosomal recessive lysosomal storage disease characterized by an inability to metabolize keratan sulfate. These patients often have normal growth and development during the first 2 years of life with abnormalities progressing rapidly after 2 years of life.[16]Atlantoaxial dislocation has been identified in up to 42 to 90% of cases of Morquio syndrome,[23], [24] likely due to an increased incidence of odontoid dysplasia (hypoplasia, aplasia, or os odontoideum) and increased ligamental laxity.[23]

## Congenital conditions associated with atlantoaxial dislocation

Congenital osseous abnormalities, in the absence of an underlying genetic abnormality, are also frequently associated with atlantoaxial dislocation. Wang et al documented a rate of 18% among 904 patients in the largest published series to date.[25] Failures in segmentation, such as occipitalized atlas, C2–C3 fusion, and asymmetrical occiput–C3 facet joints, can predispose to dislocation.[14], [26] Occipitalization of the atlas occurs when the inferior neural arch and superior spinal sclerotome form improperly. Patients with occipitalization of the atlas may also have asymmetrical occipitoatlantal facet joints because the same sclerotomal segments contribute to development of the facets.

## INFLAMMATORY CAUSES

Another notable population that is disproportionately affected by atlantoaxial dislocation is chronic rheumatoid arthritis patients, particularly adults. The atlantoaxial joint is frequently affected in rheumatoid arthritis with studies showing incidence rates ranging from 23 to 86% of patients with rheumatoid arthritis.[27], [28],[29] The rates we present here are based on traditional rates. Although it is not well documented, it is likely the rate has declined in conjunction with advancements in medical treatment of rheumatoid arthritis (i.e., biologics and immunoregulatory medications). The cervical spine often becomes involved early in the course of rheumatoid arthritis, leading to three different patterns of instability: atlantoaxial dislocation, atlantoaxial impaction, and subaxial subluxation.[30] Chronic systemic inflammation in these patients leads to chronic synovitis resulting in bony erosion and ligamentous laxity that may result in instability and atlantoaxial dislocation. In patients with rheumatoid arthritis, anterior atlantoaxial dislocation is the most frequently occurring deformity due to laxity of the ligamentous restraints.[3], [31] ,[32]

## RESULTING PATHOLOGY

When the upper cervical spine is destabilized and sagittal balance is compromised, the lower cervical spine compensates, which may lead to subaxial pathology and deformities.[1], [33] ,[34], [35] When atlantoaxial dislocation causes diminished lordosis at the C0–C2 segment, the subaxial cervical region compensates with increased lordosis to maintain balance.[33] ,[35] Some patients with end-stage changes can develop kyphosis at the occipitoaxial segment together with extreme hyperlordosis subaxially, resulting in swan neck deformity.

## BIOMECHANICS

The opposing facetal surfaces of the normal atlanto-axial joints (AAJs) are almost flat and parallel. These are horizontal in the sagittal plane and mildly sloping downwards mediolaterally in the coronal plane. Therefore they can have unrestricted movements in various planes There are 6 degrees of freedom of movements at these joints: (1) angular motion (flexion and extension); (2) rotation (right and left); and (3) linear motion and translation (anterior and posterior, right and left). However, anterior movement of the axis vertebra (C2) is restricted by the anterior arch of *Atlas* (C1) when odontoid is normal. Posterior movement is restricted by cruciate ligament (largely by transverse ligament). Rotatory movement is restricted by the alar ligaments. These AAJs form the most mobile segment of the whole spine because of which they have more chances of developing instability. Dislocation due to instability of these joints leads to compression of vital cervicomedullary neural structures that lie underneath and results in neurological disability. A minor trauma may precipitate quadriplegia, respiratory arrest and even death.

***Diagnosis:***

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 for over a century. A number of bone fusion deformities and platybasia have also been recorded. Neck pain, muscle spasms, and restriction of neck movements are frequently noted, suggesting instability of the region.

## RADIOLOGICAL CRITERIA

***Chamberlain's line***

Basilar invagination was diagnosed when the tip of the odontoid process was at least 2 mm above Chamberlain's line.[13] Measurement of Chamberlain's line on lateral sagittal reconstruction pictures of CT scan and sagittal MRI was seen to be reliable and accurate. The analysis of basilar invagination into two groups on the basis of Chamberlain's line suggested that the basilar invagination is much more severe in Group B than in Group A.

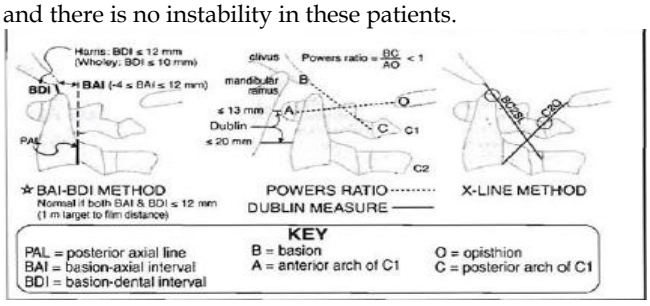
***Distance from odontoid tip to the pontomedullary junction***

The distance from the tip of the odontoid to the pontomedullary junction, as observed on MRI, was seen to

be a useful index to define the reduction of the posterior cranial fossa bone size.[13] The distance was markedly reduced in Group B patients while it was relatively large in Group A patients.

***Atlanto-dental or clivo-dental interval***

In Group A patients, it was seen that the odontoid process migrated superiorly and posteriorly into the foramen magnum and distanced itself from the anterior arch of the atlas and the inferior end of the clivus. As judged from the atlanto-dental or clivo-dental interval, there was an element of 'fixed' atlantoaxial dislocation in these patients. Actual mobility of the atlantoaxial joint on flexion and extension of the neck can be demonstrated only rarely. In our recent study, we identified patients in whom there was 'vertical mobile and reducible atlantoaxial dislocation', wherein there was basilar invagination when the neck was flexed, and the alignment was normal when the head was in extended position. Although such mobility is only rarely identified, it does indicate the need for dynamic flexion-extension studies to preoperatively assess the craniovertebral instability.[16] In Group B the alignment of the odontoid process with the anterior arch of the atlas and with the inferior aspect of the clivus remains normal and there is no instability in these patients.



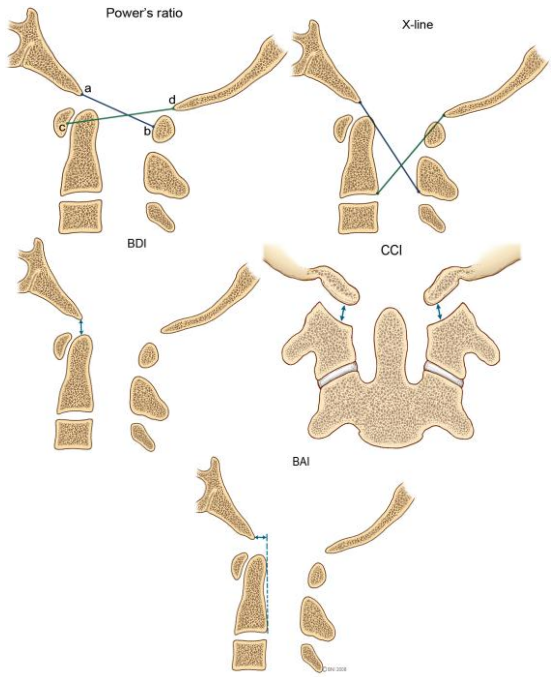




**Fig 4: Different measures of atlanto-axial instability**

***Wackenheim's clival line***

The tip of the odontoid process was significantly superior to Wackenheim's clival line in Group A patients. In Group B patients, 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 the patients, the tip of the odontoid process remained below Wackenheim's clival line[15] and McRae's line of foramen magnum.[14] The basilar invagination thus resulted from the rostral positioning of the plane of the foramen magnum in relation to the brainstem [Figure 2].

