

Macrocephaly

FAST FACTS

2–5%

of children will have macrocephaly

3 years old

is usually the age that head circumference stabilizes among children who have an otherwise normal neurological examination

Macrocephaly is defined as a head (occipitofrontal) circumference that is more than two standard deviations above average, compared to other children of the same age. OFC (Occipital Frontal Circumference) should be measured above the eyebrows and around the widest point of the occiput at all well child visits up to 24 months.

Although macrocephaly is commonly encountered in the pediatric clinical practice, it's important to distinguish harmless causes (such as benign familial macrocephaly) from more serious causes. For example, patients with macrocephaly caused by an enlarged brain (megalencephaly) may have developmental delays, seizures and other neurological problems.

ASSESSMENT

Gather any relevant family history and use special growth charts when indicated. Evaluate serial measurements of head circumference and assess the anterior fontanelle, cranial sutures and extraocular movements. Also look for prominent scalp veins.

HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS

- Rapidly increasing head circumference (averaging over 2 cm/month under one year of age or crossing two major percentile lines)
- Bulging/tense fontanelle
- Prominent scalp veins
- Paralysis of upgaze
- Failure to thrive (may be related to frequent vomiting or poor oral intake)
- Unexplained lethargy or irritability
- Developmental delays (not hitting milestones, or regression of previously met milestones)

MANAGEMENT/TREATMENT

- Measure parental head circumference to help confirm familial macrocephaly.
- Order head ultrasound if there is an open fontanelle and concern for developmental delays.

WHEN TO REFER

- Refer directly to Cincinnati Children's Emergency Department for a STAT head ultrasound or CT if the patient has multiple HPE red flags or signs of high intracranial pressure. Also place an immediate referral to the Division of Pediatric Neurosurgery.
- Refer to the Division of Pediatric Neurosurgery if the patient's head ultrasound findings are abnormal (urgency may be based on findings).

If you have clinical questions or want to refer a patient, call the Physician Priority Link® at 513-636-7997.

If you would like additional copies of this tool, or would like more information, please contact the Physician Outreach and Engagement team at Cincinnati Children's.

Macrocephaly

Child presents with macrocephaly
(Occipitofrontal circumference is >2 standard deviations above the mean)

Standard Workup

- Gather relevant family history
- Use special growth charts when indicated
- Evaluate serial measurements of head circumference
- Look for prominent scalp veins
- Assess anterior fontanelle, cranial sutures and extraocular movements

HPE (HISTORY AND PHYSICAL EXAM) RED FLAGS

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Yes **Any red flags with signs of high intracranial pressure?** No

Send to ED for STAT head ultrasound* or head CT and refer immediately to pediatric neurosurgery.

Does the patient have abnormal neurological development or delays?

No

Yes

Is there a family history of macrocephaly?

Order head ultrasound.*
If abnormal, consider neurosurgical referral (urgency may be based on findings).

No

Yes

Order head ultrasound.*

No immediate intervention required.

*Only applies to patients with open fontanelle