

## Chapter 7

# Craniocervical Developmental Anatomy and Its Implications

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### INTRODUCTION

Congenital and developmental osseous anomalies and abnormalities that affect the craniovertebral complex can result in neural compression, vascular compromise, and can manifest with abnormal cerebrospinal fluid dynamics. The basis for understanding these problems is derived from knowledge of the embryology, developmental anatomy, and the biomechanics of the region. This chapter discusses the embryology, normal development, and abnormal development of the craniovertebral junction and its implications.

### EMBRYOLOGY AND DEVELOPMENT AT THE CRANIOVERTEBRAL JUNCTION

The bony cranial base is developed by a process of enchondral ossification, in which a cartilaginous framework is first developed and subsequently resorbed with further deposition of bone, caused by distorting forces, such as eye development and brain development (10). The clivus and cranial base elongates by sutural growth at the sphenoccipital and sphenopetrosal synchondrosis and by further sutural growth along the lateral portion of the cranial base (27). On the other hand, the facial bones and the majority of the cranium develop by intramembranous ossification. This development bypasses the intermediate cartilaginous phase characteristic of the development of the bony cranial base (4, 7, 14).

During the fourth week of gestation, 42 somites are formed (9). There are four occipital somites, eight cervical, 12 thoracic, five lumbar, five sacral, and 8 to 10 coccygeal pairs (1, 10, 30). Each somite differentiates into an outer dermatome and inner myotome and a medial sclerotome. The sclerotomes are ventral-medial in their location and will form the vertebral bodies. These ventral-medial bilateral cells migrate toward the midline and surround the notochord. Each sclerotome will develop the fissure of Ebner, which is a central cleft that divides a loose collection of cells cranially from a dense cellular area caudally. In this development, the cells from the fissure of Ebner migrate toward and encase the notochord to become the precursors of the intervertebral disc (24).

The superior half of one sclerotome unites with the lower half of its neighbor and, thus, forms the earliest manifestation of the vertebral body. The first four sclerotomes, however, will not follow this course, and essentially fuse to form the occipital bone and posterior portions of the foramen magnum. Simultaneously, vascularization of the occipital bone begins and differentiation of ganglia and vascular tissue begins. The hypoglossal and first cervical arteries demarcate the caudal occipital segment (12).

Simultaneously, the nervous system begins to differentiate. During the fifth and sixth weeks, further differentiation of various parts of the brain and spinal cord occurs. The roof of the fourth ventricle, however, thins out in the midline to form the foramen of Magendie and, laterally, the foramen of Luschka (27, 30). This occurs as an opening in approximately the seventh week, when a connection between the fourth ventricle and the subarachnoid space is established.

The occipital sclerotomes correspond to the segmental nerves that group together to form the hypoglossal nerve, with a path through the individual foramina within the bone (33). The first two occipital sclerotomes ultimately form the basiocciput. The third sclerotome is responsible for the exoccipital bone, which forms the jugular tubercles. The key sclerotome in the understanding of the craniovertebral junction abnormalities is the fourth occipital sclerotome, or the proatlas. Figure 7.1 and Table 7.1 identify the embryology and the development of the craniovertebral junction. The hypocentrum of the fourth occipital sclerotome forms the anterior tubercle of the clivus. The centrum itself forms the apical cap of the dens and the apical ligament (9, 13, 25). The neural arch component of the proatlas divides into a rostral ventral segment and a caudal segment. The ventral portion forms the anterior margin of the foramen magnum as well as the occipital condyle and the midline third occipital condyle. The cruciate ligament and the alar ligaments are condensations of the lateral portion of the proatlas. The caudal division of the neural arch of the proatlas forms the lateral atlantal masses and the superior portion of the posterior arch of the atlas.

The first spinal sclerotome forms the atlas vertebra. It is modified from the remaining spinal vertebrae, wherein the centrum is separated to fuse with the axis body and, thus, forms the odontoid process. The neural arch of this first spinal sclerotome proceeds to form the posterior and inferior portion of the C1 arch (27). At times, the hypochordal bow, instead of disappearing, may survive and join with the anterior arch of the atlas to form a variant with an abnormal articulation, which then exists between the inferior clivus, the anterior arch of the atlas, and the apical segment of the odontoid process (26).

