"One of the most comprehensive and educational books on craniosynostosis."

—WALEED GIBREEL, MD

Gillette Children's Healthcare Series

CRANIOSYNOSTOSIS

Understanding and managing the condition: A practical guide for families

Ruth J. Barta, MD
Cheryl Tveit, RN, MSN, CNML
Heather Comstock, Parent

Editors

Lily Collison, MA, MSc
Elizabeth R. Boyer, PhD
Martin Lacey, MD
Tom F. Novacheck, MD

GILLETTE CHILDREN'S

Praise for *Craniosynostosis*

"This is one of the most comprehensive and educational books on the subject of craniosynostosis. I congratulate Dr. Barta and the team at Gillette Children's for putting together a thorough, detailed, and well-written book. It covers all aspects of craniosynostosis and plagiocephaly, and it is a valuable resource for providers and patients and their families. The educational material that is elegantly presented provides patients and their families with the information they need to make an informed decision, and it highlights the unique and holistic treatment approach followed by the experts at Gillette Children's."

WALEED GIBREEL, Pediatric Plastic and Craniofacial Surgeon, Mayo Clinic, Rochester, Minnesota

"I found the book very educational and easy to read. I think it will help many new families beginning the craniosynostosis journey. Very well written!"

SHELBY DAVIDSON, Parent of child with sagittal craniosynostosis, Seattle, Washington

"I had the pleasure of reading Craniosynostosis, *authored by health care providers Ruth Barta, MD, and Cheryl Tveit, RN, with parent Heather Comstock. This is a remarkably comprehensive review of all things related to craniosynostosis and is a terrific source of information for parents and families of infants and children affected by this rare and complex condition. It leaves no stone unturned. I hope it brings some level of comfort to parents and patients dealing with the challenges of craniosynostosis."*

CHRISTOPHER R. FORREST, Medical Director, SickKids Craniofacial Program, Toronto; Professor, Division of Plastic, Reconstructive and Aesthetic Surgery, University of Toronto

"This is an excellent and informative book that is both clear and factual. It serves as a valuable resource for families, providing them with essential knowledge about craniosynostosis and empowering them to engage confidently with medical and health professionals. The personal stories included offer hope and reassurance, reminding families that they are not alone in their journey. This is the book I wish I had when my son was diagnosed with sagittal craniosynostosis in 2019."

ELAINE L. KINSELLA, Parent; Chartered Psychologist and Associate Professor in Psychology, University of Limerick, Ireland

"Receiving a diagnosis of craniosynostosis can be scary; it's a condition most parents have never heard of. This book is the perfect starting point for anyone looking to understand the basics and to help them navigate the treatment options available as best practices in craniofacial surgery continue to evolve."

JEFFREY FEARON, Director Craniofacial Center, Dallas, Texas; President Emeritus of the American Society of Craniofacial Surgeons and the Texas Society of Plastic Surgeons

"This book provides an insightful exploration into the complexities of craniosynostosis, a condition where the bones in an infant's skull fuse prematurely. Through a blend of personal narrative and scientific research, it offers a comprehensive understanding of the condition's causes, symptoms, and treatment options. What sets this book apart is its compassionate portrayal of families affected by craniosynostosis. By sharing their stories, the authors shed light on the emotional challenges they face, from diagnosis to treatment decisions. Additionally, the book delves into the latest medical advancements, offering hope to families navigating this complex problem. It serves as an informative resource for those seeking knowledge about the condition; its real strength lies in its ability to inspire empathy and solidarity among readers."

EMMA CORDES, Assistant Professor, Indiana University Department of Surgery, Division of Plastic Surgery; Director of Cleft and Craniofacial Program; Director of Global Surgery Program

"Craniosynostosis *will be a valuable resource, especially for parents who are just learning of their child's diagnosis. The detailed explanation of the condition, the pronunciation of words, and the definitions are all beneficial to the reader in providing information all in one place—my favorite aspect. Incorporating personal stories gives a sense of reality, as well as comfort and reassurance often sought by those affected by craniosynostosis. I firmly believe this book will help educate and bring further awareness to the condition."*

LINDSAY WALKER, Parent of a child with nonsyndromic craniosynostosis, West Virginia

"This is a great reference book for all things craniosynostosis. It is appropriate for medical professionals as well as parents and caregivers who want to dig deeper into what they might anticipate for their child—not only in the early period of surgery but also beyond. The book is well designed and organized in such a way that the reader can more specifically find what they need. The personal stories and photographs so graciously shared by the families speak to how those having a child with craniosynostosis are affected in a way that we as providers are unable to fully comprehend. We know how to take care of these kids; families know what it is like to live it."

CHERYL HOLIHAN, Pediatric Nurse Practitioner Otolaryngology, University of Texas Southwestern, Dallas; Volunteer, Children's Surgery International

"As parents of a child with metopic CS we are certain this book will be a valuable resource to all. It provides both tactical information and human experiences through all phases of CS, from recognition to diagnosis, surgery and recovery, research, and beyond. We are so grateful for the continued research and education Gillette Children's is leading."

STACY AND ANDREW STUECK, Parents of child with metopic CS, Minnesota

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Gillette Children's Healthcare Press
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St Paul, MN 55101
www.GilletteChildrensHealthcarePress.org
HealthcarePress@gillettechildrens.com

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www.mackeith.co.uk
admin@mackeith.co.uk

To individuals and families whose lives are affected by these conditions, to professionals who serve our community, and to all clinicians and researchers who push the knowledge base forward, we hope the books in this Healthcare Series serve you very well.

All proceeds from the books in this series at Gillette Children's go to research.

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Authors and Editors

Ruth J. Barta, MD, Craniofacial and Pediatric Plastic Surgeon, Gillette Children's

Cheryl Tveit, RN, MSN, CNML, Principal Writer, Gillette Children's Healthcare Press

Heather Comstock, Parent

Lily Collison, MA, MSc, Program Director, Gillette Children's Healthcare Press

Elizabeth R. Boyer, PhD, Clinical Scientist, Gillette Children's

Martin Lacey, MD, Craniofacial and Plastic Surgeon Emeritus, Gillette Children's

Tom F. Novacheck, MD, Medical Director of Integrated Care Services, Gillette Children's; Professor of Orthopedics, University of Minnesota; and Past President, American Academy for Cerebral Palsy and Developmental Medicine

Series Foreword

You hold in your hands one book in the Gillette Children's Healthcare Series. This series was inspired by multiple factors.

It started with Lily Collison writing the first book in the series, *Spastic Diplegia–Bilateral Cerebral Palsy*. Lily has a background in medical science and is the parent of a now adult son who has spastic diplegia. Lily was convincing at the time about the value of such a book, and with the publication of that book in 2020, Gillette Children's became one of the first children's hospitals in the world to set up its own publishing arm—Gillette Children's Healthcare Press. *Spastic Diplegia–Bilateral Cerebral Palsy* received very positive reviews from both families and professionals and achieved strong sales. Unsolicited requests came in from diverse organizations across the globe for translation rights, and feedback from families told us there was a demand for books relevant to other conditions.

We listened.

We were convinced of the value of expanding from one book into a series to reflect Gillette Children's strong commitment to worldwide education. In 2021, Lily joined the press as Program Director, and very quickly, Gillette Children's formed teams to write the Healthcare Series. The series includes, in order of publication:

- *Craniosynostosis*
- *Idiopathic Scoliosis*
- *Spastic Hemiplegia—Unilateral Cerebral Palsy*
- *Spastic Quadriplegia—Bilateral Cerebral Palsy*
- *Spastic Diplegia—Bilateral Cerebral Palsy, second edition*
- *Epilepsy*
- *Spina Bifida*
- *Osteogenesis Imperfecta*
- *Scoliosis—Congenital, Neuromuscular, Syndromic, and Other Causes*

The books address each condition detailing both the medical and human story.

Mac Keith Press, long-time publisher of books on disability and the journal *Developmental Medicine and Child Neurology*, is co-publishing this series with Gillette Children's Healthcare Press.

Families and professionals working well together is key to best management of any condition. The parent is the expert of their child while the professional is the expert of the condition. These books underscore the importance of that family and professional partnership. For each title in the series, medical professionals at Gillette Children's have led the writing, and families contributed the lived experience.

These books have been written in the United States with an international lens and citing international research. However, there isn't always strong evidence to create consensus in medicine, so others may take a different view.

We hope you find the book you hold in your hands to be of great value. We collectively strive to optimize outcomes for children, adolescents, and adults living with these childhood-acquired and largely lifelong conditions.

Dr. Tom F. Novacheck

Series Introduction

The Healthcare Series seeks to optimize outcomes for those who live with childhood-acquired physical and/or neurological conditions. The conditions addressed in this series of books are complex and often have many associated challenges. Although the books focus on the biomedical aspects of each condition, we endeavor to address each condition as holistically as possible. Since the majority of people with these conditions have them for life, the life course is addressed including transition and aging issues.

Who are these books for?

These books are written for an international audience. They are primarily written for parents of young children, but also for adolescents and adults who have the condition. They are written for members of multidisciplinary teams and researchers. Finally, they are written for others, including extended family members, teachers, and students taking courses in the fields of medicine, allied health care, and education.

A worldview

The books in the series focus on evidence-based best practice, which we acknowledge is not available everywhere. It is mostly available in high-income countries (at least in urban areas, though even there, not always), but many families live away from centers of good care.

