# The Management of Disorders of the Child's Cervical Spine

Jonathan H. Phillips *Editor in Chief* 

Daniel J. Hedequist Suken A. Shah Burt Yaszay *Editors* 



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This Springer imprint is published by Springer Nature The registered company is Springer Science+Business Media, LLC The registered company address is: 233 Spring Street, New York, NY 10013, U.S.A. This book is dedicated to all children who have disorders of the cervical spine and to the people who care for them.

### Foreword

In the early 1970s, I became interested in the cervical spine, specifically congenital anomalies. That led to the publication of a report on the Klippel-Feil syndrome. I was fortunate to find a monograph entitled *Upper Cervical Spine* published in 1972. The authors, Detlef von Torklus and Walter Gehle, were from the Orthopedic Clinic and Outpatient Department of the University Hospital in Hamburg, Germany. They had done an extensive review of the literature and pathoanatomy of the cervical spine, and, importantly, nearly half of their book was devoted to children. The authors identified many normal physiologic and anatomic variations that frequently mimic pathology. Unlike the extremities, in spine issues, one cannot use a comparison X-ray of the opposite side. Their work identified variations in the pediatric spine and how they differed from the adult. This text became my go-to source for insight in complex cervical spine problems.

The Management of Disorders of the Child's Cervical Spine edited by Jonathan Phillips, Daniel Hedequist, Suken Shah, and Burt Yaszay continues that legacy. This text is comprehensive and includes an extensive review of previous literature by individuals knowledgeable in the management of children with complex cervical spine problems.

Part I, Basic Medical Science, is essential to effective diagnosis and treatment. This section contains important chapters on anatomy, biomechanics, radiology, advanced imaging, and current diagnostic techniques.

Part II, Clinical Aspects of Disorders of the Child's Cervical Spine, contains an extensive discussion of trauma to the immature spine and its potential for serious morbidity and mortality. There is a special section on cervical injury in the young athlete. The clinical aspects of many of the disorders that can affect the child's spine are presented in detail. This list is comprehensive and includes inflammatory conditions, infection, tumors, congenital anomalies, metabolic disorders, and bone dysplasias.

Part III, The Medical and Surgical Treatment of Cervical Disorders in Children, covers management—including conservative techniques such as immobilization and rehabilitation. Also included are surgical approaches, including current instrumentation, anesthesia, and neurological monitoring. There is a unique section on complications and revision surgery. The strength of this text is that it is the product of an international panel of experts, all of whom are recognized authorities. This is coupled with the skill-ful oversight of Dr. Phillips and his colleagues to create a powerful text that will be an important clinical resource for many years. This will be exceedingly helpful to those involved in the management of cervical spine problems of children, and it continues the legacy of von Torklus and Gehle.

Ann Arbor, MI, USA

Robert N. Hensinger, MD

# Preface

There is no one reason why we wrote this book. It came about, as so many different things do, by way of a conversation at the dinner table. Suken Shah, MD; Burt Yaszay, MD; and I were talking at such a dinner table in Orlando at a meeting on early onset scoliosis. We all had a big interest in children's cervical spine problems, but agreed that they were pretty rare and there wasn't much of a forum for talking about them among us orthopedic surgeons who specialize in pediatric problems.

I give Burt the credit for the statement that "peds cervical spine is the last black hole in kids' spinal knowledge" or something like that. And with that prophetic statement the seed was sown.

Suken polled the membership of the Pediatric Orthopedic Society of North America (POSNA), and within a very short time, we had a small but enthusiastic group of interested surgeons who formed the nidus of a new study group which, for now at least, is called the Pediatric Cervical Spine Study Group (PCSSG). The members of this international group have contributed most of the chapters in this text, along with their fellows and other associates. We meet a few times a year at POSNA and Scoliosis Research Society (SRS) and International Congress on Early Onset Scoliosis (ICEOS) meetings and have been supported by these organizations. I'm very happy to acknowledge their support.

One of the early topics we discussed at PCSSG meetings was the possibility of writing a text that could guide the novice surgeon in this rare but dangerous area. Both Fran Farley, MD, and Haemish Crawford, FRACS, were the initial proponents of the idea and contributed chapters. Dan Hedequist, MD, already was involved in writing a book for our publisher, Springer, and put me in touch with Kris Spring in their New York office who has been beyond patient in waiting for a long overdue final draft. Dan, Suken, Burt, and I took on editorial responsibilities for this text, so the four of us are responsible for its content.

