10-MINUTE CONSULTATION

Baby with an abnormal head

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Cite this as: *BMJ* 2014;348:f7609
doi: 10.1136/bmj.f7609

This is part of a series of occasional articles on common problems in primary care. The *BMJ* welcomes contributions from GPs.

A young couple present to their general practitioner worried that their 4 month old baby's head "doesn't quite look right."

What you should cover

What specifically is worrying the parents?—Parents sometimes get concerned that their baby's head looks smaller or bigger than it should be, is flattened on one side, or is bumpy in places; they may also be worried about the presence or size of the "soft spot" (fontanelle).

Is it posture related?—Many babies have a postural asymmetry of the head (plagiocephaly). The main risk factor is the baby's preferred resting head position, both in utero and in the first few months after birth, while the skull bones are still malleable.

Are other family members affected?—Microcephaly (head circumference <3rd centile) and macrocephaly (head circumference >97th centile) can run in families and are generally without consequence. Spontaneous microcephaly is often indicative of a serious underlying problem such as a neurodevelopmental disorder or craniosynostosis (premature fusion of sutures). Although spontaneous macrocephaly is seldom of clinical relevance, identifying secondary causes is important as this may be due to increased intracranial volume, such as in hydrocephalus or a subdural haematoma as a result of birth trauma, or more rarely congenital tumours. A wide fontanelle that is bulging or other sutures widely open can indicate clinically important macrocephaly, as would any evidence of neurodevelopmental issues such as altered tone or convulsions.

Do the parents have any concerns about the baby's development?—If so, this may indicate a serious underlying cerebral problem.

Are parents consanguineous?—Metabolic disorders and syndromic disorders occur more commonly in children from first cousin marriages, and these may lead to a primary failure of brain growth and resulting microcephaly.

What is the maternal history?—Intrauterine infections (such as toxoplasmosis), excessive maternal alcohol consumption, and use of anticonvulsant drugs during pregnancy can lead to microcephaly. In such cases, the inadequate skull growth is usually secondary to an underlying failure of brain development.

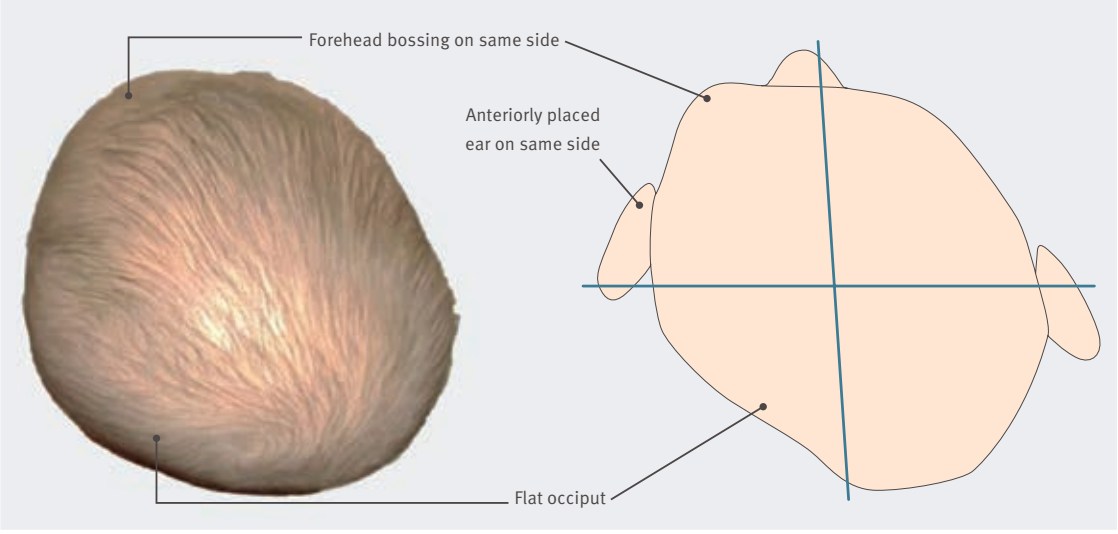
What was the gestational age at birth?—This can affect the size and shape of the head, as a premature baby often has an "elongated head" with bilateral flattening on either side.

Were there any problems at birth?—A difficult or instrumented delivery (application of forceps or ventouse) may deform the face and skull, and can result in a subgaleal haemorrhage (extensive swelling due to bleeding beneath the muscle layer not restricted by sutures) or cephalhaematoma (subperiosteal bleeding limited by suture lines).

What you should do

Look at the parents' head size and shape to see if there is any obvious familial microcephaly or macrocephaly. If so, and provided there are no other concerns, reassure the parents.

