The treatment of refractory atlanto-axial rotatory fixation using a halo vest

RESULTS OF A CASE SERIES INVOLVING SEVEN CHILDREN

We reviewed seven children with torticollis due to refractory atlanto-axial rotatory fixation who were treated in a halo vest. Pre-operative three-dimensional CT and sagittal CT imaging showed deformity of the superior articular process of C2 in all patients. The mean duration of halo vest treatment was 67 days (46 to 91). The mean follow-up was 34 months (8 to 73); at the latest review six patients demonstrated remodelling of the deformed articular process. The other child, who had a more severe deformity, required C1-2 fusion.

We suggest that patients with atlanto-axial rotatory fixation who do not respond to conservative treatment and who have deformity of the superior articular process of C2 should undergo manipulative reduction and halo-vest fixation for two to three months to induce remodelling of the deformed superior articular process before C1-2 fusion is considered.

Atlanto-axial rotatory fixation (AARF) is defined as torticollis due to dislocation or subluxation of the atlanto-axial joint and occurs most often in children.1 It commonly presents with cervical pain, stiffness and a ‘cock-robin’ position. When associated with an infection of the upper respiratory tract, it is also known as Grisel’s syndrome.2,4 The cause is often an upper airway infection such as laryngopharyngeal inflammation, or minor trauma, but occasionally there is no clear aetiology.4-8 Along with open-mouth anteroposterior (AP) and lateral radiographs, CT has been used to establish the diagnosis.9-13

The condition is usually mild and reduction can often be achieved simply with a collar or neck traction.1,6,8,14 However, treatment may be difficult if delayed.6,8,15 On rare occasions the deformity of the superior articular process of C2 may persist, resulting in refractory torticollis.16,17 In this report, we review seven cases of refractory AARF with deformity of the superior articular process C2 that had been treated with a halo vest.

Patients and Methods

Between January 1995 and December 2008, 23 319 patients registered by the Nagoya Spine Group, a multicentre study group for spinal disorders, underwent surgery of the spine. Of these, seven children with refractory AARF were treated with a halo vest (Table I). Their mean age was 6.7 years (4 to 10) and the mean period between the onset of the condition and the application of the halo vest was 84 days (31 to 169). Radiographs and CT of the upper cervical spine confirmed the diagnosis (Fig. 1). One patient had Fielding type I’ atlanto-axial rotatory subluxation while the other six had type II.

We noted the cause of AARF, the time from its onset to application of the halo vest, the presence or absence of deformity of the superior articular process of C2 on both three-dimensional (3D) and sagittal CT imaging (Figs 2 and 3), duration of halo-vest fixation, and remodelling of the superior articular process of C2 after treatment.

Under general anaesthesia, we performed closed manipulation of the atlanto-axial joint, applied the halo vest using six or eight skull pins (Fig. 4) and confirmed the reduction by CT. Follow-up included monthly radiographs for the first three months, then at six months postoperatively and then yearly. Before removal of the halo vest we performed CT to confirm remodelling of the superior articular process of C2. In this regard, the sagittal image was particularly relevant (Fig. 3). If remodelling was confirmed, we removed the halo vest without anaesthesia and applied a neck collar for two to three months. If the appropriate remodelling was not achieved, a C1-2 fusion was performed.

Results

The cause of AARF was an infection of the upper airway in four patients, mild trauma in two, and was unexplained in the other. In all
cases the deformity of the superior articular process of C2 was demonstrated on the pre-operative 3D and sagittal CT images. The mean duration of halo-vest fixation was 67 days (46 to 91) and the mean follow-up was 34 months (8 to 73). At the latest review, remodelling of the superior articular process of C2 was noted in six of the children (Figs 5 and 6). In one the torticollis recurred once after the halo vest was removed and, as the superior articular process had not remodelled, we performed atlanto-axial fusion, using a C1 lateral mass screw and a C2 pedicle screw (Stryker, Mahwah, New Jersey). In another patient (case 5), AARF recurred twice, at both one year and 1.5 years after removal of the halo vest. However, on both occasions, treatment in a collar was successful.

Discussion
In AARF, CT with 3D reconstruction is the most reliable technique to confirm the rotation of the axis and abnormality of the bony elements. Ishii et al reported 24 cases of AARF and suggested that the deformity of the superior facet of C2 in chronic AARF was a risk factor for recurrence, and that more than 20° of lateral inclination was frequently seen in cases where the condition was irreducible.
Pang and Li\textsuperscript{18} proposed a new classification using dynamic CT and described the management and outcome of patients treated with their new algorithm. Patients were divided into five groups (types I, II and III, Diagnostic Grey Zone, and Normal). This helped us to determine appropriate treatment strategies. We suggest that the sagittal CT image not only shows clearly the wedge-shaped deformity of the superior articular process of C2, but also that it is superior to 3D reconstruction in the assessment of remodelling before removal of a halo vest.

In general, patients with AARF recover either spontaneously or with conservative treatment such as a neck collar or traction.\textsuperscript{1,6,8,14} Subach et al\textsuperscript{8} found no recurrence when reduction and immobilisation of the neck were achieved within 21 days from the onset of symptoms. Fernández Cornejo et al\textsuperscript{19} reported that patients with atlanto-axial subluxation which remained unreduced for longer than three weeks are at a greater risk of recurrence or permanent deformity. Conversely, others believe that refractory and recurrent cases of AARF are due to over stretching of the ligaments, distension and laxity of the joint capsule or C2 facet deformity.\textsuperscript{6,16,17,20}

We considered atlanto-axial joint stability to be restored once the articular surface of the deformed articular process had remodelled during the two to three months in the halo vest. In the case of the patient who did not remodel successfully and required C1-2 fusion (case 4), the duration of symptoms before treatment (72 days) was no longer than the other patients, but the deformity of the superior articular process of C2 was more severe. Generally, the deformity is progressive, and halo-vest treatment should start within six months of the onset of symptoms. We suggest that another limitation to this method of treatment is a patient with refractory AARF who has irreducible bony ankylosis or severe fibrotic contracture of the C1-2 facets. In these cases, if we cannot achieve full reduction in an AARF patient while under general anaesthesia, we cannot expect appropriate remodelling of the C2 superior articular process. Fortunately, we achieved full reduction of the C1-2 subluxation in all of the cases in our study.

In conclusion, a patient with AARF who does not respond to conservative treatment and has deformity of the superior articular process of C2 should undergo manipulative reduction and treatment with a halo vest for two to three months in order to induce remodelling of the deformed facet before C1-2 fusion is considered.
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References