***Platybasia***

The association of platybasia and basilar invagination is well known. Chamberlain, in his initial study referred to both these entities as being synonymous.[13]Subsequently, various authors have questioned the clinical significance of platybasia and have considered it to be of anthropological significance only. The superior position of the odontoid process was associated with a more horizontal angulation and shortening of the clivus.[17] Klaus also identified two groups of basilar invagination on the basis of the clival line of Wackenheim.[15],[18] He noted that in basilar invagination associated with platybasia, the tip of the odontoid process almost never reaches the Wackenheim clival line, while in a steeply shelving or normal clivus, the line from the dens often reaches or even over-shadows it. Platybasia was seen in both the groups but was relatively less in number and severity in Group A. From the study of Group B patients, it appears that platybasia was as important as invagination of the odontoid process in causing the anterior concavity of the brainstem and in reducing the volume of the posterior fossa. Marin-Padilla[19] concluded from their study that the Chiari-like deformities reflect the effects of clival and occipital molding, which act mainly anteriorly. Platybasia did not directly result in any neurological symptoms, but it participated with basilar invagination in critically reducing the posterior cranial fossa volume. In our recent study,[1] we demonstrated reversal of platybasia in Group A patients following the craniovertebral realignment surgery that involved distraction of facets of atlas and axis.

***Posterior cranial fossa volume***

The Klaus' height index[18] measured on the MRI was seen to be much more accurate than the conventional measurements based on plain X-rays. The tentorium could be clearly identified on MRI and the distance of the tip of the odontoid from the line of the tentorium indicated the height of the posterior cranial fossa. On the basis of Klaus' index, the posterior fossa height was found to be markedly reduced in Group B patients while it was only moderately affected in Group A patients.

***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.[18] We described a modified Omega angle as the measurement of the angle from the vertical was affected by the flexion and extension of the neck.[11] A line was drawn traversing through the centre 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 during the movement of the neck in these 'fixed' craniovertebral anomalies [Figure 4]. 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 Group A patients while it was much larger in Group B patients. The reduction in the Omega angle in Group A patients depicted that the odontoid process had tilted towards the horizontal and was posteriorly angulated, while it was near vertical and superiorly migrated in Group B patients.

***Brainstem girth***

The effective brainstem girth measured on MRI was a useful additional parameter.[11] Whilst the brainstem girth was markedly reduced in Group A patients, the girth was only marginally affected or unaffected in Group B patients indicating thereby that there was no direct brainstem compression as a result of the odontoid process in the latter group.

The anterior concavity of the brainstem was smooth in curvature in Group B patients while it was acute in Group A patients, the angle being formed by the tip of the odontoid process. In Group A, the brainstem distortion was directly a result of indentation of the odontoid process.[11],[20]

***Occipitalization of the atlas***

The association of occipitalization of the atlas with basilar invagination was noted first by Rakitansky [cited by Grawitz 1880[8]] and has since been referred to frequently.[13],[14],[21],[22] Many authors have regarded assimilation as a characteristic feature of basilar invagination. The assimilation of atlas can be partial or incomplete.

Klippel-Feil bone anomaly refers to the triad of observations of extensive cervical vertebral fusions, low hairline, restrictions of neck movements and a short neck.

***Neck size***

Measurement of craniovertebral height can be done using a modification of Klaus's posterior fossa height index.[1],[23],[24] The cervical height was measured from the tip of the odontoid process to the mid-point of the base of the C5-vertebral body [Figure 5]. The C-5 vertebral body base was used as the inferior limit for measuring the craniovertebral and cervical heights because in several available investigations the C6 and C7 vertebrae were not clearly delineated.

On direct physical measurement, neck length was observed to increase in the majority of patients. The parameter of measurement of neck length from inion to the tip of the C7 spinous process can be useful.

## APPROACHES FOR DIAGNOSIS:

There are several different approaches to diagnosing atlantoaxial dislocation; however, no consensus exists. Atlantoaxial dislocation can be defined with radiographic measurements of atlantoaxial joint articulation using the atlantodental interval (ADI). The ADI is a small slitlike space between the posterior aspect of the anterior atlas ring and the anterior aspect of the odontoid process. Flexion and extension radiographs of the neck allow for the measurement of the ADI and to determine whether the atlantoaxial joint reduces itself in these positions.[38]

The ADI is measured from a line projected superiorly along the anterior border to the axis body to the anterior arch of the atlas (Figs. 1 and and22).[45] The ADI is normally constant in distance during movement of the head and generally does not exceed 3 mm for adults and 5 mm for children.[33], [45], [46] In this article, we define atlantoaxial dislocation as ADI greater than 3 mm in adults older than 18 years of age and greater than 5 mm in children. The majority (70%) of clinical atlantoaxial dislocation presentations are due to anterior dislocations.[41] Anterior dislocation increases the ADI, decreasing the space available for the spinal cord, which is measured from the posterior aspect of the dens to the anterior aspect of the posterior atlantal ring.[41]A decrease in the space available for the spinal cord increases the risk of spinal cord compression as well as neurologic sequelae. Of note, the space available for the spinal cord of less than 14 mm predicts the development of paralysis[47], [48] and has been shown to correlate with severity of paralysis.[48]

Neutral (left), extension (center), and flexion (right) lateral X-rays showing the atlantodental interval (ADI) anterior to the odontoid process and the space available for spinal cord posteriorly. The ADI is above the average for adults of 3 mm...

Neutral and dynamic cervical radiographs are often used to diagnose atlantoaxial instability and dislocation, but the diagnostic sensitivity is low. Some studies report decreased false-negative diagnostic rate when using dynamic radiography, with some studies also reporting the necessity for magnetic resonance imaging (MRI) to obtain information on soft tissues, joints, and the spinal cord with high sensitivity and specificity and to possibly provide early warnings signs of instability.[41] When evaluating trauma patients with concerns about cervical spine involvement, it is important to most efficiently and effectively evaluate the cervical spine to avoid missing the injury. The NEXUS study group reported reliability of a standard three-view imaging of the cervical spine that included an open-mouth odontoid view, an anterior-posterior view, and a cross-table lateral view.[49] Another study supports the use of cervical computed tomography (CT) for diagnosis given the higher specificity.

## CLASSIFICATION

Management of atlanto-axial dislocation (AAD) involves careful study of radiological findings to decide the direction and plane of dislocation, and looking for associated skeletal anomalies and Arnold *Chiari malformation* (ACM) in this region. These findings help to decide the surgical strategy. As per the direction and plane of dislocation we may have the following types of AAD: (1) Anteroposterior dislocation (mobile and hyper-mobile); (2)

Rotatory dislocation; (3) Central dislocation; and (4) Mixed dislocation (any two or three of the above).

***Anteroposterior dislocation***

a. Mobile dislocation is in one plane and one direction. This is due to laxity of the transverse ligament. Therefore C2 dislocates posteriorly in flexion and gets aligned (reduces) in extension. Its anterior movement is prevented by the intact anterior arch of atlas

b. Hypermobile dislocation is in one plane and two directions. When there is os odontoideum, the C2 body movements can not be restricted by the transverse ligament and anterior arch of atlas therefore the C2 body dislocates in both directions in the sagittal plane i.e. posteriorly under the transverse ligament in flexion and anteriorly under the arch of atlas in extension . It is important to identify this hypermobile variety before surgery. During intubation for surgery, anesthetists generally extend the neck of the patient which can result in anterior dislocation of C2 and cause cord injury. Therefore fiber-optic intubation with cervical collar *in situ* is preferred for these patients. These patients should not have flexion or extension movement of neck during intubation.

