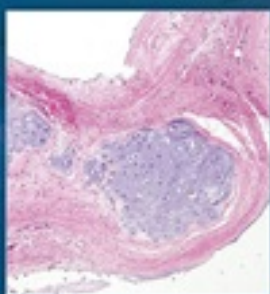
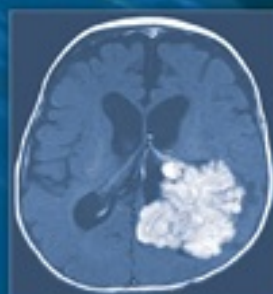


Pediatric Radiology

Practical Imaging Evaluation of
Infants and Children



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PEDIATRIC RADIOLOGY

Practical Imaging Evaluation of Infants and Children

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9 8 7 6 5 4 3 2 1

Printed in China

Library of Congress Cataloging-in-Publication Data

Names: Lee, Edward Y., editor.

Title: Pediatric radiology : practical imaging evaluation of infants and children / editor, Edward Y. Lee ; associate editors, Winnie C. Chu, Jonathan R. Dillman, Andrea S. Doria, Ricardo Restrepo, Sara O. Vargas.

Other titles: Pediatric radiology (Lee)

Description: Philadelphia : Wolters Kluwer, [2018] | Includes bibliographical references.

Identifiers: LCCN 2016039527 | eISBN 9781496380289

Subjects: | MESH: Radiography | Infant | Child

Classification: LCC RJ51.R3 | NLM WN 240 | DDC 618.92/00757—dc23 LC record available at <https://lccn.loc.gov/2016039527>

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To my parents, Kwan-Pyo and Kang-ja, and my family for their constant support and encouragement.

To the trainees and radiologists that I have met during my academic career for sharing their enthusiasm for learning.

To my colleagues in the Department of Radiology, Boston Children's Hospital, for their support and friendship. There is absolutely no better group to be with.

—Edward Y. Lee, MD, MPH

To my family, Allen, Cherie, and Paco for their love and support.

To Faculty of Medicine at the Chinese University of Hong Kong for nurturing me as a medical student, physician, radiologist, teacher, and researcher.

—Winnie C. Chu, MD, FRCR

To the children that I have had the honor to care for.

To my wife, Suzanne, and our three beautiful daughters, Kathryn, Meghan, and Claire.

—Jonathan R. Dillman, MD, MSc

To my father who has been my great mentor, advisor, and friend throughout my life.

To my mother who taught me beautiful lessons of love during the short period of time that I had the privilege to live by her side.

To my husband, son, and extended family accrued throughout the pathway of life, who have taught

me values of loyalty, honesty, companionship, and true love.

To Danny Aguilar for providing invaluable illustrations in musculoskeletal chapters.

To my patients whose journeys and battles have been the inspiration of my life.

—Andrea S. Doria, MD, PhD, MSc

To my parents, Jairo and Helena, for providing the inspiration that guided me throughout my career and also for being such incredible physician role models.

To LBS for his support and for constantly reminding me that life is beautiful and to be enjoyed in all aspects.

—Ricardo Restrepo, MD

To my colleagues in pathology, whose great care with their specimens over many years led to the diagnoses depicted herein.

To the young patients of Boston Children's Hospital, whose material enlightens us.

—Sara O. Vargas, MD

The editor and associate editors of this book would also acknowledge that all pathology images, unless otherwise specified, were provided by Sara O. Vargas, MD. Danny Aguilar, medical graphic artist and illustrator, provided schematics for musculoskeletal chapters in this book.

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PREFACE

Today, imaging evaluation is an integral component in the daily practice of managing infants and children with a variety of underlying congenital and acquired disorders. Pediatric radiology, unlike other radiology subspecialties, encompasses multiple organ systems and modalities and demands a solid foundation in the imaging of pediatric disease. To this end, the purpose of this book is to provide a comprehensive imaging review of pediatric disorders that are encountered in daily clinical practice.

The idea for this book arose from my interactions with residents, fellows, and faculty members of various specialties that I have encountered as a pediatric radiologist at Boston Children's Hospital, chair of the pediatric radiology section of the Core Examination Committee at American Board of Radiology (ABR), and visiting professor to more than 30 different countries around the world for the past 10 years. Everyone was looking for an up-to-date single volume resource for learning and reviewing the fundamentals and essentials of pediatric radiology. Such a textbook should be practical, covering both common and rare but significant pediatric disorders, while providing international perspectives to broaden its use in any part of the world. However, there was no such book available.

From this came my desire to write a pediatric radiology textbook, which increased every time I felt incomplete after giving pediatric radiology lectures to trainees and practicing radiologists, particularly during the international visiting professorships. It became clear to me that providing essential information about pediatric radiology can be best achieved by creating and organizing the material into a textbook that can reach people in every part of the world. The process of creating this textbook started with formulating an outline with five truly gifted radiology and pathology associate editors on topics that are important for the care of pediatric patients in daily clinical practice. This was later enhanced with exemplary cases that we encountered during our clinical work as well as cases provided by approximately 100 outstanding contributing authors from 40

different countries around the world. The authors who have contributed chapters to this book are recognized experts in their fields from six continents in the world, making this book truly international, current, and comprehensive, providing the most up-to-date information available. Also, many “textbook” images included in this book were given to me by “unknown” radiologists across the world that I have met during my international trips who wanted to share their cases and experience with others. Furthermore, relevant pathology of pediatric disorders is selectively included in order to enhance the understanding of the underlying disease process and radiologic imaging findings.

This book is organized into four main sections based on organ systems, which include pediatric neuroradiology, pediatric thoracic radiology, pediatric abdominopelvic radiology, and pediatric musculoskeletal radiology. The organization and presentation of this book is structured to provide accessibility to both common and less common but important pediatric disorders. Each chapter includes a discussion of practical imaging techniques, normal anatomy and variants, imaging findings, and selected pathologic features of congenital and acquired disorders in a specific section. The discussion of each disorder includes the clinical features, imaging findings, pathologic correlation in some selected cases, and up-to-date management information. Given its focus on disorders affecting the pediatric population, we have emphasized how to differentiate between the normal variants and abnormal pathology and to determine whether a certain radiologic finding is isolated or whether it is related to a genetic or malformation syndrome. In addition, when relevant and possible, differential diagnostic considerations and differentiating features, both clinical and radiologic, between disorders that produce similar imaging findings are also addressed. Every effort was made to cover the broad scope of pediatric radiology practice in a single-volume book without redundancy.

This book is intended primarily for radiology residents, radiology fellows, and practicing radiologists. However, other physicians, such as pediatricians and pediatric specialists, who frequently or occasionally encounter the pediatric patient for diagnostic imaging studies may derive valuable clinical, imaging, and some pathologic information that can be used to manage their pediatric patients.

It is my hope that readers, regardless of level of knowledge, training, and fields of specialties, will be more stimulated and interested in pediatric disorders and imaging after reading this book. If this book helps the readers to better understand the imaging evaluation of infants and children, leading to improvement in care and efficient management of pediatric patients, we will

have accomplished our overarching goal. Behind all images that were included in this book, there were living and breathing pediatric patients who were often very sick and close to the end of their promised time in this world. We must recognize that we have an invaluable opportunity to make a true difference in their lives and the lives of their loved ones.

This book has been a journey, not a destination, for me and the associate editors. We invite the readers to join us in this journey of lifelong learning, and we look forward to feedback from readers about what can be improved and added in the next edition. Until then, we hope all the readers will enjoy and learn from this book as much as we did from preparing and writing it.

