The treatment of patients with cervical spine deformities can be challenging for the spine surgeon. Often, these patients seek treatment when their deformity substantially affects their quality of life, such as a loss of ability to gaze forward or a severe neurologic deficit. If the patient seeks treatment at an advanced age, the risks of surgery versus limited, nonsurgical treatment options must be considered.

A variety of etiologies can lead to cervical spine deformity. The treating surgeon must consider both the underlying pathology and the desired outcome when planning the surgical intervention. In this chapter, the relevant anatomy, various etiologies, clinical presentation, evaluation, and the treatment of cervical spine deformity are discussed.

**Relevant Anatomy**

The cervical spine consists of the first seven vertebrae of the spine. The atlantoaxial complex (C1 and C2) has a unique articulation and serves to link the cervical spine to the occiput, providing approximately 50% of the rotary motion of the cervical spine. The atlantoaxial complex relies on the transverse ligament for stability. This strong ligament attaches through the lateral tubercles of C1. Additional stabilizers include the alar and apical ligaments.1 The lower cervical vertebrae (C3-C7) are connected by the intervertebral disks, the anterior and posterior longitudinal ligaments, the ligamentum

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**Abstract**

Cervical spine deformities pose substantial challenges for spine surgeons. The anatomy and biomechanics of the cervical spine play an important role in the decision-making process regarding treatment. The etiology of cervical deformities can be congenital, developmental, iatrogenic, degenerative, or inflammatory. Dropped head syndrome has been recently described but is poorly understood. Patients have variable presentations ranging from neck pain to an inability to maintain head position and neural compromise. Radiographic angles are important to monitor the deformity and plan the surgical correction. Treatment is focused on relieving pain, preventing and improving neurologic compromise, and improving overall spinal alignment and balance. The surgical approach and the level of fusion should be individualized on a case-by-case basis. The surgeon can greatly improve a patient’s quality of life by understanding the nature of the patient’s deformity and fully considering all treatment options.

flavum, the interspinous ligaments, and the posterior musculature. Bony stability is provided through the anterior uncinate joints and the posterior facet joints. Each of these anatomic structures makes a unique contribution to maintaining normal cervical posture. Of special importance are the semispinalis cervicis and capitis muscles, which have been shown to have a substantial pull that is required to hold the cervical spine in a neutral position.

The cervical spine averages between 14° and 15° of lordosis, with the normal weight-bearing axis lying posterior to the vertebral bodies. Biomechanical studies have demonstrated that as much as 84% of the cervical spine load is transmitted through the posterior elements. When the posterior arch-facet complex is disrupted, the normal cervical spine may have a loss of stability and a change in its load distribution, which increases the load on the anterior elements up to tenfold. (Figure 1).

**Etiology**

Many etiologies have been associated with the development of cervical spine deformities. One of the most common causes for the development of an iatrogenic deformity is postlaminectomy kyphosis. To perform a cervical laminectomy, soft-tissue structures, including the supraspinous ligament and the ligamentum flavum, are removed. These soft-tissue structures form a tension-band complex that acts to prevent excessive flexion of the cervical spine. Removal of these soft-tissue structures results in a loss of stability, with a resultant transfer of load to the anterior aspect of the cervical spine. There is a progressive loss of sagittal balance with progressive kyphosis. This puts the posterior musculature at a mechanical disadvantage, leading to pain, fatigue and, ultimately, progression of the deformity and the possibility of a recurrent neurologic deficit.

Cervical deformity also can result from degenerative disk disease. The intervertebral disks make up approximately 15% of cervical height. A loss of disk height results in an anterior shift of the weight-bearing axis of the cervical spine. The anterior translation of the center of gravity puts additional stress on the posterior elements, which can lead to balance failure and progression of the kyphotic deformity. This leads to an insidious progression of deformity and potential neural compromise. Recently, there has been increasing recognition of the role of the proximal thoracic spine as a major contributor.
to cervical alignment. Degeneration of the intervertebral disks of the proximal thoracic spine can result in proximal thoracic kyphosis, with the resultant inability of the patient to raise his or her head.

