

# **Os-Odontoideum leading to Atlanto-Axial instability — Report of surgery in four cases**

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### **Abstract**

Os odontoideum can lead to atlantoaxial instability, which can be either reducible or a fixed dislocation. We present surgical management in four patients with os odontoideum at our center. Two of these had reducible dislocations and were managed by posterior transarticular screw fixation. Other two had fixed dislocations necessitating posterior decompression and occipitocervical fixation.

### **Introduction**

Os odontoideum is the commonest anomaly of odontoid process.<sup>1</sup> It can lead to instability of the

atlantoaxial joint and places the spinal cord at significant risk for injuries even after minor trauma.<sup>2</sup> There is controversy whether it is truly congenital like in Down's syndrome or has traumatic etiology.<sup>3-5</sup> We are presenting four cases of Os odontoideum that came with atlantoaxial instability and were managed surgically. First two cases had reducible instability while last two cases had fixed dislocation at C1/C2.

### **Case Reports**

#### **Case No: 1**

A seven year old boy presented with one year history of repeated falls and progressive weakness of all four limbs.

Clinically he was able to walk but he had spasticity of limbs and there were upper motor neuron signs with brisk reflexes in all limbs. Neck movement was slightly reduced and painful at extremes. His x-rays of cervical spine revealed a reducible dislocation of atlanto-axial joint. MRI showed os odontoideum which was orthotopic in position. Posterior trans-articular screw fixation was done by Mageral's method. Post operatively patient was placed in hard cervical collar for six weeks. His recovery was un-eventful and in three months he regained full motor power and resolution of brisk reflexes. X-rays at last follow up after one year showed solid fusion.

### Case No: 2

An eight year old boy presented with history of headaches and repeated falls after minor stumbles with stiffness of all limbs recovering with few days of bed rest. His dynamic x-rays and MRI revealed a reducible atlanto-axial instability secondary to os odontoideum. He underwent posterior trans-articular screw fixation of C1/2. Post operatively he was placed in hard cervical collar for six weeks. He had an un-eventful recovery with full neurological and clinical improvement. (Fig 1). X-rays taken at six months revealed solid fusion.



Fig 1: Pre op dynamic lateral x-rays, showing os odontoideum with instability (a and b). Pre-op MRI showing os-odontoideum (c). Intra-op picture showing trans-articular screws with bone graft anchored with non-absorbable sutures (d). Post-op lateral and open mouth views showing trans-articular screws and bone graft (e and f).

### Case No: 3

An eighteen years old mentally retarded boy with delayed milestones presented with history of headache, torticollis, neck pain, repeated falls and transient

weakness of the limbs. X rays of the cervical spine showed os odontoideum with partially reducible atlantoaxial instability on flexion/ extension radiographs. MRI of cervical spine showed os odontoideum and atlantoaxial dislocation with posterior displacement of odontoid and marked compression of spinal cord. Anterior cervical spine surgery was planned which was refused and dislocation was partially reduced with traction and patient was discharged on a cervical collar. He came back with tetraparesis after a minor fall. He improved neurologically on traction but minimal reduction could be achieved. Posterior surgery was performed and trans-articular screw fixation was tried but was unsuccessful because of the fixed deformity and curvature of the spine. Gallies Fusion of C1/2 was done. Postoperatively patient developed tetraplegia while he was being transferred to X- Ray department. His X- rays showed inadequate reduction with gross compression. Immediate revision surgery with occipito-cervical fusion was planned. Old incision was opened. Wire passed around C1 posterior arch, was pulled to achieve partial reduction and decompression. Old bone graft plus BCP (bone graft substitute including tricalcium phosphate and calcium hydroxyapatite) was applied. Post operatively patient was placed in hard cervical collar for eight weeks. Intensive rehabilitation was started. On last follow up, one year after surgery, patient had muscle power of grade 4/5 and was walking with support.

### Case No: 4

A twelve year old boy presented with vague history of dizziness and repeated falls for the last one and a half year. He had severe headaches and progressive weakness of all limbs. He was admitted at a local hospital and investigated. Ultimately he was referred to our center. He had grade four power in all limbs, spasticity and brisk reflexes. X-rays of cervical spine with flexion/ extension views revealed fixed dislocation of atlanto-axial joint. MRI revealed marked compression of upper cervical spinal cord. He was placed on skull traction but dislocation could not be reduced. Trans-oral decompression/reduction followed by occipito-cervical fusion were planned but patient's relatives did not agree to anterior procedure hence only posterior surgery was done. Posterior reduction was tried but did not succeed and ultimately posterior decompression followed by occipito-cervical fusion was done. Post-operatively patient was placed in hard cervical collar for eight weeks. Child improved neurologically and ultimately gained full motor power in all limbs within six weeks (Fig 2). On last follow up after one year he was asymptomatic and had solid fusion on x-rays.



