146 Craniovertebral Junction Deformities

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SUMMARY OF KEY POINTS

- Condyle fractures can be a form of atlanto-occipital dislocation and may need to be managed surgically when there is alar ligament disruption or neural compression.
- The most sensitive and specific method of diagnosing atlanto-occipital dislocation is with the condyle-C1 interval.
- Type 1 and type 3 odontoid fractures can often be managed with external immobilization, though type 2 fractures show an increased rate of nonunion in certain patients.
- When incidentally found, os odontoideum should be closely followed with surgery offered for patients with neurologic compromise or progressive deformity.
- Rheumatoid arthritis involving the cervical spine can cause transverse subluxation and basilar impression, which should be screened for.
- Craniovertebral junction abnormalities are commonly associated with congenital disorders; in particular Down syndrome and Morquio syndrome.
- As endoscopic techniques continue to develop, the need for large open surgical procedures may decrease.
- Atlantoaxial rotatory fixation does not often require surgery if reduction can be achieved and the infection can be treated.
- The need for fusion of the craniovertebral junction in tumor resection cases is variable and can be guided by the location of resected bony elements in terms of zones.
- Atlantoaxial instability in congenital syndromes has many causes but is ultimately the result of incomplete or absent fixation of the anterior arch of C1 to the dens.

CRANIOVERTEBRAL JUNCTION DEFORMITIES

The craniovertebral junction (CVJ) is subject to deformities caused by trauma, congenital disorders, degenerative disease, infection, and tumors. The goals of management of pathology of the CVJ are to identify instability, decompress neural elements, and provide structural support for the head. Instability can be identified using a number of craniometric and morphometric indices. Many of these criteria were developed in the pre–computed tomography (CT) and magnetic resonance imaging (MRI) era, and therefore a description of new indices using "newer" technologies will be presented along with historical ones. The criteria of instability requiring stabilization differ depending on the underlying pathology. For instance, instability caused by acute trauma has tightly defined criteria for instability as opposed to the chronic instability caused by degenerative disease such as rheumatoid arthritis. The plethora of grading systems causes some confusion regarding management decisions. Attempts have been made to create treatment algorithms for pathology of this complex region; however, high-quality medical evidence relating to many important questions is not available. Therefore, treatment decisions are made with a reliance on a thorough knowledge of the biomechanics, anatomy, and physiology of the CVJ. New technology continues to drive improvement of diagnosis, management, and outcomes of CVJ disease. This chapter reviews the diagnosis and management of this complex region.

CLINICAL AND RADIOGRAPHIC ANATOMY OF THE CRANIOVERTEBRAL JUNCTION

The CVJ consists of the occiput, atlas, axis, and associated ligaments. The CVJ is a compromise between strength and flexibility. The bones and ligaments provide structural support for the head and protect the brain stem and upper cervical spinal cord. Concurrently, these structures allow for significant movement in flexion-extension, lateral bending, and rotational planes. The following discussion is limited to an overview of the anatomic landmarks and indices used to define abnormal relationships, because discussions of the embryology and anatomy of the CVJ are presented elsewhere in this text.

The normal relationships among the occiput, atlas, and axis have been studied and are well described. The use of CT with sagittal reconstruction and multiaxial MRI has greatly enhanced the understanding and definition of abnormal anatomic relationships at the CVJ. Various measurements have been described to delineate normal from abnormal pathologies. In other words, there are different indices for trauma, degenerative disease, cranial settling, and basilar impression.

TRAUMA

Injury to the CVJ can manifest as ligamentous injury or fracture of the occiput, atlas, or axis. These injuries are as follows: occipital condyle fractures, atlanto-occipital dislocation (AOD), atlas fractures or C1 burst fractures, and C2 fractures of the odontoid or pars interarticularis. Radiographic criteria have been established to help assess clinical stability. Although many of these criteria are used traditionally, they are by no means standardized criteria for each type of injury.[1] Determining the instability of these fractures is of primary importance in determining management.

