Chapter 19
Conception to Implication: Craniocervical Junction Database and Treatment Algorithm

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INTRODUCTION

The earliest anatomic description of abnormalities of the “occipital vertebrae” was attributed to Meckel in 1815 by Gladstone and Erickson-Powell (20). In 1830, Charles Bell first described the clinical and pathological development of “spontaneous atlantoaxial dislocations” resulting from destruction of the transverse ligament “that holds the process of the dentata in its place” (3). After this, there were detailed anatomic and autopsy studies reported in reference to abnormalities of the craniovertebral junction (5, 18, 27, 30, 42). Classic descriptions by Gladstone and Erickson-Powell (20) and Giacomini (19) pointed to the clinical implications. The significance became appreciated after the radiographic study on basilar invagination by Chamberlain in 1939 (6). Radiographic descriptions of the craniocervical abnormalities and their implications, together with the craniometric reference lines, aided in the diagnosis of craniocervical abnormalities on plain x-rays as well as with tomography (6, 12, 25, 26, 30, 31, 52).

Barucha and Dastur published a landmark paper on craniovertebral anomalies in 1964, and pointed to the significant morbidity and mortality to patients who underwent a dorsal procedure for decompression of the atlantoaxial dislocation (2). A year later, Dastur presented several patients who had sudden demise from “hematohydromyelia during decompression” (10). In a similar light, in 1967, Wadia presented a study of 28 patients with “myelopathy complicating congenital atlantoaxial dislocation” (59). At this time, surgical treatment of conditions affecting the craniocervical junction usually consisted of a posterior decompression by enlargement of the foramen magnum and removal of the posterior arch of the atlas vertebra. However, the mortality and morbidity associated with this treatment was high, especially for patients with irreducible cervicomedullary compression (2, 6, 22, 48, 49, 59). The early classification of atlantoaxial abnormalities was made by Greenberg in 1968 (22). By this time, the “direct anterior approach to the upper cervical spine” was presented by Fang and Ong in 1962 (14). However, this morbidity and mortality was significant. In 1967, Edmund Klaus and W Lehman (26) stated that the treatment of basilar invagination and anterior craniovertebral compression would be best managed with an anterior approach, if possible. By this time, my involvement at the craniocervical junction with similar cases had a similar result. It was predictable that an adverse outcome would occur in approximately 35 to 40% of patients treated with dorsal decompression with fixed anterior abnormality at the craniocervical junction. Thus, it was necessary to identify the causes of failures, recognize the pathology, and, as a result, have improved treatment modalities, which led to subsequent operative approaches and the necessity for stabilization (60). The need to understand the natural history of diseases affecting the craniocervical junction was critical, especially because no laboratory or bench experiments could be possible. Thus, it became evident that a long-term follow-up would be necessary to achieve these goals (Fig. 19.1A and B).

Therefore, a prospective database would have to document the imaging modalities, the operative findings, and also be capable of obtaining retrieval archives. A comparison of case studies became necessary with advancing imaging and, hence, the necessity to look again at the original procedures carried out with these patients in comparison with newer understanding with imaging techniques. A prospective treatment method and follow-up became necessary. Our first decision tree for treating abnormalities of the craniocervical junction was proposed in 1978, with eight

patients (39). Subsequently, the cohort was expanded to 17 patients and described in 1980 (33). This was the start of the craniocervical junction database evaluating all patients referred with symptomatic abnormalities at the craniocervical junction. This database currently lists 4800 patients, 4651 of whom have are analyzed for this chapter.

EMERGENCE OF IMAGING TECHNIQUES

In the 1970s, patients with suspected craniocervical abnormalities underwent plain x-rays, pleuridirectional tomograms, and, subsequently, at our behest, tomograms with dynamic motion of flexion, extension, and lateral rotation, as well as documenting the effects of cervical traction (21, 24, 39). Gas myelography supplemented these studies with pleuridirectional tomography and, subsequently, the use of Iohexol to provide contrast for the subarachnoid spaces (57). Vertebral angiography combined with motion studies identified rotational changes as well as compromised vasculature by the craniocervical abnormality (4, 17, 33, 54, 58). This subsequently gave way to the emerging technique of computed tomography (CT) in documenting osseous abnormalities. In 1983, magnetic resonance imaging (MRI) was introduced to the University of Iowa Hospitals and Clinics and an immediate change in our protocol was adapted to this imaging modality. The effects of cervical traction could now be documented with MRI during the traction (28, 33, 39, 40, 41, 57). This led to magnetic resonance angiography, computed tomographic angiography, and the detailed study of cerebrospinal fluid (CSF) dynamics. Throughout this evolution of neurodiagnostic imaging, the initial decision tree has been appropriate.

