CT Children's CLASP Guideline

Macrocephaly

INTRODUCTION

Macrocephaly is defined as head circumference greater than two standard deviations above the mean, or greater than the 97th percentile, for age, sex, race and gestation. Macrocephaly affects more than 2-3% of children. Some children with large head size have their measurement percentiles matched by height and weight, accompanied by normal neurological milestone progression. In other cases, the head size is accelerating out of proportion to the rest of the body and warrants neurosurgical evaluation.

The etiology of macrocephaly includes benign familial macrocephaly, benign extra-axial collections of infancy, hydrocephalus, subdural hemorrhage related to trauma, genetic conditions, or intracranial tumors or vascular malformations.

INITIAL EVALUATION AND MANAGEMENT

INITIAL EVALUATION:

- History: gestational/birth history reviewing prenatal/neonatal infection, prematurity, and intraventricular hemorrhage. Family history of macrocephaly or genetic disease should be questioned. Prior head circumference data should be reviewed, as should developmental milestones with attention to delays. History of vomiting/failure to thrive and or prior evaluation for brief resolved unexplained event (BRUE) should also be pursued.
- PE:
    - **Head/scalp exam**
        - Measurement of head circumference. For children born prior to 37 weeks, please ensure measurements are corrected for gestational age prior to plotting.
        - *Measurement performed just above the eyebrow to the back of the head just above the ears*

        - Palpation of the fontanelle and cranial sutures, assessing for fullness or diastasis (separation) of the sagittal suture
        - Appearance of scalp veins
    - **Neurologic exam**
        - Eye movements, assessing for strabismus or paralysis of upgaze ('sundowning')
        - Assessment of tone
        - Lateralizing motor exam
    - **Consider skin exam, assessing for**
        - Bruising of the torso, ears, or neck
        - Specific attention to the frenulum, angle of the jaw, cheeks, eyelids, and subconjunctiva

- Attention to any possible patterned bruising
- Notation of any bruising in a child under 4 months of age

- Evaluate for symptoms and signs of RED FLAGS:
  - Rapidly increasing head circumference after correction for gestational age
  - Bulging/tense fontanelle
  - Prominent scalp veins
  - Paralysis of upgaze
  - Failure to thrive
  - Altered mental status/focal neurologic findings/seizures

INITIAL MANAGEMENT:

- Monitoring by primary care is appropriate in most cases. Asymptomatic, developmentally normal children with reassuring physical examination rarely benefit from further diagnostic evaluation (see attached #5 recommendation in the attached ABIM Choosing Wisely document from the AAP Section on Neurological Surgery)
- Primary care providers may opt to obtain diagnostic imaging in selected patients. In alignment with the Choosing Wisely (**Appendix A- Choosing Wisely**) recommendation referenced above, our office discourages the ordering of cross-sectional imaging studies (CT or MRI) for macrocephaly by primary care providers. In the infant with an open fontanelle, a head ultrasound may be requested. We strongly recommend that such studies be performed by capable technicians and interpreted by a pediatric radiologist.
  - A common normal variant seen on head ultrasound is *benign enlargement of the subarachnoid space in infancy* (BESSI). These children often have a family history of macrocephaly and are typically developmentally normal with accelerated head circumference velocity crossing percentiles between 4-8 months of age after which the curve typically stabilizes/plateaus spontaneously. No intervention or follow-up imaging is required. The neurosurgery team is happy to provide guidance and/or consultation on these patients.
- For questions in advance of consultation, please do not hesitate to call our pediatric neurosurgeons, through our One Call Access Center at 833.733.7669.

WHEN TO REFER

DELAY REFERRAL (Watchful Waiting by PCP):

- Macrocephaly in proportion with height/weight measurements with no symptoms or evidence of developmental delay
- Macrocephaly with growth curve parallels established growth curve slope

ROUTINE REFERRAL (within 4 weeks) to Neurosurgeon:

- Macrocephaly crossing % and/or out of proportion with height/ weight measurements, with no red flag symptoms
- Macrocephaly with signs of developmental delay
- Parental anxiety or desire for referral

REFERRAL TO EMERGENCY DEPARTMENT WITH NEUROSURGERY CONSULT:

- Macrocephaly accompanied by loss of milestones, red flag symptoms or bruising concerning for abuse

HOW TO REFER

Referral to Neurosurgery Department via CT Children's One Call Access Center
Make a Referral - Connecticut Children's (connecticutchildrens.org)
Phone: 833.733.7669 **Fax:** 833.226.2329
Information to be included with the referral:

- Faxed copies of head circumference measurements

WHAT TO EXPECT	**What to expect from CT Children's Visit:** ▪ Neuro-imaging as appropriate ▪ Patients with an open fontanelle will undergo a cranial ultrasound examination. After closure, CT or MRI is completed. For patients with developmental delay, consultation with neurology and/or genetics as appropriate.

