

CRANIOSTENOSIS

**PREMATURE FUSING
OF
BONES IN THE SKULL**

**Esther Ost
SPED 530 – April, 2005
Minot State University**

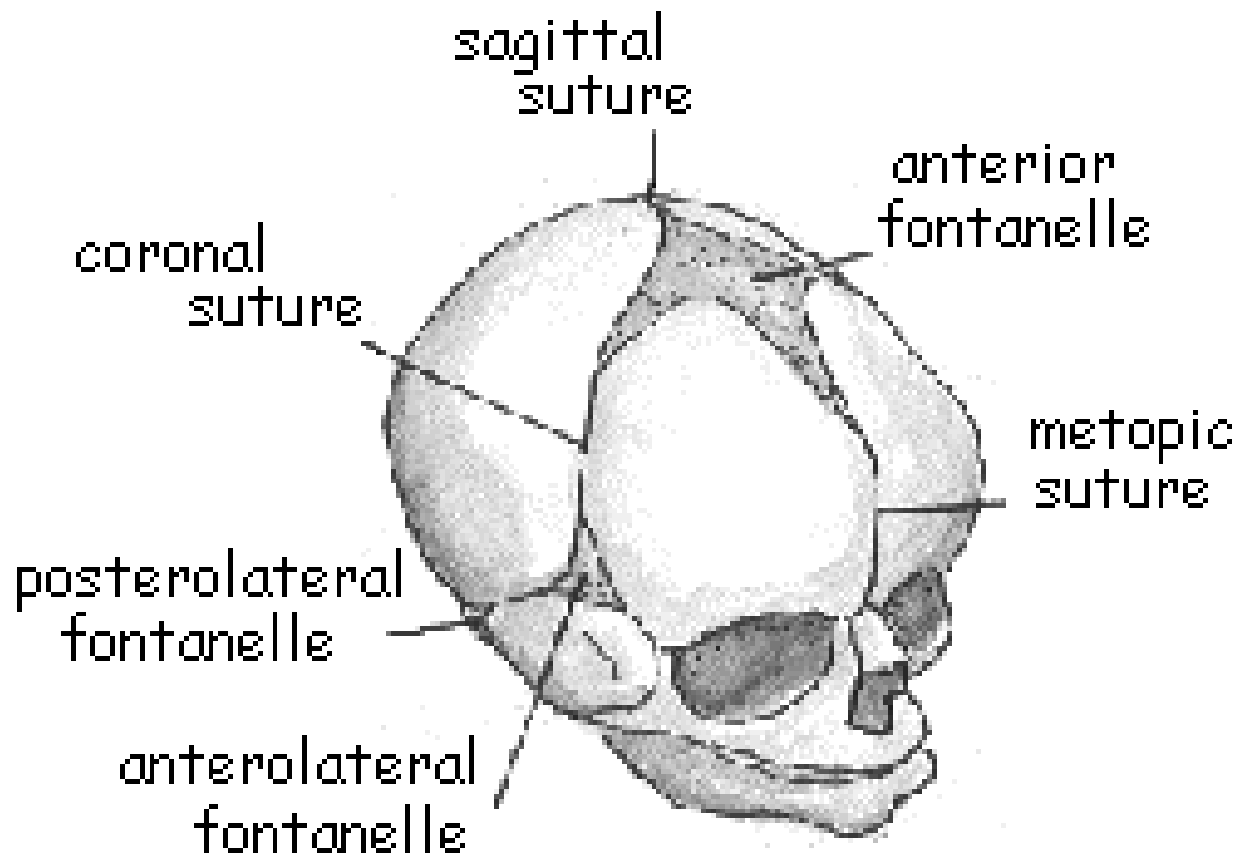
What is Craniostenosis? (Craniosynostosis)

- A condition in which one or more of the sutures in a fetus's or baby's skull closes prematurely, resulting in abnormalities of bone and soft tissues in head and face.
- The affected area of the skull does not expand normally as the brain grows.
- The brain is forced to grow in a different direction where the bones are not resisting growth.

Normal skull development

- The skull consists of five thin, curved bony plates that meet along lines called *sutures*.
- As the brain grows, it pushes against the skull, causing the sutures to stretch, allowing for rapid expansion.
- Usually a baby's brain doubles in size in the first 6 months of life and again by age 2.
- The head shape develops symmetrically.

Diagram of skull, showing location of fontanelles and sutures.



Characteristics of Craniostenosis

- Abnormal head shape, usually obvious at birth.
- Face may be flat or concave.
- Eyes may bulge because sockets are too shallow or too small to contain them.
- Outer ear (*pinna*) may be missing or only partially formed.
- Small airway may obstruct breathing.

Characteristics - cont.

