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Approach to Abnormalities in Head Shape and Size Part 2: Abnormalities in Head Shape

Developed by Lindsey Logan and Claire McNiven with Dr. Melanie Lewis, Dr. Lauren Redgate, and Dr. Peter Gill for PedsCases.com.
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Introduction:

Lindsey: Okay, we are back for part 2 of our series on head shape and size. My name is Lindsey Logan and I am a pediatric neurology resident in Toronto.

Claire: And I am Claire McNiven, a pediatrics resident at the University of Alberta. We want to thank our mentors who helped us develop and edit this podcast, Dr. Peter Gill from SickKids, Dr. Melanie Lewis from the Stollery Children's Hospital, and Dr. Lauren Redgate, a pediatrician in Calgary, Alberta.

Lindsey: Last podcast, we reviewed: head growth, skull anatomy. If you need a refresher, feel free to rewind to part 1 where we cover all of that in detail!

Lindsey: Today, our podcast is all about an approach to abnormal head shape.

Claire: The objectives we will cover in this podcast include:

1. Determine the differential diagnosis for a child presenting with an abnormal head shape.
2. Review key points on history that will support your differential diagnosis
3. Understand important aspects of physical examination to determine the etiology of the abnormal head shape
4. And finally, discuss the relevant investigations and management of children with abnormal head shapes.

Lindsey: Why don't I start us off with a case to frame our thoughts?

You are a medical student evaluating a 4 month old boy, Jayce, who was brought in by his mother after his grandparents voiced concerns about an odd head shape, specifically a prominence of the back of his head. He appears otherwise well but you do notice scaphocephaly, with an abnormally long and narrow head, indeed showing

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prominence of the occiput. When you palpate his sutures, you feel a ridge where you would expect to feel the sagittal midline suture.

Claire: Hmm. Before we decide what is going on here, let's talk about the approach to abnormal head shape, as well as the differential. Quick question - what was that term that you used there, "scaphocephaly"?

Lindsey: Oh yes, scaphocephaly! I have had to look that one up before, too. Basically, it is a fancy word for a head that is narrow and long, like it is being squished between the ears. There are also lots of other terms for variations in head shape. Some of our listeners may have heard the term plagiocephaly before. Do you know what that one means?

Claire: Plagiocephaly means flattening of one side of the head. In greek- Plagio means oblique, or slanting, and cephalo means head. So Plagiocephaly is a head where, rather than being round and circular, one side is oblique. Commonly, the flattened part of the head is the back (occipital region) from laying supine. You can also get plagiocephaly on the sides from having the head turned to a preferred side.

Lindsey: Yes, and brachycephaly is when the head is broad between the ears but flat front to back. We will talk more about these later!

Objective 1: Differential diagnosis

Lindsey: So, Claire, what is the differential for an infant with an abnormal head shape?

Claire: That is a good question. There are many possible causes, and a good place to start is the baby's age. If they have just been born, some of the changes in head shape might be from birth trauma.

Lindsey: Like we talked about before, Squishing a big head and brain through a small canal in the mother's pelvis is no easy feat!

Claire: Babies may have moulding of the head from being engaged in the pelvis during the later stages of labour. This is commonly referred to as caput succedaneum, or "caput", and it is a boggy collection of fluid right under the skin from the pressure applied by the pelvis during childbirth.

Lindsey: Caput is usually the culprit for all those newborn cone-heads. Someone once told me this is why they give all babies hats (but, of course it is also important to keep them warm!).

Claire: Haha, maybe! Another type of swelling that can happen is just over the bone underneath the periosteum. It usually happens when blood vessels in that location are ruptured and it is limited to a single bone because it can't escape the periosteal lining.

Lindsey: That means it doesn't cross suture lines. I think this rings a bell.

Claire: Correct, this swelling does NOT cross suture lines! It can also calcify, causing prominences over the bones that may persist for months.

Lindsey: Oh yes, it is called a cephalohematoma, and it is seen most commonly in the parietal regions, right?

Claire: That is spot on. The parietal bones are the most common site of injury for a cephalohematoma.

Lindsey: In addition to the swelling, after being in the birth canal, babies might have overriding sutures; in the sense that one of their bones was smushed into another causing them to overlap a little bit. This is usually not concerning and once out of the birth canal, the pressure of the growing brain allows the sutures to return to their normal position, often within the first few days of life.

