

DIAGNOSTIC LAND MINES

Related to the Spine and Head

by Michael Partington, M.D., and Joseph Petronio, M.D.

This issue of A Pediatric Perspectives discusses diagnostic "land mines" related to pediatric neurological conditions. On the surface, these conditions may appear to have simple, straightforward diagnoses; however, in some instances, they may require further exploration.

For many clinical presentations, one common, benign cause can be diagnosed, but some diagnoses are more ominous. Implementing appropriate treatment is crucial and can result in more optimal outcomes.

Below, we review several potential neurological conditions that can underlie other, more common conditions.

Minor Cervical Spine Injury

Pediatric neurosurgeons are commonly consulted regarding the problem of pain in a child or teenager with minor mechanism cervical spine trauma. Most children who are assessed in an emergency department setting will undergo cervical spine X-rays, and often no fracture is identified. It is these patients who represent potential "land mines" because, although major cervical spine trauma is rare in childhood, the immature cervical spine is more prone to ligamentous injury than to fracture.

In the acute setting, the pain and accompanying spasm may cause "splinting" of the spine and may mask the underlying unstable injury. In view of this small risk, we generally recommend keeping a child in a hard collar until the pain resolves. At that point, flexion and extension lateral views of the cervical spine should be obtained. If there is no evidence of instability or new deficit, the collar can be safely removed. If pain persists, or if instability or a new deficit is found, the child should be kept in the collar and referred for further evaluation.

Scoliosis

The majority of children with scoliosis are appropriately referred to pediatric orthopaedists and/or spine surgeons for definitive management of their spine condition. It should be

noted, however, that a number of these children harbor intraspinal pathology, which contributes to their spinal disorder. The disorder may improve if the underlying pathology is removed.

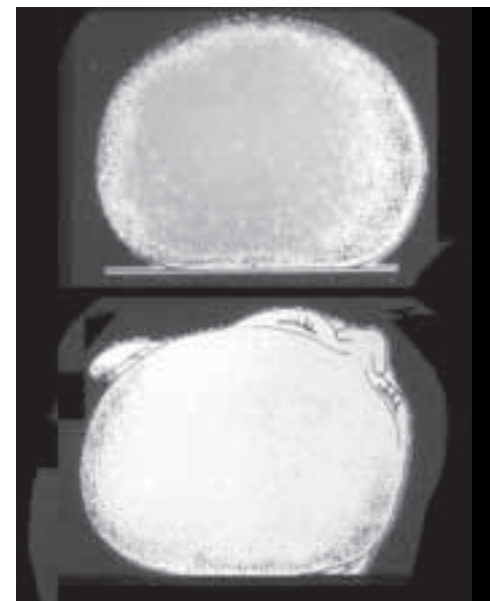
A magnetic resonance imaging (MRI) screening should be considered, not only in those cases with pain or neurological deficits, but also in cases with a severe curve, a rapid progression or early onset. The most common intraspinal finding is a Chiari malformation, type I, often with syringomyelia. Other findings include tethered spinal cord, split cord malformations, spinal lipoma and spinal cord tumor.

Plagiocephaly

The term plagiocephaly refers to lateral asymmetry of the skull.

It can occur in the anterior or posterior area of the skull. The most common cause of posterior plagiocephaly is deformational (the so-called "positional molding").

Deformational plagiocephaly can often be distinguished from pathologic conditions, such as lambdoid synostosis, by several clinical features. With the former, the skull often assumes a polygonal shape. When viewed from above, especially in more severe cases, there is often a flattening of the forehead and cheek on the side opposite the flat



Positional plagiocephaly: Like the water-filled balloon (top), the visco-elastic properties of the brain and infant skull set up gradual flattening of the dependent occipital region with rounding out of the ipsilateral forehead.

occiput. The ear on the affected side also is usually displaced or moved forward, in a deformational or molding process, or toward a closed or fused suture (craniosynostosis).

Plagiocephaly may be associated with congenital torticollis. Treatment is usually centered on treatment of the torticollis (physical therapy for range of motion, or surgical release of the sternocleidomastoid muscle in more severe cases) and repositioning of the infant. Imaging studies, such as skull films with a Towne view or a cranial computed tomography (CT) scan, are warranted in moderate to severe cases. Infants with posterior displacement of the ear on the same side as the posterior flattening, or significant bony ridge formation in the region of the lambdoid suture, should be referred for craniofacial evaluation.