- Head may grow
 - too long from front to back (boat shaped)
 - too wide
 - too tall
- One side may grow normally, the other remain smaller.
- The lower jaw may grow normally, the upper jaw at about half normal rate (sunken face).

Treatment

- The only treatment is surgical.
- The majority of operations are performed within the first three to five months after birth.
- Early (immediate) surgery is necessary when
 - The baby is unable to breathe freely
 - The baby cannot swallow

Treatment – cont.

- Surgeries that must be done, but need not occur immediately after birth:
 - Align eyes so they can focus on the same plane.
 - Adjust upper face planes so eyelids may close.
 - Bring upper face forward and reposition jaws so chewing can occur properly.
 - Provide an outer opening to ears so sound can enter and fluid is not trapped inside.

Treatment – cont.

- An important first step following the baby's birth is to seek care at a medical center that has a team that specializes in craniofacial disorders.
- Because multiple corrective surgeries may be required, it is very important that the first surgeries be done by experienced, competent surgeons as their work forms the foundation for all the surgeries that follow.

Treatment - cont.

- When surgeons are simply releasing the fused sutures, they make an incision behind the hairline, peel the skin over the back of the head, or down over the face and perforate or crack the suture seams.
- Severe deformities require extensive bone removal, recontouring and finally replacement of the skull.
- Bone “dust” may be frozen and saved for future surgeries.

Types of Craniostenosis

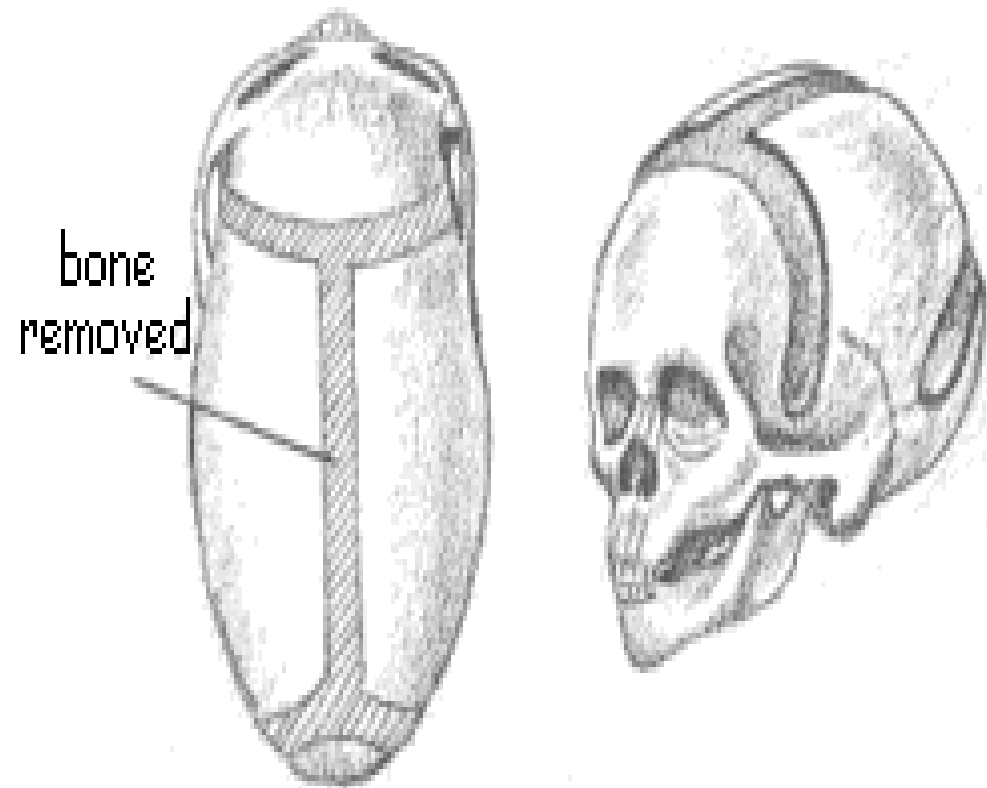
- Single suture craniostenosis
 - Only one suture is involved.
 - The shape of the head is distorted.
- Multiple suture craniostenosis
 - Two or more sutures are involved.
 - The shape of the head is distorted.
 - Usually other developmental problems occur.

Single Suture Craniostenosis

- ***Sagittal suture***: fusion of the suture between the two sides of the head. The head is long and thin. (Boat-shaped head)
- ***Metopic suture***: fusion of the suture between the two sides of the forehead plates, results in a triangular-shaped skull.
- ***Unilateral coronal suture***: fusion of the suture between one side of the forehead and side bones, resulting in a lopsided or twisted skull.

