New approach to cervical flexion deformity in ankylosing spondylitis

Case report

SARAH E. DUFF, F.R.C.S., PAUL L. GRUNDY, F.R.C.S., AND STEVEN S. GILL, M.S.

Department of Neurosurgery, Frenchay Hospital, Bristol, United Kingdom

The treatment of cervical fixed flexion deformity in ankylosing spondylitis presents a challenging problem that is traditionally managed by a corrective cervicothoracic osteotomy. The authors report a new approach to this problem that involves performing a two-level osteotomy at the level of maximum spinal curvature, thereby achieving complete anatomical correction in a one-stage procedure.

This 48-year-old woman with ankylosing spondylitis presented with a 30-year history of progressive neck deformity that left her unable to see ahead and caused her to experience difficulty eating, drinking, and breathing on exertion. On examination, she exhibited a 90° fixed flexion deformity of the cervical spine, which was maximum at C-4; this was confirmed on imaging studies.

A two-level osteotomy was performed at C3–4 and C4–5 around the area of maximum spinal curvature, and the deformity was corrected by extending the head on its axis of rotation through the uncovertebral joints. The spine was stabilized using a Ransford loop. An excellent anatomical position was achieved, as was complete correction of the deformity.

A two-level midcervical osteotomy performed at the level of maximum spinal curvature in ankylosing spondylitis enables complete correction of severe fixed flexion deformity in a single procedure. Preservation of the uncovertebral joints allows smooth and safe correction of the deformity about their axis of rotation.

Key Words • osteotomy • ankylosing spondylitis • cervical spine

ANKYLOSING spondylitis frequently results in fixed flexion deformities of the spine, typically in the thoracolumbar area; cervical deformities are less frequently encountered. Patients with severe cervical flexion deformities can be significantly disabled and may be unable to see directly ahead; in severe cases they may also have difficulty eating and breathing. These patients may be treated by means of a corrective cervicothoracic osteotomy; this procedure is usually performed at the C7–T1 level, as described by several authors.5,7,8,10 Osteotomies at other levels are rarely reported because of the significant hazards associated with such a procedure, including the risk of damage to the VA.2,3,5,11

We report a new approach for the correction of fixed flexion deformity in the cervical spine in ankylosing spondylitis. We performed a two-level osteotomy at the level of maximum curvature of the cervical spine (C-4), providing one-stage correction and stabilization by internal fixation. To our knowledge, this is the first reported case of a two-level midcervical osteotomy in ankylosing spondylitis in which a correction of approximately 90° was achieved.

Case Report

History and Presentation. This 48-year-old woman was referred to our hospital because of progressive neck deformity. She had been diagnosed as having ankylosing spondylitis 5 years previously on the basis of radiographically confirmed changes in the thoracolumbar and sacroiliac spine areas as well as a positive result on HLA B27 testing. She had never had any other joint problems, uveitis, or iritis.

Her neck deformity had begun to develop at 16 years of age following a traffic accident in which no fractures were documented. The severity of the deformity had worsened significantly during the previous 5 years, leading her to seek medical advice. At presentation she was unable to move her neck in any plane and had difficulty eating and swallowing. She was unable to open her mouth far enough to eat or drink easily; all solids had to be chewed very thoroughly and she could only drink fluids by using a straw. She was also becoming progressively short of breath during minimal exertion, stating that she “couldn’t get enough air in.” She had no neurological symptoms but was increasingly disabled socially because of the deformity.

Examination. On examination, she was found to have a...
striking fixed flexion deformity of the neck measuring approximately 90°, resulting in her chin being in contact with the lower part of her neck and slightly rotated to the left. Examination was otherwise unremarkable; in particular, she had no neurological abnormalities and no fixed flexion deformity of the hips.

Preoperative studies included plain cervical spine x-ray films, which confirmed the degree of flexion deformity, with the area of maximum flexion centered on C-4 (Fig. 1); magnetic resonance images, which revealed that the spinal cord was inclined toward the right of the canal but without evidence of compression; and three-dimensional computerized tomography scans that demonstrated the patient’s bone anatomy and which we used to plan the surgery (Fig. 2). These studies confirmed the extent of the deformity and also demonstrated significant atlantoaxial subluxation.

In view of the degree of deformity, a cervicothoracic osteotomy was not considered advisable because this procedure would create an S-bend below the area of maximum flexion. This would have resulted in the lower part of her neck projecting directly backward and would keep her chin hyperflexed, which may not have improved her airway and esophageal problems and could possibly have created spinal cord impingement.