Edward Y. Lee

Contents

Cover

TitlePage

Copyright

Dedication

Editors

Contributors

Preface

PART I PEDIATRIC NEURORADIOLOGY

Section Editor: *Winnie C. Chu*

1 Skull

Edward Yang, Sara O. Vargas, and Tina Young Poussaint

2 Brain

Sanjay P. Prabhu, Savvas Andronikou, Sara O. Vargas, and Richard L. Robertson

3 Head and Neck

Amy F. Juliano, Sara O. Vargas, and Caroline D. Robson

4 Spinal Cord

Benjamin T. Haverkamp, Peter Wittingham, Winnie C. Chu, Lisa H. Hutchison, and Paul G. Thacker

5 Vertebral Column

Esperanza Pacheco-Jacome, Kevin R. Moore, Sara O. Vargas, and L. Santiago Medina

PART II PEDIATRIC THORACIC RADIOLOGY

Section Editor: *Edward Y. Lee*

6 Lung

Bernard F. Laya, Behrang Amini, Evan J. Zucker, Tracy Kilborn, Sara O. Vargas, and Edward Y. Lee

7 Pleura

Rama S. Ayyala, Shunsuke Nosaka, Khalid Khashoggi, Janina M. Patsch, Zaleha Abdul Manaf, and Edward Y. Lee

8 Airway

Evan J. Zucker, Supika Kritsaneepaiboon, Omolola M. Atalabi, Ricardo Restrepo, Yumin Zhong, Sally A. Vogel, and Edward Y. Lee

9 Heart

Lorna P. Browne, Edward Y. Lee, Oleksandr Kondrachuk, Marielle V. Fortier, Zhu Ming, and Cynthia K. Rigsby

10 Great Vessels

Monica Epelman, Pilar Garcia-Pena, Eric J. Chong Barboza, Magdalena Gormsen, Fatma Hamza Makame, and Edward Y. Lee

11 Mediastinum

Paul G. Thacker, Kushajit S. Sodhi, I. Nimala A. Gooneratne, Claudio Fonda, Pierluigi Ciet, and Edward Y. Lee

12 Chest Wall

Dawn R. Engelkemier, Peter G. Kruk, John Naheedy, Yeun-Chung Chang, Pilar Dies-Suarez, and Edward Y. Lee

13 Diaphragm

Mark C. Liszewski, Pedro Daltro, Celia Ferrari, Gloria Soto Giordani, Fred E. Avni, and Edward Y. Lee

PART III PEDIATRIC ABDOMINOPELVIC RADIOLOGY

Section Editor: *Jonathan R. Dillman*

14 Liver, Bile Ducts, and Gallbladder

Andrew T. Trout, Daniel B. Wallihan, Alexander J. Towbin, and Daniel J. Podberesky

15 Spleen, Pancreas, and Adrenal Glands

Ethan A. Smith, Jonathan R. Dillman, Sara O. Vargas, and Peter J. Strouse

16 Gastrointestinal Tract

Sudha A. Anupindi, Andria M. Powers, Suma Kannabiran, Jonathan R. Dillman, Michael S. Gee, and Asef Khwaja

17 Kidneys and Urinary Tract

Jonathan R. Dillman and Kassa Darge

18 Male Genital Tract

Andrew Phelps, Jesse Courtier, Peter "Buzz" Marcovici, Sara O. Vargas, and John D. MacKenzie

19 Female Genital Tract

Sharon W. Gould, Sabah Servaes, Edward Y. Lee, José Ernesto Lipsich, Mohamed Issa Tawil, Sara O. Vargas, and Monica Epelman

20 Abdominal Wall, Mesentery, Peritoneum, and Vessels

Michael S. Gee, Rahul A. Sheth, Salwa M. Haidar, Dilip Sankhla, and Edward Y. Lee

PART IV PEDIATRIC MUSCULOSKELETAL RADIOLOGY

Section Editor: *Andrea S. Doria*

21 Normal Growth, Normal Development, and Congenital Disorders

Victor Ho-Fung, Adji Saptogino, Timothy Cain, Karuna M. Das, Selim Doganay, and Diego Jaramillo

22 Musculoskeletal Infectious and Inflammatory Disorders

Clara L. Ortiz-Neira, Jennifer Stimec, Marcia Torre Moreira, and Andrea S. Doria

23 Musculoskeletal Neoplastic Disorders

Hee-Kyung Kim, Jung-Eun Cheon, Sara O. Vargas, and Hye-Kyung Yoon

24 Musculoskeletal Traumatic Disorders

Mark E. Bittman, Jeannette M. Pérez-Rosselló, Donald A. Tracy, Abdusamea Shabani, and Edward Y. Lee

25 Musculoskeletal Disorders Due to Endocrinopathy, Metabolic Derangement, and Arthropathy

Ricardo Restrepo, Edward Y. Lee, Paul S. Babyn, Hadeel M. Seif El Dein, Bjorn Lundin, and Andrea S. Doria

Index

PART I

PEDIATRIC NEURORADIOLOGY

Winnie C. Chu

CHAPTER

1

Skull

Edward Yang • Sara O. Vargas • Tina Young Poussaint

INTRODUCTION

Suspected skull disorders are common indications for imaging in the pediatric age group, being easily identified by caregivers and physicians because of their superficial location. As an osseous structure, the skull manifests many of the pathologies seen elsewhere in the skeletal system, but it does so in the setting of dynamic changes of development and maturation of the skull itself as well as the brain that it protects.

This chapter reviews the radiographic anatomy and disorders affecting the skull as well as its connective tissue covering, the scalp, in the pediatric population. The chapter begins by considering the various modalities for performing an imaging examination of the pediatric skull. Next, normal imaging anatomy and skull development are discussed. Then, an overview is provided of the most commonly encountered diseases of the pediatric skull, including disorders of skull shape and integrity, infectious disorders, diffuse bone diseases, neoplasms affecting the skull and scalp, and features of skull trauma unique to the pediatric population.

IMAGING TECHNIQUES

Radiography

Radiography is the oldest imaging technique available for evaluating the skull and remains widely used because of its low cost, wide availability, and suitability in unsedated patients. In addition, it is associated with relatively low radiation dose compared to computed tomography (CT). Current indications include detection of diffuse bone abnormalities as part of a skeletal survey (e.g., metastatic disease, metabolic bone disease), screening for nonaccidental trauma (child abuse) in a neurologically intact child <2 years, and screening for craniosynostosis. Skull radiographs are also commonly used to assess surgical implants (e.g., shunts, programmable shunt valves, and cochlear implants) and to exclude the presence of radiopaque foreign bodies.

Standard skull radiography technique consists of frontal (usually posteroanterior [PA]), lateral, and Townes projections (Fig. 1.1). Whereas the petrous apices project over the central orbits in the PA projection, they are above the orbits in the Townes view because of angulation used for better visualization of the occipital bone and foramen magnum. An optional view opening up the frontal bone is the Caldwell projection where the petrous apices lie below the orbits. An age-specific dose is delivered with collimation to cover the osseous structures and overlying scalp. Typical delivered skin dose is ~1 mGy, and there is an estimated effective dose of ~0.02 mSv under these conditions.¹

Ultrasound

Sound waves penetrate osseous structures poorly and therefore ultrasound is not a widely used imaging modality for the skull in most institutions. However, the availability of ultrasound at the point of care and the lack of radiation exposure have prompted the use of ultrasound for selected indications in young (<2 years old) patients who have little hair and relatively sonolucent osseous structures. Specifically, ultrasound has been used to successfully detect skull fractures and sutural synostosis.²⁻⁵ Additionally, ultrasound has long been a first-line tool for evaluating scalp masses.^{6,7}

Ultrasound of the skull uses high-frequency (typically >8 MHz) linear transducers coupled with transducing gel. Adjustments are performed to reduce the depth and focus to accommodate the superficial structures being imaged. Typically, stand-off pads are not required. For soft tissue or vascular scalp lesions, color and duplex Doppler are added to demonstrate lesional vascularity and pulsatility of blood flow (i.e., arterial versus venous).