***Rotatory dislocation***

This type of dislocation is usually in one plane (axial) and in one direction only i.e. to the right or to the left. This is due to incompetence of the alar ligament. It usually occurs in children and is visible as the classical Cocked Robin position of the head. One can clearly see in CT that on one side the AA joint is normal but there is posterior displacement of the C2 facet in the joint at the other side.

***Central dislocation***

The opposing facetal surfaces of the normal C1-C2 joints are horizontal and parallel in the sagittal plane as explained earlier However, if these are oriented obliquely in the sagittal plane as shown in then the C2 body has a tendency to slip upwards due to the weight of the patient's head during flexion movements. Therefore the bilateral sagittal plane obliquity of these joints will result in the telescoping of the C2 body into the C1 ring and result in central dislocation. This telescoping will also invariably result in the posterior movement of C2. The CT picture of this type of dislocation is shown in. The C1 is assimilated with the occiput and there is C2-C3 (C3-third cervical vertebra) fusion. Many neurosurgeons and radiologists report this type of picture (after seeing only sagittal views) as dolichoodontoid and basilar invagination (BI). In fact it is fusion of the C2-C3 bodies, therefore the C2 body with the odontoid as a whole is mistaken for dolichoodontoid. After doing so many transoral operations I have come to the conclusion that the dolichoodontoid entity does not exist. This radiological finding is not BI either; it is central dislocation of C2. One can see the body of C2 behind the anterior arch of atlas in the axial view of the CT scan and there is also posterior dislocation of C2.

***Mixed dislocation***

This dislocation is in two planes and directions: (a) central and posterior-as already discussed central dislocation may be associated with posterior dislocation in many cases (b) posterior dislocation and rotatory dislocation .

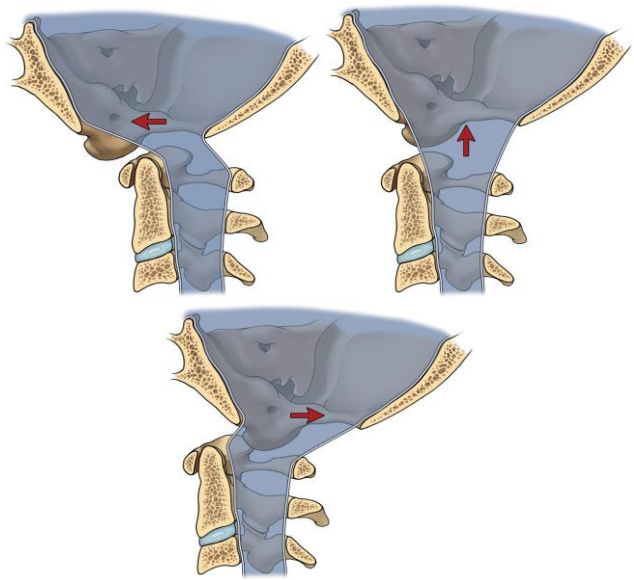


**Fig 5: Types of Dislocation**

## SURGICAL MANAGEMENT OF ATLANTO-AXIAL DISLOCATION

From the surgical point of view there are two broad categories of AAD and the categorization is based on whether C1 and C2 can be aligned or not. If C1 and C2 can be aligned by flexion/extension movements of the neck or by putting cervical traction, then the AAD is called reducible AAD (RAAD). If it is not possible to align them it is called as fixed or irreducible AAD (IAAD). The aims of the surgical treatment of AAD are: (1) All dislocations should be reduced; (2) If the AAD cannot be reduced by the closed method (traction), then the open method (i.e. by opening the joints) should be used for reducing the AAD; and (3) If the dislocation is irreducible by the above means then sufficient decompression of underlying neural structures should be done. In all cases, after achieving the above aims, arthrodesis has to be done for achieving permanent bone fusion to prevent movement between C1 and C2.

## REDUCIBLE ATLANTO-AXIAL DISLOCATION

The surgical procedures which are being discussed here are used for the internal immobilization of C1 and C2 after reduction of AAD. Bone grafts have to be used in all the procedures for ultimate bone fusion (arthrodesis).

1. ***Sub-laminar wiring***
2. ***Occipito-cervical fusion***
3. ***C1-C2 spacer jamming***
4. ***Trans-articular screw with sub-laminar wiring***

## FIXED / IRREDUCIBLE ATLANTO-AXIAL DISLOCATION

***Axial invagination***

When the AAJ becomes almost vertical in both planes as described above, the C2 body invaginates into the C1 ring. This should be called axial invagination (AI). Here it is the axis vertebra which is invaginating inside the C1 ring and foramen magnum. However, if one draws various lines which were described during the era of plain X-ray and tomography, one would call these cases as BI which would be wrong.

***Tran-oral surgery for irreducible atlanto-axial dislocation***

The generally used term 'transoral odontoidectomy' for surgery of congenital IAAD is not correct. In fact those patients who can be treated by only odontoid removal all have the reducible variety of AAD. It is actually corpectomy (the extent of which depends upon axial invagination of C2 and C3) with odontoidectomy which is required to provide sufficient decompression of the underlying cervicomedullary neural structures. This is clarified in the case shown in, wherein the C2 body is still causing severe canal compromise after insufficient odontoidectomy, which was corrected after redo transoral surgery **.** Therefore transoral odontoidectomy seems to be a misnomer. It should be called transoral decompression (TOD) and the bony decompression should be sufficient to provide space for the underlying medulla and cervical spinal cord.

The normal contour of the craniovertebral junction is funnel-shaped which becomes hourglass-shaped in true IAAD/AI. Therefore sufficient bone has to be removed for generous three-dimensional decompressions i.e. antero-posterior (z axis), transverse (x axis), and vertical (y axis) to regain this funnel shape which will then provide sufficient space for the underlying cervicomedullary structures without any compression. This should be planned before surgery on CT scan to do sufficient TOD .

***Fixed chronic/remote post-traumatic atlanto-axial dislocation***

These are usually Type II odontoid fractures which get malunited or nonunited with pseudoarthrosis in dislocated position. There may be abnormal callous formation preventing reduction of dislocation. All these dislocations can be reduced by transoral removal of the offending bone, callous and fibrous tissue.

## Classification of BI ( and other craniovertebral junction disorders) as proposed by Goel and associates:

A classification system for basilar invagination that divided it into two discrete categories. This classification helped in clarifying the understanding of the pathology and pathogenesis of the anomaly, in the selection of the surgical treatment and in prediction of the outcome. [11] The analysis was based on a study of 190 patients of basilar invagination surgically treated in our department during a ten-year period (1988-97). Based on a single criterion of the absence or presence of Chiari malformation, the anomaly was classified into Group I and II respectively. Essentially,

**Group I** included patients where there was invagination of the odontoid process into the foramen magnum and it indented into the brainstem. The tip of the odontoid process distanced itself from the anterior arch of the atlas or the inferior aspect of the clivus. The distancing of the odontoid process from the anterior arch suggested presence of instability of the region and atlantoaxial dislocation. The angle of the clivus and the posterior cranial fossa volume were essentially unaffected in these patiens.

**Group II**, on the other hand, the assembly of the odontoid process, anterior arch of the atlas and the clivus migrated superiorly in unison resulting in reduction of the posterior cranial fossa volume, which was the primary pathology in these patients. The Chiari malformation or herniation of the cerebellar tonsil was considered to be a result of reduction in the posterior cranial fossa volume. In the year 1997, we first defined the clinical implication of association of small posterior cranial fossa volume and Chiari malformation. [11]

We identified a subgroup of patients having basilar invagination where there was clear radiological evidence of instability of the region that was manifested by distancing of the odontoid process from the anterior arch of the atlas, and the radiological features matching those of Group I patients. Considering this current evaluation we have proposed a new classification for basilar invagination into two groups based on parameters that determine an alternative treatment strategy.[12]

In **Group A basilar invagination** there was a 'fixed' atlantoaxial dislocation and the tip of the odontoid process 'invaginated' into the foramen magnum and was above the Chamberlain line, [13] McRae line of foramen magnum [14] and Wackenheim's clival line. [15] The definition of basilar invagination of prolapse of the cervical spine into the base of the skull, as suggested by von Torklus, [7] was suitable for this group of patients [Figure 1].

