Flat back of head: positional or pathological?

This article reviews the aetiology and natural history of deformational (posterior positional) plagiocephaly, comparing it with the head shape seen in pathological lambdoid synostosis. The significance and treatment of both conditions are explained, with emphasis on the fact that deformational plagiocephaly is entirely avoidable.

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Keywords

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Key points

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1. Deformational plagiocephaly is not usually present at birth; it is acquired but then improves with time.
2. Lambdoid synostosis is present at birth and worsens over time.

Deformational plagiocephaly is an entirely avoidable phenomenon. It is an alteration in head shape due to application of an external deformational force on a flexible object, ie the newborn baby's head. The force is most commonly the cot mattress; babies who sleep well in the first few months of life are more vulnerable than poor sleepers who are likely to be picked up more often. Once the child can sit unaided, pressure is reduced on the back of the head and the condition will gradually improve. While the child is small, there is little hair and the head is largely looked down upon highlighting the flat back of the head. While it is harmless, the appearance tends to cause concern to family and friends. The 'Back to Sleep' campaign initiated in 1992, while having a beneficial effect with regard to sudden infant death, exacerbated the problem. Additional causes include gestational intrauterine constraints, torticollis (wry neck), hemivertebrae and neurodevelopmental delay with poor head control.

Any clinical anxiety is triggered by concern that flattening may be the result of unilateral lambdoid synostosis. While this is rare it has a more worrying natural history. Lambdoid synostosis – fusion of the lambdoid suture located at the back of the skull – can result in a very unusual head shape and the small possibility of increased intracranial pressure and its associated problems, including developmental delay. Lambdoid synostosis comprised only 0.8% of synostotic cases (two out of 237) operated on over a five-year period and makes up an even smaller percentage of the number of new referrals made to the craniofacial unit. The craniofacial unit receives approximately 250 new referrals per year but only about

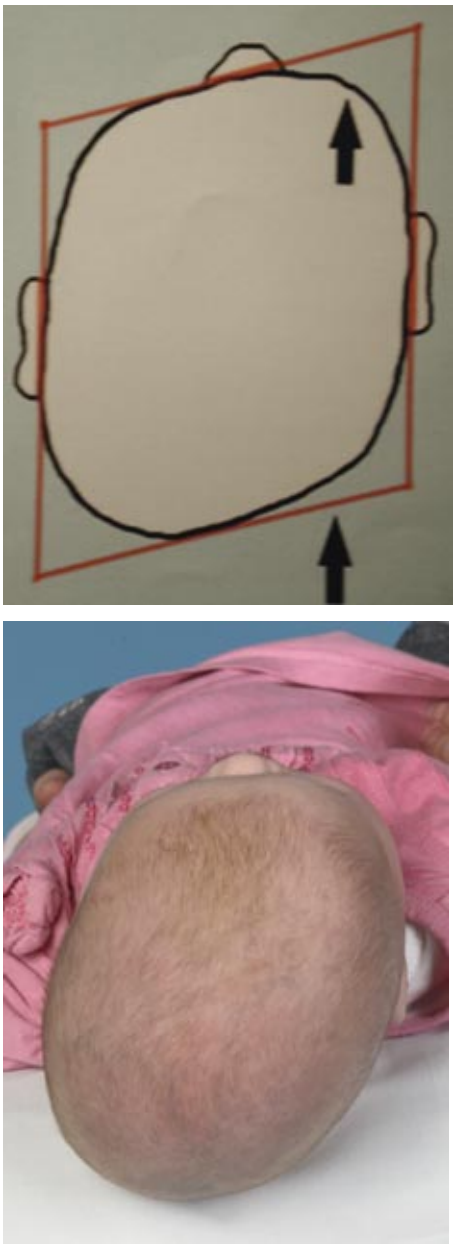

FIGURE 1 Deformational plagiocephaly, illustrating the typical features including flattening on one side of the head, ipsilateral translocation of the ear and frontal bossing on the affected side.

