Cervicothoracic Extension Osteotomy for Chin-on-Chest Deformity in Ankylosing Spondylitis

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Background: Chin-on-chest deformity is a disabling manifestation of ankylosing spondylitis. Surgical treatment consists of extension osteotomy at the cervicothoracic junction. The purpose of this study was to characterize the clinical presentation of this deformity and to determine the long-term functional and radiographic outcomes of treatment.

Methods: The medical records and radiographs of all twenty-six patients treated with cervicothoracic extension osteotomy by one of us between 1976 and 2001 were retrospectively reviewed. Three patients died during the two-year-minimum follow-up period. The remaining twenty-three patients were followed for an average of 4.5 years (range, two years to twenty-one years and ten months).

Results: The mean sagittal correction was 38°. Delayed union in two patients and additional cervical trauma in two others resulted in partial loss of the initial correction. Quadriplegia developed in one patient, who died as a result of subluxation at the osteotomy site. Five patients had irritation of the eighth cervical nerve root postoperatively.

Conclusions: Extension osteotomy can reliably improve sagittal alignment and horizontal gaze as well as decrease neck pain, eating difficulties, and neurologic abnormalities. Internal fixation is recommended to prevent subluxation, delayed union, nonunion, loss of correction, or neurologic injury. There is a risk of death or catastrophic neurologic injury from the procedure.

Level of Evidence: Therapeutic Level IV. See Instructions to Authors for a complete description of levels of evidence.

Ankylosing spondylitis is often complicated by kyphotic spinal deformity. Usually the hyperkyphosis is distributed throughout the thoracic and lumbar spine, but occasionally a patient will present with a disproportionate kyphosis in the cervicothoracic spine. The resulting chin-on-chest deformity may contribute to problems with horizontal gaze, chewing and swallowing, hygiene, neck pain, and myelopathy.

The surgical treatment to correct symptomatic chin-on-chest deformity secondary to ankylosing spondylitis is an extension osteotomy and arthrodesis at the cervicothoracic junction. To the best of our knowledge, there has been no long-term follow-up study of a large series of patients with chin-on-chest deformity who were treated surgically with one technique. Mason et al. established the importance of performing the osteotomy at the cervicothoracic junction, below the entrance of the vertebral arteries into the transverse foramina at the sixth cervical vertebra[1]. In 1958, Urist described the same surgical technique, with use of a local anesthetic in a seated, awake patient[2]. Several other authors have reported on series of patients treated with minor modifications of Urist's technique[2-7]. However, in those series, the duration of follow-up was no more than three years for any given patient or was not specified, preoperative neurologic abnormalities were not described, and the sagittal alignment of the cervical spine was not clearly defined. The purpose of this study was to characterize the clinical presentation of chin-on-chest deformity secondary to ankylosing spondylitis and to determine the long-term functional and radiographic outcomes of treatment with cervicothoracic extension osteotomy.

Materials and Methods

Between 1977 and 2001, the senior author (H.H.B.) performed twenty-six spinal osteotomies for chin-on-chest deformity (Figs. 1-A, 1-B, and 1-C) in patients with ankylosing spondylitis (see Appendix). The medical records and radiographs of each patient were evaluated retrospectively. An attempt was made to contact each patient by telephone to re-

quest that they return for an additional follow-up evaluation, but only two patients actually returned. Each patient was evaluated preoperatively; at six weeks, three months, six months, and one year postoperatively; and then annually for a minimum of two years.

Three patients died during the two-year-minimum follow-up period. Two of the deaths were late deaths that were believed to be unrelated to the osteotomy. One patient (Case 6; see Appendix), who was diagnosed with an esophageal carcinoma two months after the osteotomy, died six months after the osteotomy secondary to respiratory failure and congestive heart failure. The other patient (Case 7) died of sudden cardiac arrest, while in a nursing home, nine months after the osteotomy. No autopsy was performed. The third patient (Case 14) became quadriplegic after the osteotomy and died of pneumonia and a cardiac arrest in the immediate postoperative period. The average duration of follow-up for the remaining twenty-three patients was four years and six months (range, two years to twenty-one years and ten months). There were twenty-five men and one woman. Their average age at the time of surgery was fifty-one years (range, thirty-one to seventy-three years).

