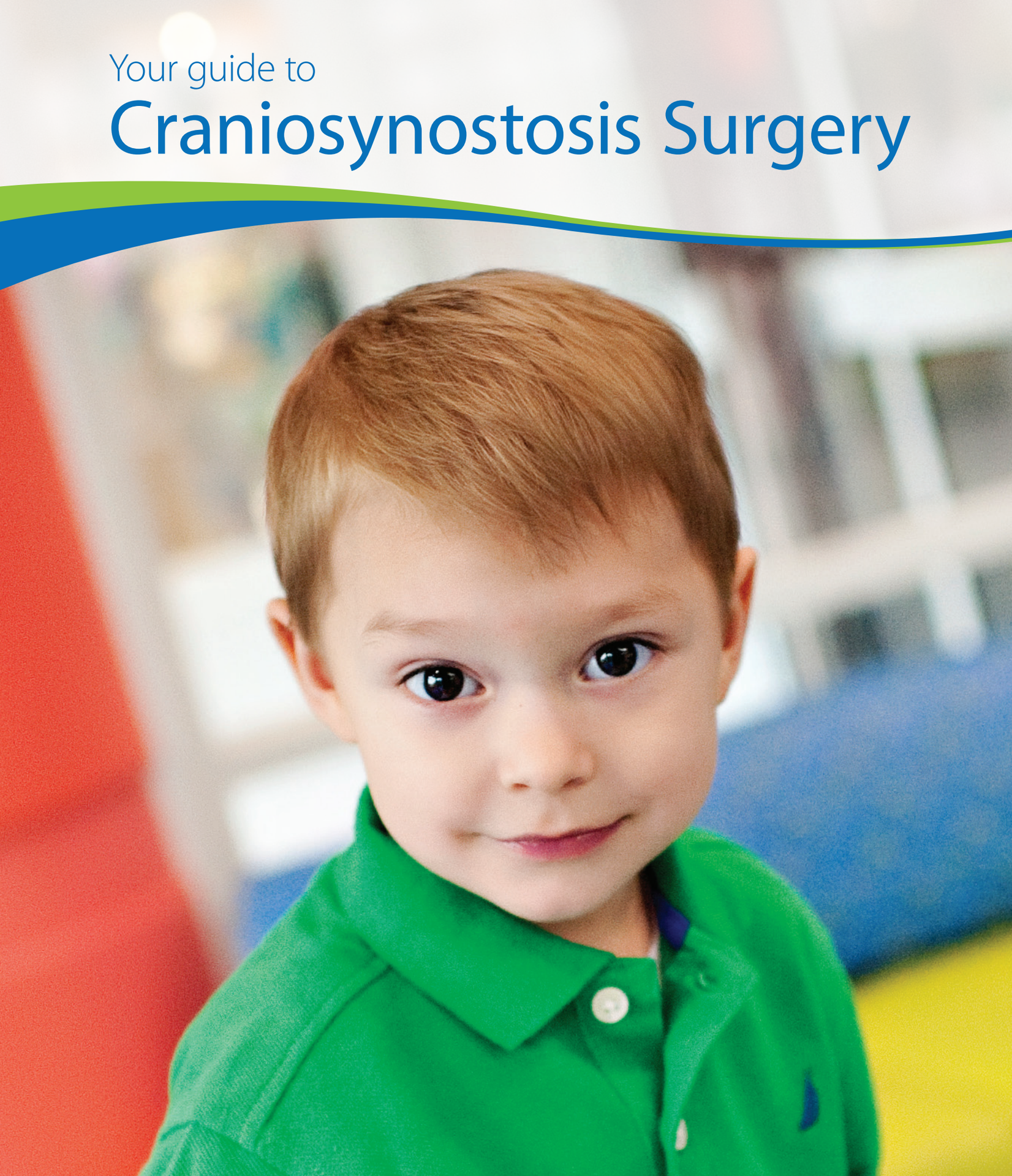
Your guide to

# Craniosynostosis Surgery



## Craniofacial Center

601 Children's Lane, Norfolk, Virginia 23507 | CHKD.org/craniofacial

# Welcome to the CHKD Craniofacial Center

You have been given this booklet because your child has been diagnosed with craniosynostosis. We understand the confusion and concern you and your family may be experiencing. Our goal is to help educate you about your child's condition, ease your anxiety and provide information to prepare you for your child's appointments and course of treatment. It is important that you understand your child's diagnosis since you are a critical member of your child's craniofacial care team.

