We present seven children with atlantoaxial rotatory fixation (AARF) of more than three months’ duration after an injury to the upper cervical spine. The deformity was irreducible by skull traction. MRI and MR angiography (MRA) of the vertebral arteries were performed in four children. The patients were neurologically intact. Thrombosis of the ipsilateral vertebral artery was noted in two patients. The deformity was gradually corrected and stabilised after transoral release of the atlantoaxial complex, skull traction and posterior atlantoaxial fusion.

Soft-tissue interposition and contractures within the atlantoaxial complex prevented closed reduction. MRI and MRA of the vertebral arteries were useful in elucidating the pathology of chronic atlantoaxial rotatory fixation.

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Atlantoaxial rotatory fixation (AARF) is uncommon in children and is usually associated with injury to the cervical spine or an infection of the upper respiratory tract. In patients who present early satisfactory correction of the deformity has been reported with non-operative management such as cervical traction and orthotic treatment, but intra- and extra-articular factors may prevent closed reduction when the diagnosis is delayed. Patients who present late at more than three months have usually been managed by in situ C1/C2 fusion after failure of reduction with traction. Reports of this technique have shown that it has not been uniformly satisfactory in correcting the deformity. We have performed clinical and radiological correction of chronic AARF by anterior release of the atlantoaxial complex, gradual skull traction and second-stage posterior atlantoaxial fusion. We found that MRI and MR angiography (MRA) of the vertebral arteries were useful investigations in the understanding and treatment of chronic AARF.

Patients and Methods

Between 1994 and 1998 we treated 43 children with AARF of whom seven (16%) presented late after injury. Details of these seven patients are given in Table I. Associated injuries included fractures of the radius and ulna (case 2) and of the clavicle (case 4).

All seven patients were neurologically intact and presented with a ‘cock-robin’ posture with the head tilted to one side and the chin rotated to the opposite side. Minor facial asymmetry was noted in two (cases 5 and 8) and the gaze was not horizontal in all patients. The sternocleidomastoid muscle on the side of rotation of the chin was in spasm and rotation of the neck to the side of the subluxation was painful and restricted in all children. Radiographs and CT of the upper cervical spine confirmed the diagnosis of AARF (Figs 1 and 2). In addition, four patients (cases 4, 5, 6 and 7) had MRI and MRA to evaluate the atlantoaxial complex and the vertebral arteries, respectively. MRI revealed interposition of fibrous tissue within the subluxed atlantoaxial joint and the atlantodental interval (Figs 3 and 4). In two patients (cases 4 and 7) thrombosis of the ipsilateral vertebral artery was demonstrated by MRA (Figs 5 and 6). The fixed rotatory subluxations were irreducible by skull traction applied for a mean of 11 days (9 to 17).

Transoral release was performed after nasal intubation with the head immobilised in skull traction with 1.9 kg of weight. An infant feeding tube was passed down the opposite nostril to retract the uvula and the oral cavity was exposed with a Doyle retractor. The hypopharynx was packed with tonsil swabs and the posterior pharyngeal wall was palpated to localise the prominent anterior tubercle of the atlas. The atlantoaxial complex was identified using an image intensifier and the posterior pharyngeal wall was infiltrated with 3 to 5 ml of 1% marcaine. The atlantoaxial complex was exposed through a midline incision extending from the anterior arch of the atlas to the C2/C3 disc space. The dissection was carried out laterally to expose the...
Table I. Details of patients with chronic atlantoaxial rotatory fixation

<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Age (yrs)</th>
<th>Mechanism of injury</th>
<th>Fielding and Hawkins classification</th>
<th>Duration of symptoms (wks)</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>6</td>
<td>Fall from tree</td>
<td>II</td>
<td>14</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>5</td>
<td>Fall from tree</td>
<td>II</td>
<td>19</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>7</td>
<td>Fall from tree</td>
<td>II</td>
<td>16</td>
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<tr>
<td>4</td>
<td>F</td>
<td>5</td>
<td>Motor-vehicle accident</td>
<td>II</td>
<td>37</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>9</td>
<td>Motor-vehicle accident</td>
<td>III</td>
<td>23</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>8</td>
<td>Fall from tree</td>
<td>II</td>
<td>29</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>11</td>
<td>Motor-vehicle accident</td>
<td>II</td>
<td>17</td>
</tr>
</tbody>
</table>

Fig. 1
A lateral plain radiograph of the upper cervical spine in AARF. The arrow shows an increased atlantodental interval type III (>5 mm) of Fielding and Hawkins.

