Combined occipitoatlantoaxial rotatory fixation

Case report

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Occipitoatlantoaxial rotatory fixation (OAARF) is a rare condition involving fixed rotational subluxation of the atlas in relation to both the occiput and axis. Atlantoaxial rotatory fixation (AARF) appears to precede OAARF in most cases, as untreated AARF may cause compensatory counter-rotation and occipitoaxial fixation at an apparently neutral head position. We report a case of OAARF in an 8-year-old girl with juvenile idiopathic arthritis. Cervical imaging demonstrated slight rightward rotation of the occiput at 7.63° in relation to C-2 and significant rightward rotation of C-1 at 65.90° in relation to the occiput and at 73.53° in relation to C-2. An attempt at closed reduction with halo traction was unsuccessful. Definitive treatment included open reduction, C-1 laminectomy, and occipitocervical internal fixation and fusion. (DOI: 10.3171/2011.5.PEDS10496)

Key Words • occipitoatlantoaxial rotatory subluxation • spine • atlantoaxial rotatory fixation • juvenile idiopathic arthritis • pediatric cervical fusion

Atlantoaxial rotatory fixation (AARF) is a nonreducible abnormal rotation of C-1 on C-2. Rarely, compensatory occipitoatlanto rotatory subluxation may develop in the setting of AARF creating a combined occipitoatlantoaxial rotatory fixation (OAARF). Atlantoaxial rotatory fixation is a familiar condition for most neurosurgeons; however, there are only 6 previous reports of pediatric OAARF in the literature. A new case is presented here, the first associated with juvenile idiopathic arthritis (JIA), and one documented by more modern imaging (3D CT reconstructions and MR imaging) than most previous case reports.

Case Report

Clinical History. This 8-year-old African American girl was referred to our neurosurgery clinic with a 9-month history of neck pain and progressively limited cervical range of motion. The discomfort began as a “crick” in her neck but gradually evolved to such stiffness that she turned her entire torso to facilitate neck rotation. No history of trauma or upper respiratory infection was present, but the family history demonstrated a father diagnosed with rheumatoid arthritis in early adulthood, a sister with rheumatoid factor (RF)–positive JIA, and a grandfather with systemic lupus erythematosus. Initial outpatient treatment by the child’s pediatrician consisted of muscle relaxants and neck exercises. When these measures failed she was referred to physical therapy. Occipitocervical manipulation by the therapist reportedly reduced her head tilt and rotatory deformity, but her pain and limited cervical mobility persisted. She was then referred to our neurosurgery clinic. Detailed review of systems revealed repetitive right knee swelling, dry eyes and mouth, occasional cervical lymphadenopathy, complaints of diffuse lower-extremity morning stiffness, and dyspnea upon minimal exertion.

Examination. The child held her head in a neutral position with vision directed forward but mobility was severely limited by pain. Fifty degrees of flexion and extension could be achieved, but only 20° of active lateral rotation to either side was possible due to pain. Twenty degrees of lateral neck flexion to the right and 30° to the left could be obtained. Neurological examination demonstrated no deficit. Dry oral mucosa, right knee swelling, hepatomegaly, and a systolic ejection murmur were also noted.
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**Radiographic Evaluation.** Cervical radiographs demonstrated basilar invagination with an obvious anterior subluxation of C-1 on C-2 (Fig. 1). Computed tomography demonstrated only slight occipital rotation to the right of midline (Fig. 2A), but significant rightward rotation of C-1 relative to both the occiput (Fig. 2B and 2C) and C-2 (Fig. 2D). Using the spinous process of C-2 as a reference, the occiput was rotated $7.63^\circ$ to the right and C-1 was rotated $73.53^\circ$ to the right, creating an occiput–C1 angle of $65.90^\circ$. Three-dimensional CT reconstructions further defined these relationships (Fig. 3) and revealed near-apposition of the occipital condyles with the superior articular surfaces of C-2. Magnetic resonance imaging demonstrated basilar invagination and cervicomedullary kinking with upper cervical stenosis (Fig. 4).

**Treatment and Posttreatment Course.** External cervical immobilization with halo traction was attempted for 7 days via 2 frontal and 2 occipital pins, each with 8 inch-pounds of torque applied. Five pounds of traction was established on Day 1 and increased to 10 pounds on Day 2, 15 pounds on Day 3, and finally 20 pounds on Day 5. Valium aided in muscle relaxation and daily lateral cervical radiographs were obtained. By Day 6 reduction of the basilar invagination was noted (Fig. 5) but only minimal correction of the C-1 rotation had been achieved. Therefore on Day 7, with 20 pounds in place and no further radiographic changes, the halo vest was attached and locked in place. The patient was taken to the operating room on Day 8.