Another type of cervical deformity is the flexible deformity referred to as dropped head syndrome (Figure 2). Also known as camptoccephalia, dropped head syndrome is characterized by a gradual increase in cervical kyphotic deformity that is correctable with passive neck extension. The deformity in dropped head syndrome is not structural but related to the forces of the cervical paraspinal muscles.

Various etiologies have been suggested, including excessive anterior muscle tension or weak cervical spine extensors. These various abnormal muscle forces may be related to systemic disease (such as systemic muscle atrophy) or may be idiopathic. This condition is commonly referred to as isolated neck extensor myopathy, a condition unrelated to systemic myelopathy. Although the exact etiology is unclear and probably multifactorial, the common theory is that abnormal muscle function leads to progressive kyphosis. Coupled with age-related tissue degeneration, it can progress to a dropped head deformity. In addition, it has been suggested that the increased kyphosis within the thoracic vertebrae can lead to an anterior shift in the center of gravity in the cervical spine. By shifting the weight anteriorly, more force is required from the neck extensors, which can become fatigued and ultimately unable to support normal posture.

Evidence also exists that links the development of flexible dropped head syndrome to myositis. Studies in the literature report signs of inflammation within biopsy samples of the extensor musculature in patients with idiopathic dropped head deformity. Other authors, however, have found noninflammatory myopathic changes and argue against an inflammatory process.

Another class of cervical deformities is related to inflammatory joint disorders, such as rheumatoid arthritis (RA) and ankylosing spondylitis. Although the advent and the increased use of antirheumatoid medications has improved the overall management of symptoms in RA, cervical spine manifestations still affect most patients with RA. Atlantoaxial instability, basilar invagination, and subaxial subluxation have been known to develop in these patients. Patients with RA and ankylosing spondylitis tend to flex forward early in the disease progression to unload the affected facet joints. In the long term, a worsening of arthritis and marginal osteophyte formation can lead to increasing rigidity of the chin-on-chest deformity.

Additional causes for cervical spine deformities include a variety of congenital deformities and their associated conditions. Klippel-Feil syndrome, Down syndrome, Morquio syndrome, and various other syndromes have been associated with spinal deformity. A variety of bony dysplasias also can lead to the development of cervical deformity. Cervical deformities can develop in patients with osteogenesis imperfecta and other type I collagen disorders, although thoracolumbar deformities are more common. Congenital scoliosis also may affect the cervical spine, including hemivertebrae or absent pedicles. Neurofibromatosis can lead to the development of cervical deformity, with as many as 44% of patients with neurofibromatosis having cervical spine anomalies. In addition, cervical deformity has developed secondary to posttraumatic, postinfectious, or oncologic etiologies.

**Presentation**

Patients who have a cervical spine deformity may have a wide range of symptoms. The loss of a forward gaze is typically the chief complaint of a patient with a cervical deformity. Patients may also report difficulty with posture, cosmesis, and neurologic symptoms. Evaluation of the deformity must include both sagittal and coronal alignment. Sagittal imbalance (chin on the chest) is far more common than coronal imbalance (ear on the shoulder); the treatment of cervical sagittal balance has been shown to have a far greater effect on patient outcomes.

With cervical kyphosis, the weight of the head falls anterior to a patient’s center of gravity. This leads to increased muscle tension within the neck and shoulders and may lead to substantial...
Spine

Neck and shoulder pain as a primary symptom. In addition, patients with cervical spine deformities may exhibit signs of low back pain because they will compensate for their postural difficulties with lumbar hyperlordosis (Figure 2). Patients may report difficulty with swallowing because the position of their head makes this action challenging. Individuals who have a cervical deformity caused by proximal thoracic kyphosis are less likely to have difficulty swallowing compared with individuals with pure cervical kyphosis (Figure 3). Patients with cervical deformities may also have varying degrees of neural compromise. A progressive kyphotic deformity can lead to draping of the spinal cord over the vertebral bodies. This can lead to myelopathy caused by an increase in longitudinal tension, because the spinal cord remains tethered by the denticulate ligament and the cervical nerve roots. It has been shown in an animal study that demyelination of the spinal cord occurs with cervical kyphosis and the resultant cord compression, increasing symptoms of myelopathy. A complete spine-related history and physical examination are essential when planning the treatment of cervical spine deformities.

