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Complex Deformities of the Cervical Spine

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Historically, complex deformities of the cervical spine have been limited to patients afflicted with congenital scoliosis, systemic inflammatory diseases, and bony dysplasias. Improved diagnosis and early medical and surgical treatment of these problems have led to better long-term outcomes and mitigation of the development of severe deformities in the cervical spine that occur as a late sequela of the disease processes. Thus, primary deformities of the cervical spine have seen a decreased incidence over the past decade. In their place is a growing incidence of postsurgical deformities that have developed as a consequence of the increased use of fusion to treat degenerative diseases of the cervical spine. Multilevel cervical fusions and long decompressions in the younger age group combined with a longer life span as a result of improvements in health care and such factors as osteoporosis have created a cohort of patients at risk for developing cervical deformity from graft subsidence and collapse, adjacent segment disease, pseudarthrosis, hardware failure, and the sequelae of chronic neurologic injury. This article focuses on primary and secondary causes of deformity in the cervical spine and the surgical approaches to treatment (Box 1).

Primary deformities of the cervical spine

Congenital scoliosis

Congenital osseous anomalies of the cervical spine are rare. On the basis of genetic data available from the evaluation of patients with Klippel-Feil syndrome, these anomalies are thought to arise from dysfunction of a family of "housekeeping"-type genes in the Hox family [1,2]. These genetic mutations lead to a failure of segmentation (type II) or a failure of formation (type I). Most of these anomalies involve congenital fusions or nonfusions of phenotypically normal-appearing vertebrae (failure of segmentation). In these instances, the importance of recognizing these syndromes lies in the awareness that the vertebral anomalies are often accompanied by developmental defects in multiple other organ systems. In patients with congenital scoliosis, the incidence of cardiac abnormalities is increased, as is the incidence of renal abnormalities (20%–30%) and intracanal abnormalities (10%–50%) [3,4]. Abdominal ultrasound or other imaging tests should be used to rule out absent or abnormal kidneys [5]. Intracanal abnormalities may include a syrinx (cyst within the cord), diastematomyelia or diplomyelia (division or reduplication of the cord, respectively), and tethered cord (presence of a tight filum terminale that does not permit the conus medullaris to migrate upward normally with growth) [6,7].

More rarely, a failure of formation can occur in a critical location in the spine, such as the

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Box 1. Causes of cervical deformity

Primary
Congenital scoliosis
Hemivertebrae
Osteogenesis imperfecta
Fibrodysplasia ossificans progressive
Neurofibromatosis

Secondary
After surgery
After trauma

Degenerative/inflammatory
Rheumatoid arthritis
Ankylosing spondylitis
Cervical spondylosis (idiopathic)

cervicothoracic junction, and leads to a progressive loss of coronal and sagittal balance that can be functionally debilitating and cosmetically unacceptable [8]. Mixed abnormalities are also found in patients with congenital scoliosis. Unilateral unsegmented bars with contralateral hemivertebrae have the greatest tendency for rapid progression and should be surgically fused as soon as the bony abnormality is evident [9]. Unilateral unsegmented bars also tend to progress [10]. Hemivertebrae have a variable prognosis for progression, depending on whether there is a contralateral hemivertebra that results in overall balance of the spine, whether there are multiple hemivertebrae on one side of the spine, and how much growth potential is predicted for each end plate of the hemivertebra [11,12]. Hemivertebrae should be observed to delineate their growth potential and progression. Patients often present with limited cervical range of motion, head tilt, torticollis, and shoulder asymmetry. Neurologic deficits can develop with the advent of adjacent segment instability, degeneration, or spinal arthritis.

External bracing is ineffective in treating congenital cervical deformities because they are often quite rigid. Historically, posterior fusion in situ has been the treatment of choice for balanced and flexible deformities [10]. Surgical treatment for patients with severe fixed deformities often requires circumferential fusion with or without resection of the hemivertebra. In some cases of hemivertebra, hemiepiphysiodesis may be performed, arresting growth on the curve convexity but permitting continued growth on the curve concavity, with resultant gradual curve correction with age [13]. This procedure has had good results in selected patients but can be unpredictable with respect to the amount of actual correction that can be achieved. In cases in which a hemivertebra is accompanied by significant coronal decompensation and compensatory growth would not be adequate to result in spinal balance, hemivertebra excision and circumferential fusion offer a good option to restore normal head alignment [11]. Resection can be done by means of a posterior-anterior-posterior approach [12]. Although this procedure is technically more demanding and has greater potential risks, it allows for better overall curve correction and improvement of coronal balance.

