METOPIC SYNOSTOSIS

Metopic synostosis is a condition where the metopic suture in a baby's skull closes too early, before the brain has fully developed.

Otherwise known as?

Trigonocephaly and metopic craniosynostosis

Signs & Symptoms

Key characteristics of metopic synostosis:

Skull Shape Differences:

- Trigonocephaly: a triangular-shaped forehead.
- Midline Ridge: a noticeable ridge along the metopic suture.
- Narrow Forehead: the forehead appears overly narrow compared to the rest of the skull.
- Bitemporal Narrowing: the temples appear pinched inward.

Facial and Eye Changes:

- Hypotelorism: eyes may be closer together than normal.
- Eyebrow irregularities: eyebrows may appear angled or raised.

Mechanism (*the how*)

Babies' skulls are made of several plates joined by flexible seams called sutures. Normally, these seams stay open during early childhood to make room for brain growth.

The skull and brain grow together. The growing brain and surrounding soft tissues help guide how the skull expands. If these growth signals or forces are altered in one area, skull growth may follow a different pattern and a suture may close earlier than expected. This is one way that specialists understand the skull shape changes seen in metopic synostosis.

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In metopic synostosis, the metopic suture — the seam that runs from the top of the head down the middle of the forehead — fuses earlier than it should. When that seam closes, growth in that direction slows, so the head grows more in the areas that are still open. This can cause a ridge on the forehead and a more triangular forehead shape.

Metopic synostosis is also understood as a pattern of bone growth involving irregular osteoblast activity. Osteoblasts are the “bone-building” cells in your child’s body; in craniosynostosis, they may not follow the usual growth pattern around the affected suture.

Causes (*the why*)

Most cases of metopic synostosis occur sporadically, meaning there isn’t a single identifiable cause. Sometimes doctors may find a genetic explanation; if so, that’s something that happens in genes naturally—it isn’t caused by anything a parent did or didn’t do. Studies have explored possible factors during pregnancy, but everyday behaviours are not known to cause this. It may help to know - you did not cause this.

1. Genetic Factors:

- Some cases are linked to genetic mutations or syndromes (e.g., Muenke syndrome, Saethre-Chotzen syndrome) and may be associated with mutations in genes such as FGR1, FGR2, FGR3 and TWIST1.
- Family history may play a role, though most cases occur sporadically.

2. Prenatal Factors:

Most cases of metopic synostosis appear sporadically and are multifactorial. Researchers have reported possible prenatal or developmental contributors, but links are not proven, and no routine pregnancy behaviour is known to cause this condition.

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Testing & Diagnosis

The diagnosis of metopic synostosis typically involves a combination of clinical evaluation, imaging studies, and sometimes genetic testing.

1. Clinical Evaluation:

- Physical Examination: A doctor will examine the infant's head shape, noting any Differences such as a triangular forehead or midline ridge.
- Symptoms and History: The doctor will ask about the pregnancy, delivery, and family history to rule out syndromes or other causes.
- Growth and Development Monitoring: The child's physical and developmental milestones may be reviewed to assess if there are any delays linked to the condition.

2. Imaging Studies:

- X-ray: An initial X-ray of the skull can sometimes show the early signs of suture fusion and abnormal skull shape.
- CT Scan (Computed Tomography): A more detailed imaging method that offers a clear view of the skull's bones, allowing confirmation of suture fusion and assessing the extent of the condition. It can also reveal any brain changes that might be present due to increased pressure or cranial deformity.

3. Genetic Testing (if needed):

- If a syndrome like Muenke syndrome or Crouzon syndrome is suspected, genetic testing might be done to look for mutations linked to craniosynostosis. This is typically performed if there are other physical Differences, developmental delays, or a family history of cranial malformations.

4. Additional Tests (if neurological concerns are present):

- MRI (Magnetic Resonance Imaging): If there are signs of intracranial pressure or neurological concerns, an MRI can provide a detailed image of the brain and help assess any impact from the different skull development.
  - ICP Monitoring: In rare cases, doctors may monitor intracranial pressure if there is concern that brain growth is being hindered.

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Treatment

The primary treatment for metopic synostosis is surgery in moderate to severe cases to address skull shape and allow normal brain growth.

The specific procedure undertaken depends on the child's age and severity of the condition, but may include cranial vault remodelling (i.e., surgery to reshape the skull) or endoscopic surgery in conjunction with springs or helmet therapy.

However, a visible or feelable ridge along the metopic suture does not always mean surgery is needed. Some children have a benign metopic ridge without the broader skull-shape changes of metopic synostosis. Parents may wish to ask their craniofacial team how they distinguish a benign metopic ridge from metopic synostosis, and what features make surgery recommended or not recommended for their child.

Internationally, including Australia, there is no single preferred type of surgery, with surgical options depending on the craniofacial team and the craniofacial surgeon's expertise and assessment of the individual child.

Individual craniofacial units and craniofacial surgeons in Australia can advise parents about their preferred surgical technique.

Did you know?

In Australia, approximately 1 baby in 10,000 births is diagnosed with metopic synostosis.*

Some Renaissance and medieval paintings depict people with prominent foreheads or triangular skull shapes. Although the artists may have exaggerated the individual's features, it is also possible that some subjects had the as yet unidentified condition of 'metopic synostosis'.

References:

Junaid, M., Slack-Smith, L., Wong, K., Bourke, J., Baynam, G., Calache, H., & Leonard, H. (2022). Epidemiology of Rare Craniofacial Anomalies: Retrospective Western Australian Population Data Linkage Study. The Journal of Pediatrics, 241, 162-172.e169 https://doi.org/10.1016/j.jpeds.2021.09.060

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How we can support you:

- Care packs
- Financial assistance
- Family support coordinator
- Connection to other families