We also acknowledge that the majority of people with disabilities live in low- and middle-income countries. Improving the lives of all those with disabilities across the globe is an important goal. Developing scalable, affordable interventions is a crucial step toward achieving this. Nonetheless, the best interventions will fail if we do not first address the social determinants of health—the economic, social, and

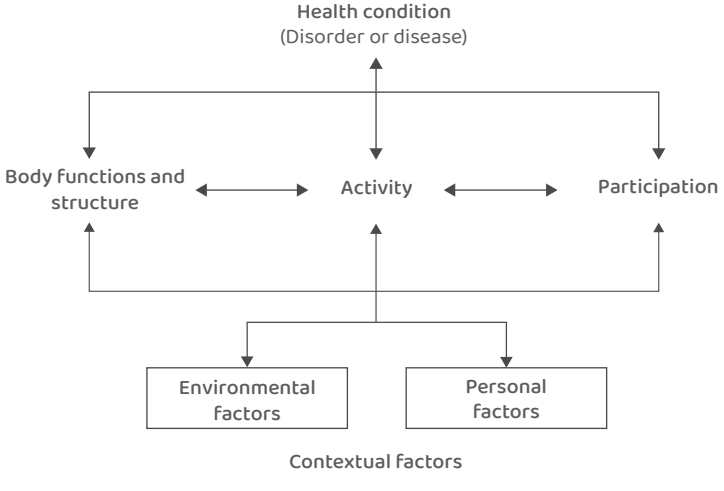
environmental conditions in which people live that shape their overall health and well-being.

No family reading these books should ever feel they have failed their child. We all struggle to do our best for our children within the limitations of our various resources and situations. Indeed, the advocacy role these books may play may help families and professionals lobby in unison for best care.

International Classification of Functioning, Disability and Health

The writing of the series of books has been informed by the International Classification of Functioning, Disability and Health (ICF).[1] The framework explains the impact of a health condition at different levels and how those levels are interconnected. It tells us to look at the full picture—to look at the person with a disability in their life situation.

The framework shows that every human being can experience a decrease in health and thereby experience some disability. It is not something that happens only to a minority of people. The ICF thus "mainstreams" disability and recognizes it as a widespread human experience.



International Classification of Functioning, Disability and Health (ICF). Reproduced with kind permission from WHO.

In health care, there has been a shift away from focusing almost exclusively on correcting issues that cause the individual's functional problems to focusing also on the individual's activity and participation. These books embrace maximizing participation for all people living with disability.

The Family

For simplicity, throughout the series we refer to "parents" and "children"; we acknowledge, however, that family structures vary. "Parent" is used as a generic term that includes grandparents, relatives, and carers (caregivers) who are raising a child. Throughout the series, we refer to male and female as the biologic sex assigned at birth. We acknowledge that this does not equate to gender identity or sexual orientation, and we respect the individuality of each person. Throughout the series we have included both "person with disability" and "disabled person," recognizing that both terms are used.

Caring for a child with a disability can be challenging and overwhelming. Having a strong social support system in place can make a difference. For the parent, balancing the needs of the child with a disability with the needs of siblings—while also meeting employment demands, nurturing a relationship with a significant other, and caring for aging parents—can sometimes feel like an enormous juggling act. Siblings may feel neglected or overlooked because of the increased attention given to the disabled child. It is crucial for parents to allocate time and resources to ensure that siblings feel valued and included in the family dynamics. Engaging siblings in the care and support of the disabled child can help foster a sense of unity and empathy within the family.

A particular challenge for a child and adolescent who has a disability, and their parent, is balancing school attendance (for both academic and social purposes) with clinical appointments and surgery. Appointments outside of school hours are encouraged. School is important because the cognitive and social abilities developed there help maximize employment opportunities when employment is a realistic goal. Indeed, technology has eliminated barriers and created opportunities that did not exist even 10 years ago.

Parents also need to find a way to prioritize self-care. Neglecting their own well-being can have detrimental effects on their mental and physical health. Think of the safety advice on an airplane: you are told that you must put on your own oxygen mask before putting on your child's. It's the same when caring for a child with a disability; parents need to take care of themselves in order to effectively care for their child *and* family. Friends, support groups, or mental health professionals can provide an outlet for parents to express their emotions, gain valuable insights, and find solace in knowing that they are not alone in their journey.

Last words

This series of books seeks to be an invaluable educational resource. All proceeds from the series at Gillette Children's go to research.

1.1

Introduction

Growth is never by mere chance;
it is the result of forces working together.
J.C. Penney

If you are reading this as a parent of a child recently diagnosed with craniosynostosis, this term may be new to you. It is pronounced "kray-nee-o-sin-os-TOE-sis" (the capitals indicate emphasis on that syllable). Craniosynostosis (CS) is a condition where the bones of the skull fuse together too early. While CS may not be noticed immediately by parents or medical professionals, it is a condition present from birth; therefore, it is known as a congenital condition.

Congenital CS is also referred to as primary CS.* It is relatively uncommon, occurring in 1 in 2,500 births.[3] In most cases, surgery in the first year of life will effectively correct it and those affected can expect a typical life (this is *nonsyndromic* CS). For a minority, CS is part of a syndrome, which is a lifelong condition (this is *syndromic* CS).

* Another type of CS, secondary CS, develops secondary to atypical brain development or other medical conditions; it is uncommon and is not included in this book.[2]

The term "craniosynostosis" comes from "cranio," meaning cranium (skull); "syn," meaning together; "ost," meaning bone; and "osis," meaning condition.

The medical definition of CS is:

> *The premature, pathologic fusion of one or more cranial sutures leading to an abnormal cranial shape that can subsequently result in facial deformities and increased intracranial pressure.*[4]

Table 1.1.1 further explains the terms used in the definition of CS.

Table 1.1.1 Terms in the definition of CS

TERM	EXPLANATION
Abnormal	Deviating from the typical expectation; atypical
Cranial	Relating to the skull
Deformities	Malformations or misshapen parts of the body
Facial	Referring to something affecting or concerning the face
Fusion	The process of joining two or more things together to form one
Intracranial pressure	The measure of pressure inside the skull
Pathologic	Referring to the involvement, cause, or nature of a condition
Premature	Referring to something occurring too early or before the usual or proper time
Suture	An immovable junction between two bones; cranial sutures are fibrous joints that connect the bones of the skull (not to be confused with surgical stitches, which are also called sutures)

The early fusion of the cranial sutures in CS causes the skull and face to become misshapen and may lead to further complications if not treated. Many of these skull shapes and sutures were first described centuries ago. Figure 1.1.1 shows early drawings of skull shapes and sutures.

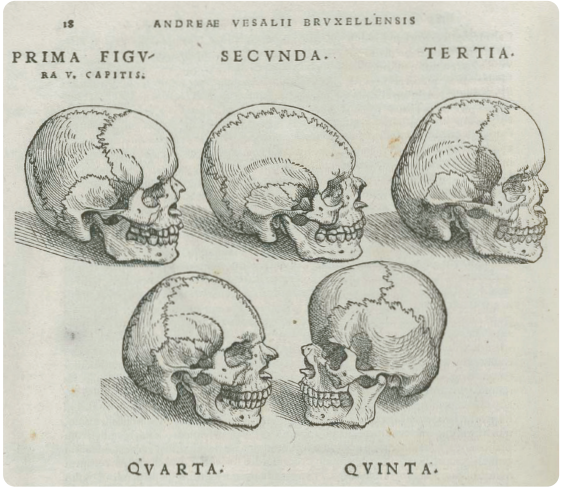


Figure 1.1.1 Early drawings of skull shapes and sutures (about 1543). Reproduced with kind permission from Syndromic craniosynostosis: from history to hydrogen bonds, by Cunningham et al. (2007) in *Orthod Craniofacial Res*, 10, 67-81.

The term "craniosynostosis" was first used by Otto in 1830. As early as 1851, Rudolph Virchow, a German physician, explained that a skull can expand evenly only when the sutures are appropriately open on all sides of the skull.[5] When a suture fuses prematurely, other areas in the skull compensate and get pushed apart to accommodate brain growth. This theory became known as Virchow's law, and it allows us to predict the skull shape based on the fused sutures.[6] Specifically, Virchow's law states:

> *When premature fusion of the cranial vault* occurs there is an inhibition of the normal growth of the skull in a direction perpendicular to the suture which is fused, which gives a compensatory growth in a direction parallel to the fused suture.*[8]

The atypical head shape in CS is caused by unequal *internal* forces on the skull (the fused sutures), but *external* forces may also contribute to an atypical head shape. When external forces are involved, it's called "deformational plagiocephaly" (pronounced as plag-jee-oh-sef-uh-lee) and is sometimes confused with CS. Indeed, deformational plagiocephaly may simultaneously occur with CS, but it is *not* CS. Deformational plagiocephaly is described in Chapter 5.

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* The cranial vault is the space that encases and protects the brain.[7]

How to read this book

This book is relevant to both types of CS: nonsyndromic and syndromic. This chapter addresses the overall condition of CS. Chapters 2 and 6 are specific to nonsyndromic craniosynostosis, and Chapters 3 and 7 are specific to syndromic craniosynostosis. The remaining Chapters—4, 5, 8, and 9—will be of interest to all readers. Throughout, medical information is interspersed with personal lived experience. Orange-colored boxes are used to highlight the personal story. Chapter 8 is devoted to vignettes from individuals and families around the globe. Chapter 9 addresses further reading and research. At the back of this book, you'll find a glossary with definitions of key terms. A companion website for this book is available at www.GilletteChildrensHealthcarePress.org. This website contains several resources, including **Useful web resources** (QR code below).