Bony dysplasias

Common bony dysplasias that can result in severe and often rigid cervical deformity include osteogenesis imperfecta (OI) and fibrodysplasia ossificans progressive (FOP). OI and its variants are caused by mutations in the type I collagen gene [14,15]. This can result in abnormal collagen or abnormal production of collagen. Clinical characteristics of OI include osteopenia, bone fragility, blue sclera, dentiginous imperfecta, hearing loss, and short stature. Most of the vertebral deformities, including scoliosis and kyphosis, occur in the thoracic and lumbar spine. Cervical deformities attributable to OI are rare but do exist. These include kyphosis, vertebrobasilar invaginations, and cervical spondyloptosis [16]. The pathogenesis of these deformities involves repetitive microfractures at the vertebral end plates that lead to abnormal spinal growth. Bracing is ineffective in treating many of these deformities. Combined anterior and posterior surgery is often necessary to correct the deformity and to arrest the progression.

FOP is an autosomal dominant connective tissue disorder caused by an abnormality in the bone morphogenetic protein (BMP) signaling pathway [17]. Patients afflicted with this disorder exhibit congenital malformation of the big toes and progressive and disabling heterotopic ossification of muscle and soft connective tissue. Most patients exhibit signs of neck stiffness before the onset of radiographic ankylosis. Vertebral anomalies include narrow vertebral bodies, enlarged pedicles, and large spinous processes [18]. No effective medical treatment exists to prevent the onset or progression of the disease. Spinal fusion is necessary to prevent deformity.

Neurofibromatosis

Neurofibromatosis (NF) is a genetic disorder of the nervous system that affects the growth of neural tissues. The disorder can be inherited, but 30% to 50% of all new cases of NF arise from spontaneous mutation. Two types of NF exist: NF-1 and NF-2. NF-1 is the most common type and is caused by a mutation in the gene for the protein neurofibromin.

Cervical anomalies occur in up to 44% of patients with NF [19]. Although cervical kyphosis is the most common finding, vertebral scalloping, foraminal enlargement, vertebral body collapse, and atlantoaxial rotatory subluxation are also commonly seen in this disease process [20]. Most kyphotic deformities are asymptomatic. Symptoms can arise if sufficient deformity leads to spinal cord compression over the apex of the kyphosis, however. Surgical treatment of these deformities often requires anterior decompression with corpectomy, followed by a posterior spinal shortening procedure. Posterior fixation, if possible, helps to achieve better deformity correction and maintenance of correction. Spinous process wires, lateral mass plates, and cervical pedicle screws have all been used successfully [21].

Inflammatory arthritic deformity

Rheumatoid arthritis

Rheumatoid arthritis is the most common form of inflammatory arthritis. It affects 3% of women and 1% of men. The same inflammatory cells that destroy peripheral joints affect the synovium of apophyseal and uncovertebral joints, causing painful instability and neurologic compromise. Up to 71% of patients with rheumatoid arthritis show involvement of the cervical spine [22,23]. The most common patterns of involvement are C1-C2 instability, basilar invagination, and subaxial subluxation [23,24]. Sudden death associated with rheumatoid arthritis, most probably secondary to brain stem compression, has been reported [25]. From 7% to 34% of patients present with neurologic problems [23,26,27]. Documentation of neurologic function can be difficult, because loss of joint mobility leads to general muscle weakness. Although many patients complain of nonspecific neck pain, atlantoaxial subluxation is the most common cause for pain in the upper neck, occiput, and forehead in patients with rheumatoid arthritis [24,28]. Symptoms are typically aggravated by motion.

