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Deformational Plagiocephaly or Misshapen Head: Integrated approach to diagnosis and treatment essential to ruling out more serious conditions and obtaining optimal outcomes

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fig. 3-

Craniosynostosis

Ear shearing is often diagnostic.



Deformational Plagiocephaly

The contralateral ear shears away from deformational plagiocephaly.

Less than 10 years ago, deformational plagiocephaly, or misshapen head, was not seen in a large number of infants. When it did occur, it was often the result of intrauterine crowding caused by conditions such as multiple fetuses, large infant size, a small pelvis in the mother, or a breech presentation. This crowding inhibited the baby from moving in the uterus, potentially resulting in the head flattening on the side pressed against the uterine wall.

Congenital muscular torticollis (CMT) has also been associated with secondary deformational plagiocephaly. With CMT, the tightening or shortening of a muscle on one side of the neck can lead to the infant holding or turning their head consistently in one direction, including against their mattress, which can cause deformation of the skull over time.

Similarly, plagiocephaly is also linked to prematurity. For one, the skull of the premature infant is exceptionally malleable (the firmness of the skull increases markedly during the final weeks of pregnancy). Secondly, infants on ventilators often have 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 head shape.

Since 1992, however, the incidence of deformational plagiocephaly has increased dramatically. This increase followed on the heels of the American Academy of Pediatrics' recommendation that babies sleep on their backs to reduce the risk of Sudden Infant Death Syndrome. The constant pressure on the back of the head caused by back-sleeping has been shown to result in plagiocephaly in some babies. By the mid-1990s, studies linking back-sleeping and plagiocephaly led to new recommendations by the AAP that parents occasionally turn their child's head during sleeping to avoid deformation.

Ruling out craniosynostosis is crucial

While plagiocephaly is often the result of consistent pressure to one side of the head, it is also a sign of craniosynostosis, or premature closure of the sutures in an infant's skull. Misdiagnosis can have serious ramifications. While brain damage is not a risk with deformational plagiocephaly, the possibility of such damage in untreated craniosynostosis is very real.

Plagiocephaly in craniosynostosis is the result of the growing brain pushing against a skull that cannot expand normally, due to suture fusion. The result is the compensatory bulging of a child's head on one or both sides. Because the skull is resistant to the pressure exerted by the growing brain, brain damage is possible from increased intracranial pressure. While there are certain distinguishing characteristics between deformational plagiocephaly and craniosynostosis (such as positioning of the ears as shown in fig. 3), only x-rays can confirm the presence of craniosynostosis. If the latter condition is diagnosed, surgery is the sole treatment option to release the sutures and remodel the skull.

The risk of misdiagnosing craniosynostosis makes an integrated physician-directed approach to assessment and treatment imperative. Consultation between the child's pediatrician and a pediatric craniofacial specialist (and a pediatric neurosurgeon if indicated) is wise before proceeding with treatment for deformational plagiocephaly.

When is it necessary to treat deformational plagiocephaly?

During the first three to four months of life, consistent repositioning of the infant's head during sleep often leads to spontaneous rounding of a skull that has mild flattening. If the deformation is significant, or fails to correct itself by four months of age, a craniofacial orthosis is an effective means of remodeling the shape of the baby's head. Nevertheless, because slight variations in head shape and size are considered normal, use of a craniofacial orthosis should be selective, and is indicated primarily in cases where failure to treat would leave the child with an enduring abnormal appearance.

The basic premise behind craniofacial orthoses goes back thousands of years, when various ancient cultures used molding devices to artificially shape an infant's head for aesthetic purposes (such as elongating the head as a sign of royal heritage). Today, craniofacial orthoses require approval from the Food and Drug Administration, meeting strict guidelines for safety, design and effectiveness of use.

The CranioCap™ is a custom-made orthosis created from a plaster-cast model of the infant's head (see fig. 4 & 5). Pads are placed inside the CranioCap™ so it fits snug in some areas and is open in others. As the baby's brain grows, the skull is slowly pushed into the open areas of the orthosis, rounding the skull. It generally takes between two and four months for the process to be completed.

The optimal age for beginning treatment with a craniofacial orthosis is between the ages of 4 and 8 months, when the skull is still quite malleable. Beginning at a later age requires a longer treatment period to achieve desired results. After the age of 12 or 13 months, an orthosis is not as effective and is less likely to change the shape of the baby's head. In most cases, corrections of head shape after 1 to 2 years of age must be done surgically.

For more information about the CranioCap™ or other craniofacial programs at Gillette Children's, call (651) 229-3905. To reach Dr. Robert Wood directly, call (651) 602-3277. Or to make a referral, call (651) 229-3944.

The making of a CranioCap™ craniofacial orthosis:

A positive plaster mold is made of the infant's head (fig. 4). A foam pad is then made for the correction and placed on the mold prior to the making of the craniofacial orthosis. Next, plastic is "blister formed" over the plaster mold and foam padding (fig. 5).

The CranioCap™ is typically worn for 4 to 16 weeks. During this period, patients are regularly scheduled for return visits with an orthotist for adjustments (fig. 6).



fig. 4



fig. 5



fig. 6