Patient Examination

The physical examination begins with an inspection of the patient’s stance. Patients with severe kyphotic deformity will have the characteristic chin-on-chest posture and difficulty in maintaining a forward gaze. Some patients may require the use of an assistive device to maintain their forward gaze (Figure 4). Often, this is seen in patients with a proximal thoracic kyphosis in whom a compensatory curve has developed. A complete neurologic examination should be conducted and clearly documented, including the flexibility or fixed nature of the deformity. Flexible deformities, such as dropped head syndrome, will be passively correctable when the patient is supine. To assess the degree of flexibility, the surgeon can place pillows underneath the patient’s head to maintain the kyphotic posture. As the patient relaxes, the pillows are

Figure 3  Illustration showing the functional classification of patients with cervical deformity and the differing treatment of patients with proximal thoracic kyphosis and cervical kyphosis. PSF = posterior spinal fusion (C2-T3-T5), PCF = posterior cervical fusion (C2-T2), ACDF = anterior cervical diskectomy and fusion.
subsequently removed to allow for passive correction of the neck and an assessment of flexibility. It should be noted that this maneuver should not be attempted in patients with a rigid deformity or in patients with ankylosing spondylitis with a suspected fracture.

Radiographic assessment should begin with a 36-inch cassette standing lateral radiograph that captures the entire spine, from the auditory canal to the level of the femoral heads. From this image, several radiographic criteria can be assessed. The lateral Cobb angle measures lordosis between C2 and C7. Of note, although correction of the Cobb angle (cervical kyphosis) is the standard of surgical treatment, there is no evidence that restoring a normal Cobb angle improves outcomes. The sagittal vertical axis (SVA), or C2 SVA, is measured by drawing a C2 plumb line and measuring the horizontal distance between this line and the posterior-superior corner of the sacrum, which shows overall sagittal plane translation. This line also can be defined regionally by measuring the distance between a plumb line dropped from the center of C2 (or odontoid) and the posterior superior aspect of C7 (C2-C7 SVA). There have been reports in the literature that a larger C2 SVA correlates with a poorer health-related quality of life (specifically, the Neck Disability Index and the Medical Outcomes Study 36-Item Short Form Physical Component Summary). The chin-brow vertical angle is measured as the angle subtended between the vertical axis and a line connecting the patient’s chin and brow. The chin-brow vertical angle provides a radiographic assessment of horizontal gaze and can be helpful in determining the size of any posterior osteotomy (Figure 5).
In flexible cervical spine deformities, such as dropped head syndrome, a thorough workup for secondary causes must be performed before definitive treatment can be initiated. The erythrocyte sedimentation rate, C-reactive protein level, creatine kinase level, thyroid function tests, and antiacetylcholine receptor antibody tests are part of the standard workup. Electromyography can help differentiate myopathic causes from other etiologies, and muscle biopsy can be performed to look for inflammation that would indicate neuromuscular diseases. Flexion and extension radiographs can be used to assess the degree and the flexibility of the deformity, and advanced imaging studies (MRI and CT) can show cord compression and detect autofused segments and arthritic joints.

**Treatment**

The treatment of cervical spine deformity is focused on relieving pain, preventing or improving neurologic compromise, and improving overall spinal alignment and balance. The treatment decisions will vary depending on the degree of the deformity, the etiology, and the patient’s specific treatment goals.

**Flexible Deformity**

**Nonsurgical Treatment**

The primary treatment goals for patients with cervical spine deformity are to restore sagittal alignment, improve horizontal gaze, decrease neck pain, and improve swallowing and respiration. Prior to surgical management, medical conditions identified during the initial workup must be addressed. Steroid treatment may sometimes be beneficial for inflammatory conditions leading to weakness of the cervical extensors, whereas other authors support early surgical intervention. Alternatively, some authors suggest reserving surgical treatment for patients with spinal cord compromise. There seems to be agreement that surgical intervention is warranted if nonsurgical management has failed and the patient is unable to maintain a forward gaze and has difficulty eating. It is critical for the surgeon to have a patient-centered, informed discussion of the risks and benefits of surgery, with the understanding that the goal of surgery is the optimization of quality of life for the patient.