The medical history of each patient had been recorded by the senior author and included information about trauma related to the onset of the deformity and the patient's subjective descriptions of neck pain, swallowing difficulties, field of vision, and neurologic symptoms. The severity of each of these problems was rated by the patient at each visit as mild, moderate, severe, or none. Twelve patients had a history of trauma preceding noticeable progression of neck pain and/or deformity. At the time of presentation, all twenty-six patients had difficulty with horizontal gaze, twenty-five had neck pain, twenty had difficulty swallowing, and thirteen patients had difficulty shaving.

The findings of the physical examination of each patient, which included a thorough assessment of the neurologic status, had been recorded by the senior author at each office visit. A Nurick grade was assigned to classify the neurologic status of each patient at each time-point[8]. The severity of the deformity was assessed qualitatively, but consistent, standardized measurements were not made. Of the twenty-six patients, sixteen were neurologically normal at presentation and ten had neurologic deficits, including seven with hyperactive reflexes and a minor gait abnormality (Nurick grade 1 or 2) and three with a major gait disturbance or quadriparesis (Nurick grade 3 or 4) (see Appendix). Two of the three patients with a Nurick

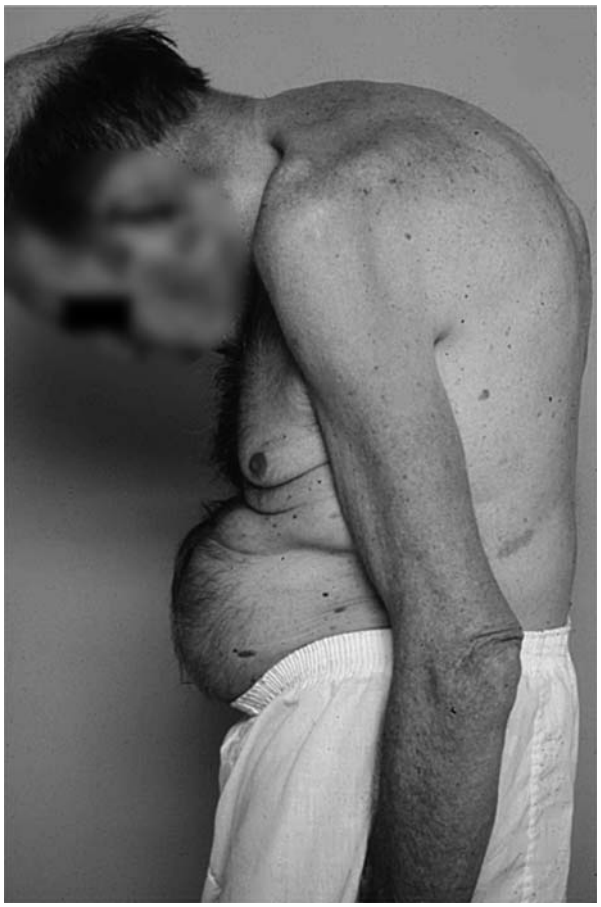

Fig. 1-A

Fig. 1-B

Figs. 1-A, 1-B, and 1-C Case 10 (see Appendix). This patient presented with a severe chin-on-chest deformity secondary to ankylosing spondylitis. He had hyperactive reflexes in the lower extremities, neck pain, difficulties with chewing and swallowing, a poor field of vision, and bilateral arm dysesthesias at the time of the initial evaluation. He had no history of trauma. **Fig. 1-A** Preoperative lateral photograph demonstrating the profound deformity. **Fig. 1-B** The chin is in contact with the sternum during acquisition of the preoperative T1-weighted magnetic resonance image with the patient supine. Also note the severe hyperkyphosis of the upper thoracic spine and the neutral to slightly lordotic alignment of the cervical spine.

grade of 3 or 4 had a history of trauma that had caused a fracture of the lower cervical spine or the cervicothoracic junction. The fractures had been treated with laminectomy and halo immobilization, with worsening of the neurologic status prior to presentation. The third patient had been treated with a halo vest for more than a year prior to presentation.

The radiographs of each patient were evaluated by orthopaedic surgeons who were not involved in the care of the patients. The deformities were measured on plain radiographs, tomograms, sagittal reconstructions of computerized axial tomography scans, or sagittal magnetic resonance images of the cervicothoracic spine. The initial degree of correction and the subsequent loss of correction were determined by comparing measurements from identical reference levels for each patient.

Fig. 1-C

An intraoperative photograph showing the laminectomy and wide foraminotomies over the eighth cervical nerve roots, with buckling of the dural sac on extension at the osteotomy site. The patient had arm dysesthesias and intrinsic hand-muscle weakness postoperatively. There was a substantial decrease in the hyperreflexia, chewing and swallowing difficulties, and neck pain and a substantial improvement in the field of vision immediately. The arm dysesthesias and intrinsic hand-muscle weakness gradually decreased, with about 80% of the sensation and strength returning over a two-year period.