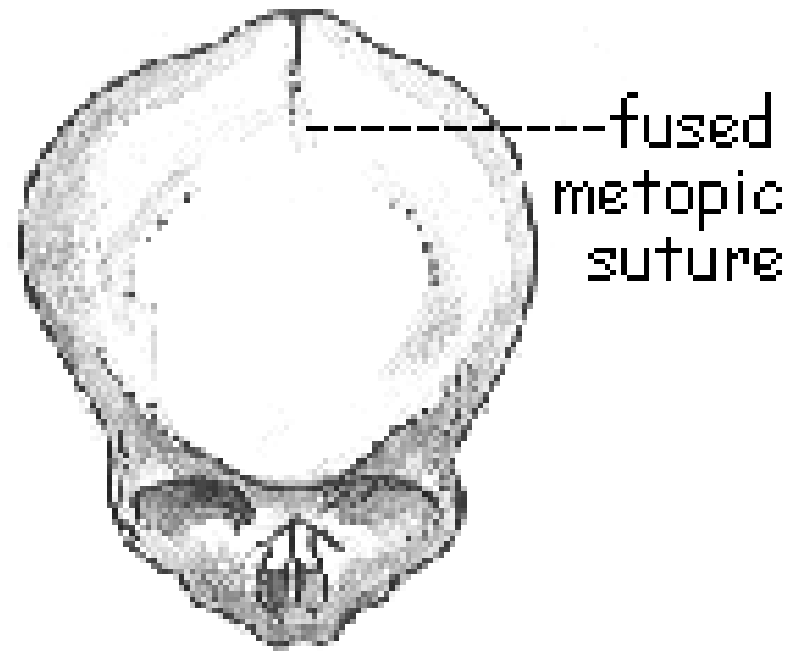
Saggital suture

- Surgery removes the suture and widens the skull by opening the coronal and lambdoid suture on the side of the head.



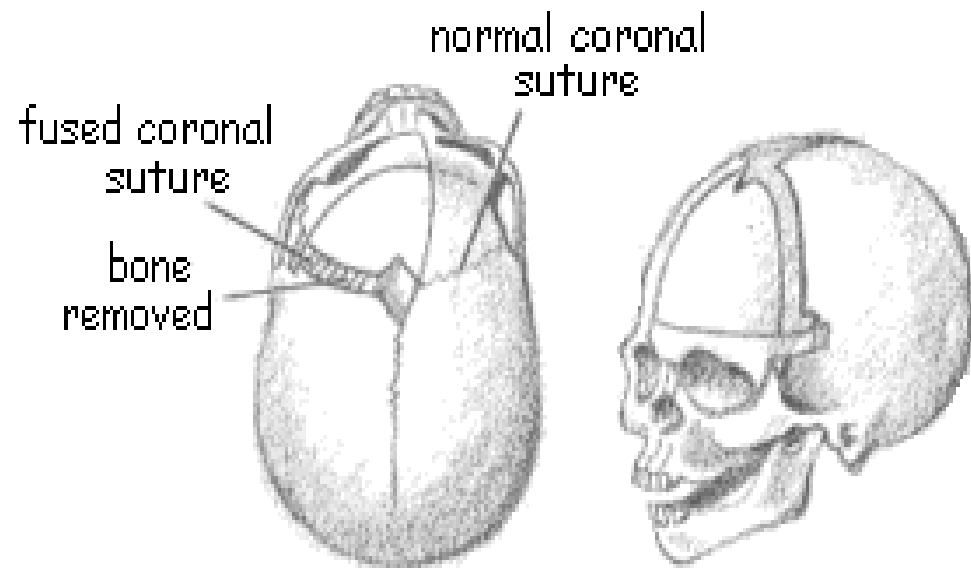
Metopic suture

- Treatment involves releasing the suture, expanding and rounding out the upper face, forehead and skull.



Unilateral Coronal Suture

- When one suture is fused, it is removed, the forehead and brow are reshaped and brought forward to match the other side. The cheek is also brought forward to reposition the eye socket.