Measure and plot the baby's head circumference to see if the head is of a normal size for (gestational) age and sex. A period of serial monitoring may be required. Measurement should be done with a non-expanding tape measure



Positional plagiocephaly: the occiput is flattened on the left and the ipsilateral ear and forehead are pushed forward with a "parallelogram" look when viewed from above

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Previous articles in this series

- A feeling of a lump in the throat (*BMJ* 2014;348:f7195)
- Tremor (*BMJ* 2013;347:f7200)
- Dental pain (*BMJ* 2013;347:f6539)
- Flashes, floaters, and a field defect (*BMJ* 2013;347:f6496)
- An adult with a neck lump (*BMJ* 2013;347:f5473)

across the forehead and over the most prominent part of the parietal and occipital bones. Take the largest of three measurements. Plot the occipito-frontal circumference on an age-appropriate growth chart, and compare the head size with other growth parameters (that is, head size relative to body length and weight).

Carefully examine the head paying particular attention to:

- *Fontanelles*—The posterior fontanelle normally closes first, at around 8-12 weeks, and then the anterior fontanelle closes, at 12-21 months.[1]
- *Sutures*—Feel with the tip of your finger for any ridging over the sutures. A smooth ridge or elevation (rather than the normal corrugated flat surface) suggests premature fusion of the suture. The sagittal suture is most commonly affected, resulting in a "boat-shaped head."
- *Shape*—Is there evidence of plagiocephaly? Positional or non-synostotic (that is, where none of the sutures is fused) plagiocephaly is the commonest cause of an unusual shaped head at this age, and this will usually resolve spontaneously by the age of 2 years.[2] Examination will reveal a flattened occiput, with the ipsilateral ear and forehead being pushed forward to produce a "parallelogram" look when observed from above (see figure).

Examine the spine for scoliosis and truncal asymmetry and look for torticollis or a sternomastoid "tumour" (tear), as muscle shortening with limited neck movement can sometimes result in positional plagiocephaly; this can be managed with the help of a paediatric physiotherapist. Remember, however, that torticollis may also be due to cervical spine skeletal anomalies, and if this is suspected a specialist paediatric neurosurgical assessment should be sought.

Examine for dysmorphism, congenital abnormalities, and abnormal neurology, such as hypotonia or hypertonia. Down's syndrome, for example, is associated with microcephaly and/or brachycephaly (a flattened head). If present, refer for a paediatric assessment.

Refer urgently to a paediatric neurosurgeon if there is microcephaly with premature closure of fontanelle(s), or sutures are prematurely closed (synostotic). Any evidence of raised intracranial pressure—such as prominent skull veins, a bulging fontanelle, or a rapidly increasing head size—also warrants urgent referral.

Management of positional plagiocephaly

- Explain that this has become more common after the success of campaigns to encourage parents to lay babies on their backs in order to reduce the risk of sudden infant death syndrome (SIDS).[3]
- Counsel parents that the vast majority of children with mild to moderate positional plagiocephaly do not need intervention. Reassure them that the head shape will naturally improve at 3-5 years of age as the child assumes a more upright posture.
- Be prepared to discuss the pros and cons of ongoing research: although it has generally been accepted that positional plagiocephaly does not result in developmental delay,[4] there is some evidence of an impact on the development of language and cognition skills.[5-8] In severe cases, some experts have suggested orthotic helmets may be used, but evidence of benefit is weak and disputed.[9 10]
- Encourage lying the baby prone when awake and instruct on varying sleep positions, in line with SIDS guidelines.[3]
- Suggest supervised "supported" sitting or lying on the side when awake, propped up with a cushion behind the back. Turning the cot position will encourage the infant to turn its head (to look at toys, for example) when lying supine, which will encourage lying on the non-flattened side.
- Parents may not be willing to accept the conservative management suggested above and may visit internet sites for expensive (up to £2500) custom made helmets. These have to be worn for up to 23 hours a day and may result in discomfort or pain from a poor fit or complications such as fungal infections of the scalp. They are therefore generally not recommended.[11]

USEFUL READING

For parents

NHS Choices. Craniosynostosis. 2012. www.nhs.uk/conditions/craniosynostosis/Pages/Introduction.aspx

NHS Choices. Plagiocephaly and brachycephaly (flat head syndrome). 2012. www.nhs.uk/conditions/plagiocephaly/Pages/Introduction.aspx

Pruitt SJ. *The truth about tummy time. A parent's guide to SIDS, the back to sleep program, car seats and more*. Authorhouse, 2011

For professionals

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Contributors: ARG and AS conceived the idea for this paper. GAS took the photo and drew the illustration. All authors were involved in drafting the manuscript.

Competing interests: We have read and understood the BMJ Group policy on declaration of interests and have no relevant interests to declare.

Provenance and peer review: Not commissioned; externally peer reviewed.