TABLE I Methods of Internal Fixation

Type of Internal Fixation	No. of Patients
None	7
Spinous process wire	2
Sublaminar wire	1
Triple wire	2
Luque rectangle with Drummond wires	14

The reference levels were determined for each patient individually, depending on which levels were clearly visible on all preoperative and postoperative radiographic studies. Generally, the reference levels were from the superior end plate of C5 to the inferior end plate of T5.

Cervical alignment was measured preoperatively in the sagittal and coronal planes from the inferior end plate of C2 to the inferior end plate of C6. The average preoperative sagittal alignment of the cervical spine was 12° of lordosis, with a range of 2° of kyphosis to 31° of lordosis. Thus, the deformity was mainly in the upper thoracic spine or at the cervicothoracic junction. Fusion status was determined with serial radiography and with either plain or computerized tomography of the cervicothoracic junction. The absence of a persistent lucent line at the osteotomy site or the presence of continuous trabecular bone across the level of the osteotomy was considered to be evidence of a solid fusion.

Three patients had had a previous atlantoaxial arthrodesis for the treatment of instability, and twenty-four patients had complete ankylosis of the cervical spine caudad to C2. No patient had evidence of scoliosis in the cervical spine.

Surgical Procedure

Surgery was performed, with the use of local anesthesia, while the patient was awake and seated and in halo traction[9] (Figs. 2-A through 2-D). Spinal cord monitoring was not used since we believe that an alert patient serves as the most accurate spinal cord monitor. Complete laminectomy was performed at the C6 and C7 levels. Complete foraminotomies were carried out bilaterally over the C8 nerve roots at the C7-T1 joints. A portion of the C7 or T1 pedicle was removed bilaterally to make room for the C8 nerve roots. If internal fixation was to be used, it was applied at this point, but it was not secured. After administration of a brief general anesthetic, the patient's head was then slowly and carefully extended while the osteotomy site was observed posteriorly. The osteotomy was completed anteriorly by means of osteoclasis. Excessive force or sudden movement was avoided by using overhead traction and an assistant to control the halo ring anteriorly. The dura generally buckled slightly on extension of the osteotomy (Fig. 1-C).

The osteotomy site was then secured with internal fixation; a wide variety of fixation methods was used (Table I). During the earlier years in which this series of patients was treated, the only internal fixation available was wire. Many early procedures were performed without internal fixation. As wiring

TABLE II Severity of Neck Pain

	No. of Patients			
	None	Mild	Moderate	Severe
Preoperative	2	5	12	7
Six weeks postoperatively	6	16	2	1
Final follow-up	10	13	2	0

became more common, it was incorporated into the technique. When the Luque rectangle (Medtronic Sofamor Danek, Minneapolis, Minnesota) became available, it was used for most of the recent operations. Once the correction and fixation had been achieved, local bone autograft generated by the laminectomies and foraminotomies was morselized and placed over the osteotomy site prior to closure. Other sources of bone graft material were generally not utilized.

The postoperative treatment included external immobilization with a halo ring and a molded plaster jacket for three months. Removal of the plaster jacket was followed by use of a rigid two-poster cervicothoracic orthosis for an additional three months. Pin-site or cast-related problems resulted in earlier conversion to the brace for four patients, but generally the orthosis was not used before two months postoperatively. Plain or computerized tomography was performed for all patients at three-month intervals to determine the position of the osteotomy site and the fusion status until the osteotomy site was observed to be fused radiographically.

Results

Two patients did not have neck pain preoperatively and remained pain-free at the time of final follow-up. Of the twenty-four patients with preoperative neck pain, twenty-one had less pain at the time of final follow-up, one who had had mild neck pain preoperatively had no change, one died in the immediate postoperative period, and one with moderate neck pain preoperatively had (unexplained) initially severe neck