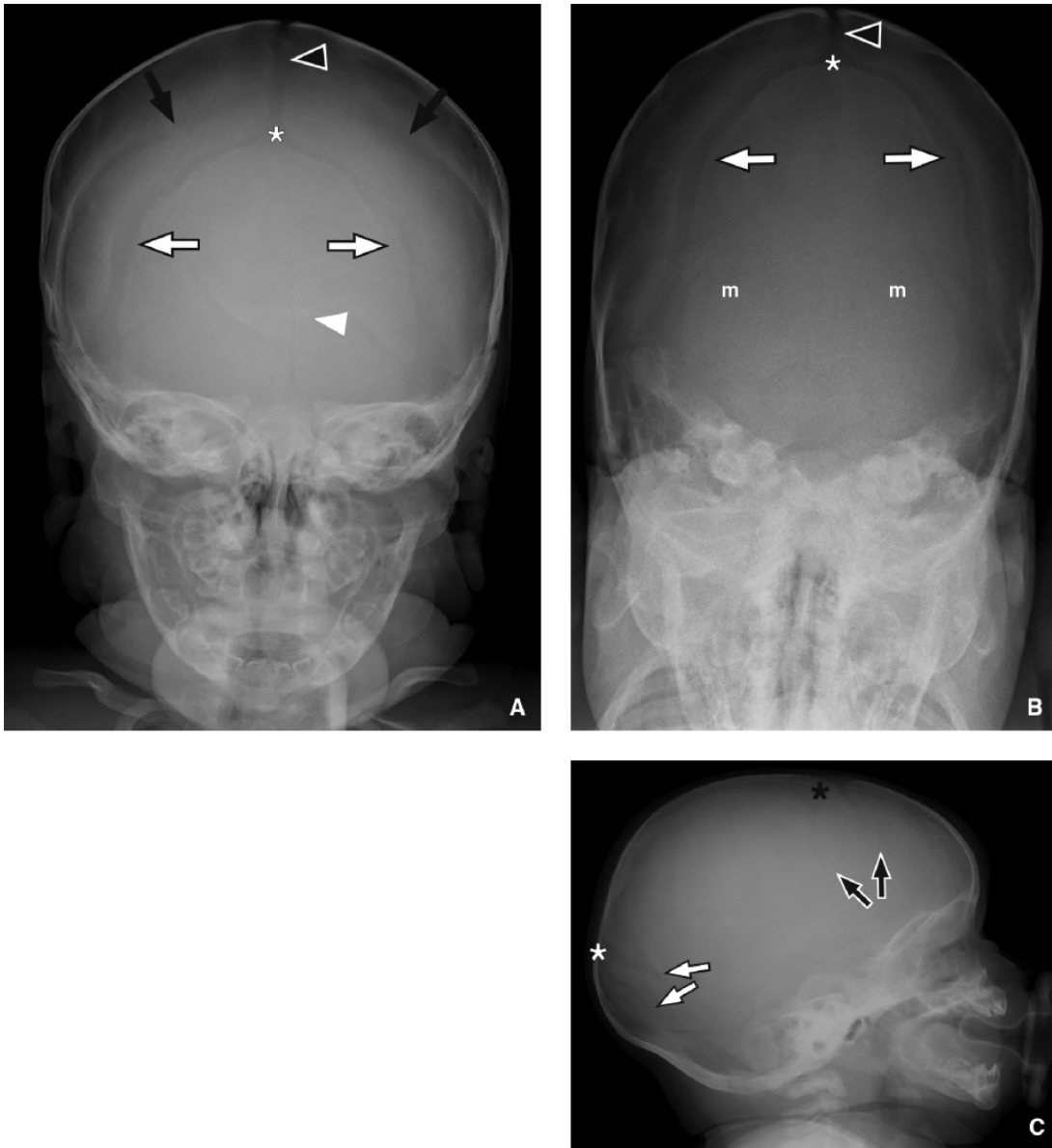


FIGURE 1.1 Skull radiographs of a normal 3-month-old boy: posteroanterior (PA) (A), Townes (B), and lateral (C) projections. *Black arrows* indicate coronal sutures, *black arrowhead* the sagittal suture, *white arrows* the lambdoid sutures, *white arrowhead* the metopic suture, *white asterisk* the posterior fontanelle, *black asterisk* the anterior fontanelle, and “*m*” the mendosal sutures.

Computed Tomography

As a high-resolution, cross-sectional technique with excellent osseous detail and soft tissue contrast, computed tomography (CT) is the current reference standard for indications such as head trauma, craniosynostosis, and characterization of focal skull lesions. Although some authors have proposed algorithms that omit

CT for single-suture synostosis,⁸⁻¹⁰ CT has superior sensitivity/specificity for detecting synostosis as it can detect even small areas of osseous bridging that may elude radiography.¹¹⁻¹³ There are also data to suggest that CT becomes increasingly cost-effective for individuals with a high pretest likelihood of synostosis, particularly involving multiple sutures. Regardless of diagnostic strategy for abnormal head shape, common clinical practice is to obtain a head CT with three-dimensional (3D) surface rendering for cases that are scheduled for surgery as the surface model assists procedural planning.^{11,14} For reasons discussed later (see “Fractures”), the American College of Radiology Appropriateness Criteria endorses an approach that integrates head CT when imaging is pursued for abusive or nonabusive head trauma.^{15,16}

Acquisition of a head CT depends on both the precise hardware available as well as the indication. For most modern multidetector CT instruments, axial acquisition of the head is followed by generation of both submillimeter (isotropic) and 3- to 5-mm-thick data using soft tissue (brain) and bone kernels. The submillimeter data can then be used to create multiplanar reformatted images and surface rendered data, the latter particularly helpful in delineating complex 3D relationships of skull lesions. In our institution, an age-specific dosage scheme¹⁷ compliant with recommendations from both the Society for Pediatric Radiology Image Gently campaign and the ACR CT Accreditation Requirements is used,^{18,19} the latter limiting the CT dose index (CTDI_{vol}) for a 1-year-old to 40 mGy compared to 80 mGy for an adult. For pediatric patients undergoing evaluation for craniofacial dysmorphism, the field of view is extended below the mandible to capture any facial bone abnormalities, and the dose is reduced to 100 kV and 50 mAs, resulting in a CTDI <5 mGy. The trade-off for this reduced dose is poorer parenchymal detail. Even with these attempts to reduce dose, it is worth noting that a standard head CT still carries an estimated dose that is 20 times a single projection skull radiograph.²⁰

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) has several advantages for imaging of the skull and overlying scalp including superior soft tissue contrast and the ability to suppress fat signal within the skull or scalp. For these reasons, MRI is often used to further characterize soft tissue masses in the scalp and discrete osseous lesions (e.g., primary bone tumors, metastases).²¹⁻²⁴ It also easily depicts communication of scalp/skull lesions with the intracranial contents, a common

concern when the differential diagnosis includes lesions such as cephaloceles, sinus pericranii, and dermoid cysts.