C0 (Occipital Condyle) Fractures

Traditionally, occipital condyle fractures are categorized by the system developed by Anderson and Montesano.[2] This system grades occipital condyle fractures according to occipital fracture (type 1), large condyle fracture (type 2), and avulsion condyle fracture (type 3). The inference from this study is that small condyle fractures represent disruption of the alar ligament. The disruption of the alar ligament has been demonstrated to increase the mobility of the C0-1 joint.[3]

Tuli and colleagues defined fractures according to evidence of ligamentous instability.[4] Type 1 represents large bony fractures, condensing Anderson and Montesano types 1 and 2 into one group. Type 2 is both small bony fractures of the condyle. The fractures are further subdivided into 2a (stable) and 2b (unstable). Instability is characterized by MRI evidence of alar ligament disruption or CT/radiographic criteria. However, use of MRI to assess disruption of the alar ligament remains controversial.[5]

To simplify the issue, Maserati and coworkers focused on the C0-1 joint.[6] Determination of instability is made using the elongation of the distance of the C0-1 joint described by Pang.[7] This method is also used to determine AOD and will be more completely described in the next section. Unstable condyle fractures are a form of AOD and need to be treated as such.[6]

However, once instability has been determined, treatment is also not standardized. Fractures without apparent ligamentous disruption can be treated conservatively with a cervical collar or halo vest. Immobilization may be performed if the fracture fragment is large enough and aligned enough to allow bony fusion[1] If the bony fragment appears small or there is an apparent alar ligament disruption, it may be necessary to perform an occipital cervical fusion because purely ligamentous injury is unlikely to heal by immobilization.[6] Further indications for surgical management may include neural compression from displaced fracture fragments or associated occipital-atlantal or atlanto-axial injuries.[8] In pediatric patients, unilateral alar ligament disruption can potentially be managed with external immobilization alone.[9]

C0-1 Fractures or Atlanto-Occipital Dislocation

In the diagnosis of AOD, vigilant clinical suspicion is most important. The deformity may reduce spontaneously because of recoil of the elastic ligamentous structures. Suspicion should be raised based on the mechanism of injury (e.g., high-velocity crash) or findings on neurologic examination (severe neurologic injury, brain stem or C1-2 level deficits), lateral cervical spine radiograph (obvious separation of the condyle-C1 joint or C1-2 prevertebral swelling), or head CT (subarachnoid hemorrhage around the brain stem or upper cervical spinal cord, or epidural/subdural blood at C1-2).[7,10,11]

AOD is determined by measurements made from normal plain radiographs. These techniques are the Powers ratio,[12] basion-axial interval (Harris),[13] Wholey dens-basion interval, Sun measurement,[14] and X line.[15] These measurements essentially infer dislocation based on measurement of structures remote from the occipital condyle–atlas joint,[12,14,15] which can lead to false-negative examinations and lack of interobserver reliability. It has been found that the diagnostic sensitivities for the common tests range from 25% to 50%, with false-negative rates of 50% to 75%. However, the diagnostic sensitivity of the nonstandard indicators (perimedullary blood, tectorial membrane damage, C1-2 extra-axial blood) is 63% to 75%.[7]

An increase in the measurement of the joint distance between the occipital condyle and C1 can be used to determine AOD. This is called the condylar distance. Thin-slice axial CT scanning allowed Pang and associates to calculate that the distance should be less than 4 mm in pediatric patients. This test has been shown in the pediatric population to have a diagnostic sensitivity of 100%.[7] Dziurzynski and colleagues showed that in adult patients a condylar distance greater than 2 mm was diagnostic of AOD. This has a sensitivity of 92% and a specificity of 95%.[16] Gire and coworkers described a revised CCI, which decreases the required measurements for diagnosis from 16 to 1. This description may find use as a rapid method of evaluation with further validation.[17]

If the patient survives the initial injury, he or she should be immediately immobilized. The use of a halo vest to immobilize the patient has been shown to be a safe and effective treatment method to prevent delayed neurologic deterioration while the patient is stabilized and prepared for definitive treatment.[18] Depending on the severity of injury, operative fixation can be performed on an elective basis.[18,19] The instability of AOD is primarily a ligamentous injury, and therefore internal fixation and fusion is recommended for definitive treatment. If reduction of the AOD is necessary, it should be done with gentle manual manipulation under fluoroscopic guidance. If the patient has a neurologic examination to follow, the reduction can be performed with the patient under mild sedation. In the anesthetized patient, somatosensory evoked responses may provide some help in determining if reduction is affecting the patient neurologically.