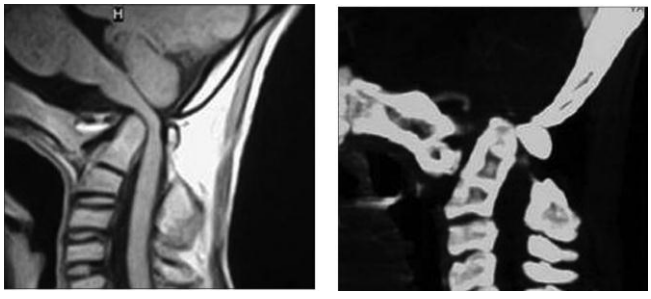


**Fig 6: Group A BI as per Goel et al.**

**Group B basilar invagination** was where the odontoid process and clivus remained anatomically aligned despite the presence of basilar invagination and other associated

anomalies. In this group, the tip of the odontoid process was above Chamberlain's line but below McRae's and Wackenheim's lines [Figure 2].

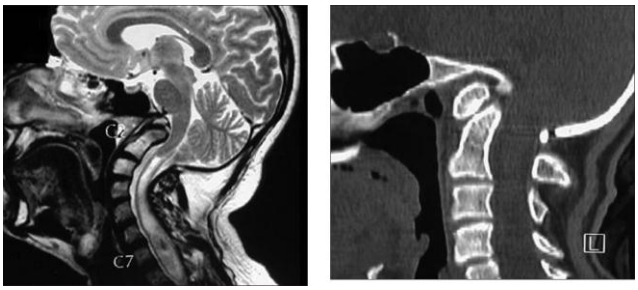



**Fig 7: Group B BI as per Goel et al**

The radiological findings suggested that the odontoid process in Group A patients resulted in direct compression of the brainstem . Essentially, in Group A basilar invagination, there was an element of instability of the region that was manifested by the tip of the odontoid process distancing itself from the anterior arch of the atlas or the lower end of the clivus. In some Group A patients there was Chiari malformation, and this feature differentiates the present classification from the earlier classification. In this group, the atlantoaxial joints were 'active' and their orientation was oblique as shown in [Figure 3], instead of the normally found horizontal orientation.

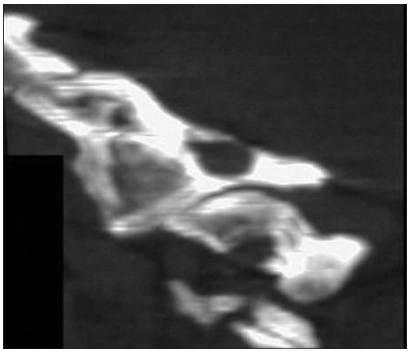


**Fig 8: Oblique orientation of the Atlanto axial joint**

We have found similarities of such a position of the C1-2 facets with spondylolisthesis seen in the subaxial spine. It appears to us that the atlantoaxial joint in such patients is in an abnormal position as a result of mechanical instability and progressive worsening of the dislocation is probably secondary to increasing 'slippage' of the facets of atlas over the facets of axis.

In **Group B**, the atlantoaxial joints were normal and were normally aligned. In some patients the joints were entirely fused. The pathogenesis of basilar invagination appears to be different in the two groups. Understanding these two types of basilar invagination is probably the most crucial factor in understanding the various involved management issues. Group A basilar invagination forms a larger subgroup of patients that are encountered in Indian subcontinent.

We had earlier speculated that patients of Group A basilar invagination and related bone and soft tissue dysgenesis may not be a congenital defect, but can be a result of mechanical 'incompetence' of the atlantoaxial joints . 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.[1] Considering that several physical features associated with this group of basilar invagination are reversible, it appears that the pathogenesis in such patients 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 cervicomedullary cord. However, it appears that it is the cord compression due to indentation by the odontoid process 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. Pain, restriction of neck movements and hyperlordosis of the neck indicate the presence of instability of the craniovertebral junction.

## Craniovertebral realignment for group A basilar invagination

The standard and most accepted form of treatment of Group A basilar invagination is a transoral decompression.[11],[12],[20] The majority of the authors recommend a posterior occipitocervical fixation following the anterior decompression.

Transoral odontoidectomy and resection of the superior half or third of the C2 body was a gratifying surgical procedure in Group A patients.[11] Following this surgery, there was a clinical recovery in all the patients in this group. 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 our current operation that involves craniovertebral realignment without any bone, dural or neural decompression. In Group A patients, the odontoid process was low-lying although posteriorly angulated. Surgery was helped by cervical traction as it reduced the posterior angulation and helped pulling the odontoid process inferiorly. The analysis of the pathology, the surgical experience and results suggest that anterior transoral decompressive surgery is relevant in Group A patients; although in our current evaluation we recommend craniovertebral realignment in these patients.

## Craniovertebral realignment for group A basilar invagination associated with syringomyelia

We had classified syringomyelia and had suggested a specific treatment protocol on the basis of the possible pathogenetic factors.[32] In this study we had suggested that

syringomyelia is a tertiary response to primary craniovertebral anomaly in the form of basilar invagination which leads to secondary Chiari 1 malformation as a result of reduction in the posterior cranial fossa volume. Accordingly, a posterior fossa bony decompression was considered optimum in the treatment of this subgroup of patients.

We have identified patients of syringomyelia where there are associated bony abnormalities of the craniovertebral region that include 'fixed' atlantoaxial dislocation or those having Group A basilar invagination. This select group of patients is treated by attempts towards reduction of the atlantoaxial dislocation and of basilar invagination and by direct lateral mass plate and screw atlantoaxial fixation by techniques described by us.[27] No bony or dural decompression or neural manipulation of any kind is done in these patients.

The majority of the patients with Chiari malformation-related syringomyelia without any bony anomaly of the craniovertebral region have hyporeflexia of the upper extremities and spastic lower extremities. Presence of spastic quadriparesis in all our patients suggests that the symptoms were related to the compression of the brainstem from invaginated dens rather than due to syringomyelia. It was observed that the patients were relatively young, neck pain formed a part of the symptom complex, and motor symptoms and ataxia were far more prominent symptoms in patients having the complex of malformation that included Group A basilar invagination, Chiari 1 malformation and syringomyelia than in patients with a similar complex but without craniovertebral region bony anomalies. It appears that when the angulation of the facets is not as acute or is only marginally affected, the progress of basilar invagination is slow and over several years, providing an opportunity for the syrinx to develop relentlessly.

It was observed that in patients of syringomyelia where there was 'fixed' atlantoaxial dislocation with or without the association of Group A basilar invagination and Chiari malformation, an attempt could be made to realign the bones in the craniovertebral junction.[12] As observed by us earlier, it appears that the atlantoaxial joint in such patients is in an abnormal position as a result of congenital abnormality of the bones, and progressive worsening of the dislocation is probably secondary to increasing 'slippage' of the atlas over the axis. The fact that there was a remarkable clinical improvement following the reduction of the atlantoaxial dislocation and of basilar invagination, it appears that the complex of atlantoaxial dislocation, basilar invagination and syringomyelia are probably secondary to the primary craniovertebral instability. The Group A basilar invagination in patients with associated syringomyelia was less severe and symptoms were longstanding as compared to patients in whom there was no syringomyelia. The conduct of surgery and joint manipulation was relatively easier in these patients.