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Accepted: 27 August 2013

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UNCERTAINTIES PAGE

Should children who have a cardiac arrest be treated with therapeutic hypothermia?

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Cite this as: *BMJ* 2014;348:f7672
doi: 10.1136/bmj.f7672

This is one of a series of occasional articles that highlight areas of practice where management lacks convincing supporting evidence. The series adviser is David Tovey, editor in chief, the *Cochrane Library*. To suggest a topic, please email us at practice@bmj.com.

The International Liaison Committee for Resuscitation recommends that comatose adult patients with spontaneous circulation after cardiac arrest are cooled to 32-34°C for 12-24 hours based on analysis of data from two randomised controlled trials and 17 observational studies.[1] However, these studies were mostly in a specific subgroup of cardiac arrest patients with witnessed, out-of-hospital ventricular fibrillation, and evidence of benefit in the general population of cardiac arrest patients has been less certain.[2] The rationale for therapeutic hypothermia is that it can reduce cerebral metabolism, attenuate biosynthesis of excitotoxic compounds, reduce free radical production, reduce inflammation, and regulate gene and protein expressions associated with necrotic and apoptotic pathways during ischaemia and reperfusion.[3]

Recommendations for treatment in children[4] (box 1) are based almost solely on adult data. However, the aetiology of cardiac arrest is very different in children,[5] possibly altering the pattern of neuronal injury. Most cardiac arrests are secondary to a respiratory cause with profound hypoxia, and primary cardiac causes of arrests, including ventricular fibrillation, are rare. In other clinical situations, therapeutic hypothermia has been seen to be both beneficial (newborns with hypoxic brain injury within 6 hours of birth)[6] and potentially harmful (traumatic brain injury).[7] It is therefore important that the question of whether children with cardiac arrest should be treated with therapeutic hypothermia is addressed.

What is the evidence of uncertainty?

Our recent Cochrane systematic review[8] searched, to December 2011, the Cochrane Anaesthesia Review Group Specialized Register, Cochrane Central Register of Controlled Trials (CENTRAL), Medline, Embase, CINAHL, BIOSIS, and Web of Science databases for randomised controlled trials comparing therapeutic hypothermia with normothermia or standard care after paediatric cardiac arrest. We also contacted international experts in therapeutic hypothermia and paediatric critical care to locate further published and unpublished studies.

We found no relevant randomised controlled trials, but identified three paediatric cohort studies: two retrospective studies[9 10] and one prospective study in abstract form only.[11] All three studies showed no difference in mortality or proportion of survivors with good neurological outcome for those treated with therapeutic hypothermia compared with standard care. Imbalance between the compared populations within studies was evident, as was heterogeneity in the cause of cardiac arrest. Patients receiving therapeutic hypothermia were sicker with longer duration of cardiopulmonary arrest, more pharmacological interventions during resuscitation, higher post-resuscitation serum lactate levels, higher multiorgan dysfunction score, and requirement for renal replacement therapy.

Prospective observational studies of children undergoing therapeutic hypothermia provide evidence that the treatment is safe and feasible, but not of its effectiveness.[12 13] Use of therapeutic hypothermia has also been reported in retrospective observational studies, but the incidence was too low to report effectiveness.[14-16]

Surveys of clinical practice in the United Kingdom and the United States show that therapeutic hypothermia is used by some paediatric intensive care and emergency medicine units after both out-of-hospital and in-hospital cardiac arrest, but there was substantial variation in practice regarding patient selection, temperature, and duration of therapy.[17 18]

Is ongoing research likely to provide relevant evidence?

Our recommendations for the framing of future research to answer this clinical uncertainty are outlined in box 2. Our search of the trials registry databases (detailed above) identified four ongoing randomised controlled trials.

The two Therapeutic Hypothermia After Pediatric Cardiac Arrest (THAPCA) trials (350 and 500 participants) will determine whether, among children aged <18 years with in-hospital cardiac arrest (NCT00880087) or out-of-hospital cardiac arrest (NCT00878644), treatment with a protocol involving hypothermia (32-34°C) for 48 hours and three days of normothermia (36-37.5°C), compared with five days of normothermia, affects neurological outcome at 12 months.

The Hypothermia for Cardiac Arrest in Paediatrics (NCT00754481) phase II (pilot) trial (n=40) will determine if children aged <18 years with in-hospital or out-of-hospital cardiac arrest treated with 48 hours of hypothermia (33-34°C) compared with normothermia (36.5-37.5°C) have different neurological outcomes at 12 months.