Fig. 2
Axial CT showing subluxation of the right atlantoaxial joint.

Fig. 3
MRI of the cervical spine. A sagittal T2-weighted image with contrast showing fibrous tissue within the atlantoaxial complex (arrow).

Fig. 4
Coronal MRI showing subluxation of the right atlantoaxial joint (arrow) and a normal occipitocervical junction.
atlantoaxial joints and the anterior arch of the atlas. The exposure of the subluxed joints should be performed with care to prevent injury to the ipsilateral vertebral artery which has an abnormal course. Malleable copper re retractors were placed immediately lateral to the atlantoaxial joints to protect the vertebral arteries. The affected joints were subluxed anteriorly and inferiorly and tethered by fibrous tissue. The anterior arch of the atlas was exposed subperiosteally. The fibrous tissue interposed between the disrupted atlantoaxial joints and the atlantodental interval was carefully removed with curved Codman curettes and ronguers. It is essential to stay within the confines of the atlantodental interval in order to prevent injury to the spinal cord when removing the fibrous tissue. The articular cartilage of the subluxed atlantoaxial joints was normal.

No attempt was made to reduce the subluxed joint intraoperatively after the release. The posterior pharyngeal wall was closed with interrupted absorbable sutures and the patients extubated 24 hours after surgery. Skull traction was maintained with 1.9 kg of weight on a split mattress until clinical and radiological evidence of reduction was observed at a mean of seven days (5 to 11). A posterior atlantoaxial fusion with sublaminar wires was performed in all patients and the neck was immobilised in a soft collar until fusion was achieved.

Results

The mean follow-up was 15 months (7 to 23). The ‘cock-robin’ posture disappeared and fusion was noted after four to seven weeks with normal alignment of the atlantoaxial joints and the atlantodental interval (Fig. 7). Two patients who had thrombosis of the vertebral artery showed no evidence of vertebrobasilar insufficiency. There were no neurological complications or sepsis with the transoral approach but one patient (case 7) developed a pressure sore over the occiput which resolved.
Discussion

Rotatory subluxation of the atlantoaxial complex in children is uncommon and is associated with a variety of conditions including congenital abnormalities, anatomical variations, trauma and infection.1-8 In children the atlantoaxial joints are horizontal and shallow and the ligaments and joint capsules have sufficient elasticity to give increased mobility without disruption. The transverse ligament allows rotation but prevents excessive anterior translation of the atlas on the axis.1,11,12 At the extremes of rotation the atlantoaxial articular surfaces disengage and further dislocation is prevented by the capsules of the facet joints.11,12 The disproportion between the size of the head and neck and the poorly developed musculature may predispose the atlantoaxial joint to excessive forces resulting in abnormal movement and atlantoaxial rotatory subluxation.12

A meticulous history, clinical evaluation, and imaging studies are essential for distinguishing AARF from other causes of torticollis. The sternocleidomastoid on the side to which the chin is rotated may show spasm in an attempt to reduce the deformity. In late presentations changes in vocal character may result from chronic pharyngeal compression and the patients may have difficulty in opening the mouth.8 Goddard et al10 reported five cases of AARF associated with fractures of the clavicle. In our study fracture of the clavicle occurred in one patient and was on the side of the dislocation. This is consistent with a fall on to the shoulder and the side of the head. The diagnosis of AARF was initially missed in three children who were involved in a motor-vehicle accident and the remaining four children did not present to hospital immediately after a fall.