In patients with a flexible deformity, a posterior approach is generally preferred. A posterior approach allows the surgeon to address more vertebral levels than an anterior approach and carries a lower risk for dysphagia, dystonia, and pseudarthrosis. Additional anterior surgery may be required if there is a degree of ankylosis anteriorly. An anterior approach is required if the deformity cannot be corrected from a posterior approach or if the deformity is secondary to radiation therapy, which has caused contractures and sclerosis of the anterior musculature. Anterior surgery also may allow for the release of contractures in the setting of neuromuscular kyphosis and may increase arthrodesis rates in poor healing environments.

Long fusions are typically used to correct cervical deformities that result from the tendency of the deformity to recur with more limited surgical procedures. The deformity of dropped head syndrome typically begins at the C4 level; however, the fusion construct should begin at C2 to provide increased stability. Fusing from C2 will be more effective in correcting the deformity.
caused by semispinalis cervicis laxity because this muscle has its main attachment at the C2 level. Extending a fusion to the thoracic spine (T3-T5) may be required if there is additional thoracic deformity (proximal thoracic kyphosis) or if substantial deformity correction has been performed to allow a solid base for the construct. Case series have shown improved outcomes with screw-rod constructs, but they have not shown decreased failure rates with the distal extension of fusion constructs. Postoperatively, hard collar immobilization is recommended for 3 months when modern screw-rod constructs are used. Limited outcome studies exist. This is primarily because there is no consensus on the optimal outcome for these patients. In one case series, seven of nine patients treated with surgery reported good to excellent outcomes based on pain relief, gait improvements, and restoration of horizontal gaze. Short-term complications are common in these procedures but do not appear to affect long-term outcomes.

Iatrogenic Deformity
As mentioned previously, iatrogenic cervical kyphosis is the most common form of cervical spine deformity, especially after multilevel cervical laminectomy. Historically, anterior cervical diskectomy, without fusion or anterior plating, was a major cause of iatrogenic cervical kyphosis. A preexisting lack of lordosis, younger age at surgery, laminectomy of four or more levels, surgery involving the C2 lamina, and increased preoperative range of motion are risk factors for the development of iatrogenic kyphosis. Laminoplasty also has been associated with the development of kyphosis, although to a lesser extent than laminectomy. Depending on the magnitude and the type of deformity, correction can be addressed by an anterior approach, a posterior approach, or a combined approach. An anterior approach offers lower rates of overall morbidity, direct decompression of the spinal cord, preservation of the posterior musculature, and generally more potential for lordosis creation. The disadvantages include less secure fixation, dysphagia, dysphonia, graft dislodgement, and the risk of esophageal injury. In addition, the fusion construct from an anterior approach has less mechanical advantage compared with posterior fusion and instrumentation because it sits anterior to the axis of rotation. When longer fusion constructs are required, anterior fusion carries an increasing rate of pseudarthrosis as the number of fusion levels increase. Posterior fusion with instrumentation provides superior fixation because the construct is located posterior to the axis of rotation and offers a mechanical advantage over anterior surgery. Stability, however, must be balanced against the disadvantages of this approach, including increased pain; a higher rate of wound infection; and in cases where the deformity is the result of a previous laminectomy, a lack of posterior coverage over the dura. Despite these potential concerns, substantial improvements in sagittal alignment have been seen with posterior-only constructs.

Abumi et al reported an improvement in kyphotic angle from greater than 28° to 5.1° with posterior surgery alone. A combination of anterior release and posterior fusion may be required for patients with substantial deformity. In addition, in patients with a fixed postlaminectomy deformity with ankylosis, a back-front-back approach may be required, whereby posterior osteotomies are performed before anterior releases followed by posterior instrumentation and fusion.

Cervical Kyphosis and Degenerative Disk Disease
Cervical kyphosis related to degenerative disk disease is another well-established disorder. The treatment of degenerative cervical disk disease is generally nonsurgical, with pain management and physical therapy. However, surgical intervention must be considered for patients who have neurologic compromise related to their degenerative deformity.

