

Craniosynostosis and Deformational Plagiocephaly: How to Differentiate the Conditions

by **Robert Wood, M.D., and Cheryl Shell, C.P.N.P. – Pediatrics**

When evaluating children who have misshapen heads, physicians should consider deformational plagiocephaly and craniosynostosis — two common skull conditions — as possible causes of the deformity. By knowing the characteristics of each condition, as well as those of associated disorders such as torticollis, primary-care physicians can determine whether children need referrals to craniofacial surgeons or neurosurgeons.

When to Refer

Early diagnosis of the conditions is crucial. When primary-care physicians diagnose craniosynostosis (premature closing of the cranial sutures), they should immediately refer the child to a craniofacial surgeon or neurosurgeon for an evaluation. Although brain damage isn't a risk with deformational plagiocephaly, the possibility of such damage in untreated craniosynostosis is very real. Moreover, successfully treating plagiocephaly requires intervention between the ages of 4 and 8 months; therefore, infants who are diagnosed with plagiocephaly should see a craniofacial specialist during this period. Before 4 months of age, primary-care physicians might recommend repositioning infants' heads whenever they're on their backs and monitoring the deformity. At the first sign of plagiocephaly, physicians also should check for torticollis (see *Associated Conditions*).

Craniosynostosis is one of the most common physical deformities of newborns, occurring in about 1 of every 2,000 births. In the past decade, the incidence of deformational plagiocephaly has increased dramatically, following the American Academy of Pediatrics (AAP) recommendation that babies sleep on their backs to reduce the risk of sudden infant death syndrome. By the mid-1990s, studies linking back-sleeping and plagiocephaly led to new AAP recommendations that parents occasionally turn a sleeping infant's head to avoid deformation.

Diagnosing the Conditions

Irregular head shapes are prominent signs of both deformational plagiocephaly and craniosynostosis. Children with craniosynostosis have distinct head patterns caused by the suture involved. Plagiocephaly results in characteristic parallelogram shapes. (See illustrations.)

In plagiocephaly, the skull has a smooth surface. The ear on the affected side is sheared forward, and facial features on the same side might be more full. Conversely, in craniosynostosis, a ridge along the skull where the suture has closed is almost always present. The ear might be pulled back or unaffected, and facial features might not be affected. (See box for other differentiating characteristics.)

The presence of such physical features, combined with a careful clinical exam of the head shape, might be sufficient to differentiate craniosynostosis and deformational plagiocephaly. When the diagnosis remains questionable, however, use a series of skull X-rays or a computed tomography (CT) scan to resolve the issue. If questions persist regarding the diagnosis, consult a craniofacial surgeon.

Craniosynostosis

Once physicians determine the type of skull deformity a child has, treatment planning can begin. If a diagnosis of craniosynostosis is confirmed, it's important to determine the type: single-suture craniosynostosis, in which one bony seam closes prematurely, or multiple-suture craniosynostosis, which affects more than one bony seam. There are four major types of single-suture craniosynostosis.

Single-Suture Types and Treatments

- Scaphocephaly (sagittal synostosis) results from a premature fusion of the bones along the middle seam at the top of the head. Children with this condition tend to have a long head that is narrowed from side to side. The forehead and back of the head might be prominent (Figure 1).

An infant with this condition should receive treatment in the first few months of life. The craniofacial surgeon and pediatric neurosurgeon will perform a corrective procedure that involves removing a plate of bone at the top of the skull. Children not treated in the first year of life might require a more complex operation — done in two separate procedures — involving the reshaping of the front and back of the head.

Figure 1 – Scaphocephaly

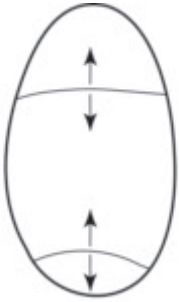


Figure 2 – Trigenocephaly

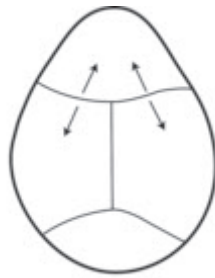


Figure 3 – Synostotic anterior plagiocephaly

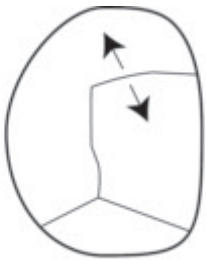


Figure 4 – Nonsynostotic deformational plagiocephaly



- Trigenocephaly (metopic synostosis) results from a premature fusion between the two bones of the forehead. Children with this condition tend to have a triangular or keel-shaped forehead. The distance between the eyes might be narrower than usual, and the outer corners of the eyes might angle upward (Figure 2).