USEFUL WEB RESOURCES

My name is Heather, and I have been married to my husband, Jason, for almost 20 years. We live in the suburbs of Saint Paul, Minnesota. We have two boys and a girl, each two years apart. Our middle child, Keegan, was born with craniosynostosis.

I have worked at a Level 1 trauma center* for the last 15 years as a physician assistant† and thoroughly enjoy helping patients and their families achieve the best outcomes with their health. Once the term "craniosynostosis" came into our lives, and our son went through his cranio journey, we developed a newfound passion: to spread craniosynostosis awareness.

Keegan was born on a Saturday, late in the evening, with temperatures outside below freezing. Our firstborn son was at home with my parents, anxiously awaiting news of his baby brother's arrival. Labor progressed similarly to my first labor until it was time to push. We anticipated a speedy delivery, but it took more pushing than expected. Keegan's heartbeat would go down with contractions, which was a bit worrisome. They decided to use a vacuum to facilitate birth, which was used for just a minute because Keegan then decided to enter the world.

When he came out, the doctor found the umbilical cord was wrapped around his neck twice. We were so relieved when we heard him cry for the first time. I recall my husband seeing him first and making a comment about having some buggy eyes. I felt slightly concerned and thought that was an odd thing to say, but by the time I was able to meet him, he could not have been more perfect. He had big, beautiful eyes that were a very deep, slate-colored blue. He stole our hearts from that moment. We did notice he had a mark on the top of his head in the shape of a heart, likely caused by bruising during delivery. At the time, we thought it was sweet. Little did we know how much it would actually come to mean to us.

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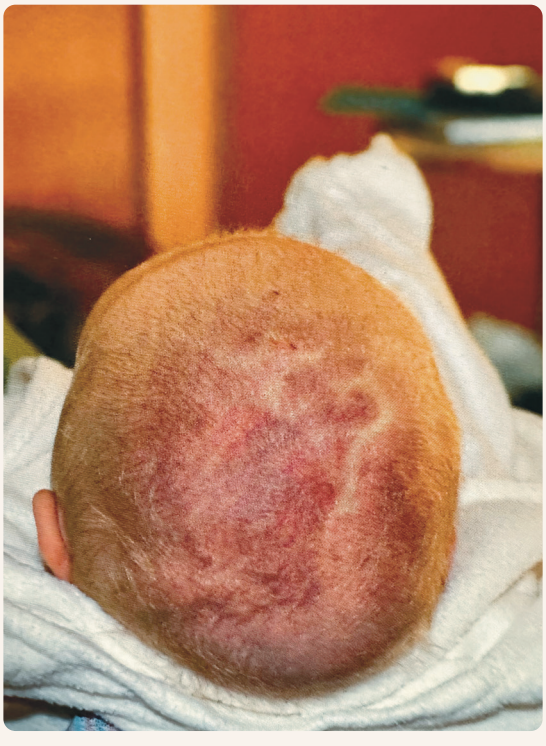
* In the US, a hospital that is capable of providing total care to patients through all aspects of a traumatic injury, as designated by the American Trauma Society.[9]

† A medical professional licensed to practice medicine in every specialty and setting in the United States and other jurisdictions. The training is modeled after medical school education, with a generalized focus, allowing them to care for all ages.[10] Physician assistants require graduate schooling and have greater medical privileges (e.g., diagnosing, prescribing) than nurses and work alongside a supervising physician.



Keegan, just after birth.

We first heard the term "craniosynostosis" when we brought Keegan in for his two-week well-child visit.* It was a new term for my husband. If I had learned about craniosynostosis in physician assistant school, it was just a mention—at most—in the pediatrics section, not enough to even trigger a memory of the term.

* In the US, well-child visits are regular appointments with medical professionals (typically as recommended by the American Academy of Pediatrics: three to five days after birth, then several times over the first 30 months, and then annually until adulthood). They often coincide with vaccination schedules and include growth and development tracking and monitoring.[11] These visits may be done in other countries, too, but may be known by other names and may occur with different timing.

1.2

Typical brain and skull development

The only reason for time is so that everything doesn't happen at once.

Albert Einstein

To understand CS, it is important to have a good understanding of brain and skull development. That includes understanding certain terms that are used throughout this book when referring to areas of the body. Table 1.2.1 lists directional terms used in anatomy, which are used specifically in this section (and generally throughout the book). (See the Glossary for a full list of terms used in this book.)

Table 1.2.1 Terms used to describe anatomical direction

TERM	EXPLANATION	VIEW
Anterior	Near the front, or front side	Front
Lateral	Away from the midline (or middle of the body), referring to the side	Side
Posterior	Near the back, or back side	Back
Superior	Above or looking down from above	Top

Embryonic and fetal skull development

An embryo is a developing human from conception up to the end of the eighth week after conception—referred to as the embryonic period. A fetus is a developing human from the eighth week after conception to birth.[12] "Prenatal" refers to the period before birth, and "perinatal" to the period around the time of birth.[13,14]

The brain and skull start to develop during the embryonic period. The cranial bones, or the skull bones, form approximately six to eight weeks after conception and the cranial sutures between 15 and 18 weeks.[12,15,16] Figure 1.2.1 shows the cranial bones in a fetus 12 weeks after conception. The gaps between the cranial bones are the areas where the cranial sutures will develop.

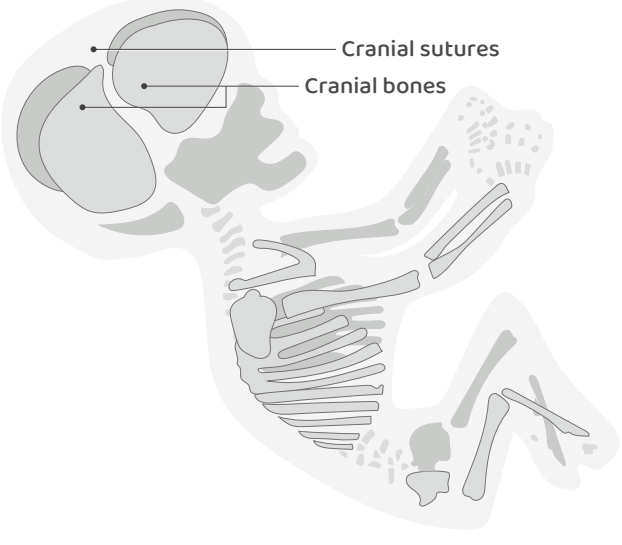


Figure 1.2.1 Fetal skeleton 12 weeks after conception.

a) Brain and skull development

At birth, the newborn* skull is made up of separate bones that are not yet fused. This allows for easier passage through the birth canal and rapid brain growth after birth. The skull protects the brain during this growth period—the brain triples in size by the time the child reaches one year of age.[5,7] This brain growth drives the growth of the skull, especially in the first few years of life. As the brain expands, it pushes the skull bones apart along the sutures, and the body creates and lays down new bone in between,[18] eventually forming the mature skull. With CS, this process is disrupted and the sutures fuse too early, which may negatively impact brain development if left untreated.

b) Cranial bones

There are many bones in the human skull, together called the cranial bones. The largest of the cranial bones make up the cranial vault, which is the space that encases and protects the brain.[7]

Figure 1.2.2 shows the newborn skull from the side (lateral view) and the top (superior view). The two frontal bones (teal) are on the skull's front (anterior); the two parietal bones (orange) are on the upper sides of the skull; the temporal bones (pink) are on the lower sides, so they are not visible in the superior view; and the occipital bone (purple) is at the back (posterior).

* A newborn is a child from birth to four weeks of age; an infant is a child from birth to one year of age.[17]

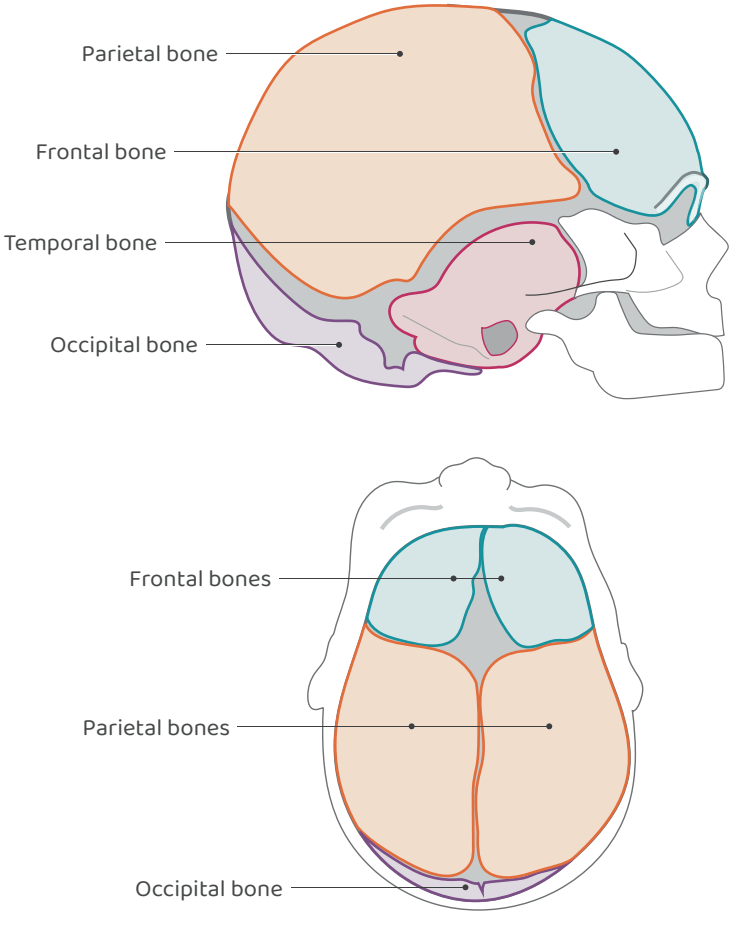


Figure 1.2.2 Cranial bones of a typical newborn: lateral view (top); superior view (bottom).