For your reference, we have included a glossary of terms that may be helpful at the end of this booklet and several additional handouts that provide detailed information about related topics.

# Table of Contents



Call our CHKD Craniofacial Center coordinator, Karen Via, at **(757) 668-7031** or **(757) 668-7713** for more information or to ask about referrals.

---


**Craniofacial Center Medical Director**
Jesus Gil B. Inciong, MD

**Craniofacial Center Coordinator**
Karen Via, MSN, RN



This guide is made possible by

American Legion Child Welfare Foundation

# Craniofacial team

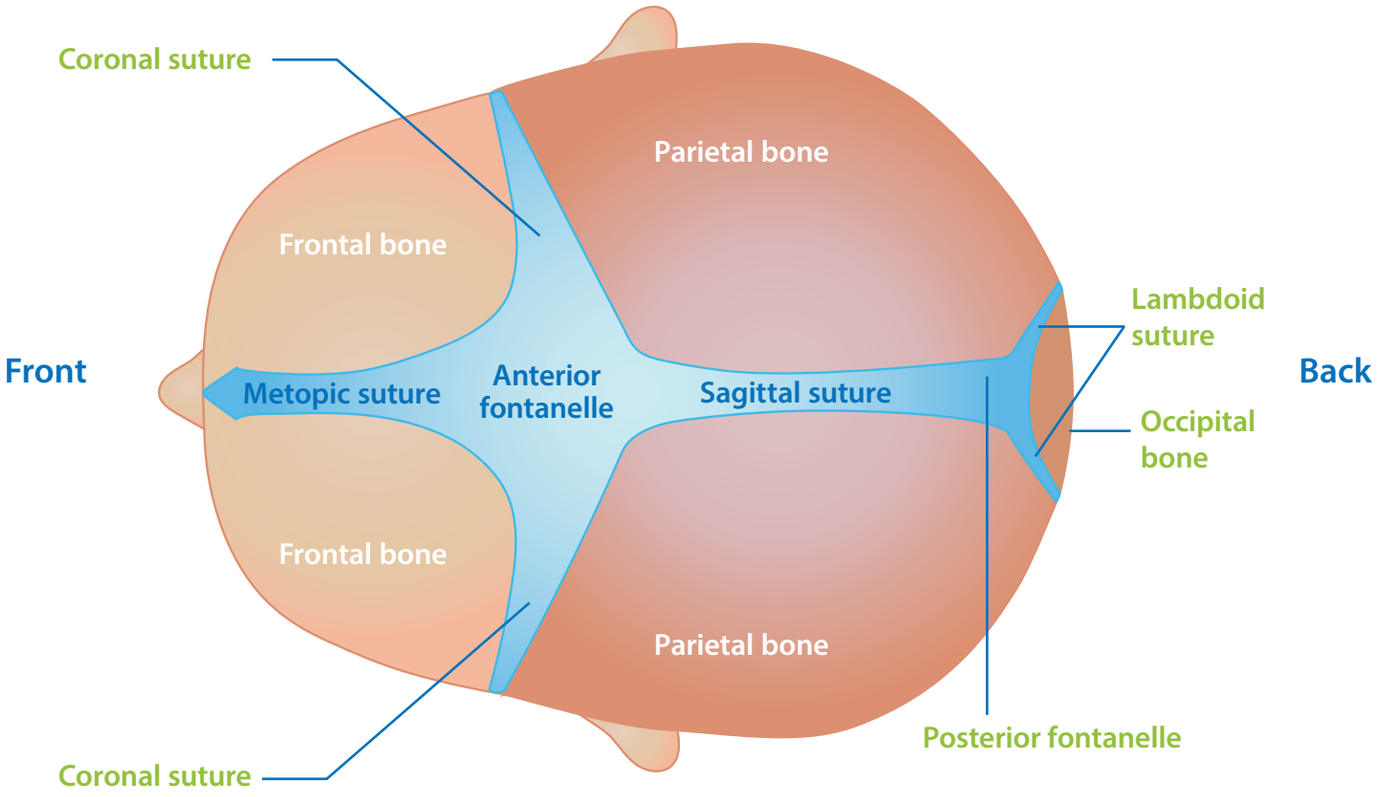
At CHKD, children with craniofacial abnormalities receive comprehensive care from a multidisciplinary team of specialists. We provide access to a full array of pediatric specialty care including pediatric plastic surgery, neurosurgery, ENT, audiology, dentistry, psychology, general pediatrics, medical genetics, ophthalmology, orthodontics, speech pathology, social work and nursing.

