Retrospective Study of Craniovertebral Junction (CVJ) Anomalies: A Clinical Profile and Outcome Analysis of Surgically Treated Patients.

Craniovertebral junction (CVJ) is a complex anatomic region providing stability and mobility to the most important part of the craniospinal axis. The purpose of this study is to analyse clinical characteristics and outcome after surgical management of patients with CVJ anomalies presented to Neurosurgery department, Bir hospital Kathmandu Nepal. A retrospective analysis of 21 patients, managed surgically for craniovertebral instability between 2013 and 2017, was performed. Imaging studies were reviewed for bony and soft tissue details. Patients managed with posterior approach alone (either occipitocervical fusion or C1-2 fusion with or without bony decompression) were included in the study. Outcome was assessed by comparing pre and post operative Nurick grade. Most common causes of CVJ instability were non union of old odontoid fracture (38.1%) and OsOdontoidium (38.1%). 76.2% had intramedullary high signal intensities in T2 weighted MRI while 90% had cervicomedullary compression. Pre operatively, 52.38% had Nurick grading scale of 4 to 5 while 47.62% had 0-3 Nurick grade scale. Post operatively, 71.42% improved clinically, 23.80% remained unchanged while one patient deteriorated. Mean duration of follow up was 20.87 months (6-60 months). Pre operative Nurick grade was significantly associated with post operative outcome (p=0.042). Early surgical intervention is associated with better clinical outcome. Larger prospective study with clinical and radiological follow up is recommended.

Key words: craniovertebral junction anomalies, outcome, posterior fusion
Craniovertebral junction (CVJ) is a complex anatomical region extending from occipital bone to C2/3 disc space. It is a transition zone between skull base and the cervical spine, which protects many neurovascular structures including medulla, spinal cord, and lower cranial nerves and vertebral arteries. Unique embryological developmental process and the highest mobility led to many congenital and acquired anomalies. Common anomalies include atlantoaxial subluxation, basilar invagination, Os odontoïdium, Chiari malformation, occipitization of atlas, and many other associated with hypoplasia or dysgenesis. Odontoid fractures are the common injuries associated with significant instability, non union and myelopathy.\textsuperscript{5,8,9,13,14}

Clinical presentation greatly varies. Most individuals present with either motor, sensory, lower cranial nerves and brain stem symptoms or vascular compromise.\textsuperscript{17} Diagnosis is made with static and dynamic x ray of upper cervical spine, computed tomography (CT) and magnetic resonance imaging (MRI) studies. Vertebral artery (VA) angiogram (either conventional catheter angiogram or CT-A) is a useful adjunct to rule out aberrant course avoiding inadvertent intra operative injury.

Various treatment strategies have been described in the literature. If the atlantoaxial joint is mobile and reducible, atlantoaxial fixation was advocated while anterior transoral decompression or posterior foramen magnum decompression was advocated for irreducible atlantoaxial subluxation. The dictum of anterior surgery for an anterior compressive lesion and posterior surgery for posterior compression was established.\textsuperscript{5,11} However, of lately, posterior approach alone with C1-2 distraction, realignment and stabilization is getting more favour. In 1994, Atul et al suggested an alternative plate and screw technique of fixation of the lateral masses of C1 and C2 vertebrae.\textsuperscript{5} Inclusion of occiput in fixation construct is also considered by some but craniovertebral junction stabilization techniques is now focusing more on atlantoaxial joint alone.\textsuperscript{6}

The purpose of this study was to analyse the clinical characteristics and outcome of patients with craniovertebral instability managed with posterior surgery with or without posterior decompression.

Materials and methods:

We reviewed 21 patients with craniovertebral junction anomalies operated at Neurosurgery department Bir Hospital between January 2013 and December 2017. We included those CVJ anomalies who were managed surgically with posterior approach alone either occipitocervical or atlantoaxial fixation and fusion. Isolated chiari malformation without atlanto-axial subluxation (AAS) or basilar invagination (BI) which was managed with posterior fossa decompression alone were excluded. Clinical and radiological reports and operative records were reviewed. All patients were clinically assessed and categorized according to Nurick grade. Imaging studies (X ray cervical spine, CT and MRI) were done to delineate bony and soft tissue details. Atlantoaxial subluxation was defined as atlanto-dental interval (ADI) 3 mm or more in a lateral flexion/extension cervical radiograph. Different craniometric lines such as McRay, Chamberlane, Wakenheimclival line, were used to define basilar invagination. All patients were followed with clinical assessment and cervical x ray to confirm the position of implants and alignment of bony elements. Outcome was assessed by comparing Pre and Post operative Nurick grade. Ethical approval was taken from institutional review board of the hospital. Statistical analysis was done using SPSS version 17.