Increasing compression of the spinal cord results in severe myelopathy with gait abnormalities, weakness, paresthesias, and loss of dexterity. From 10% to 20% of patients with rheumatoid arthritis present with radiographic subaxial subluxation [24,29,30]. Erosion of the facet joints and narrowing of the discs lead to subtle anterior subluxations that are often found on several levels [31]. This results in a characteristic "step-ladder" deformity that occurs most commonly at the C2-C3 and C3-C4 levels. Most of these deformities are flexible [32], and are thus amenable to single-stage corrections [22,27,33]. Many patients also develop osteoporosis as a consequence of inactivity, poor nutrition, and chronic steroid use. As a consequence, an effective surgical strategy in these patients is posterior or combined anterior/posterior decompression, realignment, and fusion with instrumentation [29,30,34,35].

Ankylosing spondylitis

Ankylosing spondylitis is a chronic seronegative inflammatory disease that affects the axial skeleton, especially the sacroiliac joints, hip joints, and spine. Extraskeletal involvement is found in the aorta, lung, and uvea. The incidence of ankylosing spondylitis is 0.5 to 1 per 1000 people [36,37]. Although men are affected more frequently than women, mild courses of ankylosing spondylitis are more common in the latter. The disease usually has its onset during early adulthood. Juvenile ankylosing spondylitis affects adolescents (less than 16 years of age), however, and has a predisposition toward hip involvement [38]. The human leukocyte antigen (HLA)-B27 surface antigen is found in 88% to 96%% of patients [39], and investigators have postulated that a genetic component (ie, HLA-B27) and an environmental component (eg, previous *Klebsiella* or *Chlamydia* infection) are responsible for triggering the disease process [40]. The erythrocyte sedimentation rate (ESR) is elevated in up to 80% of cases but does not accurately reflect disease activity. The serum creatine kinase (CK) level, however, is a good indicator of the severity of the disease process.

The onset of the disease is often insidious, with early symptoms including pain in the buttocks, heels, and lower back. Patients complain typically of morning stiffness, the improvement of symptoms with activity during the day, and the return of symptoms in the evening [37,41]. The earliest

changes involve the sacroiliac joints and then extend upward into the spine. Spinal disease results in loss of motion and subsequent loss of lordosis in the cervical and lumbar spine. Synovitis in the early stages leads to progressive fibrosis and ankylosis of the joints during the reparative phase. Enthesitis occurs at the insertion of the annulus fibrosis on the vertebral body, with eventual calcification that results in the characteristic ''bamboo spine.'' The pain from the inflammatory process subsides after full ankylosis of the affected joints occurs [42]. Approximately 30% of patients develop uveitis, and 30% have chest tightness [43,44]. Limited chest expansion indicates thoracic involvement. Fewer than 5% of patients have involvement of the aorta [45], characterized by dilation and possible conduction defects. In addition, patients may develop renal amyloidosis and pulmonary fibrosis [41].

Cervicothoracic kyphosis was one of the first cervical deformities to be corrected surgically [46–48]. The cervicothoracic osteotomy for the correction of flexion deformity was first described by Urist in 1958 [49]. The cervical osteotomy is performed between C7 and T1. This approach avoids injury to the vertebral artery, which usually enters the spine by way of the transverse foramen of C6. Although the procedure was initially performed under local anesthesia [50,51], the evolution of somatosensory- and motor-evoked potential monitoring of the spinal cord has permitted use of a general anesthetic for the entire case. After removal of the posterior elements, a pedicle osteotomy is performed through the neuroforamen at C7-T1, with great emphasis to enlarge the osteotomy sufficiently to prevent inadvertent compression of the C8 nerve root at the time of closure of the osteotomy [52–54]. The kyphotic deformity is then corrected with gentle posterior translation of the head. The head is held in the corrected position using internal fixation with plate-screw constructs or plate-rod constructs with adjunctive halo-vest immobilization.

Degenerative cervical deformity: cervical spondylosis

Cervical spondylosis is the predominant cause of cervical myelopathy in the United States [55]. Disc degeneration begins with changes in the extracellular matrix and proteoglycan composition. There is an increase in the ratio of keratin sulfate to chondroitin sulfate. Cross-linking of collagen fibers in the annulus decreases, with a concomitant decrease in tensile strength [56]. The disc collapses, losing height, and it can bulge posteriorly, directly causing cervical canal narrowing. The narrowing of the disc space leads to overhang of the uncovertebral and facet joints, with resultant foraminal stenosis and loss of radicular blood flow. Disc degeneration also may cause macro- or microinstability that potentiates dynamic cord compression. The abnormal motion can result in several pathologic changes. Osteophytes form around uncovertebral joints and facet joints at the insertion of the annulus fibrosis, causing foraminal, lateral recess, and central spinal canal stenosis [56]. With loss of disc height, as the spinal column shortens, the ligamentum flavum subsequently thickens and buckles. This further decreases the space available for the spinal cord, and hyperextension of the cervical spine further reduces spinal canal diameter [57]. In more advanced cases, loss of cervical lordosis or even frank kyphosis can occur. As the degenerative cascade progresses, the affected motion segments become stiffer. This phenomenon may exacerbate hypermobility at motion segments adjacent to the diseased spinal level [58].

