Pediatric congenital atlantoaxial dislocation: differences between the irreducible and reducible varieties


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Object. Clinicoradiological differences and outcome following surgery in pediatric patients (≤ 16 years of age) with congenital irreducible atlantoaxial dislocation (IAAD) and reducible atlantoaxial dislocation (RAAD) were analyzed.

Methods. Ninety-six patients (57 with IAAD and 39 with RAAD) were categorized as follows: Grade I, no deficits except hyperreflexia or neck pain (six patients); Grade II, minor deficits but independent for activities of daily living (25); Grade III, partially dependent (30); and Grade IV, totally dependent (35). Patients with RAAD underwent direct posterior fusion, and those with IAAD were treated with transoral decompression and posterior fusion. Patients with good outcomes included those who could walk unaided, with improvement in spasticity and weakness, and those who maintained Grade I status. The category of poor outcome included patients with the following conditions: postoperative deterioration or lack of improvement; inability to ambulate regardless of neurological improvement at a minimum follow-up duration of 3 months; or perioperative death.

A significantly higher incidence of C-1 assimilation, C2–3 fusion, asymmetrical occiput–C2 facet joints, and basilar invagination were seen in patients with IAAD, and os odontoideum was noted in those with RAAD (p < 0.05). A good outcome was recorded in 35 patients with IAAD and 22 with RAAD, whereas 14 with IAAD and nine with RAAD had a poor outcome (eight patients in each category were lost to follow up).

Conclusions. Radiological differences in the anatomy of patients with IAAD and those with RAAD may be due to improper segmentation of the occipital and upper cervical sclerotomes in the former and dysfunction of the transverse ligament in the latter. A significantly better outcome was noted in completely dependent patients with IAAD compared with those with RAAD. Respiratory compromise was an important prognostic factor.

Key Words • atlantoaxial dislocation • cervicomedullary junction • transoral approach • spinal fusion • pediatric neurosurgery

Clinical Material and Methods

Patient Population

Ninety-six patients with congenital AAD (those with an atlantodental interval > 5 mm) who were 16 years of age or younger were included in this study. Fifty-seven patients had an IAAD (mean atlantodental interval 9.5 ± 2.3 mm) and 39 had a RAAD (mean atlantodental interval 8.1 ± 2.8 mm) calculated from dynamic (lateral, flexion, and extension views) plain x-ray films and intrathecal contrast CT scans of the CVJ. The mean age at presentation for patients with IAAD and RAAD was 12.54 ± 3.42 and 11.03 ± 2.81 years, respectively (range 3–16 years). A definite male predominance existed in both groups (the male/female ratio was 45:12 in the IAAD and 31:8 in the RAAD groups). Patients with associated Chiari malformation Type I and those with associated genetic syndromes (Down or Morquio syndrome, achondroplasia, and so on) were excluded from the study. Mean values are represented as the means ± standard deviations.

Abbreviations used in this paper: AAD = atlantoaxial dislocation; CSF = cerebrospinal fluid; CT = computerized tomography; CVJ = craniovertebral junction; IAAD = irreducible AAD; Oc = occiput; RAAD = reducible AAD.
Clinical Examination

The patients were graded according to a system developed by Jain, et al.,²⁶ that consists of the following categories: Grade I, independent without deficits except for hyperreflexia or neck pain (six patients); Grade II, with minor deficits but independent for activities of daily living (25); Grade III, partially dependent on others for daily needs (30); and Grade IV, totally dependent (35) (Tables 1 and 2). The mean duration of symptoms at presentation in patients with IAAD and RAAD was 6.3 ± 2.1 and 4.1 ± 3.2 months, respectively. Respiratory distress (defined as breath-holding time < 10 seconds and single breath count < 10) was present in 10 patients with IAAD and in 11 with RAAD (Table 1). The radiological features of the disorder are summarized in Table 3.

Perioperative Management

The patients with RAAD were treated with posterior fusion alone. In patients with IAAD, reduction was attempted using Crutchfield cervical traction, starting with 7 to 8% of body weight, with a gradual increase in the traction weight to a maximum of 7 kg. Sequential lateral cervical radiographs were obtained to monitor reduction.¹⁴,¹⁵,¹⁷,²² Further increases in weight were stopped when reduction of AAD was accomplished (in one patient) or when distraction of the subaxial spine was noted on plain lateral x-ray films of the CVJ. Incremental additions to the traction weight had to be discontinued in a patient in whom radicular pain and neurological deterioration developed in the form of a transient increase in spasticity, with a change of grade from IV to III, which reversed within 4 hours of removal of traction.

