

## Diagnosing and Treating Deformational Plagiocephaly, Torticollis and Craniosynostosis in Infants

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Today, primary care providers are more likely than in years past to see infants who have irregularly shaped heads. One of the most common conditions is posterior deformational plagiocephaly, in which a flat spot develops on the back of a baby's head—sometimes as the result of an infant sleeping exclusively on his or her back (in keeping with the American Academy of Pediatrics' sleep position recommendation intended to reduce the risk of sudden infant death syndrome).

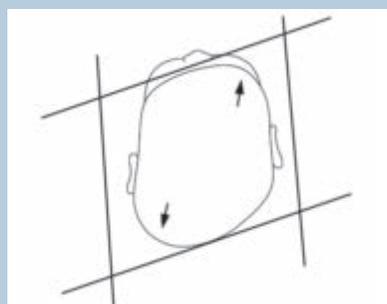
Posterior plagiocephaly may also be related to or accompanied by muscular torticollis, a shortening and tightening of the sternocleidomastoid muscle on one side. A recent retrospective study evaluated patients seen by the author between 1999 and 2012. Of the patients studied, 9,683 were diagnosed with plagiocephaly, and 5,150 (53.2 percent) of those patients also had torticollis.<sup>1</sup>

Craniosynostosis, a congenital condition caused by premature fusion of the cranial bones, is more serious than plagiocephaly, which occurs because of external environmental factors. Craniosynostosis is relatively rare and occurs in approximately one of every 2,000 births.<sup>2</sup> Craniosynostosis can be associated with significant functional and cognitive issues. Head shape issues and associated conditions such as torticollis generally have excellent outcomes when they are appropriately diagnosed and promptly treated.

### Diagnosing Deformational Plagiocephaly

Although the heads of babies who have deformational plagiocephaly may appear flat on one side (e.g., the back), the skull's shape does not affect brain function. In plagiocephaly, typically the sutures are not ridged. The ear on the affected side is sheared forward, and facial features on the same side might be more full. Plagiocephaly results in the characteristic parallelogram shape shown in Figure 1.

**Fig. 1 Nonsynostotic  
Deformational Plagiocephaly**



### KEY INSIGHTS

- Craniosynostosis, a condition in which an infant's cranial bones fuse prematurely, should be repaired when the child is 3 to 6 months old.
- Both craniosynostosis and deformational plagiocephaly can cause a baby's head to appear flat on one side. However, only craniosynostosis has the potential to cause functional and cognitive issues.
- Approximately 50 percent of children who have deformational plagiocephaly also have torticollis, so if plagiocephaly is found, the patient should be evaluated for torticollis.

## Diagnosing Torticollis

Because plagiocephaly and torticollis are so frequently seen in tandem, if plagiocephaly is found, torticollis should specifically be sought. Torticollis might result from the child's position in utero or from trauma to the muscle, but often the cause is unknown.

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 may result 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 who have torticollis might tilt the head noticeably and be unable to turn their heads 90 degrees (chin over shoulder) on the affected side.

## Diagnosing Craniosynostosis

Craniosynostosis occurs when one or more of the sutures between the bones of the cranial vault fuse prematurely. With craniosynostosis, the skull becomes deformed, and brain growth and development may be impaired. If craniosynostosis is not corrected, increased intracranial pressure, seizures and developmental delays can occur.

### Single-Suture Craniosynostosis

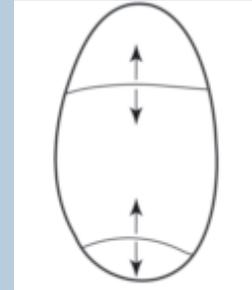
Patients who have single-suture craniosynostosis have distinct head shapes that correlate with the specific suture that has closed. A palpable ridge along the suture is typically present and can be felt upon physical examination. Growth occurs perpendicular to the closed suture, yielding the associated head shape. The four types include scaphocephaly (sagittal synostosis), trigonocephaly (metopic synostosis), synostic anterior plagiocephaly (unilateral coronal synostosis), and synostic posterior plagiocephaly (lambdoid synostosis). See Figure 2a-2d.