Following surgery, the alignment of the odontoid process and the clivus and the entire craniovertebral junction improved towards normalcy. We could obtain varying degrees of reduction of the basilar invagination and atlantoaxial dislocation. The atlantoaxial alignments changed towards normalcy and the tip of the odontoid process receded in relationship to Wackenheim's clival line and Chamberlain's line suggesting reduction in the basilar invagination and of atlantoaxial dislocation. The posterior tilt of the odontoid process, as evaluated by modified omega angle, was reduced after the surgery.[11],[12] All patients had a sustained neurological improvement of varying degrees suggesting the effectiveness of the operation. Stainless steel plates, non-locking variety of screws and custom-made spacers were used due to the higher costs of branded material. Due to the type of metal used in the procedure, the effect on syringomyelia not be confirmed.

## Reversibility of musculoskeletal changes following surgery

We had recently analyzed the possibility of reversal of longstanding musculoskeletal changes in patients with Group A basilar invagination after surgical decompression and stabilization.[1] The implications of such an analysis in understanding the pathogenesis of a number of features that are characteristically associated with basilar invagination were evaluated. One hundred and seventy selected patients with basilar invagination who underwent atlantoaxial joint distraction-fixation surgery between 1999 and April 2008 were evaluated in that study. In this series, prior to surgery there were several physical changes such as reduced neck length, torticollis, exaggerated lordosis of the cervical spine, and reduced craniospinal angulation. Other findings included reduced disc-space height, significant posterior cervical osteophytes formation, assimilation of atlas (72%), single level (29%) or multiple-level (3%) cervical fusions, and an increase in spinal subarachnoid space both above and below the level of maximum neural compression at the tip of the odontoid process. After surgical decompression of the region, there was remarkable recovery in craniospinal alignment, and an increase in neck length (maximum 42 mm) was obvious on physical and radiological examination in 85% of patients. The disc-space height increased and there was a reversal of altered cervical lordosis, craniospinal angulation (maximum up to 36 degrees), and torticollis. From our study we concluded that a number of physical spinal changes characteristically associated with basilar invagination such as a short neck, exaggerated neck lordosis, torticollis, cervical spondylotic changes and fusions are potentially reversible after decompression and stabilization of the craniovertebral junction.

Many authors have opined that congenital and developmental musculoskeletal abnormalities of the craniocervical junction complex can cause varying degrees of neural compression. Based on our analysis, it appears that neural compression at the craniovertebral junction is primary and the majority of physical and osseous

abnormalities are probably secondary in nature. A variety of changes that probably occur in the bones and soft tissues appear to be directly related to the natural adjustments that operate to minimize the compression of the cord by the indentation of the odontoid process and the instability of the region. The restriction of neck movements, reductions of neck length, and alterations in the craniospinal angulation probably lead to the presence of large subarachnoid spaces in relationship to the cervical cord and to the brainstem. All these natural responses probably allow the cord a relatively stretch-free traversal over the indenting odontoid process. Reduction of the disc spaces, osteophyte 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. The reduction in the disc-space height and fusions are more prominently seen in the upper cervical vertebrae. It appears that cervical fusions and assimilation of the atlas may be related to longstanding and progressive reduction in the disc-space height.

## Foramen magnum decompression for group B patients

It appears from the analysis of the results in our series, that patients in Group B benefited by foramen magnum bony decompression. The procedure resulted in amelioration of symptoms and at least an arrest in the progression of the disability. None of the patients in this group had a delayed worsening in the neurological condition following a foramen magnum decompression. Driesen reported that during operations for craniovertebral anomalies, he often had to remove noticeably thickened pieces of bone from the posterior edge of the foramen magnum.[25] In our patients, the suboccipital bone and posterior rim of the foramen magnum and the dura overlying the herniated cerebellar tissue were thin in a significant number of patients. This probably was related to the chronic pressure changes secondary to the reduced posterior cranial fossa volume. The bulbous lipping of the posterior rim of the foramen magnum represents the rudiments of the posterior arch of the atlas assimilated into the occipital bone.[4] Various authors have recommended that to achieve maximal decompression, it is necessary to open the dura mater and to cut all constrictive dural and arachnoidal bands. Some authors have recommended leaving the dura open while others have recommended the placement of a graft. Current papers do not recommend resection of the herniating tonsils[33] or even sectioning of adhesions around it. The fact that dural opening was not necessary whilst performing posterior fossa or foramen magnum decompression was first described by us in 1997.[11] This was based on the understanding that the dura is an expansile structure and can never be a compressive factor.[27],[34] Opening of the dura is not only unnecessary but also subjects the patient to an increased risk of cerebrospinal fluid fistula. It turns an otherwise simple surgery into a relatively complex and dangerous surgical maneuver. The treatment of syrinx in the presence of Chiari malformation is also controversial. Logue reported 75 patients treated with craniovertebral decompressions for Chiari malformations and syringomyelia.[35] The patients were divided into two groups: one treated with decompression only, leaving the arachnoid intact, and the other treated with Gardner's procedure of opening the fourth ventricle and plugging the upper cervical canal. They concluded that muscle plugging did not seem to change the results. Levy *et al.*, also concluded on the same lines.[36] Logue noticed that there was no significant need for performing a syringosubarachnoid shunt following craniovertebral decompression.[35] Various subsequent studies have questioned the need for a syrinx drainage surgery following foramen magnum decompression. Di Lorenzo *et al.*, concluded from their study that 'conservative' craniocervical decompression should be considered the first option in the treatment of syringomyelia-Chiari 1 complex.[37] Our experience suggests that only bony decompression of the foramen magnum is sufficient even in patients with syringomyelia.

## SURGICAL OPTIONS IN DETAIL:

### Posterior C1-C2 Stabilization

Trauma is among the most frequent indications for posterior C1-C2 stabilization. Traumatic injuries that are amendable to posterior C1-C2 fixation include certain subsets of Type II and Type III odontoid fractures.

Although most Type II odontoid fractures can be treated either with immobilization or with anterior odontoid screw fixation,[9] there are several subsets of this fracture pattern which are not amendable to these treatment measures. These include Type II odontoid fractures associated with fractures of the atlantoaxial joint, Type II odontoid fractures with oblique fractures in the frontal plane that preclude odontoid screw placement, Type II odontoid fractures with significant displacement which may not heal in immobilization (and are too displaced to place an odontoid screw), Type II odontoid fractures with an associated Jefferson fracture, and Type II odontoid fractures with a ruptured transverse ligament.[9]

In addition, patients with a very large thoracic kyphosis or a very large barrel chest preclude the appropriate angle for anterior odontoid screw placement, and must be treated with a posterior C1 and C2 stabilization procedure.[9]

Even when there is a Type II odontoid fracture that might heal with immobilization, there are certain cases where immobilization is not practical. Elderly patients in particular do not heal well with immobilization. They have a higher rate of nonunion due to osteoporosis and have increased respiratory morbidity when placed in halo vests.[8]

In addition, all patients initially treated with immobilization who develop a pseudoarthrosis are not good candidates for subsequent attempts at anterior odontoid screw fixation because of the pseudoarthrotic

material occupying the fracture line which prevents contact of the decorticated fracture surfaces.[9]

For patients who have failed immobilization and are no longer good candidates for anterior odontoid screw fixation, C1 and C2 fixation is the only remaining treatment option.

Type III odontoid fractures with atlantoaxial joint fracture combinations and Type III odontoid fractures with associated Jefferson fracture are also unstable and are often best treated with a posterior C1 and C2 stabilization procedure.[9]

Congenital malformations of C2 (i.e. os odontoideum and odontoid agenesis), degenerative diseases, inflammatory diseases, tumors, and infections can also result in instability of the atlantoaxial complex. Specifically, rheumatoid arthritis can often result in atlantoaxial subluxation or superior migration of the odontoid into the foramen magnum (with compression of the brainstem and upper cervical spinal cord) necessitating a posterior occipitocervical decompression and fusion (with or without transoral resection of the odontoid).