Surgical technique:

Cervical traction was set up prior to general anaesthesia and endotracheal intubation. Patient was placed prone with chest and pelvic support with head end of the table elevated to 25-30 degrees. Suboccipital region and upper cervical spine were exposed through longitudinal skin incision from inion down to C4 spinous process. Occipital bone and the posterolateral elements of C1 and C2 exposed. Lateral mass of C1 vertebra was exposed by doing subperiosteal dissection. A portion of inferior part of posterior arch of C1 was drilled and the venous plexus and C2 dorsal root ganglia retracted caudally for better visualization of lateral mass. Bleeding from venous plexus was controlled with sustained compression using surgicel (oxidised regenerated cellulose) and gelfoam. Under fluoroscopy guidance, lateral mass screw on C1 and pars or pedicle screw on C2 were place bilaterally. Occipital plate was used for occipito cervical fusion. Rods were loosely fixed between the screw heads, axial traction from the head end was applied to reduce the deformity and then odontoid was pushed anteriorly (under fluoroscopy) before the final fixation. We were able to achieve satisfactory reduction by this technique as confirmed by intra operative live fluoroscopy images. Posterior margin of the foramen magnum was removed in cases where decompression was not adequate. Corticocancellous bone graft was harvested from posterior iliac crest and laid down along the decorticated posterolateral elements of C1 and C2 for fusion.

Results:

Of the 21 cases, majority were male patients (66.66%). Mean age group was 40.71 years (16 to 73 years).
## Cranovertebral Junction (CVJ) Anomalies

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Associated anomaly</th>
<th>Operation</th>
<th>Follow up period (mos)</th>
<th>Nurick grade</th>
<th>Pre op MRI</th>
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*Table 1 Patients’ characteristics.  * IHSI-Intramedullary High Signal Intensity, ** CMC-Cervicomedullary Compression*
Table 1 shows the clinical data of 21 patients managed surgically. Most common causes of upper cervical instability were old odontoid fracture with non union (n=8, 38.1%), Osodontoidium (n=8, 38.1%) followed by basilar invagination (n=3, 14.28%), potts’ spine of C2 vertebra (n=1,4.76%) and congenial atlantoaxial subluxation (n=1, 4.76%) (Figure 1,2 3). The average duration of symptoms was 35.76 months (5-120 months).

Disability was evaluated with Nurick grade. 52.38% were dependent while 47.62% were independent pre operatively. Imaging study showed intramedullary high signal intensity (IHSI) in 76.2% and cervicomedullary compression in more than 90% cases. All patients were managed with posterior approach: C1-2 fusion (n=17) and occipitocervical fusion (n=3). Follow up period ranges from 6 months to 5 years (Mean= 20.87 months). On follow up, 71.42% (n=15) had clinical improvement, 23.80% (n=5) remained static while 1 patient deteriorated (Table 3). Mean duration of illness among improved was 28.5 months while it was 40.8 months among those who did not show clinical improvement. Among improved, 66.67% had intramedullary high signal intensity while it was present in all cases among not improved. All patients with pre operative Nurick grade <=3 had some clinical improvement while only 45.45% improved from among Nurick grade >=4. Unadjusted regression analysis did not show any significant correlation of outcome with age, sex, duration of illness, intramedullary high signal intensity except for pre operative Nurick grade (p=0.042). Most common complication was occipital neuralgia (n=6). One patient required revision surgery due to malposition of screw (Table 2).

Discussion

In our study, most common causes of upper cervical instability were non union of odontoid fracture and the Osodontoidium (76.19%). Clinical improvement was
observed in 71.42% while 23.8% remained static and one patient deteriorated after surgical intervention. Patients with long duration of illness, intramedullary high signal intensity in T2 MRI image and patients with high Nurick grade at presentation tend to have unfavourable outcome. In unadjusted regression analysis, no variables (age, sex, diagnosis, duration of illness and high T2 signal intensity changes) showed statistical correlation with outcome except pre operative Nurick grade. Patients with low Nurick grade at presentation had better clinical outcome ($p=0.042$)

Song G-Chang et al analyzed 32 patients with CVJ instability, managed surgically and followed up for mean duration of 38 months. They found that 78% had clinical improvement, 28.1% remained static and in 93% good fusion was achieved which was consistent with our findings in which patients were followed up for mean duration of 21 months.
Paudel et al