Claire: The last head deformity related to birth trauma is the one we tend to worry about the most, a subgaleal hematoma. The subgaleal space is in between the skull and the scalp. It encircles the whole head and provides a lot of potential space for blood to collect, usually over hours after delivery. It can be really dangerous as a baby can lose a significant amount of blood there, and can be an emergency. The use of a vacuum at delivery can precipitate rupture of veins in this space.

Lindsey: These kids need to be monitored from a hemodynamic standpoint. They are also at risk of jaundice because of significant blood breakdown.

Claire: Yes! Now that we have covered birth-related cranial deformities, let's move on to other things that we may see as children get older: In older children lumps and bumps on the head can often be from lymph nodes, especially in the occipital region. Other lumps that show up may be more concerning and could require imaging like an ultrasound or a CT, or even a referral to a neurosurgeon.

Lindsey: As babies grow, we may see other changes in their head shape. One of the most common of these is flattening of the head from being placed supine on their backs for a prolonged period of time. This called positional plagiocephaly.

Claire: “Plagiocephaly”, as we mentioned before, just means flattening of one side of the head - in this case, it is the back part!

Lindsey: Preventing plagiocephaly is one of the reasons why tummy time - placing babies prone while observing them closely for periods of time- is so important!

Claire: Yes, tummy time is not only great for developing strength, but also for preventing flattening of the back of the head.

Lindsey: Another, more worrisome cause of an asymmetrical head as kids get older is craniosynostosis.

Claire: The term craniosynostosis means premature closure of the sutures while the head is still growing. Usually this is just a single suture. This happens sporadically, and most commonly, the underlying cause is uncertain.

Lindsey: However, there are also associated syndromes, some of which can be more severe or may involve fusion of bones elsewhere in the body. These are referred to as “syndromic craniosynostosis”, and include Crouzon syndrome and Apert syndrome.

Apert syndrome involves syndactyly (fusion of the fingers) and premature closure of the coronal suture, between the ears. This results in brachycephaly, which means an abnormally broad head with a recessed lower forehead.

Claire: Ok! I think that is good for a differential. Now, what would we ask on our history to differentiate these causes of abnormal head shape?

Objective 2: History

Claire: As with any encounter, it is very important to take a complete history including a full review of past medical, family, as well as social history.

Lindsey: Here we will review some pertinent details to ask.

Claire: In terms of the history of presenting illness, one of the first things to understand is the timeline of the concern in head shape. For anything present at birth or soon after, it is important to ask about instrumental deliveries and any history of birth trauma.

Lindsey: If the parents only noticed it more recently, or if the child is older, you can ask them when they first noticed and if it is getting better or worse. It is also important to ask

about any other abnormalities, anywhere else on the body. This might help point towards a syndrome.

Claire: Also review growth parameters other than the head (such as height and weight), and development: is the child continuing to gain developmental skills as expected or are they plateauing or losing skills (called regression). Regression is a big red flag.

Lindsey: For positional plagiocephaly, don't forget to ask how much time they spend on their back vs on their tummies, or if they have a positional preference to lie on a specific side.

Claire: You can also ask about torticollis, which is latin for "twisted neck". It refers to babies that hold their necks to one side due to tight muscles, uterine constraints, or other factors.

Lindsey: Let's quickly review some of the pertinent points on history for our case:

Claire: Remember, this is Jayce, a 4 month old term infant whose parents are bringing him in due to concerns voiced by the family about a progressive prominence of his occipital bones. Starting with birth history: the child was born by spontaneous vaginal delivery at term and had no head deformities at birth. He has had no other health concerns thus far.

Lindsey: He is meeting his developmental milestones, specifically, he has very good head control and is able to keep his head up during tummy time, which he enjoys. He is also rolling front-to-back. Jayce's parents say he is able to track objects across midline, bring his hands together, reach for objects and that he can shake a rattle. He responds to his name, laughs, and enjoys vocalizing during conversations.

Claire: He lives with his parents but sees his grandparents often and is an only child. He is fully breast-fed and feeds well with good urine and stool output. His growth has been tracking nicely on his growth curve. There is no history of trauma. On family history, there is no mention of anyone having odd head shapes or any cranial conditions.