After traction, patients with IAAD underwent transoral decompression followed by posterior fusion.²,¹⁴,¹⁵,²³,²⁶,³¹ After completion of transoral and posterior fusion surgeries for IAAD, the patients were treated with elective ventilation therapy overnight in an intensive care setting and were extubated on the morning of the following day.

Operative Technique for Transoral Decompression

Anterior decompression was performed via the transpalatal, transpharyngeal route.⁴,⁶,⁷,¹⁴,¹⁵,₂₆,³¹ For this approach, traction was continued and the patient was placed supine with the head in a slightly extended position. The patient’s mouth was opened and the tongue retracted using a Boyle–Davis mouth gag. The palate was incised in the midline, skirting around the uvula. The anterior arch of the atlas was used as a guide to the midline. The posterior pharyngeal wall was incised in the midline raphe, exposing the anterior or longitudinal ligament. Then, using the monopolar cautery, the anterior surfaces of the clivus, the C-1 anterior arch, odontoid, and the body of C-2 were exposed up to the C1–2 facet joints on either side. Using a high-speed drill, the anterior arch of the atlas, the clivus, the medial part of the C1–2 joints, the odontoid, and the C-2 body were prepared. The tip of the odontoid was lifted off the dura mater, the apical and alar ligamentous attachments were incised, and the odontoid removed. The posterior longitudinal ligament and the tectorial membrane were also incised, exposing the dura mater. Hemostasis was achieved and the posterior pharyngeal wall and palate were repaired with 2-0 Vicryl in two layers by using muscle and mucosal sutures.

An adequate vertical decompression during the transoral procedure was ensured by excising the part of the C-2 that appears above the Wackenheim clival canal line where it projects into the cervical canal.¹⁴,¹⁵ Lateral decompression

### TABLE 1

<table>
<thead>
<tr>
<th>Symptom</th>
<th>IAAD (%)</th>
<th>RAAD (%)</th>
<th>p Value†</th>
</tr>
</thead>
<tbody>
<tr>
<td>pyramidal signs (weakness, spasticity)</td>
<td>55 (96.5)</td>
<td>29 (74.4)</td>
<td>0.09 (NS)</td>
</tr>
<tr>
<td>ps t column deficits</td>
<td>38 (66.7)</td>
<td>22 (56.4)</td>
<td>0.308 (NS)</td>
</tr>
<tr>
<td>spinohalamic tract demeragement</td>
<td>26 (45.6)</td>
<td>16 (41)</td>
<td>0.656 (NS)</td>
</tr>
<tr>
<td>lower CN dysfunction</td>
<td>3 (5.2)</td>
<td>4 (10.3)</td>
<td>0.355 (NS)</td>
</tr>
<tr>
<td>cerebellovestibular signs</td>
<td>6 (10.5)</td>
<td>9 (23)</td>
<td>0.096 (NS)</td>
</tr>
<tr>
<td>sphincter disturbances</td>
<td>9 (15.8)</td>
<td>14 (35.9)</td>
<td>0.023</td>
</tr>
<tr>
<td>respiratory distress</td>
<td>10 (17.5)</td>
<td>11 (28.2)</td>
<td>0.21 (NS)</td>
</tr>
<tr>
<td>torticollis</td>
<td>23 (40.4)</td>
<td>11 (28.2)</td>
<td>0.22 (NS)</td>
</tr>
<tr>
<td>specific stigmata (high arched palate, low hairline, short neck)</td>
<td>18 (31.6)</td>
<td>10 (25.6)</td>
<td>0.529 (NS)</td>
</tr>
<tr>
<td>transient LOC &amp; weakness after mild trauma</td>
<td>6 (10.5)</td>
<td>5 (12.8)</td>
<td>0.729 (NS)</td>
</tr>
</tbody>
</table>

* CN = cranial nerve; LOC = loss of consciousness; NS = not statistically significant; pst = posterior.
† According to a test of proportion; probability values less than 0.05 suggest a significant difference between the two groups.

