Atlantoaxial stabilization in rheumatoid arthritis

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Atlantoaxial subluxation in patients with rheumatoid arthritis is common. Operative stabilization is clearly indicated when signs and symptoms of spinal cord compression occur. However, many recommend early operative fusion before evidence of appreciable neural compression occurs because 1) the myelopathy in these patients may be irreversible; 2) the overall prognosis is poor once symptoms of cord compression are present; and 3) the risk of sudden death associated with atlantoaxial subluxation is increased even in asymptomatic patients. The authors believe that rheumatoid arthritis patients in relatively good health without advanced multisystem disease and less than 65 years of age should be considered for operative stabilization if mobile atlantoaxial subluxation is greater than 6 mm. Seventeen patients with severe rheumatoid arthritis and atlantoaxial subluxation treated with a posterior arthrodesis are presented. A new method of fusion, devised by the senior author (V.K.H.S.), was utilized in all cases. Indications for operative therapy in these patients included evidence of spinal cord compression in 11 patients (65%) and mobile atlantoaxial subluxation greater than 6 mm but no signs or symptoms of cord compression in six patients (35%). Thirteen patients developed a stable osseous fusion, two patients a well-aligned fibrous union, one patient a malaligned fibrous union, and one patient died prior to evaluation of fusion stability.

The details of the operative technique and management strategies are presented. Several technical advantages of this method of fusion make this approach particularly useful in patients with rheumatoid arthritis. Because of multisystem involvement of this disease, a high rate of osseous fusion is often difficult to achieve.

KEY WORDS • atlantoaxial subluxation • cervical spine • rheumatoid arthritis • spinal fusion

RHEUMATOID involvement of the cervical spine was first reported by Garrod in 1890. Of the 500 patients with rheumatoid arthritis without the benefit of radiographic confirmation whom he studied, 36% had clinical evidence of cervical spine involvement. Cervical spine malalignment and instability in rheumatoid arthritis occur secondary to synovial proliferation and destruction of the surrounding bone and supporting ligamentous structures. The three most common abnormalities of the cervical spine found in patients with rheumatoid arthritis are atlantoaxial subluxation, subaxial subluxation, and superior migration of the odontoid. The incidence of atlantoaxial subluxation alone or in combination with other lesions of the cervical spine is reported to occur in 19% to 71% of patients with rheumatoid arthritis. Suboccipital pain secondary to compression of the C-2 root is the most common symptom associated with atlantoaxial subluxation. The presence and severity of an associated myelopathy correlate with the degree of subluxation, the extent of upward migration of the odontoid, and the severity of associated pannus formation. While progressive subluxation over time is typical, the rate and extent of progression may be unpredictable in individual patients. Radiological progression may be more common than the concomitant progression of neurological symptoms.

Most authors agree that atlantoaxial subluxation associated with progressive neurological symptoms is an appropriate indication for operative intervention; however, the indications for operative treatment of patients without myelopathic deficits is still controversial. Patients with severe rheumatoid arthritis often have complex multisystem medical problems associated with the disease process, in addition to complicating side effects related to drug therapies. The operative mortality rate associated with cervical spine stabilization procedures ranges from approximately 5% to 15%. The failure rate for atlantoaxial (C1-2) fusion may reach as
TABLE 1

Classification of functional capacity*

<table>
<thead>
<tr>
<th>Class</th>
<th>Definition</th>
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<tbody>
<tr>
<td>I</td>
<td>complete ability to carry on all usual duties without handicaps</td>
</tr>
<tr>
<td>II</td>
<td>adequate for normal activities, despite handicap or discomfort or limited motion at one or more joints</td>
</tr>
<tr>
<td>III</td>
<td>limited to little or none of duties of usual occupation or self-care</td>
</tr>
<tr>
<td>IV</td>
<td>incapacitated, largely or wholly, bedridden or confined to wheelchair; little or no self-care</td>
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*Classification according to Steinbrocker, et al.27

high as 50%. Nonetheless, many authors believe that “prophylactic” C1–2 fusion should be performed in select patients even without evidence of a cervical myelopathy.3,4,11,12,17,24,25

Several fusion techniques have been utilized for posterior stabilization of the atlantoaxial complex. Internal fixation typically involves a bone graft with either midline wires (Galileo-type fusion),4,8,12,18,21 lateral wires (Brooks-type fusion),23,10,17 or posterior-lateral clamps (Halifax-type fusion).2 A new C1–2 fusion technique, devised by the senior author (V.K.H.S.), was used in 17 patients with severe rheumatoid arthritis and atlantoaxial instability. Our experience is described here.