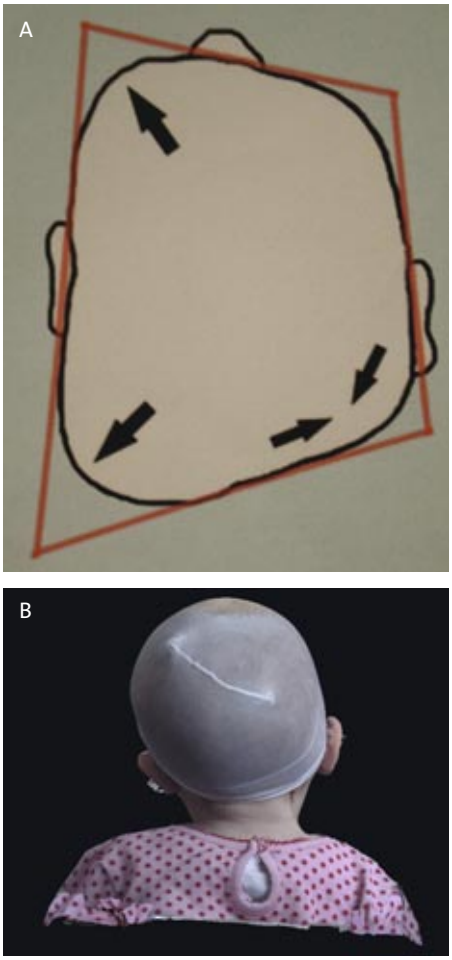

FIGURE 2 Right lambdoid synostosis. A) An illustration showing flattening on the affected side, posterior translocation of the ear on the right, parietal bossing on the left. B) In a patient, flattening on the right with ipsilateral posterior inferior displacement of the ear.

40% are accepted for surgery. The remainder of the referrals are a mixed collection of concerns but consist predominantly of non-synostotic deformational plagiocephaly patients. Thus there are a comparatively large number of referrals of patients with deformational plagiocephaly compared with patients with lambdoid synostosis.

Deformational plagiocephaly is associated with a number of typical features including flattening on one side of the head, ipsilateral translocation of the ear and frontal bossing on the affected side. This results in a parallelogram shaped skull (**FIGURE 1**).

Lambdoid synostosis may present with variable findings that can mimic the findings in deformational plagiocephaly and thus it can be challenging to diagnose (**FIGURE 2**). Concerns that a flat head may be synostosis rather than positional trigger several referrals to the craniofacial unit.

Assessment

Clinical history is the most important factor in determining the aetiology of a deformed skull. Questions regarding pregnancy, mode of delivery, head shape at birth, feeding[1,2], sleeping, neurodevelopment and presence of torticollis help to elucidate likely pathology. Charts can be used to attempt to describe the likely diagnosis (**FIGURE 3**).

Examination in the clinic includes overall assessment of:

- head shape
- presence of ridged sutures on the skull
- head circumference
- anthropometric measurements, including anterior posterior skull length, biparietal diameter (skull width), measurement of the oblique angles (right frontal to left posterior parietal and vice versa).

Three-dimensional photographs are taken to further document head shape. X-rays and CT scans are rarely performed as the diagnosis is usually obvious clinically, but if there is any doubt a CT with 3D reconstruction is carried out (**FIGURE 4**).

Management

Deformational plagiocephaly is managed with reassurance that it will get better over time. Information leaflets are provided. If the baby is less than six months' old, a pressure relieving mattress and 'tummy time' may be helpful. These measures take pressure off the back of the head and therefore allow the flattened side to fill out. Torticollis should be treated with physiotherapy. The unit does not recommend helmets as the condition improves with time and there is little information about the complications of moulding therapy. Surgery is not necessary. If the deformation is severe the child may have follow-up in a nurse-led clinic.

Lambdoid synostosis is managed with posterior cranial vault remodelling at approximately 12-14 months of age, depending on age at presentation. The rationale for surgery is the desire to normalise head shape and the small risk of raised intracranial pressure, which may cause developmental delay, if the condition is left untreated. Patients with lambdoid synostosis are seen in the multidisciplinary craniofacial clinic and are entered onto the comprehensive pre-operative assessment and post-operative follow-up pathway. This includes psychological and speech