A primary-care physician can monitor some minor forms of this condition, which might improve in time. Other forms are treated by reshaping the central portion of the forehead. This operation is usually scheduled for children 4 to 6 months of age.

- In synostotic anterior plagiocephaly (unilateral coronal synostosis), the spaces between the bones on one side of the head close prematurely. That causes a flattened forehead, an elevated and widened eye on the affected side, a tilted nose, and a cheek swept back posteriorly on the affected side (Figure 3).

Surgical correction for this condition is done when a child is 4 to 6 months of age. It involves repositioning the forehead and the area above the eyes and straightening the nose.

- Synostotic posterior plagiocephaly (lamboid synostosis) is caused when the bones on the side and back of the head fuse. It results in a flattened area on the side of the synostosis. The back of the head shifts to the unaffected side, and the area near the ear (mastoid process) might bulge. The condition is rare and can be confused with deformational posterior plagiocephaly (Figure 4).

Multiple-Suture Types and Treatments

Craniosynostosis is usually more severe when more than one suture closes too early. The forms of multiple-suture craniosynostosis are often associated with at least 70 different syndromes. Apert, Crouzon, Pfeiffer and Saethre-Chotzen are the most common.

Multiple-suture craniosynostosis significantly restricts the ability of the skull to expand as the brain grows. As pressure builds on the brain, infants might develop:

- A full or bulging fontanelle
- Scalp veins
- Bulging eyes
- Increasing head circumference
- Apnea
- Seizures
- Developmental delays

The pressure also might cause infants to vomit, become sluggish, sleep more, play less, and become irritable. In addition, such infants can have breathing problems and difficulty following sounds. Intracranial pressure in single-suture synostosis is generally increased in 13 percent of cases; when multiple sutures are involved, 42 percent of patients experience increased intracranial pressure.

Because the problems associated with multiple-suture craniosynostosis are so diverse and medically complex, a craniofacial surgeon or neurosurgeon should take part in developing a treatment plan tailored to a patient’s individual conditions and needs.

Deformational Plagiocephaly

In deformational plagiocephaly, the skull sutures don’t fuse. Unlike craniosynostosis, which is caused by internal mechanisms, deformational plagiocephaly occurs because of environmental factors. They include sleeping on the back, a restricted intrauterine environment, muscular torticollis and premature birth. Infants who need ventilators often keep their heads in a fixed position over a long period of time, which can lead to flattening — typically of the sides of the head — and a long, narrow shape. Physicians can usually diagnose plagiocephaly after conducting a thorough physical examination; X-rays or CT scans aren’t typically necessary.

Treating Deformational Plagiocephaly

Treatment of deformational plagiocephaly generally includes positioning and/or orthosis therapy. During the first three to four months after birth, consistent repositioning of a sleeping infant’s head often leads to spontaneous rounding of the skull with mild flattening. Increasing the time infants spend lying on their stomachs or sides — while supervised — is helpful as well.

In moderate to severe cases of deformational plagiocephaly, or when a trial of counter-positioning has failed, physicians might recommend a cranial remodeling orthosis. The orthosis assists the skull-molding process by removing pressure over the flat area, allowing the skull to grow into the space provided. Orthoses are most effective for children 4 to 8 months of age, which is when the skull is still quite malleable. Beginning at a later age requires a longer treatment period. After children reach 12 or 13 months, an orthosis is less effective and less likely to change the shape of the head. In most cases, correcting head shape in children older than 1 to 2 years requires surgery.

There are many cranial orthoses available. At Gillette, we use the CranioCap™ orthosis (Figure 5), which our craniofacial surgeon and orthotists designed. Fittings and moldings for these orthoses take place at Gillette.

Figure 5

The CranioCap™ is typically worn for four to 16 weeks. During this period, patients are regularly scheduled for return visits with an orthotist for adjustments.