C1 Fractures

Fractures of the atlas (C1) can manifest in multiple ways: isolated ventral or dorsal arch, burst, and lateral mass fractures. Isolated arch fractures are a controversial diagnosis because it is unlikely that a ring can have a fracture in one place without fracturing in another, although such occurrences have been described.[20] An axially directed force that translates into C1 through the wedge-shaped occipital condyles causes burst fractures of the atlas. These fractures were first described by Geoffrey Jefferson in 1920.[21] These fractures are detected with an open-mouth odontoid radiograph demonstrating spread of the lateral masses of C1 beyond the lateral borders of the C2 lateral masses. Assessment of the integrity of the transverse ligament is critical for determining the treatment of C1 burst fractures. Initial assessment of the competence of the ligament was made by a cadaveric study performed by Spence and colleagues[22] in 1970. The researchers showed that the transverse ligament typically failed if the spread between lateral masses was 6.9 mm or more. When corrected for the magnification of the radiographs, this distance should be increased to 8.1 mm.[23] This allows for indirect determination of rupture of the ligament based on plain radiographs. Again, the advent and widespread use of CT and MRI have allowed for direct visualization of ligament integrity. Dickman and associates used MRI to evaluate the transverse ligament and found an abnormal atlantodental interval of 3 mm or more implies the incompetence of the transverse ligament.[24] A ruptured transverse ligament was found in cadaver studies to produce hypermobility at C1-2, increasing flexion-extension (42%), lateral bending (24%), and axial rotation (5%).[25-27]

There is not enough evidence to provide standardized treatment guidelines, but there are recommendations for this treatment of C1 fractures.[28] Isolated ventral or dorsal ring fractures may be treated with cervical immobilization (collar or halo) for 8 to 12 weeks with good results. C1 burst fractures without ligamentous injury can be treated with collar or halo immobilization for 12 weeks. C1 burst fractures with rupture of the transverse ligament may be treated with halo immobilization for 12 weeks or with internal fixation of C1 to C2 with fusion.

C2 Fractures

C2 fractures can be broadly divided into odontoid, C2 body, and pedicle/pars fractures. Odontoid fractures are classified by the system devised by Anderson and D'Alonzo.[29] Type 1 fractures are rare and are at the distal tip of the odontoid process. Type 2 fractures occur at the base of the odontoid where it meets the body of the axis. Type 3 fractures occur through the body of the axis. The management options for odontoid fractures depend on the type of fracture, the degree of subluxation of the cranial fragment, and the status of the transverse ligament. Type 1 and type 3 fractures are often managed by external immobilization alone, collar, or halo.

Type 2 fractures can be managed by immobilization or operative intervention, depending on patient factors and the degree of subluxation. An increased rate of nonunion has been associated with patients older than 50 years, subluxation greater than 4 to 6 mm, and dens displacement greater than 5 degrees after closed reduction.[30-32] Nonunion rates can be as high as 28%. Type 2a fractures, characterized by comminution of the C2 body, are associated with lower healing rates without surgery.[33,34] C2 pars and pedicle fractures may require surgical intervention, depending on the degree of angulation and distraction between the fragments (see the subsequent discussion).[31,34]

Os odontoideum is defined as an ossicle of cortical bone in the position of the odontoid process often attached to the C2 body by a cartilaginous segment. The cause remains unclear. There is some evidence to suggest that this is a consequence of old trauma, often at an early age.[35] It is unlikely that this condition is a failure of fusion during development, because the normal somite pattern of development of the axis does not normally have a site of fusion where the axis meets the body.[36] However, os odontoideum is associated with congenital disorders, such as Down and Morquio syndromes, and spondyloepiphyseal dysplasia. Patients who have neurologic compromise are offered surgical decompression and fusion. Patients with gross instability or narrow canal diameter are also offered surgery. The treatment of incidentally found os odontoideum is controversial. Most authors recommend close follow-up, with surgery reserved for the development of symptoms or radiographic evidence of instability or progressive deformity.[36,37]