There are many others who have put up with the long process of writing, notably our families, of course. But I would also like to acknowledge the help of my colleagues at Arnold Palmer Children's Hospital in Orlando in disciplines apart from orthopedics, namely, neurosurgery, ENT, general surgery, and physiatry, who have written chapters which complete the scope of this book. The final and most important thank you of all goes to my secretary and friend of 20 years, Mary Regling, BA, who has been the "den mother" of the PCSSG from its inception and the driving force behind getting this work published. Without her, the project would have foundered and failed.

Orlando, FL, USA

Jonathan H. Phillips, MD

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### Introduction

This book was written for a wide audience. Some of its readers will be familiar, or even expert, in the care of children with neck and cervical spine disorders. Others will be completely new to the subject. Though its emphasis is on the orthopedic and neurosurgical approach to children's cervical spine, there are chapters that are contributed by other disciplines. Thus, an ENT surgeon who may be called upon to perform an anterior trans-oral approach to the dens will be reassured by the account of this technique in Chapter 19. Chapter 21 focuses on non-spinal disorders which may present to physicians and others encountering children with neck problems in their clinics. Knowing what their significance is and which consultant to engage with in their management is important.

While it is unwise to try to be all things to all people, it is hoped that this is a reference that can be dipped into by the occasional reader looking for something specific and also be a comprehensive guide to the young surgeon embarking on a career which may include pediatric cervical spine surgery.

The area we cover is quite rare and quite dangerous for the unprepared. Yet with the changing demographics of childhood trauma and increasing survival of children with previously lethal syndromes, we are encountering these rare diagnoses with greater frequency.

The reader is encouraged to approach the text in a traditional fashion. We are all anxious to know "how to do it," but such enthusiasm must be tempered by acquiring the building blocks of "why." Thus, we start with basic science, and it cannot be overemphasized how important a thorough knowledge of the anatomy (both normal and abnormal), pathology, biomechanics, and radiology is to treating these rare disorders. The chapters on clinical assessment and presentation of the multitude of problems in this area follow. Only after these basic areas are covered do we embark on accounts of the surgical and nonsurgical management of the problems encountered.

Each chapter is written by experts in the area and can be taken as standalone treatises. It is hoped, however, that the whole will be greater than the sum of its parts.

> Jonathan H. Phillips, MD Daniel J. Hedequist, MD Suken A. Shah, MD Burt Yaszay, MD

# Part I

**Basic Medical Science** 

# Embryology and Anatomy of the Child's Cervical Spine

Jonathan H. Phillips

#### **Embryology and Definitions**

The process of embryological development and maturation of the fetus can be described in various stages known as Carnegie stages. These refer to levels of development rather than gestational age or crown-rump length in millimeters. Though the three systems overlap, we will use the Carnegie stages as much as possible in this discussion.

The terms rostral and caudal and ventral and dorsal—while intuitive in embryology—are used less often in descriptive surgical anatomy, and the terms superior and inferior and anterior and posterior are used interchangeably in this chapter. In addition, descriptive names such as basiocciput, atlas, and axis are interchanged with skull, C1, and C2, which better describe the approach of the surgeon in the operating theater to ensure accurate surgical instrumentation at correct levels.

Metamerism is an important concept that relates to the general pattern of segmental repetition of similar structures in the developing embryo. It is this basic symmetrical template which is modified by localized gene expression to form region-specific structural changes which result in highly differentiated anatomical areas in vertebrates. Nowhere is this specialization more apparent than in the upper cervical spine of the human. The formation of the skull base and upper two cervical vertebrae is unique in the axial human skeleton and departs quite markedly from the lower cervical, thoracic, lumbar, and sacral morphology where there are more structural similarities than differences. We will see that the particular embryology of this rostral area of the spine has highly complex origins.

Segmentation describes a phenomenon of division of building blocks of tissues into repeating units and is similar in concept to metamerism. There is a further twist to the idea of segmentation in the human spine, however, because a process of resegmentation occurs during embryogenesis in which the caudal and rostral parts of adjacent segments fuse together to form the completed vertebrae. When this process is corrupted, the vertebrae are malformed. In the so-called hemimetameric shift, for instance, the process of resegmentation can fail unilaterally, resulting in the appearance of a hemivertebra and resulting in congenital scoliosis. This occurs most frequently in the thoracic spine, where coronal plane decompensation is an expected outcome for a fully segmented coronal hemivertebra, depending on the specific pattern of malformation. It occurs also in the cervical spine, both in the coronal and sagittal planes. Sagittal plane abnormalities are more common than coronal, the prototypical example of which is that seen in

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Klippel-Feil syndrome, a failure of segmentation rather than a hemimetameric shift, though this last can and does occur in the child's neck, resulting in cervical congenital scoliosis.

