Posterior occipitoaxial fusion for atlantoaxial dislocation associated with occipitalized atlas

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Congenital atlantoaxial dislocation (AAD) is caused by the malformation of the basioccipital bones, atlas, and axis and their supporting ligaments. Some patients with this abnormality lead a precarious existence in which even a trivial trauma could precipitate severe morbidity; other patients may face chronic myelopathy caused by the prolonged instability and secondary narrowing of the diameter of the foramen magnum. The clinical manifestations of AAD may be myriad. The patient may exhibit acute transitory attacks or progressive neurological deficits caused by upper cervical and/or bulbar paralysis. Often only cervical pain or restricted neck movements may be apparent. Atlantoaxial dislocation may also be associated with multiple bone and soft-tissue abnormalities.

Usually in cases of AAD, posterior C1–2 fusion is performed to provide stability. However, 40% of patients with congenital AAD have associated occipitalization of the atlas and some patients have a thin or deficient posterior arch of the atlas. These patients require an occipitocervical fusion for stability because a posterior C1–2 fusion is not feasible.

Although the first occipitocervical fusion using onlay bone graft was performed by Foerster in 1927, to date no consensus has been reached regarding a single method for the attainment of occipitocervical stability.

In this article we present our experience with a technically simple method of occipitoaxial posterior fusion described by Jain, et al., in which a rigid internal fixation is achieved by fusing an artificial arch of atlas, created from the occipital bone along the margin of the foramen magnum, with the axis by means of sublaminar wire and interposed strut and lateral onlay bone grafts.

Clinical Material and Methods

Patient Population

The fifty patients included in the study suffered from congenital AAD with occipitalized atlas (46 patients) or thin or deficient posterior arch of the atlas (four patients). Between 1989 and 1994 these patients underwent occipitocervical fusion performed using the technique described by Jain, et al.
tion was 27.8 years (range 5–70 years). There was an overwhelming male predominance (male/female ratio 42:8) in this group of patients.

**Clinical Examination**

The neurological presentation of the patients included pyramidal signs (weakness, spasticity) in 46 patients (92%), focal distal upper-limb wasting in five (10%), posterior column deficits in 40 (80%), spinothalamic tract derangement in 27 (54%), bulbar signs (fifth, seventh, ninth, and 10th cranial nerve palsies) in six (12%), cerebellovestibular signs (unsteady gait, trunk and limb ataxia, nystagmus) in eight (16%), sphincter disturbances in 10 (20%), and respiratory discomfort in one (2%). Twenty-six patients (52%) had restricted neck movements. Specific stigmata (short neck, low hairline, facial or hand asymmetry, high arched palate) were present in nine patients (18%). Thirteen patients (26%) had a history of clinical deterioration following minor trauma, and two patients (4%) had transitory unconsciousness. The duration of symptoms experienced by the patients as of the time of presentation averaged 9 months with a range of 20 days to 10 years.

The preoperative neurological status of the patients was assessed by the method used by Di Lorenzo. Ten patients (20%) were totally dependent on family assistance (severe disability; Grade IV); 21 (42%) were partially dependent for their daily needs (moderate disability; Grade III), and 14 (28%) were independent with minor pyramidal signs (diffuse hyperreflexia and spasticity) (minor disability; Grade II). Five patients (10%) were neurologically intact (Grade I) and presented after experiencing an attack of transient quadripareisis (Table 1).

**Radiological Examination**

All of the patients underwent plain radiographs of the craniovertebral junction (lateral view, flexion and extension and open mouth view of the atlantoaxial joint) and dynamic tomograms (in flexion and extension). Multiplanar computerized tomographic myelography or magnetic resonance (MR) imaging was performed in 32 patients (64%) for further evaluation of soft-tissue abnormalities and cervicomedullary compression. Arnold Chiari malformation was detected in eight patients (16%) and cervical syrinx in two (4%). Other associated bone anomalies included basilar impression (40%), Klippel–Feil syndrome (56%), dens dysplasia (ossiculum terminale, os odontoideum and hypoplasia–aplasia) (26%), condylar hypoplasia (18%), and platybasia (6%). Based on flexion–extension views of the craniovertebral junction, patients were categorized into two groups, those having AAD reducible on extension and those with irreducible AAD.

