CRANIOSYNOSTOSIS

Surgical Specialty:	Plastics and Craniofacial Surgery
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Background:

- General Considerations
    - Craniosynostosis occurs as a consequence of premature closure of one or more cranial suture(s), resulting in asymmetric growth of the cranial vault.
    - Simple craniosynostosis is more common, seen in 80% of patients, generally involves only one suture, and is not associated with an underlying genetic syndrome[1].
    - Complex craniosynostosis often involves multiple sutures and is associated with an underlying genetic syndrome.
    - Management of craniosynostosis is largely surgical, with the age of surgical correction generally ranging from 3 to 18 months of age, depending on the suture involved and surgical technique.
- Patient Considerations
    - If left uncorrected, children are at risk of developing raised intracranial pressure (ICP), cognitive impairment, and developmental delay.
        - 40-70% of syndromic craniosynostosis may present with increased ICP, due to a variety of factors including hydrocephalus and abnormalities in cerebral venous drainage[2].
    - Surgical correction is performed on an elective or time-sensitive basis, weighing the risks and benefits of early versus delayed intervention[1].
        - Benefits of earlier surgical intervention include softer bone leading to increased malleability, and facilitation of ongoing brain growth in a reconstructed cranial vault.
- Associated Comorbidities/Syndromes
    - Three autosomal dominant syndromes are classically associated with complex craniosynostosis[1].
    - Apert Syndrome
        - Associated with maxillary hypoplasia, cleft palate, and complex syndactyly.
        - Difficulties with airway management and intravenous access should be anticipated.
        - Preoperative echocardiography may be considered, as approximately 10% of children are born with congenital heart disease.

- Crouzon Syndrome
  - Associated with midface hypoplasia, upper airway obstruction, and cervical spine abnormalities.
  - Difficult ventilation and intubation should be anticipated, and an evaluation of the cervical spine should be completed prior to surgery.
- Pfeiffer Syndrome
  - Associated with varying degrees of midface hypoplasia, broad and medially deviated thumbs, and significant proptosis.
  - Difficult airway is less common than in Crouzon or Apert syndrome, but raised intracranial pressure may be more common.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Identification of an underlying genetic syndrome and any additional comorbidities, such as increased intracranial pressure, hydrocephalus, and history of prematurity, will influence intra-operative management and post-operative disposition.
  - Baseline hemoglobin and hematocrit allow for an estimation of allowable blood loss, while a review of the chemistry panel may indicate pre-existing acid/base disturbances secondary to hypoventilation and obstructive or central sleep apnea.
  - If available, a review of coagulation studies may reveal underlying abnormalities, guiding blood product replacement, especially in medically complex patients.
  - Children undergoing craniectomy for open anterior/posterior vault reconstruction should have a current type and screen with blood products immediately available.
  - A pre-operative echocardiogram to evaluate for congenital heart disease may be appropriate in syndromic craniosynostosis, especially in children with Apert or Pfeiffer syndrome.
  - Discussions with family should include the possibility of blood product transfusion, pediatric intensive care unit (PICU) admission, postoperative intubation, and mechanical ventilation.
- Specific or Unique Room Set-up Requirements
  - Airway
    - Difficult airway equipment should be immediately available in complex or syndromic craniosynostosis.
    - Orotracheal intubation is generally appropriate, sometimes with the use of RAE preformed endotracheal tube, depending on the surgical approach.
    - Nasotracheal intubation may also be considered for greater stability, particularly if the patient will be in the prone position.
    - A skilled assistant should be available during induction to assist with airway management.
    - Prior to securing the endotracheal tube, confirming appropriate tube position with the head both flexed and extended will reduce the risk of unintentional mainstem intubation (with flexion) or extubation (with extension) required for surgical positioning.
  - Drugs/Infusions
    - Institutional practice varies with regard to the maintenance of anesthesia, but a strategy optimizing cerebral perfusion in the setting of potentially increased intracranial pressure is paramount.
      - Options include total intravenous anesthesia versus balanced anesthetic with volatile agent and opioid infusion.

        - Anti-fibrinolytics, such as tranexamic acid, should be administered as part of a strategy to minimize the need for transfusion[3].
        - Medications to treat increased ICP, such as 3% hypertonic saline or mannitol, should be immediately available.
        - A vasoactive infusion suitable for peripheral venous administration, such as norepinephrine or phenylephrine, should be available.
    - Monitors
        - In addition to standard American Society of Anesthesiologists monitors, invasive arterial monitoring is generally indicated for more extensive cranial vault surgeries.
        - Central venous pressure monitoring may be considered to guide fluid administration and intravascular volume resuscitation, although evidence for its use or its effect on improving outcomes is lacking.
        - A Foley catheter should be placed for patients undergoing vault reconstruction with the potential for significant fluid shifts.
        - The use of a precordial Doppler may be helpful in the detection of venous air embolism for high-risk patients.
    - Blood Availability
        - An active type and cross with immediate blood availability is required for open cranial vault surgeries.
        - Institutional practice varies with regard to less invasive approaches, but the potential for rapid and significant blood loss should always be considered during any type of craniosynostosis surgery.
    - PICU Bed Availability
        - PICU admission should be anticipated for patients with complex medical comorbidities, and those undergoing anterior, posterior, and total vault reconstructions.
        - Post anesthesia care unit disposition is generally appropriate for minimally invasive approaches.