Somites, or more properly their derivatives, sclerotomes, are the building blocks of the spine. They appear in increasing numbers during embryogenesis, and the number of these segmental tissue blocks correlates with the anatomical staging of the embryo. Somites are just one part of the mesoderm layer of the three-layered early embryonic disc. This disc, a few days old, has an outer epidermal layer facing the amniotic cavity, a middle layer of mesoderm, and an endodermal layer facing the yolk sac. This pattern is apparent by about 3 weeks postfertilization. The mesoderm is itself divided into three parts, medial, intermediate, and lateral mesoderm. The most medial band is called the paraxial mesoderm and once again divides into three, this time from dorsal to ventral. The area nearest the dorsal surface is the dermatome, next the myotome, and further to the center of the embryo is the sclerotome. All of these areas are arrayed surrounding two structures which carry powerful molecular signaling properties-the notochord in the very center of the embryo and the neural tube which by now (stage 10 or about 4 weeks) has formed from the original neural plate and which lies right behind the notochord on its dorsal aspect. The notochord will regress quickly, but not before the ventral

cells of the somitic mesoderm have spread toward this structure, which induces the formation of the sclerotome. The sclerotomal segments (and this tissue mass is segmented) will form the vertebrae, whereas the notochord, under the negating influence of the neural tube, remains in the mature human only as the nucleus pulposus of intervertebral discs and the alar and apical ligaments of the craniocervical junction. This segmented system develops in a rostral to caudal direction (Fig. 1.1).

Somite count increases from about one to four at age 20 days, first appearing at the head of the embryo, to 34-35 at age 30 days toward the tail. Ultimately, 44 pairs of somites occur and form the left and right half of the sclerotome. The other two parts of the somites go on to form muscle and skin. The remaining parts of mesodermal layer lateral to the somites form splanchnic structures. These include gut, vascular, and urological structures. Insult to the embryo at this stage can affect all these systems and explains the concomitant appearances in clinical practice of multi-system congenital formation failure. The best known example of this is VACTERL syndrome in which heart, gut, renal, and vertebral malformations coexist.

At about the 5- to 8-week period, or Carnegie stages 15–22, the emerging pattern of spinal formation is becoming evident. However, the contribution of somites to their sclerotomal structures is



Fig. 1.1 The relationship and control of somatic mesoderm to the notochord and neural tube (Reproduced with permission from Gilbert [7]; © Sinauer Associates, Sunderland, MA)

highly complex at the cranial extent of the vertebral column. There are designated pairs of sclerotomes inasmuch as the upper four form the basiocciput, the next eight form the cervical vertebrae (of which there are only seven, but with eight spinal nerves), and the more caudal pattern (12 thoracic, five lumbar, and five sacral, variable coccygeal) is more easily understood based on the gross anatomy of the fully formed human skeleton. It is the complex variation from the typical pattern of vertebral development which gives rise to the unique shape and function of the atlas and axis. These two vertebrae share a common origin with the basiocciput, and as such should be considered as an embryological, anatomical, and functional unit very different from the subaxial spine. This unit is uniquely designed to transmit the termination of the brainstem and emerging spinal cord in a highly flexible protective tube that allows very roughly 50% of the total movement of the skull on the spinal column. The remaining cervical motion is distributed over the five lower cervical segments. All of these cervical vertebral segments except C7, however, carry the responsibility of transmitting the vertebral arteries, a function solely attributed to neck vertebrae. Once again the pattern of the vertebral arterial anatomy is radically different at the atlas and axis, and a thorough understanding of this arterial anatomy is fundamental to safe posterior surgical approaches to the upper cervical spine.