Fractures of the C2 pars interarticularis are called hangman's factures because of the similarity to those seen in judicial hangings.[38] These fractures are also called C2 traumatic spondylolisthesis fractures. They have been classified into three types by Effendi and colleagues, though the Francis classification has also been used.[39,40] Effendi type 1 fractures are displaced less than 2 mm and minimally angulated, and the C2-3 disc space remains intact. Type 2 fractures have a displaced and angulated body of the axis and a disrupted C2-3 disc space. Type 3 fractures are like type 2 fractures with locked C2 and C3 facets, and the body of the axis is ventrally displaced.

Decisions regarding the treatment of C2 pars fractures are primarily guided by the degree of subluxation of C2 on C3, disruption of C2-3 disc space, or inability to achieve fracture alignment with external immobilization.[33] A type 1 fracture without significant ligamentous injury can be treated with immobilization. A halo ring can be used to achieve reduction by extension and capital flexion, reversing the mechanism of fracture. When significant ligamentous injury exists, care must be taken with the use of traction to avoid iatrogenic separation of C2 and C3. In type 2 or type 3 fractures, if there is displacement greater than 3 mm, operative intervention may be indicated for reduction and fixation.[30,40]

C2 transverse process fractures do not cause instability, but potential injury to the vertebral artery is an area of concern. It is unclear whether aggressive imaging or treatment of these injuries affects patient outcomes, and decisions should be individualized depending on patient symptoms and anatomy.[41]

In cases of combination C1 and C2 fractures, there is an increased incidence of neurologic deficit compared with either isolated C1 or C2 fractures. The type of axis fracture typically determines the management.[42]

DEGENERATIVE DISEASE

Abnormalities of bone metabolism, degeneration of synovial joints, or abnormal stresses placed on the CVJ can result in basilar impression. The principles of diagnosis and treatment remain the same, regardless of the cause.

Rheumatoid arthritis (RA) is the most common degenerative disorder of the CVJ. RA is characterized by destruction of synovial joints. The disease is estimated to affect 0.8% of the Caucasian adult population in the United States, about 2.2 million people. The cervical spine is the second most commonly involved region of the body.[43] The degenerative changes seen in the cervical spine are progressive in nature. Translational subluxation of C1-2 occurs first, followed by vertical subluxation of C1 on C2.[44,45] Compression of the spinal cord and brain stem occurs as the lateral mass joints are eroded by inflammatory synovitis and the odontoid ascends through the atlas and the foramen magnum. Oda and colleagues found a predictable progression of transverse subluxation to reducible vertical subluxation to irreducible vertical atlantoaxial subluxation.[44] Fujiwara and colleagues redemonstrated this progression and also noted an association between the severity of RA and the progression of subluxation. Patients with less severe RA develop transverse subluxation, those with RA of moderate severity develop a combination of transverse and vertical subluxation, and those with more severe RA develop vertical subluxation.[45] Basilar impression is the ascension of the odontoid process into the posterior cranial fossa and is defined by an abnormal position of the dens with respect to the foramen magnum. As the dens ascends into the posterior fossa, variable symptoms, which include but are not limited to myelopathy and lower cranial nerve deficits, develop. Although motor weakness and sensory changes due to myelopathy are the most common signs, the earliest sign of spinal cord dysfunction is posterior column function.[46] The incidence of cervical fusion in patients with rheumatoid arthritis has been decreasing possibly due to the use of new medical therapies.[47]

Determination of transverse C1-2 instability is performed using the anterior dental interval (ADI) and posterior anterior dental interval (PADI). An ADI greater than 3 mm and a PADI less than 14 mm is considered to be abnormal.[48,49] Vertical subluxation is measured using the Ranawat method. The vertical distance between the center of the pedicles on the axis to a line connecting the ventral and dorsal arches of the atlas is measured. If this distance is less than 13 mm in men and 15 mm in women, vertical subluxation is diagnosed.[44]