Surgical treatment through a standard posterior midline approach exposed the occiput, C-1, and C-2. Bilateral 3.5 mm × 16 mm C-2 pars screws were placed and an occipital fixation plate was secured with bicortical 10-mm midline keel screws and bilateral 8-mm screws to either side; a C-1 laminectomy followed. Easy visualization on the right allowed placement of a 3.5 mm × 14 mm C-1 lateral mass screw, but the obscured anatomy precluded safe placement of a C-1 lateral mass screw on the left (Fig. 6). Bilateral connector rods were contoured and secured; the right C-1 screw required an offset connector to achieve

![Fig. 1. Lateral cervical radiograph showing anterior subluxation of C-1 on C-2.](image)

![Fig. 2. Axial cervical spine CT scans. A: Note slight rightward rotation of the occiput and posterior projection of dens. B: Occipitoatlantoaxial rotatory subluxation can be seen with significant rightward rotation of C-1 compared with the occiput and C-2. C: Note continued rotation of C-1 on C-2. The right occipital condyle is also visible anterior to the right of the C-1 lateral mass. D: Axial cervical spine CT. Significant angulated anterior subluxation of left C-1 lateral mass relative to C-2 is evident. Midline location of the C-2 spinous process demonstrates neutral alignment of axis.](image)

![Fig. 3. Three-dimensional cervical spine CT reconstruction demonstrating significant C-1 subluxation.](image)
appropriate alignment (Fig. 7). Exposed bony surfaces of the occiput, C-1, and C-2 were decorticated and an autograft/allograft mixture was applied for arthrodesis.

The patient tolerated the procedure well and was discharged home on postoperative Day 4. External immobilization via rigid cervical collar was continued for 8 weeks postoperatively. Follow-up at 6 weeks demonstrated neutral head position, complete resolution of neck pain, normal findings on neurological examination, and 50% of normal cervical range of motion. Postoperative MR imaging demonstrated improvement of the basilar invagination compared with preoperative imaging (Fig. 8). Postoperative CT at 3 months demonstrated the early formation of a fusion mass across the occiput, C-1, and C-2 (Fig. 9).

Additional Evaluation and Diagnosis. Rheumatological workup included referrals to rheumatology, ophthalmology, and general surgery clinics. Laboratory testing included the following: complete blood count; comprehensive metabolic panel; antinuclear antibody (ANA), RF, erythrocyte sedimentation rate, C-reactive protein, gamma-glutamyl transpeptidase, angiotensin-converting enzyme, and lysozyme assays; urinalysis; human leukocyte antigen profiling; and cyclic citrullinated peptide antibody assay. Only mild elevations of the erythrocyte sedimentation rate and gamma-glutamyl transpeptidase, aspartate aminotransferase, and alanine aminotransferase levels were noted. Echocardiographic findings were normal; abdominal CT and MR imaging demonstrated focal nodular hyperplasia of the liver. The constellation of her clinical course, persistent joint complaints (which progressed to bilateral knee pain and swelling), and family history have led to the diagnosis of JIA by her rheumatologists.

Discussion

Atlantoaxial rotatory fixation is a relatively familiar condition to pediatric neurosurgeons, whereas combined OAARF is much less common. Only 6 previous reports of
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pediatric OAARF exist, with the first having been published in 1959 and none published in the last decade. This is in contrast to AARF, which was first described in 1830 and has been well studied in the last 20 years.

Injury to the atlantoaxial joint capsules or ligaments can result in atlantoaxial rotatory subluxation. If reduction of this subluxation does not occur, ligamentous or joint capsule contraction can result in AARF. Atlantoaxial rotatory fixation typically causes the head to be held slightly flexed, tilted 20° to one side, and rotated 20° in the opposite direction from the head tilt—the “cock robin position.” Despite this knowledge, the exact definition of how much rotation is abnormal remains under debate. Wortzman and Dewar declared asymmetry of the dens between the atlantal articular masses that does not correct with rotation as the true definition of AARF. Pang and Li theorized that AARF is not defined by any absolute angle of C1–2 separation but instead by abnormal “stickiness” of the C1–2 articulation during rotation.

