Atlantoaxial Stabilization by Posterior C1 and C2 Screw-Rod Fixation for Various Pathologies: Case Series and Comprehensive Review of Literature

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We retrospectively analyzed atlantoaxial dislocation (AAD) of various pathologies, namely, rheumatoid arthritis (RA), os odontoideum, and trauma. Various techniques were discussed in relation to C1-C2 stabilization. The study aims to share our clinical experience in a series of six cases of C1-C2 instability that underwent posterior C1-C2 fusion, with free hand technique and limited fluoroscopy. The clinicoradiological presentation for each patient is described. We reviewed different literatures related to our case vividly and focused on the basic neuroanatomy involved in the atlantoaxial joint. All patients of AAD had evidence of severe canal compromise and chronic compressive spinal cord changes. In our study, the patients age ranged from 28 to 52 years. The study included four males and two females. Out of six patients of AAD, three had history of trauma, two had os odontoideum, and one had chronic inflammatory condition (RA). From our case series, we concluded that the Goel–Harms technique is the most versatile and surgeon friendly technique for C1-C2 fixation. Early recognition and surgical intervention of atlantoaxial joint instability is essential to prevent catastrophic neurological complications.

Keywords
- atlantoaxial dislocation
- C1-C2 fixation
- os odontoideum
- odontoid fracture
- rheumatoid arthritis
- neuroanatomy
- C1 lateral mass
- C2 pedicle

Abstract

Introduction

Traditionally, there are several surgical management techniques used to manage C1-C2 instability such as the Gallie, Brooks–Jenkins, or interspinous methods.1-3 Unfortunately, studies have shown that the nonunion rates for these techniques are as high as 80% (range, 3–80%) even with the use of postoperative immobilization such as the halo vest.4 These unsatisfactory outcomes led to the development of newer techniques for C1-C2 fusion instrumentation, including the use of C1-C2 transarticular screws.5 Although biomechanically superior to the wiring techniques, the Magerl technique is contraindicated in obese patient and in patients with increased thoracic kyphosis.6 Further, this technique also requires preinstrumentation reduction of any subluxation. In 1994, Goel and Laheri reported the use of plates and C1 lateral mass and C2 pars interarticularis screws.7 In 2001, Harms and Melcher further popularized the technique of posterior C1-C2 fusion with C1 lateral mass screw and C2 pedicle screw.8 Although this technique is technically challenging, but it has showed superior biomechanical and clinical results. The advantages of the new technique (Goel–Harms) are safer trajectory of the screws, potential for postinstrumentation reduction, and avoidance of damage to the C1-C2 facet joint, and this makes it superior to other C1-C2 fusion in terms of reduction of C1-C2 subluxation, which leads to improved indirect decompression of the spinal cord. Harms
and Melcher reported a 100% fusion rate. Preoperative bony anatomy of cervicomedullary junction and vertebral artery are mandatory in planning the procedure to avoid catastrophic complications.

This study aims to share our clinical experience in a series of six consecutive cases of various pathologies of atlantoaxial dislocation (AAD), which underwent posterior C1-C2 fusion, using the C1 lateral mass screw and C2 pedicle/pars screw with free hand and under limited fluoroscopy. We used titanium polyaxial screw and rod systems, and the length of the screw was determined preoperatively from CT scan. This technique was found to be surgeon friendly, statistically reliable, and led to reproducible results if bony anatomy of craniovertebral junction (CVJ) was conducive to the surgery. We described the clinicoradiological presentations of each patient and also reviewed the literatures related to our case comprehensively.

Illustration of Cases

C1-C2 Instability in Rheumatoid Arthritis (RA)
Patient 1 (†Fig. 1)
A 46-year-old female presented with progressive neck pain and minimal difficulty in walking for the past 5 years. The pain was radiating to bilateral posterior shoulders and more onto her left arm. The neck pain worsened with movement, but she had no complaints of numbness, tingling and gross weakness in extremities. On examination, it was found that she had normal higher mental function, tone was increased in both upper limbs, power of the limb was normal, tandem walk was not possible, and Hoffman's sign positive, more marked on left side. There was an exaggerated reflex in bilateral upper and lower limbs, but jaw jerk was absent. Patient was on medication for RA prior to surgery (†Fig. 1A, B); X-ray of C-spine showed mild increase in atlantodental interval (ADI) and osseous erosion at C1 and C2 level (†Fig. 1D); atlantodental distance was apparently increased with abnormal hypointense soft tissue and abnormal spinolaminar line at CVJ (†Fig. 1C, E). CT scan shows erosion, sclerosis, partial fusion of left atlantoaxial facet, mild increased ADI 4.4 mm, and cortical irregularities, with bone formation noted in the anterior arch atlas. There were end plate irregularities and sclerosis at multiple vertebral levels too. No evidence of basilar invagination (†Fig. 1F): postoperative C-spine.