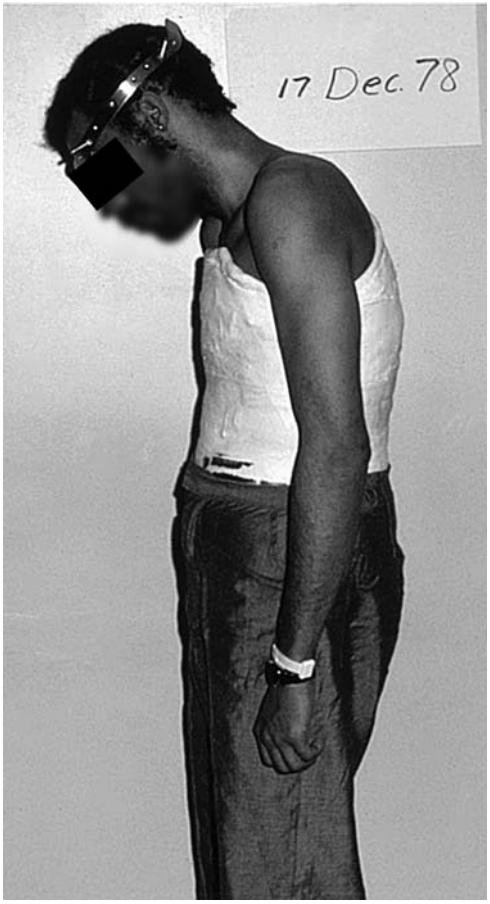

Fig. 2-A

Fig. 2-B

Figs. 2-A through 2-D Case 18 (see Appendix). This patient presented with neck pain, myelopathy, and a chin-on-chest deformity secondary to ankylosing spondylitis. He had no history of trauma. **Fig. 2-A** Preoperative lateral photograph showing the cervicothoracic deformity. The halo ring and plaster jacket were applied preoperatively. **Fig. 2-B** Intraoperative photograph showing the laminectomy and wide foraminotomies over the eighth cervical nerve roots. Repeat arthrodesis with interspinous wiring was required because of delayed union at four months after the osteotomy.

