Craniosynostosis: Caring for Infants and Their Families

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Craniosynostosis is a developmental anomaly with premature closure of the cranial sutures causing an abnormally shaped skull in an infant. Recommended surgical treatment involves cranial vault reconstruction to open the closed suture, increase intracranial volume, and allow the brain to grow normally. Parents work with a multidisciplinary team during the evaluation process and face various preoperative and postoperative stressors. Critical care nurses can improve the care of the infants and their families by being knowledgeable about the anatomy, assessment, and surgical and nursing management of infants with this anomaly and its impact on the patients' families. This article discusses the definitions, diagnosis, and treatment of craniosynostosis and support for parents of infants with this malformation. (*Critical Care Nurse*. 2013;33[4]:39-51)

Craniosynostosis is a craniofacial malformation in which one or more sutures of the cranial vault are fused prematurely.[1] The cranial vault evolves during embryonic life, and malformations become evident at birth. Craniosynostosis occurs as an isolated condition or as part of a syndrome. It manifests itself in association with 130 different syndromes, but most patients are nonsyndromic.[2,3] Syndromic craniosynostosis is accompanied by other body deformities and involves multiple systems. Genetic and environmental factors such as single gene mutations, exposure to teratogens, and fetal constraint due to biomechanical forces play a role in the development of craniosynostosis.[1,3] Secondary craniosynostosis occurs when suture fusion is associated with other disorders such as thalassemia, hyperthyroidism, hematologic or metabolic syndromes, or placement of a cerebral shunt. In these instances, abnormal formation of the skull may lead to sutural obliteration or failure of brain growth, or intracranial decompression with shunting for hydrocephalus can lead to collapse of the cranial vault and early fusion of sutures.[1,4,5]

CNE Continuing Nursing Education

This article has been designated for CNE credit. A closed-book, multiple-choice examination follows this article, which tests your knowledge of the following objectives:

1. State the physical examination and radiologic findings used to diagnose craniosynostosis
2. Describe the postoperative nursing care for children undergoing surgical repair of craniosynostosis
3. List 4 potential complications following surgical repair of a craniosynostosis defect

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The incidence of craniosynostosis is 1 in 2000 to 3000 births.[2,6] The premature ossification of cranial sutures results in several morphological abnormalities, such as a dysmorphic cranial vault and facial asymmetry.[1] The sagittal suture is the most commonly affected suture (60% of children); next, in order, are the coronal (25%) and metopic (15%) sutures. Lambdoid craniosynostosis is the rarest form of the disease (2% of children).[6,7] Additional consequences of all forms of craniosynostosis include deformities in the cranial base, midface deficits, dental malocclusion, visual impairment, potential increase of intracranial pressure, and problems with self-esteem and cognitive deficits.[1,7] Critical care nurses involved in treatment of infants with craniosynostosis can improve the care of the infants and the infants' families by being knowledgeable about the anatomy, assessment, and surgical management of craniosynostosis and the impact of the malformation on the patients' families. In this article, I discuss the definitions, diagnosis, and treatment of craniosynostosis and support for parents of infants with this malformation.

Craniosynostosis is a craniofacial malformation in which one or more sutures of the cranial vault are fused prematurely.

Pathophysiology

The cranial bones form during the first weeks of fetal life through a process of membranous ossification (the hardening or calcification of soft tissue into bonelike material) of the mesenchyme (connective tissue derived from the embryological layer called the mesoderm). This process continues during the second and third months of fetal life.[1,8] Most other bones in the body form by the ossification of a preexisting cartilaginous matrix (endochondral ossification).[9]

The skull in the fetus consists mainly of 4 bones: the frontal, the parietal, and the squamosal and anterior parts of the occipital bone. The cranial bones undergo intramembranous ossification from a layer of mesenchyme located under the dermal mesenchyme and above the meninges covering the brain.[6-8] The mesenchymal layer differentiates into an outer layer in which the ossification centers form skull bones. Ossification (the area of primary bone growth) proceeds from the central ossification centers outward toward the edges of the bones.[8,9]

Cranial sutures are fibrous tissues that unite the cranial bones as the bones approximate with one another during craniofacial development. The cranial sutures have 2 important functions: maintenance of head malleability during passage through the birth canal and continuance of separation of the calvarial bones during intrauterine and early perinatal life.[1,7,10] Sutures serve as secondary bone growth sites, adaptive regions at which bone remodeling takes place. Bone growth does not occur until a stimulus to make bone is provided by some external signal such as the expanding (growing) brain. As the brain expands and the cartilaginous bone-growth centers lengthen, the sutures respond by adding intramembranous bone at the edges of the bone fronts, such that the sutures remain approximately the same width and the cranial vault increases in size to accommodate the enlarging brain.[6,8-10] The sutures allow expansion of bone during brain growth and function as signaling centers, regulating the growth and remodeling of cranial bone.[1,6-9]

Fontanelles are gaps between the bones and are located at the junction of the cranial sutures. The posterior fontanelle, located at the junction of the sagittal and the lambdoid sutures, is closed by 2 to 4 months after birth. The anterior fontanelle, located at the junction of the sagittal, metopic, and coronal sutures, remains open and unossified, with a fibrous union, until approximately 18 months after birth. The sutures close at different times during a child's life. The metopic suture (located in the forehead) is the first suture to close, by age 1 year. By age 22 years, the rest of the sutures begin to close with solid-bone union. Sutures within the facial bones can take up to 70 years to fuse.[10]