Multiple Suture Craniostenosis

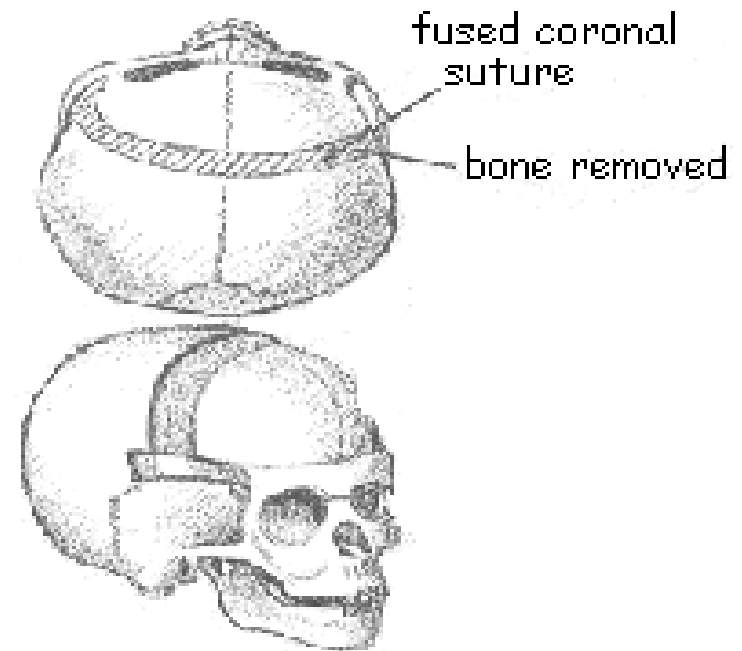
- Multiple suture craniostenoses are called syndromes (Apert, Crouzon, Pfeiffer, Saethre-Chotzen) and usually include a combination of distinctive features including
 - Head and facial distortion
 - Bulging eyes
 - Misalignment of jaws and teeth
 - Cleft palate
 - Webbed fingers and/or toes
 - Vision and hearing problems

Multiple Suture - Cont.

- Intelligence is **usually not impaired** when fusion is treated early.
- Mental retardation may occur in Apert syndrome about 50% of the time.

Bilateral Coronal Fusion

- The head is short and wide.
- Eyes bulge and skull bulges around the ears.
- When both coronal sutures are fused, they are removed, the skull narrowed and lengthened. Cheekbones are brought forward to reposition the eye socket.



Breathing Problems

- **Choanal Stenosis** – soft tissue or bone blocks the back of the nasal opening.
- **Underdeveloped Midface** – too little space in the upper jaw region causes airway to be blocked by adenoids, soft palate and soft tissue.
- **Underdevelopment of Lower Jaw (Hypoplasia)** – tonsils and adenoids block opening into the larynx and esophagus.
- A tracheotomy may need to be performed immediately after birth. As the child grows his airway may become large enough for air flow.

Eating Problems

- Babies who are not able to suck and/or swallow may need to be “tube fed”.
 - A *naso-gastric* tube is inserted through the nose, esophagus and into the stomach.
 - A *gavage* tube is inserted through the mouth and into the stomach.
 - A *gastrostomy (G-tube)* is inserted through an incision in abdomen directly into the stomach.
- Naso and gavage are short-term measures.

Vision Problems

- *Protruding or bulging eyes:* An eye-ball may be dislodged from its socket if the child is bounced too hard or hit in abdomen.
- *Widely spaced eyes* that cannot focus together because of different plane positions.
- *Strabismus* – eye do not track together. One may turn up (*hypertropia*), one may turn out (*extropia*) or one may turn in (*esotropia*).
- *Corneal irritation or cracking* due to dryness when eyelids cannot completely cover the eye surface because of facial distortion.

Hearing Loss

- The ear canals may be very small or completely closed.
- Fluid may build up in the middle ear because the eustachian tube and throat are not properly formed.
- The outer ear (*pinna*) may be missing or only partially formed.
- Inner ear canal may be narrow or absent.
- Sensorineural disfunctions may contribute to the child's hearing loss.

Facial Differences/ Craniofacial anomalies

- All types of craniostenosis cause facial differences.
- Severe facial differences may interfere with parent-child bonding, especially at feeding times when the parent finds it difficult to look at the child's face.
- Most facial differences do not affect the child's intellectual abilities.

Responding to Others' Responses to Your Child's Facial Differences.

- Parents and their child often must endure stares, double-looks, pointing, thoughtless and even cruel remarks and questions.
- People often are caught off-guard and embarrassed by their own first responses.
- They may not interact normally with someone with a facial difference:
 - Begin and end conversations more abruptly.
 - Ask intrusive questions.
 - Make eye contact for shorter periods.

Responding – Cont.

- Strangers often assume a person with facial differences is mentally deficient.
- The parent sets the tone for peoples' response.
- When someone is staring, look that person in the eye and smile. (It's disarming.)
- Parent is the model for their child's reactions.
- Learn to forgive stranger in their ignorance.
- Tell people the child was born this way and will require/or has had corrective surgeries.

Responding – cont.

- A child with facial differences needs to develop social skills that go beyond those required by most people.
 - This may be extra difficult because they may have a hearing and/or speech difficulty.
- Help the child to come away from the situation feeling a sense of integrity.
 - You may feel sad or depressed that the situation arose, but you can feel good about how you handled it. Your child needs this example.

Parents Must Advocate for Children with Facial Differences

- Professionals may not treat them appropriately.
- People often assume a child is mentally retarded because of their appearance or speech or hearing impairment.
- Laws that provide standards may not be interpreted properly to provide for your child's needs.
- Insurance may be unwilling to cover procedures they perceive to be for cosmetic reasons.