Spinal cord compression can occur from loss of normal sagittal alignment of the cervical spinal column. Thoracic hyperlordosis can cause compensatory cervical kyphosis and spinal cord impingement as the spinal cord is stretched and compressed over a kyphotic segment. Cervical stenosis can also occur when degenerative changes are superimposed on congenital anomalies of the cervical spine. These can include a congenitally narrow cervical spine (achondroplastic dwarfs), Klippel-Feil deformities, and craniocervical deformities. Patients with athetoid cerebral palsy often develop severe cervical spondylosis as a result of continuously unchecked neck motion.

Symptoms and signs

Cervical radiculopathy in spondylosis can be quite complex, with nerve root involvement seen at one or more levels and occurring unilaterally or bilaterally. The onset may be acute, subacute, or chronic, and impingement on the nerve roots may be from osteophytes or disc herniation. With radiculopathy, sensory involvement in the form of paresthesias or hyperesthesia is more common than motor or reflex changes. Several dermatomal levels may be involved, with radiation into the anterior chest and back. The chief complaint is radiation of pain into the interscapular area and

the arm. Typically, patients have proximal arm pain and distal paresthesias.

Cervical myelopathy has a variable clinical presentation, given the complex pathogenic mechanisms involved. These include static or dynamic canal impingement, facet arthropathy, vascular ischemia, and the presence of spondylotic transverse bars. In addition, given its neuronal topography, the cord may be affected in dramatically different ways by relatively minor differences in anatomic regions of compression. The clinical course of myelopathy is usually progressive, leading to complete disability over a period of months to years with stepwise deteriorations in function.

Patients often present with paresthesias; dyskinesias; or weakness of the hand, the entire upper extremity, or the lower extremity. Deep aching pain of the extremity, broad-based gait, loss of balance, loss of hand dexterity, and general muscle wasting are found in patients with advanced myelopathy. Impotence is not uncommon in these patents.

Hyperextension injuries of the spondylotic cervical spine can precipitate a central cord syndrome in which motor and sensory involvement is typically greater in the upper extremities than in the lower extremities. Recovery from this injury is usually incomplete. Complete quadriplegia can also occur if the preexisting stenosis is severe. In this setting, the 1-year mortality rate approaches 80% [59,60].

Deep tendon reflexes can be hyporeflexic or hyperreflexic, with the former seen in anterior horn cell (upper extremity) involvement and the latter seen in corticospinal tract (lower extremity) involvement. Hyporeflexia is found at the level of compression, whereas hyperreflexia occurs at the level below. Long-tract signs, such as the presence of Hoffmann's reflex or Babinski's reflex, indicate an upper motor neuron lesion. Clonus is often present, although asymmetric. Upper extremity involvement is often unilateral, whereas lower extremities are affected bilaterally. High cervical spondylosis (C3-C5) leads to complaints of numb and clumsy hands, whereas myelopathy of the lower cervical spine (C5-C8) presents with spasticity and loss of proprioception in the legs. Abdominal reflexes are usually intact, enabling the clinician to differentiate spondylosis from amyotrophic lateral sclerosis, in which reflexes are often absent. Multiple compressions of the spinal cord cause more severe deterioration functionally and electrophysiologically than single-level compression does.

Surgical treatment

Surgical intervention should be considered if the patient does not respond to a conservative treatment protocol or shows evidence of deteriorating myelopathy or radiculopathy. The spinal cord can be effectively decompressed by an anterior, posterior, or combined approach [61,62].