Moderate to severe cases may require an orthotic molding helmet, such as the Gillette CranioCap™. Craniosynostosis, a pathologic condition in which the sutures fuse prematurely, usually requires surgical correction.

Sacral Dimples

A variety of birthmarks, found overlying the spine in the midline, mark an underlying occult dysraphism (a congenital spinal cord abnormality that causes progressive neurologic dysfunction). These include dimples, dermal sinus tract openings, hemangiomas, lipomas, tails and hairy patches.

Of these, only the sacrococcygeal dimple can be safely ignored. The sacrococcygeal dimple in humans represents the end of the embryo. It's the point where the ectoderm and the skeleton come together. In all other mammals — except for the great ape — it's also the point where the tail attaches to the body. To be considered a benign sacrococcygeal dimple, it must be below the top of the gluteal cleft, have a shallow, visible base, and be located at the level of the coccyx. Furthermore, there should be no accompanying neurological or skeletal abnormalities.

A useful clinical feature for the examiner to note is that when the skin around a sacrococcygeal dimple is pulled upward (i.e. cephalad), the dimple closes. Downward traction opens the mouth of the dimple. The converse is true for higher, non-benign dimples, which overlie some form of occult dysraphism or may be the opening of a dermal sinus tract.

If the exam features are consistent with a benign sacrococcygeal dimple, no further work-up is needed. If any other feature is identified, obtain an MRI of the entire spine and a pediatric neurosurgical referral. One further caveat: an open tract should never be probed or flushed, because of the risk of transmitting skin bacteria into the central nervous system if the tract extends intradurally.

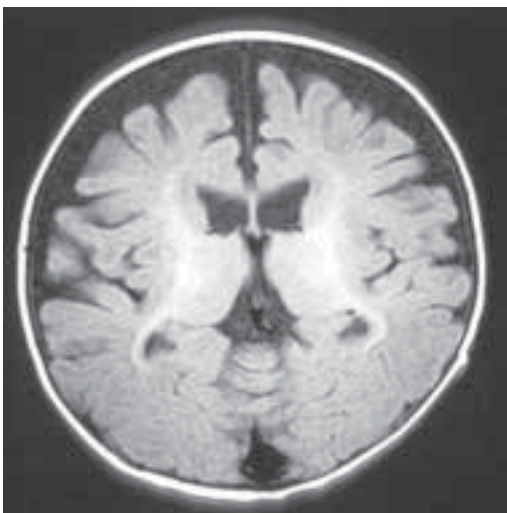
Macrocephaly

There are many known causes of macrocephaly. The most common one seen in routine pediatric practices is the so-called “benign macrocrania.” Infants with this condition often have a head circumference at or above the 95th percentile for their age. Neurologically and developmentally, these infants may lag a few months behind their peers with regard to gross motor skills. Nevertheless, they continue to make developmental progress.

Imaging often reveals enlargement of the spinal fluid spaces within the skull, including the extra-axial subarachnoid spaces (over the surface of the brain) and



This atypical sacral dimple is too high to be considered benign. An MRI diagnosed a spinal lipoma.



This MRI image shows symmetrical bifrontal subarachnoid cerebral spinal fluid collections, typical of benign external hydrocephalus.

the cerebral ventricles (within the brain). These extra-axial collections are always subarachnoid as opposed to subdural in location (including those related to child abuse or prior meningitis). They can be distinguished radiographically by their tendency to symmetrically involve the spinal fluid spaces between the two hemispheres of the brain.

Benign macrocephaly usually stabilizes by the second to third year of life, with the head circumference curve approaching the 95th to 98th percentile by age 3. Children with mild macrocrania, who are normal developmentally and neurologically, often can be followed closely with careful attention to developmental progress and neurologic examinations.