The factors taken into consideration for treatment of craniocervical abnormalities were (39): 1) reducibility; 2) determining the manner of encroachment on the cervicomedullary junction; 3) identifying vascular compromise; 4) associated neural abnormalities; and 5) the status of the developing craniocervical junction.

REPRESENTATIVE CLINICAL MATERIAL

Patient 1

This 12-year-old male patient presented with vertebrobasilar migraines and episodic loss of vision in his left homonymous visual field. This occurred more frequently with head rotation. On examination, the striking feature was limitation of neck movement and spastic quadriparesis. Pleuridirectional frontal tomography demonstrated atlas assimilation with advanced basilar invagination (Fig. 19.2A). Selective vertebral angiography revealed occlusion of the right vertebral artery by the invaginated odontoid process (Fig. 19.2B).

Patient 2

This 14-year-old individual had the hallmarks of Klippel-Feil syndrome and difficulty swallowing, repeated aspiration pneumonia, and diminished gag reflex. He had marked weakness in his upper extremities with significant spasticity in the lower extremities. In 1978, a pleuridirectional tomography with gas myelography demonstrated atlas assimilation with odontoid invagination through the center of the foramen magnum, indenting the inferior medulla (Fig. 19.3). He underwent transpalatopharyngeal resection of the clivus and the odontoid process with a subsequent dorsal occipitocervical fixation.

Patient 3

A 76-year-old man with advanced rheumatoid arthritis was referred after being seen at several institutions. He had rheumatoid odontoid vertical migration into the posterior fossa, 3 cm above the plane of the foramen magnum. The odontoid tip was intradural and adherent to the vertebral and basilar arteries (Fig. 19.4A and B). The odontoid process was removed, leaving the apex intra-arachnoid and, thus, preventing a CSF leak. Unfortunately, cervical traction was of no avail, and a fracture occurred at the odontoid base before resection. This was one of the earliest descriptions of rheumatoid invagination.

Patient 4

This 14-year-old female patient presented with difficulty swallowing and slurred speech caused by vagal, glossopharyngeal, and hypoglossal palsy. She had a spastic brainstem myelopathy. MRI scans identified a proatlas segment abnormality with invagination into the mid medulla (Fig. 19.5A, B, and C). The advanced imaging technique of 3-dimensional (3-D) CT correctly identified the osseous abnormality. The patient was treated with transpalatopharyngeal resection of the clivus and the offending pathology. A dorsal occipitocervical fusion completed her treatment. Subsequent MRI showed resolution of the cervical syrinx and decompression of the medulla (Fig. 19.5D). Her neurological status reverted to normal.

Patient 5

This 19-year-old individual had a craniocervical chordoma invading the lower medulla and upper cervical spinal cord (Fig. 19.6A, B, and C). A far lateral-posterolateral approach to the craniocervical junction allowed for resection of the tumor, and a subsequent ventral transoral procedure completed the tumor resection.

J.H.

A 36-year-old patient with spastic quadriparesis was recognized to have a grossly unstable dystopic os odontoideum with sagittal and coronal plane instability on 3-D CT and motion studies (Fig. 19.7A, B, and C). The patient was treated with cervical traction followed with documentation of relief of the compression on the cervicomedullary junction by x-rays. He underwent transarticular screw fixation between the axis and the atlas vertebra and an interlaminar rib graft fusion (Fig. 19.7D).

SYMPTOMATIC CRANIOVERTEBRAL ABNORMALITIES—1977 to 2004 DATABASE ANALYSIS OF 4651 PATIENTS

A primary basilar invagination was identified in 728 patients (35). Of these, an atlas assimilation was recognized in 560 patients, the Klippel-Feil syndrome in 502 patients, and hindbrain herniation consistent with the so-called Chiari I malformation in 340 patients (13, 37). The occurrence of syringohydromyelia in one-third of the patients is interesting (3).

