A Retrospective Analysis of the Utility of Head Computed Tomography and/or Magnetic Resonance Imaging in the Management of Benign Macrocrania

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Objective To assess whether computed tomography (CT), magnetic resonance imaging (MRI), and neurosurgical evaluations altered the diagnosis or management of children diagnosed with benign macrocrania of infancy by ultrasonography (US).

Study design We queried our radiology database to identify patients diagnosed with benign macrocrania of infancy by US between 2006 and 2013. Medical records of those with follow-up CT/MRI were reviewed to determine clinical/neurologic status and whether or not CT/MRI imaging resulted in diagnosis of communicating hydrocephalus or required neurosurgical intervention.

Results Patients with benign macrocrania of infancy (n = 466) were identified (mean age at diagnosis: 6.5 months). Eighty-four patients (18.0%) received subsequent head CT/MRI; of these, 10 patients had neurologic abnormalities before 2 years of age, of which 3 had significant findings on MRI (temporal lobe white matter changes, dysmorphic ventricles, thinned corpus callosum). One patient without neurologic abnormalities had nonspecific white matter signal abnormality (stable over 6 months) but no change in management. None required neurosurgical intervention. Another 9/84 patients had incidental findings including Chiari I (3), small subdural bleeds (2), arachnoid cyst (1), small cavernous malformation (1), frontal bone dermoid (1), and a linear parietal bone fracture after a fall (1).

Conclusions Children diagnosed with benign macrocrania of infancy on US without focal neurologic findings do not require subsequent brain CT/MRI or neurosurgical evaluation. Decreasing unnecessary imaging would decrease costs, minimize radiation and sedation exposures, and increase clinic availability of neurology and neurosurgery specialists. *(J Pediatr 2017;182:283-9).*

Macrocrania in infancy, observed as rapidly enlarging head circumference (HC) or a HC >95th percentile, can be due to both benign and pathologic causes. Published literature would suggest a lower percentage of cases are pathologic. In 1 study of 255 children with macrocrania, 6% had significant abnormalities on ultrasonography (US) including hydrocephalus or congenital malformations.[1] Alternatively, benign macrocrania of infancy is a more common and less concerning cause of macrocrania described as enlarging extra-axial fluid spaces leading to an expansion of HC around 4-6 months of age, with HC measurements crossing percentile lines and often reaching above the 95th percentile.[1-5] Typical radiologic findings can be seen in **Figure 1**.[5-7] Although macrocrania may persist into adulthood, the enlarged extra-axial cerebrospinal fluid spaces often resolve by 2-3 years of age.[8-11] Other terms have been used for this condition including benign external hydrocephalus or benign familial macrocrania. Even though the incidence of benign macrocrania of infancy in the general population is not clear, studies have consistently shown it is more prevalent in male patients compared with female patients (~2:1) and is often familial.[12] The etiology of benign macrocrania of infancy is not clear, but a common hypothesis is that delayed maturation of arachnoid villi leads to decreased absorption of cerebrospinal fluid and expansion of extra-axial spaces.[13] Because of the compliant skull in young infants, the result is enlarged HC without increased intracranial pressure.[14]

Benign macrocrania of infancy can be accompanied by mild developmental delays (speech, gross, or fine motor delays), which often, but not always, resolve.[8,11,12,15] Because macrocrania in infants can result from other more serious conditions (including communicating hydrocephalus, mass lesions, or vascular malformations resulting in obstructive hydrocephalus), all infants whose HC rapidly crosses percentile lines should undergo neuroimaging.[16] US is an appropriate screening tool in this age group and is sufficient for diagnosing benign macrocrania of infancy.[1,17]

CT	Computed tomography
CCHMC	Cincinnati Children's Hospital Medical Center
GA	Gestational age
HC	Head circumference
MRI	Magnetic resonance imaging
US	Ultrasonography
WHO	World Health Organization

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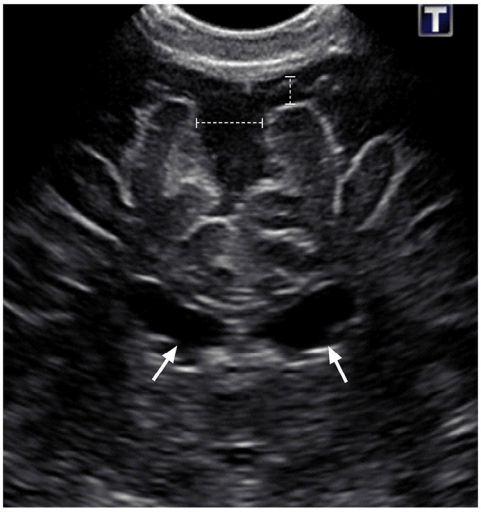

Figure 1. Example image of benign macrocrania on a head US. Coronal image through the brain demonstrates symmetric enlargement of extra-axial spaces (*dotted lines*) and mildly prominent frontal horns of the lateral ventricles (*arrowheads*).