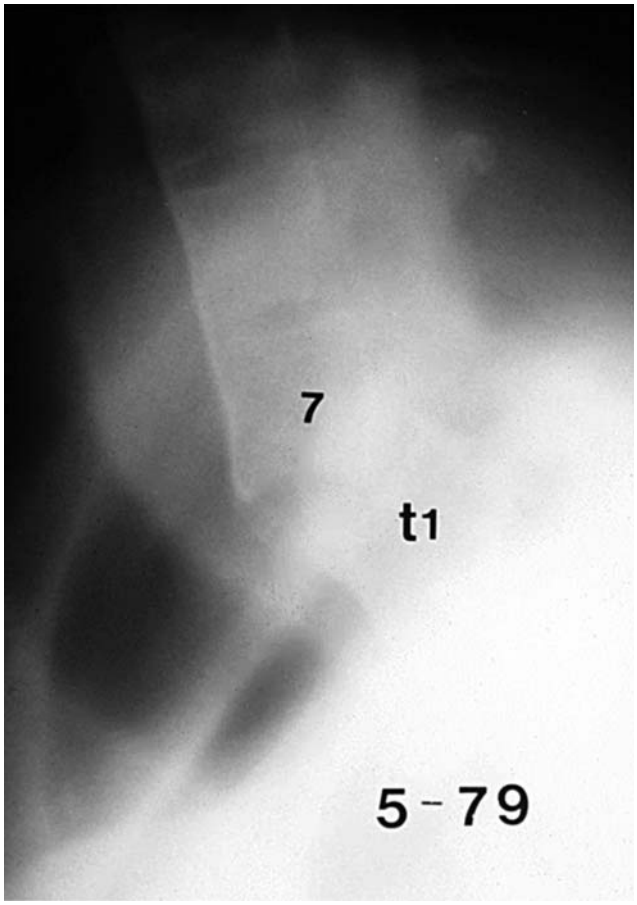

Fig. 2-C

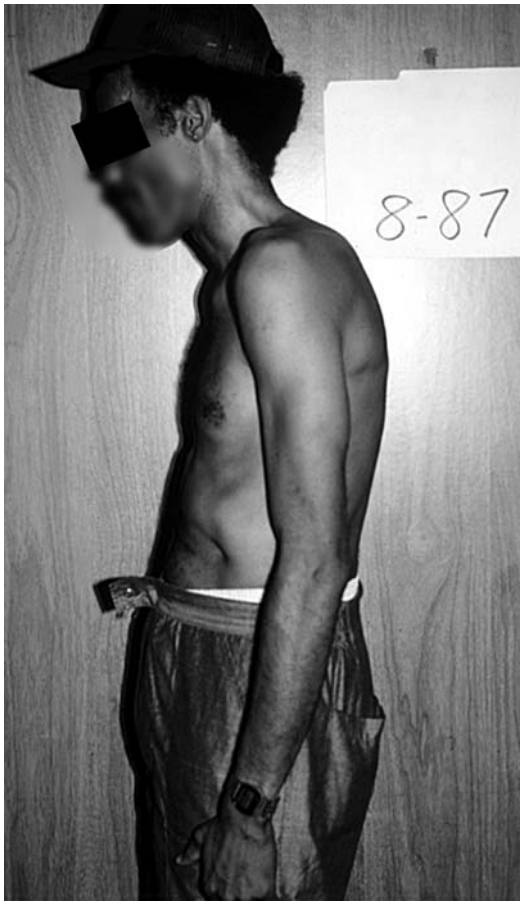

Fig. 2-D

Fig. 2-C Postoperative lateral tomogram showing anterior osteoclasis and correction of the kyphosis at the cervicothoracic junction. **Fig. 2-D** Lateral photograph made eight years and three months postoperatively, showing excellent alignment of the head. The patient had dysesthesias in the right hand postoperatively, but these symptoms resolved after three months. He reported a decrease in neck pain and weakness and an improvement in the field of vision, balance, and overall quality of life at the time of final follow-up, at twenty-one years and ten months postoperatively.

pain postoperatively, which was moderate at the time of final follow-up (Table II). Of nineteen patients who had difficulty swallowing preoperatively, eighteen reported an improved swallowing ability at the time of final follow-up and one, who had no improvement, was ultimately diagnosed with esophageal carcinoma. Transient dysphagia occurred in ten patients in the immediate postoperative period, and it resolved completely by the six-week follow-up evaluation in all cases. Horizontal gaze and the field of vision were not systematically measured, but they were subjectively improved in all patients.

Of the sixteen patients who had been neurologically normal preoperatively, one became quadriplegic, one had transient arm dysesthesias that quickly resolved, and fourteen remained neurologically normal. Of the ten patients with preoperative neurologic deficits, one still had hyperactive reflexes and clonus at the time of follow-up but the remaining nine patients had improvement of one, two, or three Nurick grades (see Appendix).

One patient died one week postoperatively. Of the twenty-five remaining patients who were evaluated with plain or computerized tomography at three months postoperatively, twenty-two were seen to have fusion at the osteotomy site. Two patients were found to have fusion when they were re-evaluated at six months, and one patient underwent repeat bone-grafting and fixation with a single interspinous wire four months postoperatively and subsequently had a successful fusion. Four patients had 5 mm of anterior subluxation at the osteotomy site at the time of the initial follow-up, without adverse sequelae. The anterior opening wedge correction (the osteoclasis) was at the C7-T1 level in twenty-three patients, at the C6-C7 level in two, and at the C5-C6 level in one. There were no adverse consequences related to the corrections that were inadvertently performed cephalad to C7-T1.

The mean initial sagittal correction was 38°, with a range of 15° to 84°, and there was a mean loss of correction of 2.6°, with a range of 3° of improvement to a 20° loss of correction, at the time of final follow-up. Of the five patients who lost ≥5° of the initial sagittal correction, two had a delayed

union and the other two had sustained additional trauma to the cervical spine that had resulted in some loss of correction in the interval before the final follow-up evaluation. In all remaining patients, the correction seen at the time of final follow-up was within 5° of the initial correction.

Neurologic Complications

A variety of neurologic complications occurred. The most devastating occurred in a patient (Case 14, see Appendix) who was found to be quadriplegic while he was still in the operating room immediately after the osteotomy. He underwent urgent reexploration with repositioning of the obviously subluxated osteotomy site and had almost complete neurologic recovery over the next three days. However, he ultimately died of pneumonia and cardiac arrest while still in the intensive care unit. Radiographs made at the time suggested that the osteotomy site had resubluxated despite immobilization in a halo cast.

Five patients had new upper-extremity pain, dysesthesias, or weakness in the immediate postoperative period. These symptoms completely resolved within six months in three patients, whereas they decreased over a period of about two years in the remaining two patients, who never regained a normal status. One patient (Case 1, see Appendix) had a seizure in the immediate postoperative period and had altered mental status with left-sided weakness that persisted for five days; she recovered without any permanent neurologic sequelae.