Lindsey: Last, very importantly, the family says that they only really started noticing the bony bump at the back of his head a few months ago, and it seems to be getting worse.

Claire: So, Jayce's history is overall unremarkable except for the fact that he was **not** born with any bump at the back of his head, and now it seems to be getting worse. Let's move on to physical exam and see what kind of things to look out for!

Objective 3: Physical Exam

Lindsey: For a child with abnormal head shape, there are a number of things we want to pay attention to on physical examination. Of course, it is always important to complete a full physical examination, especially if a baby looks syndromic and you are suspicious about other abnormalities.

Here we will discuss specific things to focus on related to head shape.

Claire: First, observation: just looking at the infant's head may tip you off to abnormalities in shape. Take off their hat if they are wearing one! Look at their head from all angles. An especially good angle to assess plagiocephaly and determine if it is positional versus craniosynostosis is from the top.

Lindsey: Ahh yes, like a birds eye view.

Claire: Yes. Looking from the top helps identify the area of flattening, the overall shape, and if there's any displacement of the ears (forward or backward). These are important clues that can aid in your diagnosis. For example, positional plagiocephaly is often described as looking like a parallelogram - as one side of the back of the head flattens, the ear on that side is pushed forward. Sometimes the forehead and eye on that side can shift forward too, creating asymmetry in the face. This differentiates positional plagiocephaly from premature fusion of the lambdoid suture - also called lambdoid synostosis- where the ear on the affected side is actually displaced backward. Babies with positional plagiocephaly might also be lacking hair along the backs of their head where they tend to rub it against a surface.

Lindsey: It is also very important to palpate the head! Premature fusion of the sutures - craniosynostosis - often produces a palpable ridge. In other cases, you may feel splaying of the sutures if there is pressure pushing them apart, or perhaps overriding from them being pushed together. An inability to palpate cranial bones is also worrisome and can tip you off to an underlying bone disorder, such as rickets.

Claire: In the case of a newborn baby, palpation will help you feel whether a swelling or bulge extends across sutures, if it is boggy and soft or if it is tight.

Lindsey: While you are palpating you can also observe the child's face and movements. Are they dysmorphic? Are they appropriately moving, and are movements symmetric, or is there an indication of a nerve injury or paralysis? Is the range of motion of their neck limited?

Claire: Last but not least, it is important to measure their head! (Good thing we already learned how!) Small and large head sizes are important to detect, which we will cover in more detail in our next podcast.

Lindsey: So, what does our physical exam show in little 4 month old Jayce?

Claire: Jayce is an active boy who is behaving in a developmentally appropriate manner on our exam. His growth and head circumference parameters are normal. His neurological examination is symmetric with normal cranial nerves, reflexes, strength, and tone. However, as we mentioned previously, when we take a closer look at his head we notice scaphocephaly. His head is abnormally long and narrow, with a prominence of the occiput. Upon palpation of his sutures, we feel a ridge-like prominence along where you would expect to feel the sagittal midline suture.

Lindsey: Hmm, that examination does make me concerned about possible craniosynostosis of his midline sagittal suture.

Claire: Exactly - me too! Let's move on and talk about the next steps - investigations and management.

Objective 4: Investigations and management

Lindsey: Okay! Now for investigations and management. Obviously, it depends on what you observe as to whether any investigations are needed.

Claire: For birth trauma, often you can palpate a baby's head and tell whether swelling is from caput, a cephalohematoma, or a subgaleal hemorrhage.

Lindsey: In the case of a subgaleal, the mass can be fluctuant and soft. These babies need to be monitored carefully as they can actually bleed out their entire blood volume into this space! So watch for changes in vital signs and responsiveness.

Claire: And, as we mentioned earlier, babies may require phototherapy or other measures to assist with jaundice from the breakdown of red blood cells in these areas.

Lindsey: For other types of head bulges, an ultrasound is always a good first step depending on the level of concern.

Claire: If small, you may watch and wait, but if large or rapidly changing, imaging is warranted.

Lindsey: Now for flattening and plagiocephaly:

If a baby has positional plagiocephaly you will often be able to tell based on the history and physical examination.

