

Infant Heads

Too Big, Too Small, Misshapen

Joseph H Piatt, Jr, MD, FAAP

Professor of Neurosurgery and Pediatrics, Drexel University College of Medicine
Chief, Section of Neurosurgery, St Christopher's Hospital for Children

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Skull deformities are by far the single most common reason for referral of new patients to the pediatric neurosurgeon. Although parents bring to the first appointment the most alarming mental images of the surgical reconstruction of their baby's skull, the great majority of cases can be managed with less aggressive measures. A basic understanding of the causes and treatments of infantile skull deformities enables the primary physician not only to make an appropriate referral in a timely fashion but also to shape parental expectations appropriately.

The infant skull is composed of 7 flat membranous bones joined along their borders by condensations of dura and pericranium called "sutures." The sagittal suture forms the border between the paired parietal bones. The metopic suture lies between the frontal bones. Between the frontal and the parietal bone on each side is the coronal suture. The lambdoid sutures separate the parietal bones from the occipital bone. A squamosal suture separates the parietal bone from the temporal bone on each side. Sutures also join the bones of the skull vault with the cartilage-derived bones of the skull base. At the intersection of sutures are larger membranous areas called fontanel. The largest 2 fontanel occur at the intersection of the sagittal, coronal, and metopic sutures – the anterior fontanel – and at the intersection of the sagittal and lambdoid sutures – the posterior fontanel.

The membranous bones of the calvarium grow and take shape in response to pressures; they have little intrinsic programming to develop into predetermined forms. The most important pressure is the internal pressure applied by the growing brain. The pressure of the growing brain causes tension in the overlying

calvarium that results in the deposition of new bone along the sutures and over the external surface of the skull. Furthermore, the pulsations of the growing brain cause resorption of bone along the internal surface of the skull. The growth of the brain is the principal determinant of the size and shape of the calvarium. In the absence of pathological constraints, the calvarium expands diffusely into a normal ellipsoidal shape. If the growth of the calvarium is restricted in a certain dimension by one of the processes described below, the brain will push outward in the direction of least resistance and cause one of a relatively small set of distinctive patterns of calvarial deformity.

Craniosynostosis

Premature ossification of a suture, or synostosis, is sometimes attributed to intrauterine restraint of head growth by twinning or by early engagement of the head in the pelvis. It can occur in the setting of complex, eponymic, inherited craniofacial disorders such as Crouzon, Apert, Pfeiffer, or Saethre-Chotzen syndromes, to mention the most common, or in the setting of fetal exposure to phenytoin, valproic acid, or other teratogens. Most often, however, there is no identifiable cause. Involvement of a single suture is the rule in sporadic cases, but in syndromic cases more complex patterns of involvement are common, particularly bilateral coronal synostosis.

The expansion of the head is driven from within by growth of the brain, and craniosynostosis causes deformity by restricting expansion of the head in the dimension perpendicular to the affected suture. Compensatory pressure by the growing brain in the unrestricted dimensions further complicates the

associated deformity. Involvement of each particular suture causes one of a small number of characteristic patterns of deformity that are readily recognized by the trained eye. Thus the diagnosis of craniosynostosis is based on physical examination. Skull radiography and computed tomographic (CT) scanning of the skull are not only unnecessary, but their misinterpretation is a common source of confusion and alarm for parents. The least expensive and most conclusive strategy for investigating a skull deformity is referral to an experienced pediatric neurosurgeon or craniofacial plastic surgeon.

Without surgical intervention, the skull deformities caused by craniosynostosis persist through life. The trained eye can spot adults with never-treated craniosynostosis in large crowds in public places: Sagittal synostosis causes elongation and narrowing of the skull that can attain grotesque proportions in childhood and are not completely mitigated by normal enlargement of the facial skeleton and thickening of the neck muscles later in life. Unilateral coronal synostosis causes asymmetries of the forehead and orbits and rotation of the nose that do not disappear with maturation. (The natural history of nonsyndromic metopic synostosis is uncertain. Older children and adults with trigonocephaly are not seen, and some surgeons have abandoned the treatment of this condition.)