The relative somatic contributions to the spinal column are shown in Fig. 1.2. The upper four sclerotomes form the basiocciput but also borrow from somite five, which is a cervical one, thus the intimate relationship of the atlas to the skull base embryologically. Sclerotome five (a cervical one) forms both the posterior arch of the atlas and occipital condyles. The anterior arch of the atlas has an origin in the hypochordal bow which appears ventral to the notochord and undergoes chondrification and fusion with the posterior neural arch elements. There is a transient proatlas centrum which is dissolved. There is no vertebral body in C1 under normal circumstances. Teratogenic influences at this stage have been shown in mice. Interference with Hox genes by retinoic acid (most commonly used in the human for the treatment of acne) has been shown to



**Fig. 1.2** Relative somatic contributions to the spinal columns (Redrawn with permission from Muller and O'Rahilly, © 1994 Wiley Publishing)

cause caudal or rostral homeotic transformations [1]. The Hox-4.2 gene expression can transform occipital bones into neural arches [2]. Finally, transgenic mice can be found to exhibit a third occipital condyle fusing the skull base to the dens [3], and rostral vertebral shifts have been seen after heat exposure.

Though murine and avian genetic models should be interpreted with caution in the human, it is easy to imagine that altered expression of these homeobox genes may be the basis for wellknown malformations at the upper cervical spine such as assimilation of the atlas, which can occur posteriorly and anteriorly.

The formation of the axis is in many ways perhaps the most bizarre in the human axial skeleton. A review by Muller and O'Rahilly in 2003 explains the process well [4]. The fact that two, not one, sclerotomes form the posterior neural arch of C2 explains why it is so massive



**Fig. 1.3** Ossification centers of C2. *L* is the dentocentral synchondrosis; *J* is the neurocentral synchondrosis. There are two *Is*, two *Cs*, and one *A*, totaling five primary centers. *G* and *H* are secondary centers of ossification (Reproduced with permission from Bailey [8]; <sup>(C)</sup> The Radiological Society of North America)

(and therefore ideally suited to the placement of translaminar screws during cervical instrumentation). It also helps us to understand the sometimes confusing radiological appearance of the synchondroses of C2 in the immature child (Fig. 1.3), an important goal to achieve since these areas are often misinterpreted as fractures. Perhaps mutations of gene expression in this area can also explain the retroflexed dens seen in Chiari malformation and congenital types of basilar invagination.

The third cervical vertebra and its subjacent levels exhibit the so-called typical cervical morphology. As noted above, this is still distinguishable from thoracic and lumbar vertebrae but approximates more closely to the general pattern of vertebral development.

There are three primary ossification centers at C1. The anterior center, derived from the hypochordal bow, fuses with the posterior/dorsal elements of the neural arches at the neurocentral synchondrosis. This junctional area fuses completely around age 7. The spinous process uniting the left and right neural arch growth centers unites at age 3. Thus, radiographically there appears to be a spina bifida occulta present in the toddler, though usually the laminae meet at a complete cartilaginous bridge. The same appearance may be present in more caudal vertebrae also.

At C2 there is predictably a much more complex arrangement consequent upon its development from three sclerotomes. Five ossification centers appear and there are also two secondary centers (the tip of the dens and the ring apophysis of the inferior/caudal aspect of the body of the axis). These centers result in two radiographically significant synchondroses (see Chap. 4). The dentocentral synchondrosis represents the fusion of two sclerotomes at the level of the future body of C2. However the fusion level, though less distinct, may also be apparent in the young child, most commonly on CT scan or MRI reformatted in the sagittal plane. As mentioned above, this may be a source of concern in the injured child as a potential fracture line [5]. The possible relationship of these synchondroses to later formation of an os odontoideum is discussed elsewhere (see Chap. 4). The neurocentral synchondrosis represents the junction of two sclerotomes anteriorly (ventrally) with the left and right neural arches derived dorsally from the same tissue. There are therefore two of these junctions left and right, and they are best seen in coronal imaging modalities. The growth centers of C1 and C2 are represented graphically in Figs. 1.3, 1.4, and 1.5. These describe the prototypical arrangements, but it must be emphasized that many anatomical variations can occur, which may be confusing on imaging studies of the young child. This point is well made by Karwacki and Schneider in their 2012 analysis of atlas and axis growth center variability based on 550 CT scans of children aged 0–17 years [6].

Growth centers are present at various stages in the human embryo. Initially seen as chondrification centers, they become ossified and visible radiographically by birth and early childhood, though the adult pattern is not achieved until final vertebral physeal closure in the 20s.

#### **Muscles of the Neck**

The musculature of the neck has a complex arrangement predicated on the function of high mobility of the skull on the spinal column. The most superficial muscle posteriorly is the trapezius. This huge triangular muscle arises from the superior nuchal line of the skull all the way to the spinous process of T12. Its lateral attachment is on the spine of the scapula. Thus it is, strictly speaking, a muscle of the upper limb. The true deep muscles of the neck lie deep to trapezius and comprise five groups. The groups are splenius, erector spinae, transversospinal, interspinal, and intertransverse muscles.