Intraoperative Considerations:

- General
    - Difficulty with airway management should be anticipated in complex craniosynostosis patients with an associated genetic syndrome.
    - Large bore intravenous access, appropriate for the child's age and weight, must be obtained.
    - Techniques to reduce bleeding and the need for transfusion should be employed in patients undergoing surgical vault reconstruction.
- Induction
    - Premedication may be appropriate for otherwise healthy patients to facilitate separation from caregiver but should be administered cautiously in patients with pre-operative increased intracranial pressure.
    - An inhalational induction with the placement of intravenous access following induction is usually appropriate for most infants and children.
    - If there is a concern for difficult airway management, an assessment of the benefits of maintaining spontaneous ventilation versus the potential benefits of neuromuscular relaxation should be considered.
- Positioning
    - Positioning varies with surgical approach and affected cranial suture(s).

- Anterior cranial vault/frontal orbital advancement may be performed in the supine position with the bed rotated 90 or 180 degrees from the anesthesia workstation.
  - Posterior cranial vault may be performed in the prone or sphinx position, with the bed also rotated away from the anesthesia workstation.
  - Positioning may require significant head flexion or extension.
  - Ensuring adequate padding of pressure points, and avoiding excessive flexion/extension of neck is critical to prevent post-operative neurological deficit.
- Maintenance
  - Several techniques are appropriate, with the general goals of maintaining adequate cerebral perfusion, minimizing brain bulk during exposure, ensuring adequate analgesia, and facilitating emergence and extubation in the operating room.
- Hemodynamic/Physiologic goals
  - In patients with elevated intracranial pressure, avoiding hypotension and maintaining adequate cerebral perfusion pressure is critical.
  - During exposure of dura, mild hyperventilation and reverse Trendelenburg position may be requested by the surgical team in order to reduce brain bulk and facilitate surgical exposure.
    - Care should be taken to avoid excessive hyperventilation and hypocarbia, which may compromise cerebral perfusion.
  - Strategies for maintenance of normothermia should be employed, including warming the operating room, use of a forced air warmer, and the use of a blood/fluid warmer.
- Surgical Considerations
  - Correction of sagittal craniosynostosis generally involves extended strip craniectomy or total cranial vault remodeling, depending on the age of diagnosis.
  - Anterior cranial vault remodeling and fronto-orbital advancement are performed in children with metopic and coronal craniosynostosis.
  - Posterior cranial vault remodeling is performed in cases of bi-coronal or lambdoidal craniosynostosis.
  - Minimally invasive techniques include spring-assisted cranioplasty and endoscopic suture release and are associated with a lower risk of bleeding and transfusion, with a shorter hospital length of stay[1,4].
    - Minimally invasive approaches rely on subsequent brain and skull growth and are therefore performed at 3-6 months of age.
    - During spring-assisted cranioplasty, a sagittal strip craniectomy is done, with the placement of springs across the defect. These springs are removed during a subsequent procedure at a later date.
    - Endoscopic suture release is usually performed in conjunction with postoperative helmet molding.
- Emergence/Disposition
  - For either an open surgical approach or a less invasive technique, emergence and extubation in the operating room are generally attempted, although this may vary depending on institutional practice.
  - Significant intraoperative fluid shifts, particularly in the prone position, may also affect the decision of whether or not to attempt extubation in the OR.
  - Disposition to PACU versus PICU will vary, depending on institutional practice, surgical approach, and patient comorbidities.
- Post-op Care
  - Achieving adequate postoperative analgesia may be challenging while balancing the risk of oversedation and hypoventilation.

    - Institutional practice may vary, but generally, intravenous acetaminophen and an as-needed intravenous opioid are appropriate until a child is able to tolerate PO medication.
  - Post-operative hyponatremia may occur in up to 30% of patients following cranial vault remodeling, associated with pre-operative increased ICP and greater volume of intra-operative blood loss[5].
    - Avoid administration of hyponatremic intravenous fluids.

Case-Specific Complications/Pitfalls

- Hypothermia
  - Hypothermia may occur quickly in younger infants and children undergoing craniectomy for vault reconstruction, and this can be very difficult to correct.
  - An esophageal or rectal temperature probe will better estimate core temperature than skin or axillary site.
  - Ensuring appropriate surgical draping will help prevent soakage of forced air warming blanket with surgical prep and irrigation fluid.
- Major Hemorrhage
  - Estimation of blood loss during cranial vault remodeling is often difficult, due to insidious bleeding in the surgical field and drapes, especially during the scalp dissection portion of the procedure.
    - Therefore, close communication with the surgical team regarding bleeding is critical.
  - Younger infants and children are at especially high risk of intraoperative hypothermia, and this may contribute to ongoing coagulopathy.
  - The availability of point-of-care testing allows for rapid correction of associated electrolyte and acid-base disturbances as consequences of bleeding, fluid shifts, and large-volume resuscitation.
  - Viscoelastic testing may be helpful in identifying the etiology of coagulopathy, guiding blood product choice during resuscitation.
- Venous Air Embolism
  - Venous air embolism may occur if dural sinuses or other venous sinusoids are exposed and violated during dissection.
  - Although the incidence of venous air embolism in craniosynostosis repair has been estimated to be as high as 80%, most are clinically insignificant[6].
  - Early detection and communication with the surgical team are paramount in guiding the immediate management of hemodynamically significant venous air embolism, which can rapidly progress to obstructive shock and cardiovascular collapse.
  - Clinical manifestations of a significant venous air embolism may include a decrease or loss of end-tidal CO_2 trace, an increase in the fraction of inspired nitrogen, arrhythmia, and hypotension.
  - Management includes immediate notification to the surgical team, application of 100% oxygen, flooding the surgical field with saline, and lowering the head to a level below the heart.
    - Calling for assistance and resuscitation equipment is critical, as a large venous air embolism may rapidly progress to cardiac arrest.
    - If an appropriately positioned central venous catheter has been placed, this may be used to attempt aspiration of venous air, although in practice this may be challenging.
    - Use of a cognitive aid should be employed to guide management in the case of an intraoperative crisis or emergency

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