A higher approach was planned, with two osteotomies centered around the area of maximum flexion: C3–4 and C4–5. This approach entailed the risk of damaging the VAs, but we believed that it would allow us to achieve a more anatomically appropriate correction by addressing the deformity directly, thus resulting in a greater degree of correction.

Despite the relatively high risks involved in the procedure, the patient was so disabled by her deformity that she consented to surgery.

Operation. Surgery was performed by the senior author (S.S.G.). The patient underwent fiberoptic intubation while awake and a state of anesthesia was induced. She was positioned prone on the operating table with her head held on blocks in a Mayfield three-pin head fixator, and she was strapped and taped to the table so that her body would be completely immobilized before the head end of the table was elevated. Electrophysiological spinal cord monitoring was performed intraoperatively using somatosensory evoked potentials.

A midline incision was made from the external occipital protuberance to T-2. The patient’s posterior cervical musculature was considerably atrophied; the muscles were dissected from the occiput, spinous processes, and C1–T2 laminae in the standard fashion. A C-1 laminectomy was performed and intersegmental decompressions with removal of the ligamentum flavum were performed at all levels down to T1–2 to allow for placement of instrumentation. Significant blood loss was encountered from the epidural veins at this stage because of an elevated venous pressure, probably caused by the patient’s prone position, and from abdominal compression by the positioning blocks. Ideal positioning was difficult because of the patient’s short stature and her cervical deformity. A C4–5 wedge osteotomy was performed through the facet joints, and the C-5 nerve roots were exposed. The laminae and spinous processes of C-4 and C-5 were preserved. The ankylosed disc space was exposed on the left side between the uncovertebral joint and the theca medially, the VA laterally, and the nerve root superiorly. This space was drilled through using a 2-mm cutting burr with the aid of image intensifier screening, thus preserving the uncovertebral joints. Correction was performed by releasing the head fixator and extending the head gently, allowing correction about an axis of rotation on the uncovertebral joints. During this procedure, electrophysiological monitoring revealed normal parameters. A sublaminar wire was passed and used to hold C-4 and C-5 firmly together in the corrected position, so that they would act as a single unit when the correction procedure was performed again at the higher level. The wedge corrective osteotomy was repeated in the same manner at C3–4. Occipital and sublaminar wires were passed and a Ransford loop was secured from the occiput to T-1. A bone graft was obtained from the spinous processes and the C-1 lamina was packed around the osteotomies. Two air emboli occurred during the procedure; they were detected by a fall in the end-tidal carbon dioxide level and were managed by performing a Valsalva maneuver and covering the bleeding vessels with damp packs. The wound was closed in layers with interrupted Vicryl sutures and staples were applied to the skin.

Postoperative Course. The patient required intensive care support for 10 days postoperatively because of respiratory difficulties secondary to bibasal collapse, probably caused by a combination of a large volume transfusion, supine positioning, and the length of the procedure. Postoperative radiographic studies revealed correction of the deformity and satisfactory positioning of the Ransford loop (Fig. 3). The patient developed transient dysphagia...
and hoarseness, then made an excellent recovery and was delighted with the correction achieved. She was discharged home 17 days postoperatively independently mobile, with a mild weakness of shoulder abduction on the right side that resolved within 3 weeks.

Discussion
The indications for surgery in cervical spine deformity were first described by Law.3 These include: 1) correction of severe deformity by elevating the chin from the sternum and enabling forward vision; 2) prevention of atlantoaxial subluxation and dislocation; 3) relief of esophageal and tracheal compression; and 4) prevention of spinal cord kinking or undue traction on the nerve roots resulting in neurological disturbance. Candidates for these procedures are also often severely socially and professionally disabled by the deformity, and surgery may be contemplated for this reason.

The first osteotomy for flexion deformity in ankylosing spondylitis was performed in the lumbar spine by Smith-Petersen, et al.,9 in 1945. Treatment of the rarer cervical deformities has proved more difficult. Mason and colleagues5 in 1953 described performing a cervical osteotomy through the body of T-1 via an anterior approach followed by gradual correction of the deformity by using Crutchfield tongs. This procedure was performed after the patient had received a local anesthetic agent and nitrous oxide; the patient lay in the lateral decubitus position. These authors also emphasized the relative safety of performing the osteotomy at the C7–T1 level, thus avoiding the VAs.5 In 1958, Urist10 reported using a posterior approach to treat the same condition, with the patient sitting and the surgery performed using a local anesthetic. In these cases, the correction was made by the use of adjustable braces during the postoperative period. Simmons7,8 went on to modify the method introduced by Urist in a large series of patients. He again used a local anesthetic and placed the patient in the sitting position; the osteotomy was performed at the cervicothoracic junction and the correction was made by extending the neck after the osteotomy and full nerve root decompression had been accomplished. Stabilization was achieved using a halo jacket.