The optimal MR protocol for evaluation of a scalp or cranial vault lesion includes at least two planes of fat-suppressed T2-weighted MR imaging (usually, short tau inversion recovery [STIR]) and two planes of contrast-enhanced T1-weighted MR imaging with fat suppression. Advanced imaging techniques such as diffusion-weighted imaging have also been applied to the skull to assess for pathology such as metastatic disease and osteomyelitis.^{25,26} In combination with standard brain MRI sequences, these additional sequences make intrinsic marrow signal abnormalities, focal osseous lesions, and soft tissue masses of the scalp conspicuous as the background signal from fat is suppressed. However, erosion of the skull and skull fractures remain poorly seen compared to CT because of the comparatively low spatial resolution of MR.^{27,28} New MRI techniques are attempting to address these limitations but are not yet widely used outside of research settings.^{29,30}

Nuclear Medicine

Positron and gamma ray emitting radiotracers are rarely used specifically to detect skull pathology. In the case of skull fractures (inflicted or accidental), it is widely accepted that skull fractures do not reliably appear on technetium-based bone scans because of poor callus formation though bone scintigraphy remains commonly performed for detection of fractures elsewhere.^{31,32} Whether introduction of ¹⁸F-fluorine PET improves sensitivity awaits further investigation.³³ For infections of the skull, there is adult literature that suggests gallium and technetium radiotracers may have some utility particularly for skull base infections,^{34–36} but MRI can provide similar information while simultaneously evaluating for intracranial pathology. For oncologic indications, several radiotracers remain in wide use for screening of metastatic disease and can therefore suggest skull involvement. These include staging for tumors common to the cranial vault such as Langerhans cell histiocytosis (technetium 99m methylene diphosphonate [Tc99m-MDP] and ¹⁸F-fluorodeoxyglucose [FDG])^{37,38} and neuroblastoma (Tc99m-MDP, ¹²³I-metaiodobenzylguanidine [MIBG], and FDG).^{39,40}

NORMAL ANATOMY

Major Sutures and Fontanelles

Sutures are fibrous articulations between bones of the cranial vault that develop by intramembranous ossification. The skull base (sphenoid, ethmoid, nonsquamous temporal bone, and occipital bone below the superior nuchal line) develop with enchondral ossification, and therefore the articulations of the skull base ossification centers are called *synchondroses*.⁴¹⁻⁴⁴ The major sutures of the cranial vault include the *sagittal suture* separating the paired parietal bones, the *metopic suture* separating the two halves of the frontal bone, the *coronal suture* separating the frontal and parietal bones, and the *lambdoid suture* separating the occipital and parietal bones (Figs. 1.1 and 1.2).

Major openings present at birth include the *anterior fontanelle* at the junction of the coronal/sagittal sutures (called the *bregma* when closed) and the *posterior fontanelle* at the junction of the lambdoid sutures (called the *lambda* when closed).

Accessory/Minor Sutures

A number of minor sutures are seen in the vicinity of the squamous temporal bone, including the *sphenotemporal (sphenosquamous) suture* anteriorly, *temporoparietal (squamous) suture* superiorly, and the *occipitomastoid suture* posteriorly (Fig. 1.2).⁴⁵ The H-shaped junction where the sphenoparietal suture meets the coronal/frontosphenoid sutures anteriorly and the sphenotemporal/temporoparietal sutures posteriorly is called the *pterion*. The complexity of occipital bone development (at least eight ossification centers) gives rise to a number of transient sutures in this region (Fig. 1.3). Common transient sutures include midline *superior and inferior occipital fissures* at the lambda and foramen magnum, respectively; *mendosal sutures* including forms dividing the squamous occipital bone transversely; and paired *anterior/posterior intraoccipital synchondroses* that straddle the anterior and posterior extent of the foramen magnum at the skull base.⁴⁶⁻⁴⁸

Normal Development and Timing of Suture Closure

The initial ossification of the skull begins at a few months of gestation and is largely complete by term delivery.⁴⁹ The sutures and skull base synchondroses are patent at birth and then begin to close at variable rates. However, two useful

rules of thumb are that no major suture should close in the first year of life and no suture should undergo mature fusion in childhood except for the metopic suture, which normally closes at 3 to 9 months.^{44,50-54} As the patient ages, the sutures become more serrated at their outer table though they remain smooth along the inner table.^{55,56} For cases where there is subjective narrowing, normative neonatal data have been published for selected sutures based on CT data.⁵⁷ Important synchondrosis milestones include closure of frontosphenoid and intersphenoid synchondroses by 1 to 2 years of age and of the sphenoccipital (clival) synchondroses during adolescence.⁵⁸ Closure of synchondroses and sutures within the occipital bone follow a more complicated sequence. The synchondroses at the foramen magnum typically close within the first few months of life though other sutures including mendosal sutures and inferior occipital fissure may persist through the first few years of life^{47,48,58,59}; variants where these sutures/synchondroses persist into adulthood are occasionally encountered. The anterior and posterolateral fontanelles typically close by the end of the 2nd year. The posterior fontanelle closes much earlier, typically by 3 to 6 months.⁶⁰

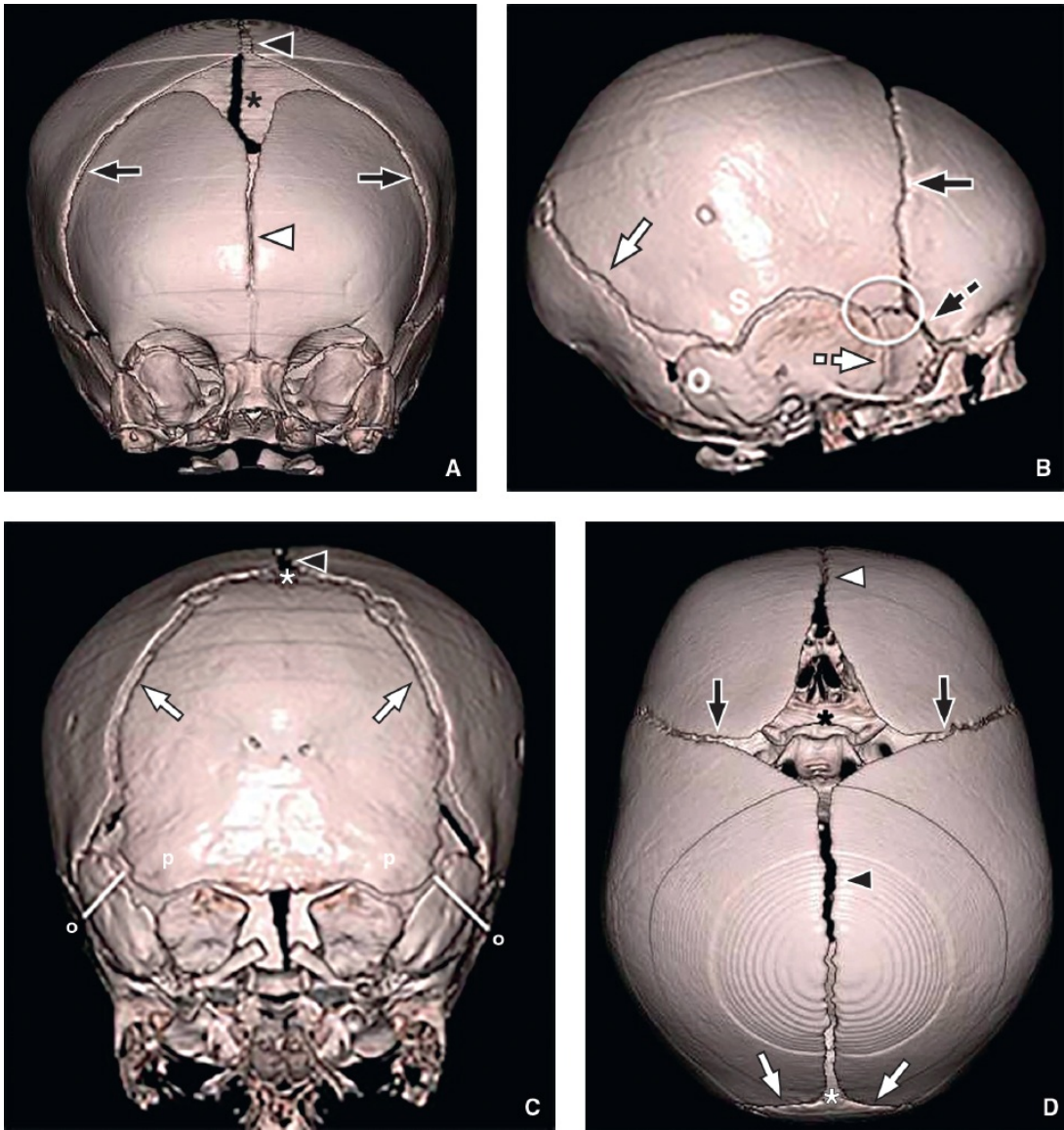


FIGURE 1.2 Three-dimensional surface renderings of a head CT from a normal 3-month-old boy evaluated for craniosynostosis: frontal (A), lateral (B), posterior (C), and superior (D) views. Annotations as for Figure 1.1 with additional markings of pterion with a circle, squamous (temporoparietal) suture with “s,” sphenotemporal suture with *white dashed arrow*, frontosphenoid suture with *black dashed arrow*, occipitomastoid suture with “o,” and posterior intraoccipital synchondrosis with “p.”