Post-surgical instability relating to C1 and C2 laminectomies with or without removal of adjoining facets is another indication for posterior C1-C2 fixation. We have performed posterior lateral approaches to remove retro-odontoid degenerative masses and also to remove tumors within the spinal canal that inherently destabilize the C1-C2 complex and require posterior C1-C2 fixation.

Patients may also have ligamentous laxity and have resultant C1 and C2 instability. Ligamentous instability of C1/2 is identified with measurements of the atlanto-dental interval on flexion and extension views. Normally this interval should not exceed 2 to 4 mm.[10],[11] When the atlanto-dental exceeds 5 mm in nonrheumatoid patients and when it exceeds 8 mm in rheumatoid patients, there is instability of the C1-C2 complex and posterior C1/2 fixation is indicated.[12],[13],[14],[15]

Furthermore, atlanto-axial rotatory dislocations are also an indication for C1 and C2 fixation. This problem can be treated via a posterior reduction and fusion approach or via an anterior transoral reduction and C1-C2 fixation.

## METHODS OF POSTERIOR C1-C2 FIXATION

**Posterior C1-C2 fusion with interlaminar clamps**

Posterior interlaminar clamps can be used if the C1-C2 lamina are intact. The technique cannot be used if there are significant degenerative changes or if osteoporosis of the posterior elements of C1 and C2 present. In addition, in cases where there is a Jefferson's fracture or a Hangman's fracture this technique cannot be used.

The clamps are used by placing hooks on the superior surface of the C1 lamina and hooks on the inferior surface of the C2 lamina. The hooks are tightened and preferably a bone graft is placed between the two lamina before the laminar clamps are tightened (Figure 1).

Biomechanically, posterior laminar clamps have excellent stability with flexion and extension maneuvers. However, in rotational motion the clamps are not as effective as other techniques involving posterior screws or wires.[16]

If the posterior clamp construct loosens before bony fusion is achieved, then further surgical intervention will be required.

Consequently, patients are placed in a hard cervical collar or a halo following posterior clamp fusion.

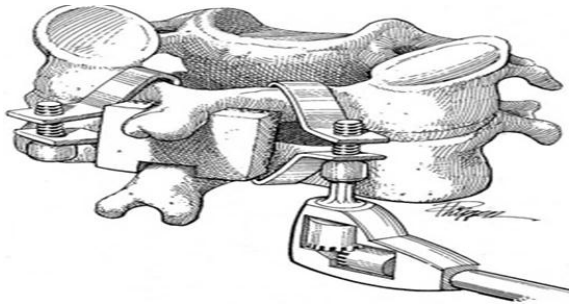


**Fig 10: laminar clamps**

## POSTERIOR WIRING TECHNIQUES

The posterior wiring techniques, like the posterior clamping technique, require an intact posterior arch of C1 and C2. They cannot be utilized if there are fractures of the C1 or C2 posterior elements (including Hangman's or Jefferson's fracture), or if posterior decompression of the C1-C2 complex is required, or if there is significant osteoporosis.

Unlike the interlaminar clamping techniques, the posterior wiring techniques require sublaminar passage of a cable and have the potential for injury to the dura or spinal cord during this maneuver.

When we perform posterior wiring techniques, we prefer to use Atlas double braided titanium cables (Medtronic Sofamor Danek, Memphis, TN) because they are more flexible than steel wire and have less chance of causing dural or neural injury. Gallie fusion Gallie first described posterior C1-C2 sublaminar wire fixation in 1939 with the use of steel wire.[17] In the Gallie technique, a single autograft harvested form the iliac crest is notched inferiorly and placed over the C2 spinous process and leaned against the posterior arch of C1. The graft is held in place by a sublaminar wire that passes beneath the arch of C1 and then wraps around the spinous process of C2. Passage of the sublaminar wire under the lamina of C2 is avoided in order to decrease the risk of neural or dural injury.

The **Gallie** fusion offers good stability in flexion and extension. However, like interlaminar clamping it offers very poor stabilization and rotational maneuvers. Consequently, the rate of nonunion with the Gallie fusion has been reported to be as high as 25%.[18]

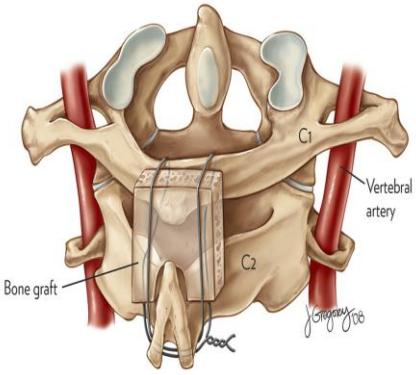



**Fig 11: Gallie's fusion**

### *Brooks-Jenkins Fusion*

In the Brooks-Jenkins fusion technique, unlike the Gallie fusion technique, two separate iliac crest autografts are placed between C1 and C2. Each autologous iliac crest graft is beveled superiorly and inferiorly and wedged in between the C1 and C2 lamina on each side of the midline. One sublaminar cable is then passed on each side of the midline under both the C1 and C2 arches and wrapped around each bone graft respectively. The cables are then tightened around the grafts and secured and crimped in place.

The Brooks-Jenkins fusion technique provides more rotational stability than does the Gallie technique.[19] The Brooks-Jenkins technique has similar stability in flexion and extension as does the Gallie fusion technique.[20] The rate of fusion after this technique has been reported to be as high as 93% and is improved by the use of halo immobilization.[4]

The disadvantages of the Brooks-Jenkins fusion technique include the need for passage of bilateral sublaminar cables beneath both C1 and C2. This entails a higher potential rate of neurological or dural injury than does the single cable passage under the posterior C1 arch for the Gallie technique.[19]

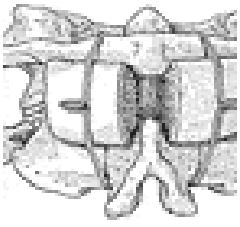


Fig 11: Brooks fusion

### *Sonntag Posterior C1-C2 Technique*

The Gallie technique was modified by Volker Sonntag in the early 1990s. Sonntag's modified technique improves the rotational stability of the Gallie fusion technique while avoiding the bilateral sublaminar C1-C2 cable passage of the Brooks-Jenkins technique.

In the Sonntag technique, a sublaminar cable is passed under the posterior C1 arch from inferior to superior. Next a notched iliac crest is placed in between the spinous process of C2 and wedged underneath the posterior arch of C1 (Figure 2) (unlike the Gallie technique where the bone graft is notched over the spinous process of C2 and simply leaned onto the posterior arch of C1). Both the superior aspect of the C2 spinous process and the inferior arch of C1 are decorticated before graft placement.

The cable is then looped over the iliac crest autograft and placed into a notch created on the inferior aspect of the C2 spinous process. The cable is then tightened and crimped.

In patients treated with a wiring procedure only, Sonntag recommends the use of a halo to immobilize patients for three months after surgery and the use of a rigid cervical collar for an additional one to two months after that. With this kind of immobilization he has demonstrated a 97% fusion rate with the technique.[4]

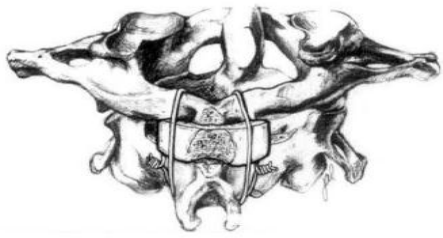


**Fig 12: Sonntag method for wiring**

### *C1-2 Transarticular Screw Technique*

1n 1979, Magerl and Jeanneret described transarticular screw fixation for the treatment of odontoid fractures. [21],[22]

One advantage of the C1-2 transarticular screw technique is the complete obliteration of rotational motion of the atlantoaxial joint. However, the disadvantages of this technique are the steep learning curve and the potential for serious complications including errant screw placement leading to spinal cord injury, hypoglossal nerve injury, or vertebral artery laceration.