The exact mechanism of normal fusion of cranial sutures is not known. Fusion of cranial sutures and maintenance of patency depend on the interplay of transcription factors, cytokines, growth factor receptors, and extracellular matrix molecules.[8] Craniosynostosis most likely is due to disequilibrium between proliferation and differentiation of the bone-forming cells of the cranial

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sutures caused by abnormalities in signaling, tissue interactions, or a combination of both elements.[8]

According to another theory, the cranial base is the primary locus of abnormality in children with craniosynostosis and the altered cranial base transmits the tensile forces through the dura. In craniosynostosis, the premature closure of any of the cranial sutures prevents separation of the cranial bones. This lack of separation produces a restriction of growth vectors, leading to a morphological change in calvarial shape.[7,10]

The absence of one or more of the sutures of the skull hampers normal bone growth in one or more directions, fostering compensatory growth along other open sutural areas. If a suture line is prematurely ossified, no growth occurs in the direction perpendicular to the involved calvarial suture. The compensatory brain growth causes the skull to become misshapen in a way that is predictable, depending on which suture is closed.[6-10] Figure 1 shows the deformity in the skull according to the closed suture.

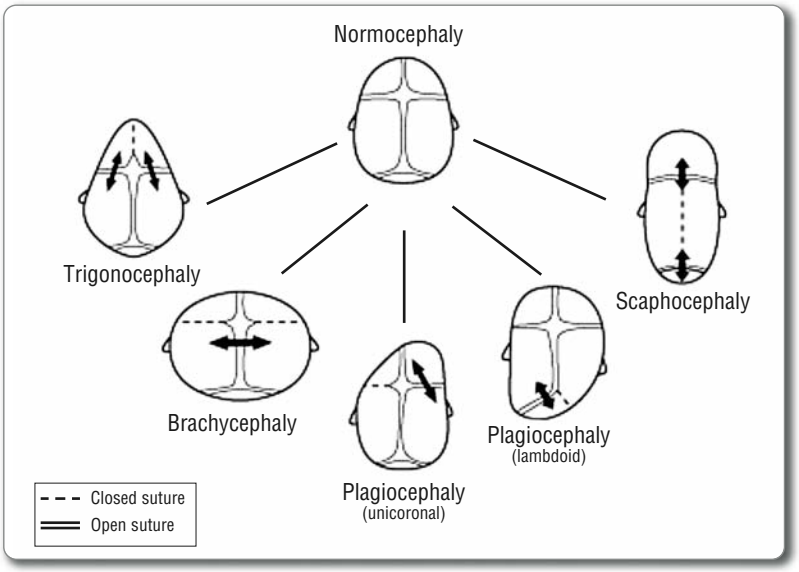

Figure 1 Types of craniosynostosis. When a suture fuses prematurely, skull growth is restricted perpendicular to that suture. Arrows show compensatory growth along open sutures.

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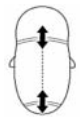
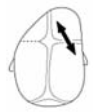
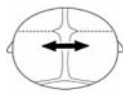
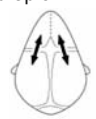
Diagnosis

A skull deformity or an abnormally shaped skull in an infant or child is a common reason for referral to a pediatric neurosurgeon, craniofacial surgeon, and/or a craniofacial center. A diagnosis may be made at birth if sutures are already closed or within several months of age, depending on whether the suture is completely closed or not. The diagnosis of craniosynostosis is based on the findings of physical examination. Questions about developmental milestones, irritability, headaches, emesis, and visual complaints must be investigated for a detailed history.[10]

On examination, special attention is paid to the alertness of the infant or child, the head circumference, the pupils, the fontanelles, and the shape and characteristics of the deformity. The physical examination, with a focus on the skull, includes palpation of the anterior and posterior fontanelles for size, shape, position, and fullness. The symmetry of the frontal bones and occiput is examined. The width of the biparietal diameter is compared with the anteroposterior length of the skull. Each suture is palpated for a ridge; a ridge is significant in the diagnosis of craniosynostosis and does not occur with normal closure of the suture.[10,11] The shape of the forehead, slope of the cranium, and relative position of the ears are assessed. The frontal view allows an assessment of eye symmetry and position and twisting of the nasal tip.[2,10] In severe cases of craniosynostosis, exorbitism (ie, protrusion of the eyeballs) can result.[10,11] The level (symmetry) of the skull base and any vertical displacement of the ears are assessed from the posterior view. The cranium has a specific shape depending on which suture or sutures are closed.[1,2,7,10,11] Table 1 gives the clinical findings of each type of craniosynostosis.