Deviations from normal sutural closing (apparent sutural widening) are present in conditions of poor bone mineralization (osteogenesis imperfecta, rickets, hypothyroidism) and can be misinterpreted as sutural diastasis from elevated intracranial pressure. Other reported causes of apparent widening of normal sutural width include recovery from chronic malnutrition, in utero renin-

angiotensin system disruption (Fig. 1.4), achondroplasia, trisomy 21, and treatment with prostaglandins for prematurity.^{61–65}

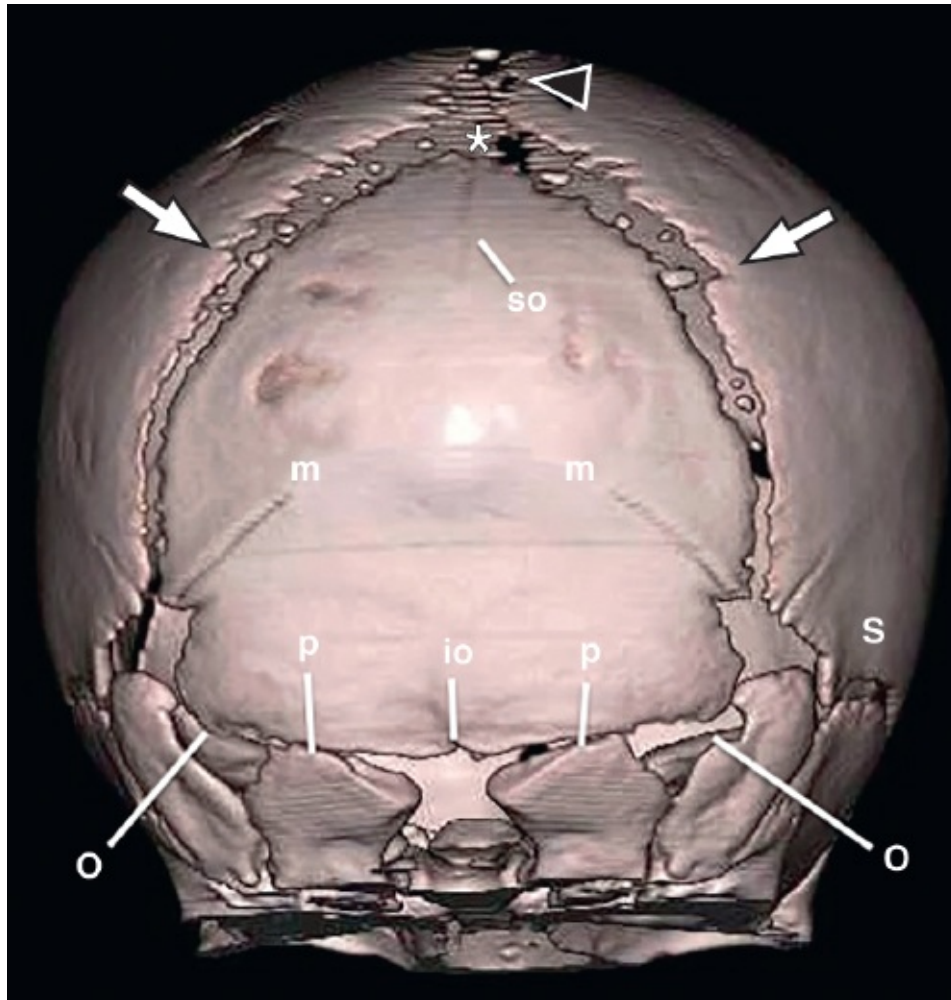


FIGURE 1.3 Transient sutures of the occipital bone visualized on a three-dimensional surface rendering of a 1-month-old boy's head CT (posterior view). Markings are as for Figures 1.1 and 1.2 but additional sutures marked include "so", "m", and "io" for superior occipital, paired mendosal sutures, and inferior occipital sutures, respectively.

SPECTRUM OF SKULL DISORDERS

Congenital and Developmental Anomalies

Craniosynostosis

Craniosynostosis refers to premature closure of the cranial sutures (segmental or

total) with resultant deformity. Craniosynostoses are usually classified as *primary* or *secondary*, secondary forms representing the consequence of an identifiable cause unrelated to suture development, such as metabolic bone disease, bone dysplasia, or loss of intracranial volume (e.g., shunting, brain injury). The primary craniosynostoses are further divided into *single* versus *multiple suture* forms and *syndromic* versus *nonsyndromic (isolated)* forms.

The overall incidence of craniosynostosis is low, ~3 to 10 cases per 10,000 live births.^{13,66} Craniosynostosis typically presents in the neonatal period and occasionally in utero though certain secondary craniosynostoses may present much later in childhood.⁶⁷ Upward of 80% of craniosynostosis cases are of the primary nonsyndromic (isolated form), ~75% to 80% being single-suture involvement and 20% to 25% being multiple sutures.^{68,69} Excluding rare instances of secondary craniosynostosis, the remaining cases consist of syndromic synostoses that typically involve multiple sutures. Craniosynostosis is seen more commonly with advanced parental age, multiparity, extremes of fetal weight, and (except for unilateral coronal synostosis) male gender.^{66,70,71} Although nonsyndromic causes of synostosis are generally viewed as idiopathic/sporadic, it is worth noting that a minority (~10%) of nonsyndromic synostosis has familial transmission (i.e., genetic cause); these familial nonsyndromic cases have a majority (2/3) of bicoronal synostosis compared to the nonsyndromic, nonfamilial cases where unicoronal synostosis constitutes 2/3 of cases.^{66,71-73} This fact may be explained by the increasing detection of mutations responsible for syndromic synostosis in “nonsyndromic” synostosis patients, and therefore many authorities advise genetic screening in coronal synostosis patients.⁷⁴⁻⁷⁶