In analyzing the symptomatic rheumatoid patient with involvement of the craniocervical junction, 1532 patients were documented (7, 8, 29, 38, 41, 45, 49, 55). An atlantoaxial dislocation was observed in 630 patients, cranial settling with odontoid upward migration in 706 patients, and rheumatoid granulation masses causing ventral compression of

the craniocervical junction in 108 patients. A complex picture of cranial settling with fracture and rotary dislocation was observed in 88 patients. Cervical traction corrected the atlantoaxial dislocation and the cranial settling in 81% of patients (8, 41). This is especially crucial for management because these fragile individuals could then be treated with only a stabilization procedure after the reduction was made (9, 43, 49). The acute serous effusion in the granulation mass representing destructive rheumatoid disease was identified on T2-weighted MRI as fluid within the mass around the odontoid process, which was enhanced with gadolinium injection (32). When this was recognized, a dorsal occipitocervical fusion was made after cervical traction showed improvement in the compression. In these individuals, it was recognized that the ventral granulation mass receded after the immobilization and stabilization of the craniocervical junction. For patients with symptomatic rheumatoid patient with involvement of the craniocervical junction who have a significant co-morbidity, our subsequent protocol has called for a Minerva-type brace without an operative intervention. This has obviated the necessity for operating on these fragile individuals.

Inflammatory states affecting the craniocervical junction were observed in 221 patients (23, 32, 33, 47). A postinfectious dislocation at the craniocervical junction was observed in 122 patients; pseudogout, as with calcium pyrophosphate deposits, causing craniocervical masses in 43 individuals (38, 61); ankylosing spondylitis with dislocation at the occipitoatlantoaxial articulation was observed in 35 patients; psoriatic changes akin to rheumatoid was observed in 13 patients; and regional ileitis as well as Reiter's syndrome affecting the craniocervical junction in 8 patients.

Tumors at the craniocervical junction were divided into 382 osseous tumors and 426 neural tumors. This will be separately presented. Spondyloarthropathies occurred in 222 individuals (32, 34, 46, 56). Achondroplasia had a significant role in 42 patients. In patients with achondroplasia, we identified the infolding of the dural shelf dorsally, compressing the cervicomedullary junction, as well as paramesial invagination leading to apnea and difficulty with swallowing in patients between the ages of 6 months and 2 years. In addition, it was recognized that the posterior arch of the atlas folded inward, causing a dorsal compression. The members of the pediatric radiology service also placed the subsequent widening at the foramen magnum in the natural history of patients with achondroplasia. Bone-softening states associated with Paget's disease, Rickets, hypoparathyroidism, and osteogenesis imperfecta form the large group in this series. Sixty patients, between the ages 1 to 14 years, with spondyloepiphyseal dysplasia were evaluated. Patients with mucopolysaccharidosis formed a smaller group. Down's syndrome was recognized in 104 patients, Goldenhar's syndrome in 46 patients, and craniofacial dysostosis causing craniovertebral junction abnormalities in adolescence was recognized (34, 53). Fifty percent of the patients with Down's syndrome and an abnormal craniovertebral junction had an occipitoatlantal instability, and the remainder had an atlantoaxial instability. Failure to distinguish between the involvement of the occiput with C1 and mere fusion of the atlantoaxial articulation led to unfavorable results.

Major traumatic lesions (1) at the craniocervical junction resulting in occipitoatlantoaxial dislocation occurred in 104 patients, rotary luxation in 152 patients, and a complex ligamentous injury without odontoid fracture in 119 patients (57).

There were 540 odontoid abnormalities. A dystopic os odontoideum was observed in the majority of the patients with odontoid abnormalities (15, 16, 33, 36). Early investigation into the genesis of os odontoideum was made in each individual regarding cervical trauma in early childhood (36, 44, 51). In one-third of the individuals, material was available documenting an intact odontoid process at the time of initial evaluation for cervical trauma with subsequent

development of the os odontoideum. The category of miscellaneous pathology included fibrous dysplasia of the clivus and occipital condyles, fibrocartilaginous herniation at the foramen magnum and C1, and meningocele of the sphenoid and clivus.