Previous studies have speculated on the mechanism of fixation in AARF.2,11,13,14 Muscle spasm and swollen capsular and synovial tissues which occur in the early stages may prevent reduction. Ligamentocapsular contractures occur in the late stage.2 It has been suggested that adherent and inflamed synovial fringes may also impede reduction.11 Although Wortzman and Dewar11 postulated that there was damage to the articular surfaces of the atlantoaxial joint in these injuries, this was not seen in our study. CT tomographic scans have made a significant contribution to the understanding of the patterns of instability within the atlantoaxial complex. The transverse cuts can show the disengagement of the facet, injury to the ligamentous and bony complex and any congenital variants. MRI and MRA, which clearly defined the soft-tissue pathology, have not been previously reported in the assessment of chronic AARF without neurological deficit. Non-concentric alignment of the atlas and axis reduces the space available for the cord and may predispose the patient to neurological damage.2,7,16

Thrombosis of the ipsilateral vertebral artery after atlantoaxial rotatory fixation can result in infarction of the brainstem. The absence of neurological deficit after thrombosis of the vertebral artery in two of our patients may be attributed to the younger age group and the possible establishment of an adequate collateral circulation. MRI and MRA were not available during the earlier part of the study (cases 1, 2 and 3).

Although the natural history of the untreated condition is not known, Fielding and Hawkins2 reported a follow-up for eight years of two patients who refused surgery. Spontaneous correction of the clinical deformity occurred in one and in the other the deformity was unchanged. Satisfactory outcomes have been reported after conservative treatment with traction and collars in acute AARF.2,4,9 Subach et al8 noted no recurrence when reduction and immobilisation were achieved within 21 days of symptoms. We reported seven children who presented between two and six weeks with type-II AARF.16 The displacement was reduced in six after traction, but instability was noted on flexion/extension radiographs at three months in five patients and necessitated atlantoaxial fusion. Philips and Hensinger11 reviewed five patients who presented between one and three months with type-II AARF. The subluxation recurred in three patients after traction and immobilisation and in the remaining two reduction was not achieved. Traction and neck manipulation of chronic AARF have not been considered to be an adequate or safe method for overcoming the fibrosis and scarring which result from chronic displacement.4

Previous studies have recommended an in situ fusion for irreducible AARF but our results and those reported by others after this procedure have not been uniformly satisfactory.2,4 Chronic AARF is associated with facial asymmetry and the development of a squint as the child tries to maintain a horizontal gaze. The apparent spontaneous correction of the clinical deformity may be attributed to compensatory changes occurring either at the occipitocervical junction or at the subaxial spine. Fixed rotatory subluxation may result in adaptive and early degenerative changes in the subaxial spine because of compensatory rotation in the lower spine to realign the head to the neutral position.8,12 Studies have also reported compensatory counter occipitoatlantal subluxation to correct the deformity in chronic AARF.15,17 Clark et al17 reported a child who presented with a normal occipitocervical junction and chronic AARF. The preoperative halo traction and physiotherapy resulted in a partial correction of the fixed deformity. However, the partial correction which was due to a compensatory subluxation of the occipitoatlantal joint necessitated an occipitocervical fusion.

The lateral retropharyngeal and transoral routes have been described for exposure of the atlantoaxial complex.18,19 The operation must be performed by surgeons familiar with these approaches in order to avoid potential neurovascular complications. Adequate lighting and magnification are essential to the performance of a safe and satisfactory procedure. The lateral retropharyngeal approach gives an excellent exposure of the ipsilateral atlantoaxial joint, but access and adequate visualisation of the joint on the opposite side are not always satisfactory.
We have used this approach to drain cold abscesses affecting the upper cervical spine in order to avoid wound sepsis which may follow the transoral route. In chronic AARF it is essential not to reduce the joint after the transoral release. Fang and Ong reported a case of chronic atlantoaxial dislocation in which a blunt hook was used to reduce the joint. Haemorrhage occurred on several occasions and despite adequate resuscitation the patient developed a fever and died nine days after transoral surgery. A tear of the vertebral artery and vein was revealed at post-mortem.

Although AARF is uncommon, early recognition and treatment may prevent subsequent instability and deformity. MRI of the atlantoaxial complex and MRA of the vertebral arteries are essential before surgery in patients with chronic AARF. A staged procedure was safe and useful in correcting the clinical and radiological deformity in chronic AARF after trauma.

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References