### TABLE 2

<table>
<thead>
<tr>
<th>Grade &amp; Definition</th>
<th>IAAD (%)</th>
<th>RAAD (%)</th>
<th>p Value†</th>
</tr>
</thead>
<tbody>
<tr>
<td>I: independent w/o deficits except hyperreflexia or neck pain</td>
<td>2 (3.5)</td>
<td>4 (10.5)</td>
<td></td>
</tr>
<tr>
<td>II: independent for activities of daily living but w/ minor deficits</td>
<td>16 (28.1)</td>
<td>9 (23.1)</td>
<td></td>
</tr>
<tr>
<td>III: partly dependent on others for daily needs</td>
<td>17 (29.8)</td>
<td>13 (33.3)</td>
<td></td>
</tr>
<tr>
<td>IV: totally dependent on others for daily needs</td>
<td>22 (38.6)</td>
<td>13 (33.3)</td>
<td></td>
</tr>
</tbody>
</table>

* Graded according to Jain, et al., 1996.
† According to a test of proportion; probability values less than 0.05 suggest a significant difference between the two groups.

### TABLE 3

<table>
<thead>
<tr>
<th>Radiological Feature</th>
<th>IAAD (%)</th>
<th>RAAD (%)</th>
<th>p Value†</th>
</tr>
</thead>
<tbody>
<tr>
<td>arch of atlas</td>
<td>41 (71.9)</td>
<td>11 (28.2)</td>
<td>0.002</td>
</tr>
<tr>
<td>occipital</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>hypoplastic/bifid</td>
<td>7 (12.3)</td>
<td>5 (12.8)</td>
<td>NS</td>
</tr>
<tr>
<td>normal</td>
<td>9 (15.8)</td>
<td>23 (59.0)</td>
<td>0.002</td>
</tr>
<tr>
<td>odontoid</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>os odontoidude</td>
<td>3 (5.3)</td>
<td>11 (28.2)</td>
<td>0.002</td>
</tr>
<tr>
<td>hypoplastic (dens ≤ 10 mm)</td>
<td>3 (5.3)</td>
<td>1 (2.6)</td>
<td>NS</td>
</tr>
<tr>
<td>basilar invagination</td>
<td>38 (66.7)</td>
<td>1 (2.6)</td>
<td>0.002</td>
</tr>
<tr>
<td>C2–3 fusion</td>
<td>35 (61.4)</td>
<td>7 (17.9)</td>
<td>0.002</td>
</tr>
<tr>
<td>aplasia of C–2 pst arch</td>
<td>1 (1.7)</td>
<td>1 (2.6)</td>
<td>NS</td>
</tr>
<tr>
<td>incurring pst margin of FM</td>
<td>3 (5.3)</td>
<td>2 (5.2)</td>
<td>NS</td>
</tr>
<tr>
<td>asymmetrical C1–2 joints‡</td>
<td>19 of 21</td>
<td>3 of 7</td>
<td>0.007</td>
</tr>
<tr>
<td>platybasia</td>
<td>1 (1.7)</td>
<td>1 (2.6)</td>
<td>NS</td>
</tr>
</tbody>
</table>

* FM = foramen magnum.
† According to a test of proportion; probability values less than 0.05 suggest a significant difference between the two groups.
‡ The C1–2 lateral joints were visualized in only 21 patients with IAAD and in seven with RAAD.
was ensured by drilling the odontoid and axis beyond the lateral dural margins on either side. Thus, a normal funnel-shaped dimension of the upper cervical canal was established.

Operative Technique for Posterior Fusion

In 29 patients, posterior C1–2 fusion was performed using the modified Brooks technique. In 52 patients with an occipitalized posterior arch of the atlas and in 12 with a hypoplastic posterior C-1 arch, an Oc–C2 or an Oc–C3 fusion (with C2–3 block vertebrae in 42 patients) was performed by placing sublaminar stainless-steel wiring between an artificial arch created on the occipital bone and C2–3 laminae. A Ransford contour rod fixation was done in four patients with IAAD and in three with RAAD. Among these patients, two had congenital aplasia of the laminae of the axis, for which an Oc–C4 fusion was required. In the remaining five patients the incurving margin of the foramen magnum required posterior decompression, which necessitated a contour rod fixation (Table 4).