The anterior approach allows multilevel discectomy, vertebrectomy, foraminotomy, and fusion with tricortical iliac crest bone grafts or strut grafts [63–65]. Newer instrumentation techniques, such as cervical plates and expandable cervical cages [66], alleviate the need for halo immobilization. Supplemental posterior fixation and fusion should be added if more than three vertebral levels are decompressed anteriorly, however. Posterior fixation minimizes the risk of anterior dislodgement of the graft even in the presence of solid anterior fixation [67]. Anterior interbody fusion after decompression for a herniated cervical disc has a high success rate [63,64].

Cervical disc replacement prostheses have also been developed to provide a motion-sparing alternative to anterior cervical discectomy and fusion. By maintaining existing motion or restoring motion to a diseased motion segment, these prostheses have the potential to decrease the rate of symptomatic adjacent segment degeneration. Currently, US Food and Drug Administration (FDA)–approved clinical trials are underway to assess the efficacy of these devices against the outcomes that can be obtained with anterior cervical discectomy and fusion [68].

The number of involved levels may be important in deciding which of the surgical approaches to use. Patients with cervical myelopathy and involvement of more than three vertebral body levels may be best managed by a posterior approach. Multilevel laminectomy or laminoplasty has shown excellent results [67]. If laminectomy is performed, the facet joints and capsules should be preserved to minimize the chance of postlaminectomy deformity [69]. Late swan-neck deformities after laminectomy can be avoided with simultaneous posterior fusion using lateral mass plates. Laminoplasty is advantageous in that the cervical spinal cord can be decompressed without the high risk of developing late deformity. In addition, the morbidity associated with instrumentation and fusion can be avoided. Operative treatment in cases of cervical spondylotic radiculopathy and myelopathy must be individualized for every patient.

Prognosis

Cervical spondylosis is generally a progressive chronic disease process. In a study of 205 patients with neck pain, Gore and colleagues [70] found that many patients had decreased pain at the 10-year follow-up but that those with the most severe involvement did not improve. Conservative measures may retard the disease process in its early stages. If myelopathy or radiculopathy becomes clinically evident, surgical intervention is often necessary. For disease involving less than three vertebral levels, early anterior decompression and fusion have improved the clinical outcome, particularly in the elderly individual with cervical myelopathy.

Secondary (iatrogenic) cervical deformity

Secondary (postsurgical) cervical deformity is more common than primary and degenerative deformity. Although the development of better instrumentation to achieve and maintain spinal alignment has likely decreased the overall incidence of postsurgical deformity, the presence of spinal instrumentation often creates a significant surgical challenge when attempting to revise a rigidly fixed deformity. Some devices, such as the dynamic plating systems, occasionally compromise loss of spinal alignment (anterior settling) to allow for improved fusion rates.

Adjacent segment disc degeneration has been estimated to occur at a cumulative rate of 3% per year after fusion [71,72]. In the midcervical spine, it is controversial as to whether short-segment (<3 levels) fusion contributes significantly to the natural rate of disc degeneration that occurs at these levels. Adjacent segment degeneration at the cervicothoracic junction can lead to significant loss of cervical sagittal balance [73]. Symptoms often include C8 level radicular symptoms with or without myelopathy; loss or decrease in horizontal gaze; and occipital headaches, which often arise from O-C1 impingement as the patient tries to compensate for the cervicothoracic kyphosis at the O-C1 articulation.

Postlaminectomy deformity can occur after excessive facet resection or, more commonly, after long posterior cervical decompression. The incidence of postlaminectomy deformity is as high as 21% [69]. Although long laminectomies are still performed to treat multisegmental cervical stenosis, several key points must be considered to minimize the occurrence of this known complication. Facet resection should be avoided or minimized. Extensor muscle attachments to C2 and C7 should be spared if possible. Laminectomies without concurrent stabilization should be avoided in patients with anterior column compromise or in patients who have lost their cervical lordosis. In these cases, adjunctive instrumented fusion or laminoplasty (if lordosis is maintained) should be considered.