The cranial bones protect the brain and other structures in the skull. In Figure 1.2.3, notice the rounded frontal bones and orbital sockets (the openings for the eyes in the skull). One key function of the frontal bones is to protect the eyes. The supraorbital rim (the upper edge of the orbital socket) of each eye is formed by the frontal bone, and this design offers protection by creating brow bones that provide a cover over the eyeballs. In some forms of CS, this protective feature can be lost, leaving the eyes susceptible to injury.

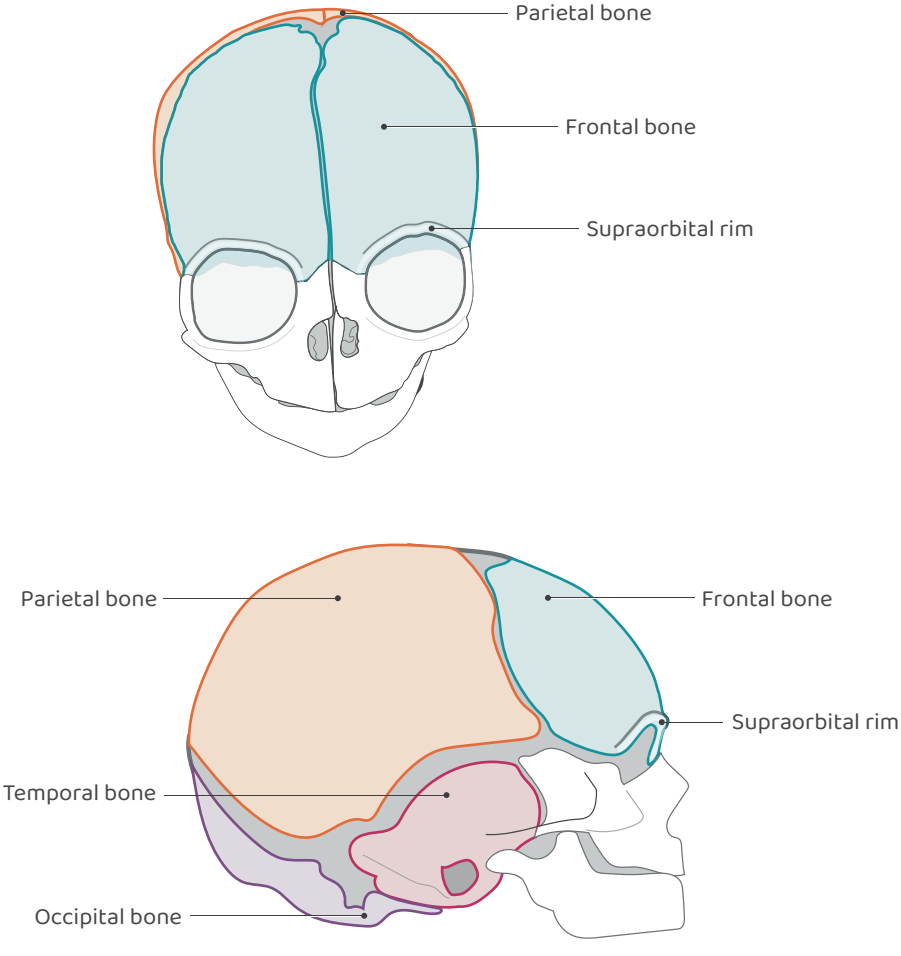


Figure 1.2.3 Skull of a typical newborn: anterior view (top); lateral view (bottom).

c) Cranial sutures

Cranial sutures are the fibrous joints that connect the bones of the skull. These are divided into major and minor sutures. While the minor sutures can also fuse prematurely, they rarely need surgical correction and are, therefore, not described here.

The major sutures are shown in Figure 1.2.4:

- **Sagittal suture:** Where the two parietal bones meet
- **Metopic suture:** Where the two frontal bones meet
- **Coronal suture:** Where one frontal bone and one parietal bone meet; there is a right coronal suture and a left coronal suture
- **Lambdoid suture:** Where one parietal bone and the occipital bone meet; there is a right lambdoid suture and a left lambdoid suture

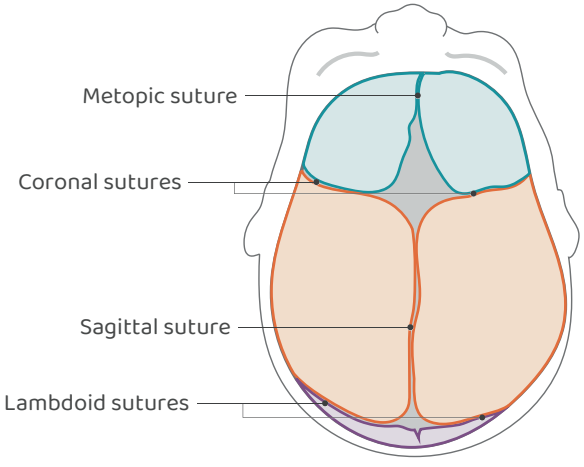


Figure 1.2.4 Cranial sutures of a typical newborn (superior view).

Suture fusion is the gradual process of ossification (turning into bone) of the fibrous material that makes up the sutures and the bones joining together. A suture is considered fused when all that remains is a thin line where the bones are joined. Figure 1.2.5 shows two views of a newborn skull, with open sutures, and two views of an adult skull, with fused sutures, for comparison.

Figure 1.2.5 Lateral and superior views of a newborn skull (top) and an adult skull (bottom).

The sutures generally fuse in order from front to back (anterior to posterior) and from the sides to the midline.[19]

- The metopic suture is the first suture to fuse and the only major suture to normally close in infancy. It sometimes starts fusing as early as 3 months of age and is typically completely fused by 9 to 12 months of age.[5,7,20]
- The sagittal, coronal, and lambdoid sutures begin fusing between the ages of 20 and 29 years.[5,21]

The premature fusion of these sutures in newborns or infants, as occurs with CS, can impact head shape and other aspects of health if left untreated, especially if more than one suture is involved.

d) Cranial fontanels

Cranial fontanels are the areas where the "corners" of the skull bones meet. You can feel them in newborns and young infants, and they are sometimes called "the soft spot" because they feel soft compared to the rest of the head. As the infant grows, the fontanels close from the edges to the middle, as the body creates new bone, eventually covering them completely. It's important to note that the disappearance of fontanels is not the same as fusion of the bones or suture fusion. However, certain changes to the fontanels, such as persistent bulging or an atypical shape, may raise concern.[22]

There are several fontanels in the newborn skull. Figure 1.2.6 shows the two main ones.

- The anterior fontanel is toward the front of the head and is the largest fontanel, making it easy to find. It is at the junction of the coronal, metopic, and sagittal sutures. The anterior fontanel typically fills in between 12 and 18 months.[22,23]
- The smaller posterior fontanel is toward the back of the head. It is at the junction of the lambdoid and sagittal sutures. This fontanel typically fills in by approximately two months of age.[23]

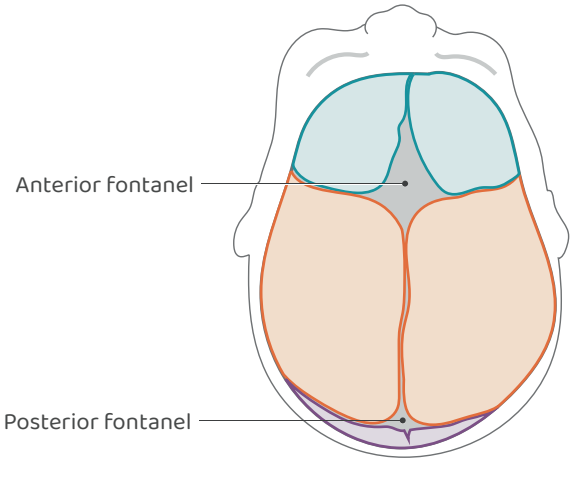


Figure 1.2.6 Cranial fontanels of a typical newborn (superior view).

I had two miscarriages before our first child was born. Both were early in the pregnancy. After our first child was born, we became pregnant again and I was cautiously optimistic, but I also was mentally preparing for another miscarriage. The pregnancy did indeed last, without any prenatal concerns. Every checkup and ultrasound showed Keegan growing as expected. No mention of craniosynostosis occurred during the pregnancy.

1.3

Classifications of craniosynostosis

Divide each difficulty into as many parts as is feasible and necessary to resolve it.