The orthosis — which has Food and Drug Administration approval — is created from a plaster-cast model of the infant’s head. Pads inside the CranioCap help it fit snugly in some areas while remaining open in others. As the infant’s brain grows, the skull slowly pushes into the open areas of the orthosis, rounding the skull. It generally takes two to four months to complete the process.

Because slight variations in head shape and size are considered normal, craniofacial orthoses are used primarily in cases where failure to treat would leave the child with an enduring abnormal appearance.

Associated Conditions

Torticollis

Once a diagnosis of deformational plagiocephaly is made, physicians should check for associated conditions, such as torticollis, which is present in about 60 percent of children who have plagiocephaly. Muscular torticollis is present when the sternocleidomastoid muscle is shortened or tightened on one side.

The condition occurs most frequently on the right side. It might result from the child’s position in utero or from trauma to the muscle.

The cause of the majority of cases of torticollis is unknown. Medical specialists might refer to these cases as postural torticollis. Such infants have the clinical features of torticollis, but don’t demonstrate tightness or mass in the sternocleidomastoid muscle.

In torticollis, a tight muscle pulls the head laterally to the affected side, limiting the infant’s ability to turn the head toward the affected shoulder. Such repeated and prolonged positioning results in the flattening of one side of the head. For example, if the right side of the neck is tight, the infant will prefer to turn to the left, resulting in a flat appearance to the left side of the head.

Torticollis becomes more evident at about 4 months of age, when infants develop better head control. Most 4-month-olds can hold up their heads when placed in a sitting position and can follow an object placed in their line of vision. Infants with torticollis might tilt the head noticeably and be unable to turn their heads 90 degrees (chin over shoulder) on the affected side. They often compensate by leaning back to view an object.

Treatment

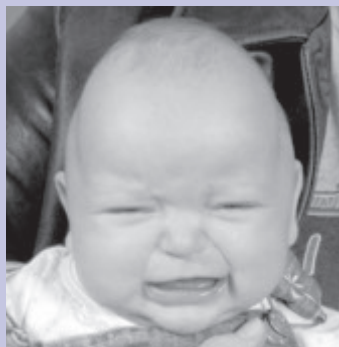
In the majority of cases, physical therapy — which usually takes place once a week — is successful in treating torticollis. Therapists help stretch the muscle on the affected side and strengthen the muscles on the opposite side. After developing a plan of care, therapists teach parents at-home exercises to help infants gain better mobility. Therapy usually lasts for two to three months. In rare cases, when torticollis is persistent and unresponsive to therapy, surgery becomes necessary. In some cases, botulinum toxin injections are effective in relieving torticollis.

Differentiating Characteristics of Craniosynostosis and Deformational Plagiocephaly		
	Craniosynostosis	Deformational Plagiocephaly
Head shape	Asymmetrical	Asymmetrical
Fusion of cranial sutures	Premature fusion of cranial suture(s)	Normal cranial sutures
Diagnosis	Made with X-rays and CT scans	Usually made without X-rays or other imaging studies
Treatment	Surgery	Positioning and/or orthosis therapy
Causes	Some genetic; many unknown	Sleeping on the back, a restrictive intrauterine environment, muscular torticollis, prematurity

Case Studies:

Craniosynostosis

This infant girl presented with sagittal craniosynostosis (premature closure of the sagittal suture of the skull). Children with unrepaired craniosynostosis have a higher risk of increased intracranial pressure, which can result in developmental and functional problems, as well as severe skull deformity.



At 3 months of age, this child underwent a nearly total calvarial remodeling. There were no complications from surgery, and she was discharged after four days. Four and a half months later, she maintains an excellent result, and her scar is nearly undetectable.



Deformational Plagiocephaly

This infant boy presented at 6 months of age with significant flattening of the right posterior of his head. Examination was positive for deformational plagiocephaly. A craniofacial orthosis was made, based on a positive plaster mold of the child's head.



After four months of wearing the orthosis, the child had a normalized head shape.



Volume 14, Number 3
2005

A Pediatric Perspective focuses on specialized topics in pediatrics, orthopaedics, neurology and rehabilitation medicine.

