Pt scheduled for Primary Craniosynostosis Repair

Schedule Craniofacial and PASS Clinic Visit 1 – 2 weeks prior to surgery

Craniofacial Clinic visit includes:
- Review history and complete physical assessment
- Patient education on procedure and recovery
- Information regarding directed donor blood
- Surgical consents signed and scanned into EPIC
- Patient Road Map provided
- Determine need for additional consults

PASS Clinic visit includes:
- Review history and complete physical assessment
- Patient education on procedure and recovery
- Information regarding directed donor blood
- Surgical consents signed and scanned into EPIC
- Patient Road Map provided
- Determine need for additional consults

Preoperative Assessment in the PASS Clinic includes:
- Review history and complete physical assessment
- Assess for difficult airway
- Confirm with blood bank that blood products are available
- Determine need for additional consults
- Type and Cross obtained if patient has been transfused in the last three months, a type and cross must be completed within three days of surgery.

Preoperative ENT consult

Additional Consults Needed?

Preoperative Blood Orders:
- 2 units RBCs for patients >10 kg
- 1 unit of RBCs for patients <10 kg
- Split unit if patient <5 kg
- 10 ml/kg of fresh frozen plasma (FFP)

Laboratory Assessments:
- Additional routine preoperative labs should not be completed unless patient age <6 months and/or warranted by clinical history (e.g. cardiopulmonary disease, prematurity and postmenstrual age <60 weeks, family history of bleeding disorder, syndromic end organ disease, unknown pertinent aspects of clinical history).
- Type and Cross:
  - If patient has been transfused in the last three months, a type and cross must be completed within three days of surgery.

Difficult airway assessed?

Yes

Preoperative ENT consult

No

Clinical history warranting additional preoperative labs?

Yes

Order labs based upon specified need noted in clinical history

No

Additional Consults Needed?

Consults coordinated and ordered in EPIC

May need 2nd PASS Clinic visit

Continue to Day of Surgery Algorithm

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Quality and Outcomes Management, Texas Children’s Hospital
Day of Surgery Management of Patients Receiving Primary Craniosynostosis Repair

Evidence-Informed Pathway

Preoperative Huddle
Topics of discussion
- Positioning
- Postoperative disposition (Request PICU bed if needed)
- Challenges Anticipated

Anesthesia induction
Surgical Antibiotic Prophylaxis
Baseline labs (Hgb, Platelet, INR, Fibrinogen)

Verify blood product availability
Retrieve Blood Bank Lab STAT Pack

Blood Product Administration
Blood products should be administered in the following situations:
1. Once two lab values are out of range OR one abnormal lab value and active bleeding.
2. Hemodynamic instability as determined by the operative team.

Directed Donor Blood
For patients with directed donor blood, give whole blood at 20 ml/kg then administer platelets at 5 ml/kg

Blood Transfusions
Platelets <100
INR >1.5
Fibrinogen <150
Hemoglobin <7

Administer 5 - 10 ml/kg Platelets
Administer 10 ml/kg FFP
Administer Cryoprecipitate
Administer PRBCs 10 ml/kg

Repeat labs in 30 - 60 minutes after transfusion complete

Emergence
Transfer to PACU
Check labs in recovery room

Postoperative Monitoring and Patient Disposition
Most patients should be monitored in the acute care setting. Patients that meet the following criteria may need intensive care monitoring:
- Hemodynamic instability assessed after surgery in the recovery room
- Continued transfusion requirement
- Syrnosti
- Oxygen requirement
- Complex repair
- Cardiopulmonary disease
- Prematurity and postmenstrual age less than 60 weeks
- End organ disease
- Family history of bleeding disorder

Surgery Cancellation Criteria
- Active upper respiratory infection
- NPO violations
- Active infections
- Fever within the last 24 hours
- Emesis within the last 24 hours

Prerenal Fluid Replacement
- Lactated Ringer's or Plasmalyte should be used for intraoperative fluid replacement.
- Albumin should be considered for patients with hypotension, hypovolemia and/or hemodynamic instability (e.g. pulse pressure variation of 10-12% on arterial line.)

Notify pathologist that labs are being directed donor blood

Directed Donor Blood

Give 20 ml/kg RBCs then administer FFP 10 ml/kg and Platelets 5 ml/kg
Labs 30 – 60 minutes after transfusion complete

Does the patient meet criteria for intensive care monitoring?