Many indices are used to screen for basilar impression from plain radiographs. These indices use bony anatomic landmarks and are the Clark station, McRae line,[50] Chamberlain line,[51] McGregor line,[52] Redlund-Johnell criterion,[53] Ranawat criterion,[54] Fischgold-Metzger line,[55] and Wackenheim line.[56] Riew and associates evaluated the sensitivity and specificity of these standard screening measurements.[57] The most sensitive measurements (the tests with the fewest false-negative results) are the Wackenheim line, at 88%, and the Clark station, at 83%. The Redlund-Johnell criterion is the most specific measurement (it has the fewest false-positive results) at 76%. The Redlund-Johnell measurement has the highest positive predictive value (PPV) of 68%. The Wackenheim line has a positive predictive value of 48%. The Fischgold-Metzger line has a negative predictive value of 100%. The McRae line has the lowest negative predictive value of 75%. The study also found that identification of bony landmarks is difficult and precludes accurate application of these measurement techniques in many cases. Riew and associates have recommended a combination of tests to screen for basilar invagination: the Clark station, the Redlund-Johnell criterion, and the Ranawat criterion.

The goals of treatments of basilar impression are to decompress the brain stem and spinal cord and to reestablish support for the head. Decompression of the neural elements can be achieved either directly or indirectly. Indirect decompression is performed via closed reduction through the use of traction and manual manipulation. Long-standing lesions are unlikely to be reducible. However, a trial of craniocervical traction is warranted. Use of a halo ring provides multiple points of skull fixation and allows for fixation to the thoracic vest once the deformity is reduced. Traction is started with a weight of approximately 7 pounds and is increased to a 15-pound maximum. Attempted reduction is generally limited for 5 to 7 days because the likelihood of further benefit is limited after this period and complications related to immobilization increase.[43]

Special beds have been designed to help prevent complications of immobilization.

The ventral rheumatoid pannus often resolves once the C1-2 junction is fused, which can indirectly decompress the brain stem and spinal cord.[58,59] However, occasionally a ventral transoral approach needs to be used to obtain adequate decompression.[60-64] Intraoperative image guidance may be used to help with the decompression in a region where anatomic landmarks have been distorted.[65-67] Endoscopic techniques have emerged to avoid the complications associated with the transoral approach.[68]

CONGENITAL DISORDERS

CVJ abnormalities are a common component of many congenital disorders. These disorders can be broadly grouped as connective tissue disorders (Down syndrome), Klippel-Feil syndrome, osteochondrodysplasia (e.g., achondroplasia), mucopolysaccharidoses (e.g., Morquio and Lesch-Nyan syndromes), and skeletal dysplasia (i.e., osteogenesis imperfecta), as well as other disorders of development including Goldenhar syndrome, Conradi syndrome, and the Klippel-Feil triad.[69-73] CVJ abnormalities are most commonly seen in Down syndrome (20%) and Morquio syndrome (50%).[74] The anatomic abnormalities of the craniovertebral junction observed in these disorders and their natural history can help guide treatment decisions.

The following discussion reviews the more common disorders of Down syndrome, Morquio syndrome, and achondroplasia to help delineate pertinent concepts. Description and management of other mentioned syndromes can be found in listed references.[36,73,75-77]

CVJ abnormalities in these patients have variable causes. Atlantoaxial instability is a complex entity that has many causes. No matter the cause, the problem requiring fixation is the lack of fixation between the dens and the anterior arch of C1.[78] Laxity of the normal ligamentous structures in the CVJ is one such cause and is likely the major cause in patients with connective tissue disorders, such as Morquio syndrome[79] and Lesch-Nyhan syndrome.[71] It is also a component of Down syndrome.[80] Aberrant ossification of the dens occurs; this could be due to ligamentous laxity and disturbances in blood supply during development because of inordinate mobility[79,81] Patients with skeletal dysplasia, such as osteogenesis imperfecta, have abnormal collagen deposition, resulting in brittle bones that easily develop multiple microfractures. Accumulation of these microfractures leads to ascension of the dens and medial skull base, causing basilar invagination.[70]