Of greater concern to parents than the persistence of deformity into adulthood are social issues during the school years. Children who are different in any way tend to become the objects of ridicule and ostracism by their peers, and the skull deformities caused by craniosynostosis do not escape the attention of schoolmates. Parents are usually willing to subject their infant children to surgery in order to remove obstacles to social integration, but such an indication for surgery places heavy demands on the surgical team with respect both to safety and aesthetic results. Treatment of craniosynostosis requires not only surgical expertise but also the highest quality support in pediatric anesthesia and pediatric intensive care.

The goal of the modern surgical treatment of craniosynostosis is immediate correction of the skull deformity. In the past surgical intervention was limited to synostectomy, so-called "reopening" of the affected suture. The rationale for this approach was that removal of the synostotic constraint on skull development would allow subsequent brain growth to remodel the skull. Treatment by synostectomy put a very high premium on early diagnosis, in order to take maximal advantage of remaining brain growth potential. However sound the theory may have been, the results were poor, and contemporary surgeons have abandoned synostectomy in favor of a variety of techniques for active remodeling of the skull. Patients now look better in the recovery room. This shift in surgical philosophy has been boosted by an explosion

of new instruments and materials that have allowed intervention not at the earliest possible moment but at the point in skull development mechanically most satisfactory for reconstruction. Postponement of surgery until 6 to 9 months of age increases the margin of safety as well.

Although the data are lower quality than one might wish, craniosynostosis involving a single suture does not seem to present any obstacle to the growth and development of the brain. On the other hand, involvement of 2 or more sutures, as in Crouzon, Apert, and the related syndromes, is commonly associated with elevated intracranial pressure due to insufficient accommodation of brain growth by the deformed skull. These eponymic craniofacial syndromes may be complicated by hydrocephalus as well. Careful clinical surveillance is required both before and after surgical treatment in order to prevent blindness and cognitive disabilities due to insidious intracranial hypertension. Periodic neurosurgical and ophthalmological follow-up is indicated from birth at least until school age.

The management of children with craniosynostosis often calls for a multidisciplinary approach. Deformities of the orbits and other portions of the facial skeleton require the expertise of the craniofacial plastic surgeon. Orbital deformities are frequently associated with strabismus, which engages the attention of the pediatric ophthalmologist. Syndromic forms of craniosynostosis entail underdevelopment of the midface with airway insufficiency and Eustachian tube dysfunction that involve the pediatric otolaryngologist. Developmental delays, speech disturbances, and feeding disorders complicate syndromic cases as well.



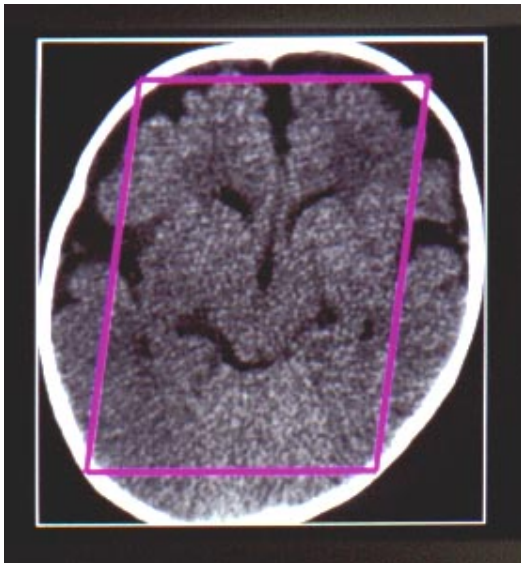
At St Christopher's Hospital for Children, patients with craniosynostosis and other skull deformities receive multidisciplinary care through the Craniofacial Clinic (pictured from left to right: pediatric dentist David Metroka, DDS; coordinator Brenda Allgood, MSN, CRNP; pediatric neurosurgeon Joseph Piatt, MD, FAAP; and craniofacial plastic surgeon Paul Glat, MD, FACS).

Positional Skull Flattening

The mechanical difference between an infant's head and a water balloon is only a matter of kinetics. A water balloon laid on a table conforms to the contour of the table instantly. The infant head laid on a firm mattress takes weeks to conform, but if the infant is sufficiently immobile, the result is the same.

Flattening of the posterior aspect of the skull in early infancy is by far the most common reason for referral of new patients to the pediatric neurosurgeon; it is more common than craniosynostosis by an order of magnitude. The literature mentions this condition by a number of names: posterior plagiocephaly, nonsynostotic occipital plagiocephaly, posterior positional plagiocephaly, and others. Use of the term positional skull flattening is particularly suitable for communication with families because it avoids jargon and conveys a mechanism.