Your child's diagnosis will determine his craniofacial team members, and a craniofacial coordinator will serve as a facilitator between your family, referring physicians, and your CHKD craniofacial team. We know you will have many questions throughout this process, and we are here to provide answers. We encourage open, ongoing dialogue and welcome questions via email or phone at any time.

# The newborn skull

To understand craniosynostosis, it is helpful to understand the anatomy of a newborn's skull. A child's skull is made up of several major bones that are connected by sutures. The space between the bones of an infant's skull, where the sutures intersect, is called a fontanelle.

Below is a diagram highlighting the major bones, fontanelles and sutures of a newborn's skull.




**Sutures** give the skull flexibility to move during the birth process. They then act like a growth center, allowing the skull to expand symmetrically and evenly as the child's brain grows.

Over time, the sutures close, the bones fuse together, and the skull becomes a solid piece of bone. The anterior fontanelle is the area on the top of the head, often called the soft spot, where the two frontal bones of the skull and two parietal bones eventually meet. This area remains soft until about 18 months to 2 years of age. The posterior fontanelle is a small soft spot near the back of the skull where the large parietal bones meet the occipital bone at the very back of the head. This smaller fontanelle near the back of the head usually closes during the first few months of an infant's life.

# Craniosynostosis

## What is craniosynostosis?

Craniosynostosis occurs when one or more of the sutures of the skull close too early, causing problems with skull growth and skull shape. The condition is not uncommon – it occurs in 1 in every 2,000 children born in the United States and affects boys slightly more often than girls. Craniosynostosis is usually noticeable at birth and becomes more apparent during the first few months of your baby's life.

The following may be signs of craniosynostosis:

- Misshapen skull.
- Abnormal or missing soft spot (anterior fontanelle).
- Development of a raised, hard ridge along one of the skull's sutures.
- Slow or no growth of the head.

*Tucker, 5 months old, before surgery*

*Tucker, 7 months old, after surgery*

## What causes craniosynostosis?

The first thing to understand is that you did not do anything to cause your child's condition. In most cases, the cause of a child's craniosynostosis is unknown. In other cases, certain genetic syndromes can affect your baby's skull development. If necessary, your pediatrician or craniofacial team will order genetic testing.

### *Cooper, 22 months*

*– Newport News, VA*

When Cooper was 3 months old, his mom, Brittany, became concerned about the shape of his head. At the recommendation of a family member, she brought Cooper to CHKD where he was diagnosed with metopic craniosynostosis. Three months later, Cooper underwent reconstructive surgery. Today, Cooper is a happy, healthy toddler.

*"It's amazing. To look at him today, you would never be able to tell he had this major surgery."*

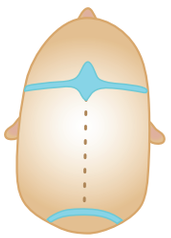
*– Brittany, Cooper's mom*

# Types of craniosynostosis

There are many types of craniosynostosis. Different names are given to the various types based on which suture(s) are involved and how the shape of the skull is affected. Common types of craniosynostosis are illustrated below. The dotted lines represent the sutures that have closed prematurely, while the areas in blue represent the sutures that are open.

## Scaphocephaly (sagittal synostosis)

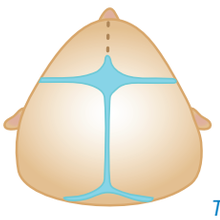
This is the most common type of craniosynostosis. The skull is long from front to back and narrow from ear to ear. It is caused by the closing of the sagittal suture, which runs front to back, down the middle of the top of the head.



*Scaphocephaly*

## Trigonocephaly (metopic synostosis)

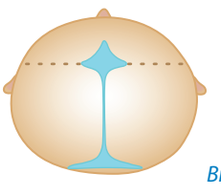
This type of craniosynostosis causes a vertical ridge to develop on the forehead. The eyes may be close together, and the forehead may look pointed and narrow. It is caused by fusion of the forehead (metopic) suture. This suture runs from the top of the head down the middle of the forehead, toward the nose.