Fig 2: Pre-op dynamic x-rays of cervical spine showing fixed atlantoaxial dislocation with os-odontoideum (a and b), Pre-op MRI (c). Intra-op picture showing occipito-cervical fusion with two recon plates of 3.5mm each with posterior decompression at level of foramen magnum and posterior arch of atlas (d). Post-op x-rays (e and f).

## Discussion

Atlanto-axial instability is defined as more than three millimeters of motion at level of C1/C2 joint on lateral flexion/ extension x-ray films.<sup>1</sup> It is of three types: type 1 which is due to odontoid anomalies, type 2 is due to ligament deficiencies and type 3 is rotatory fixation.<sup>6</sup> Congenital anomalies of Odontoid process include aplasia, hypoplasia and Os Odontoideum. In Os odontoideum, Odontoid remains as a separate ossicle from axis. It can remain at its normal place (orthotopic) or migrate near foramen magnum (dystopic).<sup>6</sup> Os odontoideum usually remains asymptomatic and is brought to notice after minor trauma leading to atlanto axial instability, as demonstrated in our cases. Once there is clear cut atlanto axial instability manifested by abnormal movement at atlanto axial joint, invariably

operative treatment is required. There are various methods of stabilization described in literature. If reduction can be achieved pre-operatively by traction, Magerl's transarticular screw fixation is a reasonable fixation method and is a superior biomechanical construct as compared to wiring techniques.<sup>7</sup> If there is associated peg fracture, anterior trans-articular screw fixation with peg screw has been described.<sup>8</sup> However, if reduction cannot be achieved, then in persistent cord compression, anterior decompression by trans-oral or antero-lateral cervical approach has been suggested. Harms has described C1/2 fixation by multi-axial pedicle screws fixed with a rod. Harm's technique is useful in incompletely reduced dislocations. However it is technically demanding as there is a lot of bleeding from the plexus at posterior articulation of C1/C2, endangering the 2nd posterior root.<sup>9</sup> Recently successful anterior reduction of fixed dislocation by soft tissue release followed by posterior occipito-cervical fixation has also been considered.<sup>10</sup> Our third and fourth cases show that in difficult cases, posterior decompression followed by occipito-cervical fixation can be a valid option.

## References

1. Fielding JW, Hensinger RN & Hawkins RJ. Os Odontoideum. *J Bone & Joint Surgery*, 1980 62; 376-83.
2. William CWJ. Pediatric Cervical Spine. In: Canale ST Ed. *Campbell's Operative Orthopaedics*. 10th Ed. Philadelphia: Mosby, 2003, pp 1715-1750.
3. Riaz S, Drake JM, Hedden DM. Images in spine surgery: atlantoaxial instability in Down syndrome. *J Pak Med Assoc*. 2007; 57:213-15.
4. Sankar WN, Wills BP, Dormans JP, Drummond DS. Os odontoideum revisited: the case for a multifactorial etiology. *Spine* 2006; 31: 979-84.
5. Fagan AB, Askin GN, Earwaker JW. The jigsaw sign. A reliable indicator of congenital aetiology in os odontoideum. *Eur Spine J*. 2004; 13:295-300. Epub 2004.
6. Fielding JW, Hawkins J, Ratzan SA. Spine fusion for atlanto-axial instability. *J Bone Joint Surg Am*. 1976; 58:400-07.
7. Reilly CW, Choit RL. Transarticular screws in the management of C1-C2 instability in children. *J Pediatr Orthop*. 2006; 26:582-88.
8. Reindl R, Sen M, Aebi M. Anterior instrumentation for traumatic C1-C2 instability. *Spine*. 2003; 28: E329-33.
9. Harms J, Melcher RP. Posterior C1-C2 fusion with polyaxial screw and rod fixation. *Spine*. 2001; 26:2467-71.
10. Wang C, Yan M, Zhou HT, Wang SL, Dang GT. Open reduction of irreducible atlantoaxial dislocation by transoral anterior atlantoaxial release and posterior internal fixation. *Spine*. 2006; 31:E306-13.