Anterior graft collapse can occur in the context of pseudarthrosis or can occur from anterior graft collapse. The average rate of anterior pseudarthrosis after anterior discectomy and fusion ranges between 5% and 30% depending on the number of levels fused [74–77], the use of spinal instrumentation, and the use of osteoinductive agents (eg, BMP). When detected early and in the presence of rigid fixation, spinal alignment is often unaffected by the presence of pseudarthrosis. When detected late, however, spinal collapse and hardware migration or failure are often seen. Anterior graft collapse can also occur in the presence of longitudinally dynamizing plates, especially the slotted-type devices. In these cases, the spine may fuse but does so in a suboptimal alignment. More rarely, in the presence of poor bone stock, further settling of the spine after instrumentation can lead to screw failure in the intermediate fixation segments within the fusion.

Salvage of postsurgical cervical deformities requires consideration of many factors. These include technical factors, such as assessment of the areas of neural compression, magnitude and location of the deformity(ies), flexibility of the deformity, presence of spinal hardware, areas of pseudarthrosis, and areas of previous surgery (anterior, posterior, or both). Most cervical deformities can be successfully managed through a single-stage or two-stage approach [78,79]. In the absence of a spontaneous posterior fusion, kyphosis from anterior graft collapse can be successfully managed with revision anterior decompression, osteotomies, and reconstruction (Fig. 1). The surgeon should be aware of patients with developmental spinal stenosis when lordosis restoration is planned in the absence of multilevel corpectomy or preexisting posterior decompression. In these special cases, restoration of lordosis can exacerbate the developmental stenosis and lead to postoperative worsening of the myelopathy. Interbody segmental reduction and careful plate contouring are powerful tools to restore cervical lordosis and to translate the cervical spine anteriorly. Posterior fusion can be done in a staged

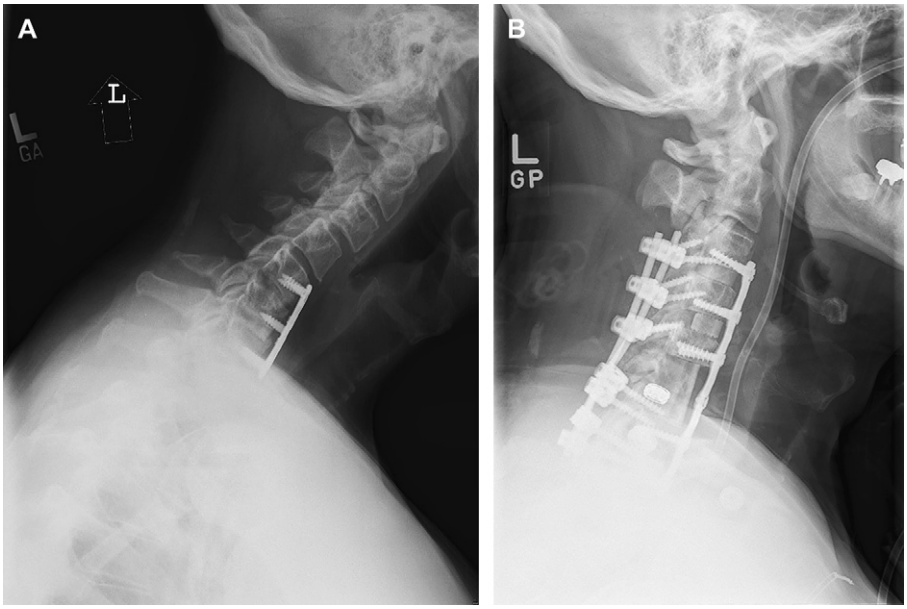

Fig. 1. (*A*) Lateral radiograph of a 63-year-old man with a prior two-level correction to anterior cervical discectomy and fusion with progressive kyphotic deformity. (*B*) Lateral postoperative radiograph after a two-staged procedure, including C6 corpectomy, C3-T1 ACDF, and C3-T1 posterior spinal fusion, demonstrates near correction of prior deformity.

fashion in patients who have a high risk for pseudarthrosis. If the anterior construct is mechanically secure (as in the case of intercalary segment fixation), adjunctive posterior fusion can be avoided. Often, patients can be followed over time, and, typically, only one motion segment, if any, exhibits a delayed union. This problem can be successfully managed with a short-segment posterior augmentation and internal fixation. This treatment scheme avoids the morbidity that is associated with long posterior cervical exposures.