Summary of Cases

Clinical Presentation

Seventeen consecutive patients with severe rheumatoid arthritis and atlantoaxial instability underwent operative C1–2 fusion using the technique described. This group included 11 females and six males with a mean age of 60 years (range 39 to 79 years). The clinical severity of rheumatoid arthritis (excluding neurological dysfunction) at the time of presentation was classified according to the criteria introduced by Steinbrocker, et al.27 (Table 1). Five patients had a functional capacity of Class II, seven Class III, and five Class IV.

Fifteen patients (88%) presented with complaints of severe suboccipital pain consistent with irritation of the C-2 root. Ten patients (59%) had symptoms of a progressive myelopathy, consisting of progressive quadriparesis and/or spasticity. One patient complained of Lhermitte’s sign associated with flexion of the neck. The remaining six had no presenting signs or symptoms suggestive of spinal cord compression. However, it is often difficult to assess the severity of neurological involvement in patients with severe rheumatoid arthritis due to the presence of a concomitant peripheral neuropathy, muscle atrophy, and/or joint deformities.

Medical Therapy

Medical therapy at the time of presentation included exogenous steroids in 10 patients, methotrexate in six,

Plaquenil (hydroxychloroquine sulfate) in three, and gold injections in two. Four patients had significant pulmonary disease which required maintenance bronchodilator therapy. Six patients were receiving antihypertensive medications. Two patients were hypothyroid, corrected with thyroid replacement. All patients were intermittently given a variety of nonsteroidal anti-inflammatory agents.

Radiographic Findings

Five patients had a combination of atlantoaxial instability and cranial settling with irreducible anterior subluxation that required a transoral resection of the odontoid and associated pannus. This procedure was typically followed by a C1–2 fusion approximately 7 to 10 days later. In the remaining 12 patients, the degree of subluxation, as measured from the atlanto-odontoid interval on lateral cervical spine x-ray films in flexion and extension, ranged from 6 to 14 mm (Fig. 1). Flexion and extension magnetic resonance (MR) images were obtained in most patients to assess the degree of spinal cord compression (Fig 2). Initial transoral decompression was performed on those patients who had persistent anterior neural compression as demonstrated on dynamic MR images.

Operative Procedure

All patients underwent extensive preoperative evaluation with particular attention to associated pulmonary and cardiovascular disease. Following induction of general anesthesia, a halo body jacket is applied and adjusted to achieve optimal radiographic C1–2 alignment. Somatosensory evoked potentials are monitored throughout this period and the remainder of the operation. The patient is placed in a prone position, the halo apparatus is secured to the operating table using the halo-Mayfield attachment,7 and a final preoperative x-ray film is obtained to assure proper alignment (Fig. 3).
A midline posterior cervical incision is made from the midecciput to the spinous process of C-3. Sharp subperiosteal dissection is used to expose the occipital squama, the posterior ring of C-1, and the lamina of C-2. The unipolar electrocautery is used sparingly to avoid devascularizing the cortical bone. Connective tissue and pannus, which is often quite thick in patients with rheumatoid arthritis, is sharply removed along the superior and inferior margins of C-1 and the superior margins of C-2. The posterior arch of C-1 may migrate cephalad and rotate, thereby requiring removal of the lower lip of the foramen magnum or the upper edge of the C-1 ring for proper exposure. The graft site is initially prepared by decortication of the inferior edge of the ring of C-1 and the superior portion of the C-2 lamina and spinous process using a Kerrison rongeur or high-speed drill.

A tricortical bone graft approximately 4 cm long is obtained from the top edge of the posterior iliac crest. The rounded cortical side of the graft is removed creating a bicortical strut. The strut is slightly curvilinear due to the shape of the posterior iliac crest. The graft is fitted horizontally between C-1 and C-2, and the concave cortical margin is opposed to the dural surface approximating the curve of the posterior ring of C-1. This configuration maximizes the contact surface between the graft and posterior arch of C-1. A midline notch is fashioned in the inferior cancellous portion of the graft to match the contour of the spinous process of C-2. To approximate the upward slope of the C-2 spinous process, the graft is notched deeper anteriorly than posteriorly (Fig. 4).