In our hands, recent computed tomographic (CT) evaluation of the atlas has shown that several ossification centers are present in the atlas development (26). However, the lateral atlantal masses must be present at birth. A complete ring must form by 3 years of age. Abnormal development is observed with the skeletal dysplasias, such as spondyloepiphyseal dysplasia; achondroplasia; Goldenhar's syndrome; and in genetic abnormalities, such as Down's syndrome.

During embryogenesis, the hypocentrum of the second spinal sclerotome disappears. The axis body is formed by the centrum, and the division of the neural arch forms the facets and the posterior arch of the axis vertebra (15). Thus, the dens appears from the first sclerotome, whereas the terminal portion of the odontoid process arises from the proatlas. The most inferior portion of the axis body is formed by the second spinal sclerotome (Fig. 7.2). At birth, the odontoid process is separated from the body of the axis vertebra by a cartilaginous band that represents a vestigial disc and is later referred to as the neural central synchondrosis (27, 38). This is crucial in the understanding of the formation of os odontoideum. The neural central synchondrosis lies below the level of the superior articular facets of the axis and does not represent the anatomic basis of the dens. This synchondrosis is present in most children younger than 3 to 4 years of age, and disappears by age 8 (29). The odontoid process is observed at birth but does not fuse to the base of the axis. The tip of the odontoid is not ossified at birth and is not observed on lateral x-rays. It is represented by a separate ossification center, usually observed at 3 years of age, and fuses with the remainder of the dens by 12 years of age.

Posterior fossa expansion occurs because of enchondral resorption, sutural growth, and bony accretion. Growth of the basal aspect of the clivus elongates the basiocciput and lowers the frontal margin of the foramen magnum. Synchondrosial growth occurs until 16 to 18 years of age. This is comparably matched by a resorbative drift downward and backward at the opisthion because of cerebellar downward displacement together with rotation of the occipital and temporal lobes of the brain. The bony abnormality in hindbrain herniation syndrome has a significance here. Lack

of posterior fossa volume results in herniation of the cerebellar tonsils through the foramen magnum, resulting in tonsillar ectopia (28).

Significant muscle development takes place dorsal and lateral to the cervical spine to provide for the top-heavy cranial end of the fetus (27). The stability of the craniovertebral articulation, with its forward inclination, is dependent on maintaining the geometry of the articular surfaces of the craniovertebral junction and the ligamentous attachments and, more importantly, the heavy musculature. Thus, it can be observed that the ideal fusion at the craniocervical junction should be dorsal.

The discovery of developmental control genes has led to significant advances in furthering the understanding of the craniocervical junction. Two families of regulatory genes have been implicated in the development of the sclerotomal parts of the somites during their resegmentation to form the specific identity of each vertebra (7, 19). They promote proteins that modulate morphogenesis by influencing the transcription of specific downstream genes. Teratogen-induced disturbance of HOX gene expression and mutation in the HOX genes can cause alterations in the identity or number of cervical vertebra that are formed. Inactivation of the *hox-d3* gene results in mutant mice with assimilation of the atlas to the basiocciput (5). The sensitivity of the occipitocervical junction to disturbances in this gene expression might prove to be the underlying cause of malformations in this region. PAX genes are expressed in diverse cell types and contribute to the development of the early nervous system (18). Control of resegmentation of the sclerotomes to establish vertebral boundaries seems to be independently controlled by two regulatory genes in the PAX family.

#### IMPLICATIONS OF CRANIOVERTEBRAL ABNORMALITIES

Table 7.2 provides a practical classification of the most frequently encountered congenital cervical anomalies, which are divided into those that are present at birth (congenital) and those that are developmental, which have an abnormal embryology leading to symptomatic abnormalities during early childhood and into adulthood (26). The immediate relevance of this classification is the understanding of the basis of abnormalities that occur with atlas assimilation, remnants of the occipital sclerotomes, fusion abnormalities, os odontoideum, basilar invagination, segmentation failures at the occiput, atlas and axis vertebra, and, more importantly, the natural history. Thus, a wide variety of abnormalities exist, which can occur singularly or in multiples in the same individual; involving both the osseous and neural structures. An insult to both of the structures may occur between the fourth and seventh week of intrauterine life, resulting in a combination of abnormalities consisting of failures of segmentation, failure of fusion of different components of each bone, hypoplasia, and ankylosis.