*Trigonocephaly*

## Brachycephaly (bicoronal synostosis)

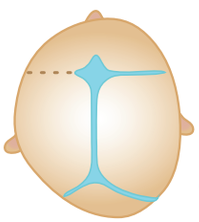
When both of the coronal sutures fuse prematurely, your baby's head may appear taller, but flatter and wider, and most commonly with a forehead tilted forward.



*Brachycephaly*

## Plagiocephaly (unicoronal synostosis)

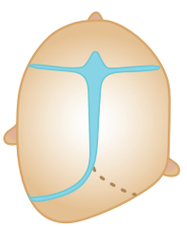
Premature closing of one of the coronal sutures that run from the ear to the very top of the skull may cause the forehead to flatten on the affected side and bulge on the unaffected side.



*Plagiocephaly*

## Lambdoid synostosis (posterior plagiocephaly)

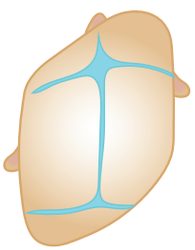
In lambdoid synostosis, the lambdoid suture prematurely closes causing a flattening of the back of the head on the side where the suture has fused. This is the rarest type of craniosynostosis.



*Lambdoid synostosis*

## About positional plagiocephaly

In positional plagiocephaly, the infant has an asymmetrically shaped head with the skull flattened in one area. This is caused by pressure on the bones of the skull before or after birth and does not require surgery to correct. In this condition, unlike craniosynostosis, the skull bones have not fused prematurely. This is often caused by positioning babies on their back for sleep, as is recommended by all pediatricians.



*Positional plagiocephaly*

## Other symptoms of craniosynostosis

If your pediatrician suspects your child may have craniosynostosis, you will be referred to a specialist for further examination. Some additional indications of the condition may include:

- Asymmetrical facial features.
- Increased intracranial pressure.

  This will be more significant if more than one suture is fused, but your child's brain is usually spared any damage from increased pressure because the other skull bones will expand to accommodate the growing brain.

## How is craniosynostosis diagnosed?

A CT scan is needed to confirm a diagnosis of craniosynostosis. (See the handout on CT scans included in this folder.) If your pediatrician has not already ordered a CT scan, the craniofacial team at CHKD will provide you with an order and take care of getting the insurance authorization needed for the CT scan.

We will make every attempt to schedule the CT scan and your follow-up appointment with the pediatric plastic surgeon on the same day.

***Waylon, 15 months***

*– Topping, VA*

At Waylon's 4-month well-baby checkup, his pediatrician noticed his forehead appeared abnormally pointed. He referred Waylon to CHKD's Craniofacial Center. A CT scan confirmed Waylon had metopic craniosynostosis - a fusion of the suture running from the top of his head down to his nose. At 8 months old, Waylon underwent surgery to correct his condition. Today Waylon is meeting all his developmental milestones, and his parents are very satisfied with the results of his procedure.

*"The whole care team at CHKD was wonderful. I can't thank them enough for helping us through this experience."*

*– April, Waylon's mom*

## What to expect at your first appointment

CHKD's Craniofacial Center team will schedule or provide a thorough exam during your first appointment. The team includes the pediatric plastic surgeon, neurosurgeon, nurse/clinical coordinator and social worker.

At this first meeting, please be prepared to share your child's full medical history, including any:

- Bleeding disorders within the family.
- Medications your child is currently taking.
- Allergies to medication, foods, latex (rubber) or the environment.

During this appointment, you will meet the pediatric plastic surgeon and neurosurgeon who will work together during the surgery. The surgeons will provide you with details about the procedure, review the CT scan with you and answer any questions you may have. They will present the options available for your child and help you choose the most appropriate procedure depending on your child's age and the specifics of their individual case.

You will also be given the option of scheduling a preoperative tour to better prepare you and your child for surgery.

# Surgery

## Scheduling surgery and a tour

The key to treating craniosynostosis is early detection and treatment. Treatment depends on each child's symptoms, the severity of their condition, their age and general health.

It is usually best for babies to have the surgery before their first birthdays, while the bones of the skull are still very soft. Most surgeries can be done as early as 3 to 8 months. Your surgeon will provide you with instructions on how to prepare your child during the weeks and months leading up to surgery.

The surgery scheduler for the pediatric plastic surgeon will be responsible for scheduling the surgery and obtaining insurance authorization. It can take several weeks to coordinate the schedules of the pediatric plastic surgeon and the neurosurgeon before finalizing a date for your child's surgery. Call (757) 668-7713 to speak with the surgery scheduler if you have questions about the timing of your child's surgery.