René Descartes

There are multiple classifications of CS. Generally, it is classified based on the cause of the condition, the number of sutures involved, and the sutures that fused prematurely, resulting in a specific head shape.

Classification by the cause

The most important classification of CS is based on cause: *nonsyndromic* versus *syndromic*.

- **Nonsyndromic CS:** CS that is not associated with a syndrome but is instead its own medical condition that has no known cause. Nonsyndromic CS accounts for 85 percent of cases of CS.[24]
- **Syndromic CS:** CS that is associated with a syndrome (a group of characteristics that consistently occur together).[25] Syndromic CS usually has a genetic cause and accounts for 15 percent of cases of CS.[24]

Classification by the number of sutures

- **Single suture CS:** The premature fusion of one suture. Most forms of nonsyndromic CS are single suture.[3]
- **Multisuture or multiple suture CS:** The premature fusion of more than one suture. Most forms of syndromic CS are multisuture[26] and the treatment is more complex than for single suture CS.

Classification by the suture that fused prematurely

- **Scaphocephaly:** The boat-shaped head typical of sagittal CS, which is caused by the premature fusion of the sagittal suture ("scapho" means boat).[27]
- **Trigonocephaly:** The triangular-shaped head typical of metopic CS, which is caused by the premature fusion of the metopic suture ("trigono" means triangle).[27]
- **Anterior plagiocephaly:** The skewed head shape in which the front of the head is flat on one side typical of unicoronal CS, which is caused by the premature fusion of one of the coronal sutures ("plagio" means oblique or slanted or at an angle; "uni" means one).[27]
- **Brachycephaly:** The short and wide head shape with a prominent forehead typical of bicoronal CS, caused by the premature fusion of both coronal sutures ("brachy" means short; "bi" means two).[27]
- **Posterior plagiocephaly:** The skewed head shape in which the back of the head is flat on one side typical of lambdoid CS, which is caused by the premature fusion of one of the lambdoid sutures.[27]

Figure 1.3.1 identifies various head shapes and the suture that fused prematurely to cause the head shape. The top image shows a skull without CS (normocephaly; "normo" means normal). The teal arrows indicate where the expanded compensatory growth will occur. The orange arrows indicate the area where growth is restricted due to the fused suture. The orange dotted lines indicate the fused suture.

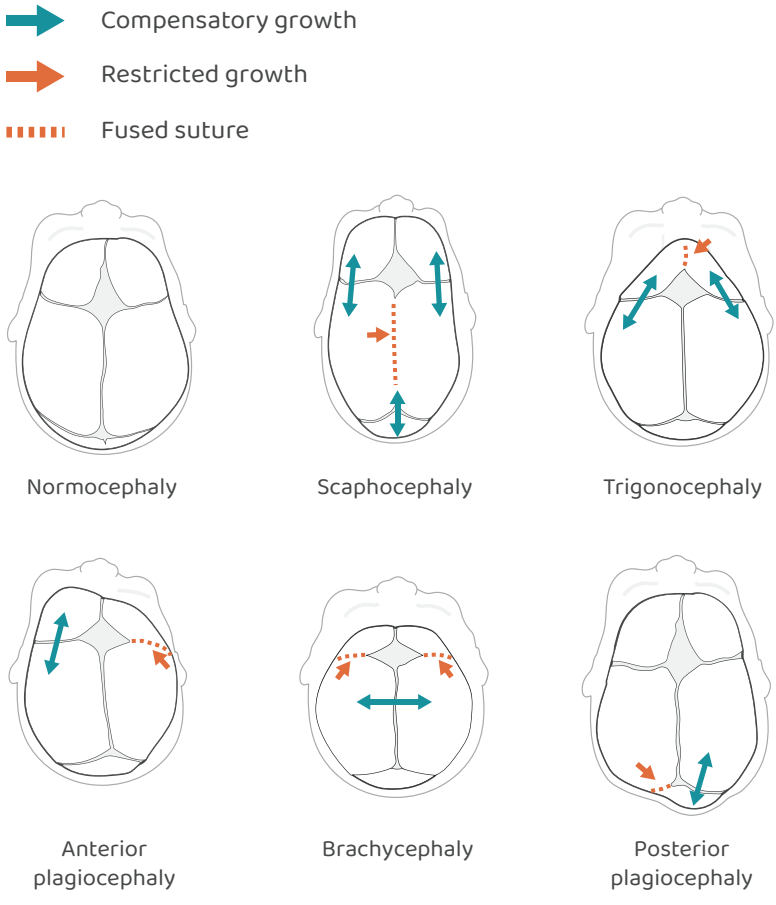


Figure 1.3.1 Classifications of CS by suture that fused prematurely and resulting head shape.

Table 1.3.1 summarizes key information about nonsyndromic CS and syndromic CS. Chapter 2 describes the types of nonsyndromic CS, and Chapter 3 describes the five most prevalent types of syndromic CS.

Table 1.3.1 Key information about nonsyndromic CS and syndromic CS

	NONSYNDROMIC CS	SYNDROMIC CS
Single suture	Frequent	Less frequent
Multisuture	Less frequent	Frequent
Progressive suture fusion after birth	Less frequent	Frequent
Head shape	Scaphocephaly: sagittal CS Trigonocephaly: metopic CS Anterior plagiocephaly: unicoronal CS Brachycephaly: bicoronal CS Posterior plagiocephaly: lambdoid CS	Depending on which sutures fuse, varying shapes can result.
Present at birth (congenital)	Yes	Yes
Prevalence	85%[24]	15%[24]

Figure 1.3.2 shows classification of CS beginning with the most important classification on the left side moving toward other classifications on the right. The size of the rectangles in the figure are generally proportional to prevalence.

1.6

Why treatment is important

In times of stress, the best thing we can do for each other is to listen with our ears and our hearts and to be assured that our questions are just as important as our answers.

Fred Rogers

The treatment for CS is surgical repair. Most often, particularly for nonsyndromic CS, a single surgery before a child is one year of age is all that is required to completely correct the condition. Others, however, may require additional surgeries (both for those with nonsyndromic and syndromic CS), and children with *syndromic* CS may require surgeries for other body systems. Surgical management of CS is described in Chapter 4.

There are several goals in the treatment of CS:

- **Preventing increased intracranial pressure and permanent brain damage:** Brain damage from increased intracranial pressure could

result in cognitive impairment, seizures,* and life-threatening symptoms.[49,51] It is estimated that at least 15 percent of individuals with nonsyndromic CS and up to 60 percent with multisuture CS, as often occurs with syndromic CS, have increased intracranial pressure.[61,62] However, there is no way to predict which individuals with CS will develop this condition, which is why a surgical repair is always recommended. Surgical repair promotes normal brain growth by releasing the restriction in the cranial vault caused by the prematurely fused sutures.

- **Preventing papilledema:** Papilledema is the swelling of the optic nerve, which can result from increased intracranial pressure. Untreated, it can lead to permanent vision loss.[5,63]
- **Correcting head shape and facial symmetry:** Surgical repair can correct the head shape and face to improve appearance, which has been shown to positively affect emotional and social well-being.[5] It also allows for better fitting of protective headgear such as helmets for sports. Surgical repair is best done early, as it is more challenging and may not be possible after skull growth is complete.[64]
- **Protecting eyes and correcting eye abnormalities:**
  - **Supraorbital rim (brow bone) abnormality:** When the brow bone, which protects the eyes, is altered, the eyes are more susceptible to injury. Figure 1.6.1 shows an infant with metopic CS before and after corrective surgery.
  - **Hypertelorism/hypotelorism:** Hypertelorism refers to the spacing between the eyes being wider than typical; hypotelorism refers to the spacing being smaller than typical ("hyper" means over or overly; "hypo" means below or low).
  - **Proptosis (exorbitism):** This describes bulging eyes and may prevent the eyes from closing completely. This condition puts the cornea (the transparent layer at the front of the eye) at risk for injury and may lead to issues with vision.[54]
  - **Strabismus:** This is the misalignment of the eyes, sometimes appearing as crossed eyes. It may occur due to the change in space between the eyes, abnormal anatomy, or abnormal muscles that control eye movements.[54] If it is left uncorrected, it can

* A seizure is "a sudden, uncontrolled, abnormal burst of electrical activity in the brain that may cause changes in the level of consciousness, behavior, memory, or feelings."[59] Seizures may occur in individuals diagnosed with epilepsy, which is a neurological disorder in which brain activity becomes abnormal.[60] Seizures may also occur without epilepsy and may be caused by factors within the brain, such as increased intracranial pressure.

lead to what is commonly called lazy eye (amblyopia), where one eye becomes stronger and the other, the "lazy" one, has reduced vision.[65]

- **Managing a Chiari malformation:** A Chiari malformation is a condition in which brain tissue extends into the spinal canal instead of staying within the skull. It occurs because the base of the skull becomes too small and forces some of the brain tissue downwards.[66,67,68] If an infant has CS, having a neurosurgeon evaluate for a Chiari malformation is recommended. Some types of CS are more susceptible to a Chiari malformation, especially those that involve the lambdoid suture.[66]