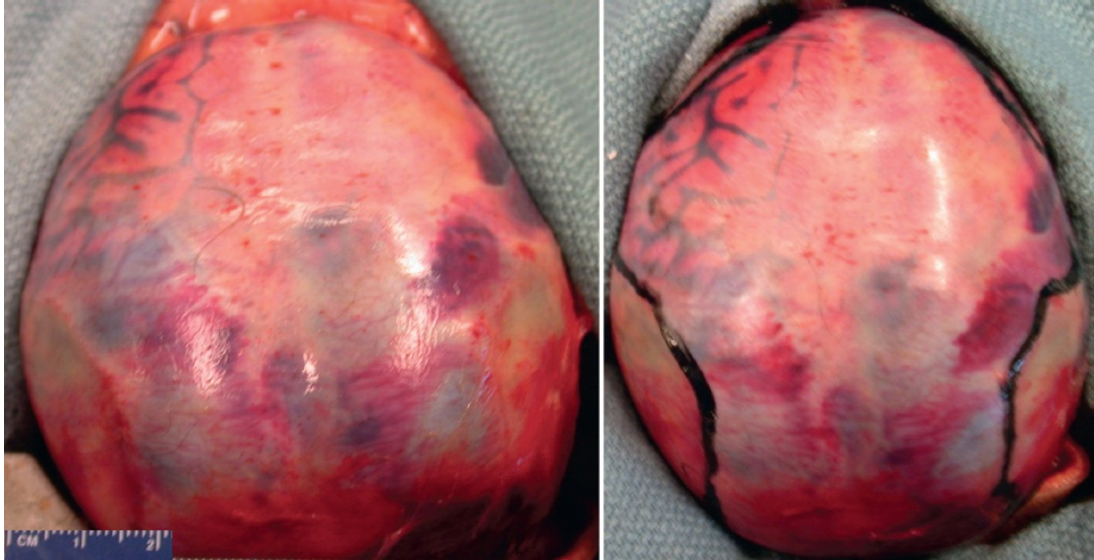


FIGURE 1.4 Markedly widened cranial sutures (hypocalvarium) in a 1-day-old girl with renal tubular agenesis, pulmonary hypoplasia, and other skeletal anomalies suggesting in utero exposure to renin–angiotensin system blockers or an inborn error in the renin–angiotensin system. Reflection of the scalp at autopsy shows only a thin membrane covering much of the cerebrum. In the right panel, the margins of the fontanelles are outlined in black ink.

As first recognized by Virchow in the mid-19th century,⁷⁷ premature fusion of a suture results in constriction of skull growth in the direction perpendicular to the affected suture and compensatory elongation of the skull in dimensions parallel to the abnormal suture. This simple principle explains the many patterns of deformity seen with craniosynostoses, recently reviewed in detail elsewhere.⁷⁸

Sagittal synostosis is the single most common craniosynostosis, accounting for roughly half of the nonsyndromic cases of craniosynostosis.^{66,68,70} Sagittal synostosis causes *scaphocephaly* or transverse narrowing with anteroposterior elongation of the skull, usually with some associated ridging at the site of fusion (Fig. 1.5).

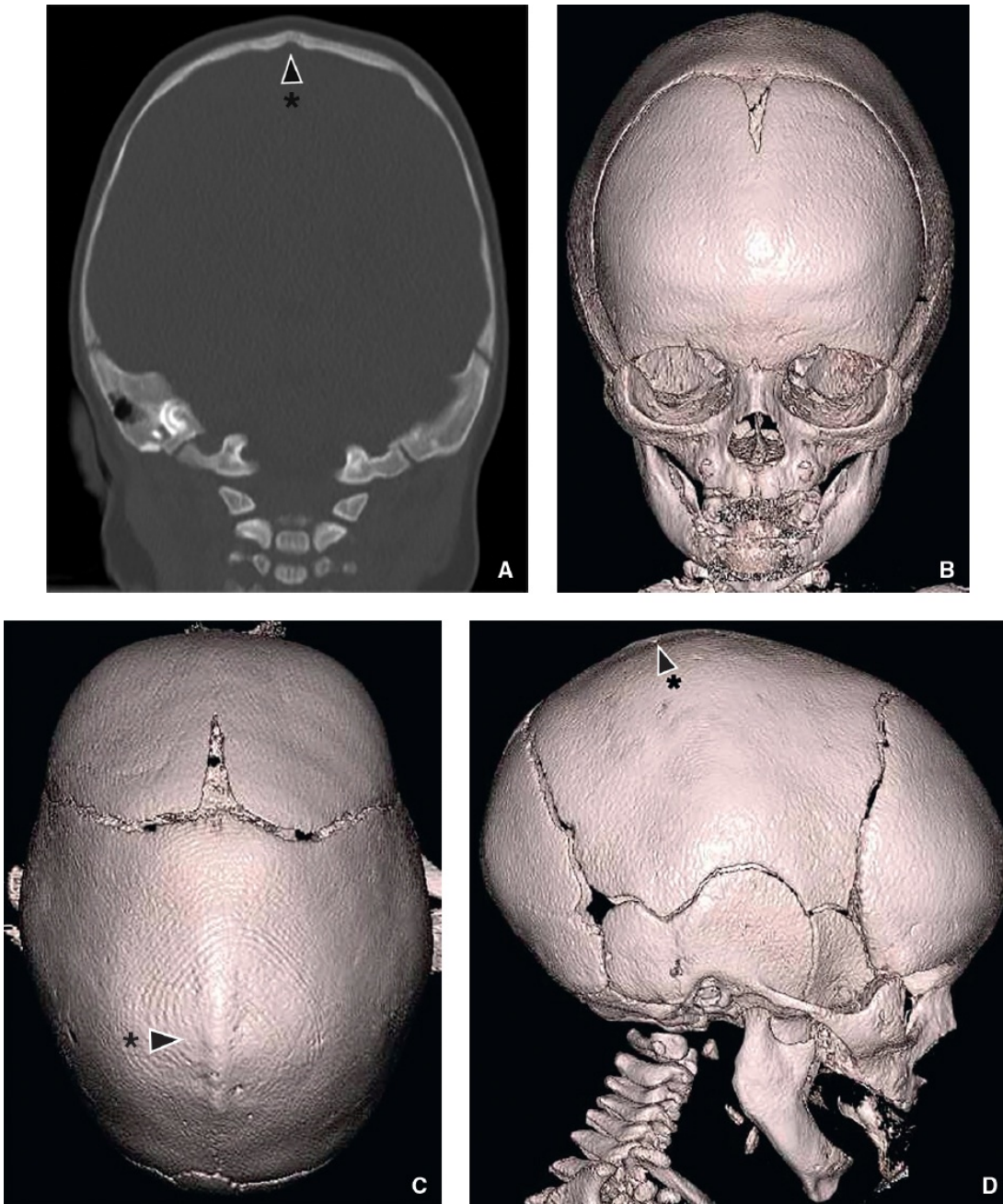


FIGURE 1.5 Sagittal synostosis in a 3-month-old boy with dolichocephaly and midline osseous ridging. **A:** Coronal reformatted bone window CT image shows premature fusion of the sagittal suture accompanied by osseous ridging (*asterisked black arrowhead*). **B–D:** Frontal (**B**), superior (**C**), and right lateral (**D**) surface rendering CT images confirm the osseous ridging and depict the elongated AP dimension of the skull consistent with clinical impression of dolichocephaly. When accompanied by osseous ridging, dolichocephaly is called scaphocephaly, a characteristic appearance for sagittal synostosis. Note that the metopic suture has already undergone fusion without deformity, consistent with normal closure.