Discussion

Most patients with ankylosing spondylitis do not have a loss or reversal of cervical lordosis alone. Rather, the most severe kyphotic deformity is found at the cervicothoracic junction and in the upper thoracic spine. Thus, extension osteotomy should be carried out not in the cervical spine per se, but at the cervicothoracic junction. Such intervention does not change the thoracic kyphosis, but it does correct the sagittal alignment of the cervical spine and the head in relation to the thoracic spine.

Neurologic abnormalities are frequent in patients with ankylosing spondylitis who have a chin-on-chest deformity. In this series of twenty-six patients, ten had neurologic deficits preoperatively, with findings ranging from mild myelopathy in seven patients to severe myelopathy in three. Spinal cord dysfunction may be due to the exaggerated cervicothoracic kyphosis causing relative lengthening or distraction of the posterior spinal column and draping of the spinal cord over the posterior vertebral bodies. Neurologic signs and symptoms decreased with successful cervicothoracic extension osteotomy in nine of the ten patients.

McMaster reported a mean sagittal correction of 54° (range, 30° to 71°) with an average loss of correction of 5.7° in a series of fifteen patients treated with a cervicothoracic extension osteotomy for chin-on-chest deformity secondary to ankylosing spondylitis[6]. In our series, the mean sagittal correction was 38° (range, 15° to 84°) with a subsequent mean loss of correction of 2.6° at the time of final follow-up. The excellent maintenance of correction in our patients may have been a result of the increased use of instrumentation, aggressive external immobilization, and achievement of a high fusion rate. The initial correction of sagittal alignment was maintained over time, with the notable exceptions of patients who had a postoperative fracture, delayed union, or subluxation.

Successful fusion of the osteotomy site was readily achieved in this series of twenty-six patients; one patient died immediately postoperatively and the remaining twenty-five had a solid fusion at six months, including one who had a reoperation because of delayed union. Simmons reported two nonunions in forty-two patients who had been treated with anterior arthrodesis[5]. McMaster reported nonunion in two of fifteen patients who had been treated with anterior arthrodesis[6]. All forty-two of the patients in Simmons' series and twelve of the fifteen patients in McMaster's series were treated without internal fixation. Delayed union occurred in three patients in our series, two of whom had been treated without internal fixation and one of whom had been treated with a single interspinous wire for fixation. One of the patients treated without internal fixation underwent subsequent repeat bone-grafting and interspinous wiring, whereas the other two did not require additional surgery.

Anterior subluxation of ≥5 mm occurred in five of the twenty-six patients in our series, with one patient sustaining a severe neurologic injury. McMaster reported subluxation in four of the fifteen patients in his series[6]; nonunion developed in two of them and irritation of the eighth cervical nerve root, which resolved, developed in the other two. All of the subluxations in the current series and in McMaster's series occurred in patients who had been treated without internal fixation. We therefore recommend the use of secure internal fixation to prevent subluxation. A Luque rectangle with Drummond wires (Zimmer, Warsaw, Indiana) appears to be sufficient. Other methods, such as lateral mass or pedicle screw fixation, may be suitable, but generally there is not enough room posteriorly to expose the second thoracic vertebra with the plaster jacket in place. External immobilization with a halo is advisable in the setting of rigid internal fixation because of the biomechanical difficulty of immobilizing the stiff, long segments of the ankylosed spine and the generally osteoporotic bone that is characteristic of these patients.

Post-osteotomy complications related to irritation of the eighth cervical nerve root in these patients are usually dysesthesias, which tend to resolve uneventfully over time. However, when intrinsic hand-muscle weakness occurs it can persist. Care must be taken during closure of the posterior osteotomy site, and a wide foraminotomy and pediculectomy are essential. As emphasized by Simmons, it is helpful to perform the osteotomy in an awake patient so that, if necessary, adjustments can be made in the final position of the head in the controlled surgical environment[5]. A disadvantage of rigid internal fixation in this setting may be that it prevents such minor adjustments.

When considering cervicothoracic osteotomy for a given patient, the surgeon must weigh the risks of neurologic injury

against the functional improvement and pain relief that can be achieved with successful correction of the spinal deformity. The likelihood of relieving neck pain, improving altered neurologic status, achieving fusion at the osteotomy site, improving horizontal gaze, and improving swallowing ability is high.

Appendix

eA Tables presenting details of the patients' history, radiographic characteristics, neurologic function, and postoperative complications are available with the electronic versions of this article, on our web site at jbjs.org (go to the article citation and click on "Supplementary Material") and on our quarterly CD-ROM (call our subscription department, at 781-449-9780, to order the CD-ROM). ■

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