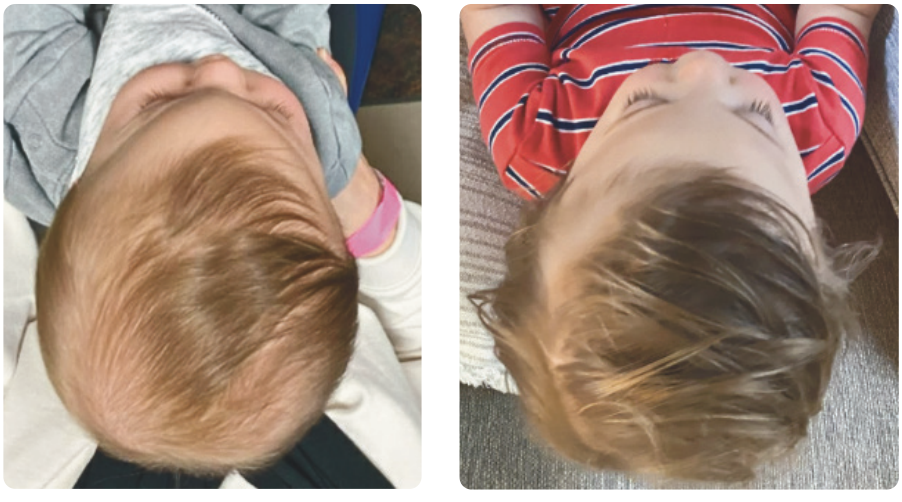


Figure 1.6.1 Infant with metopic CS before (left) and after corrective surgery (right). In the left image, the angle of the frontal bone is not rounded and the angle of the supraorbital rim is not protective of the eyes. In the right image, the frontal bone is rounded and eye protection by the supraorbital rim is restored.

Besides these surgical corrections, children with CS may require treatment for cognition-related issues. "Cognition" refers to the process of acquiring knowledge and understanding (e.g., memory, learning, and planning).[69] Specific issues related to cognition and behavior are described in both Chapter 2 (nonsyndromic) and Chapter 3 (syndromic).

Keegan was referred to an ophthalmologist to monitor eye pressure, and he was found to have very mild ptosis (drooping of the upper eyelid).* This has never required intervention. He continues with yearly eye appointments, including dilation of the eyes, which have never shown any problems.

* Some forms of syndromic CS are associated with a high risk of ptosis and are described further in Chapter 3. Ptosis can also occur without CS.

Key points Chapter 1

- CS occurs when the sutures in an infant's skull fuse and cause the bones to join prematurely. This condition results in an atypical head shape. Surgical repair is the treatment for CS.
- CS occurs in 1 in every 2,500 births worldwide.
- Because the brain grows along with the skull, when the skull growth is disrupted, there is a risk of injury to the brain.
- CS is a condition that is present at birth, but it is not usually detected before birth.
- CS can be either nonsyndromic (accounting for 85 percent of CS cases and not associated with any other conditions) or syndromic (accounting for 15 percent of CS cases and associated with a condition, known as a syndrome).
- CS may involve a single suture fusing or multiple sutures fusing.
- No single cause of nonsyndromic CS has been identified.
- Syndromic CS is often linked to a genetic cause.
- A physical exam along with imaging studies such as CT, MRI, X-ray, or ultrasound are often done to confirm the diagnosis of CS.
- CS is best managed by a multidisciplinary craniofacial team in a hospital experienced in the treatment of CS, using a shared decision-making model.

2.2

Sagittal CS

There isn't a way things should be.
There's just what happens, and what we do.
Terry Pratchett

Sagittal CS, or sagittal synostosis, occurs when the sagittal suture prematurely fuses. The sagittal suture is located where the two parietal bones meet, in the midline (middle) of the skull. This suture normally begins fusing at 22 years of age.[21] Sagittal CS is the most common type of CS, responsible for 40 to 55 percent of all cases.[5] It occurs more frequently in males than in females, at a 4:1 ratio.[3]

When the sagittal suture fuses early, the parietal bones cannot be pushed apart, and the skull cannot expand from side to side. Therefore, the skull expands front to back instead to accommodate brain growth. As shown in Figure 2.2.1, this causes scaphocephaly, or a boat-shaped, elongated head with a very prominent forehead and protruding occipital region (back of the head). In this infant skull, the metopic suture is fused as well, which is a typical finding in infants older than three months.

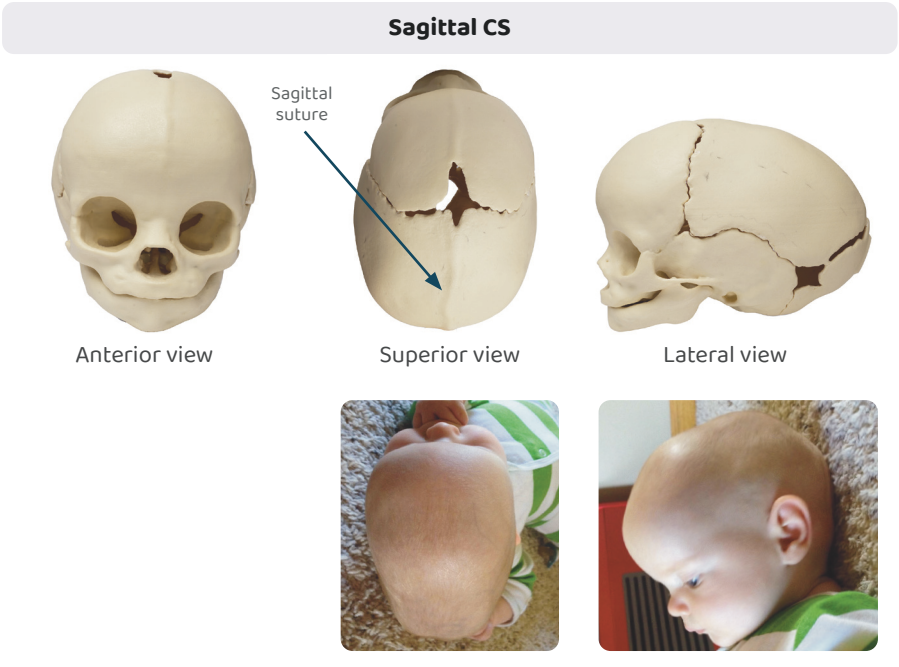


Figure 2.2.1 Anterior, superior, and lateral views of an infant skull showing fused sagittal suture (top). Superior and lateral photos of infant with sagittal CS at 10 weeks (bottom).

Key points Chapter 2

- Nonsyndromic CS is present as an isolated condition, is not part of a syndrome, and accounts for 85 percent of all CS cases.
- Nonsyndromic CS typically involves only one suture, and in the majority of individuals, a single surgery before the age of one will permanently correct the condition.
- Different types of nonsyndromic CS are sagittal CS, metopic CS, coronal CS (unicoronal or bicoronal), and lambdoid CS. Sagittal CS is the most prevalent form of nonsyndromic CS.
- Deformational plagiocephaly is a condition where an area of the head becomes flat due to external pressure, typically from positioning. It often results in a similar head shape seen with lambdoid CS and therefore needs to be distinguished from CS. It is not CS and does not require surgery.
- Issues related to cognition and behavior may be present in children with nonsyndromic CS and appear to be suture-dependent and may be related to the timing of the surgical repair.
- Neuropsychological testing or other screening may be recommended. This can help in securing additional resources and monitoring the outcomes of any recommended early interventions.
- Children with nonsyndromic CS are at an increased risk of speech and language concerns compared to the general population. Referrals to speech and language specialists may be needed.

Key points Chapter 3

- Syndromic CS usually has a genetic cause and is part of a syndrome (a syndrome is a group of characteristics that consistently occur together). Syndromic CS accounts for 15 percent of all CS cases.
- There are almost 200 syndromes associated with CS. Five of the most prevalent are Apert, Crouzon, Pfeiffer, Muenke, and Saethre-Chotzen.
- Syndromic CS often involves the premature fusion of multiple sutures.
- Increased intracranial pressure, hydrocephalus, and Chiari malformation occur in individuals with syndromic CS at higher rates than in those with nonsyndromic CS and may require treatment or additional surgeries, such as the placement of a shunt to treat hydrocephalus.
- Airway management, feeding, and eye closure are initial management concerns in individuals with syndromic CS and may need to be addressed prior to surgical repair of CS.
- Syndromic CS may be accompanied by concerns related to cognition, learning, and behavior. These may result in intellectual disabilities and developmental delays. These are generally more prevalent in children with syndromic CS compared to children with nonsyndromic CS.

Helmet therapy after endoscopic surgical repair helps correct the shape of the skull. The helmet, or helmet orthosis, is custom-made so the protruding part of the head is confined while open areas are left to allow for growth that will produce a symmetrical head shape. Helmet orthoses are usually made of lightweight, plastic or carbon fiber (see Figure 4.3.2). The helmet is worn 23 hours a day for several months until the desired skull shape is achieved, typically until the child is at least 12 months. Because the brain and skull grow rapidly in this period, the helmet will need to be adjusted frequently.

Figure 4.3.2 Helmet orthosis.

Another technology that can be paired with the endoscopic approach is the use of surgical springs to shape the head instead of a helmet. After the abnormal bone is removed, springs are placed where the fused bone used to be. Over time, the springs slowly push the bones apart to create more space for the brain and a more normal head shape. This requires a second surgery to remove the springs, and the technology is not available at every center.

Open surgical repair

Open surgical repair is done through a larger incision compared to an endoscopic surgical repair. It involves remodeling of the bones, not just removing the affected suture.

