Management of Atlanto-axial Dislocation, A Case Based Approach: An Institutional Experience

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Abstract
Introduction: Although many different techniques have been described for management of AAD, fixed or mobile varieties, no single technique is sufficient to manage all types of AADs as anatomical variations are many a times different in patients resulting in the tailoring the procedure for the associated deformity correction. Although radiological imaging allows diagnosis of abnormality, many times planned procedure is modified. In our series we describe the various techniques of management as well as rescue operations in case of failure of planned procedure.

Methods: It is a retrospective study extended over a period of 10 years i.e. from January 2007 to December 2017. The clinical parameters which were used are demographics, visual analogue scale for assessment of pain & Nurick’s grading for neurological deficit. Apart from dynamic flexion & extension neck X-ray & CT scan, MRI craniovertebral junction, 3D CT angiogram of neck vessels was used for detailed anatomy & pathological assessment. In a patient with AAD associated with congenital anomalies, 3D print was used in perioperative period. Improvement in the neurological deficit is assessed & compared after 6 month follow up.

Results: Total 154 patients of AAD are operated over a period of 10 years. Most of these patients were presented with neck pain and occipital headache (86.5%) followed by mono or hemi or quadriparesis (66.7%). In initial period of experience, most of these patients were managed by simple sub-laminar techniques like Gallie’s or Brooke’s fusion technique. But with gain in experience, there is gradual shift to more robust construct techniques like C1 lateral mass and C2 pedicle screw and rod technique or occipito C2-C3 fusion technique or Magerl’s technique. Vertebral artery injury occurred in two patients while placement of C1-C2 transarticular screw while one patient had a vertebral artery injury while placement of C2 pedicle screw. Though these techniques give more robust fixation, due to anatomical variation and intraoperative failure of planned procedure, other conventional and bailout techniques like occipitocervical fixation using Hartshill and sub-laminar wires or Writer’s technique or Brooke’s technique are used.

Conclusion: Each case of AAD has to be evaluated with CT images with 3D reconstruction. 3D printing aids in decision making and also in cases with congenital anomalies like Klippel-Feil syndrome where there is high risk of vertebral artery injury due to anomalous course. Though there are many techniques described for the management of AAD, it is essential to have all basic techniques in an armamentarium as these procedures may be useful as bail-out techniques.

Abbreviations
AAD: Atlanto Axial Dislocation; BI: Basilar Invagination; 3D CT: 3 Dimensional Computed Tomography; MRI: Magnetic Resonance Imaging; CMJ: Cervicomedullary Junction; CVJ: Craniovertebral Junction; CSF: Cerebrospinal Fluid; VA: Vertebral Artery

Introduction
Atlantoaxial dislocation is one of the difficult pathology which has been managed by different techniques over a period of time. The atlantoaxial joints can lose stable articulation from traumatic, inflammatory, idiopathic, or congenital abnormalities. There was a gradual evolution in the diagnostic techniques as well as therapeutic modalities in past few years. There are many studies which describe the various operative techniques for the treatment of Atlantoaxial Dislocation (AAD). Most of the studies describe the C1-C2 fusion by posterior approach. There are some studies which dealt the AAD via anterior approach. But no particular study has determined which approach or technique is more effective in achieving the aims of AAD treatment, such as relieving cervicomedullary junction (CMJ) compression, restoring the normal alignment of CVJ with reconstruction of stability. Here...
we share our experience of ten years in the management of AAD which incorporates gradual evolvement of various techniques from sublaminar wiring to C1-C2 fusion using rod & screw construct.