Claire: In some cases, you will be able to counsel the parents to initiate more observed “tummy time” during the day. The CPS statement on positional plagiocephaly recommends 10-15 minutes, at least 3 times daily.

It is important to note, however, that as per CPS guidelines, infants should still be placed on their backs to sleep, to reduce the risk of Sudden Infant Death.

Lindsey: A helpful trick to make tummy time a little more tolerable is to put a rolled towel under the child’s chest, and allow their arms to stretch out in front of them. You can also put interesting things for them to look at that helps motivate them to lift their head.

Claire: For sleeping, you may try alternating the end of the crib that parents place the baby’s head at, since children often like to look out into a room. You can also put colourful crib-safe toys or mirrors in the crib to help encourage a child to look in different directions.

Lindsey: If the positional plagiocephaly is severe, or if these tips don’t work and the baby is nearing 6 months, they may need to be referred to a special head shape clinic where they can be seen by physical or occupational therapists and fitted for a helmet or headband. Helmets are expensive, require frequent reassessment and can cause skin issues, so they are only recommended in severe cases. Helmet therapy increases the initial rate of improvement, but has not been shown to change to final outcome in head shape.

Claire: If a child has torticollis, a referral to a physical therapist is indicated.

Lindsey: And if you are concerned about craniosynostosis, or premature fusion of the sutures, a referral to neurosurgery or plastics craniofacial clinic would be a good first step.

Claire: Let’s go back to our case: As a reminder, Jayce is 4 months old and was brought into the clinic because of concerns about a prominent occiput, as well as a narrow head between the ears. The sagittal suture felt ridge-like on exam.

Lindsey: Hmm, it doesn’t sound like it is positional plagiocephaly. And, when we discussed the physical exam, we did mention that craniosynostosis can cause ridges or prominences of the sutures.

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Claire: Exactly! This case warrants a referral to a craniofacial clinic to see a specialist (usually a neurosurgeon or plastic surgeon), who may perform a CT and decide whether the craniosynostosis needs to be surgically released.

Lindsey: Sounds like a plan! And now, Let's discuss another classic case in peds, that is really important to highlight.

Claire: This type of case will honestly be what family physicians and pediatricians see the most:

Parents bring you their 2 month old infant because of concern about flattening of the back of the head. When you examine this baby boy, you note he is developmentally appropriate however has a spot on his occiput where all of his hair is worn down. When you try to put him on his tummy his mom remarks "oh, he really hates tummy time"

Lindsey: Sounds like a classic case of positional plagiocephaly to me. I would encourage more tummy time, trying the roll under the arms, and also maybe some of the other tactics we discussed to get the child to move their head to the left or right when supine!

Claire: I agree. If that doesn't work we can always refer to physical or occupational therapy, or consider a head shape clinic for a helmet or head band if the plagiocephaly is severe.

Lindsey: And that wraps up Part 2 of our podcast on approach to abnormal head shape. Let's quickly review what we covered today:

Review

- After birth, children may have abnormal head shapes associated with birth trauma or moulding, as well as any instrumented deliveries.
- Caput succedaneum is superficial moulding of the skull and will resolve quite quickly.
- Cephalhematoma is under the periosteum so does not cross suture lines, and can harden into the bone before going away.
- Subgaleal hematoma is a boggy collection under the skin of the scalp and must be closely monitored.
- After birth, it is important to track head circumference and head shape at regular pediatric visits.

- If the sutures between bones close too early it leads to a condition called craniosynostosis, which can result in abnormal head growth and abnormal head shape. This often requires neurosurgical surgical referral.
- Plagiocephaly is another term for abnormal head shape. It essentially means flattening of one side of the head!
- The most common cause of plagiocephaly is positional - when babies spend too much time on their backs or with their head to one side.
- Depending on the severity of positional plagiocephaly, it is possible to initiate environmental modifications and exercises like tummy time. The CPS recommends three times per day for 10-15 minutes. If these do not work, there are often head shape programs at pediatric centres to determine whether further intervention such as physiotherapy and/or custom helmets are indicated.
- Okay, that is it for our overview of head growth, head shape, and measuring head circumference.
- Be sure to stay tuned for parts 3 and 4 of our podcast, which discuss an approach to big and small heads - macro and microcephaly.

Thanks for listening!

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