A clinical examination can reveal associated abnormalities in the digits, toes, and spine of an infant with syndromic craniosynostosis. Abnormalities such as syndactyly and polydactyly of the hands and feet, broad thumbs and toes, and hypoplastic thumbs and digits are diagnostic for syndromes such as Apert and Pfeiffer syndromes. Widespread and shallow orbits, jaw malocclusion, and cervical spine anomalies are associated with Crouzon syndrome. Soft-tissue and skeletal involvement, airway obstruction, dental abnormalities, hearing loss, and cardiac anomalies are also associated with syndromic craniosynostosis.[2,5,10]

Table 1 Characteristics of various types of craniosynostosis

Type	Head shape	Clinical features
Sagittal 	Scaphocephaly or dolichocephaly	Frontal bossing, elongated cranium (boat-shaped), prominent occiput, palpable keel ridge, normal head circumference, reduced biparietal diameter (skull longer in anteroposterior diameter), reversed slope of cranium Anterior fontanelle may be patent or not
Coronal 	Unilateral plagiocephaly	Marked craniofacial asymmetry: flattened forehead on affected side, flat cheeks, nose deviation to normal side, higher supraorbital margin leads to harlequin sign on radiographs and outward rotation of orbit
	Bilateral; brachycephaly, acrocephaly	Vertical, broad, flattened forehead, possible hypoplasia of midface and progressive proptosis, choanal atresia, high-arched palate, and poor dental occlusion Skull shorter in anteroposterior diameter, biparietal diameter increased A ridge possibly detected via palpation over coronal suture Nasolacrimal duct possibly narrowed, visual acuity possibly decreased
Metopic 	Trigonocephaly	Pointed or triangular forehead and prominent midline ridge, lateral supraorbital recession, fontanelle usually absent, hypotelorism
Multiple	Oxycephaly	Tower skull with shallow orbits

The diagnosis of craniosynostosis is confirmed by findings on 3-dimensional computed tomography (CT) of the brain and skull.[1,10] The images obtained highlight the sutures, shape of the skull, and, possibly, any other skull abnormalities and are particularly useful in showing the general volume relationships and overall shape of the underlying bony structures. Computer-assisted models can be used preoperatively to assist in planning osteotomies. The axial views of a CT scan show any associated brain anomalies. Magnetic resonance imaging is not usually included in the diagnostic process unless the CT scan shows abnormal congenital findings.[2] Some surgeons may recommend head sonography to assist in the diagnosis to avoid the radiation associated with CT scans. However, CT scans are the standard of care.[12] CT findings of craniosynostosis are shown in Figure 2.

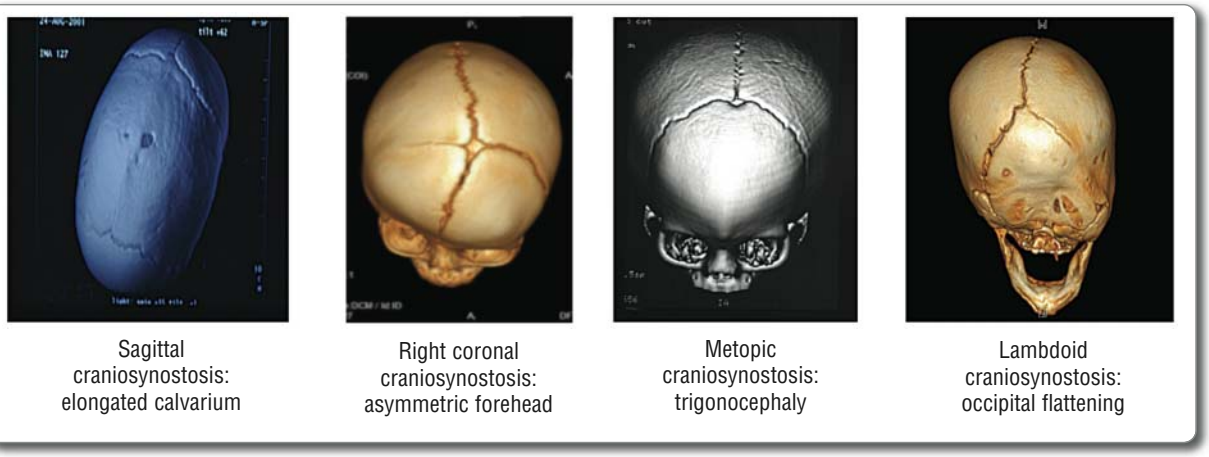


Figure 2 Computed tomography findings for various types of craniosynostosis.

Preoperative Discussion and Evaluation

Preparation for surgery starts with examination of the infant or child in a neurosurgery or craniofacial clinic. A careful history is obtained, and a physical examination is done. Because some craniosynostoses are syndromic and familial, a detailed family history is essential. The preoperative discussion with the patient's parents involves information on the abnormality, review of the CT scan of the brain, and indications for surgery. Clinical correlation can be easily made and understood by a patient's family after viewing the CT scans. The parents are told about the details of surgery, the need for blood transfusion, and complications of surgery. They are informed that the aesthetic outcome for the child will depend on the growth of the brain and the effect of the growth on the expansion of the skull.

Most brain growth occurs in the first year of life. The deforming vectors of the continually growing brain result in progression of the deformity with increasing age.[10,11] Uncorrected craniosynostosis is associated with an increase in intracranial pressure; however, few studies have revealed increased intracranial pressure in nonsyndromic single-suture craniosynostosis. The sequelae of craniosynostosis include both physical deformity and insufficient cranial volume to permit normal cerebral growth and development.[10,11] Osseous defects undergo reossification more completely when surgery is performed before the child is 1 year old than they do when surgery is performed at a later age. A delay in surgery beyond the first 9 to 12 months of life leads to progressive deformity of the cranial base, resulting in abnormal facial growth and asymmetry of the maxilla and mandible. The calvarium in a child 3 to 9 months old is still malleable and therefore quite easy to shape.[10] Thus, the timing of surgery is birth to 6 months for cranial vault decompression and suture release for sagittal craniosynostosis, 6 months to 3 years for fronto-orbital reshaping and advancement for coronal and metopic craniosynostosis, and 4 to 8 years for correction of midface deformities associated with cranial vault procedures and adjunct procedures for patients with syndromic craniosynostosis.[2]