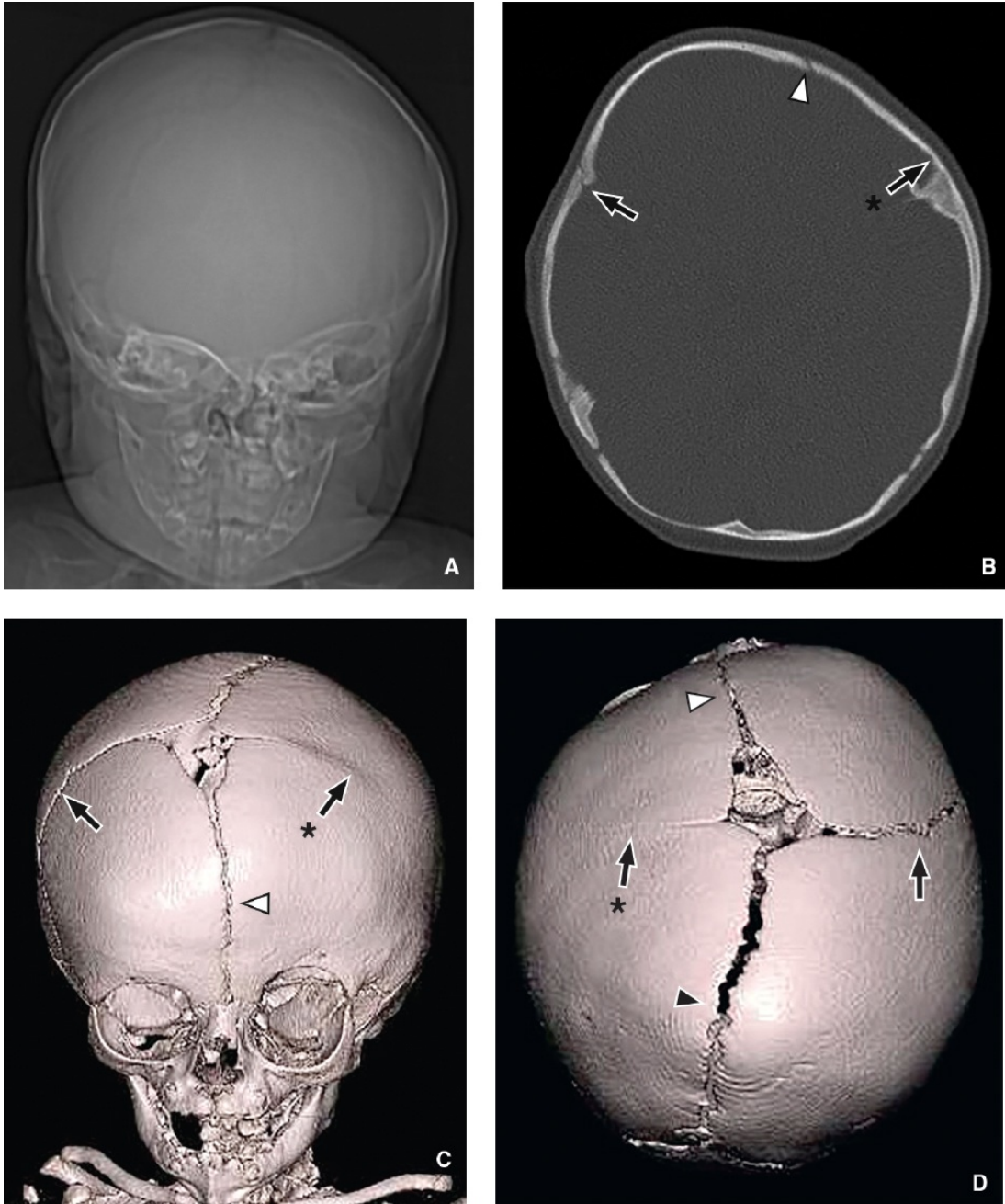


FIGURE 1.6 Unilateral coronal synostosis in a 4-month-old boy with abnormal head shape. **A:** Frontal projection CT topogram demonstrates elevation of the left orbital rim. **B:** Axial bone window CT image demonstrates premature fusion of the left coronal suture (abnormal side *asterisked arrow*). **C and D:** Three-dimensional surface rendering CT images in the frontal (**C**) and superior (**D**) perspective demonstrate left frontal plagiocephaly and right frontal bossing with retraction of the metopic suture to the left. Labeling conventions as for [Figure 1.2](#) with abnormally fused left coronal suture denoted by an *asterisked black arrow*.

Coronal synostosis can be either unilateral or bilateral. For unilateral coronal

synostosis, there is flattening of the frontal bone (anterior plagiocephaly) with some compensatory bulging of the contralateral frontal bone and skewing the metopic suture to the ipsilateral side. There is also upward slanting of the lateral orbital rim (“harlequin” deformity) from retraction of the ipsilateral frontal bone and orbital roof (Fig. 1.6). In cases of bilateral coronal synostosis, the harlequin orbit deformity is bilateral and the overall anteroposterior dimension of the skull is reduced, a morphology known as *brachycephaly* (Fig. 1.7).

Metopic synostosis causes narrowing of the frontal bone into a beak-like configuration termed *trigonocephaly*. Historically, metopic synostosis has been thought to be relatively rare at ~5% to 10% of nonsyndromic craniosynostosis, but multiple studies of the past 10 to 15 years have documented an increase of up to 20% of nonsyndromic craniosynostosis.^{70,79,80} The classic findings of metopic synostosis include retraction of the supraorbital ridges medially, bossing of the parietal bones, hypoplastic ethmoid sinuses, and a W-shaped metopic notch of the endocranial surface of the fused metopic suture (Fig. 1.8).^{54,78,81}

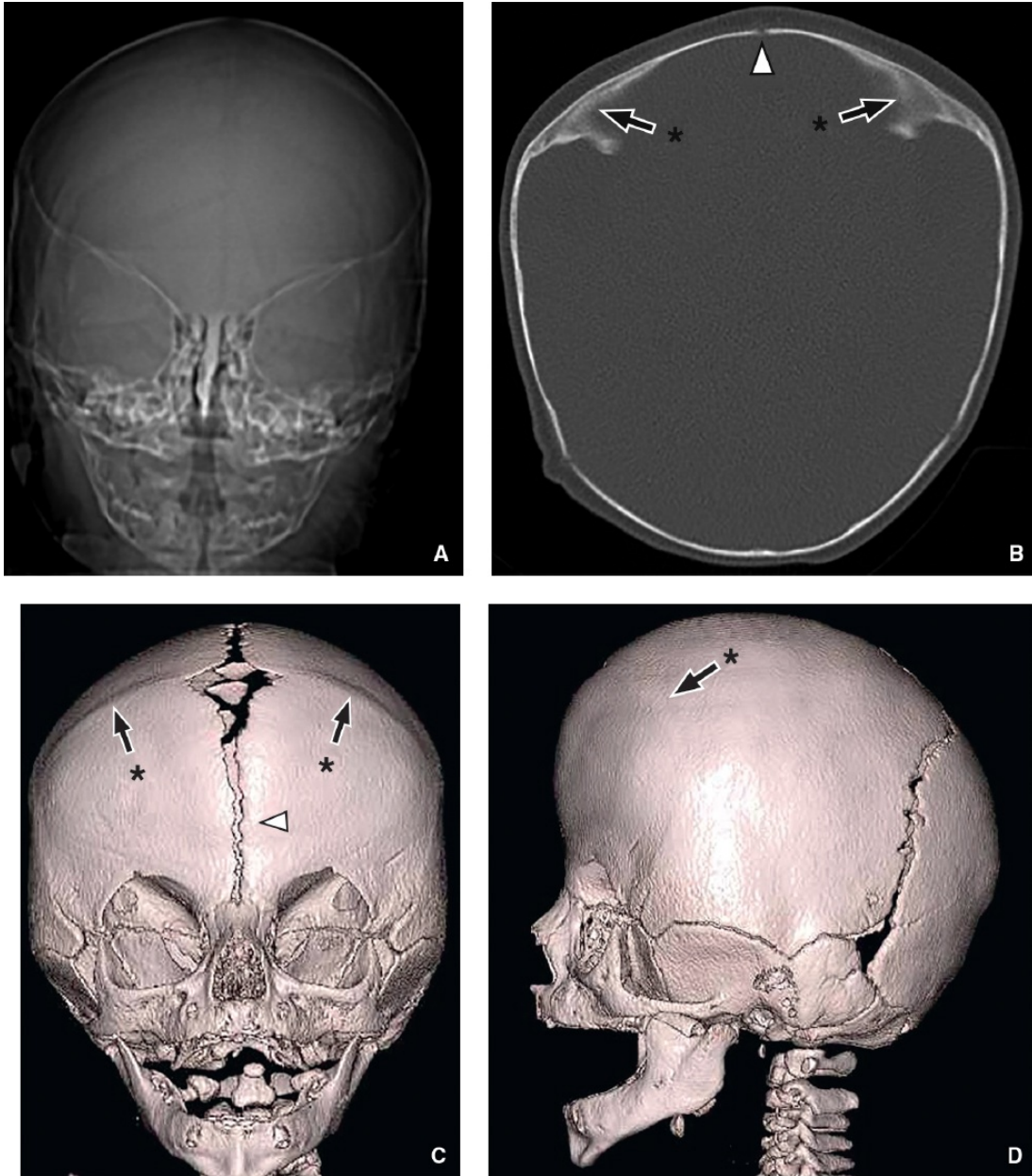


FIGURE 1.7 Bicoronal synostosis in a 2-month-old boy with skull deformity. A: Frontal projection CT topogram shows bilateral harlequin deformities of the orbits. **B:** Axial bone window CT image demonstrates bilateral anterior plagiocephaly and premature fusion of both coronal sutures (*asterisked black arrows* with metopic suture denoted by *white arrowhead*). **C and D:** Surface rendering CT images confirm premature fusion and ridging of the coronal sutures with upward retraction of the orbits in the frontal view (**C**) as well as brachycephaly on the lateral view (**D**).