The goal of open surgical repair is to increase the cranial vault volume and make more space to accommodate the growing brain. The cranial vault is the space that encases and protects the brain. This repair for CS is generally referred to as cranial vault remodeling (CVR). Several

techniques may be used, depending on which sutures are involved. The basic principle of each repair is the same; the cranial bones are cut and/or removed, reshaped, and often secured with resorbable plates and screws. (Resorbable plates and screws are composed of materials that break down and are absorbed into the body, so they don't have to be removed.)

Open surgical repair produces more immediate results compared to endoscopic surgical repair and does not require helmet therapy afterwards.[7] However, it is a longer surgery, requires a longer hospital stay, more frequently requires blood transfusions, and costs more.[165,166,167]

Different open cranial vault remodeling surgical repair techniques are described.

a) Posterior cranial vault remodeling

Posterior cranial vault remodeling addresses areas at the back of the head and is typically used to correct lambdoid CS (see Figure 4.3.3). The occipital bone of the skull is removed, reshaped, and put back in place with resorbable plates and screws. This surgical repair is typically done at four to six months of age.[5]

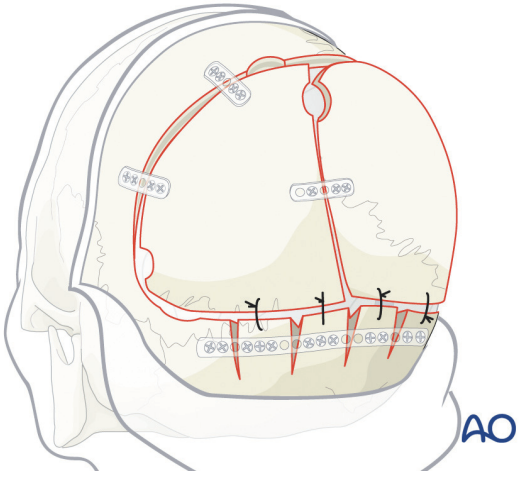


Figure 4.3.3 Skull showing posterior cranial vault remodeling for lambdoid CS. AO Surgery Reference, www.aosurgery.org. Reproduced with kind permission. Copyright by AO Foundation, Switzerland.

b) Anterior cranial vault remodeling with fronto-orbital advancement

Anterior cranial vault remodeling addresses areas in the front of the head. This is typically performed with the fronto-orbital advancement surgical repair, which corrects the supraorbital rim and upper eye socket area by moving the forehead forward.

Figure 4.3.4 shows a skull view following fronto-orbital advancement in a child with metopic CS. The remodeled forehead improves eye protection and reshapes the eye sockets. Typically, this is used to correct metopic CS, unicoronal CS, and bicoronal CS, and is usually done after six months of age.

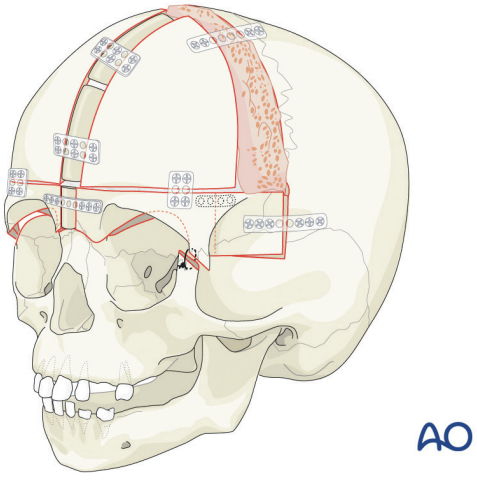


Figure 4.3.4 Skull showing fronto-orbital advancement. AO Surgery Reference, www.aosurgery.org. Reproduced with kind permission. Copyright by AO Foundation, Switzerland.

c) Near-total cranial vault remodeling

Near-total cranial vault remodeling, also known as a midvault remodel, addresses both the front and back areas of the head. It may be used to correct sagittal CS, and it is typically done between four and eight months of age. Figure 4.3.5 shows a near-total cranial vault remodeling in a child with sagittal CS.

Key points Chapter 4

- The treatment for CS is surgical repair.
- Most children with nonsyndromic CS will need a single operation sometime before their first birthday.
- Children with syndromic CS will often require more than one surgery.
- Collaboration with other specialties is needed as well, especially for syndromic CS, and the child may see several team members.
- Surgical repair of CS may be either endoscopic (a less invasive surgery done through small incisions with specialized tools) or open (removal and remodeling of the skull bones through a larger incision).
- The type of surgical repair selected is based on the age of the child at diagnosis as well as the type of CS.
- Risks of surgery include the need for blood transfusion, risks associated with anesthesia, and other more serious risks such as cerebrospinal fluid leak, brain damage, stroke, air embolus, infection, or very rarely, death.

Nobody gets to live life backward.
Look ahead—that's where your future lies.
Ann Landers

In this chapter, people share stories of living with CS, both nonsyndromic and syndromic CS, and one is from a family whose child had deformational plagiocephaly. The stories come from families who are early in their journey as well as those whose children had surgery many years ago. They are the voice of the parents and of an adult living with CS.

Raising a child with a condition such as CS is a journey, and each individual and family will have different needs. Understanding the journeys of others, as described in these stories, may be helpful, but don't judge your own journey against others.

Sheila, mother of four-year-old Evelyn, from Minnesota, US

Evelyn was born with unicoronal craniosynostosis. We noticed right away she had one eye that looked more open than the other. My husband, Brian, thought something was wrong. I just thought because she was breech she was a bit squished inside for so long that it would take a little while to straighten out. The nurses seemed to agree that it would get better in the days after her birth, but we didn't see much change.

At her five-week well-child check, the pediatrician felt a ridge on her skull above her left eye that ran down toward her ear. She had also developed a flat spot on one side of the back of her head. After discussing this with the pediatrician, we decided to take Evelyn to see a craniofacial surgeon to make sure everything was okay. On the way to that appointment, we thought in the worst-case scenario she would need a helmet. We had no idea what we were in for!

Evelyn had a CT scan that showed her sutures were fused, causing one of her eye orbits to be misshapen and her skull to be held back. This news was so scary and devastating. I felt a lot of guilt about things I had done during my pregnancy: Did I have too many chocolate shakes while pregnant? Did this happen because I opted to stay on my antianxiety medication? I also blamed myself for *not* doing some things. The baby had been breech, and I should have been more active. I thought maybe because she was stuck, her head grew wrong! I should have tried to get her to turn more. No matter what anyone said, I really felt deep down that I caused the problem.

I tried to take Evelyn to a new-baby class, but the questions from other moms about diapers and burping just felt so small compared to what I was dealing with. I didn't want anyone to know she had an issue. We didn't even share the diagnosis with extended family right away, as we didn't want people to pity our beautiful new daughter. I wanted to protect her from that.

We had to wait until she was six months old for surgery. The anxiety of waiting during this time was really difficult. I just wanted the surgery over so her poor little brain wouldn't be squished in her skull. I also kept staring at her perfectly beautiful head. It was hard to believe there was a problem underneath the beautiful skin. I didn't want to believe that my husband and I created something that was flawed. I didn't want anything to change for her. She was beautiful the way she was to me.

Between the time of Evelyn's diagnosis and shortly after her surgery, I found it helpful to write about our journey to share with select family and friends through an online blog. We were also able to get connected with other parents who had been through this journey before, and they were so helpful in answering my questions.

Some excerpts of my blog posts help to convey my stress and feelings of anxiety as we neared the surgery day.

> Surgery is only 12 days away. We got some information about the surgery from a nurse yesterday…preparations, what to bring, what to expect. The call lasted a good 30 minutes and was very overwhelming. This mama is feeling anxious. Mainly I don't want her to get sick before surgery so we don't have to postpone. The

waiting has been hard. Luckily, I have a rock-solid hubby and a sweet little two-year-old to hug me all day long. The dog lies by us for protection and support, too.

Surgery is six days away. I'm very worried about Evelyn getting sick and having to postpone. I have decided to quarantine us for the remainder of the week to minimize exposure. It would really stink to have to wait another few weeks after waiting five months for this surgery. This mama's heart could not handle it. I keep thinking of the surgeon's words telling me that she will look different and will have lumps and bumps for the next year or so. It will take a little while to get used to. I am in a bit of mourning over her beautiful head which right now is smooth, perfect, and scarless.

Tomorrow is the day. We have to be at the hospital early in the morning. Her surgery will last three to four hours and after that she will spend the night in the pediatric intensive care unit (PICU). Send up some prayers for us in the morning. I took a picture of her beautiful head of hair. I'm hoping it will grow back fast. I'm trying not to think about tomorrow, but it's all I've been thinking about for a long time. Other moms I have talked to who have gone through this have mentioned the feeling of relief once the surgery is over. I cannot wait for that feeling and to be on the other side of this.

Day of surgery

She is out. I'm holding her as she sleeps, hoses and all. She looks pale and her voice is hoarse. But she is ok.

The surgery Evelyn had was a cranial vault remodeling with a fronto-orbital advancement. It went well, and she had her first haircut by the surgeon! I was so relieved when I was able to hold her afterwards, even though she had so many lines coming out of her tiny body.