Depending on the institution, the surgeon, and the clinical criteria for surgery, surgery is done via open craniectomy or via an endoscopic approach.[13,14] At Advocate Children's Hospital, Oak Lawn Campus, Oak Lawn, Illinois, open craniectomy is used for surgical reconstruction of the cranial vault via surgical resection of the closed suture and the creation of osteotomies, bone grafts, and/or use of bioresorbable plates to reconstruct the vault. The goal of surgery is reconstructive, to allow the brain to grow naturally, and is not intended to be purely cosmetic. Minimally invasive endoscopic reconstruction for craniosynostosis is an alternative approach at some medical centers for patients who meet certain requirements. An endoscope is used to resect the closed suture, and techniques for osteotomies and use of bioresorbable plates have been developed. Surgery to correct the cranial or craniofacial deformity is performed in infants less than 3 months old up to 6 months old who have less complex defects. This procedure does not allow the surgeon to intraoperatively alter the calvarial shape or cephalic index. The patients must wear helmets for 6 to 8 months after surgery to help mold the cranium.[10,13,14]

> Congenital defects involving an infant's face and skull seem to evoke particularly strong emotional responses from the infant's parents.

The goal of surgery via open craniectomy is to excise the fused suture and reconstruct the cranial vault to increase intracranial volume, avoid intracranial hypertension, and allow the brain to grow in a natural pattern. Complications of surgery[10,12,15-17] are listed in Table 2. Although unlikely, stroke and even death can occur as a result of surgery. Mortality is unusual, but rates can be as high as 1.1% to 2.2%.[16]

In addition to the aesthetic outcome of surgery, the impact of craniosynostosis on the physical and emotional condition of the child and on learning disabilities are discussed with the patients' families. The stigma attached to facial and bodily disfigurements can lead to socioemotional problems, and children with craniofacial anomalies are considered at high risk for negative self-perceptions, withdrawal, social inhibition, anxiety, and peer rejection. Growing up with a abnormally shaped head can be devastating. Even mild deviations from typical facial appearance can have marked impact on psychological adjustment.[18,19] A child with craniosynostosis can experience social and emotional consequences due to verbal victimization by peers about the child's physical appearance.[20-22]

Craniosynostosis is associated with a 3- to 5-fold increase in risk for cognitive deficits or learning or language disabilities.[2,23,24] The causal basis of this association

Table 2 Complications of surgery to repair craniosynostosis

Complication	Cause	Recognition	Prevention	Management
Hemorrhage	Venous sinus hemorrhage Scalp vessel hemorrhage Bone dissection Epidural or subdural hematoma Cerebral contusion	Tears during surgery Decrease in hematocrit Unstable vital signs Prolonged prothrombin time or partial thromboplastin time Hypotension Coagulopathy	Caution when removing closed suture from dura Packed red blood cells available in operating room	Administer blood transfusion Monitor hematocrit Maintain fluid balance
Cerebrospinal fluid leak	Torn dura	Leaking cerebrospinal fluid	Surgical technique	Suture dura
Meningitis or infection	Dura torn during surgery Wound breakdown	Fever Wound infection Dehiscence	Surgical technique	Administer antibiotics in 30 min before start of surgery and after surgery
Pulmonary edema	Reaction to blood transfusion	Difficulties in ventilation	Assessment of ventilatory parameters Arterial blood gas analysis	Manage airway pressure Use oscillator
Air embolism	Osteotomies	Decrease in oxygen saturation Increase in end-tidal carbon dioxide Findings on precordial Doppler monitoring	Bone wax Positioning	Assess vital signs and hypotension Arterial blood gases

is unclear, and the roles of increased intracranial pressure and secondary cerebral deformation continue to be studied.[24] Children with syndromic craniosynostosis characterized by multiple-system involvement are often developmentally delayed.[10,25] Children with single-suture craniosynostosis often do not seem to have marked developmental delays; however, multicenter studies[24] consistently have revealed lower mean neurodevelopmental scores in these children at 36 months of age than in control groups without the abnormality. Children with metopic and sagittal craniosynostosis have minor delays in learning and speech. Neurodevelopmental testing is recommended to obtain information about the cortical functioning before and after remodeling of the cranial vault.[10]

A particular stressor for parents of children undergoing surgery for craniosynostosis is anxiety about how the child will look after surgery.

Parents

Children with craniosynostosis benefit from a multidisciplinary-team approach, which is used in most craniofacial clinics affiliated with major pediatric medical centers.[2] The specialists usually include plastic surgeons, neurosurgeons, otolaryngologists, dentists, audiologists, ophthalmologists, speech pathologists, developmental pediatricians, neuropsychologists, medical geneticists, social workers, and nurses. Specialists (ie, cardiologists and gastroenterologists) are consulted for management of associated defects and clearance for surgery. Making appointments to meet all the specialists and processing all the information discussed can be overwhelming for patients' parents.