Lambdoid synostosis is the most rare of the single-suture synostoses, estimated at <5% of nonsyndromic craniosynostoses.⁷⁰ After successful implementation of the “Back to Sleep” program by pediatricians in the 1990s,

there was a spike in the diagnosis of lambdoid synostosis, which later proved to be due to misdiagnosis of posterior deformational plagiocephaly (see below).^{82,83} In addition to posterior plagiocephaly, true lambdoid synostosis features compensatory enlargement of the ipsilateral frontal bone and contralateral occipital bone as well as skewing of the occipital protuberance towards the side of the synostosis (Fig. 1.9A–C).⁸⁴

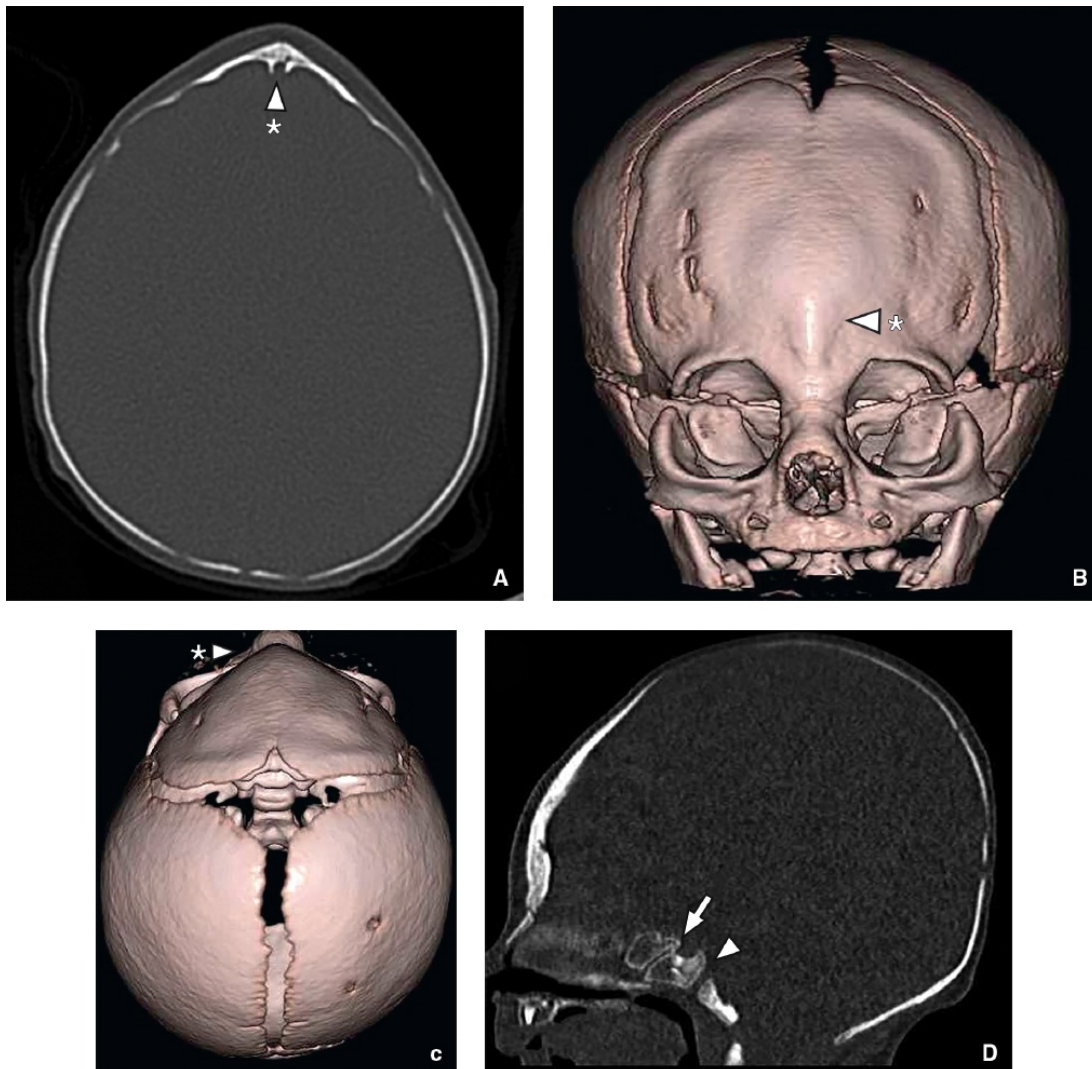


FIGURE 1.8 Metopic synostosis in a 24-day-old boy with trigonocephaly. **A:** Axial bone window CT image confirms trigonocephaly with prominent osseous ridge and metopic notch (*asterisked white arrowhead*) at the site of the prematurely fused metopic suture. **B and C:** Frontal and superior surface rendering CT images (**B and C**) show retraction of the medial orbital, hypotelorism, and the contour deformity of the skull. **D:** Sagittal reformatted CT image demonstrates an incidentally noted persistent craniopharyngeal canal (*arrow*) anterior to the normal spheno-occipital synchondrosis (*arrowhead*).

Craniosynostosis involving multiple sutures can result in unusual shapes.⁸⁵ *Turricephaly/oxycephaly* result from bilateral lambdoid synostosis with craniocaudal elongation of the calvaria and bulging at the vertex; this appearance can also be encountered with bicoronal synostosis (Fig. 1.10). When lambdoid, coronal, and sagittal sutures are all fused, there is bulging of the cranial vault where unconstrained by synostosis; this is known as the cloverleaf skull or *kleiblattschädel* (Fig. 1.11).

Syndromic craniosynostosis presents with characteristic additional clinical examination or radiographic findings. The features of the more common syndromic synostoses are summarized in Table 1.1. The major areas of differentiation between the syndromes are the findings outside of the cranial vault, namely findings in the extremities. Bicoronal synostosis (with or without additional sutures), midface hypoplasia, hypertelorism, and varying degrees of exorbitism (proptosis) are commonly seen in all the syndromic craniosynostoses (Figs. 1.10 to 1.12). These similarities are shared at a molecular level as many of the classically recognized syndromes share abnormalities in fibroblast growth factor receptor signaling.

Secondary craniosynostoses are rare, but the most frequent causes in routine clinical practice include overshunting and massive brain injury.⁸⁶ In both instances, the intracranial volume contracts, resulting in overlapping sutures and premature fusion with osseous ridging (Fig. 1.13). Other reported associations include underlying disorders of bone metabolism, such as rickets; bone dysplasias, such as achondroplasia; metabolic disorders, such as mucopolysaccharidoses; in utero compression; hematologic disorders, such as sickle cell and polycythemia; endocrine disorders, such as hyperthyroidism; and any cause of poor underlying brain growth.^{87–96}