Perioperative Period and Follow-Up Evaluations

The patients were mobilized as early as possible, with their cervical movements stabilized using a hard cervical collar. Clinical assessment was conducted on the 7th postoperative day (at discharge), and at regular follow-up intervals at 3 months, 6 months, and then at yearly intervals. The radiological assessment included plain radiographs of the CVJ at 3 months and then at 6-month intervals to determine the alignment of the CVJ and a stable bone union. The bone union was determined by the presence of a good occipitocervical construct and the visualization of the bone graft in close approximation to the occiput and upper cervical spine, with or without new bone formation. Relative movements between the C-1 posterior arch/artificial arch of atlas and the C2–3 laminae were also evaluated. At follow-up review, a good outcome included patients who could ambulate unaided and who had postoperative neurological improvement in terms of spasticity and weakness. Patients who had no preoperative neurological deficits (Grade I) were also considered to have a good outcome if they maintained their preoperative neurological status after surgery. The outcome was considered poor when there was either postoperative deterioration or lack of improvement; when the patient was nonambulatory without aid, regardless of the neurological improvement in spasticity at a minimum follow-up duration of 3 months; or when there was a postoperative death.

<table>
<thead>
<tr>
<th>TABLE 4</th>
<th>Surgeries performed in 57 children with IAAD and 39 with RAAD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary Op</td>
<td>IAAD (%)</td>
</tr>
<tr>
<td>C1–2 fusion</td>
<td>1 (1.8)</td>
</tr>
<tr>
<td>Oc–C2 fusion</td>
<td>0</td>
</tr>
<tr>
<td>transoral decompression &amp; C1–2 fusion</td>
<td>7 (12.3)</td>
</tr>
<tr>
<td>contour rod fusion</td>
<td>4 (7.0)</td>
</tr>
<tr>
<td>Oc–C2 fusion</td>
<td>45 (78.9)</td>
</tr>
<tr>
<td>contour rod fixation</td>
<td>3 (7.7)</td>
</tr>
</tbody>
</table>

Statistical Analysis

The Z test of proportion was used to compare the incidence of variables in the two groups.

Results

Clinical Spectrum

In 96.5 and 74.4% of patients with IAAD and RAAD, respectively, we found pyramidal tract signs, and in 66.7 and 56.4%, respectively, there were posterior column deficits. Spinothalamic involvement was significantly less in both the groups. Respiratory distress and sphincteric disturbance were more frequent in the RAAD than in the IAAD group. Specific stigmata pointing to the existence of CVJ anomaly were nearly equal in both the groups; however, torticolis was seen more frequently in the IAAD group. Transient loss of consciousness and transient quadriplegia after mild trauma were nearly equal in both groups (10.5 and 12.8% in the IAAD and RAAD groups, respectively; Table 1). A significant number of patients in both groups (68.4 and 66.6% in the IAAD and RAAD, respectively) were categorized in the poor grades (Grades III and IV) at presentation (Table 2).

Radiological Spectrum

An occipitalized arch of the atlas and C2–3 fusion were present in significantly more patients with IAAD (71.9 and 61.4%, respectively, in IAAD and 28.1 and 17.9%, respectively, in RAAD, p < 0.05), whereas os odontoideum was significantly more common in patients with RAAD (5.3 and 28.2% in IAAD and RAAD, respectively; p < 0.05) (Figs. 1–3). As shown in Table 3, in five of these patients with RAAD, there was both an anterior and posterior C1–2 dislocation during flexion and extension movement of the neck (hypermobile AAD). Five patients with an incurring posterior margin of the foramen magnum required posterior decompression before stabilization. The C-2 posterior arch was absent in two patients; this was associated with C2–3 listhesis. A majority of the patients with fixed AAD had improvement in spasticity and respiratory distress after initiation of cervical traction, although reduction was achieved in only one patient. One patient deteriorated after an increase in traction weight, as has already been mentioned.

Postoperative Outcome

Eight patients each in the irreducible and reducible categories were lost to follow up. Thirty-five patients with IAAD and 22 with RAAD met the criteria for good outcome at a mean follow-up duration of 11 ± 3.1 months (range 3–36 months). Fourteen patients with IAAD and nine with RAAD had a poor outcome. In this study, there was a significantly worse outcome in patients with RAAD who were nonambulatory, compared with those who had IAAD (Fig. 4). This was so despite the fact that these two groups were evenly matched with respect to age, duration of preoperative symptoms, and the fact that the patients with IAAD had undergone two surgical procedures (transoral decompression and posterior fusion) compared with only one procedure (posterior fusion) in the mobile AAD group (Table 5). There were three cases of construct failures and four cases of wound infection. The other patients available
at follow-up visits had either evidence of new bone forma-
tion or an adequate occipitocervical construct without C1–2
mobility, visualized on follow-up radiographs.

