Craniosynostosis

Premature closure of cranial sutures which may lead to increased ICP due to inability of skull to accommodate growing brain tissue; may be an isolated defect or part of malformation syndrome including Apert's or Crouzon's, both of which may be associated with facial dysmorphic features and a potentially difficult airway

ANESTHETIC CONSIDERATIONS:

- Pediatric patient
- Syndromes associated with craniofacial anomalies → **risk of difficult airway**
- Associated syndromes:
  - **Crouzon's** (AD, craniosynostosis, maxillary hypoplasia, shallow orbit with proptosis)
  - **Apert's** (AD, craniosynostosis, syndactyly of hands / feet)Potential for **increased ICP**
- Potential for **massive fluid and blood replacement** (bleeding d/t incision size and vascularity of the bone)
- Risk of **VAE**
- Considerations of prone positioning → airway maintenance despite lack of access to the face

ANESTHETIC GOALS:

- Ensure appropriate examination of airway and necessary equipment / personnel to manage
- Vigilant monitoring of intraoperative blood loss and early transfusion (helpful to calculate transfusion trigger)
- In case of increased ICP, relaxed brain is essential, by hyperventilation, diuresis, adequate venous drainage
- Prompt recognition of intraoperative VAE, coagulopathy, massive hemorrhage

HISTORY

- Standard pediatric history
- History of previous anesthetics including history of difficult intubation
- Obtain history as to whether a single suture involved for cosmetic reasons as opposed to multiple sutures in a patient with congenital syndromes and increased ICP
- History of congenital heart defects (ASD, TOF, PDA etc.)
- Previous intubations, history of tracheal stenosis / tracheobronchomalacia
- History of OSA
- History suggestive of increased ICP
  - Vomiting, lethargy, seizures
- Other neurological history (hydrocephalus, seizure disorders, mental retardation)

PHYSICAL

- **VITALS** – including room air SpO_2
- **HEENT** – examination of airway including large tongue, micrognathia, short neck
- **CVS** – precordial examination for murmurs / irregular rhythms suggestive of congenital heart disease
- **RESP** – signs of tracheal stenosis / tracheobronchomalacia (tachypnea, sternal retractions, accessory muscle use
- **CNS** – screening neurologic exam

INVESTIGATIONS

- **Labs**
  - Cross-match with blood available
  - CBC (baseline Hb)
  - E-lytes (electrolyte abnormalities)
  - ABGs
- **Imaging**
  - ECG
  - ECHO / cardiac catheterization (as suggested by history / physical exam)

OPTIMIZATION

- Ensure cross-matched and blood available
- ICU consult for postoperative disposition

ANESTHETIC OPTIONS

- GETA is the only option for this procedure

ANESTHETIC SETUP

- **Drugs**
  - Standard emergency drugs
- **Equipment**
  - Standard CAS monitors including temperature
  - Single suture in healthy child needs only single well functioning IV (20-22 Ga)
  - Multiple sutures involved:
    - At least 2 large bore IVs
    - Arterial line
    - Consider central line
    - Urinary catheter
    - Precordial Doppler

- Warming equipment available
- Airway
  - Surgeon for tracheostomy if known difficult and previous attempts unsuccessful
  - AFOI a possibility
  - Numerous laryngoscope blades, airways, masks available
  - Reinforced ETT

MANAGEMENT OF ANESTHESIA

- **Induction**
  - Normal airway in healthy child → an inhalational or IV induction
  - Increased ICP → IV induction without ketamine
  - May need to modify induction depending on syndrome involved
- **Maintenance**
  - Blood loss can be > 150% of blood volume, requiring massive transfusions
  - Coagulopathies, electrolyte abnormalities, acidosis, and inadvertent extubation are possible → low threshold for intraoperative monitoring of CBC / INR / PTT / ABG
  - Warming devices as necessary
- **Emergence**
  - A short procedure (one suture) with minimal blood loss can be extubated in the OR
  - Numerous suture involvement associated with significant blood loss may be extubated if warm, hemodynamics as preoperatively
  - For the difficult airway, plan to have necessary equipment
  - For procedures below the orbital ridge, may need to leave intubated until edema resolves

DISPOSITION & MONITORING

- HDU
- In the postoperative period strict surveillance of vital signs and HCT is mandatory
- Other major concerns are the airway and problems with oxygenation and ventilation
- If patient remains intubated, the need to sedate and ventilate and to decide when and how to extubate can be discussed by the surgeons, anesthesiologists, and intensivists
- Bleeding and metabolic abnormalities may occur in the postoperative period also

COMPLICATIONS

- Airway difficulties
- Massive blood loss
- VAE

PATHOPHYSIOLOGY

- Incidence of 0.4-1.0 per 1000 live births
- Premature closure of the sagittal suture alone (~57%) is the most common manifestation, other sutures, singly or in combination, may be involved
- Degree of deformity depends on the number of sutures & time when fused
- Sagittal suture fusion: scaphocephaly
- Unilateral and bilateral coronal fusion: plagiocephaly and brachycephaly
- Multiple sutures associated with syndromes such as Apert's and Crouzon's, that affect cranium, cranial base and face (Crouzon's associated with strabismus and MH)

