Abnormal head shape in infants

"His head is all squished on one side. Can you prescribe one of those helmets I've seen or should I take him to the cranial osteopath?" One of the downsides of the great success of the 'Back to Sleep' campaign to reduce cot deaths is that there has been an increased incidence in positional skull deformities.

Parents worry if their baby has an asymmetrical head. Most commonly, this will be a normal phenomenon related to babies preferred positioning (positional plagiocephaly) or an inherited tendency towards a small or large head. However, rarely, it may be an indication of a more serious underlying problem. Two articles, a BMJ 10-minute Consultation and a more recent BMJ Practice Pointer, have considered how we should approach this problem in primary care (BMJ 2014;348:f7609, BMJ 2023;381:e073906).

This article was updated in September 2023.

History

Establish exactly what concerns the parents have. Is it just a cosmetic concern, or are there other issues relating to development and growth (BMJ 2023;381:e073906)?

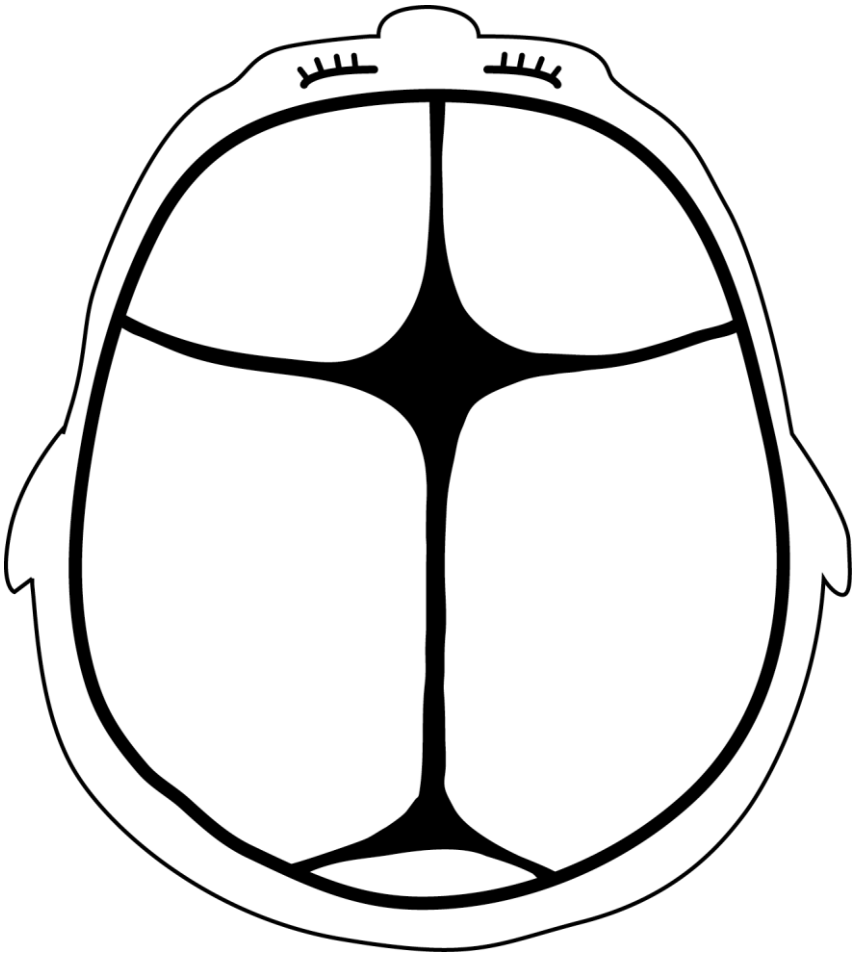
- Ask when the head shape became a concern.
- Has head shape changed? Craniosynostosis will be present from birth.
- Ask about baby's sleep position and posture. Is there a favourite side? Do the eyes close fully during sleep?
- Ask about family history of microcephaly (head circumference <3rd centile), macrocephaly (head circumference >97th centile) and neurodevelopmental conditions.
- Take a developmental history and ask about other concerning features, e.g. seizures, developmental delay, irritability, sleep disturbance, breathing or feeding problems (choking or vomiting).
- Take pregnancy and birth history. Prematurity, instrumental deliveries, etc. can all have an impact.

Examination

Here, we are looking for features which may suggest raised intracranial pressure, premature fusion of skull bones (synostosis) or developmental abnormalities – if these are found, refer. Advice on how to do this is below (BMJ 2023;381:e073906).

- Measure and plot the baby's head circumference. Compare this with previous measurements, with weight and length, and consider taking serial measurements over a number of months. Change in centiles is as important as absolute centile. *Do you routinely record head circumference at the 6w check?*
- If possible, compare head size with parental head size!
- Examine the head, looking from the top, side and front:
    - Check the fontanelles – posterior closes 8–12w, anterior closes 12–24m. Early or late closure or bulging may indicate an underlying problem.
    - Check the sutures – a ridge or elevation may indicate early fusion.
    - Check the shape – use the table below to help identify the cause of the change in head shape compared with the normal head shape shown in the first image.
- Check the spine and neck looking for scoliosis, truncal asymmetry and torticollis.
- Look for dysmorphic features and abnormal tone which may suggest underlying disorders, e.g. Down syndrome.

In the majority of cases, no concerning features will be identified and a diagnosis of positional plagiocephaly can be made.



Craniosynostosis

Craniosynostosis occurs in approximately 1/1695 live births, and was summarised in a BMJ Practice Pointer (BMJ 2023;381:e073906).

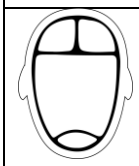
Fusion can occur in a single suture line (78% of cases) or multiple suture lines (22% of cases). Usually, fusion of the cranial sutures will have occurred before birth, but the changes are not easily visible on antenatal ultrasound scan.