Morbidity and Mortality

There were seven deaths in the series (in three patients
with RAAD and four with IAAD). All these patients had
been categorized in the poor preoperative grades. Five of
them could not be weaned from the ventilator after surgery.
Of these, three patients had significant preoperative respi-
ratory distress (single breath count < 10 and breath-holding
time of < 10 seconds). Two patients in whom CSF leakage
developed died of septicemia after their CSF leak. Nev-
evertheless, their blood and CSF cultures grew no organisms.
The complications and their management are listed in Ta-
ble 6.

Discussion

Management Protocol and Role of Cervical Traction

In this series, patients with RAAD underwent a direct
posterior fusion; those with IAAD underwent a trial of cer-
vical traction followed by transoral decompression and pos-
terior fusion during the same session of anesthesia, thus
avoiding any morbidity due to postoperative instability and
also obviating the need for a prolonged postoperative trac-
tion.

There has been a difference of opinion regarding the util-
ity of preoperative traction in patients with fixed AAD.
Tuite, et al., reported a reduction of IAAD with traction in
80% of children younger than 14 years of age. Menezes, et
al., however, have stated that congenital fixed AAD rarely
reduces with traction. In agreement with the latter observa-
tion, after traction therapy in the present study, reduction of
IAAD was achieved in only one of our patients, in whom
the need for an additional transoral procedure was avoided.
Our other patients with IAAD, however, showed consider-
able improvement after traction therapy, due to straighten-
ing of spinal curvature (in which the direction of the dislo-
cated, posteriorly directed odontoid changed to a vertical
one); and distraction of odontoid from the foramen mag-
num. Thus, spinal canal diameter increased, resulting in an
improved neurological status that was sustained after sur-

gery. Irreducibility in AAD can only be indisputably es-
tablished by placing the patient in significant traction and
attempting to reduce the dislocation by applying tongs
slightly anterior to midposition to create both extension and
distraction. The transient deterioration in motor strength
in one patient after an increase of traction weight may have
been due to excessive distraction of the vertebral column,
because no other difference in the radiological findings
from the rest of the cases of IAAD was noted.

Radiological Findings and Difference in Origins

There was a significantly higher incidence of occipital-
ization of the atlas, C2–3 fusion, asymmetrical Oc–C2 fac-
et joints, and basilar invagination in patients with IAAD
compared with those with RAAD. We propose that these
radiological differences are a manifestation of the differ-
ences in the genesis of IAAD and RAAD.

Irreducible atlantoaxial dislocation may be the result of
Irreducible and reducible atlantoaxial dislocation in children

Fig. 2. Sagittal reconstructed intrathecal contrast CT scans of the CVJ. Left: The CVJ in flexion showing AAD with os odontoideum. Right: The CVJ in extension showing reduction of AAD with retrolisthesis of os odontoideum.

improper segmentation of the occipital and upper cervical sclerotomes. Thus, the failure of the rostral–ventral component (that forms the foramen magnum) and the caudal–dorsal component (that forms the superior portion of the posterior arch of the atlas) of the neural arch of proatlas, and the first spinal sclerotome (that forms the inferior portion of the posterior arch of C-1), leads to occipitization of the atlas. Segmentation defects of the second spinal sclerotome lead to C2–3 fusion.

Because the ventral–rostral subdivision of the neural arch of the proatlas is also responsible for the development of occipital condyles, its dorsal–caudal division for the development of lateral atlantal masses, and the neural arch of the second spinal sclerotome for the development of the facets of the axis, a failure of development also results in a disproportionately higher incidence of asymmetrical occipitoatlantal facet joints in patients with IAAD. The significantly higher incidence of basilar invagination in these patients is usually due to maldeveloped Oc–C2 joints that cause a central dislocation of the body of the axis and the odontoid into the foramen magnum.

In RAAD, however, in the majority of cases the facet joints are symmetrical, and there is a very low incidence of occipitalized atlas, C2–3 fusion, or basilar invagination. In these patients perhaps incompetent ligaments rather than bone maldevelopment may be responsible for the C1–2 dislocation. The significantly higher incidence of os odontoideum in this category of patients may actually be due to unnoticed cervical trauma sustained during childhood, resulting in malunion of the fractured odontoid segment rather than a developmental anomaly. The radiographic abnormality always has a hypoplastic dens and the sphenoccipital synchondrosis is a definite, visible entity. Thus, os odontoideum cannot be congenital. The hypertrophied rounded ossicle and hypoplastic dens leads to incompetence of the cruciate ligament, causing mobile AAD.