Splenius covers the deeper muscles of the back of the neck and has capitis and cervicis divisions (Fig. 1.6). The splenius capitis and cervicis

arise from the ligamentum nuchae and spinous processes C7 to T6. Cervicis inserts into the posterior tubercles of the upper two or three cervical vertebrae, and capitis has a more proximal insertion on the mastoid process and the lateral part of the superior nuchal line. Contraction of the splenius rotates the head toward the side of the muscle acting, and bilateral contraction extends the head and neck. Innervation is from dorsal rami of C2 to C6. Deep to it lie erector spinae and semispinalis. Anteriorly the sternocleidomastoid inserts more superficially to the capitis division at the mastoid. This last muscle arises from both clavicle and sternum and opposes splenius rotating the head to the opposite side of the muscle contracting, flexes, and laterally flexes the neck.

The erector spinae muscle group is represented in the neck as iliocostalis cervicis, longissimus cervicis, and spinalis cervicis and capitis—in other words, three subgroups (Fig. 1.7). Iliocostalis is lateral; spinalis medial and longissimus are in between the other two. The muscles lie in the costovertebral groove.



Fig. 1.4 Ossification centers of C1. Not unusually, the body center is bipartite and occasionally occurs in three or other multiple parts (Reproduced with permission from Bailey [8]; © The Radiological Society of North America)



**Fig. 1.5** Primary (stippled) and secondary (striped) ossification centers of the typical cervical vertebra. There are three. Note the ring apophyses (*G*) at superior and inferior parts of the body. These fuse late in life, sometimes in the early 20s (Reproduced with permission from Bailey [8]; © The Radiological Society of North America)

Fig. 1.6 Dorsal neck muscles, superficial layer

Iliocostalis cervicis arises form upper ribs and inserts onto transverse processes of the lower cervical vertebrae. Longissimus cervicis arises from the uppermost ribs and inserts into the C2 to C6 transverse processes. Spinalis cervicis is a variable muscle often not well defined. The erector spinae muscles laterally flex and extend the neck.

Of the transversospinal group, one muscle is important and forms the largest single muscle of the posterior neck. It is the semispinalis capitis and arises from transverse processes of the upper thoracic and seventh cervical vertebrae and the articular processes of C6 to C4 (Fig. 1.7). It inserts onto the undersurface of the skull base posteriorly and is a powerful extensor of the neck. Semispinalis cervicis is contiguous with its thoracis component and passes from thoracic transverse processes to spinous processes several levels higher, ultimately reaching the posterior axis.

Interspinal and intertransverse are small segmental muscles represented by such groups as multifidus and rotatores arising from transverse processes of adjacent vertebrae. All are segmentally innervated and perform functions of local stabilization and small rotations at segmental levels.

At the base of the skull lies a unique triangular arrangement of muscles which form the suboccipital triangle (Fig. 1.8). These suboccipital







Fig. 1.8 Suboccipital musculature

muscles are part of the transversospinal group. The four muscles are rectus capitis posterior major and minor and superior and inferior oblique muscles of the head, as seen in Figs. 1.2 and 1.5. In the floor of this triangle lies the posterior atlanto-occipital membrane deep to which the vertebral artery passes of the arch of the atlas into the foramen magnum. The suboccipital (C1) and greater occipital nerve (C2) arise, respectively, above and below the posterior arch of the atlas. The C2 root overlies the lateral mass of the atlas and can obstruct the placement of a screw in this

structure during posterior C1 instrumentation (Fig. 1.9). Occasionally the nerve must be sacrificed for this reason. A prolific venous plexus also lies in this region and can cause troublesome bleeding during C1 lateral mass instrumentation.

The vertebral artery arises as the first branch of the subclavian. It passes upward in the posterior part of the pyramidal space above the apex of the lung. It enters the cervical spine through the foramen transversarium of C6, not C7, and ascends to C2 where it passes backward then medially and then forward in a wide loop that



Fig. 1.10 Arteries of the anterior and lateral neck

allows for movement between atlas and axis (Fig. 1.10). As it passes anteriorly toward the foramen magnum, it leaves a groove on the superior surface of the atlas. It is highly vulnerable in this area to damage during surgical exposure of the occipital and atlantoaxial region. Its position lateral to the midline plane effectively limits the lateral dissection in surgery of this area.