### Multiple-Suture Craniosynostosis

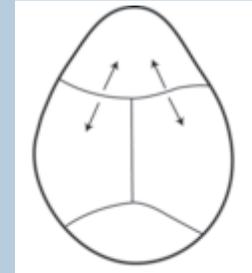
Craniosynostosis is more severe when more than one suture is involved. Multiple-suture craniosynostosis is associated with as many as 70 syndromes. Apert, Crouzon, Pfeiffer, Saethre-Chotzen, and Muenke syndromes are the most common.

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, prominent scalp veins, proptosis, increasing head circumference, apnea, seizures and developmental delays. Left untreated, severe increased intracranial pressure may cause blindness and death. A physical exam is often sufficient to make a preliminary diagnosis of craniosynostosis. A CT scan may be used to confirm the diagnosis.

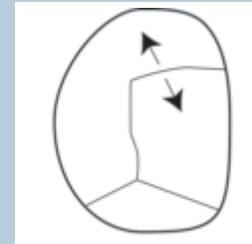
### Types of Craniosynostosis



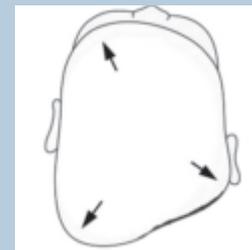
**Fig. 2a**  
Scaphocephaly



**Fig. 2b**  
Trigonocephaly



**Fig. 2c**  
Synostotic anterior plagiocephaly



**Fig. 2d**  
Posterior-lambdoid synostosis

## Treating Deformational Plagiocephaly

For mild cases of deformational plagiocephaly, treatment 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 a 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. The usual length of treatment is two to three months. After children reach 12 or 13 months, an orthosis is less effective and less likely to change the shape of the head.

There are many cranial orthoses available. At Gillette Children's Specialty Healthcare, we use the Gillette CranioCap® orthosis (Figure 3), which Robert Wood, M.D., and Gillette's orthotists designed. Fittings and moldings for these orthoses take place at Gillette.



**Fig. 3** The Gillette CranioCap® orthosis is typically worn for four to 16 weeks. During this period, patients regularly return to an orthotist for adjustments.

## Treating Torticollis

Approximately 50 percent of children who have plagiocephaly will also have torticollis. For most children who have torticollis, physical therapy—which usually takes place once a week—is a successful treatment. 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.

## Treating Craniosynostosis

Typically, craniosynostosis is repaired when the child is 3 to 6 months old. A craniofacial surgeon and neurosurgeon reconstruct the cranial vault. First, a wavy “stealth” incision is made. If the patient has sagittal synostosis, a strip craniectomy will be performed and the bone will be reshaped in situ. For patients with other forms of synostosis, bone will be removed, reshaped and fixed into the native skull using resorbable fixation and screws. Resorbable fixation has greatly reduced operative time. Currently, the procedure can be performed in one to two hours. Resorbable fixation is safe and is associated with few complications and greatly improved outcomes. Prior to 1996, steel or titanium plates were used instead of resorbable material.

A study published in *Plastic and Reconstructive Surgery* in 2004 describes a combined prospective and retrospective analysis of 1,883 patients who had craniosynostosis. The patients were younger than 2 years of age and treated by 12 surgeons from seven geographic locations over a five-year period. The author was one of the participating surgeons. In all cases, the same type of resorbable bone-fixation device (poly-L-lactide/polyglycolic copolymer) was used. The analysis found that only 0.2 percent of patients experienced significant infectious complications, 0.3 percent experienced device instability (primarily resulting from postoperative trauma), and 0.7 percent had self-limiting local foreign-body reactions.<sup>3</sup> At Gillette, we have had no incidents of brain injury or mortality in craniosynostosis repair. Within nine to 15 months after surgery, the polymer fixation will be completely absorbed and replaced by the child's own cranial bone.