In addition, congenital defects involving an infant's face and skull seem to evoke particularly strong emotional responses from the infant's parents, who must contend with a host of potentially stressful events and circumstances, including the infant's unusual appearance, potentially life-threatening surgeries and other medical procedures, and the possibility of future neuropsychological and educational problems. All of these factors can potentially affect parents' responsiveness and adaptation to an infant who has a craniofacial abnormality.[25] In studies[25-28] of the impact of syndromic craniosynostosis on quality of life, parents' scores for children with syndromic or complex craniosynostosis were significantly lower than scores for parents of children without the abnormality. Mothers of children with the abnormality reported greater stress if they perceived their child's condition as noticeable to others. Visibility of the anomaly should be considered a risk for increased stress for mothers of infants with craniosynostosis.[28]

Preoperative Nursing Interventions

The surgical team's advanced practice nurse assists in counseling families of children with craniosynostosis and in providing holistic care to integrate the medical, surgical, and genetic data.[2] The nurse offers support before surgery by assisting in coordinating consultations and addressing developmental and psychosocial issues. He or she assists in coordinating preoperative testing, insurance verification, blood donation, and setting up conversations with other families who have experienced similar surgeries. Information about the specific suture involved and what information parents need to share with their families is provided. Internet resources are shared. Expectations of hospitalization are explained: preoperative and postoperative care, length of surgery, length of stay in the pediatric intensive care unit, and plans for discharge. Parents are encouraged to room in at the hospital, and mothers are encouraged to breastfeed in the times before and after surgery. Parents are told to bring familiar toys and objects such as blankies and music to provide comfort, because the patient's eyes may be swollen shut for 1 to 3 days postoperatively. The parents are told that they will receive updates during surgery from the advanced practice nurse. Depending on the suture involved, surgery can take up to 8 hours. Parents are informed that they will talk with the surgeon or surgeons immediately after surgery, who will explain the plan of care, and are encouraged to prepare any questions that they may have.

Most important, the surgical team's nurse is available to address concerns that the families have about their infants. Parents look at before-and-after pictures to see what their child will look like without surgery and possibly after surgery. Many parents want to believe that they are doing what they think is best for the child.[20,24] The main influence for surgery for many parents is that their child is starting to look strange. As one parent remarked, "It was hard watching the head become more distorted and seeing adults stare at my child." Parents most often choose surgery because of concern for brain damage, aesthetics, or both and expect improvement in quality of life to be high.[28]

Providing education to a patient's family members empowers them as they make choices about their child's care. Families require education about their child's condition and their role in providing care. They need to prepare themselves and their child for surgery and postsurgical care. A family's level of adaptation of cohesion and emotional health influences the ability to nurture a child. Parents' confidence in their ability to provide care and support for their child will decrease emotional distress.[2]

In the immediate preoperative period, nurses orient the parents and child to hospital policies; obtain samples for hematocrit, hemoglobin, electrolytes, and coagulation studies; and address questions the parents may have. Intravenous access is established by the nurses for intravenous hydration, and the child receives nothing by mouth after midnight. Parents accompany their child to the preoperative holding area, where they meet with the anesthesiologist and with the neurosurgeon, the craniofacial surgeon, or both.

Intraoperative Period

Intraoperative preparation for the craniotomy includes placement of additional intravenous or central venous catheters, arterial catheters, and a urinary catheter; intubation; and positioning of the patient either supine for coronal or metopic craniosynostosis or prone in the sphinx position for sagittal craniosynostosis. For orbitofrontal procedures, the patient's eyelids are sutured shut to protect the cornea from being exposed to the air during the surgery; the sutures are removed after the surgical procedure. The hair is shaved along the incision line, which is placed in a zigzag fashion from tragus to tragus.

Brain protection is mandated to prevent violation of the underlying dura and protect the large venous sinus from hemorrhage.

During the operative procedure, the scalp and muscle layers are dissected from the bony surface, and the fused suture is visualized. Landmarks for removal of the abnormal bone are established, burr holes are placed for incision, and the diseased bone is removed carefully. Brain protection is mandated to prevent violation of the underlying dura and to protect the large venous sinus that runs parallel to the closed suture. For sagittal craniosynostosis, a piece of bone 4 to 6 cm in diameter is removed from the fontanel to the inion of the skull. For coronal and metopic craniosynostosis, the orbital rim and the frontal bones are removed. The removed bone is recontoured and reshaped. The dura is protected with a thin layer of coagulable material. As the removed bone

is prepared, barrel-staved osteotomies are placed to expand the skull. Recontoured grafts may be placed and secured to the skull with absorbable plates and screws.[10,12,15,16,29-32] A drain is left in place above the skull but is pierced through the scalp as the scalp is sutured back together with 2 layers of stitches.

Blood loss during surgery may be acute, and transfusions are usually necessary because of the insidious losses throughout the surgical period. Transfusion requirements can be 15% to 90% of the patient's circulating red cell volume.[33,34] Numerous techniques intended to reduce intraoperative blood loss during craniosynostosis surgery have been studied, including use of autologous blood obtained before surgery, short-term nomovolemic hemodilution, and intraoperative blood salvage.[12,15] Tranexamic acid is a synthetic antifibrinolytic drug that competitively decreases the activation of plasminogen to plasmin and suppresses fibrinolysis by inhibiting plasminogen and the binding of plasmin to fibrin. Tranexamic acid has been used to decrease intraoperative blood loss during cardiac surgery. In a randomized double-blind study[33] published in 2011, patients undergoing surgical correction of craniosynostosis who were treated with erythropoietin before surgery and with tranexamic acid during the operation sustained significantly lower volumes of blood loss and received less transfused blood than did the control patients.