Once fitted, the graft is removed and the wires are placed. A loop of No. 24 twisted wire (three turns per centimeter, which corresponds to a maximal tensile strength) is passed from inferior to superior under the arch of C-1. This is a "two-handed process," and involves simultaneously feeding and pulling the wire to avoid pushing it anteriorly into the dura and underlying spinal cord and also to avoid pulling the C-1 ring back.
causing a posterior subluxation. Because the C-1 ring may be eroded and osteoporotic, care should be taken not to fracture the bone with this maneuver. The free loop of wire is then passed over the ring of C-1 and secured under the spinous process of C-2. A notch may be fashioned along the inferior lateral margins of the C-2 spinous process with a Kerrison punch to seat the wire.

The bone graft is then placed so that it is trapped between the free ends of the wire anteriorly and the loop of wire posteriorly. One free end of the wire is placed below the spinous process of C-2 in a notch previously created with a bone punch, and the entire contrast is tightened snugly. This procedure is performed by two people: one manipulates the individual segments of the wire loop so that they fit snugly, while the other tightens the end with a wire twister. The total C-1 graft/C-2 complex should function as a single unit. An intraoperative x-ray film is obtained at this time to assure appropriate C1–2 alignment (Fig. 5). Several areas of the exposed lamina of C-2, the posterior ring of C-1, and the cortical portion of the graft are decor- ticated with a high-speed drill and covered with fragments of cancellous bone previously harvested from the iliac crest. The wound is routinely closed in layers.

Postoperative Management

All patients are immobilized in the halo orthosis for 12 weeks postoperatively. Stable osseous fusion is then assessed with flexion and extension lateral cervical spine films with the halo disarticulated from the vest (Fig. 6). If stable osseous fusion is not achieved at 12 weeks, the halo orthosis is continued for an additional 4 to 8 weeks.

Operative Results

Thirteen patients (76%) developed a long-term stable osseous fusion. The mean follow-up period was 8.7 months (range 3 to 18 months). At the 12-week postoperative evaluation, three patients were found to have developed a fibrous union; these wore the halo vest for an additional 4 weeks. One of these patients, a 59-year-old man, developed a well-aligned fibrous union that has remained stable and, at his 1-year follow-up examination, showed signs of delayed ossification. The second patient, a 39-year-old woman, developed a well-aligned fibrous union, but at 1 year postoperatively showed approximately 2-mm movement between the flexion and extension films. The third patient, a 57- year-old woman, developed a 6-mm malaligned stable fibrous union. She has no associated neurological deficits and her preoperative symptoms of Lhermitte's disease have resolved. One patient, a 79-year-old woman with a severe progressive myelopathy, died several days after the operation. She underwent transoral resection of the odontoid and associated pannus followed 2 weeks later by a C1–2 fusion. Postoperatively, she developed a deep venous thrombosis for which the initial treatment was systemic anticoagulation; this was complicated by a bleeding diathesis and subsequent death.

There were no cases of neurological deterioration related to the operative procedure. Significant operative morbidity prolonged the hospital stay in two patients. One patient developed respiratory failure and a retropharyngeal abscess related to a previous transoral procedure. He required a tracheostomy and eventually made a good recovery after a prolonged course of antibiotic therapy. The second patient had pre-existing pulmonary disease and developed postoperative respiratory distress. She also required a tracheostomy for a short period and made a full recovery.

Three patients developed local inflammation at a halo apparatus pin site, which cleared after the pin position was changed and a short course of oral antibiotics was given. No other complications were related to the halo orthosis.
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**Discussion**

*Natural History*

In general, the severity of cervical spine involvement parallels the severity of peripheral joint involvement and overall systemic disease. The occurrence of atlantoaxial subluxation may be variable, related to such factors as the extent of disease, the interval until diagnosis, and the age of the patient; it has been reported in 19% to 71% of various groups of patients evaluated with rheumatoid arthritis.\(^1,3,19,23,25,26\)

