"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

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GILLETTE CHILDREN'S

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).

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]

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.

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.

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* 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]

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.

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]

At Keegan's two-week well-child doctor appointment, the pediatrician greeted us with pleasantries, but I noticed he was quite focused on looking at Keegan. He stepped out saying he would be right back, and when he returned, he brought with him a figure chart. He felt Keegan's head and said there was a ridge on top from where the front soft spot area was supposed to be toward the back of his head. Keegan had a prominent forehead, and the back of his head was quite narrow in comparison. We knew Keegan's head had a different shape but did not feel that was out of the ordinary compared to other babies, as not many have perfectly circular heads after vaginal deliveries.

The pediatrician said he suspected craniosynostosis. My immediate thought was that he would just need to wear a helmet. But the pediatrician then told us that surgical correction is usually needed. He recommended following up for further evaluation in two weeks, at the one-month well-child doctor appointment. The news was not exactly something we wanted to hear, but we remained optimistic.

During the wait for the next appointment, I researched online the new words we had learned: "scaphocephaly" and "craniosynostosis." Each website I looked at reiterated the need for surgery and the possibility that craniosynostosis could be linked to different syndromes and developmental issues.

At the one-month visit, Keegan was happy, healthy, and gaining weight like crazy. However, his head shape was still concerning, and the pediatrician referred us to a craniofacial surgeon for evaluation. I broke down and cried, feeling this confirmed that our sweet little innocent, precious baby would need surgery. I'll never forget the sound of my tears falling on the paper liner on the exam table.

"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."

—CHRISTOPHER R. FORREST, MD

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 Dr. 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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