Removal of drainage fluid can be uncomfortable for the child, and pain medications may be given before the fluid is drained.

Postoperative Care

After surgery for craniosynostosis, patients return to the pediatric intensive care unit for monitoring of hemodynamic status and level of consciousness. When the duration of surgery is long or blood loss is marked, the endotracheal tube may be left in place. Oropharyngeal and laryngotracheal edema may exacerbate preexisting obstructive apnea in patients with syndromic craniosynostosis and may critically worsen an already difficult intubation for patients with midface deficits.[2,15] Patients are positioned with the head of the bed elevated, and pulse oximetry is used to monitor oxygen saturation.

Vital signs are monitored for indications of hypovolemia. Infusions of isotonic solutions are maintained until fluid shifts are complete, and hemodynamic, hematologic, and neurologic conditions are monitored until the risk for bleeding has passed. Fluid is removed from the subgaleal surgical drain every 4 to 6 hours. Continued blood loss via drains is replaced by transfusion of packed red cells or isotonic fluid, depending on the patient's hemodynamic status and hemoglobin level. Drainage from the subgaleal drain can be up to 100 mL of sanguinous fluid per draining event and will diminish to less than 20 mL of serosanguinous fluid within 1 to 2 days before the drain is removed. The amount of drainage fluid varies among patients. The head dressing is monitored for concealed blood losses. Coagulopathy may be consumptive or dilutional and may require specific correction with fresh-frozen plasma, platelets, or cryoprecipitate.[3,33,34]

Intravenous solutions with dextrose and physiological saline are administered to maintain adequate intake and output balance and prevent electrolyte imbalances. Blood samples are obtained for laboratory tests to monitor the hematocrit level and the serum level of sodium; alterations in the sodium concentration can precipitate seizures. Pain medications such as acetaminophen, morphine, and fentanyl are administered when needed. Pain and discomfort can be anticipated from not only the head incision but also the facial and periorbital edema that develops postoperatively. The subgaleal drain decreases periorbital edema and ecchymosis around the eyes. The compression bandage on the head also helps limit scalp edema. Removal of drainage fluid can be uncomfortable for the child, and pain medications may be administered before the fluid is drained. Pyrexia (38°C) is not unusual for the first 72 hours because of atelectasis. Continuing pyrexia, persistent swelling, and potential cellulitis should be further investigated.[10] Urine output is monitored, and adequate intake and output balance is calculated. Anticipated postoperative complications and interventions[2,10,14,16,34] are listed in Table 3.

A CT scan of the brain is scheduled soon after surgery to obtain a postoperative baseline and to monitor for complications such as epidural or intracranial hemorrhage. The scan also provides an accurate method for documenting the osteotomy sites. In addition, the surgeon can confirm postoperatively whether the appropriate symmetry and frontal advancement were achieved intraoperatively.[10] The 3-dimensional CT scan shows the complexity of the reconstruction. The CT scan is shown to the patient's parents; seeing the 3-dimensional image

Table 3 Nursing care after surgery to repair craniosynostosis

Complication	Cause	Prevention	Indications	Management
Altered level of consciousness	Air embolism, stroke Hypovolemia Electrolyte imbalance	Assess level of consciousness, cranial nerves Do sensory and motor evaluation	Lethargy, unresponsiveness	Do neuroassessment hourly
Hemorrhage	Scalp drainage Bone dissection Intracranial hemorrhage	Monitor tachycardia Monitor complete blood cell count Monitor subgaleal drainage output Monitor coagulopathy	Low hemoglobin level Pallor Tachycardia Low blood pressure Increased drain output Findings on computed tomography	Monitor complete blood cell count Replace scalp drainage output milliliter for milliliter with packed red blood cells or physiological saline
Pain	Surgical incision Periorbital edema	Give acetaminophen or narcotics as needed Elevate head of bed 30°	Tachycardia Restlessness Irritability	Administer medication Assess after medication given
Stridor	Intubation	Assess respiratory status	Noisy airway Drooling	Administer racemic epinephrine Administer steroids
Apnea	Midface deficits Laryngotracheal intubation	Assess respiratory status	Respiratory distress Hypoxia	Start mechanical ventilation Provide oxygen support
Periorbital swelling	Reflection of scalp during surgery Postoperative swelling	Maintain head of bed elevation at 30° Position side to side Place compression bandage on scalp	Swelling of both eyelids and face Eversion of eyelids	Encourage parent to hold infant Implement canthotomy for extreme edema behind orbit
Electrolyte imbalance	Intraoperative fluid imbalance Hypervolemia Hypovolemia	Monitor electrolyte levels	Decreased serum level of sodium Decreased urinary output	Administer boluses of physiological saline
Seizures	Electrolyte imbalance Hypoxia	Monitor electrolyte levels Monitor arterial blood gases	Neuroassessment hourly	Administer anticonvulsants Assess causes
Infection	Osteomyelitis	Monitor body temperature	Pyrexia	Administer acetaminophen
Blood transfusion reaction	Intraoperative blood administration	Monitor body temperature Monitor for pulmonary edema	Pyrexia Hematuria	Provide ventilatory support

of the brain helps them understand the goal of surgery and the changes in the cranial shape (Figures 3-5). The first dressing change can be frightening because some of the hair has been shaved off and the incision from ear to ear is markedly apparent, yet parents are eager to see the outcome of surgery. Parents can see the difference in the shape of the head immediately.