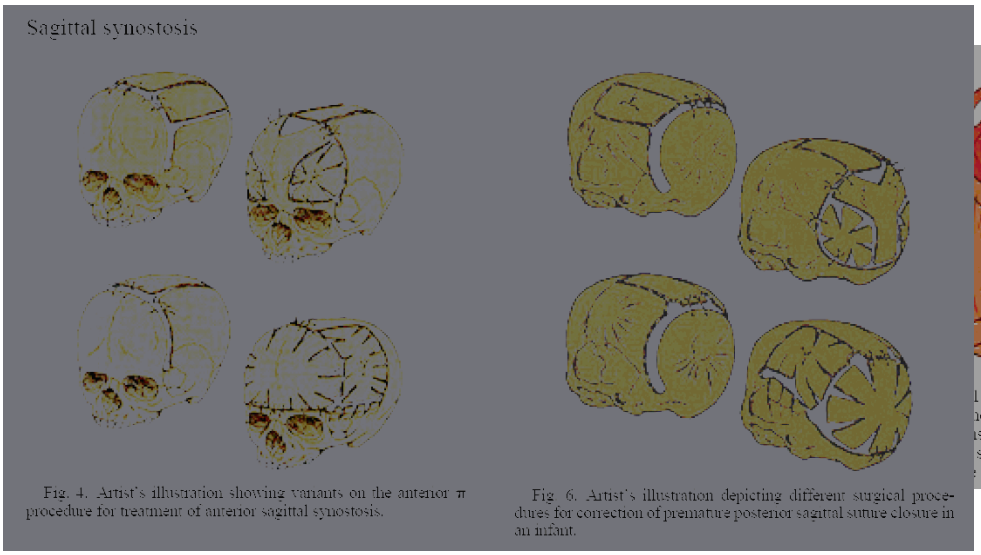
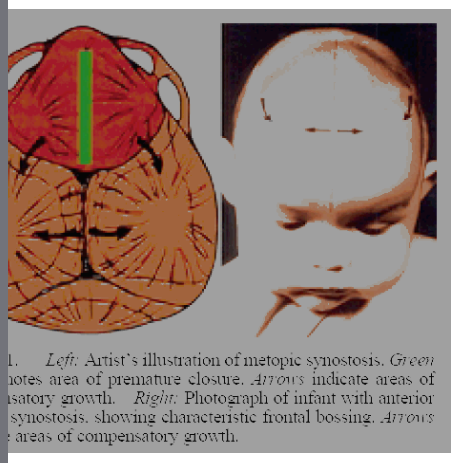


Fig. 4. Artist's illustration showing variants on the anterior π procedure for treatment of anterior sagittal synostosis.

Fig. 6. Artist's illustration depicting different surgical procedures for correction of premature posterior sagittal suture closure in an infant.



1. *Left:* Artist's illustration of metopic synostosis. *Green* notes area of premature closure. *Arrows* indicate areas of satory growth. *Right:* Photograph of infant with anterior synostosis, showing characteristic frontal bossing. *Arrows* areas of compensatory growth.

- May also be classified by 58 different syndromes: monogenic syndromes that are AD, AR, X-linked, or with an unknown inheritance pattern
  - Other syndromes are chromosomal, environmental induced, or of unknown cause
  - Teratogens that cause craniosynostosis are diphenylhydantoin, aminopterin, methotrexate, retinoic acid, oxymetazoline, and valproic acid
  - Can also be d/t metabolic and hematologic disorders
- Relationship of craniosynostosis to neurological deficit and hydrocephalus is not clear
  - Appears to be agreement that total synostosis (cloverleaf skull) may cause elevation of ICP during rapid brain growth periods
  - Single suture involvement → effect on the brain is uncertain

- Felt by many that the goal of operative management is primarily cosmetic
- Best result is achieved when surgery is performed at an early age (before three months of age and even at four to six weeks of age)
- Anesthetic considerations: include large rapid blood loss, blood transfusion, airway maintenance despite lack of access to the face and patient positioning which frequently includes the prone position
- Intraoperative blood loss:
    - Assessment of blood loss difficult:
        - Efforts to weigh sponges, use graduated suction cylinders and colorimetric methods to measure hemoglobin concentration may be inadequate for the pediatric patient
        - Portion of small blood volumes forms unmeasurable stains on surgical gowns and drapes
        - Irrigating fluid and cerebrospinal fluid alter sponge weight and suction trap volume
    - Several factors influence the amount of blood lost during this procedure
        - Controlled ventilation, induced hypotension, scalp infiltration and hemostatic scalp sutures all may reduce blood loss
        - Periosteum not scalp is major source of blood loss therefore scalp infiltration with LA containing epinephrine may not be justified
    - A HCT of 0.30 remains a reasonable postoperative goal → recommend an allowable blood loss to a HCT of 0.30 with crystalloid replacement solutions
        - Further blood loss should be replaced with packed red blood cells
    - Type of surgery (sutures involved) most predictive of blood loss (not duration):

TABLE III Duration of surgery

Suture	*Mean duration in minutes (± SD)*
Sagittal	74 ± 26
Unicoronal	105 ± 26
Metopic	78 ± 19
Bicoronal	135 ± 30
Others	91 ± 39

TABLE IV Blood loss (per cent EBV)

Suture	*Blood loss (mean ± SD)*
Sagittal	24.1 ± 14.7
Unicoronal	20.9 ± 11.9
Metopic	42.0 ± 12.0
Bicoronal	64.7 ± 35.2
Others	22.9 ± 16.1

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