Progression of subluxation is typical, although stabilization and, rarely, spontaneous fusion may occur. In a 5-year study of 106 patients with rheumatoid arthritis reported by Pellicci, *et al.*,\(^{23}\) 46 had radiographic evidence of an atlantoaxial subluxation at the initiation of the study. Eighty percent of those patients with subluxations demonstrated radiographic progression, and 20% of the patients without initial C1-2 instability developed radiographic evidence for new atlantoaxial subluxation. Thirteen percent of patients with initial subluxation developed neurological deterioration and required surgery. In an additional follow-up study by Smith, *et al.*,\(^{26}\) over a mean period of 4\(\frac{1}{2}\) years, 40% of patients with atlantoaxial subluxation showed significant radiological progression. In 45% of patients with subluxation between 3.5 mm and 5 mm the deficit progressed to 5 mm to 8 mm, and 10% of these progressed to more than 8 mm. Patients with systemic disease severe enough to require maintenance steroid therapy progressed even more rapidly.

Davis and Markley\(^4\) first documented the relationship between atlantoaxial subluxation in rheumatoid arthritis and sudden death. Martel and Abell\(^13\) and several others have subsequently emphasized that increased risk of sudden death occurs even in patients without antecedent evidence of neural compression. A postmortem series of 104 consecutive patients with rheumatoid arthritis revealed 11 with atlantoaxial subluxation and cervicomedullary compression.\(^13\) Seven of these 11 died suddenly, and the cause of death was thought to be fatal medullary compression. Once clinical evidence of a cervical myelopathy is established, the prognosis is even worse. Meijers, *et al.*,\(^{19}\) reported a 5-year follow-up study of 43 rheumatoid patients with cervical subluxations and associated myelopathy. All of their nine patients with myelopathy who were managed nonoperatively died within 1 year; four of the deaths were directly attributed to neural compression. Of the 34 patients treated operatively, 10 were alive at 5 years and only two deaths were related to fusion failure and cord compression. Marks and Sharp\(^{14}\) reported 31 patients with cervical myelopathy and rheumatoid arthritis; 19 of these died, 15 within 6 months of presentation. Four of these deaths were primarily due to neurological compromise. Death occurred in all of the untreated patients and half of those treated with only a cervical collar. Only operative fusion improved the opportunity for survival in these patients.

*Indications for Surgery*

Most authors agree that rheumatoid patients with atlantoaxial subluxation resulting in progressive myelopathy are appropriate candidates for surgical stabilization. Ranawat, *et al.*,\(^{24}\) first recommended early operation before appreciable neurological involvement occurs because 1) the myelopathy may be irreversible, 2) the prognosis is poor once symptoms of cord compression occur, and 3) the risk of sudden death is increased even in asymptomatic patients. They sug-
TABLE 2
Criteria for atlantoaxial stabilization in patients without neurological symptoms

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Subluxation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ranawat, et al., 1979</td>
<td>&gt; 8 mm</td>
</tr>
<tr>
<td>Conaty &amp; Mongan, 1981</td>
<td>&gt; 6 mm</td>
</tr>
<tr>
<td>Heywood, et al., 1988</td>
<td>&gt; 10 mm</td>
</tr>
<tr>
<td>Kourtopoulos &amp; von Essen, 1988</td>
<td>&gt; 6 mm</td>
</tr>
<tr>
<td>McCarroll &amp; Robertson, 1988</td>
<td>&gt; 8 mm</td>
</tr>
<tr>
<td>Santavita, et al., 1988</td>
<td>&gt; 9 mm</td>
</tr>
<tr>
<td>Clark, et al., 1989</td>
<td>&gt; 8 mm</td>
</tr>
</tbody>
</table>

Suggested that if mobile atlantoaxial subluxation is greater than 8 mm, patients should be considered for operative fusion.

In a retrospective examination of the x-ray films of 151 patients with rheumatoid arthritis and atlantoaxial subluxation, Weissman, et al., found that the probability of having clinical evidence of a myelopathy increases exponentially once the subluxation reaches approximately 9 mm. Concomitant cranial settling decreases the tolerance for atlantoaxial subluxation even further. This finding suggests that early "prophylactic" operative stabilization should be performed before atlantoaxial instability progresses to this severity.

Clark, et al., stated that, given the probability of progressive subluxation, an indication for cervical arthrodesis is radiological evidence of "impending neurological deficit," which they defined in the atlantoaxial region as a subluxation of 8 mm or more. Several other authors supporting early "prophylactic" fusion define between 6 and 10 mm of mobile atlantoaxial instability as the point at which operative stabilization should be considered (Table 2).