Treatment of these lesions is guided by the symptoms and syndrome involved. For example, many children with Down syndrome have asymptomatic atlantoaxial instability.[80,82] Large cohort studies have not demonstrated increased rates of neurologic injury in children with Down syndrome and atlantoaxial instability as compared with their peers without abnormal ADIs. These studies also do not demonstrate a protective effect of restricted activity.[78,82,83] Reduction and stabilization of the CVJ in these cases is not indicated unless there are clear signs of brain stem or upper cervical spinal cord compression.[80,84] Generally, more than 7 mm of atlanto-occipital subluxation has been used as a benchmark for recommending fusion. This is, in part, based on the work of Browd and associates,[85] which demonstrated that atlanto-occipital instability in these patients with > 7 mm of subluxation is often progressive.

Morquio syndrome and other forms of skeletal dysplasia are often found to have an os odontoideum and ligamentous laxity.[86] The cartilaginous os odontoideum deforms with flexion and extension. Radiographically, C1-2 instability manifests as changes in the ADI and is a late finding in affected children. By the time this condition is seen, myelopathy is nearly always present. These patients benefit from prophylactic fusion prior to the onset of myelopathy. The high prevalence of cervical myelopathy demands careful discussions between the surgeon and anesthesiologist as intubation is anything but benign in these cases.[87] The ideal age for this operation has not been determined. Ransford and coworkers[86] have suggested that the surgery be performed at 4 years of age unless myelopathic signs develop earlier. Dorsal occipitocervical fusion can result in complete ossification of the dens, which supports the role of ligamentous laxity in the formation of os odontoideum.[79] It is important to note that other spine abnormalities and instability are not prevented by occipitocervical fusion and must be screened for in this population.[88]

The treatment of patients with other congenital disorders of the CVJ varies depending on the syndrome, symptomatology, and relevant anatomy. The natural history of the disorder as well as the individual anatomic characteristics present in each patient take precedence in treatment decisions.[75] Traction can be used if there is a suspicion that the lesion is reducible. Use of a halo ring allows for the application of corrective forces and subsequent fixation when reduction is completed.[89] If a lesion proves to be irreducible, either after a trial of reduction or radiography, surgery is indicated to decompress the brain stem and spinal cord and to stabilize the CVJ. Traditionally, open surgical fixation, from an anterior or posterior perspective, has been the mainstay of treatment.

Open posterior cervical fixation carries with it a significant risk of injury to the vertebral artery, requiring increased attention to its course on preoperative studies.[90] In the future, endoscopic surgery may provide options for less invasive techniques for accessing the occipitocervical junction, thereby allowing simpler but equally effective means of anterior surgical reduction or fixation to become more common.[91-94]

INFECTION: ATLANTOAXIAL ROTATORY SUBLUXATION AND FIXATION

Infection may lead to a rare syndrome termed *atlantoaxial subluxation* or *atlantoaxial rotatory fixation*. It was originally described by Bell[95] in 1830 but was named after Grisel,[96] a French otolaryngologist who described this syndrome after upper respiratory infection. Children with atlantoaxial subluxation often present with torticollis, holding their head in a "cock-robin" position. There is no clear mechanism of pathogenesis for this entity, although it is associated with infection, trauma, head and neck surgery, RA, Down syndrome, Morquio syndrome, and other congenital cervical anomalies.[97] Battiata and colleagues[98] hypothesized a baseline ligamentous laxity along with an inflammatory response to an infectious process. Pang and associates hypothesized that rather than ligamentous laxity, there is increased friction of the C1-2 joints.[97]

The diagnosis is made by the clinical presentation along with findings of rotation of C1 on C2 seen on axial CT imaging. It is important to differentiate the presentation from muscular torticollis that has other etiologies.[99,100] Using dynamic CT scans, Pang and associates developed a grading system related to the degree of subluxation.[97,101] Type I shows movement of less than 20% with movement of the head away from the affected side. Type II is less sticky, and C1 moves on C2 greater than 20%. Type III shows movement of C1 on C2 past midline but remains abnormal compared with normal controls. MRI can be performed to evaluate for infectious etiology. This grading scale correlated with the difficulty and duration of treatment, with type I atlantoaxial rotatory fixation being much more difficult to treat.[102] Delayed treatment was also associated with greater difficulty and longer duration of treatment for these patients.