Please send your questions or comments to:

A Pediatric Perspective
Marketing Communications
200 University Avenue East • St. Paul, MN 55101
651-229-1744

Editor-in-Chief.....Steven Koop, M.D.
Editor.....Beverly Smith-Patterson
Designer.....Kim Goodness
Photographer.....Anna Bittner

Copyright 2005, Gillette Children's Specialty Healthcare.
All rights reserved.



Gillette Children's
Specialty Healthcare

200 University Avenue East
St. Paul, Minnesota 55101
651-291-2848
TDD 651-229-3928
1-800-719-4040
www.gillettechildrens.org

Nonprofit
Organization
U.S. Postage
PAID
St. Paul, MN
Permit No. 5388

Upcoming Conferences

Referral Information

Gillette accepts referrals from physicians, community professionals and outside agencies. Contact the Admitting manager at the number listed below. Physicians who are on staff can admit patients through Admitting from 7 a.m. to 4:30 p.m. Physicians who aren't on staff should contact the Admitting manager.

Admitting Manager	651-325-2145
Admitting	651-229-3944
Center for Cerebral Palsy	651-290-8712
• Infant and Toddler Clinic	651-229-3917
Center for Craniofacial Services	651-325-2308
Center for Pediatric Neurosciences	651-312-3176
• Epilepsy Clinic	651-290-8712
• Neuromuscular Clinic	651-312-3176
Center for Pediatric Orthopaedics	651-229-1758
Center for Pediatric Rehabilitation	651-229-3915
Center for Pediatric Rheumatology	651-229-3914
Center for Spina Bifida	651-229-3878
Gillette Lifetime Specialty Healthcare	651-634-1920

Children Who Have Special Needs

A Focus on Diagnoses and Care of Low-Incidence Populations

Parent Workshop – Wednesday, Nov. 2, 2005

Professional Conference – Thursday, Nov. 3, 2005

Best Western – Bigwood Event Center
925 Western Ave., Fergus Falls, Minn.

For course content information, go to our Web site at www.gillettechildrens.org, or contact Cynthia Smith, outreach coordinator, at 218-237-3020 or csmith@gillettechildrens.com. For registration information, contact Amy Schall at 651-229-1721, 800-719-4040 (toll-free), or aschall@gillettechildrens.com.

Sponsored by: Gillette Children's Specialty Healthcare and Minnesota Children With Special Health Needs

13th Annual Pediatric Orthopaedic Update

With hands-on workshops – A course for primary-care physicians

Friday, Dec. 2, 2005

7:30 a.m. to 5 p.m.

Gillette Children's Specialty Healthcare, St. Paul, Minn.

For more information, visit our Web site at www.gillettechildrens.org, or contact Patrick Cavanaugh at 651-229-1758, 800-719-4040 (toll-free), or pcavanaugh@gillettechildrens.com.

Gillette is accredited by the Minnesota Medical Association to provide continuing medical education for physicians.

Online CME Available

A Pediatric Perspective and additional case studies are available for continuing medical education (CME) credit online. To access our online CME, visit www.gillettechildrens.org.

If you're interested in obtaining back issues of *A Pediatric Perspective*, log on to our Web site at <http://www.gillettechildrens.org/default.cfm/PED=1.7.8.1>. Issues from 1998 to the present are available.

Authors' PROFILES



Robert Wood, M.D., is a craniofacial surgeon and medical director of Gillette's Center for Craniofacial Services.

Wood received his medical degree from the University of Minnesota Medical School and completed a general surgery residency at Hennepin County Medical Center in Minneapolis. He trained in plastic surgery at Emory University in Atlanta and completed a fellowship in craniofacial surgery at New York University Medical Center.

Wood is a member of the American Cleft Palate and Craniofacial Association and the American Society of Maxillofacial Surgeons. In addition to being certified by the American Board of Surgery and the American Board of Plastic Surgery, Wood is a fellow of the American Academy of Pediatrics.



Cheryl Shell, C.P.N.P. – Pediatrics, is a pediatric nurse practitioner at Gillette. She provides assessments, and plans and implements care, for children with craniofacial disorders and plastic-surgery needs.

Shell earned a bachelor's degree in nursing from Wichita State University in Kansas and a master's degree — with a focus on advanced practice pediatric nursing — from the University of Minnesota. She is certified by the National Certification Board of Pediatric Nurse Practitioners and Nurses and is a member of the National Association of Pediatric Nurse Associates and Practitioners.