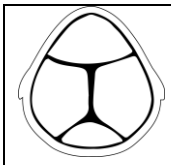
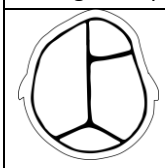
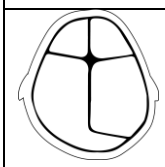
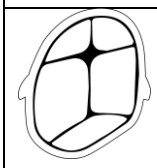
In around 1/3 cases, the craniosynostosis will be part of a wider clinical syndrome, meaning that the majority of cases seen in primary care will be single suture line, non-syndrome related.

The complications of craniosynostosis originate from raised intracranial pressure and can include headaches, irritability, sleep disturbances, seizures and damaged vision. Parents are often concerned about developmental delay; however, craniosynostosis in the absence of raised intracranial pressure is not believed to affect development.

Subtypes of craniosynostosis

Depending on which of the cranial sutures has fused, the appearance of the baby will differ (BMJ 2023;381:e073906). The commonest presentation is sagittal craniosynostosis, which affects 1/2000–1/5000 children and is more common in boys.

Head shape	Craniosynostosis type	Features
 Boat-shape head	Sagittal	• Frontal bossing. • Biparietal narrowing. • Sagittal ridge. • Large head circumference. • Triangular anterior fontanelle.

 Triangle-shape head	Metopic	• Narrow forehead with vertical prominent ridge in middle of forehead. • Bitemporal flattening. • Biparietal widening. • Triangular anterior fontanelle.
 Oblique at front, trapezium-shape head	Unicoronal	• Flattened forehead and brow ipsilaterally. • One wider eye. • Fullness to contralateral forehead. • Nose/chin deviating away from side of affected forehead. • Shortened ear-to-orbit distance on affected side.
 Oblique at back, trapezium-shape head	Unilamboid	• Flattened posterior skull unilaterally. • Bulging unilateral mastoid. • Ear displaced posteriorly or inferiorly. • Windswept appearance of the posterior skull.
 Parallelogram-shape head	Positional plagiocephaly (no craniosynostosis)	• Flattening to posterior skull unilaterally. • Ipsilateral frontal fullness. • Ipsilateral ear anteriorly displaced. • No mastoid bulging.

Management

Management is highly specialised and will take place at one of five UK tertiary centres (BMJ 2023;381:e073906). Primary care can and should refer directly to avoid delay in treatment. If uncertain about the need for referral, you can contact your local unit and share photographs for clinical advice.

- Alder Hey Children's Hospital Liverpool – craniofacial service.
- Birmingham Women and Children's Hospital – craniofacial unit.
- Oxford University Hospitals - craniofacial unit.
- Great Ormond Street – craniofacial unit.
- NHS Scotland – craniofacial unit.

Imaging is not required before referral. X-rays are unhelpful and the gold standard imaging is CT, which will be requested in secondary care.

Surgery is likely to be offered to correct head shape and reduce risk of raised intracranial pressure. The risk of surgically remodelling the cranial bones rises with age so earlier referral (ideally under 6 months) is preferable.

Positional plagiocephaly – advice for parents

Management of positional plagiocephaly was outlined in a BMJ 10-minute Consultation article (BMJ 2014;348:f7609):

- This is common and usually self corrects by 3–5y.
- It does not affect brain development.
- Encourage periods of supervised tummy time and supported sitting, and vary head position when sleeping and in cot using toys, moving the cot, etc.
- There is no evidence that helmet therapy is superior to conservative management. To pursue it privately can be costly (up to £2500!), and helmets have to be worn 23h a day and may cause discomfort and rashes.

Helmet therapy for positional skull deformities

This timely BMJ pragmatic single-blinded RCT looked at how effective helmet therapy was in improving skull shape (BMJ 2014;348:g2741). It enrolled 84 infants aged 5–6m with moderate to severe positional skull deformities and no underlying condition, and randomised them to 6m helmet therapy compared with expectant management (allowing the skull deformity to correct itself over time). It followed them to 24m of age.

- There was no difference in changes in extent of skull deformity in the two groups over the study period.
- In both groups, full recovery to a 'normal' skull shape was achieved in about 25% by 24m.
- Helmet therapy did not influence motor development, sleep or crying times.
- All parents in the helmet group reported one or more side-effect, e.g. skin irritation, pain associated with helmet.

This study suggests that helmet therapy should not form part of standard treatment for positional skull deformities as it is no more effective than allowing nature to take effect, is more expensive and has more side-effects. However, the authors urge that it is repeated in a larger sample size because this study was relatively underpowered.

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- Take a history, measure and plot head circumference, and examine fontanelles and sutures.
- Refer babies with micro- or macrocephaly, signs of raised intracranial pressure or premature fusion of the fontanelles or sutures (felt as ridges), or if there are associated development concerns, etc.
- Babies with positional plagiocephaly and torticollis should be referred to paediatric physiotherapy.
- Helmet therapy is no more effective than nature in treating positional skull deformities. It is more expensive and has more side-effects, and should not be used routinely.

Do you record head circumference at 6w? Could you add this to your template?

Useful resources for patients:

Websites (all resources are hyperlinked for ease of use in Red Whale Knowledge)

- GOSH - information leaflet on positional plagiocephaly for patients
- Headlines Craniofacial Support - charity
- Alder Hey Children's Hospital Liverpool – craniofacial service
- Birmingham Women and Children's Hospital – craniofacial unit
- Oxford University Hospitals - craniofacial unit
- Great Ormond Street – craniofacial unit
- NHS Scotland – craniofacial unit

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