Study by Sindgikar P et al found that 55 (13%) patients (out of 414) required re surgery after primary CVJ surgeries. A larger proportion of patients with irreducible Atlantoaxial dislocation (IrAAD) underwent reexploration compared to the patients with reducible atlantoaxial dislocation (RAAD) (45 vs 10, OR = 2.67, 95% CI = 0.51–14.03; P = 0.23). Among IrAAD, those associated with BI had a higher likelihood of undergoing resurgery (OR = 2.27, 95% CI = 0.54–9.47; P = 0.25). Common causes of revision surgery were implant related (32.7% - breakage of sublaminar wiring, hardware associated stenosis), infection, Osodontoidectomy with reducible AAD, and vertebral artery injury. Within the first year of primary CVJ surgery, infection was the major reason for revision while implant failure was more common after one year. Breakage of sublaminar wire was associated with 1.5 times higher risk of implant revision.

In contrast to their finding in which infection constituted 23.2% of resurgeries, 19% of our patients had superficial surgical site infection associated with pressure sore which was managed with regular dressing, frequent repositioning and local suture application.

Anterior vs. Posterior approach

Surgical approaches to CVJ region have been broadly categorized as anterior decompression only, or anterior decompression with posterolateral fusion or posterolateral fixation and fusion based on reducibility of atlantoaxial subluxation. The standard and most accepted form of treatment of Group A basilar invagination is a transoral decompression while the majority of the authors recommend a posterior occipitocervical fixation following the anterior decompression. Goel A et al have been advocating that posterior approach alone if done properly would be sufficient to reduce the BI and maintain the alignments. The atlantoaxial joints were maintained in a distracted and reduced position with the help of bone graft and spacers. Atlantoaxial joint forms the fulcrum of all movements and stabilizing the joint presents the best opportunity for fixating and arthrodesis. Direct atlantoaxial fixation provides the biomechanically strongest method of stabilization and the treatment protocol should shift towards craniovertebral realignment and fixation-arthrodesis rather than decompression and bone removal.

Occipitocervical vs. C1-2 fusion and Pseudoarthrosis

There are different opinions whether to include occiput in the fusion or not. It was shown that occiptoatlantoaxial fusion has high implant failure rate and high risk of infection compare to C1-2 fusion alone. Risk of implant failure might be attributable to asymmetric occipital squama, the long lever arm of the bony construct that led to mechanical failure or the contiguous involvement of the occipitoatlantoaxial joints in the former, rather than a single joint (C1-2) in the latter procedure. Similarly, inclusion of occiput, was associated with a 1.23 times risk of implant infection. This was usually due to dehiscence of the thin skin or development of a pressure sore at the level of external occipital protuberance.

Implant failure has been seen even after 2 decades of surgery suggesting that pseudoarthrosis is one of the important cause of disability after primary surgery. Inadequate bone decortication, failure to maintain sufficient bony contact between the autologous bone graft and the occipitocervical bone, inadequate neck immobilization, graft lysis, insufficient quantity of trabecular bone with inadequate osseous progenitor cells, or loosening of the metal construct due to improper bone purchase, osteoporosis, or subclinical infection have been suggested as risk factors for pseudoarthrosis.

Recent literature suggests that the inclusion of occipital bone in the fixation construct provides a suboptimal form of fixation, as it involves inclusion of a normal joint in the fixation assembly. Direct atlantoaxial fixation provides a segmental stabilization at the point of fulcrum of all movements, and hence the occipito cervical fusion may not be required in all cases to stabilize and decompress the cervicovertebral junction.

Limitation: This study has small sample size limiting adjusted regression analysis to establish statistical correlation of different variables with outcome. Because of financial and logistic constrains, detail imaging studies like CT scan and MRI could not be done in follow up for proper evaluation of bony morphology and fusion and adequacy of neural decompression. Preoperative vertebral artery angiogram was not done in all cases which would have helped to plan surgical technique and prevent inadvertent arterial injury. However, none of our patient had intraoperative vertebral artery injury or post operative posterior circulation stroke.

Conclusion

Craniovertebral junction anomalies could be managed with posterior approach achieving adequate decompression and stabilization. Early surgical intervention was associated with better clinical outcome in our study. Prospective study with larger sample size and detailed pre and post operative imagining studies would help for proper evaluation of clinical outcome and fusion after surgery.

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