Primary treatment of atlantoaxial rotatory fixation involves traction and muscle relaxants. Once a reasonable amount of clinical reduction has been achieved, the patient can be placed in a cervical collar and treated with muscle relaxants for 2 weeks. Any infection needs to be treated completely to help prevent recurrence.[102] Patients whose condition does not reduce or continues to recur may need more aggressive treatment. Closed reduction under general anesthesia can be used in selected cases. Operative C1-2 fixation and fusion can be used to permanently prevent recurrence. Surgical intervention is controversial. None of the patients in Menezes' series of 54 patients required fusion.[62] It is also worth noting a report of intraoperative reduction in patients with subacute and chronic atlantoaxial rotation that obviated the need for preoperative traction, a treatment not often tolerated by pediatric patients.[103]

TUMORS

Tumors of the CVJ produce signs and symptoms of neural element compression and mechanical instability (pain and progressive deformity). The treatment of these tumors depends on prognosis, symptoms, and anatomic configuration. The most common presentations of adults are myelopathy, radiculopathy, and occipitocervical pain.[104] The presentations of children, in order of descending frequency, are occipitocervical pain, paresthesias or dysesthesias of the hands, cranial nerve palsies (most commonly diplopia), and myelopathy.[105] Chordomas and meningiomas are the most frequent tumors in this location, but other primary tumors—osteoblastoma, eosinophilic granuloma, plasmacytoma, chondrosarcoma, and Ewing sarcoma—also occur. Metastatic tumors, including breast tumors and paragangliomas, have also been encountered.

Treatment of patients with CVJ tumors involves decompression of neural structures and a determination of the presence or absence of instability at the CVJ *after* tumor resection. The first step is to determine the direction of the surgical approach. Piper and Menezes divided the axis into four zones that help guide the surgical approach for tumor resection (Fig 146-1).[104] Zone I tumors are in the ventral midline involving the axis, atlas, and lower clivus and are best accessed by the transoral approach, with or without division of the palate or the mandible. Zone 2 tumors are more ventrolateral and often involve the lateral mass of C1 and C2. They are best accessed using a retropharyngeal approach. Zone 3 tumors are located dorsal to the lateral mass and may extend into the occipital condyle or dorsal fossa. These tumors are best accessed using a far lateral approach. However, this approach may not be good for later instrumentation in the case of a fusion. Zone 4 tumors are in the dorsal midline and are resected through a standard midline approach. The need for fusion is then determined based on the contribution of the resected portions of the CVJ to its stability and is not amenable to a black and white algorithm at this time.

Increasingly, endoscopic techniques are being utilized in the resection of tumors at the CVJ, especially in zones 1 and 2.[91,94,106-111] Such endoscopic techniques provide the advantage of increased illumination in narrow, deep operative fields with the additional advantage of increasing the number of working angles available within the confines of the narrow approach. Specific benefits also include limitation of bony removal (in some cases, reducing the need for fusion), early identification of neurovascular structures, and postresection visualization of the resection cavity to ensure complete tumor resection. Despite these advances, resection of tumors at the CVJ remains a microsurgical procedure best accomplished via the visualization provided by an operating microscope. The endoscope is best used as an assistive device in most cases.[111]

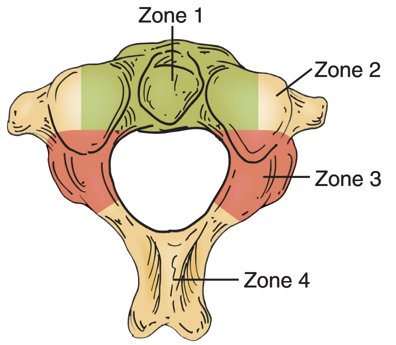

Figure 146-1. Surgical approaches and anatomic zones for tumor resection at the craniocervical junction as described by Menezes and Piper. Zone 1 tumors are in the ventral midline involving the axis, atlas, and lower clivus and are best accessed by the transoral approach, with or without division of the palate or the mandible. Zone 2 tumors are more centrolateral and often involve the lateral mass of C1 and C2. They are best accessed using a retropharyngeal approach. Zone 3 tumors are located dorsal to the lateral mass and may extend into the occipital condyle or dorsal fossa. These tumors are best accessed using a far lateral approach. However, this approach may not be good for later instrumentation of the spine. Zone 4 tumors are in the dorsal midline and are resected through a standard midline approach.