Os Odontoideum
Patient 2 (†Fig. 2)
A 35-year-old female patient, with history of fall in the bathroom 9 months back, presented to us with progressive neck pain radiating to bilateral shoulder and also complained of numbness and progressive weakness in her both upper limb (Medical Research Council [MRC] grade ⅘) and lower limbs (MRC grade 5/5) for the last 3 to 4 weeks. Patient had no bladder and bowel involvement.

CT and MRI scan of spine done revealed os odontoideum dystopic type with atlantoaxial dislocation along with T2 hyperintensity at C2 suggestive of myelomalacia. X-ray C-spine shows os odontoideum (†Fig. 2A). CT C-spine shows os odontoideum (†Fig. 2B). MRI C-spine shows atlantoaxial dislocation with spinal cord compression and T2 hyperintensity suggests myelomalacia (†Fig. 2C).
X-ray C-spine anteroposterior view with C1-C2 fusion and stabilization (►Fig. 2D).

Patient 3 (►Fig. 3)
A 52-year-old male manual laborer presented with neck pain following sudden lifting of weight on his head. But for the last 2 months, pain severity had increased significantly with restriction of neck movements. Also, patient had numbness and weakness in bilateral upper limbs, which had progressed to both the lower limbs over the course of 2 months. On examination, there was hypertonia, exaggerated reflexes, Hoffman’s positive both sides, and Nurick grade 3. C-spine X-ray (lateral–flexion and extension view) was suggestive of atlantoaxial dislocations (►Fig. 3A, B). CT scan revealed os odontoideum with hypertrophied and sclerotic anterior arch of atlas. C1/C2 subluxation was noted with sclerosis and cystic changes in right half of C1 and vertebral body (►Fig. 3C, D). MRI revealed smooth well corticated ossicle of size 9.4 × 8 mm superior to dens. Anterior subluxation of atlas over axis was noted with atlantodental distance, narrowing of spinal canal at the same level with thinning of spinal cord and T2 hyperintensity at CVJ and upper cervical canal level (►Fig. 3E,F). ►Fig. 3G shows postoperative image.

Odontoid Fracture and Atlantoaxial Dislocation (AAD)

Patient 4
A 46-year-old male presented to us with history of fall following slippage and sustained neck trauma for 3 months. Patient had neck pain and numbness on both upper limbs and was progressive for the last 1 month. On examination, there was hypertonia, Hoffman’s sign positive and bilateral plantar extensor. The gait was spastic, and there was no bladder and bowel involvement. The patient was subjected to investigation and C spine X-ray showed atlantoaxial instability be (AAI). CT CVJ showed odontoid fracture involving the base of the odontoid with anterior displacement of proximal fracture fragment and spinal canal narrowing (odontoid fracture—type III). MRI revealed fracture odontoid without evidence of myelomalacia (►Fig. 4A). MRI sagittal showed fracture at the base of odontoid process and posterior subluxation causing spinal canal stenosis and cord compression (►Fig. 4B). CT sagittal section of spine revealed odontoid fracture at the base of odontoid process with anterior displacement of proximal fracture fragment and spinal canal narrowing (►Fig 4C); Goel–Harms technique.

Patient 5
A 37-year-old male patient came to our hospital with a history of fall 8 years back with neck pain, features of myelopathy, and Nurick grade 3. CT sagittal spine shows anterior displacement, angulation of odontoid fracture segment and distal segment displaced posteriorly causing compression at CVJ (►Fig. 5A).
Fig. 4 (A–C) Images of patient 4.

Fig. 5 (A–C) Images of patient 5.

Fig. 6 (A–D) Images of patient 6.

►Fig. 5B shows displaced fracture of odontoid through the base, and fracture segment displaced 5 mm in sagittal image with narrowing CVJ. X-ray C-spine shows screw and rod in situ (►Fig. 5C).