There is a high incidence of both anterior and posterior spina bifida of C1 as well as os odontoideum in connective tissue diseases, such as mucopolysaccharidosis, Down's syndrome, and Morquio's syndrome. This subsequently results in the atlantoaxial subluxation. It is possible that, because of the abnormal excessive head movements in the embryo between the 50th and 53rd day, the process of chondrification is impaired, resulting in anterior and posterior spina bifida of C1 (27). This has previously been alluded to in the development of the atlas. Spinal trauma in children younger than 8 years of age is mainly centered at the craniovertebral border because of the high fulcrum of neck motion (25). This results in ligamentous injuries more than fractures. However, odontoid fractures in this age group are usually observed as avulsion injuries with separation of the neural central synchondrosis (25).

Growth of the posterior fossa and especially the clivus continues past late adolescence and certainly provides a rationale for the need to continue observing children who have undergone an occipitocervical stabilization or craniovertebral decompression. The downward growth of the brain and the elongation of the posterior fossa and clivus may recreate a ventral bony abnormality later in life, despite a satisfactory previous ventral decompression at the craniovertebral junction performed during the first two decades of life. We have observed this occur, although infrequently, in our patients with ventral or dorsal posterior fossa decompression.

Abnormalities of the craniovertebral junction must be suspect in infants with Goldenhar's syndrome, skeletal dysplasias, and the Conradi syndrome. It should be suspected in infants who present with torticollis. Diseases such as Down's syndrome have a 14 to 20% incidence of atlantoaxial dislocation (35). Once the stage is set by congenital craniovertebral anomalies, the developmental and acquired phenomenon may supervene, producing atlantoaxial instability and, subsequently, basilar invagination. This is more common in developing countries where heavy loads are carried on the head from childhood. An erroneous diagnosis of "congenital dislocations" thus appears in their literature. Likewise, upper respiratory infections can cause stiff neck, torticollis, and ligamentous instability, and may come to attention later in developing countries than in places where medical attention is readily available. For this reason, it seems that abnormalities of the craniovertebral junction are more frequently encountered in the populous and less advantaged countries.

In 1981, Marin-Padilla demonstrated that the basichondrocranium of fetuses with hindbrain malformations, such as the Chiari syndrome, is shorter than normal and elevated in relation to the axis of the vertebral column (22). The shortness of the basichondrocranium of these fetuses is attributed to underdevelopment of the occipital bone, especially noticeable in its basal component. The defect results in a short and small posterior fossa, inadequate to contain the developing nervous structures of that region. The elongation of the odontoid process, referred to as the "dolicho-odontoid process," is explained by the depression of the basiocciput, resulting in the basilar impression often observed in the clinical Chiari malformations. These changes have been experimentally reproduced in pregnant hamsters by a single dose of vitamin A early in the morning of the eighth day of gestation, thus, inducing a typical Chiari II malformation as well as various types of axial skeletal dysraphism (21).

#### Proatlas Segmentation Failures or Manifestations of Occipital Vertebrae

Malformations and anomalies of the most caudal of the occipital sclerotomes are caused by proatlas segmentation failures. These abnormalities surround the foramen magnum and usually involve the posterior arches of C1 (9, 12, 27, 32, 36). A hindbrain herniation is associated in 33% of individuals (28). At times, the proatlas component of the dens may fail to separate from that portion that forms the basiocciput of the clivus. Thus, the anterior arch of the atlas comes to rest above the axis body. At times, the proatlas abnormality is united with the clivus, grossly distorting the cervicomedullary junction ventrally. Variations of this may be observed in the midline ventrally, laterally, and, at times, dorsally. Thus, one may see paramesial invagination.

In our series of 70 patients, 85 to 90% presented between the first and second decade of life. The earliest presentation was 3 years of age and the oldest was 23 years. A spastic quadriplegia was a presenting symptom in 80% of the patients and lower cranial nerve palsies in 33% of the patients. Vascular presentation of vertebrobasilar system dysfunction was observed in 40%, and trauma presentation in 60% of individuals.