Ventral Odontoid Resection

Dickman and colleagues performed a biomechanical study of transoral odontoidectomy and concluded that resection of the ventral C1 ring, odontoid, and transverse ligament causes increased motion of C1 on C2 and acute/chronic instability.[61] The conclusion from this study was that ventral odontoid resection requires subsequent dorsal fixation. Menezes[112] reported that a minority of patients undergoing odontoidectomy could go without dorsal stabilization. Disagreement also exists regarding the timing of dorsal stabilization. Menezes delayed fixation for 1 week postsurgery and maintained patients in a halo to allow for wound healing and assessment of instability. Crockard and Stevens[81] recommended performing the dorsal stabilization at the time of ventral resection. An increased incidence of infection has not been seen due to immediate dorsal surgery after a transoral resection.[113]

Lateral Condyle Resection and Lateral Mass Resection

Biomechanical studies have shown a significant increase in hypermobility with resection of greater than 50% of the condyle. Resection of the condyle affected the stability of the C1-2 junction as well.[114] Based on their retrospective case series, Shin and associates recommended an occipital-cervical fusion if greater than 50% of a condyle is resected or if the C1 or C2 lateral masses are resected.[115]

DEFORMITY REDUCTION

Deformity can be reduced by either closed or open methods depending on the anatomic configuration. Successful closed reduction with axial traction can sometimes obviate the need for open ventral decompression, or in some cases of trauma, such as traumatic spondylolisthesis, closed reduction can potentially obviate the need for surgical intervention. Closed reduction is achieved with the use of axial traction with a halo fixator. In some cases, manual manipulation under fluoroscopic guidance is used for reduction. The halo is applied carefully, and the force of pin application is tailored to patient age and underlying pathology. Children younger than 2 years or with underlying pathology affecting the skull may not be candidates for a halo ring, and the use of a custom-built Minerva device may be preferable. Children 2 to 4 years of age should have an eight-point halo fixation with an MRI-compatible device.

The pins should be tightened to between 1 to 1½ pounds of torque.[116] The maximum pin torque is 4 pounds for children who are 5 years of age. Traction should be initiated with low weight (4 pounds) in 5-year-old children, and it should not exceed 7 pounds. Halo application and traction are performed under general anesthesia or moderate sedation in some cases. Fluoroscopy, plain radiographs, or even CT or MRI may be used to determine if reduction has been achieved.[117]

Patients who have long-standing degenerative disorders or tumors are generally not candidates for preoperative reduction. These patients require direct surgical decompression via the appropriate surgical route. This can be aided with the use of computerized stereotactic navigation.[55,118] After the decompression, the deformity can be reduced by direct manipulation and appropriate shaping of implants and postoperative immobilization.

SUMMARY

Surgical pathology of the CVJ is quite complex. Treatment of traumatic causes of deformity is primarily guided by clinical suspicion and the identification of instability followed by stabilization. Management of degenerative disease is guided by the natural history and symptomatology. Congenital abnormalities also are guided by the natural history of the disease and symptoms. Tumor management is guided by tumor location and management of subsequent instability related to tumor debulking. The emergence of more advanced endoscopic techniques provides an adjunct with advantages that may be significant. Further development of these techniques will allow increased incorporation of endoscopes into the surgical treatment of CVJ abnormalities. Although technically challenging, appropriate preoperative consideration of the biologic, biomechanical, and pathophysiologic factors associated with CVJ surgery will increase the likelihood of a favorable clinical outcome.

The complete list of references is available online at ExpertConsult.com.

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