## Conclusion

Although craniosynostosis is the most serious infant head shape condition, it is also the most rare. Deformational plagiocephaly, which is often accompanied by torticollis, has become more common in recent years. Outcomes are usually excellent for these conditions as long as treatment is begun within the first three to six months of the child's life.

## Author PROFILES



### **Robert Wood, M.D.**

Robert Wood, M.D., is a craniofacial surgeon and program director for the Center for Craniofacial Services at Gillette Children's Specialty Healthcare. Prior to joining Gillette, he served as director of the Emory

Egleston Center for Cleft and Craniofacial Anomalies in Atlanta, Ga. One of the most experienced craniofacial surgeons in the country, he has particular expertise in treating craniosynostosis, plagiocephaly, torticollis, and cleft lip and palate.

Active in craniofacial research, Wood helped pioneer current techniques in endoscopic surgery to remove facial masses. He also developed an intra-oral distractor and has an international reputation for his experience with resorbable fixation. He worked closely with orthotists at Gillette to develop the CranioCap® orthosis to correct deformational plagiocephaly in infants.

He is board-certified in plastic surgery by the American Board of Plastic Surgery. He is a Fellow of the American Academy of Pediatrics (FAAP), a Fellow of the American College of Surgeons (FACS), and a member of numerous other professional societies. In addition, he is a clinical associate professor at the University of Minnesota.

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<sup>1</sup> Wood, R, Chibbaro G, McGrory M. Plagiocephaly, torticollis, lambdoid craniosynostosis: A spectrum of disease. A review of 9,683 patients with posterior plagiocephaly. Abstract presented at the International Society of Craniofacial Surgery, Sept. 10-14, 2013.

<sup>2</sup> Jane JA Jr, Wyler AR. Surgery for craniosynostosis. *Medscape Reference*. Available at: <http://emedicine.medscape.com/article/248568-overview#a0199>. Accessed May 3, 2012.

<sup>3</sup> Eppley B, Morales L, Wood R, et al. Resorbable PLLA-PGA plate and screw fixation in pediatric craniofacial surgery: clinical experience in 1883 patients. *Plast Reconstr Surg*. 2004;114(4):850-6.

## Study of Nearly 10,000 Patients Who Have Head Shape Conditions Yields Valuable Insights

Gillette's craniofacial program launched nearly 15 years ago under the guidance of Robert Wood, M.D. Our program serves patients from the upper Midwest and across the nation who have complex congenital conditions ranging from cleft lip and palate to craniosynostosis. We also care for patients who have rare syndromes that affect facial features and head shapes, including Apert, Pfeiffer and Crouzon syndromes.

Because we have one of the largest programs in the country—treating nearly 10,000 patients in 15 years—we have the opportunity to gather extensive retrospective data. The author presented these findings at the 23rd annual meeting of the American Society of Craniofacial Surgery:

- From 1999 to 2012, we saw 9,683 patients (mean age of 7.4 months) who had plagiocephaly.
- A total of 5,792 patients with torticollis were seen during this time frame.
- Of the 9,683 patients who had plagiocephaly, 5150 (53.2 percent) also had torticollis.
- A total of 36 patients (0.4 percent) were noted to have lambdoid craniosynostosis.
- Of the patients who had lambdoid craniosynostosis, 30 (83 percent) also had torticollis.

The preponderance of evidence points to the relationship between torticollis and posterior plagiocephaly. Lambdoid craniosynostosis is rare, but within this population, torticollis is common.

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## NEWS & NOTES

### ***Gillette Welcomes Barbara Joers***

In her new role as CEO of Gillette, **Barbara Joers** looks forward to contributing to Gillette's reputation for providing the best care to children who have disabilities and other complex conditions. She plans to reach out to community leaders to enhance their awareness of Gillette's services and reputation.

### ***Craniofacial and Plastic Surgery Care Provided at All of Gillette's Metro Clinics***

Gillette's team of craniofacial specialists treats pediatric patients who have cleft lip and palate, craniosynostosis, deformational plagiocephaly, hemangiomas and vascular anomalies, and torticollis. Patients can receive care at our clinics in Burnsville, Maple Grove, Minnetonka and St. Paul.