**Perioperative Management**

In all patients having irreducible AAD, reduction was attempted using Crutchfield’s tong traction starting with 7% to 8% of body weight with graded increases to a maximum of 7 kg. Sequential lateral cervical radiographs were obtained to monitor the reduction. In 29 patients the dislocation remained irreducible during traction for 2 weeks. The final treatment plan was decided at this stage. The surgical procedures undertaken are summarized in Table 2. Posterior fusion alone was performed in patients with AAD reducible on extension or after traction (21 patients, 42%).

In patients in whom the deformity was irreducible with or without associated basilar impression (22 patients, 44%), posterior fusion was preceded by ventral decompression (transoral–transpharyngeal odontoidectomy). However, in seven patients (14%) who showed marked clinical improvement on traction, despite radiological evidence of persisting AAD, direct occipitocervical fusion was performed without anterior decompression.

**Operative Technique**

In the initial 15 cases occipitocervical fusion was performed strictly according to the method described by Jain, et al. In subsequent patients, a few modifications were made in the technique.

General anesthesia is induced, and surgery is performed with the patient lying prone with the head resting on a horseshoe head rest. Perioperative reduction and stability

### TABLE 1

<table>
<thead>
<tr>
<th>Preop Grade &amp; Disability†</th>
<th>No. of Preop Patients</th>
<th>No. of Patients Available at Follow Up</th>
<th>Neurological Status at Follow Up</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Improved</td>
</tr>
<tr>
<td>I. independent</td>
<td>5</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>II. independent</td>
<td>14</td>
<td>13</td>
<td>9</td>
</tr>
<tr>
<td>with minor disability</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>III. partially dependent</td>
<td>21</td>
<td>19</td>
<td>13</td>
</tr>
<tr>
<td>IV. totally dependent</td>
<td>10</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>total (%)</td>
<td>50</td>
<td>43</td>
<td>31</td>
</tr>
</tbody>
</table>

* Seven patients were lost to follow up.
† Grade according to the scale of Di Lorenzo.

### TABLE 2

<table>
<thead>
<tr>
<th>Surgical Indications</th>
<th>Occipitocervical Fusion</th>
<th>Transoral Odontoidectomy &amp; Occipitocervical Fusion</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. (%)</td>
<td>No. (%)</td>
<td>No. (%)</td>
</tr>
<tr>
<td>reducible AAD</td>
<td>21 (42)</td>
<td>0 (0)</td>
<td>21 (42)</td>
</tr>
<tr>
<td>irreducible AAD</td>
<td>2 (4)</td>
<td>7 (14)</td>
<td>9 (18)</td>
</tr>
<tr>
<td>AAD with basilar</td>
<td>5 (10)</td>
<td>15 (30)</td>
<td>20 (40)</td>
</tr>
<tr>
<td>impression total</td>
<td>28 (56)</td>
<td>22 (44)</td>
<td>50 (100)</td>
</tr>
</tbody>
</table>