Use of a local anesthetic for cervical flexion deformity was considered to be very important in the early days of surgery. It avoided the problem of a technically difficult or impossible intubation, avoided the risk of postoperative respiratory complications (to which these patients are prone), and allowed important feedback from the patients regarding any neurological symptoms they might experience during surgery and correction. However, pain relief was frequently less than ideal and the use of a local anesthetic in some cases led to cardiac arrest, toxicity reactions, and hypotension.7 Technical improvements, in particular the use of the fiberoptic bronchoscope,4 allowed for easier intubation in these patients, often via the nasotracheal route while the patient was awake. Thus surgery is now usually performed using a general anesthetic for greater patient safety and comfort. Routine use of intraoperative electrophysiological monitoring allows important feedback during surgery, which in the past would have come from the awake patients’ responses.

When using a local anesthetic the patient was traditionally placed upright, using the halo jacket and traction to provide head control when the deformity was corrected. However, we believed that in this case the sitting position would not allow sufficient surgical access for a two-level osteotomy and might increase the risk of air emboli; therefore, the patient was placed prone. However, this position is not without difficulties, in particular excessive intraabdominal pressure and elevated venous pressure, which may cause bleeding problems.

Gradual correction of deformity by using adjustable fixation devices has fallen out of favor in the wake of improvements in intraoperative monitoring and internal and external fixation techniques. It is now more common to perform correction as a single-stage procedure and to achieve stabilization with a combination of fixation methods. Internal fixation alone may run the risk of implant failure or cut out through osteoporotic bone, which is often encountered in patients with ankylosing spondylitis.

Correction at two levels, as described here, enabled a
large degree of angulation to be made (approximately 90°). Previous cervicothoracic osteotomies have achieved much less correction: McMaster describes a range of 30 to 71°, with a mean of 54°; Law reports a range of 20 to 30°; and Weale and Marsh a range of 15 to 50°, with a mean correction of 31°.

Preservation of the laminae at the corrected levels provides additional stability and facilitates fixation of the corrected levels posteriorly with sublaminar wires. A long occipitocervicothoracic fixation in which the surgeon uses a Ransford loop not only provides stable fixation for the osteotomized levels but should also prevent recurrence of the flexion deformity, which might otherwise happen if the weight of the head causes reshaping of the fused cervical spine.

Many complications have been described in association with corrective osteotomies for fixed flexion deformities in ankylosing spondylitis. In the early days of corrective surgery mortality was 10%1, often because of damage to the VAAs. Other complications include neurological damage (for example, spinal nerve injury), bone nonunion, pseudarthrosis and subluxation at the osteotomy site, and respiratory and cardiovascular problems. In our patient, the major complication encountered was intraoperative bleeding and two air emboli created during surgery despite the prone position. These were treated rapidly and resulted in no long-term sequelae. In the postoperative period, the patient developed pulmonary collapse and transient dysphagia and hoarseness; these resolved with conservative treatment.

Conclusions

We describe a novel approach to the difficult technical challenge presented by a patient with a severe fixed flexion deformity of the cervical spine in association with ankylosing spondylitis. The two-level corrective osteotomy was performed at the level of maximum curvature of the spine rather than at the cervicothoracic junction, allowing anatomical correction. We believe that it is important during correction of the deformity to allow rotation around the axis of the uncovertebral joints. The correction reported here led to adequate stabilization with internal fixation alone. This approach provides an anatomical correction of the deformity and stabilizes the entire cervical spine, thus reducing the risk of subluxation at the osteotomy site. However, great care must be taken not to damage the VAAs during the procedure.

References


Manuscript received October 4, 1999. Accepted in final form June 5, 2000.
Address reprint requests to: Sarah E. Duff, F.R.C.S., 115 Howard Road, Westbury Park, Bristol BS6 7UZ, United Kingdom. email: saraheduff@aol.com.

S. E. Duff, P. L. Grundy, and S. S. Gill

Fig. 3. Postoperative lateral cervical spine radiograph confirming anatomical correction and adequacy of fixation.