A working classification of abnormalities at the craniocervical junction is presented in Table 19.1 (34).

CRANIOCERVICAL PATHOLOGY PUBLISHED FROM THE DATABASE ANALYSIS

This large-volume patient base spans from birth to 90 years of age (50). It has allowed the understanding of the natural history and the pathology of abnormalities at the craniocervical junction; thus the adaptation of treatment modalities to the following disease states: 1) rheumatoid arthritis; 2) atlas assimilation; 3) os odontoideum; 4) Down's syndrome; 5) skeletal dysplasias; 6) bone-softening states, such as osteogenesis imperfecta; and 7) tumors of the craniocervical junction. Thus, the surgical approaches have been modified to individually treat each patient and disease state. In a similar manner, fusion techniques involving the craniocervical junction have evolved to include instrumentation and the evaluation of the effects of fusion throughout the age span.

A significant number of patients with osseous abnormalities of the craniocervical junction have a hindbrain herniation that is mainly the result of reduction in posterior fossa volume. The recognition of osseous and neural abnormalities and a prospective mode of treatment is presented elsewhere in this book.

TREATMENT OF CRANIOVERTEBRAL ABNORMALITIES

A surgical physiological approach to treating abnormalities of the craniocervical junction was first proposed in 1977 and has undergone only slight modifications. The factors that influence the specific treatment of craniovertebral junction abnormalities are as follows: 1) the reducibility of the bony lesion (i.e., the ability to restore anatomic alignment, thereby relieving compression of the neural structures); 2) the mechanics of compression and the direction of encroachment; 3) the cause of the pathological process as well as the presence of hindbrain herniation, syrinx, and vascular abnormalities; and 4) the presence of abnormal ossification centers and epiphyseal growth plates.

The primary aim of treatment is to relieve compression at the cervicomedullary junction (Fig. 19.8). Stabilization is paramount in reducible lesions to maintain the neural decompression. Irreducible lesions require decompression at the site where the compression has occurred; these can be subdivided into ventral and dorsal compression states. In ventral compression states, the operative procedure is ventral decompression through a transpalatopharyngeal route, a LeForte dropdown maxillotomy, or lateral extrapharyngeal route (11, 32). In dorsal compression states, posterolateral decompression is required. If instability is present after decompression, posterior fixation is mandated for stability (50). Therefore, it is necessary to select the operation or combination of operations for each individual patient on the basis of a clear understanding of the pathophysiology and the functional anatomy.

Cervical Traction

The crown halo is preferred to a complete ring because the complete ring causes distortion on MRI. For children less than 2 years of age, 8- to 10-point cranial fixation is used. Pin fixation pressures are individualized by age, cranial thickness, and disease state. At 2 years of age, only finger tightening is used; at 4 years of age, 3 pounds of pin

pressure is used; at 8 years of age, 6 pounds of pressure is used; and above age 10 to 12 years, 8 pounds of pressure is used. In an adult, traction starts at 7 pounds and does not exceed 15 pounds for the craniocervical region. It is important that the crown halo be placed below the equator of the cranium. Reducible lesions that are the result of inflammatory states or recent trauma will respond to conservative management with external immobilization once reduction is achieved. The healing is usually ligamentous and may include bony reconstitution. If this does not occur, or if the condition is not the result of trauma or infection, a bony fixation is mandated.

In diseases such as spondyloepiphyseal dysplasia, mucopolysaccharidosis, osteogenesis imperfecta, Goldenhar's syndrome, and allied situations in young children, the problem of instability at the craniocervical junction is a difficult one. It is essential that the treating physician be able to recognize the epiphyseal growth plates, if present, using thin-section CT with 3-D reconstruction. When there is an absence of growth plates, development can be allowed to occur by supporting the occipitocervical region with a custom-built cervical orthosis, revised every few months. The young child or toddler is reevaluated periodically with diagnostic procedures aimed at identifying the status of the developing craniocervical junction. If the components of the craniocervical junction are not present by 3 to 4 years of age, then a surgical approach with fusion is necessary.

FUTURE INVESTIGATIONS

The future of solving problems at the craniovertebral junction lies with understanding the control genes, the chemical and environmental influences, and our responses to them. Epiphyseal maturation has expanded our understanding of the problems recognized at birth and prevention of neurological deficits.