The decision regarding operative stabilization in patients without symptoms of neural compression should be made based on: the probability of progressive atlantoaxial subluxation, with an increased risk of irreversible neurological deficit and sudden death; the risk of the operative procedure; and the predicted life expectancy for the individual patient. As a group, patients with rheumatoid arthritis have a life expectancy 10 years shorter than the general population, irrespective of cervical subluxation. These patients typically have advanced systemic disease involving the endocrine, pulmonary, and cardiovascular systems, which additionally increase the risk of perioperative complications. Maintenance medications such as exogenous steroids and antimitobolites may also increase potential perioperative complications. Despite advances in anesthetic technique and perioperative medical management, the contemporary mortality rate associated with operative stabilization ranges from approximately 5% to 15%. Given these considerations, we believe that patients in relatively good health without advanced multisystem disease and aged less than 65 years should be considered for operative stabilization if mobile atlantoaxial subluxation is greater than 6 mm.

Operative Technique and Postoperative Care

Failure of stable osseous fusion after C1–2 arthrodesis may occur in as many as 50% of patients with rheumatoid arthritis. The underlying disease process, concomitant with the use of steroids and antimitobolites, decreases ossification and wound healing. The typical site of osseous failure is the interface between the bone graft and the posterior arch of the atlas. There is often upward migration and rotation of the posterior ring of the atlas with associated bone erosion and pannus formation related to the disease process. Once the posterior ring is exposed and the inferior portion is decorticated, a minimal surface area may be available for the graft interface. Stability at this interface is particularly susceptible to rotational strain.

This new method of atlantoaxial fusion, a single horizontal curvilinear strut wedged bilaterally between C-1 and C-2, provides immediate segmental and rotational stability. The bicortical orientation of the bone graft allows significant compressive force applied by the wire loop with maintenance of height and axial stability. The Brooks-type fusion may also provide immediate stability; however, the placement of C-2 sublaminar wires may inadvertently injure the spinal cord, and the placement of two bone grafts as opposed to one allows additional rotational movement.

The postoperative use of a halo orthosis provides superior rotational stability and improved fusion rates in rheumatoid arthritis patients. We routinely use a halo orthosis for 12 weeks postoperatively. Nonetheless, three (18%) of our 17 patients developed a fibrous union, which was probably partially attributable to the poor neo-ossification as previously mentioned. Despite efforts to minimize rotational movement, the site of the fibrous union was at the C-1 graft interface in two patients and at the C-2 graft interface in one patient. However, all three of these patients were managed early in this series when a tricortical bone graft was used. This resulted in cortical graft contact at both the C-1 and the C-2 interface as opposed to cancellous graft contact as indicated in the description of our current technique. Since this modification, we have achieved 10 consecutive osseous fusions.

Conclusions

Management of patients with rheumatoid arthritis and atlantoaxial instability may be challenging for many reasons. The decision regarding appropriate therapy should be individualized. Once signs and symptoms of an associated myelopathy occur, the prognosis worsens. Operative stabilization, which is almost always indicated for patients with a progressive myelopathy, provides the best opportunity for neurological recovery and survival. The indications for fusion in neurologically asymptomatic patients remain controversial. Several authors have previously suggested that, considering the typical progression of atlantoaxial subluxation in rheumatoid patients, the poor prognosis associated with
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the onset of a myelopathy, and the associated risk of sudden death, operative stabilization should be considered before the onset of neurological symptoms. We believe that patients under the age of 65 years without severe multisystem disease should be considered for operative stabilization if mobile atlantoaxial subluxation is greater than 6 mm. If operative stabilization is decided upon, this new fusion construct, in conjunction with a postoperative halo orthosis, offers a number of technical advantages and has yielded good results in this limited series.

References


Manuscript received July 3, 1989. Accepted in final form May 25, 1990. Address for Dr. Papadopoulos: Section of Neurosurgery, University of Michigan, Ann Arbor, Michigan. Address reprint requests to: Volker K. H. Sonntag, M.D., c/o Editorial Office, Barrow Neurological Institute, 530 West Thomas Road, Phoenix, Arizona 85013.