Patient 6
A case of a 28-year-old male, with a history of motor vehicle accident 1 year back, presented to us with features of myelopathy and progressive neck pain. MRI T2 sagittal shows exaggeration of cervical lordosis; blunting of C2 vertebra was noted with evidence of dorsiflexion of C2 vertebral body, causing compression of spinal cord (►Fig. 6A). Altered signal intensity and thinning of spinal cord were noted at CVJ. ►Fig. 6B shows fracture involving odontoid process with proximal fracture segment displaced anteriorly, lying in close contact with posterior margin of arch of atlas and distal C2 displaced posteriorly, causing severe spinal cord compromised. C-arm view of C1-C2 screw and rod fixed in situ (►Fig. 6C). 3D CT reconstruction image depicting course of vertebral artery and anatomy of C1-C2 is shown in ►Fig. 6D.

Surgical Outcome
The age of the patients ranges from 28 to 52 years (►Table 1). Four male and two female patients were selected in the study. Out of six patients of AAD, three had history of trauma,
two had os odontoideum, and one had chronic inflammatory condition (RA). No major intraoperative complications were noted. Five patients were fused with C1 lateral mass and C2 pedicle, and in one patient, we used pars screw instead of pedicle. The screw placement was satisfactory. No implant failure noted. There was an adequate increase in diameter of spinal canal and alignment. In all the patients, significant improvement was noticed in terms of symptoms, particularly neck pain and numbness. There was considerable increase in Nurick grade in 5 patients which was reviewed after 6 months in outpatient department (OPD) assessment. However in one patient, the improvement was not up to the mark, as there was delay in surgical intervention.

**Discussion**

Anderson and D’Alonzo classification of odontoid fractures was based on the anatomic location of the fracture. There are three types of C2 odontoid fractures. Type I: An oblique fracture through the upper part of the odontoid process. Type II: Fracture at the base of the odontoid, as it attaches to the body of C2. Type III: When the fracture line extends through the body of the axis. Type I fracture is seen in less than 5% of cases, and Type II fracture is seen in more than 60% of the cases. Hadley and colleagues modified this classification and described type IIA odontoid fractures, where there were additional chip fracture fragments at the anterior or posterior portion of the fractured base of the dens. It is invariably unstable and leads to nonunion without surgical intervention. Type III fracture is seen in approximately 30% of the cases. It extends into the vertebral body and is relatively stable unless severely displaced. The classification systems are helpful to decide the type of treatment to be chosen. The rate of union of the fractures is negatively affected by age of the patient, the extent of fracture displacement, comminution of the fracture, and delay in diagnosis. The decision becomes difficult to decide the type of treatment, when a type II fracture extends inferiorly to consider a type III fracture. Such intermediate fractures have been referred to as “shallow” or “high” type III fractures. Therefore, these fractures are considered to be in a gray zone between type II and type III fractures for which some surgeons advice fixation as the choice of treatment. Thus, this difference in opinion leads to confusion. Grauer et al proposed a classification system for type II fractures—type II-A, II-B, and II-C, which is also treatment-oriented. Type IIA is defined as a minimally or undisplaced type II fracture with no comminution. These fractures are generally treated with external immobilization. Type IIB is a displaced fracture, extending from anterior–superior to posterior–inferior, or a transverse fracture. These fractures are to be treated with anterior screw fixation following fracture reduction, assuming adequate bone density. Type IIC is a fracture line extending from anterior–inferior to posterior–superior or a fracture with significant comminution. These fractures are generally treated with posterior atlantoaxial stabilization.

The unique characteristics of CVJ are anatomical and biomechanical properties, which distinguish it from the subaxial spine. It contains vital neural and vascular structures. The neurologic deficits occur as a result of injury to brain stem, lower cranial nerves, and vertebral artery.

The atlas lacks a vertebral body and instead articulates with the odontoid process or dens, a bony protuberance extending superiorly from the vertebral body of the axis. The atlas also communicates inferiorly with the axis by flat, wide articular facets. The odontoid process and horizontal facets permit rotation of the skull, the predominate motion of the C1–C2 vertebral junction. The transverse ligament of the atlas constrains the dens within 3 mm of the anterior ring of the atlas by bounding the dens posteriorly.

The atlantoaxial instability due to traumatic rupture of transverse atlantal ligament (TAL) can lead to increase in the ADI. The ADI is the space between posterior portion of anterior arch of C1 and to the anterior portion of the odontoid process. In adults, an ADI less than 3 mm is considered to be normal. An ADI greater than 5 mm suggests disruption of both the TAL and accessory ligaments.