Positional skull flattening is not difficult to recognize. Like the various forms of craniosynostosis, positional flattening causes a characteristic pattern of deformity that the trained eye can distinguish without radiological investigations. Gravity exerts a deforming force on the entire skull, not just on the dependent, flattened surface. The contralateral frontal aspect of the skull often becomes somewhat flattened as well, and prominences develop in the contralateral occipital region and the ipsilateral frontal region. Viewed from above the skull looks like a parallelogram.



The ipsilateral frontal prominence is occasionally the feature of the deformity that families find most distressing even though the posterior flattening is more severe. Other components of the facial skeleton may become deformed: The ear ipsilateral to the flattening is displaced anteriorly compared to the other

ear. The nose feels the tug of gravity and rotates in the plane of the face. Like the ipsilateral ear the ramus of the mandible on the side of the flattening may be pushed forward, and the jaw may be asymmetrical. Concerns have been expressed about the possibility of dental malocclusion later in childhood, but the actual prevalence of this problem is unknown. The differential diagnosis of positional skull flattening is synostosis of the lambdoid suture. Fortunately for the busy primary physician, because lambdoid synostosis is so vanishingly rare, for practical purposes all predominantly posterior skull deformities in infants are positional.

Infants are occasionally born with posterior flattening, which implies earlier immobilization of the fetal head in a tight place somewhere, most likely against the sacral promontory. More often the head is round (or normally molded) at birth and becomes flat in the first couple months of life as the infant lies supine for naps in compliance with the recommendations of the American Academy of Pediatrics's "Back to Sleep" campaign. Conjunction of developmental immobility with a stiff neck – so-called "congenital torticollis" – leads to asymmetrical flattening, which is the implication of the term plagiocephaly. Once flattening develops, it perpetuates itself, as the infant must overcome an even greater moment of inertia to rotate the head off the flat surface. In the second half of infancy, with the developmental acquisition of independent sitting and active rolling from supine to prone, the deformity ceases to progress. Subsequent brain expansion remodels the skull and reduces the flattening somewhat, and the asymmetry that persists is mitigated further by disproportionate enlargement of the neck muscles and the facial skeleton and by hair growth. Whether even the most severe positional skull flattening ever leads to significant deformity in developmentally normal school age children is doubtful.

The best treatment is prevention. In the first few months, sleep position should be varied by propping the infant slightly to one side or the other with a folded towel or blanket. The infant who has already developed flattening should be propped to the opposite side. Behavioral interventions to encourage active rotation of the head off the flattened aspect can be helpful as well. For instance, most right-handed parents carry small infants in the crook of the left arm so that the infant must rotate the head to the right to gaze at the parent's face. Likewise, most right-handed parents lay infants down with the head to the parent's left, forcing the infant to rotate the head to the right to look out into the room. Probably not coincidentally, the great majority of asymmetrical infant heads are flattened on the right side. These right-handed routines should be varied. Infants quickly learn to defeat positional manipulations beyond 4 or 5 months of age. Asymmetrical positional flattening is almost always associated with a degree of congenital torticollis, and

passive range of motion exercises for the neck are useful at any age. These exercises can be taught and reinforced by a physical or occupational therapist, and they should be performed several times daily, as with diaper changes.

Externally applied forces cause positional flattening, and externally applied forces can be utilized to correct the deformity. The "orthotic helmet" is a padded polypropylene band that is custom fabricated to fit snugly over the prominent aspects of the infant skull and to leave daylight over the flattened aspects.



The infant wears the helmet continuously with interruptions only for bathing, and over the course of several months, as the brain grows, the head expands into the daylight and becomes more rounded. Minor adjustments in the fit of the helmet are required on a roughly bi-weekly basis to prevent pressure injuries to the scalp. Data suggest that orthotic helmet therapy is no more effective than positioning and exercises in the management of mild to moderate deformities, and the frequent visits to the orthotist are a burden to families from remote communities, so the author reserves this intervention for the most severely affected patients.