Parents are encouraged to hold their infant for comfort after surgery, because the infant's eyelids will be swollen shut and remain closed for 2 to 3 days not only to provide comfort but also to help decrease periorbital edema. Familiar items, such as blankets and music, and being held in the mother's arms provide comfort to the child. Postoperative feedings are encouraged as soon as the effects of anesthesia have dissipated. Mothers can breastfeed with the assistance of a nurse in positioning the child with the child's electrodes and drains.

Postoperative Stressors and Nursing Interventions

Parents experience various stressors upon seeing their child in the intensive care unit, such as the child's appearance, sights and sounds of the unit, procedures, the child's behavior and emotions, and staff communications.[35] The facial swelling and the presence of drains in a child who has had surgery for craniosynostosis can be

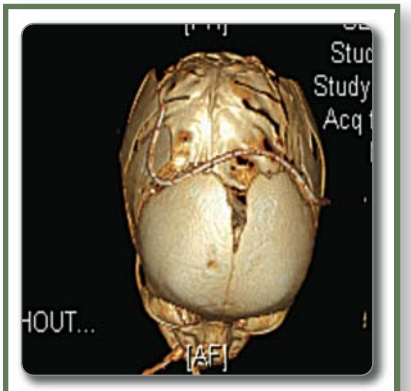


Figure 3 Sagittal strip craniectomy for repair of sagittal craniosynostosis.

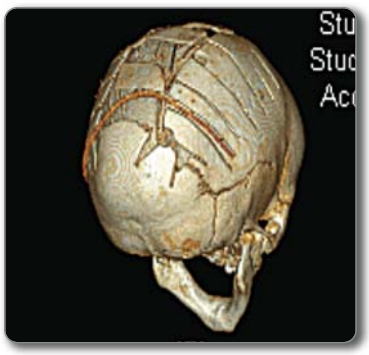


Figure 4 Sagittal strip craniectomy with autologous bone grafts for repair of sagittal craniosynostosis.

Figure 5 Orbital frontal advancement and multiple osteotomies for repair of coronal craniosynostosis.

overwhelming. The swelling and the amount of drainage increase during the first few days and may deter the parents from holding the child. Parents see other children who are critically ill and interact with other anxious parents. The parents may witness the death of another child and experience the reactions of the affected family.

Nurses can use strategies to decrease stress in parents with a child in the pediatric intensive care unit, such as providing unrestricted access to the child and honest and straightforward information. In surveys[35-37] of parents, these interventions have been described as the highest needs. Providing the patient's family members with ongoing daily information is an important priority throughout a child's stay in the intensive care unit. Family conversations with the same person each day, involvement of the family in patient rounds, provision of information frequently and in terms the family can understand are important needs of families. Receiving explanations of all tubes, equipment, and procedures and being able to participate in their child's care are important.[35-37] Paramount to parents are trust in the caregiver and knowledge that their child is receiving excellent care. Cultural identity, rituals, values, and behaviors are also important aspects of family systems that should be incorporated into the care of the child.[36] Including family members in the daily care of the child lowers their levels of stress and increases their satisfaction.[36,37] Use of these strategies can decrease parents' anxiety and enhance their trust in the care their child is receiving.

Stressors vary among families, and the degree of stress is influenced by factors other than the child's hospitalization.[37] In addition to the recovery from surgery, a particular stressor for parents of children undergoing surgery for craniosynostosis is anxiety about how the child will look after surgery. Parents need to be informed that the facial and periorbital eye swelling will increase for the first few days and then dissipate, and only then will they be able to appreciate the outcome of surgery. The first time they see their child's head without a dressing can be alarming. The change in the head shape immediately after surgery will be marked; however, the final aesthetic outcome may take years to develop. Discussing these changes will reassure parents that they have made the right decision for surgery.[20]

Discharge

Discharge teaching includes instructions for caring for the incision and watching for signs of infection. Parents are told that the incision tends to itch as it heals. Ointments are not recommended. Keeping the incision covered to keep the child from scratching it and administering acetaminophen are helpful. Getting an infant back on schedule with naps and feedings may take a couple of days as the child recovers from the stress of hospitalization. Some pain may be confused with the pain of teething, because teething begins around 6 months of age. The incisional sutures are absorbable and do not need to be removed; dressings are changed for the first 2 weeks. Parents are instructed to dab the incision with povidone-iodine solution every other day and to keep it covered with a gauze dressing and a netting hat for 1 to 2 weeks. As the postoperative swelling diminishes, the parents will be able to feel the several soft spots (cranial defects) created by the surgery. They will feel these spots when they begin to shampoo the baby's head in 2 weeks. The defects will fill in gradually from the edges of the

bone inward, and parents will feel the sharpness of the defect soften with time as the areas fill in. Ossification within the orbit takes place within 1 year after surgery. It may take 2 to 3 years for the remaining defects to fill in with a semismooth finish to the bone.[10] Pictures taken at several intervals at home can help monitor the progress.