**Materials and Methods**

One fifty four patients with AAD (118 males and 36 females, aged 8 to 69 years with an average age of 38.7 years) underwent C1-C2 or occipito-C2-C3 fusion under general anesthesia (Table 1). Each surgical procedure was performed by the same experienced senior neurosurgeon. Our indication to perform surgery was neurologic deficit (myelopathy) with CMJ compression. The clinical presentations are summarized in Table 2. Neurologic deficit duration was between 6 and 55 months (average ~ 23 months). Anteroposterior, lateral and dynamic plain X-rays, Computed Tomography (CT) scans with 3D reconstruction views of CVJ, CT angiogram and Magnetic Resonance Imaging (MRI) were obtained in all patients preoperatively. Chamberlain, McRae, and Wackenheim lines were surveyed, and all measured data met the diagnostic criteria of AAD with BI. Awake intubation using fiber-optic bronchoscope in neutral position was done in all patients. Skeletal traction was applied to all patients in neutral position (Figure 1). For mobile variety, C1-C2 fusion has done using sublaminar wiring (Brooke’s technique) or transarticular screw placement (Magrel’s technique). If there is significant anterior compression, transoral odontoidectomy performed followed by posterior fixation. For non-mobile variety, C1-C2 fusion carried out using C1 pars screw & C2 pedicle screws & rods (Goel&Harms). In patients, where screw placement failed, a C1-C2 sublaminar wiring with fixation was carried out. Occipito-cervical fixation with hartshill & wires was also used as a rescue operation. All of the patients were assessed clinically to observe neurologic recovery in terms of improvement in Nurick’s grade following surgery. Improvement in Nurick’s grade was analyzed at 6 months interval. Complications were documented in the form of intraoperative, immediate & delayed ones.

**Results**

Total 154 patients of AAD were operated in 10 year period with male to female ratio of 3.1:1. Most of these patients were between 10 to 30 years of age group. Main symptom at the presentation was neck pain & occipital headache (86.5%) followed by neurological deficit in the form of monoparesis or hemiparesis or quadriparesis (66.7%) (Table 2). Sensory abnormalities seen in 63.6% patients. Short neck, low hairline and limitation of neck motion is observed in 30.2%. Head tilt is observed in 42.6% patients. Most of the patients were in Nurick’s grade 1 or 2 of neurological deficit (Table 3). At the initial phases of surgical management of AAD, most of the patients were managed by Gallie’s fusion [1]. In Brooke’s method of C1-C2 fusion, two bone grafts are fashioned to wedge between C1 & C2 after decortication of C1 arch & C2 lamina. But with gaining more experience, there was a gradual shift from sublaminar wiring technique to more robust construct like C1-C2 fusion using rods and screws over last few years (Table 4). In last 2 years, there were only 4 patients who were operated by Brooke’s technique (Table 5). Out of these four patients, two patients had complex CV junction anatomy, one patient was switched from conventional C1-C2 screw & rod fixation to sublaminar wiring due to vertebral artery injury & one patient had a significant morbidity prompting to reduction in surgical time & selection of more simple & safer technique of sublaminar wiring. Out of 29 patients operated by sublaminar wiring, two patients had a dural tear intraoperatively which was managed.
success...
Eight patients had wound infection, seven of which were managed conservatively while one patient had undergone Hartshill implant removal. This patient was managed without re-implantation as there was evidence of C1-C2 fusion in follow up period. Five patients had implant failure. Four of which are wire related & one due to failure of C1 pars screw. Three out of four patients had breakage of sublaminar wire (Figure 6) while one patient had a breakage of wire used for Hartshill fixation.

Two patients died in follow up period by cause unrelated to the procedure & twelve patients lost to follow up period (Table 6). There was significant improvement in the visual analogue scale in patients having moderate to severe neck pain in follow up period of six months (Table 7). 119 patients showed improvement in Nurick’s grade in six month follow up period while 14 patients remained same as preoperative status. Eight patients found to be deteriorated in post-operative follow up period (Table 8, 9). 11 patients out of 14 who were remained same were primarily treated by sublaminar wiring & three were treated by occipito-C2-C3 fusion. Six out of eight patients who had deteriorated were having traumatic atlanto-axial dislocation & were not found to be associated with any specific mode of surgery. Two of three patients who had vertebral artery injury while placement of transarticular screws while one patient had a vertebral artery injury while placement of C2 pedicle screw. In this patient, screws are placed only on the side of vertebral artery injury & procedure abandoned for further evaluation of injured vertebral artery (Figure 7).