Occipitoatlantoaxial rotatory fixation seems to be a compensatory result of uncorrected AARF as patients chronically attempt to correct for their abnormal head position and neck pain. Pang and Li noted significantly increased rotation of the occiput on C-1 in chronic AARF (greater than 3 months’ duration). In their studies, patients with acute AARF demonstrated a mean occiput–C1 angle of 5°, those with subacute AARF a mean angle of 9.1°, and those with chronic AARF a mean angle of 31.2°—a statistically significant difference to a probability value of < 0.0000001. Even further, calculations by Pang and Li demonstrated that 64.5% of compensation to a vision-forward head position in chronic AARF occurs via the occipitoatlanto joints and only 35.5% occurs via the subaxial spine. Risk factors for chronic or irreducible AARF include ligamentocapsular contractures, fibrous formations within the synovial joint, inflamed adherent synovial surfaces, osseous union between C-1 and C-2, and abnormal facet deformities.

Occipitoatlantoaxial rotatory fixation has been reported in children aged 9–17 years following traumatic injuries (fall from a horse, break dancing, and chiropractic manipulation), infections (pharyngitis and upper respiratory infection), and surgical treatment of protruding ears (Table 1). Five of 6 previous descriptions of OAARF, as well as this most recent report, identified a prolonged presence of symptoms (range 2–9 months), as diagnosis was often delayed or simply initially missed. This is in contrast to AARF where patients typically present within 17 days of symptom onset. Reports of OAARF have described head positions ranging from neutral (2 previous reports and this case), to a slight cock-robin position, to a pronounced cock-robin position. Diagnosis has been traditionally made via cervical radiographs and CT scans. Magnetic resonance imaging was used in only 2 of the previously reported cases. No cervical vascular injury has been described in
articular arthritis is comprised of 7 subtype groups: systemic arthritis, oligoarthritis, RF-positive polyarthritis, RF-negative polyarthritis, enthesitis-related arthritis, psoriatic arthritis, and undifferentiated JIA. Diagnosis is based on clinical presentation. Subtypes are separated primarily by number and location of joints involved over the first 6 months of symptoms, the presence of other systemic symptoms, and antibody profiles. Specific laboratory markers may aid in subtype classification, but only 10% of JIA patients are RF-antibody positive and 30%–60% are ANA positive. The presented patient likely demonstrates oligoarthritis, which affects 4 or fewer joints during the first 6 months of symptoms and typically occurs in female patients in an asymmetrical distribution over the lower extremities.

No treatment consensus exists for OAARF but the previous strategies are similar to those typically used for AARF. However, while external immobilization and closed reduction can be successful in AARF, they have been universally ineffective in OAARF. This mirrors the decreased rates of success in closed reduction of AARF. Attempts at closed reduction for OAARF have included manual reduction under anesthesia, Halter traction with cervical bracing for 6 months, and Gardner-Wells traction or halo immobilization. Standard treatment for nonreducible AARF without basilar invagination involves open posterior reduction with internal fixation and fusion. Typical methods of open reduction involve exposure of C-1 and C-2; mobilization of the C-2 nerve root; disimpaction of the C1–2 articular surfaces with care taken to avoid the vertebral arteries; placement of C-1 lateral mass and C-2 pars, pedicle, or lamina screws; and “toggling” at the screws to achieve bony reduction. Decompression via laminectomies may be used depending on the degree of stenosis. Alternative reduction techniques may involve an extreme-lateral approach on the side with the anterior-most C-1 facet or utilization of a temporary transverse bar across C-1 to allow improved anchoring during reduction. Anterior transoral atlantoaxial joint release can be used as well. Only one treatment regimen for OAARF featured nonsurgical management, and that was the one described in