To avoid complications we routinely perform pre-operative CT scanning of the cervical spine (often with three-dimensional remodeling sequences on a Stealth station (Medtronic Sofamor Danek, Memphis, TN) to identify an anomalous vertebral artery course (Figure 3), destruction of bone at the intended site of screw fixation, or an unacceptably small C2 pars. In addition, we recommend and routinely use pre-operative MRI scanning in order to assess the degree of neural compression and the integrity of the transverse atlantal ligament prior to performing this procedure.

In order to minimize the risk of vertebral artery injury with transarticular facet screw placement, we only place the screw if the preoperative CT confirms the normal anatomic position of the vessel.[23],[24],[25] In cases with aberrant unilateral vessel position, we place the screw only on the normal side. In case of a vertebral artery injury during screw placement, our preference is to place the screw to tamponade the bleeding. The contralateral screw should not be attempted in this setting in order to avoid the risk of bilateral vertebral artery injury. The patient should be taken to angiography to assess the injury which has occurred and to potentially sacrifice the injured vessel angiographically.

To perform the C1-2 transarticular screw technique, we position the patient prone in a Mayfield head holder (OMI, Inc., Cincinnati, Ohio). We leave the neck in a neutral position and flex the head on the neck in a "military tuck" position (Figure 4A). This affords posterior translation and reduction of the C1-C2 complex allowing the surgeon access to the desired C1-2 trajectory. The patient is prepared and draped to expose from the suboccipital to the midthoracic area. Before making a skin incision, we use lateral fluoroscopy we determine the planned entrance site on the skin for the screw trajectory (usually the paramidline area near the T1 spinous process) in order to ensure that enough of the thoracic spine is prepped into the surgical field. We create a separate midline incision to expose the posterior elements of C1-C3.

Next we identify and palpate the bony limits of the C2 lateral mass. The superior and medial aspect of the C2 pars are exposed and palpated with a Penfield dissector. Using the Penfield 4 dissector, the C2 nerve is undermined with gentle sweeping movements in a rostral direction. The Penfield 4 dissector is then passed medially and superiorly over the pars and pedicle of C2 to determine the angle for the screw. There is typically a robust epidural venous plexus on the medial aspect of the pars of C2 which can be controlled by bipolar cautery. We do not dissect on the lateral portion of the pars of C2, as this is not necessary and can increase the risk of bleeding from the paravertebral venous plexus.

In patients with significant C1-C2 subluxation, a reduction of C1 onto C2 must be performed before the screw path can be drilled. The reduction can be accomplished by simple manipulation of the Mayfield head holder, or it can be accomplished by first creating a posterior interspinous tension band with a Sonntag interspinous construct.

With fluoroscopic guidance or with intraoperative neuronavigation with a Stealth station, the proper trajectory is confirmed by placing the drill or similar instrument adjacent to the neck outside of the incision. This trajectory should cross the C1-C2 facet joint and at the anterior arch of the atlas. We usually find that the percutaneous entrance site for the drill lies approximately 2cm lateral to the T1 spinous process (Figure 4B) (and this is why we prepare and drape the upper thoracic spine into the surgical field). We create stab incisions approximately 1cm from the midline bilaterally.We place a guide tube with an obturator through the stab incision site and into the open surgical site at C1-C3. The tip of the guide tube is docked at the C2 entry site. The C2 entry site is identified by locating the inferior medial angle of the C2-C3 facet joint. The entry site is approximately 34 mm rostral and 3-4 mm lateral to this point (i.e., from the inferior medial facet joint of C2-3 "go up 3 mm and out 3 mm''.

With a high speed drill the cortical bone is pierced to mark a Kwire entry site. The K-wire trajectory is typically 15 degrees medial with the superior angle visualized by fluoroscopy. The K-wire is directed down the C2 pars and pedicle complex and across the C1-C2 joint, aiming at the anterior tubercle of C1. The tip of the K-wire is advanced to a point 3-4 mm posterior to the anterior C1 tubercle (to avoid penetration of the wire into the retropharyngeal area).

While the K-wire is drilled, subtle changes may be perceived in resistance as the K-wire subsequently traverses the 4 cortical surfaces along its path into the C1 lateral mass. The cortical surfaces include the posterior C2 entry point, the superior articular surface of C2, the inferior articular surface of C1, and the anterior cortex of C1 ring.

After the K-wire is placed, a cannulated drill bit is passed over the K-wire and drilled to the same target point. Care must be taken to avoid advancing the K-wire further as the drilling is performed. We typically have an assistant hold the K-wire with a needle driver as the drill bit is advanced in order to avoid advancing the K-wire beyond the anterior arch of C1 and into the soft tissues of the neck.

The pilot hole is then tapped over the K-wire through the C1-C2 facet joint and into the lateral mass of C1. Subsequently, a fully threaded 3.5 mm or 4 mm cortical screw is placed over the Kwire (Figure 5). The necessary screw length can be measured directly from the drill or from the K-wire inserted. The screw is usually 1-3 mm shorter than the actual measured length since some degree of compression of the C1-2 joint occurs with screw placement. (We typically use screws 34-44 mm in length). An oversized screw may breach the anterior cortical margin and injure the pharyngeal soft tissue. On the other hand, a screw that is too short may compromise purchase.

This same technique is repeated on the opposite side as well. If not already performed, we usually supplement transarticular screw fixation with a Sonntag posterior interspinous wiring procedure for added stability.[21,22, 26]

We typically mobilize patients in a hard cervical collar for three months after performing the procedure. We prefer to perform the C1-2 transarticular fixation with either Axis screws or UCSS screws (Medtronic Sofamor Danek, Memphis, TN). More recently, we have incorporated a C1/2 transarticular screw into a multilevel cervical construct using the VERTEX polyaxial screw-rod system (Medtronic Sofamor Danek, Memphis, TN).

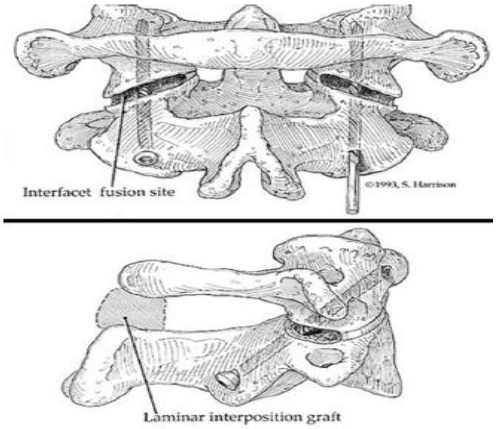



**Fig 13: Transarticular screw fixation screw placement and patient position for surgery**

**Goel's C1 lateral mass screw with C2 pars or pedicle screw fixation**

The technique of segmental atlantoaxial fixation and fusion using C1 lateral mass screw and C2 pedicle screw and plates was pioneered by Prof Goel et al.[6] The authors achieved 100% fusion with minimal rate of complications.[7] The authors advocate bilateral sacrifice of C2 ganglia in order to prepare the atlantoaxial facet joints for arthrodesis.