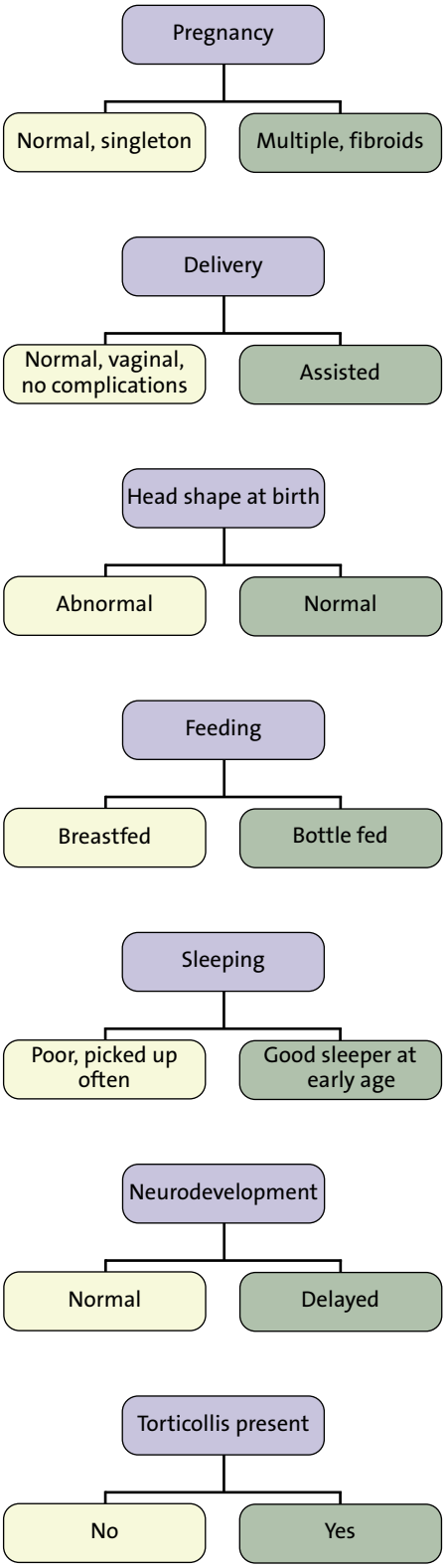

FIGURE 3 A schematic diagram to aid diagnosis via history. Predominantly yellow answers indicate craniosynostosis; predominantly green answers indicate deformational plagiocephaly. A baby who has minimal risk factors for an external force causing deformational plagiocephaly (green boxes) but has an abnormal head shape is more likely to have craniosynostosis. A normal head subjected to the factors in the green boxes, is more likely to develop deformational plagiocephaly.

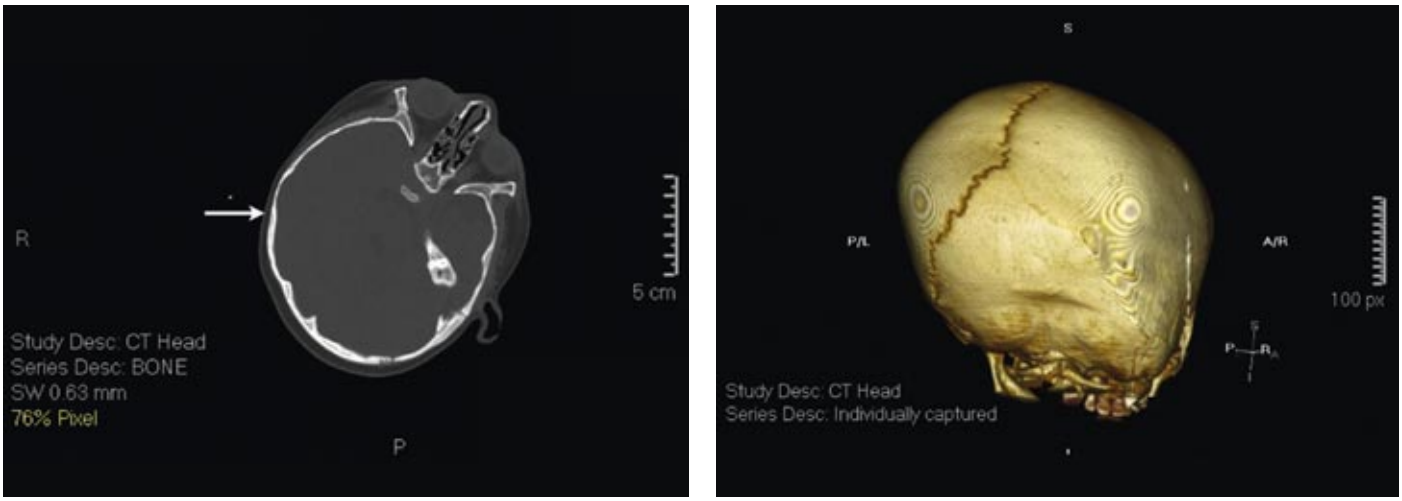

FIGURE 4 CT scans of the same patient seen in figure 2. Left: an axial bone window CT scan showing an absent right lambdoid suture (arrow). Right: A 3D CT scan showing the absent right lambdoid suture.

and language assessments along with a review by a geneticist.

Summary

Deformational plagiocephaly is a common condition that improves over time without any intervention but measures to relieve pressure from the back of the head may improve outcome. The key to diagnosis is the history and examination. Lambdoid synostosis is rare but requires surgical treatment. If there is any concern regarding head shape a referral should be made to the regional craniofacial service at:

- Alder Hey Children's Hospital, Liverpool
- Birmingham Children's Hospital
- Oxford Children's Hospital
- Great Ormond Street Hospital, London.

References

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