Child life specialists at CHKD host evening tours for patients and parents to help prepare you for a successful surgery experience. The staff will answer your questions, provide a tour of the facilities and explain medical equipment to help take the anxiety out of surgery day. Please call the child life tour line at (757) 668-6748 for further information.

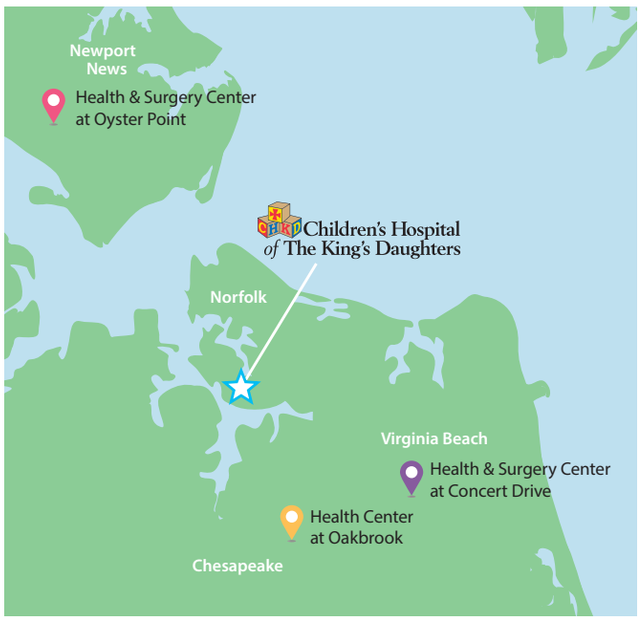
***Zyniq, 12 months***

*– Norfolk, VA*

Zyniq was diagnosed with Apert syndrome and bicoronal craniosynostosis. CHKD plastic surgeon, Dr. Jegit Inciong, examines Zyniq and explains her upcoming procedure to her mom, Natosha, at an appointment in the CHKD Craniofacial Center.

# Preparing for surgery

Before surgery, your child will have another appointment with the surgeon. At this time, there will be a discussion regarding the surgical procedure and consents will be signed. Your surgeon will order lab work which you will need to complete at least five working days prior to the surgery date. You can go to the outpatient lab at any of the following CHKD locations. No appointment is necessary.




- CHKD Main Hospital,
  601 Children's Lane, Norfolk,
  Monday–Friday, 8 a.m. - 8 p.m.
- CHKD Health and Surgery Center at Oyster Point,
  11783 Rock Landing Drive, Newport News,
  Monday–Friday, 8:30 a.m. - 5 p.m.
- CHKD Health Center at Oakbrooke,
  500 Discovery Drive, Chesapeake,
  Monday–Friday, 8:30 a.m. - 5 p.m.
- CHKD Health and Surgery Center at Concert Drive,
  2021 Concert Drive, Virginia Beach,
  Monday–Friday, 8:30 a.m. - 5 p.m.

If you have other children, please attempt to make arrangements for their care on surgery day. No other children are allowed in the pre-op and recovery areas. If you are having difficulty making these arrangements, contact the scheduling office at (757) 668-7332.

You will get a call a day or two before surgery to give you important information about the time you must arrive at the hospital the morning of surgery. Instructions for drinking and eating before surgery will also be given to you. **Your child's stomach must be completely empty before surgery for safety reasons related to anesthesia. Any food, water or formula increases the risk of serious complications during surgery, so it is important you write down the instructions and follow them.** If for some reason you do not receive a call by the day before surgery, please call (757) 668-7332.

Please plan for your child to spend three to five days in the hospital after surgery. On the morning of surgery, be sure to bring the following with you to the hospital:

- Your photo ID.
- All papers from your physician, guardian papers and insurance card.
- The name of any medications and doses your child may be taking.
- Change of clothes for discharge home.
- Patients with feeding tubes, tracheostomies or other special medical needs, should bring any necessary medical equipment .
- A favorite toy, blanket, music box or pacifier.
- Favorite bottle or cup for after surgery.

# Morning of surgery

When you arrive at CHKD, go to the welcome desk in the main lobby to obtain a visitor's badge. Only two adults may accompany your child to the day surgery room and recovery unit. One of these adults must be the child's legal guardian. If there are additional adults accompanying you the morning of surgery, they may wait for you in the surgical waiting room on the third floor.