Patients with focal kyphosis or less severe deformity can be treated with anterior cervical diskectomy and fusion. Patients with severe deformity requiring more than three levels of correction may require a posterior fusion or a combination of an anterior and posterior fusion construct. When planning posterior fusion levels, the most common construct extends from C2 to T2 because the rates of adjacent level degeneration are higher in constructs ending at the cervicothoracic junction. The placement of C7 lateral mass screws can be technically difficult because of the small size of the lateral mass at that level, although many surgeons will use a pedicle screw at C7 as an alternative foundational support. Ending the construct at the cervicothoracic junction (C7) is typically not recommended because this can lead to mechanical failure resulting from the tremendous kyphotic stress typically seen at this location.

Fixed Deformities
Cervical spine deformity that is related to inflammatory arthropathies, such as RA and ankylosing spondylitis, require
Separate consideration because they produce unique deformities and surgical challenges. Cervical deformity is exceedingly common in RA, with as many as 86% of patients with RA exhibiting cervical spine disease. RA can result in a variety of typical anatomic deformities, with atlantoaxial subluxation being the most common deformity in this group. This subset of patients should be managed with atlantoaxial fusion.

For this procedure, Magerl transarticular screws have demonstrated good outcomes (97% fusion rates). Increasingly, surgeons are becoming familiar with the techniques of placing C1 and C2 screws.

Less common cervical deformities in patients with RA include atlantoaxial impaction and subaxial subluxation. Atlantoaxial impaction can be managed with halo traction if this treatment can be tolerated by the patient, or posterior decompression and occipitocervical fusion, if halo traction is not an option or is unsuccessful. Subaxial subluxation can be managed with posterior fusion, and complex deformities should be managed with extensive fusion of the cervical spine extending to the upper thoracic vertebrae. As a general rule, more extensive fusions should be considered in patients with RA because they are at higher risk for hardware failure because of their osteoporotic bone. Other complications include adjacent level disease and subaxial subluxation after occipitocervical fusion.

Patients with ankylosing spondylitis may have varying degrees of autofusion associated with the deformity. The levels of ankylosis must be considered when planning the surgical intervention. A completely ankylosed cervical spine is ideally corrected to 10° to 20° of flexion to allow the patient to see the front of his or her body, as well as his or her feet. Too straight a gaze makes it difficult for patients to walk down stairs or see objects near their feet. If there is retained motion, the chin-brow vertical angle should be normalized as much as possible. If there is a thoracolumbar deformity, correction of the deformity often restores horizontal gaze. Treating the cervical spine deformity first can lead to overcorrection. A pedicle subtraction osteotomy may be performed at the C7 level, with the fusion carried up to the C2 level or the occiput, depending on whether the patient has mobility in the occipitocervical and atlantoaxial segments. In cases of a rigid cervical kyphosis with neurologic compromise, when the spinal cord is tethered over the kyphotic segment, a circumferential cervical osteotomy should be performed. The procedure begins with a posterior laminectomy, a facetectomy, and screw placement. The surgeon must then perform an anterior decompression and release, followed by posterior fixation and fusion.

Other Causes

Additional causes of cervical deformity include postoperative infection, posttraumatic causes, tumors, and congenital deformity. Each patient requires individual consideration, depending on the presenting symptoms and the specifics of the deformity. The requirement for specific osteotomies, resections, and levels of fusion must be considered carefully and generally requires advanced imaging for preoperative planning. These patients are often medically complex, and complications are common. Detailed conversations with patients and their families can often help manage expectations. The treatment should be tailored to the functional needs of the patient.

Summary

Deformities of the cervical spine are complex and can result in difficulty with ambulation, a loss of horizontal gaze, and difficulty with swallowing and respiration. Such difficulties can have a substantial effect on a patient’s quality of life. There are many etiologies for cervical spine deformity, each having its own outcome and treatment challenges. Regardless of the underlying cause, the treating physician’s goal must be to restore balance and function and prevent associated neurologic deficits. By understanding the nature of a patient’s deformity and fully considering all treatment options, the physician may greatly improve a patient’s quality of life.

References

Advances in the Understanding of Cervical Spine Deformity

Chapter 36