Outcome Assessment

Poor results are often seen in patients in a poor preoperative grade. Concomitant failure of the brainstem and upper spinal cord, gliosis of the nuclei at the cervicomedullary junction and demyelination of long tracts of this region due to repeated trauma, and hypoxia due to venous stasis or occlusion of the vertebral arteries have been implicated as the probable causes of poor outcome after surgery. In our series, however, a number of patients in poor grades (20 with IAAD and 10 with RAAD) had a good outcome at follow-up evaluation regardless of the duration or degree of myelopathy. It is therefore imperative to increase the spinal canal diameter at the CVJ either by transoral decompression and posterior fusion in IAAD, or by C1–2 reduction in RAAD in every case, because surgery remains the best therapeutic option available to these patients. Improvement brought about by surgery, even if it is just one grade, confers on them a huge benefit in terms of their quality of life.

Patients with IAAD who had a longer mean duration of symptoms (6.3 ± 2.1 months) than did those with RAAD (4.1 ± 3.2 months). They also underwent two surgical procedures (transoral decompression and posterior fusion), compared with a single procedure in the RAAD group. Consequently, the duration of surgery and the hospital stay in the first group was significantly longer than in the second. Nevertheless, an interesting finding was the significantly better outcome in severely myelopathic, completely dependent, and nonambulatory patients with IAAD compared with those who had RAAD. This apparent contradiction may perhaps be explained by the differing mechanisms for the development of myelopathy in patients with these two kinds of AAD.

Patients with IAAD experience neurological deficits because of a persistently narrow spinal canal. Myelopathy develops due to the recurrent tenting of the cord against a posteriorly directed odontoid during flexion and extension movements of the neck. Patients with RAAD, however, may deteriorate because of a different mechanism. The repeated backward movement of the odontoid during flexion of the neck (which is relieved on extension) causes spinal cord trauma in these patients. Thus, the amplitude of relative C1–2 movement permits greater energy transfer to the spinal cord in RAAD. In patients with IAAD, the magnitude of the primary injury sustained by the spinal cord may be much less than in RAAD. After transoral decompression, therefore, patients with IAAD improve in terms of neurological deficits because of the establishment of a capacious spinal canal; however, the repeated injury sustained by the cord in patients with RAAD precludes an early recovery, especially in those with severe deficits. An increased incidence of sphincteric and respiratory dysfunction in RAAD compared with IAAD in our patients may also be due to this fact.

An important factor leading to death in our pediatric patients with AAD was respiratory distress. In fact, five pa-
Patients could not be weaned from the ventilator after surgery due to a lack of sustained respiratory effort and the existence of sleep apnea. When an attempt was made to wean these patients from the ventilator, there was a gradual rise in PCO$_2$, so that they often had to be reintubated and again placed on ventilatory support. Poor nutritional status, compromised diaphragmatic function, and weakness of accessory respiratory muscles also played a major role in causing this morbidity.\textsuperscript{14,18} Careful observation of these patients with poor respiratory effort is therefore mandatory. Failure to observe this precaution may lead to a sudden and catastrophic apnea and hypotension with neurological deterioration once the patients are exhausted by their voluntary respiratory efforts.\textsuperscript{14}

Transient postoperative deterioration occurred in six patients in the form of an increase in spasticity and deterioration in strength from Grade I to II, which improved by the 7th postoperative day. In four of these patients with IAAD, neurological deterioration could have been due to trauma to the spinal cord during the transoral decompression or the sublaminar wiring (both were performed during the same session of anesthesia). In two patients with RAAD, the sublaminar wiring may have been responsible for neurological deterioration. Geremia, et al.,\textsuperscript{11} have also alluded to injury caused during sublaminar wiring. The biomechanical advantage and the low cost made us persist in using sublaminar wiring in our series according to the modified Brooks\textsuperscript{3} or Jain\textsuperscript{16} technique. Moreover, a large number of patients with fixed AAD had asymmetrical facet joints, which made transarticular C1–2 stabilization difficult. An interesting