Patients have follow-up examinations 2 weeks after surgery, then every 4 to 6 months for the first year, and then yearly for the next 3 to 4 years. The closure of the defect is monitored clinically and, as needed, with imaging. The need for possible future surgeries is discussed as the child progresses after surgery. Patients whose defects remain patent and do not close will require surgery to close the defect with hydroxyapatite paste or titanium mesh by age 4 to 5 years. Patients with syndromic craniosynostosis may require midface advancement procedures at ages 4 to 7 years.[10]

Parents of children who have had surgery for craniosynostosis may be concerned about the possibility of trauma to the surgical site from roughhousing between the child and the child's siblings. The parents are instructed to continue with the same preventive or well-child care they would provide for any child with a head injury. Helmets for protection are not recommended because they actually interfere with the expansile force of the brain and may hamper the aesthetic effect of surgery on the cranial vault. According to reports,[38,39] parents are satisfied with the aesthetic results of surgery. Because of the potential delays in cognitive and emotional development associated with craniosynostosis, follow-up with neuropsychologists or developmental pediatricians or both should be maintained.[29,38,39] Bellew et al[24] reported that children with sagittal craniosynostosis who had surgery had an improvement in locomotor skills postoperatively, suggesting that surgery might have an impact on development.

Summary

The challenges of providing care to children who have had surgery for craniosynostosis and to the children's families are to provide expert care to the patient and to support and allay anxiety for the parents. Expectations about hospitalizations help parents cope with surgery. Establishing a trusting relationship with parents and providing care that parents acknowledge as compassionate and competent are important. Efforts to allow parents to participate in their child's care are paramount. Continuity of care after discharge is needed for follow-up of the aesthetic and neurodevelopmental outcomes of the child. The Centers for Disease Control and Prevention[40] recommend research into long-term outcomes of surgery for craniosynostosis, health care utilization, psychological adjustment, social relationships, and factors that improve outcomes for families with an affected child. CCN

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To learn more about pediatric critical care, read "The KIDS SAFE Checklist for Pediatric Intensive Care Units" by Ullman et al in the *American Journal of Critical Care*, January 2013;22:61-69. Available at **www.ajcconline.org**.

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CNE Test Test ID C134: Craniosynostosis: Caring for Infants and Their Families

Learning objectives: 1. State the physical examination and radiologic findings used to diagnose craniosynostosis 2. Describe the postoperative nursing care for children undergoing surgical repair of craniosynostosis 3. List 4 potential complications following surgical repair of a craniosynostosis defect

1. Which of the following sutures is most commonly affected with craniosynostosis?
a. Sagittal
b. Metopic
c. Coronal
d. Lamboidal

2. The underlying premise of the pathophysiology for craniosynostosis is most likely related to disequilibrium of which of the following factors?
a. Failure of the unification of bone growth at the metopic suture
b. Interplay of cytokines coupled with premature closure of the sutures
c. Failure of bone growth to accommodate the increased brain size
d. Abnormalities in signaling, tissue interactions, or a combination of both

3. Which of the following diagnostic studies is used to confirm a diagnosis of craniosynostosis?
a. Multiple failed developmental milestones
b. Nonpalpable fontanels in an infant
c. Magnetic resonance imaging of the brain
d. 3-dimensional computed tomography of the brain and skull

4. Which of the following periods is the optimal timing for surgical intervention for the most common type of craniosynostosis?
a. 6 months to 3 years
b. 9 to 12 months
c. Birth to 6 months
d. 4 to 8 years

5. According to one study, the most common reasons parents offer for pursuing surgery for their child with craniosynostosis include concern for brain damage, aesthetics, and which of the following?
a. Decreased need for future orthodontia
b. Increased IQ score
c. Normal visual acuity
d. Improved quality of life

6. Which of the following is the rationale for administering tranexamic acid intraoperatively?
a. Reduce intraoperative blood loss and transfusion requirements
b. Decrease the amount of fresh frozen plasma to be administered
c. Eliminate the need for erythropoietin to be administered preoperatively
d. Eliminate the need for autologous transfusions

7. Which of the following interventions is appropriate to treat postoperative pain in children?
a. Administer Toradol, maintain the compression bandage, and elevate the head of the bed to 30°
b. Administer morphine, apply ice to the eye lids, and maintain the bed flat
c. Administer fentanyl, administer acetaminophen, and elevate the head of the bed to 30°
d. Administer Dilaudid, maintain the compression bandage, and do not reposition

8. Which of the following complications is most likely occurring in a child with fever, crackles, and hematuria following a craniosynostosis repair?
a. Blood transfusion reaction
b. Osteomyelitis
c. Hypovolemia
d. Pulmonary edema

9. Which of the following is an appropriate nursing intervention for pyrexia occurring on postoperative day 1?
a. Assist with performing a lumbar puncture
b. Send a urine culture
c. Encourage pulmonary toilet
d. Remove the central catheter

10. Which of the following strategies minimizes parental stress in the pediatric intensive care unit?
a. Restricting visitation
b. Offering honest and straightforward information
c. Minimizing parent participation on rounds
d. Not allowing parents to hold their child

11. When should parents be told to expect notable changes in their child's head shape?
a. Immediately following surgery
b. At the 2-week follow-up appointment when the dressing is removed
c. One month following surgery
d. About 1 year following surgery

12. Follow-up with which of the following specialists is recommended for children who have undergone surgery for craniosynostosis?
a. Plastic surgeon
b. Oral surgeon
c. Neuropsychologist or developmental pediatrician
d. Psychiatrist

Test answers: Mark only one box for your answer to each question. You may photocopy this form.

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