Salvage surgery

Fixed cervical deformities that involve anterior and posterior fusions often require a three-stage (posterior-anterior-posterior or 540°) approach; however, published literature on this subject is lacking. Posterior removal of spinal hardware, followed by multilevel posterior osteotomies and insertion of screws (without the rod), is performed initially. The patient is then placed in a cervical collar between stages. The second stage involves anterior removal of hardware, discectomies, anterior osteotomies at the apex of the deformity, and restoration of the cervical lordosis using intercalary segment reduction and anterior plating. Patients who have significant translational deformity of the cervical spine often require extension of the fusion across the cervicothoracic junction to correct this problem. The third stage involves reexploration of the posterior wound and augmenting the stability of the anterior correction using posterior instrumentation. This approach results in a gradual restoration of lordosis and good correction of anterior translation (Fig. 2). In the authors' experience at the University of California, San Francisco, they have demonstrated an average of 28° of angular correction and 31% translational correction as measured from C2 to C7 using the approach described previously (B. Tay, MD, unpublished data).

Discussion

Complex cervical deformity can arise from multiple causes. Unfortunately, the most common type of cervical deformity remains that occurring after surgery. Treatment of these complex problems is challenging and requires a clear understanding of the deformity and of the patient. The treating surgeon must be comfortable with remobilizing the spinal column anteriorly and posteriorly and with methods of anterior and posterior correction of deformity using instrumentation. Gentle multisegmental restoration of sagittal

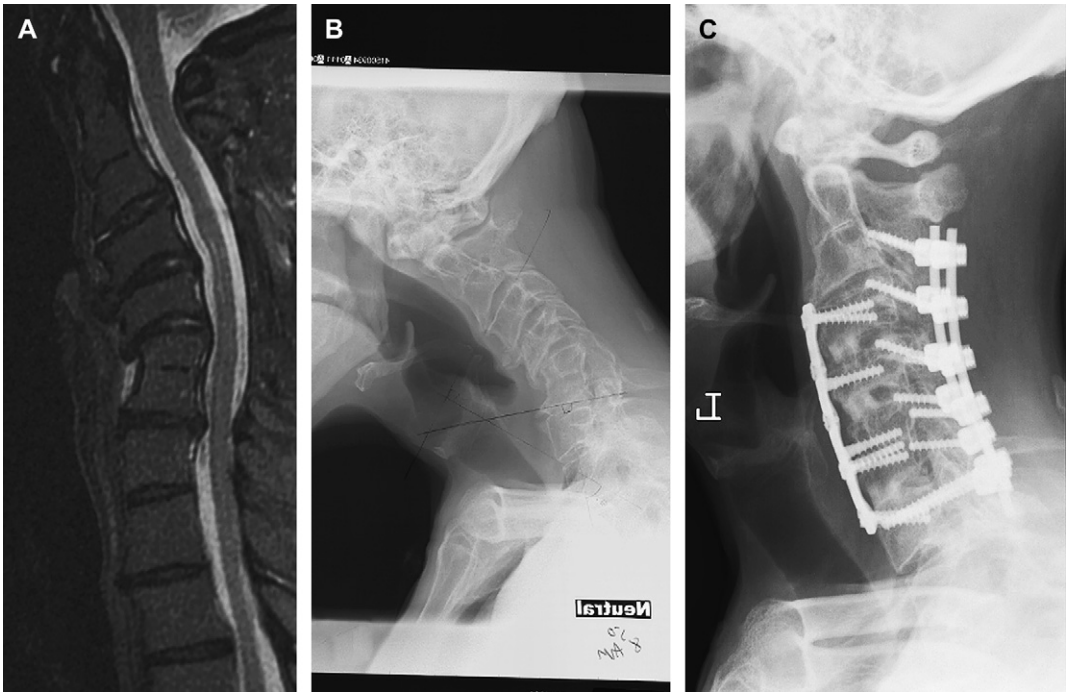

Fig. 2. Sagittal MRI (*A*) and lateral radiograph (*B*) of a 60-year-old man with a prior laminectomy and Klippel-Feil malformation at C2-C3 shows progressive fixed kyphotic deformity. (*C*) Lateral postoperative radiograph after a three-staged procedure, including posterior release, C3-T1 posterior spinal fusion, and C3-T1 correction to anterior cervical discectomy and fusion, shows marked reduction of fixed deformity.

balance is favored over acute angular correction from C6 and higher because of the potential for injury to the vertebral artery.

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