Surgical interventions have virtually no place in the management of positional skull flattening. "Unipolar" or "bipolar release" of the sternomastoid muscle has been recommended for congenital torticollis caused by scarring and contracture due to birth injury. The distinction between functional and contractural shortening of the sternomastoid is not very clear in practice, so the indications for surgery have always been indefinite, and the superiority of surgical release to physical therapy or to the untreated natural history

of this condition has never been demonstrated. The surgical treatment of posterior skull flattening has a dubious history as well. In the 1980s and early 1990s surgeons at a few centers were advocating a variety of synostectomies and reconstructive procedures for posterior plagiocephaly under the diagnostic label of "lambdoid synostosis." The Center for Disease Control actually investigated an epidemic of lambdoid synostosis in the Rocky Mountain region, concluding discretely that there were disagreements about the use of this diagnostic term. As the nature of posterior plagiocephaly has become better understood, and as the generally benign natural history has been appreciated, surgical indications have all but disappeared.

Microcephaly

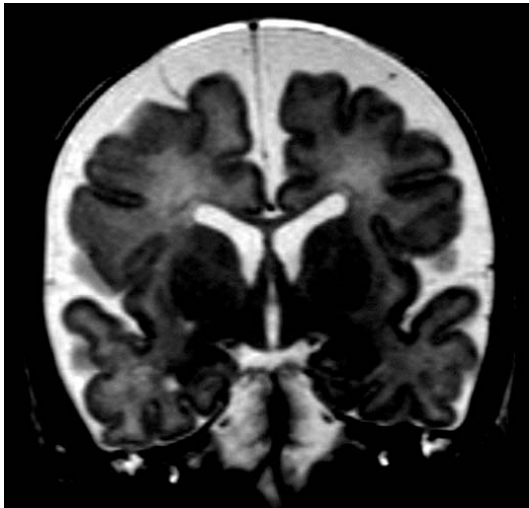
There are no diseases of the skull that cause global, diffuse failure of head growth without any associated deformity. A symmetrical, normally shaped head that is failing to grow contains a brain that is failing to grow. "Microcephaly," although commonly used and widely understood, is not such an appropriate term for this situation as "micrencephaly." Not all micrencephalic infants are abnormal in any more than a statistical sense: 5% of all normal infants have heads smaller than the 5th %ile. Unfortunately, if newborn screening has eliminated the possibility of congenital hypothyroidism, the remainder of micrencephalic infants have brain diseases for which diagnoses are seldom available and treatments almost never. Micrencephaly lies within the scope of pediatric neurology, not pediatric neurosurgery.

Macrocephaly

The volume of the contents of the calvarium determines the size of the head. The brain and the cerebrospinal fluid (CSF) are the normal contents of the calvarium, and too much of one or the other accounts for most cases of macrocephaly. As is true of microcephaly, many macrocephalic infants are not abnormal in any more than a statistical sense. "Constitutional" macrocephaly is fairly common and tends to be familial; there are no treatment implications, of course. There are a small number of very rare diseases of the brain, such as Alexander's disease and Canavan's disease, that cause macrocephaly from proliferation of glial tissue or from deposition of the product of a metabolic disturbance. These conditions are seen in the context of catastrophic developmental regression, and they are untreatable.

Most cases of macrocephaly are attributable to excessive volumes of CSF. The excessive CSF may reside within the brain, in the ventricular cavities, or outside the brain, in the subarachnoid spaces. The former condition is ordinary, "internal"

hydrocephalus. It generally requires neurosurgical treatment with a CSF shunt, and because of it is the subject of another article in this set, it will receive no further discussion here. The latter condition has been known by a variety of names including - among others less appropriate - "benign enlargement of the subarachnoid spaces" and "external hydrocephalus."



In this coronal, T2-weighted magnetic resonance (MR) image of an infant's head, the light gray space outside the brain is the subarachnoid space, filled with the CSF in which the brain floats. This infant has external hydrocephalus, and the subarachnoid space is much deeper than normal. The brain does not fill the cranial cavity. At the top of the image, the thin, white crescents are probably very tiny chronic subdural hematomas.