Goel et al have highlighted the advantage of their method over other constructs.[7] One of the advantages of the C1 lateral mass in combination with C2 screw technique is that anatomic alignment of the C1-C2 complex is not necessary prior to instrumentation. In addition, this technique can still be utilized in cases where there is an aberrant vertebral artery. The plates act as tension-band, providing stability in flexion/extension and hence a midline procedure by Gallie's fusion or Brooks fusion is not necessary. The procedure is technically demanding and precise and an exact three-dimensional understanding of the anatomy of the region and of the vertebral artery is mandatory. Large venous plexuses in the lateral gutter need to be handled appropriately. Goel et al have reported that sacrifice of the C2 ganglion provides a wide exposure to the region for the conduct of the surgery and does not lead to any significant neurological symptom. Biomechanical testing has shown that C1 lateral mass screw with C2 pars screw construct allows an average of 0.6 degrees more motion than does a C1-2 transarticular screw.[29]

Recently several authors including us have modified the technique and use polyaxial screws and top loading rods and do not sacrifice the C2 ganglia.[26], [27]

## SURGICAL TECHNIQUE

The patient is positioned prone using a Mayfield head holder (OMI, Inc, Cincinnati, OH). The neck is kept neutral and the head is placed in the military tuck position. The arms are tucked at the sides. The shoulders are retracted caudally using tape. A midline incision is made extending from the suboccipital area to the spinous process of C3. The C2-C3 facet joints are exposed and the dorsal arch of C1 is exposed laterally exposing the vertebral artery in the vertebral groove on the superior aspect of the C1 arch (sulcus arteriosis). The C2 nerve root is identified and is either sacrificed or mobilized inferiorly. Bipolar cautery and haemostatic agents such as gel foam are used to control bleeding from the venous plexus surrounding the C2 nerve root and also surrounding the vertebral artery. The lateral mass of C1 inferior to the C1 arch is exposed after the C2 nerve root has been sacrificed or mobilized inferiorly. The medial wall of the C1 lateral mass is identified using the forward angle curette to palpate the medial limit of screw placement. The medial aspect of the transverse foramen at C1 and C2 can also be identified and serve as a lateral limit for screw placement.

The entry point for the C1 lateral mass screw is identified at the centre of the C1 lateral mass. Another suggested entry point by Goel et al[30] is at the junction point of the midpoint of the C1 lateral mass midpoint and the inferior aspect of the C1 arch (Figure 6). It should be noted that the vertebral artery often runs in a sulcus on the superolateral aspect of the C1 arch and care should be taken to avoid drilling in this area (Figure 7).

Using fluoroscopy, a 3 mm drill bit and guide are used to drill a hole with 10-15 degrees medial angulation to penetrate the anterior cortex of C1. On lateral fluoroscopic imaging the drill is aimed towards the anterior tubercle of C1 so that the drill penetrates the ventral cortex of the lateral mass midway between the superior and inferior facets of C1. The hole is tapped and subsequently,a C1 lateral mass screw is placed (Figure 8).

We then turn our attention to placing a screw at C2. The C2 screw can either be placed in the pars of C2 or in the pedicle of C2. The pars of C2 is defined as the portion of the C2 vertebra between the superior and inferior articular surfaces. A C2 pars screw is placed in a trajectory similar to that of a C1-C2 transarticular screw except that it is much shorter. The entry point for the C2 pars screws 3 mm rostral and 3mm lateral to the inferior medial aspect of the inferior articular surface of C2. The screw follows a steep trajectory paralleling the C2 pars (Often 40 degrees or more) (Figure 9). We are usually able to achieve this trajectory through an incision that extends down to C4 without using a percutaneous stab incision at T1 (Figures 10 and 11). The

screws are passed with 10 degrees of medial angulation. Screw length is typically 16 mm, which often stops short of the transverse foramen (Again, we confirm this with pre-operative CT scanning). Vertebral artery injury is still a risk with C2 pars screws but the risk is not as high as with transarticular screws.

The C2 pedicle, on the other hand, is the portion of the C2 vertebrae connecting the dorsal elements with the vertebral body (The C2 pedicle is anterior to the C2 pars). The trajectory of the C2 pedicle screw is different than that of C2 pars screw. The entry point for a C2 pedicle screw is in the pars of C2, lateral to the superior margin of the C2 lamina. This point is usually 2 mm superior and 2 mm medial to the entry point for the C2 pars screw that we have just described. Goel's C1 lateral mass screw with C2 pars/ pedicle screw fixation (Figure 6). The screw is placed with 15-25 degrees of medial angulation. The thick medial wall of the C2 pedicle will help redirect the screw if necessary and prevent medial wall break out and entry into the spinal canal. The screw is placed after a drill is used to create the entry hole and after the hole is tapped. The trajectory of the C2 pedicle screw is 20 degrees up angle and 15-25 degrees medial from the entry point. The screws are tightened over a plate or rods are top loaded onto the screw heads (our modification).

The versatility of this technique for the treatment of various craniovertebral disorders has been highlighted by Goel et al in their numerous publications.[31],[35]

Recently, various authors have reported variations of this technique by substituting C1 or C2 laminar hooks or by using C2 translaminar screws.[36]

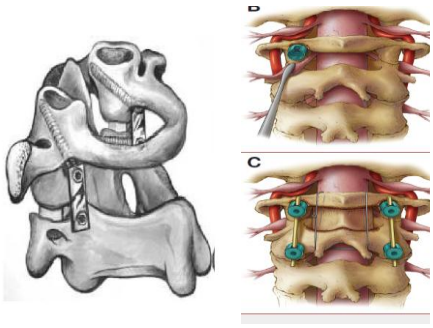


**Fig 14: Goel fusion technique**

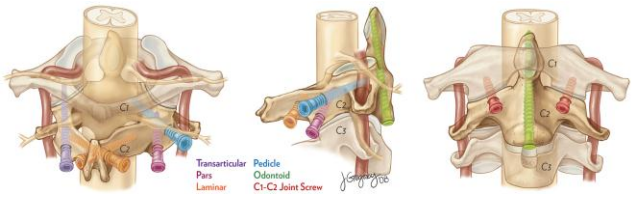



Fig 14: summary of the fixation techniques

**Anterior C1 and C2 fixation technique**

Transoral instrumentation for unstable craniovertebral junction was described by Goel et al in 1994.[36] Harms and colleagues have recently used transoral technique to fixate the anterior cervical spine in patients undergoing transoral approaches to the odontoid for rotatory dislocations, tumors, or infections. The advantage of this technique is that the patient undergoing transoral decompression with anterior fusion can avoid a subsequent posterior neck incision for C1-2 fixation.

The technique is performed with the use of a "T-plate" (Depuy Spine, Raynham, MA). The horizontal portion of the plate is placed over the C1 lateral masses anteriorly and screws are placed through the plate apertures into the anterior C1 lateral mass to achieve a bicortical purchase. The vertical portion of the plate rests on the body of C2 inferior to the base of the dens. Two vertebral body screws are then placed just superior and parallel to the C2-3 disc space. This procedure requires an extensive posterior pharyngeal "C" shaped flap via the transoral approach. Midline linear posterior pharyngeal exposures are not recommended if this "T-plate" is used because they are prone to wound breakdown due to the profile of the "T-plate" causing pressure on the pharyngeal incision.

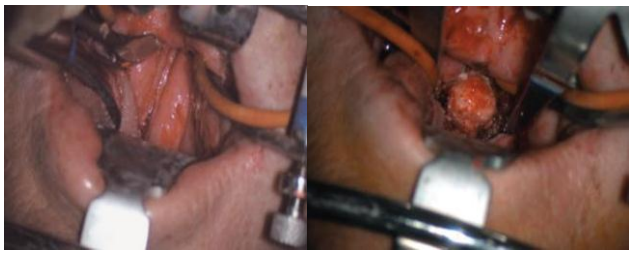


**Fig 15: transoral dissection of the posterior pharyngeal wall with the underying odontoid**

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