The best definition of the abnormality was on three-dimensional (3-D) CT combined with magnetic resonance imaging. Earlier in the series, pleuridirectional tomography was used in the frontal and lateral projections (25). A hindbrain herniation was present when the posterior fossa volume was reduced, especially by distortion of the vertical height of the posterior fossa. Thus, the treatment of this condition rests with precise definition of the anatomic and pathological abnormality, relief of neurovascular compression, and prevention of recurrence by stabilization. The surgical approach depends on the manner of encroachment on the neurovascular structures. The patient shown in Figure 7.3A–C required a transpalatopharyngeal decompression of the ventral cervicomedullary junction with a dorsal occipitocervical fixation. The patient shown in Figure 7.4A–E required a posterior approach for decompression of the dorsolateral cervicomedullary junction and a fusion procedure. However, the patient shown in Figure 7.5A–C was managed with a posterolateral decompression of the foramen of the occipital condyle on the left and a simultaneous midline approach on the right, decompressing the medial condyle, and a fusion procedure.

#### Assimilation of the Atlas and Klippel-Feil Syndrome

This results from failure of segmentation between the fourth occipital sclerotome and the first spinal sclerotome (11, 13, 38). It may be unilateral, segmental, focal, or bilateral. In most instances, it occurs in conjunction with abnormalities such as the Klippel-Feil syndrome. Basilar invagination is a secondary phenomenon. The finding of atlas assimilation was present in more than 500 individuals who were evaluated for craniovertebral abnormalities (29). A hindbrain herniation occurred in 38 to 40%, caused by reduced posterior fossa volume. Segmentation failures of the second and third cervical vertebra, in association with atlas assimilation, leads to an excessive load on the atlantoaxial motion segment, which subsequently becomes unstable (Fig. 7.6A–C). Initially, a reducible atlantoaxial instability is present. The sequence of events is that there is pannus formation around the odontoid process with the reducible dislocation. However, as the child grows, there is grooving behind the occipital condyles caused by the upward migration of the axis. This is followed by a reducible basilar invagination up to approximately the age of 14 to 15 years. As these children age, the lesion becomes an irreducible basilar invagination. During the phase of reducibility and partial reducibility, a prolific granulation tissue mass crowns the odontoid process in an attempt to reduce the excursion. This, in turn, compounds the compression of the cervicomedullary junction.

In the irreducible basilar invagination, there is an associated horizontally oriented clivus, and the abnormal grooving that occurs behind the occipital condyles also pushes up the cranial base, resulting in platybasia and a short horizontal clivus (Fig. 7.7A–C). This leads to complete irreducibility of the lesion (29). Thus, a child who comes for evaluation between 4 and 16 years of age has a better chance of having a reducible atlantoaxial dislocation or a reducible basilar invagination than a fully grown adult. The upward migration of the cranial base and the reduction of the vertical height of the posterior fossa leads to an acquired hindbrain herniation syndrome (Fig. 7.8A–C). Hence, an operative procedure that relies on posterior decompression without addressing the potential for instability below the age of 20 can lead to subsequent unfortunate results.

Torticollis has been a presenting symptom in children with unilateral atlas assimilation (26). This is critical with the head manipulation that occurs during general anesthesia for patients who require placement of drainage tubes in the tympanic membrane and for adenoidectomy. In this situation, the trunk seems to stay in one position while the head is rotated 90 degrees to either side. As a result, the patients present with a rotary dislocation of the atlas on the axis. It thus behooves the treating physician to be aware of these problems, especially in children with the Klippel-Feil syndrome.

The Klippel-Feil syndrome has a classic triad of short neck, webbed neck, and a low hairline with limitation of neck motion. In this syndrome, deafness, high arch palate, facial palsies, and cardiovascular abnormalities are common. Abnormal rib fusions and scoliosis are observed, and 30% of individuals have genitourinary tract abnormalities.

Thus, the treatment of the reducible atlantoaxial dislocation or the reducible basilar invagination is stabilization; and, should a hindbrain herniation be present, a posterior fossa decompression should be made with the fusion. If, on the other hand, irreducible basilar invagination is present, ventral decompression is warranted. Unfortunately, if a posterior procedure for decompression is made in the face of ventral compression, 30% of individuals would have an unfavorable outcome (25). This is because the ventral abnormality acts as a "peg" impinging on the cervicomedullary or pontomedullary junction when the patient is positioned for the prone procedure. Additionally, fusion in a flexed position compromises the ability to perform a satisfactory ventral decompression. It is important to reiterate that the ability to reduce the invagination is age related. The presence of syringohydromyelia with hindbrain herniation and basilar invagination should not sway the neurosurgeon to perform a posterior operative procedure alone. In most instances, the syringohydromyelia disappears once the ventral abnormality has been corrected.