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs)</th>
<th>Duration of Sx</th>
<th>Head Position</th>
<th>Cause</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Washington, 1959</td>
<td>11</td>
<td>4 mos</td>
<td>neutral</td>
<td>pharyngitis</td>
<td>traction &amp; Calot jacket</td>
</tr>
<tr>
<td>Clark et al., 1986</td>
<td>16</td>
<td>8 mos</td>
<td>obvious cock-robin</td>
<td>trauma: fall from horse</td>
<td>traction, occiput–C2 arthrodesis, then halo bracing</td>
</tr>
<tr>
<td>Altongy &amp; Fielding, 1990</td>
<td>9</td>
<td>2 mos</td>
<td>obvious cock-robin</td>
<td>trauma: break dancing</td>
<td>traction, atlantoaxial arthrodesis, then halo bracing</td>
</tr>
<tr>
<td>Cowan &amp; Inglis, 1996</td>
<td>13</td>
<td>9 mos</td>
<td>obvious cock-robin</td>
<td>postsurgical</td>
<td>traction, atlantoaxial arthrodesis</td>
</tr>
<tr>
<td>Hettiaratchy et al., 1998</td>
<td>13</td>
<td>3 mos</td>
<td>neutral</td>
<td>upper respiratory infection</td>
<td>traction, C1–2 transarticular fixation, then halo bracing</td>
</tr>
<tr>
<td>Bouillot et al., 1999</td>
<td>17</td>
<td>&lt;1 wk</td>
<td>slight head tilt</td>
<td>trauma: chiropractic manipulation</td>
<td>traction, occiput–C2 arthrodesis</td>
</tr>
<tr>
<td>present case</td>
<td>8</td>
<td>9 mos</td>
<td>neutral</td>
<td>JIA</td>
<td>traction, occiput–C2 fixation &amp; fusion, then cervical collar</td>
</tr>
</tbody>
</table>
Combined occipitocervical rotary fixation

the earliest report (from 1959). Surgical OAARF treatments have included open reduction followed by C1–2 arthrodesis,2,6 occiput–C1–2 arthrodesis,4,5 and C1–2 transarticular screw fixation.8 Postsurgical immobilization was maintained with halo fixation for 3 months in 3 cases2,5,8 and not described in 2 others.4,8 In the majority of cases, a neutral head position was achieved following treatment2,5,6,8,20 (one report did not describe the final head position). Long-term fusion rates were universally not provided.

Nonreducible AARF with basilar invagination and cervicomedullary stenosis is typically treated via anterior transoral C1–2 joint release and odontoidectomy followed by a posterior fusion.19 Direct posterior reduction of basilar invagination and C1–2 subluxation can be achieved without anterior decompression. Typically, this involves the placement of occipit and C-2 screws and subsequent distraction applied between them.15 Anterior decompression was not used in the presented case, as the basilar invagination reduced with halo traction and neurological symptoms were absent. Postoperative MR imaging (Fig. 7) demonstrated some persistent basilar invagination and stenosis, but improvement from the initial presentation was evident. In the absence of neurological symptoms and with a stable fixation construct in place, further surgical intervention was not deemed necessary. Early postoperative CT demonstrated onset of osseous fusion between C-1 and C-2, further strengthening the belief that adequate fixation was provided.

If open reduction cannot be achieved intraoperatively, many AARF treatment reports call for occipitocervical fixation and fusion.1 However, among the previously reported cases of surgically treated OAARF, only 2 cases included the occiput in the fusion construct4,5 while 3 did not.2,6,8 This seems to reflect a belief that surgical treatment of the primary atlantoaxial instability alone could result in resolution of the secondarily reactive occipitoatlanto rotation with simple postoperative external immobilization. If true, this would be appealing because inclusion of the occiput in the fusion construct significantly limits cervical motion. Unfortunately, long-term outcomes are not provided in previous OAARF cases to allow comparison. Therefore, no data exist to support this theory, and we believe occipitocervical fusion is required in the treatment of OAARF to ensure adequate stability. Specific reasons occipitocervical fixation and fusion were felt necessary in the present case included an inability to fully reduce the occipitoatlantoaxial rotation through halo traction or intraoperative techniques, suspected further underlying pathological instability due to JIA, and the increased potential for neurological injury with continued instability due to the degree of cervicomedullary stenosis.

Conclusions

This report describes the first case of OAARF in a child with clinically confirmed JIA. While the child’s head position appeared neutral, a significant amount of occipitoatlantoaxial rotation was present. Initial treatment with halo traction improved the basilar invagination but did not correct the rotational deformity. Ultimate treatment required occipitocervical internal fixation and fusion. Pediatric neurosurgeons should always be aware of possible occipitocervical instability while evaluating AARF, particularly in the presence of a neutral head position or in patients who present in a delayed fashion (after at least 3 months’ symptom duration).

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: Fusco. Analysis and interpretation of data: Fusco. Drafting the article: Fusco. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Fusco. Administrative/technical/material support: Fusco.

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Manuscript submitted November 2, 2010. Accepted May 27, 2011.
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