After you receive your badge, please go to the day surgery department on the third floor where you will register for surgery. Your child will be given a short exam and hospital pajamas to change into. You will meet the anesthesiologist and the surgeons before the procedure begins. (See the handout on anesthesia included in this folder.)

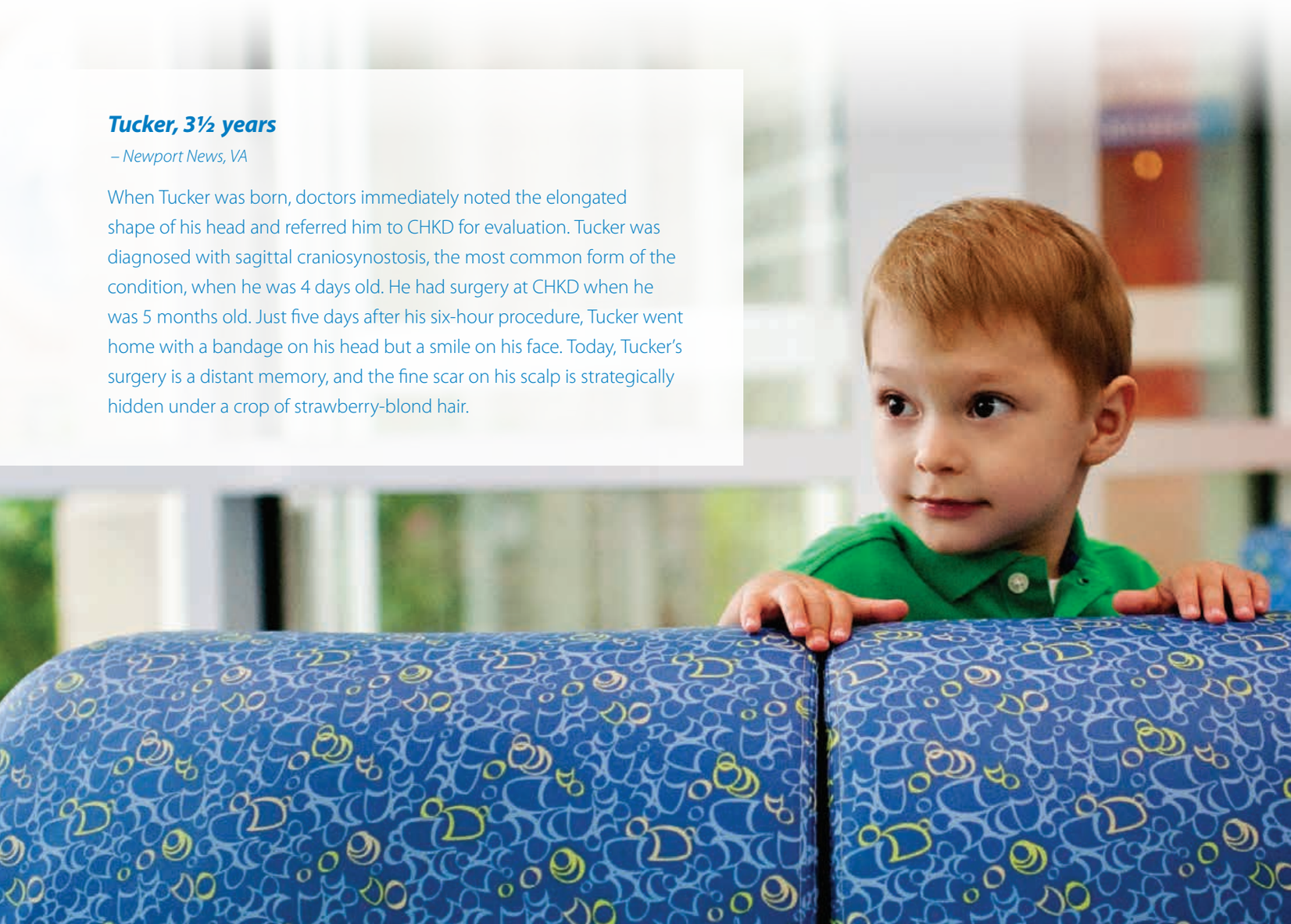
When it is time for surgery, your child will be carried into the operating room in the arms of a nurse. At the entrance to the hall leading to the operating room, you will have a chance to give your child a hug and a kiss before the surgery.