The day after surgery

Today she is a little swollen. They say she will look swollen for the next three weeks. She can still open her eyes, so that's good. She will need a blood transfusion today. There were a lot of alarms going off overnight, mostly with her moving around and kicking. The best thing that happened overnight was the 4 a.m. party. Evelyn threw it for herself… babbling and kicking and playing with toys. It seemed like the old Evelyn was back. This morning she has been enjoying snuggles and sleeping on mom and dad. She is so sweet. I'm hoping she will want to drink milk again soon. She did eat oatmeal for me. These things take time, they say.

Two days after surgery

No more drains!* Yay! Evelyn is detached from everything and only has one IV needle taped to her right hand. She started to nurse again. Woohoo! She is doing well with her pain and swelling. She has been sleeping great, although staying in the hospital can mix everybody's nights and days up. I'm so grateful and proud today. Evelyn has handled this whole thing with such grace. We have a lot to be thankful for.

Three days after surgery

Home! Evelyn did so well that we got to take her home this morning. Her stitches are dissolvable so that is good. We have a follow-up in three weeks and then again in about six months. We are so happy that things will return to normal. We have to keep her big brother careful and gentle around her, but overall we're back to normal. Amazing.

* Surgical drains may be used following surgery. They are small plastic tubes/devices to collect fluid from around the incision area (draining it instead into the tube). Most are removed two to three days after surgery.

Two weeks after surgery

> We have been home for a while now and Evelyn is healing nicely. She does have some lumps and bumps, but she is overall cheerful and off all pain medicine. She is back to eating solid foods, but her sleep is back to newborn status; awake every two to three hours. Evelyn does not want to sleep without being held right now, understandably. It makes for a tired mama.

Evelyn's recovery continued without any issues. At around age two, we had to bring her back to see the craniofacial surgeon because her head didn't seem to be growing much and she was having some problems sleeping. However, everything checked out okay and she has not had any other issues since.

Today, Evelyn is a happy, healthy four-year-old. She loves reading, dancing, princesses, music, art, and rainbows. She is highly coordinated and can already pedal a bike with no training wheels. She has done many things at an earlier age than her older brother did. She will need one last surgery to correct a couple of small areas on either side of her forehead. Small amounts of filler will be put in to even out the appearance of her skull. Luckily, this is a short, one-night stay and we will get this done before she starts kindergarten. She is amazingly optimistic and the toughest person we know.

Evelyn, six months old, the morning of her surgery.

Éamon, father of 10-year-old Éanna, from Limerick, Ireland

We call Éanna our little miracle man.

We had been trying to have children for over eight years when Éanna was born. At that time, I was nearly 50 years old and Rosemarie was in her mid-40s. We had gone through the plethora of fertility treatments—IVF,* donor egg, etc.—to no avail and had all but given up hope of being parents. But then one day, a month or so before my birthday, Rosemarie walked into the house and announced she was pregnant. We were over the moon and bursting with excitement and anticipation.

The pregnancy was by and large a good one with regular scans showing no issues or concerns until week 18 when bleeding near the placenta was noticed and again at week 30. Our consultant physician informed us that the rate of growth had slowed and was not following the normal trajectory. She recommended a series of tests, including an amniocentesis (a test that collects a sample of the amniotic fluid that surrounds the baby during pregnancy), which caused some worry for us as it was used to screen for various syndromes.

Going through this was difficult for both of us. For me, the week of waiting for the results was the longest and most agonizing of my life, with little or no sleep and fearing the worst. Thankfully, these tests didn't identify any specific syndromes but still didn't explain the slowdown in growth. So, while there was a certain relief with the results, we were still beset with constant worry.

What followed next was weekly scans with close monitoring and eventually Rosemarie being admitted to the maternity hospital at 36 weeks pregnant. Éanna was born at 38 weeks gestation by cesarean section, weighing just 5 lb, 9 oz (2,534 grams). Seconds after he was born, I was allowed into the operating room where the cesarean section was performed and was relieved to hear the wailing of a newborn baby, one with a beautiful and angelic face. However, immediately we could see

* IVF, or in vitro fertilization, is a type of fertility treatment that is done by manually combining egg and sperm in a laboratory dish to create an embryo. The embryo is then transferred to the uterus.[227]

that Éanna's head was relatively small and odd shaped, having a long head, chin to top, as if he had a chef's hat on. Within hours of his birth, he needed to be transferred to the children's hospital where he spent the next 10 days undergoing all sorts of tests. His poor little foot was like a pincushion with all the blood samples he had to provide. However, through it all, when I looked at his little face, I sensed a resilience and a business-like attitude as we went from appointment to appointment. He was alert, engaging with his surroundings, and he bonded with me immediately (Rosemarie had to stay in the maternity hospital for medical reasons, and the separation was naturally very distressing). I instinctively sensed there and then that everything was going to be okay.

The tests revealed that Éanna had craniosynostosis. In Éanna's case, his type of craniosynostosis involved multiple sutures, namely the metopic, coronal, and sagittal sutures. The resulting skull shape was trigonoturry-brachycephaly. We learned that Éanna's craniosynostosis was most likely due to a gene mutation.

The doctors explained to us that craniosynostosis can affect a child's brain and development. If left untreated, it can lead to increased pressure in the skull and serious developmental problems, such as not being able to read and requiring intensive special education. (In that respect we have been very lucky as Éanna's remaining issues are primarily cosmetic.)

When Éanna was a few months old, Rosemarie enrolled him in an early-intervention program near our home where he continued for several years. This program helped with his gross and fine motor skills, speech, and general development. Éanna progressed so much that they recommended discharging him when he was seven years old. In fact, Éanna himself by then was questioning why he had to attend the sessions.

Éanna attends mainstream school and is just above the middle of the pack academically, which is exactly where we would like him to be, craniosynostosis or no craniosynostosis. In other aspects he has outperformed his peers, being able to both swim and ride a two-wheeled bike at just three and a half years old.

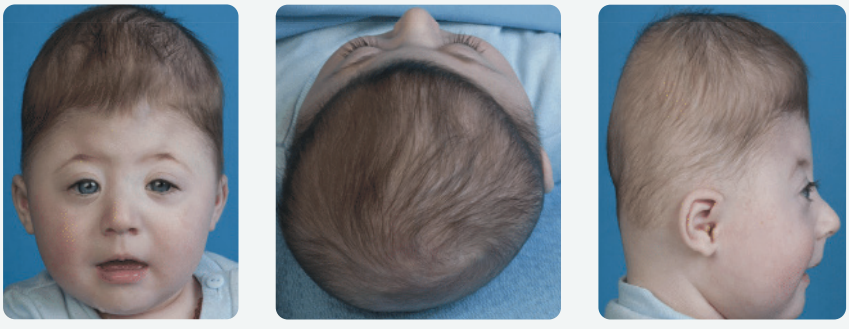
He has had three surgeries on his head: the first two were to create room for his brain to grow and the third was mainly cosmetic. It is only now that I can bring myself to read the detail of those operations. I deliberately chose not to know what was involved in the surgeries at the time. After each, his head was bandaged, his eyes were swollen, and his surgical wounds needed monitoring and cleaning.

Fortunately, the first two surgeries were completely successful. The third one achieved its objective in terms of reshaping his head, but it stretched his scalp too much and killed some hair follicles, resulting in a bald patch at the back of his head. A final operation, called "tissue expansion," is scheduled in summer 2024 to address this. If successful, we hope Éanna will be like any other young boy going into secondary school, both in appearance and intellect.

Éanna continues with regular monitoring for any impact on his eyes, ears, nose, and hormone levels. These regular follow-up appointments will continue until he is in his late teens.

Éanna is now 10 years old. Throughout the surgeries, follow-ups, and various other appointments, he has shown resilience and determination way beyond his years. He attends a great school with brilliant and caring teachers. One of our fears was that he would be bullied or teased because of his appearance, in particular the bald patch at the back of his head, but to this point that has not been an issue. He is very popular and has very good friends who know him as the soccer-mad little man with the hat! The odd time when he is outside his immediate familiar environment, some people have passed a remark, but his attitude has always been "It doesn't bother me. If they have a problem, it is their problem, not mine."

We are so, so proud of him, love him dearly, and are so thankful to have him in our lives.



Éanna at six months of age.

Éanna at 10 years of age with his parents, Rosemarie and Éamon.

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Abbreviations used in index: CS craniosynostosis, DP deformational plagiocephaly. Figures and tables indicated by page numbers in italics.

"The book I wish I had when my son was diagnosed with sagittal craniosynostosis."

—ELAINE L. KINSELLA, PARENT

"A remarkably comprehensive review of all things related to craniosynostosis."

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Craniosynostosis is a condition where the bones of an infant's skull fuse together too early. In most cases, surgery in the first year of life will effectively correct it and the child can go on to expect a typical life. For a minority, craniosynostosis is part of a syndrome, which is a lifelong condition. This practical guide explains how craniosynostosis develops and the evidence-based, best-practice treatments. It also includes the lived experience of families.

The writing of *Craniosynostosis* was led by Ruth Barta, MD, Craniofacial and Pediatric Plastic Surgeon at Gillette Children's, a world-renowned center of excellence for the treatment of brain, bone, and movement conditions. *Craniosynostosis* is part of the **Gillette Children's Healthcare Series**, a series of books for families who are looking for clear, comprehensive information. Health care professionals, researchers, educators, students, and extended family members will also benefit from reading *Craniosynostosis*.

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