External hydrocephalus may be almost as common as ordinary hydrocephalus, and like constitutional macrocephaly, it is commonly familial with autosomal dominant inheritance. The prominence of the subarachnoid spaces is a transient, developmental phenomenon that usually resolves by school age. The macrocephaly, however, is permanent, although it does not attain grotesque extremes. External hydrocephalus does not require surgical intervention; however, the failure of the brain to fill the cranial cavity in infancy creates an unstable biomechanical situation that parents must understand: Infants with external hydrocephalus have a low threshold for subdural hemorrhage in response to minor impacts to the head. If the disproportion between the volume of the brain and the volume of the cranial cavity is marked, a protective helmet may even be indicated during the early stages of the acquisition of ambulation.

The most menacing element of the differential diagnosis of macrocephaly is chronic subdural hematoma. Blood spilled in the subdural space may be

degraded and reabsorbed spontaneously, but in some patients it stimulates the formation of a capsule made up of cellular elements derived from the dura above and, not so vigorously, from the arachnoid membrane below. Blood degradation products in the hematoma cavity have fibrinolytic activity, promoting continuous microscopic hemorrhage from the fragile capillaries in the reactive capsule. Thus if the initial trauma is unrecognized or unreported, chronic subdural hematoma may become a self-propagating, progressive condition that can cause insidious enlargement of the unclosed infant skull with few or no overt symptoms of intracranial pressure. The treatment of chronic subdural hematoma entails irrigation of the hematoma cavity to eliminate the fibrinolytic substances that promote its expansion and drainage of the cavity - either temporary external drainage or permanent internal drainage with a shunt. The prognosis depends on the neurological condition of the infant at the time of treatment: If macrocephaly is the sole presenting complaint, the long-term outlook may be quite good. Infants who come to attention because of macrocephaly and are found to have chronic subdural hematoma must be managed as suspected victims of child abuse.

The imaging investigation of macrocephaly in infancy is problematic. The tendency of many primary practitioners is to request simple, relatively inexpensive tests that, unfortunately, are often inconclusive or misleading. Computed tomography (CT) demonstrates extracerebral fluid reasonably well, but only the very highest quality scans permit confident distinction between external hydrocephalus and chronic subdural hematoma and thereby obviate the requirement for MR imaging. Head ultrasound (US) is a satisfactory initial study for the macrocephalic infant, so long as it can be performed with Doppler recordings. Head US demonstrates extracerebral fluid in the interhemispheric fissure and over the medial convexities, and if the Doppler flow signals from bridging veins can be detected passing through the extracerebral spaces, the diagnosis of external hydrocephalus is established. Alternatively, if the extracerebral fluid is full of debris, and if the Doppler signals from the cerebral veins are present only adjacent to the surface of the brain, the diagnosis is chronic subdural hematoma. Head US can demonstrate the ventricular enlargement of ordinary hydrocephalus, but unless the cause of the hydrocephalus is known from the clinical context, MR imaging will be required to complete the evaluation. MR imaging is by far the most sensitive study for investigation of macrocephaly, and if high-quality ultrasonography is not available or possible, MR imaging is the modality of choice.

"Abnormalities" of the Anterior Fontanel

The anterior fontanel ordinarily closes sometime in the second half of infancy. Closure of the anterior fontanel

can be accelerated by micrencephaly and by craniosynostosis. It can be retarded in the settings of macrocephaly and a variety of rare dysmorphic syndromes. As an isolated observation, however, on a head that is normal in shape and size, the early disappearance or late persistence of the anterior fontanel is not a sign of disease. The status of the anterior fontanel is never, by itself, an indication for diagnostic investigation.

Management Suggestions for the Primary Physician

- The most expedient and inexpensive investigation of a skull deformity in infancy is referral to an experienced pediatric neurosurgeon or craniofacial plastic surgeon without preliminary imaging studies.
- Infants with craniosynostosis, particularly when associated with craniofacial syndromes, frequently have disturbances of

vision, hearing, speech, and development. They deserve multidisciplinary care coordinated through an organized craniofacial program.

- Microcephaly without skull asymmetry or other deformity is not a surgical condition. Consultation with a pediatric neurologist may be indicated.
- High-quality head ultrasonography with Doppler is an adequate diagnostic study for most infants with macrocephaly. Magnetic resonance imaging is the most sensitive imaging modality.
- On a head that is growing normally without deformity, the early disappearance or late persistence of the anterior fontanel is of no concern.

See PowerPoint presentation:

[“The Child with an Abnormally Shaped Head”](#)

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