Children who are at greater risk for developing severe macrocephaly and need to be referred for a neurosurgical evaluation include those with other signs and symptoms of intracranial hypertension. These symptoms include a persistently bulging anterior fontanelle or separated cranial sutures, abnormal eye movements, seizures, failure to meet developmental milestones, unexplained lethargy, vomiting and irritability. If untreated, chronic intracranial hypertension can result in neurologic disabilities, including cerebral palsy, seizures, blindness or even death.

Headaches

Headaches are common in children of certain age groups, especially those ages 10 to 17, and may be attributable to a number of causes, including infections such as pharyngitis, sinusitis and meningitis. Other causes may be allergic reactions and vascular conditions, including childhood migraines.

Certain headaches warrant an imaging study. They include those that:

- Are worse in the morning
- Occur when lying down
- Occur regularly or daily
- Are accompanied by recurring nausea or vomiting
- Involve alterations in levels of consciousness
- Result in abnormal neurologic exams, including focal deficits, papilledema and ataxia

These symptoms could indicate elevated intracranial pressure or a structural abnormality, such as hydrocephalus, brain tumor, Chiari malformation or intracranial infection.

In addition, headaches are significantly less common in children under the age of 10. Any child with recurrent or frequent headaches should receive a thorough neurologic exam. Any abnormal findings or lack of an identifiable source of the headaches should mandate an imaging study.

Authors' Profile

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Michael Partington, M.D., and Joseph Petronio, M.D., are pediatric neurosurgeons at Gillette Children's Specialty Healthcare in St. Paul, Minn.

Partington graduated from the University of Minnesota Medical School and served a residency in neurosurgery at Mayo Graduate School in Rochester, Minn. He held a research fellowship and a clinical fellowship in pediatric neurosurgery at Children's Memorial Hospital in Chicago and has practiced at The Children's Hospital in Denver. Partington has practiced at Gillette since 1998 and is the Spina Bifida Program medical director.

In his practice, he has concentrated on patients with brachial plexus injuries and lesions, congenital hydrocephalus, congenital quadriplegia (cerebral palsy), and spina bifida.



Petronio graduated from Northwestern University Medical School in Chicago and served residencies at the Hospital of the University of Pennsylvania in Philadelphia and Children's Hospital of Philadelphia. He held a clinical fellowship in neuro-oncology and a postdoctoral fellowship in neurosurgery at the University of California in San Francisco. He also was a postdoctoral research fellow in neurosurgery/human molecular biology and genetics and a clinical fellow in pediatric neurosurgery at University of Utah/Primary Children's Medical Center in Salt Lake City. Petronio has practiced at Emory University Affiliated Hospitals and Scottish Rite Children's Medical Center in Atlanta. He began practicing at Gillette in 1999 and treats patients with congenital hydrocephalus, cerebral palsy, spina bifida and craniofacial anomalies.

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Kevin J. Sheridan, M.D., is now a full-time physician at Gillette Children's Specialty Healthcare in St. Paul, Minn. He also will conduct clinics at Gillette's Lifetime Specialty Care Clinic, which provides specialized health care to adults with cerebral palsy.

An internal medicine and pediatric specialist, Sheridan has specialty certification in adult internal medicine and pediatrics and subspecialty certifications in adult and pediatric endocrinology. He has a special interest in the role of preventive health care in people of all ages who have chronic conditions.

Sheridan graduated from the University of Minnesota Medical School and completed a fellowship in endocrinology there. He has attended pediatric patients and consulted on inpatient pediatric endocrine and adult issues at Gillette since 1996, shortly after he joined St. Paul-Ramsey Medical Center.

Teresa Schultz, C.N.P. - Pediatrics, received her bachelor's degree in nursing from the College of St. Catherine's in St. Paul, Minn. She attended graduate school at the University of Minnesota, completing her degree as a pediatric nurse practitioner with concentrations in children with special health-care needs and disability policy. Schultz works with Michael Partington, M.D., and Joseph Petronio, M.D., in their neurosurgical practice.



At Gillette, her role includes assessing patients before and after surgery, monitoring patients' progress while they are in the hospital, ordering diagnostic tests, and educating patients and families. Her professional affiliations include membership in the Minnesota Board of Nursing, The National Association of Pediatric Nurse Associates and Practitioners, and the American Association of Neurological Surgeons.