* Associated posterior decompression was performed in 11 patients (22%), associated C1–2 lateral joint fusion in 10 (20%), and one-stage transoral odontoidectomy and occipitocervical fusion in seven patients (14%). AAD = atlantoaxial dislocation.
are maintained by means of skull traction. The occipital bone and the C-2 lamina are exposed by sharp subperiosteal dissection. Using a drill, a 3 × 1-cm groove is made in the occipital bone 1 cm superior to the foramen magnum. The depth of the groove reaches the posterior fossa dura. Thus, a bony ridge of occipital bone and occipitalized atlas (termed the “artificial arch of atlas” by Jain, et al.23) is created along the posterior margin of the foramen magnum. The artificial arch and the C-2 lamina are decorticated using a drill. One midline and two lateral notches are made on the superior border of the artificial arch and corresponding notches are made along the inferior border of the C-2 spine in the midline and the C-2 lamina laterally. A loop of No. 22 stainless steel braided wire is passed through the midline under the artificial atlas and the lamina of the axis, and two No. 2 silk sutures or wires are placed on each side of the braided wire. The braided wire is anchored in the midline notches (on the artificial arch and the axis spine) and the silk sutures or wires in the lateral notches on the artificial arch and the axis lamina (Fig. 1). A bone graft harvested from the rib or posterior iliac crest is wedged between the artificial arch and the C-2 lamina. The traction weights are removed and the stainless steel wire is tightened, approximating the occipital bone to C-2, thereby exerting a vertical compression force and correcting the rotational dislocation of the axis. Onlay bone grafts are also held in place by silk sutures or wires on the occipital bone and the C-2 lamina on either side of the tightened steel wire. Before occipitocervical fusion, lateral C1–2 joint arthrodesis was performed in a few patients by drilling the facets, curetting the articular cartilage, and inserting bone chips into the joint space in expectation of additional stability.

Postoperative Course. Postoperatively, the patients were outfitted with a hard cervical collar to be worn for 3 months and mobilized as early as possible.

Results

Neurological and Physiological Outcome

Follow up ranged from 3 months to 1 year. After discharge from the hospital, seven patients were lost to follow-up review. The neurological outcome of the other 43 patients available at follow up is summarized in Table 1. Thirty-one patients (72.09%) improved at least one grade from their preoperative neurological disability, seven patients (16.28%) remained the same, and five (11.63%) deteriorated in comparison to their preoperative status. Osseous fusion was achieved in 38 (88.3%) of the 43 patients who were seen at follow up.

Postoperative Complications

There was postoperative deterioration in strength in two patients, perhaps caused by spinal cord trauma as a result of sublaminar wiring. Five patients underwent reoperation using the same technique. This was necessitated by radiological evidence of loosening or breaking of the wire in three patients and fracture of the artificial arch in two. All of these patients deteriorated neurologically after an initial postoperative improvement. Following occipitocervical restabilization, only two improved. One patient developed retinal ischemia due to pressure exerted on the globe while the patient was lying prone on the horseshoe head rest. Careful positioning avoided this complication in subsequent procedures. Two patients developed stitch-line infection. There was no operative mortality in the series.

Discussion

The use of numerous techniques for occipitocervical stabilization indicates that no particular technique is entirely satisfactory. The earliest proposed method of occipitocervical fusion uses a simple onlay graft and external
The strength of the present technique of occipitocervical fusion lies in its simplicity and its ability to achieve stable internal fixation. The artificial atlas and posterior elements of C-2 are held firmly in contact with the bone graft by the encircling wire. Additional onlay grafts are also placed on the decorticated occipital bone and C-2 and arthrodesis of C1–2 facet joints may be performed, as we did in 10 patients. The procedure provides excellent bone-to-bone contact and prevents all potential movement between the occipital bone and the axis. Tightening of the wire between the artificial atlas and the axis lamina produces a vertical compression force that prevents postero-superior dislocation of the axis into the canal (Fig. 2). It also corrects anteroposterior and rotational dislocation of the axis, thus providing a three-dimensional reduction of AAD. The use of autologous bone graft ensures a physiological occipitocervical arthrodesis. We practice an early postoperative mobilization of our patients while they wear a hard cervical collar. That a stable union is achieved in our patients is evident by the high osseous union rate (88.3%). Stainless steel prosthesis causes image distortion in MR imaging. Wires or interlaminar clamps made of titanium can be used in the present method to allow undistorted postoperative images of the spinal cord at the level of fixation.