In a significant fraction of cases, despite US findings suggesting benign macrocrania of infancy and normal neurologic examination results, patients are referred for neurosurgical evaluation and additional neuroimaging by computed tomography (CT) or magnetic resonance imaging (MRI). This referral pattern may result from misguided perceptions that US misses important findings that would be evident on CT/MRI, that the degree of acceleration in head growth requires specialty evaluation, or that macrocrania with developmental delay defies the definition of "benign" macrocrania and mandates further evaluation. Does referral for additional neuroimaging and specialty evaluation result in a diagnosis of communicating hydrocephalus or other concerning diagnosis or result in neurosurgical intervention? To answer this question, this study examines a cohort of 466 patients from 2006 to 2013 at Cincinnati Children's Hospital Medical Center (CCHMC) with or without CT/MRI following an US diagnosis of benign macrocrania of infancy.

Methods

With institutional review board approval, we queried our radiology database (Softek Illuminate Insight, Prairie Village, Kansas) to identify patients diagnosed with benign macrocrania of infancy by head US from January 2006 through June 2013. Patients were separated into 2 groups: those with follow-up CT or MRI and those without. Patients were excluded from either group if (1) indication for the head US included something other than macrocephaly or a synonymous term; (2) patient had previous head or spine imaging for an indication other than macrocrania (however, patients with a previous normal head US performed under a neonatal intensive care unit screening protocol were not excluded); (3) descriptive US findings were not consistent with the final diagnosis of benign macrocrania of infancy assigned to the patient; (4) on chart review, a history of confounding systemic or genetic illnesses, intracranial trauma, child abuse, or congenital anomalies were identified; (5) the indication for the CT or MRI was specifically to follow-up incidental findings on the US ; and (6) the age at time of first CT or MRI was >24 months.

Using Epic software (Verona, Wisconsin), demographic and health data were collected including zip code, date of birth, gestational age (GA), imaging reports, developmental assessments, neurologic assessments, referrals to relevant specialty services, HC measurements, and medical management including medications, surgeries, or other therapies, etc. Personal health information was collected, stored, and protected according to the Health Insurance Portability and Accountability Act of 1996 regulations and policies and in accordance with institutional review board safeguards.

Definitions of Collected Health Information

Developmental delay was defined as social, gross motor, fine motor, or speech delay reported during annual well-child visits, therapy evaluations, neurology, neurosurgery, genetics, or developmental or behavioral clinics. Neurologic deficits were focal/regional motor or sensory deficits, seizures, or abnormal movements/posturing not attributable to non-neurologic causes. Hypotonia was reported separately and not considered a focal neurologic deficit. GA was rounded down to the nearest full week gestation. A patient was considered premature if their GA was <37 weeks as defined by the World Health Organization (WHO).[18]

HC Measurements and Growth Curves

HC measurements were collected if the GA was known. For any premature infant, the HC was plotted on the Fenton Preterm Growth Chart[19] until they reached 40 weeks gestation. After 40 weeks GA, a corrected age was calculated to plot the HC on the WHO growth chart for 0-2 years of age.[20] The corrected age in weeks is calculated as the (number of weeks from birth) – (number of weeks premature). HC measurements from 0 months up to but not including 1 month were averaged and plotted at 0.5 months on the WHO growth chart with its accompanying 95% CI. The same method was used for each 1-month bin up to 24 months of age. The percentile lines (50th, 75th, 90th, and 95th) from the WHO and Fenton preterm growth charts were added to our HC figures for reference.[21] Precalculated L (estimates of the power of the box-cox transformation), M (median), and S (generalized coefficient of variation) values published by the WHO were used to calculate z scores from each HC measurement between 0 and 24 months of age following previously published methods.[21-24]

Imaging Collection and Review of Head Ultrasound, Head CT, and Brain MRI

Study inclusion criteria dictate that all patients in the study have an initial diagnosis of benign macrocrania of infancy by US; therefore, the head US reports were reviewed for confirmatory terms such as enlarged extra-axial spaces, prominent ventricles, or both. If these terms were absent, the patient was excluded