#### Os Odontoideum

The ossiculum odontoideum is an independent bone in the place of the dens and is cranial to the axis (38). It should not be considered as an isolated dens but exists apart from a small hypoplastic dens. Radiographically, the os has smooth borders and a small dens is always present. It is located in the position of the odontoid process near the basiocciput, where it may fuse with the clivus. The gap between the axis and the free ossicle usually extends above the level of the superior facets of the axis, thus making this an acquired abnormality rather than a congenital abnormality (Fig. 7.9A–C). The entire complex leads to an incompetence of the cruciate ligament and, subsequently, to atlantoaxial instability (25).

In our series, the evidence for os odontoideum has pointed to trauma between the ages of 1 and 4 years in cases where there is a previously recognized normal odontoid process (29). At times, os odontoideum may be associated with an unrecognized fracture in children less than 5 years of age with previously normal odontoid structure, as observed in a large number of patients in our series (8, 25). Os odontoideum may also be associated with nontraumatic situations with ligamentous laxity, such as Down's syndrome and Morquio's syndrome (34). In our series, symptomatic patients were found to have instability in all planes. The biomechanics are complex. It is different for each individual, in that, in some situations, the flexed position may be the best to relieve compression of the cervicomedullary junction, whereas in others, it is the extended position. Thus, the biomechanics must be carefully studied.

Irreducible dislocation was found to be caused by 1) pannus and 2) a cruciate ligament that slipped behind the os odontoideum and in front of the superior portion of the dens. Thus, both the dens and the axis body create the compressive mechanism on the ventral cervicomedullary junction; not the ossicle alone. In severe chronic dislocations, the os may become fixed with severe basilar invagination. At its worst, os odontoideum has significant implications regarding compression of the cervicomedullary junction (Fig. 7.10A–C). I strongly think that all patients with recognizable instability at the craniocervical junction and associated os odontoideum should undergo stabilization. We presently use lateral mass screw/rod fixation or transarticular screw fixation between C2 and C1. In

some circumstances, an occipitocervical arthrodesis may be necessary. If a fixed irreducible abnormality is found, a decompression is first performed in the manner in which compression occurred.

#### Atlas Abnormalities

Failure of development and failure of segmentation of the atlas result in abnormal articulation between the clivus, the atlas, and the odontoid process. Variations may consist of partial absence of the posterior arch of the atlas and a bifid atlas anteriorly or posteriorly. A bifid anterior or posterior atlas results in the two halves of the atlas vertebrae acting like a complex Jefferson fracture with lateral displacement (2, 3, 6, 17, 20, 23, 31). This condition should be considered grossly pathological if it is present beyond the age of 3 years and should be addressed (Fig. 7.11A and B).

I have reviewed 160 normal CT scans of the craniocervical junction in subjects between the ages of birth and 4 years of age. Our conclusion was that the atlas should be a complete ring by 3 years of age. In cases where the atlas persists as being separate with abnormal dynamics, an operative intervention or fusion should be performed. Neurological deficit was observed in 16 infants with a bifid anterior and posterior arch of the atlas or with absent anterior and posterior arches, except for preservation of the lateral atlantal masses. Placement in a custom-built cervical collar and bracing through 3 years of age has allowed for reformation of the anterior atlas arch and stabilization of the craniocervical junction in 60% of children. It is thought that continued motion in an abnormal situation will prevent the formation of the absent segments and, thus, lead to further neurological deficit.

A persistent bifid anterior and posterior arch of the atlas beyond the age of 3 to 4 years is observed in skeletal dysplasias, Down's syndrome, Goldenhar's syndrome, Conradi syndrome, and atlas assimilation. In our series, the presentation was torticollis and plagiocephaly. Neurological dysfunction presented as paresis and apnea and failure to thrive.

The ideal form of imaging is 3-D CT and magnetic resonance imaging (37). As previously mentioned, the treatment in our series consisted of bracing until 4 years of age with repeat 3-D CT on a yearly basis (Fig. 7.12). If instability was still present beyond age 4 years, an occipitocervical bony arthrodesis was accomplished.