Inferior and superior crura arise from the transverse ligament, as it crosses the dens, attaching to the body of C2 and

### Table 1 Patient profile

| Patient number | Age (in years) | Sex | Diagnosis | Procedure | Myelomalacia | Preoperative symptoms | Remarks |
|----------------|---------------|-----|-----------|-----------|-------------|----------------------|---------|
| 1              | 46            | F   | AAD, RA   | C1 lateral mass screw + C2 pedicle + spacer | Neck pain, numbness upper limb | Improved |
| 2              | 35            | F   | Os odontoideum + AAD | C1-C2 Fusion | Yes | Neck pain, NG-III | Improved |
| 3              | 52            | M   | Os odontoideum + AAD | C1-C2 Fusion | Yes | Neck pain, NG-III | Improved |
| 4              | 46            | M   | Os odontoideum + AAD | C1-C2 Fusion | Yes | Neck pain, NG-III | Improved |
| 5              | 37            | M   | #Odontoid, type-III | C1-C2 Fusion | Yes | Neck pain, NG-III | Improved |
| 6              | 28            | M   | #Odontoid + AAD | C1-C2 Fixation | Yes | Neck pain, NG-IV | Improved |

Abbreviations: AAD, atlantoaxial dislocation; F, Female; M, male; NG, Nurick grade, RA, rheumatoid arthritis.

Note: the symbol “#” denotes fracture.
the anterior foramen magnum, respectively. Taken altogether, the transverse ligament and its crura form the cruciform or cruciate ligament. The transverse ligament contributes substantially to the stability of the CVJ, preventing the dens from folding into the midbrain during flexion. The alar ligaments arise from the anterolateral aspect of the dens and attach to the medial aspect of the occipital condyles, inferior to the foramen magnum. The primary function of the alar ligaments is to restrict rotation of the cranium. The alar ligaments are also critical to maintaining stability at the CVJ. Arising from the subclavian artery, the bilateral vertebral arteries progress cephalad via the transverse foramina of the cervical spine. After exiting the transverse foramina of C1, the vertebral arteries course along the superior surface of the posterior ring of the atlas before turning ventrally to pass through the atlanto-occipital membrane, medial to the superior articular facet.

The atlantoaxial junction is a delicate anatomic complex that involves an intricate relationship between the atlas and the axis to allow flexion, extension, rotation, and lateral bending. The interrelation of osseous, ligamentous, neural elements and articulation make the atlantoaxial junction vulnerable to instability and potentially devastating neurologic complications (quadriplegia, diaphragmatic paralysis and sudden death).

The lateral masses have large, concaved and superior surfaces to articulate with the occipital condyles. They are directed upward, medially and backward and form a cup for the corresponding condyles. The inferior surfaces are circular and directed downward and medially to articulate with the superior articulating facet of C2 and form the zygapophyseal joints, which allows flexion-extension, side bending, and rotational movements. The atlantoaxial joints are the articulation between C1 and C2 and have a range of motion in the transverse plane for rotation. This rotation is facilitated by the odontoid process, which acts as a pivot joint for the transverse plane for rotation. This rotation is equated with instability or instability with neurologic development or prior unrecognized trauma, requires the same radiologic and clinical workup. Surgical stabilization of the upper cervical spine has been proposed for progressive symptomatology, including intractable neck pain, transient or progressive myelopathy, vertebral artery insufficiency and worsening instability. From the consecutive case series of 71 RA patients by Yan et al., the instability of the atlantoaxial joint, including anterior atlantoaxial subluxation, posterior atlantoaxial subluxation and anterior-posterior atlantoaxial subluxation were found in 68 cases (95.7%), while rotation subluxation was presented in 37 cases (52.1%). Vertical migration of the odontoid was seen in 11 cases (15.5%). Rheumatoid factor was positive in 18 cases (25.3%). Patients who have os odontoideum

Static stability is conferred by both osseous and ligamentous contributions consisting primarily of the facet articulations, dens and fovea dentis, the facet capsule, and the TAL. Dynamic stability arises from the multiple muscular attachments of the anterior arch and transverse process. Trauma, congenital malformations, inflammatory arthritides (RA), and malignancy have been implicated in the development of atlantoaxial instability. Since the first description of surgical treatment by Mixter and Osgood in 1910, multiple techniques have been described to provide atlantoaxial stability in an effort to protect the space available for the spinal cord and prevent basilar invagination.
with a reducible atlantoaxial dislocation can be effectively treated with single-level posterior fusion and stabilization. The differential diagnosis for os odontoideum typically includes ossiculum terminale and type II odontoid fracture.\textsuperscript{9}\textsuperscript{a} The former appears as a secondary ossification center of the dens between 3 and 6 years of age and normally fuses by the age of 12 years. Posterior C1–C2 screw and rod fixation with spacer or autologous corticocancellous bone is regarded as safe and efficient technique for fixation of AAD in terms of complications, biomechanical stability and fusion rate. Yeom et al showed that 8 of the 39 (20.5\%) C2 pedicle screws caused C1–C2 fusion in atlantoaxial instability, 21 (54\%) vertebral artery groove violation, and two of the 39 (5.1\%) C1 lateral mass screws showed evidence of cortical violation at C1 with one undetected vertebral artery injury.\textsuperscript{10}