In this series, an anterior decompression via the transoral–transpharyngeal route was normally performed in the presence of a ventral, irreducible deformity at the craniovertebral junction. Because ventral excision of the osteoligamentous components is bound to produce instability, either clinical or latent, this was always followed by posterior fusion. However, seven of our patients who had ventral compression showed marked clinical improvement on traction despite radiological evidence of persisting AAD. These patients underwent direct occipitocervical fusion. Their improvement can be attributed to a mild distraction of the odontoid process from the foramen magnum and to a change in the direction of the posteriorly directed odontoid process to a more vertical one due to the pull of traction. After arthrodesis, these patients continue to maintain their improved neurological status and receive regular follow-up evaluation. The option of a transoral odontoidectomy is still open in case they exhibit any deterioration. In the last seven patients, anterior decompression and posterior stabilization were performed under the same anesthetic, as has been practiced by Crockard, et al. This was successful in alleviating the potential hazard of subluxation in the interval between staged anterior and posterior surgeries and has also decreased the morbidity caused by traction.

A few complications were observed. Snapping or loosening of wire was circumvented by using braided steel wires, which are stronger and less liable to slip. Fracture of the artificial arch occurred when the bridge had been made too thin. The neurological deterioration observed could be attributed to either spinal cord trauma during sublaminar wiring or to anterior bowing of the wire, causing sagittal narrowing of the spinal canal. Jain, et al., have recommended the Gallic type of posterior fusion or use of the Halifax interlaminar clamp to avoid this complication. However, these techniques lack the biomechanical advantage provided by sublaminar wiring and orthosis. Although technically simple, it has been associated with the complications of prolonged immobilization in a Minerva jacket or halo brace. Another technique, traditionally performed by tightening a wire that is passed between posterior occipital burr holes and beneath the C-2 lamina or through or below the C-2 spine with a wedged bone graft has been associated with the risk of redislocation and neurological deterioration when the wire is tightened. Use of acrylic augments fusion by increasing the area of contact between the metal construct and the surfaces of vertebrae. However, acrylic predisposes the patient to a higher risk of infection and compromises wound healing.

The newer techniques include using occipitocervical plates and screws, contoured loops and rectangular rods. These methods are technically demanding. The variable thickness of occipital bones makes occipital screws prone to loosening, pulling out of the bone or perforating the inner cortex causing cerebrospinal fluid leakage. Considerable preoperative molding of the construct may be required to adjust to the variable contours in the occipitocervical region. The long lever arm associated with the instrumentation construct may create significant stress and degenerative changes at motion segments below the lowest level instrumented. Occasionally, a lack of direct contact between a segment of the construct and bone results in an unacceptable degree of laxity.
wedge graft. Constant attention must be directed to the eyes of the patient while the head is resting prone on a horseshoe head rest because any direct pressure on the globe may cause blindness from increased intraocular pressure and consequent retinal ischemia.

In our series, the neurological status of 12 patients (27.91%) did not improve at follow up. Similar results have been obtained in treatment of congenital malformations of the craniocervical junction by Di Lorenzo and colleagues and by Malcolm and coworkers in 1994. The lack of improvement may be due to inadequate decompression or concomitant congenital developmental failure of brainstem and spinal cord or because of repeated impaction of bony elements against the cord, which causes permanent axonal loss or microcirculatory changes.

Conclusions

The technique of occipitocervical fusion that creates an artificial atlas in patients with congenital AAD with an assimilated or deficient posterior arch of the atlas is technically simple, provides excellent bone-to-bone contact, and ensures a three-dimensional correction and maintenance of stability. It does not share the disadvantages of other techniques—the risk of redislocation associated with occipitocervical fusion by making occipital bur holes; wound infection and compromised wound healing that occurs with acrylic; plate and screw loosening; inner cortex perforation; or stress at motion segments below the lowest level instrumented—associated with occipitocervical plates and screws. Rather, it provides a stable internal fixation and early patient mobilization.

References


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