#### Axis Spondylolysis

The rare situation of axis spondylolysis has only recently been recognized, because of the improved neurodiagnostic imaging of 3-D CT and the improved motion dynamics with magnetic resonance imaging. A persistent pars defect led to abnormal bone formation in an older individual. In a very young patient, I have observed significant kyphosis and canal compromise at the affected pars defect at the axis vertebrae. The stability of the craniocervical unit is dependent on the geometry of the articular surfaces of the occipital condyles, and the lateral atlantal and axial masses. The integrity is also maintained by the ligamentous complex in the anterior and the middle column. However, the dorsal column is unstable, with a freely mobile segment of bone beyond the pars that may cause hypertrophy at this junction, causing lateral as well as dorsal compression. Decompression and stabilization has been performed in the younger individual.

In an older patient, a pincers-like bony ingrowth circles the spinal cord behind the axis vertebra (Fig. 7.13A–D). Thus,

decompression for axis spondylolysis was mandated by an anterior procedure going through the entire vertebral body and resecting the area of bony compression at the level of the pars defect. This complex situation was encountered in the patient shown in Figure 7.13.

## CONCLUSIONS FOR DEVELOPMENTAL ANATOMY AND ITS IMPLICATIONS

A large database of 4700 patients has provided for the understanding of the natural history of many entities. This database has allowed treatment protocols to be established that have stood the test of time. In addition, the previous presentation in this manuscript confirms the embryology of the craniocervical junction via a clinicopathological correlation. It is important to obtain newer imaging techniques to unravel the complexities of the craniocervical junction.

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Fig. 7.1 Artist's illustration of embryology and development of the craniovertebral junction.

Fig. 7.2 Development of the axis vertebrae.

Fig. 7.3 A, lateral cervical spine and craniocervical x-ray. This 12-year-old patient had a long-standing history of severe spastic quadriplegia with difficulty swallowing. The open arrow points to a bony abnormality within the foramen magnum. The anterior atlas arch is abnormal. B, midsagittal T2-weighted MRI of the brain and upper cervical spinal cord. Note the bony indentation into the ventral cervicomedullary junction. C, a T1-weighted MRI of the craniocervical junction outlines a bony continuation of the clivus into the ventral cervical canal impinging on the cervicomedullary junction. There is no odontoid process present. This is a proatlas segmentation abnormality.

Fig. 7.4 A, lateral cervical spine and craniocervical x-ray in a 9-year-old patient with headaches and occipital neck pain. He had episodic hemiparesis with exertion. Note the abnormal location of the anterior atlas arch and a bony mass descending from the exoccipital bone at the posterior aspect of the foramen magnum. B, Town's view of the cranium. There is a downward descent of the exoccipital bone on the left. C, Axial CT scan through the plane of the foramen magnum and the anterior arch of the atlas. The bony abnormality from the left occipital exoccipital bone indents into the subarachnoid space from the left. D, midsagittal T2-weighted MRI of the craniocervical junction through the plane of the fourth ventricle. Note the dorsal cervicomedullary compression by the bony proatlas segmentation abnormality. E, 3-D CT of the craniocervical junction viewed from within the foramen magnum. A left bony mass descends from the exoccipital bone on the left with absence of the left posterior atlas arch. The right posterior arch of the atlas is seen.

Fig. 7.5 A, midsagittal T2-weighted MRI of the craniocervical junction. There is a bony mass indenting into the lower medulla. B, 3-D CT scan of the craniocervical junction viewed from within the foramen magnum. Note the medially approximating occipital condyles, which are grossly abnormal. This is part of the proatlax segmentation abnormality. C, coronal T2-weighted MRI through the plane of the occipital condyles. The cervicomedullary junction is being pinched by the abnormal occipital condyles, which form part of the proatlax segmentation abnormality.

Fig. 7.6 A, 3-D CT of the craniocervical junction with midsagittal thin section. There is assimilation of the anterior atlas arch and the atlantal masses to the occipital bone. Note the atlantoaxial dislocation. B, midsagittal T1-weighted MRI demonstrating ventral compression of the lower medulla and cervicomedullary junction by the dislocated odontoid process. C, postoperative lateral cervical spine x-ray demonstrating reduction of the atlantoaxial dislocation with bilateral transarticular C2–C1 screw fixation and interlaminar rib graft fusion.