Accompanying the vertebral artery is the vertebral vein or more properly a plexus of veins which pass both inside and outside of the foramina transversaria. One branch exits at C6 and another at C7 transverse foramen, and both drain into the subclavian vein.

Anterior vascular anatomy dictates the surgical approach to the anterior cervical spine (see Chap. 18). The carotid sheath contains the common carotid artery, the internal jugular vein, and the vagus nerve. It extends from the base of the skull to the aortic arch and is tightly opposed to the posterior surface of the sternocleidomastoid muscle above the sternoclavicular joint. At C3 level the common carotid bifurcates. The internal carotid has no branches in the neck and passes up into the skull through the carotid foramen just anterior to the jugular foramen, which itself contains the internal jugular vein and lies just deep to the external acoustic meatus. The carotid sheath and its contents, lying deep to the anterior border of sternocleidomastoid, form the posterior border of the anterior surgical approach to the mid cervical spine. Anteriorly the esophagus and trachea are retracted laterally to allow exposure of the anterior cervical vertebral bodies, and their discs thus form the anterior border of this exposure (see Chap. 14, 18).

#### **Cervical Osteology**

The atlas C1 is a gracile, almost circular, ring of bone with articular facets above for the occipital condyles and below for the axis (Fig. 1.11a, b). The neural arches are massively enlarged to form the lateral masses, which constitute the only substantial bony elements allowing surgical screw purchase. Their axes pass from posterior lateral to anterior medial. Above are the deeply concave kidney-shaped articular facets for the occipital condyles; below are the more circular facets for articulation with the axis. Lateral to the masses lies the foramen transversarium, formed from both neural and costal elements. Anteriorly on the arch of the atlas is a tubercle to which the anterior longitudinal ligament attaches. There is no centrum; the proatlas has dissolved. In addition, the anterior arch of the atlas is formed not from the centrum remnant as would be imagined, but rather from the hypochordal bow. This structure is important in cervical spine embryology, but exists elsewhere only as the ligamentous fascicle running deep to the anterior longitudinal ligament joining two rib heads. Thus the hypochordal bow of the atlas is the ossified ligament joining its two costal elements. It often shows failure of complete ossification in the child, as does the posterior arch of the atlas, which is more conventionally formed from neural arch elements. The course of the vertebral artery over the superior surface of the posterior arch has been described. One other anomaly is of interest. The articular elements of the upper two cervical vertebrae are in series with the tiny synovial uncovertebral joints on the lateral aspects of the subaxial cervical vertebrae and not their more massive and functional cervical articular facets aligning more posteriorly from C3 to C7. Thus the first and second cervical nerves send their anterior primary rami behind the joints and not in front as the lower vertebrae do. The resulting obstruction to surgical approaches to C1 has been mentioned.

The axis C2 has much more massive proportions than its cephalad neighbor (Fig. 1.12a, b). We have seen that it is derived from a larger number of sclerotomes and not only has retained, but also co-opted, a greater centrum contribution embryologically. Its characteristic features are the upward pointing dens which articulates with the posterior surface of the anterior arch of C1,



Fig. 1.11 (A, B) First cervical vertebra (atlas). (a) Cranial and (b) caudal



Fig. 1.12 (A, B) Second cervical vertebra (axis). (a) Cranial and (b) anterior



Fig. 1.13 (A, B) Typical cervical vertebrae showing a body, neural arch, spinous, and complex lateral processes

the large lateral masses, and the huge spinous process, which even in small children may be big enough to allow passage of translaminar screws. In contrast to the atlas, it may have discrete pedicles, though their size in children is variable and often does not allow for accommodation of a true pedicle screw. Alternative techniques of C1-C2 arthrodesis, such as a Magerl screw, are discussed in later chapters of this text. In addition, sublaminar wiring techniques have a long history in orthopedics and utilize the posterior arch of C1 and the lamina or spinous process of C2. Again, see later chapters.

The orientation of the articulations between occiput and atlas and atlas and axis allows for a very large range of motion, nodding at the former and rotation at the latter. Approximately 50% of rotation is lost by atlantoaxial arthrodesis, but the effect on atlanto-occipital fusion is less obvious because of the large flexion extension range of the subaxial spine.

From C3 to C7 the vertebral morphology is more typical and reproducible. Usually the costal elements are limited to the contribution to the foramen transversarium, but occasionally true cervical ribs appear at C7 or are represented by sometimes troublesome fibrous bands, causing thoracic outlet symptoms. In the case of properly formed cervical ribs, the brachial plexus may exhibit precession arising from C4 to C8 instead of C5 to T1.