It is only through recognition of the natural history of developmental and acquired states that we can propose early intervention, before neurological disaster occurs. Surgical treatment of problems at the craniocervical junction presently involve designing extensile approaches to the craniocervical junction, minimally invasive techniques, and the understanding of the biomechanics at the craniocervical junction using digital and computer generated models. This allows for the design of newer constructs at the craniocervical junction. Neurodiagnostic imaging has certainly given us a better understanding of the disease and our ability to treat it.

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FIG. 19.1 *A*, midsagittal T1-weighted MRI in a 16-year-old male patient with basilar invagination and odontoid compression of the ventral medulla. Note the hindbrain herniation. *B*, midsagittal T1-weighted MRI 6 years after posterior fossa decompression for the so-called Chiari I malformation. Note the marked worsening of the basal upward invagination, now indenting into the pontomedullary junction by the invaginating clivus odontoid articulation and secondary hydrocephalus. The patient had marked worsening of his neurological status.

FIG. 19.2 *A*, frontal pleuridirectional tomogram through the plane of the odontoid basilar invagination. This 12-year-old male patient has atlas assimilation. He presented with spastic quadriparesis and visual loss in a homonymous field. *B*, frontal projection of right vertebral angiogram demonstrating occlusion of the vertebral artery at the formation with the basilar artery by the odontoid invagination.

FIG. 19.3 Midline pleuridirectional tomogram with gas myelography. The odontoid process occupies the equator of the foramen magnum with severe medullary compression.

FIG. 19.4 *A*, composite of midline sagittal pleuridirectional tomogram of the craniocervical junction (left), with interpretation on the right. This represents odontoid vertical migration in advanced rheumatoid disease. *B*, composite of axial CT myelogram with Iohexol enhancement of the CSF. The odontoid process is seen ventral to the medulla and attached to the junction of the two vertebral arteries. The odontoid process has now achieved a subarachnoid location.

FIG. 19.5 *A*, midsagittal T1-weighted MRI of brain and cervical spinal cord. There is a bony mass indenting into the mid medulla with cerebellar tonsillar ectopia. *B*, midsagittal section through 3-D CT of craniocervical junction. There is an extension of the clivus (proatlas segmentation abnormality) into the ventral foramen magnum. *C*, composite of T1-weighted (left) and T2-weighted (right) MRI of the posterior fossa and cervical spinal canal. The proatlas ventral bony abnormality is seen indenting into the medulla with a hindbrain herniation present. Note the cervicothoracic junction syrinx. *D*, composite of T1-weighted and T2-weighted MRI of the posterior fossa and cervical spine made 6 months after transpalatopharyngeal decompression of the ventral craniocervical abnormality and a dorsal occipitocervical fixation. The medullary compression has been relieved and the syrinx disappeared.

FIG. 19.6 *A*, composite of preoperative midsagittal T1-weighted MRI of craniocervical region in the midsagittal and parasagittal planes. There is a large clivus chordoma invaginating into the posterior fossa and upper cervical spinal canal with significant neural compromise. Note the presence of tumor at the anterior craniocervical border. *B*, composite of axial MRI with gadolinium enhancement through the level of the foramen magnum (left) and 2 cm below the foramen magnum. The ventrally located chordoma is well seen with medullary compression. *C*, postoperative T2-weighted midsagittal MRI after posterolateral decompression and resection of the chordoma. A residual portion, present at the anterior craniocervical border, was subsequently removed via a transoral approach.

FIG. 19.7 *A*, composite of lateral cervical spine x-rays in the extended (left) and flexed (right) positions. There is a dystopic os odontoideum present with grossly abnormal motion between the atlas and the axis vertebrae. *B*, frontal view of 3-D CT of the craniocervical junction. Note the hypoplastic dens and the atlantoaxial side-to-side dislocation. *C*, composite of 2-D CT in the coronal, sagittal, and axial planes. The dystopic os odontoideum and the dislocation are well visualized. *D*, postoperative lateral cervical spine x-ray demonstrating restoration of sagittal alignment by transarticular C2-C1 screw fixation and bilateral interlaminar rib graft fusion.

FIG. 19.8 Treatment of craniovertebral abnormalities. The primary aim of treatment is to relieve compression at the cervicomedullary junction.