Fig. 7.7 A, midsagittal T1-weighted MRI of craniocervical junction in a 13-year-old male patient with atlantoaxial dislocation and atlas assimilation. This patient had a 1-week history of quadriplegia after a football spearing accident. There is significant compression of the cervicomedullary junction. B, composite of parasagittal and far lateral pleuridirectional tomograms through the craniocervical junction. On the right, it is evident that there is atlas assimilation and segmentation failure of C2 and C3. There is a groove behind the occipital condyle and the lateral atlantal masses into which the superior articular facet of C2 sits. This leads to irreducible atlantoaxial dislocation with the basilar invagination. C, postoperative T1-weighted MRI made 1 week after ventral odontoid resection and cervicomedullary decompression. Further reduction was obtained with traction and subsequent dorsal occipitocervical fusion.

Fig. 7.8 A, this 9-year-old patient had intermittent quadriplegia with worsening severity for 3 months. Midsagittal T2-weighted MRI of craniocervical junction demonstrates odontoid invagination into the posterior fossa with ventral compression of the inferior medulla and a dorsal compression by the posterior rim of the foramen magnum. B, midsagittal reconstruction of 2-D CT of the craniocervical junction. There is atlas assimilation with a dolicho-odontoid process, which occupies the ventral half of the foramen magnum. There is significant basilar invagination. C, 3-D CT of craniocervical junction viewed within the foramen magnum. There is gross ventral compression. The patient responded to a transpalatopharyngeal resection of the ventral craniocervical junction and a dorsal occipitocervical fusion.

Fig. 7.9 A, lateral cervical spine x-ray demonstrating gross atlantoaxial dislocation. The odontoid process is not seen. This patient presented with weakness in the upper extremities and inability to walk. B, midline pleuridirectional

tomogram through the craniocervical junction demonstrating dystopic os odontoideum that is fused to the clivus. There is gross atlantoaxial dislocation. C, postoperative lateral craniocervical x-ray. There is a completed dorsal occipitocervical fusion and the anterior arch of the atlas; the os odontoideum and the superior aspect of the axis body has been resected.

Fig. 7.10 A, midsagittal T2-weighted MRI of craniocervical junction. This 13-year-old became quadriplegic in a wrestling event. He recovered during a 4-week span. There is a ventral soft tissue mass behind the os odontoideum and the axis body. A hyperintense signal is seen in the upper cervical cord corresponding to the axis body and not the os odontoideum. B, open mouth view of the atlantoaxial articulation. There is an os odontoideum present with a hypoplastic dens. C, lateral cervical x-ray made 6 months after dorsal occipitocervical fusion with loop instrumentation and rib graft. The patient made a full recovery.

Fig. 7.11 A, 3-D CT of craniocervical junction in a 14-month-old patient with quadriparesis. There is atlantoaxial dislocation. A previous attempt at dorsal occipitocervical fusion failed. B, axial CT through the atlas demonstrating absence of the anterior arch of the atlas and a bifid posterior arch. There is atlantoaxial dislocation present. This patient responded to cervical traction and a repeat dorsal occipitocervical fusion.

Fig. 7.12 Frontal view of 3-D CT of the craniocervical junction. Note the absence of the right lateral mass of the atlas from the midline backward. There is a hypoplastic dens. This 4-year-old patient was initially evaluated at 6 months of age for torticollis and plagiocephaly. The atlas abnormality was recognized on the initial 3-D CT with subsequent bracing. At age 4 years, a dorsal occipitoatlantoaxial fusion was made with a rib graft.

Fig. 7.13 A, composite of midsagittal and parasagittal T2-weighted MRI of the craniocervical region. There is a ventral bony indentation at the level of the mid axis body indenting into the cervical cord. The posterior arch of the atlas is ventrally displaced. B, composite of a 3-D CT, viewing the cervical canal from within. There is a spondylolysis of the axis vertebra. A bony mass also protrudes into the cervical canal. A dorsal location of the posterior atlas arch is visualized. C, composite of axial CT through the plane of the C2 spondylolysis. Note the abnormal configuration of the axis body as well as the facets. There is spondylolysis with a pincers-like inward bony growth encircling the ventral spinal cord. D, composite of an axial CT through the plane of maximum compression at C2 (spondylolysis segment) and 1 cm below. Note the cervical cord compression at the affected segment. This patient underwent ventral decompression of the axis body and the bony mass impinging into the ventral cervical canal. A dorsal fixation between the posterior arch of C1, C2, and C3 was made. The patient had marked improvement of neurological symptoms.