You can then move to the nearby surgery waiting room and check in with the receptionist at the desk. This is a great time to get something to eat, but please do not leave the facility. The operating room staff will call the surgery waiting area with updates for you every hour.

***Tucker, 3½ years***

*– Newport News, VA*

When Tucker was born, doctors immediately noted the elongated shape of his head and referred him to CHKD for evaluation. Tucker was diagnosed with sagittal craniosynostosis, the most common form of the condition, when he was 4 days old. He had surgery at CHKD when he was 5 months old. Just five days after his six-hour procedure, Tucker went home with a bandage on his head but a smile on his face. Today, Tucker's surgery is a distant memory, and the fine scar on his scalp is strategically hidden under a crop of strawberry-blond hair.

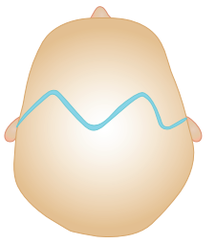


# During the procedure

The surgeon will make a zigzag incision over the top of your child's head, from ear to ear, in order to remove a portion of the skull. This will be separated into several pieces, which are reshaped and then placed in their new positions with the help of tiny absorbable plates and screws.

During surgery, it is standard procedure to have blood available for transfusion if needed. If you would like, you may be able to donate your blood for your child. (See the handout on these types of blood transfusions in this folder.)

**Bicoronal incision**



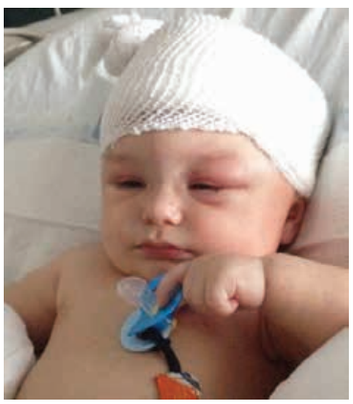
The surgeons will estimate the time it will take to complete the surgery. You will be provided with an update on the surgery once every hour. There are many activities that take place in the operating room, so if your child's procedure takes a little longer than expected, there is no need to be concerned. Your surgical team will contact you as soon as your child is recovering.

# After surgery

After surgery, patients go to the post-anesthesia care unit (PACU) to be monitored as the anesthesia wears off. As soon as they become appropriately alert, parents are able to join them in the PACU. The PACU nurses will be constantly monitoring your child's intravenous fluids (IV fluids) and vital signs (such as blood pressure, pulse, temperature and breathing). Your child will receive medication through the IV. Once your child is stable, he will be transferred to the pediatric intensive care unit (PICU).

Your child will have a gauze dressing around the skull which resembles a turban. This dressing will remain on for two days. In addition, significant swelling called periorbital edema is expected around the face and eyes, which lasts for several days. The eyes become swollen over the first 12-24 hours and may be swollen shut for two to three days. Parents' voices and other familiar music and sounds may help comfort your child during this time. Your child will also have several IV lines and a urinary catheter.

*Tucker, age 5 months old, after surgery*



*Day 1*

*Day 2*

*Day 4*

# Staying in the hospital

Children spend the first night and occasionally the second night after surgery in CHKD's pediatric intensive care unit (PICU). (See Welcome to PICU Fact Sheet included in folder.) There are a number of people who may care for your child in addition to the surgeon and the bedside nurse in the PICU, including the pediatric intensive care doctor, a resident physician, nursing assistants, respiratory therapists, social workers and child life specialists.

Only two visitors per patient are allowed in the PICU. PICU nurses will monitor your child's vital signs, oral intake, output and IV fluids. Your child will be given an antibiotic through an IV and frequent pain medications either by mouth or IV to ensure comfort. Your child's heart will be monitored through sticky pads on the chest and a red light on a finger or toe will check the oxygen level in your child's blood. Every six hours for up to two days, a blood sample will be taken to make sure your child is recovering as well as expected. The nurses will assist you in holding your baby on the first day after surgery. You will also be able to feed your baby by breast or bottle at this time.

In most cases, your child will be moved to the general floor the day after surgery. Beds are available in these rooms for one parent to sleep at the bedside. On the second day after surgery, the head dressing will be removed, and the nurses will begin instructing you on how to care for the incision on your child's scalp. It is not uncommon for your child to have a low-grade fever of 99 to 100 degrees for a few days after surgery.