![Fig. 3. Upper Left and Right: Plain lateral radiographs of the cervical spine in flexion (upper left) showing AAD with occipitalized atlas and C2–3 fusion. On extension (upper right), there is no reduction of AAD. Irreducibility can be indisputably established only by placing the patient in significant traction with tongs affixed slightly anterior to midposition to create both extension and distraction. Center Left: Sagittal reconstructed intrathecal contrasted CT scan demonstrating IAAD with occipitalized atlas. Center Right: Coronal CT image demonstrating gross asymmetry of facet joints in IAAD. Lower Left: Postoperative lateral x-ray film showing transoral decompression of the odontoid with occipitocervical fusion.]

![Fig. 4. Line graph showing a significantly worse outcome in nonambulatory patients (Grade IV) with RAAD than in those with IAAD.]

<table>
<thead>
<tr>
<th>TABLE 5</th>
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<tbody>
<tr>
<td>Outcome at follow up for children with IAAD and RAAD*</td>
</tr>
<tr>
<td>Preop Grade</td>
</tr>
<tr>
<td>---</td>
</tr>
<tr>
<td>IAAD</td>
</tr>
<tr>
<td>I</td>
</tr>
<tr>
<td>II</td>
</tr>
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<td>III</td>
</tr>
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</tr>
<tr>
<td>III</td>
</tr>
<tr>
<td>IV</td>
</tr>
<tr>
<td>total</td>
</tr>
</tbody>
</table>

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* The mean follow-up duration in patients with IAAD was 11 ± 3.1 months (range 3–36 months), and for RAAD it was 11 ± 2.5 months (range 3–36 months). Abbreviation: FU = follow up.
Irreducible and reducible atlantoaxial dislocation in children

<table>
<thead>
<tr>
<th>TABLE 6</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complication</td>
</tr>
<tr>
<td>transient postop deterioration</td>
</tr>
<tr>
<td>CSF leak</td>
</tr>
<tr>
<td>construct failure (breaking or loosening of wires)</td>
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<tr>
<td>delayed deterioration due to incomplete reduction of RAAD after post fusion</td>
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<tr>
<td>velopharyngeal insufficiency</td>
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<tr>
<td>wound infection</td>
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* Wound allowed to granulate with regular dressing and not restitched once infection cleared.

cause of postoperative neurological deterioration was the presence of an incompletely reduced odontoid (causing spinal canal compromise) after posterior fusion was performed in a patient with RAAD. This patient showed improvement after a transoral decompression was subsequently performed. None of the other patients with RAAD underwent this operation.

Neck Movement Restriction

Sixty percent of axial rotation occurs exclusively between C-1 and C-2; for rest of the axial rotation, the subaxial spine makes a significant contribution. None of the pediatric patients who underwent an exclusive C1–2 fixation complained of restriction of neck movements; however, all patients with simultaneous occipitocervical stabilization and subaxial fixation by a contour rod were troubled by their neck movement restriction in the initial months, until they had adjusted to this by compensatory torso rotation.

Parisini, et al., have observed the development of subaxial kyphosis after C1–2 posterior fusion in a few of their patients. All of them underwent a spontaneous realignment (known as Toyama remodeling) over a longer follow-up period so that none of them had a swan neck deformity or cervical kyphosis at follow-up evaluation. In our patients, a longer follow up would be necessary to study the effects of transoral decompression and cervical stabilization on the growth and curvature of the growing cervical spine.

Conclusions

A significantly higher incidence of C-1 assimilation, C2–3 fusion, asymmetrical Oc–C2 facet joints, and basilar invagination were seen with IAAD, and odontoidectomy was also seen in conjunction with RAAD. We propose that these radiological differences are a manifestation of the differences in the genesis of IAAD and RAAD. Because IAAD is usually associated with asymmetrical Oc–C2 facet joints, occipitalized atlas, and C2–3 fusion, the condition may be caused by an improper segmentation of the occipital and upper cervical sclerotomes. Reducible atlantoaxial dislocation is usually associated with a well-formed posterior arch of the atlas and with symmetrical lateral joints. In the latter condition, therefore, C1–2 dislocation may be the result of dysfunction of transverse ligaments.

A significantly better outcome was noted in severely myelopathic, completely dependent, and nonambulatory patients with IAAD than was found in those with RAAD. Respiratory compromise was an important prognostic factor affecting the surgical outcome.

References

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