In our patient 1, who was a female of fifth decade the C1–C2 instability was due to RA, and there is clear evidence of neurologic deficits. The RA is a slow and progressive chronic inflammatory pathology where anatomical supports at the CVJ lost over the time. C1–C2 anterolisthesis and retrolisthesis may cause cervicomedullary injury both from anterior and posterior aspects and commonly leads to reducible AAD.

The patient of os odontoideum in adult variety presented to the hospital with trivial history of trauma or fall, which may be often ignored as cause of neurological deterioration. The os odontoideum is a congenital anomaly/posttraumatic event of C2 vertebra, in which the odontoid process is separated from the body of the axis by a transverse gap, and it is a smooth, well-corticated ossicle.\textsuperscript{18} An “orthotopic” type is closely approximated to C1 anterior arch and moves in unison with it, and a “dystopic” one is intimately related to the clivus. In our case series following posterior C1–C2 fixation (polyaxial screw and rod) ± fusion, we found the best short-term clinical outcomes, and sufficient literatures support the satisfactory fusion rate over time. However, successful outcome depends upon preoperative assessment and meticulous surgical technique. As mentioned earlier, we found satisfactory screw placement, alignment and reductions intraoperatively, which were confirmed by postoperative images too. We found current technique most versatile, simple and technically sound, which allows intraoperative direct manipulation of C1–C2 and does not require continuous fluoroscopic imaging. One more important aspect of this technique is there is less risk of vertebral artery injury because of trajectory of screw directed toward medial and superior and no facet joint destruction when we compared with transarticular screw fixation (Magerl). Also, transarticular screw fixation is technically demanding and requires continuous fluoroscopy.

There is general consensus that type I and type III odontoid fracture can be managed with hard cervical collar (rigid cervical orthosis) in most cases. Type II odontoid fracture is usually associated with AAD (most common is type-IIA), having lower union rate. The current treatment modalities for type-II odontoid fracture include anterior (transoral odontoiodectomy, odontoid screw) and posterior instrumentation (C1–C2 arthrosis, occiput cervical fusion). Other modalities of treatment include rigid cervical orthosis and halo vest immobilization. In young patients with acute type-II odontoid fracture, posterior C1–C2 fixation without fusion may be sufficient for fracture healing. The factors affecting healing of type II odontoid fracture included age, bone density, types and classification of fracture (degree of displacement and comminution), old fracture, and smoking.

The purpose of cervical stabilization is to provide symptoms free life, stability of craniospinal axis and alignment, and halt the progression of neurological deficits by relieving the compression to spinal cord.

A purely traumatic AAD in the absence of odontoid fracture and associated other osseous injury is rare. Traumatic AAD is due to high velocity injury following disruptions of transverse ligaments. If we compare occipitocervical fusion (OCF) and atlantoaxial fusion (AAF), AAF provides greater range of motion at C0–C1 segment and better subjective satisfaction to the patients. However, clinical outcome, complication rate and infection study conducted in elderly population in between above two procedures appear to be same.\textsuperscript{15}

Again, deciding the treatment for type II odontoid fracture remains upon the type of fracture, local pathologies, and surgeon’s preference. To date, there are no well-controlled trials and studies to infer the true merits and demerits of the different treatment and approaches. So, no single ideal technique is found to be so effective for the treatment of odontoid fracture.\textsuperscript{36}

\section*{Conclusion}

Posterior C1–C2 screw and rod fixation (Goel–Harms) can be regarded as safe and efficient technique for fixation of AAD in terms of minimal complications, more biomechanical stability, increased pull out strength and fusion rate. This technique may be considered as procedure of choice when it satisfies the criteria. Early detection of atlantoaxial dislocation is the key for successful management of the atlantoaxial dislocation in terms of symptomatic and neurological improvement and halting the progression of the spinal cord disease process.

\section*{Conflict of Interest}

None declared.

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