Typical cervical vertebrae show a body, neural arch, spinous, and complex transverse processes which contain the vertebral artery and its veins (Fig. 1.13a, b). The lateral part of the body at the interface with the intervertebral disc shows an upward turn into the uncus, and it is here that tiny uncovertebral synovial joints exist. They limit lateral flexion and, described by Lushka, only exist in the cervical spine. Pedicles are much better formed in the typical cervical vertebrae and allow screw instrumentation. In addition, the greatly broadened lateral masses, which exhibit superior and inferior articulations that sandwich the bony masses, allow for strong screw fixation, again explained later in this text. The choice of lateral mass or medial pedicle trajectory is predicated on the position of the vertebral artery which lies directly anterior to the lateral masses and thus precludes a straight anteriorward approach in surgical fusions. However, purchase under the substantial laminae of C3 to C6 is available, if not as stable in fusion constructs. At C7 the pedicle is usually so well formed, even in children, that it has become the preferred location for spinal instrumentation at this level. Between C3 and C6, careful analysis of the specific anatomy with advanced imaging is mandatory for safe placement of instrumentation, indeed all levels should be examined with CT preoperatively, and this idea is discussed in Chap. 4.

#### Ligamentous Support of the Child's Cervical Spine

In the lower cervical spine, a familiar pattern is found. The anterior longitudinal ligament supporting the anterior vertebral bodies, with intervertebral disc annulus and posterior longitudinal ligament at the posterior margin of discs and bodies, is very similar to the thoracic and lumbar pattern. At C2 and above a very different construct exists, uniquely suited to the previously mentioned function of allowing large degrees of motion between the head and neck. The cruciform ligament joins the posterior body of the dens to the base of the occiput at the anterior margin of the foramen magnum, bypassing the atlas. Its strong transverse ligament component captures the dens axis against the posterior part of the anterior arch of the atlas, where a synovial joint and significant bursa exists. The apical ligament is the continuation of the cruciform into the skull. Inferiorly the stem of the "cross" is adherent to the inferior posterior body of the axis below the dens. One additional and important connective tissue structure also adds stability to the skull-to-atlas-to-axis complex. The tectorial membrane passes from the posterior rim of the anterior lip of the foramen magnum to the posterior axis body. It is continuous with the posterior longitudinal ligament and blends with the dura on its deep or posterior surface. It can be imaged with MRI and if ruptured in this modality implies instability at the occipitoatlantal level (Figs. 1.14 and 1.15).



Fig. 1.15 Axial view showing transverse, alar, atlantodental ligaments



Fig. 1.14 Occipito-cervical level, showing ligaments and membranes, sagittal view

#### Summary

The development of the human neck shows unique aspects of specialization to fulfill the functions of great flexibility and range of motion not seen elsewhere in the spinal column. The uppermost two vertebrae show major departures from the pattern seen subaxially. A common embryological origin of the skull base and parts of the atlas and axis explains their unique shape and the fact that these parts function as a unit.

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# Biomechanics of the Growing Cervical Spine

2

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#### Normal Cervical Spine Biomechanics

The cervical spine functions to provide motion of the head atop the axial skeleton and to protect the neural elements of the spine as they traverse the neck. The cervical spine can be divided into two distinct segments, the occipitocervical junction extending from the occiput to C2 and the subaxial cervical spine from C3 to C7, each having distinct anatomic and biomechanical features. Together, they provide several degrees of motion for the head on the axial skeleton, including flexion, extension, and lateral rotation and bending to the right and left; distraction and compression are theoretical and not desirable. The normal active range of motion of the child's cervical spine is slightly greater than that of an adult, with average values of 60° of flexion, 90° of extension, 45° of lateral bending in each direction, and 70° of axial rotation in each direction [1, 2]. Reasons for this include maturing osseous structures and increased ligamentous laxity in children, which will be discussed further throughout the chapter. Because of

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B. Yaszay (⊠) Children's Specialty, San Diego, CA, USA e-mail: byaszay@rchsd.org their distinct differences, the two regions of the cervical spine are discussed separately. A thorough discussion of the anatomy and embryology of the growing cervical spine can be found in Chap. 1, but here we discuss the osseous and ligamentous structures of the cervical spine as they relate to its kinematics and stability.