Discussion

Surgical treatments are required in most of the patients with atlanto-axial dislocation with continuing neurological deficit [6-13]. Though there is improvements in the surgical techniques have brought some encouraging results over the years, there is no agreement among specialists regarding the appropriate surgical procedures that should be used for treating CVJ anomalies and reconstructing...
stability. Simple posterior sublaminar wiring techniques 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. When we perform posterior wiring techniques, we prefer to use double braided titanium wires because they are more flexible than steel wire and have less chance of causing dural or neural injury.

Gallie in 1939 first described posterior C1-C2 sublaminar wire fixation with the use of steel wire [1]. In the Gallie technique, a single autograft harvested from the iliac crest is notched inferiorly and placed between the C2 spinous process and the posterior arch of C1. The graft is held in place by a sublaminar steel wire that passes beneath the C1 arch and then wraps around the spinous process of C2. By avoiding the Passage of the sublaminar wire under the lamina of C2, there is decrease the risk of neural or dural injury. There is increased rate of non-union in Gallie fusion up to 25% since it offers good stability only in flexion & extension and there is poor stability in rotational maneuvers [14]. Unlike Gallie fusion technique, in Brooks and Jenkins fusion, two separate iliac grafts are obtained & wedged between C1 arch and C2 lamina on either side of the midline. These grafts are then secured in place by placing the sublaminar cables on each side wrapping around these grafts. The cables are then tightened around the grafts and cramped in place. This technique not only provides stability in flexion and extension but also more rotational stability than Gallie’s fusion technique. Therefore this procedure has more fusion rate as high as 93% and is improved by halo immobilization [15-18]. But there is increased risk of dural tear or neurological injury in this procedure as compared to Gallie’s, because two wires are passed beneath the C1 arch and C2 lamina [16]. Transarticular screw fixation was first described by Roy Camille in 1972 [19]. But it is popularized by Magelr & Jeanneret. In 1979, Magerl described this technique for odontoid fractures. But it has widely being used for C1-C2 inflammatory disease, infections, congenital anomalies and acquired postsurgical deformities [2,3]. The main advantage of this technique is complete obliteration of rotational movement at C1-C2 joint. As these screws provide adequate internal immobilization, external immobilization is not required. Disadvantage of this technique is steep learning curve and there is a risk of vertebral artery injury, spinal cord or hypoglossal nerve injury. This risk significantly increases in patient with anomalous vertebral artery course as in patients with congenital anomalies like Klippel-Feil syndrome. Vertebral artery injury is still a risk with C2 pars screws but the risk is not as high as with transarticular screws. If a fusion is to be performed: the posterior arch of C1 and the C2 lamina are decorticated with a drill. Only fusion substrate is then placed, taking care not to compress the dura. Joint surface is drilled and bone is packed within the C1–2 joint. Coagulation around C2 ganglion can prevent venous bleeding. Cutting C2 ganglia facilitates opening and packing the C1–2 joint with bone. Stepped approach with 2 mm drill bit under fluoroscopic guidance helps in preventing wrong trajectory for screw placement. Occipitocervical fusion technique can be carried out using Hartshill & sublaminar wires or by occipital keel plate, screw and rod construct. In Hartshill and sublaminar wire fusion

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technique avoids vertebral artery injury and relatively easier bail out technique if there is difficulty in placement of C1-C2 screws or due to abnormal anatomy at CV junction. In this technique, Hartshill is moulded according curvature at the CV junction in neutral position and is fixed translaminar screws [25, 26] 1 year stability appears to be less than C2 pedicle screws when used for subaxial fusions, but was as effective for axial fusions (C1–2 or C1–3) [27]. May be useful as a “bailout” for subaxial fusions when the C2 pars diameter is too small for pedicle screws or if there is pedicle compromise during C2 screw placement [27].

Conclusion

Each case of AAD has to be evaluated with CT images with 3D reconstruction. 3D printing aids in decision making and also in cases with congenital anomalies like Klippel-Feil syndrome where there is high risk of vertebral artery injury due to anomalous course. Though there are many techniques described for the management of AAD, it is essential to have all basic techniques in an armamentarium as these procedures may be useful as bail out techniques.

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