**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 hydrocephalus, benign extra-axial collections of infancy, cortical malformations of the brain, craniosynostosis or intracranial tumors.

**INITIAL EVALUATION AND MANAGEMENT**

**INITIAL EVALUATION:**
- History, cranial and neurological exam
- Evaluate for symptoms and signs of increased intracranial pressure
- Evaluate for developmental delay

**INITIAL MANAGEMENT:**
- The choice of imaging is challenging and it is preferred that the burden of choice be shifted from the PCP to the neurosurgeon.
- If you choose to image prior to referral, consider the following:
  - **CT** leads to radiation exposure.
  - **Ultrasound** shows hydrocephalus well but images small extra-axial collections and the posterior fossa poorly.
  - **MRI** provides the best imaging, but is also the most costly and requires sedation in some children.
- For questions in advance of consultation, please do not hesitate to call our pediatric neurosurgeons, Drs. Kanev or Martin, 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 signs of increased intracranial pressure or developmental delay

**ROUTINE REFERRAL (within 4 weeks) to Neurosurgeon:**
- Macrocephaly out of proportion with height/weight measurements, with no signs of increased intracranial pressure
- Macrocephaly with signs of developmental delay
- Parental anxiety or desire for referral

**REFERRAL TO EMERGENCY DEPARTMENT WITH NEUROSURGERY CONSULT:**
- Macrocephaly accompanied by any vomiting, lethargy or focal neurological signs or symptoms

**HOW TO REFER**

Referral to Neurosurgery Department via CT Children’s One Call Access Center
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.