Choosing Wisely®

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American Academy of Pediatrics

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Section on Neurological Surgery

Five Things Physicians and Patients Should Question

1 Do not perform routine imaging for evaluation of infant head shape.

Routine imaging for the evaluation of infant head shape is not necessary. It exposes the child to unnecessary radiation. Positional plagiocephaly can be diagnosed on clinical examination. Most craniosynostosis presentations can also be discerned on clinical examination. Imaging may be obtained by specialists to make the diagnosis in complex cases and, if necessary, for surgical planning.

2 Do not obtain imaging of the cervical spine following trauma in an awake and alert patient without considering the use of clinical decision making (CDM) tools for cervical spine clearance.

Consideration should be given to avoid unnecessary radiation exposure when appropriate. For instance, CDM tools incorporate 3 or more variables from history, physical examination, or simple clinical tests to guide patient management. Results from the National Emergency X-Radiography Utilization Study (NEXUS) and the Pediatric Emergency Care Applied Research Network (PECARN) provide a high negative predictive value for significant cervical spine injuries in pediatric patients. Low-risk criteria from NEXUS include: no posterior midline cervical spine tenderness; no evidence of intoxication; normal level of consciousness; no focal neurological deficit; and no painful distracting injuries. PECARN developed a model that was highly sensitive for a normal cervical spine in the absence of: altered mental status, focal neurologic findings, neck pain, torticollis, substantial torso injury, conditions predisposing to cervical spine injury, high-risk motor vehicle crash, and diving. In comparison to NEXUS, the PECARN model takes into account mechanism of injury and specific extent and location of other associated injuries.

3 Do not routinely perform imaging or routine elective procedures requiring sedation or general anesthesia for very young children with low-risk asymptomatic lesions.

Low-risk asymptomatic lesions such as small rubbery scalp masses representing dermoid cysts or shallow midline sacral dimples do not routinely require intervention as a young infant. Routine magnetic resonance imaging requiring anesthesia is typically not recommended. Given the US Food and Drug Administration's Drug Safety Communication on pediatric anesthesia www.fda.gov/Drugs/DrugSafety/ucm532356.htm) warning that general anesthesia and sedation drugs used in children younger than 3 years for anesthesia of more than 3 hours or repeated use of anesthetics may affect the development of children's brains, risks and benefits of elective imaging or procedures should be carefully weighed (http://smarttots.org/). If imaging is necessary, consider approaches such as feed-and-wrap for MRI in infants or referral to specialists to develop a clinical follow-up plan and timing of intervention as appropriate.

4 Do not perform routine imaging for evaluation of VP shunt function in a patient without signs or symptoms of shunt malfunction.

Routine imaging to evaluate ventricle size in an asymptomatic patient with hydrocephalus is not necessary. When imaging is needed, performing a rapid brain MRI is a recommended option to prevent radiation exposure to the child. Surveillance imaging, if needed, should only be ordered by specialists who treat hydrocephalus.

5 Do not routinely obtain a CT or MRI scan for developmentally normal, clinically asymptomatic infants with macrocephaly.

Most infants with macrocephaly do not have abnormalities that require neuroimaging or neurosurgical evaluation. Imaging should generally be reserved for infants with clinical concerns such as abnormal neurological examination findings, significant developmental delay, or rapidly increasing head circumference measurements (such as those crossing growth curves). When imaging is indicated, head ultrasonography should typically be considered as the first-line test for infants with an open fontanelle.

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How This List Was Created

The pediatric neurosurgery Choosing Wisely topics were chosen after discussion among the Section on Neurological Surgery (SONS) Executive Committee members regarding the most common pediatric neurosurgery issues and treatments general pediatricians see in their practices. Various expert committees and sections of the AAP reviewed and approved the list. The AAP Executive Committee then granted final approval of the list.

AAP's disclosure and conflict of interest policy can be found at www.aap.org.

Sources

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About the ABIM Foundation

The mission of the ABIM Foundation is to advance medical professionalism to improve the health care system. We achieve this by collaborating with physicians and physician leaders, medical trainees, health care delivery systems, payers, policymakers, consumer organizations and patients to foster a shared understanding of professionalism and how they can adopt the tenets of professionalism in practice.

To learn more about the ABIM Foundation, visit www.abimfoundation.org.

About the American Academy of Pediatrics Section on Neurological Surgery

The American Academy of Pediatrics is an organization of 67,000 primary care pediatricians, pediatric medical subspecialists and pediatric surgical specialists dedicated to the health, safety and well-being of infants, children, adolescents, and young adults.

The Section on Neurological Surgery addresses issues common to pediatric neurosurgery to improve the care of infants, children, adolescents, and young adults with neurosurgical disorders.

For more information, visit www.aap.org.

For more information or to see other lists of Things Clinicians and Patients Should Question, visit www.choosingwisely.org.