# Functional Anatomy and Normal Biomechanics

#### **Occiput to C2**

#### **Osseous Anatomy**

The craniocervical junction is made up of the base of the occiput, C1 (the atlas), and C2 (the axis) that function as a unit to control head movement on the subaxial spine. The primary motion of the occipitoatlantal joint is flexion and extension, with the atlantoaxial joint contributing primarily axial rotation. The underside of the occipital bone includes the foramen magnum, through which the spinal cord passes into the cervical spine. The anterior midline of the occiput is known as the basion, and the posterior margin is known as the opisthion. The transverse diameter of the foramen magnum is slightly less than the anterior posterior diameter. On the lateral side, just anterior to the midline are the occipital condyles, which are convex in the sagittal plane but oblique and rest on the concave lateral mass of

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C1, or the atlas, allowing for flexion and extension at the O–C1 articulation. In the coronal plane, the articulation is angled slightly medially, allowing for a small amount of lateral bending while resisting lateral translation.

The atlas is a ring-shaped bone, lacking the vertebral body and spinous process of other vertebrae and acting as a dished washer between the occiput and C2. The two thick lateral masses (which are the morphologic corollary to the transverse processes in the sub-cervical spine), act as the articulating surfaces of C1. The superior surface articulates with the occipital condyles as previously mentioned and the inferior surface with C2. The inferior facets are relatively flat, with a slight convexity and lateral tilt to allow axial rotation between C1 and C2. This rotation occurs around the odontoid process, which is a cephalad projection of the body of C2 and is a significant stabilizer of the C1-2 articulation, as discussed below. The body of C2 is larger than that of C1 and is connected on each side by a neural arch that includes an inferior and superior facet. The superior facets sit lateral and just posterior to the dens, are slightly concave, and receive the convex inferior facets of C1. The lateral tilt limits lateral translation while allowing significant amounts of rotation. The inferior facet of C2 sits posteriorly on the neural arch and has an orientation similar to the subaxial articular facets of the cervical spine.

#### **Ligamentous Anatomy**

The limited articular and osseous constraints of the craniocervical junction place significant importance on the ligamentous structures to provide stability while still allowing for a very extensive range of motion. The tectorial membrane is a cranial continuation of the posterior longitudinal ligament that travels posterior to the body of C2 and anchors to the base of the skull at the anterior rim of the foramen magnum (see Chap. 1). It controls extension by becoming taught when the head is extended and limits flexion at C1/2 when it is tightened as the skull tilts anteriorly on C1 [3]. A recent investigation argued that the tectorial membrane may act less as a true stabilizing structure and more as a reinforcement to prevent impingement of the odontoid on the neural elements, which secondarily stabilizes the occipitocervical junction [4]. The alar ligaments extend from the dorsolateral surface of the dens to the medial aspect of the occipital condyles, each one limiting lateral rotation to contralateral side. They also act as a check ligament to limit the amount of axial rotation between C1 and C2, further discussed below. The cruciate ligament consists of a transverse and ascending/descending portion. The transverse ligament is the thickest and most important portion and connects between the two condyles of C1, stabilizing the dens between them. The ascending/descending portion extends from the anterior edge of the foramen magnum to the body of C2. The apical ligament, the atlantodental ligament, and the anterior and posterior atlantooccipital membrane are biomechanically insignificant [5, 6].

#### **Kinematics**

The occipitocervical complex provides approximately 40–50% of flexion and extension and 60% of axial rotation of the cervical spine. Much less lateral bending is allowed at these two articulations, most of which comes from the lower cervical spine. The primary motion between the occiput and C1 is flexion and extension, contributing approximately 25° total [7]. The cupshaped articulation limits rotation with reports ranging from 0° to 8° [7–9] and lateral bending ranging from 2° to 8° [7, 10, 11].

Axial rotation is the primary motion of the C1–C2 articulation, with up to 65° of motion in one direction in adults [5]. The joint also contributes approximately 20° of flexion and extension and, similar to the occipitoatlantal joint, contributes only approximately 5° of lateral bending [7, 10]. In their attempts to further understand lateral axial rotatory subluxation in children, Pang and Li have performed a thorough CT evaluation of the kinematics of the C1-2 articulation in normal children. In the early phase of lateral rotation, C1 moves with the head, while C2 is left stationary, a phase which they termed the single motion phase. At approximately 23° of rotation, the alar ligaments begin to tighten and C2 rotates with C1, but at a different rate, termed the double