## Care at home

Your child will be discharged four or five days after surgery. You will be given a prescription for an antibiotic and for pain medication. You will be told how to care for the incision and will be given the supplies to continue the incision care for one week after surgery. Things to remember:

- You can wash your child's incision each day with a mild shampoo, but do not soak the area of the incision in the tub.
- The incision will need about two weeks to heal.
- Cover your child's head when going outside and use sunscreen at all times as the incision will easily get sunburned. Sunscreen protects from harmful burns and prevents the scar from darkening.
- The sutures are absorbable and will not have to be removed by the surgeon.
- We recommend you keep your child home from school or daycare until after your first follow-up visit.

Once your child is home, it is fine to resume a regular diet and activity level. Remove low-lying furniture with sharp edges such as coffee tables to prevent possible head injuries.

## Scars

Scars may seem to get more noticeable before they get better. For about six weeks after surgery, the scar will continue to become red, firm and hard. Over the next four months, it will soften and lose the redness. This is the body's normal process of scarring. Although scars remain forever, typically the scar will blend into the normal skin creases so that it is hardly noticeable six months after surgery. Every scar is different, and some may not follow this exact timeline. It can take up to two years for some severe scars to fully heal. (See handout on scars in this folder.)

## Follow-up visit

Your child will have a follow-up visit with the surgeon approximately 10 days after surgery, followed by another appointment six weeks after surgery. You can contact the craniofacial office at (757) 668-7031 or (757) 668-7713 to schedule these appointments. The swelling around your child's head and eyes should decrease each day.

### If you notice any of the following complications, contact your surgeon:

- Fever greater than 101 degrees.
- Frequent vomiting.
- Increased irritability.
- Decreased alertness.
- Increased redness, swelling or drainage from the incision.
- Confusion or excessive sleepiness.

*"Tucker is now 5 years old and thriving. We're so fortunate to live near CHKD and have this amazing resource for our children. I'm forever grateful."*

*– Kristin, Tucker's mom.*

# Glossary of terms related to craniosynostosis

**Acetaminophen:** A pain reliever and a fever reducer.

**Anesthesia:** A medication used to induce a temporary loss of awareness.

**Antibiotic:** Medicine used in the treatment and prevention of bacterial infection.

**Arterial line:** A thin catheter inserted into an artery to monitor blood pressure directly and in real-time.

**Asymmetry:** Absence of symmetry or when two halves do not match.

**Bicoronal incision:** Ear-to-ear incision.

**Coronal suture:** The suture that goes from ear to ear.

**Craniofacial reconstruction:** A procedure used to reshape or repair the skull.

**Cranioplasty:** A surgical repair of a bone defect in the skull.

**Cranium:** The part of the skull which encloses the brain.

**CT scan:** A computerized tomography scan combines a series of X-rays to provide more detailed information than plain X-rays.

**Catheter:** A tube inserted into the bladder to drain urine.

**Frontal bossing:** An unusually prominent forehead.

**Intravenous fluid:** The administration of fluids through a blood vessel, usually a vein.

**Intubation:** A process to place a tube into the airway during surgery.

**Lambdoid suture:** The suture that goes across the back of the head.

**Medical geneticist:** A specialist who provides diagnosis and treatment for patients with genetic disorders.

**Metopic suture:** The suture that goes from the top of the head down the middle of the forehead.

**Narcotic:** Medicine used to relieve pain.

**NPO:** "Nothing by mouth," medical instruction to withhold food and fluids from a patient, especially prior to surgery.

**Ophthalmologist:** Surgeon who specializes in medical and surgical eye problems.

**Otorhinolaryngologist (ENT specialist):** A physician who specializes in ear, nose, and throat.

**PACU:** Post-anesthesia recovery room, provides care for patients recovering from various types of anesthesia.

**Periorbital edema:** Swelling of the tissues around the eyes that may appear after surgery.

**PICU:** Pediatric intensive care unit, provides care for children who are critically ill or have had major surgery.

**Pulse oximeter:** Device placed on a fingertip, earlobe or foot (for an infant) to monitor blood oxygen levels.

**Sagittal suture:** The suture that goes from the front of the head to the back, down the middle of the top of the head.

**Speech pathologist:** A therapist who specializes in language, speech and voice.

**Syndrome:** A set of medical symptoms that are correlated with each other.