The use of a strict normothermia group in these three trials aims to eliminate a confounding effect of hyperthermia in the control group, as was noted in about 25% of the European trials of hypothermia after cardiac arrest in adults.[19] Hyperthermia (≥38°C) is harmful and associated with unfavourable neurological outcome after hypoxic brain injury.[20]

Box 1 | Recommendations for use of therapeutic hypothermia after paediatric cardiac arrest (International Consensus on Cardiopulmonary Resuscitation and Emergency Cardiovascular Care Science with Treatment Recommendations[4])

- Therapeutic hypothermia (32-34°C) may be beneficial for adolescents who remain comatose after resuscitation from sudden, witnessed, out-of-hospital, ventricular fibrillation cardiac arrest
- Therapeutic hypothermia (32-34°C) may be considered for infants and children who remain comatose after resuscitation from cardiac arrest

Box 2 | Recommendations for future research

The key components of the research required to reduce our clinical uncertainty in the use of therapeutic hypothermia after paediatric cardiac arrest

Population

- Infants and children experiencing an in-hospital or out-of-hospital cardiac arrest at risk of neurological injury.
- Inclusion of sufficient patients to allow subgroup analysis of the impact on outcome of:
  - Cause of cardiac arrest—asphyxia induced or of cardiac origin
  - Presenting cardiac rhythm at time of cardiac arrest—shockable (ventricular fibrillation or ventricular tachycardia) or non-shockable (asystole and pulseless electrical activity).

Intervention

- Therapeutic hypothermia administered within a defined dose:
  - Time to initiation of therapeutic hypothermia
  - Time to reach target temperature
  - Specify method used for cooling
  - Duration of at least 24 hours, up to 72 hours
  - Temperature depth in the range 32-34°C
  - Controlled rewarming rate no faster than 0.25°C per hour.

Comparison

- Strict normothermia (36-37°C), actively avoiding hyperthermia (≥38°C).

Outcome

- Long term survival, neurocognition, and developmental ability.
- Safety of intervention (rate of infections, bleeding, arrhythmias, metabolic derangement).
- Potentially using early biomarkers if validated with longer term clinical outcomes.

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- Should women with HIV, or at high risk of contracting HIV, use progestogen-containing contraception? (*BMJ* 2013;347:f6695)
- How should we manage fear of falling in older adults living in the community? (*BMJ* 2013;346:f2933)
- Should inpatient hyperglycaemia be treated? (*BMJ* 2013;346:f134)
- Does routine oxygen supplementation in patients with acute stroke improve outcome? (*BMJ* 2012;345:e6976)
- Does gluten sensitivity in the absence of coeliac disease exist? (*BMJ* 2012;345:e7907)

The fourth ongoing study is the Duration of Hypothermia for Neuroprotection After Pediatric Cardiac Arrest (NCT00797680) phase II (pilot) trial (n=40) determining if children aged <18 years after in-hospital or out-of-hospital cardiac arrest treated with 72 hours of hypothermia (32-34°C) compared with 24 hours of hypothermia (32-34°C) develop less brain injury (assessed by plasma biomarkers and magnetic resonance spectroscopy).

Clinical trials in paediatric critical care are challenging because of the low incidence of critical illness in children. As a result, paediatric trials often adopt wide inclusion criteria. The heterogeneity of patients, particularly among those with out-of-hospital cardiac arrest, may result in mixed inclusion of hypoxia induced cardiac arrests (such as asthma) and cardiac causes (such as primary arrhythmias), potentially masking a treatment effect in a particular subgroup. The current studies will not provide definitive evidence for the optimal duration of therapeutic hypothermia (24, 48, 72 hours, or longer), rewarming rates after hypothermia, timing of the start of treatment, or method of temperature manipulation. As two of the four trials are small pilot studies, these also will not be able to provide evidence of effectiveness.

What should we do in the light of the uncertainty?

Managing a comatose child or infant after return of spontaneous circulation after cardiac arrest is a rare event outside specialist centres. Early consultation with a paediatric intensive care specialist is strongly recommended in all cases. In the absence of definitive evidence it is reasonable to initiate therapeutic hypothermia for comatose adolescent survivors from a ventricular fibrillation or pulseless ventricular tachycardia cardiopulmonary arrest, based on the evidence from adult clinical trials (box 1).[4] Therapeutic hypothermia may also be considered for infants and children who remain comatose after resuscitation from cardiac arrest. In all cases avoid hyperthermia (≥38°C) and when possible consider enrolment in ongoing randomised controlled trials or registration in existing national databases such as the Paediatric Intensive Care Audit Network (www.picanet.org.uk) and UK National Cardiac Arrest Audit (www.icnarc.org).

Contributors: BRS conceived of and drafted the article. The other authors contributed to writing the article and checking content.

Competing interests: BRS is currently working as a co-investigator of the THACPA-IH (Therapeutic Hypothermia After Pediatric Cardiac Arrest - In-Hospital) trial to potentially enable UK patient participation.

Provenance and peer review: Not commissioned; externally peer reviewed.

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Accepted: 31 October 2013