Transfer to ICU

Transfer to Acute Care Floor
Critical Points of Evidence

Evidence Supports

- Post-operative monitoring for patients <2 years of age receiving a primary repair for non-syndromic craniosynostosis should be provided in the acute care setting and include continuous cardiac and pulse oximetry monitoring, hematocrit level within 12-24 hours of postoperative care based on procedure type, and monitoring of drain output every four hours. Patients meeting the requirements below should be monitored in the intensive care unit postoperatively.\(^{(1-6)}\) — Strong recommendation, Low quality evidence
  - Hemodynamic instability assessed after surgery in the recovery room
  - Continued transfusion requirement
  - Syndromic
  - Oxygen requirement
  - Complex repair
  - Cardiopulmonary disease
  - Prematurity and postmenstrual age less than 60 weeks
  - End organ disease
  - Family history of bleeding disorder

- Lactated ringer’s or Plasmalyte should be used for intraoperative fluid replacement. Albumin should be considered for patients with hypotension, hypovolemia and/or hemodynamic instability (e.g. pulse pressure variation 10-12% on arterial line)\(^{(7-13)}\) — Strong recommendation, moderate quality evidence

- The use of a Goal Directed Transfusion Pathway should be employed to decrease blood product transfusion in patients with craniosynostosis repair.\(^{(13-22)}\) — Strong recommendation, low quality evidence

- A type and cross should be obtained on patients scheduled for craniosynostosis repair. Additional routine preoperative labs should not be completed unless patient age <6 months and/or warranted by clinical history (e.g. cardiopulmonary disease, prematurity and postmenstrual age less than 60 weeks, family history of bleeding disorder, syndromic, end organ disease, unknown pertinent aspects of clinical history).\(^{(23-31)}\) — Strong recommendation, very low quality evidence

- An alternative transfusion strategy of 2:1 ratio of red blood cells (RBCs) and fresh frozen plasm (FFP) should be administered to perioperative craniosynostosis patients requiring blood products. (For every 20 mL/kg of RBCs, a dose of 10 ml/kg of FFP and 5 ml/kg of platelets should be given.) For patients with directed donor blood, whole blood should be administered and once a transfusion amount of 20 ml/kg is reached give 5 ml/kg of platelets.\(^{(32-37)}\) — Strong recommendation, low quality evidence

Evidence Lacking/Inconclusive

- Transfusions of FFP, Platelets, Cryoprecipitate and/or Fibrinogen should be based upon abnormal lab values and active bleeding. Blood product administration should be administered as noted below once two laboratory values are abnormal or there is active bleeding with one abnormal lab value.\(^{(36-38)}\) — Consensus recommendation
  - Platelets <100 then give 5 – 10 ml/kg Platelets
  - INR >1.5 give 10 ml/kg FFP
  - Fibrinogen < 150 give 1 unit/10 kg Cryoprecipitate

Remarks: Laboratory values should be rechecked after blood product administration to evaluate if additional blood product is necessary.

- The use of cell saver to decrease the rate of transfusion\(^{(36,39-43)}\) — Unable to make a recommendation.
  Remarks: More information is needed to determine if cell saver is an appropriate intervention for use in this population as some hemostatic agents used during surgery may be incompatible with the use of this equipment.

Evidence Against

- Erythropoietin and iron should not be used to decrease the rate of transfusion in preoperative craniosynostosis patients.\(^{(6,36,37,43-47)}\) — Strong recommendation, low quality evidence

- Preoperative vitamin K should not be used to decrease the rate of transfusion in perioperative craniosynostosis patients.\(^{(48)}\) — Strong recommendation, low quality evidence

- Controlled hypotension should not be used to decrease the rate of transfusion in perioperative patients receiving craniosynostosis patients.\(^{(56,49)}\) — Strong recommendation, moderate quality evidence

- Acute normovolemic hemodilution should not be routinely used to decrease the rate of transfusion in patients receiving perioperative craniosynostosis patients.\(^{(56,50)}\) — Strong recommendation, low quality evidence


Clinical Standards Preparation
This clinical standard was prepared by the Evidence-Based Outcomes Center (EBOC) team in collaboration with content experts at Texas Children's Hospital. Development of this clinical standard supports the TCH Quality and Patient Safety Program initiative to promote clinical standards and outcomes that build a culture of quality and safety within the organization.

Craniosynostosis Evidence Informed Pathway Content Expert Team
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EBOC Team
Andrea Jackson, MBA, RN, Evidence-Based Practice Specialist
Charles Macias, MD, MPH, Director

Development Process
This clinical standard was developed using the process outlined in the EBOC Manual. The literature appraisal documents the following steps:
1. Review Preparation
   - PICO questions established
   - Evidence search confirmed with content experts
2. Review of Existing Internal and External Guidelines
   - Parameters of Care for Craniosynostosis, National Foundation of Facial Reconstructive Surgery
   - Craniosynostosis, Seattle Children's Hospital
   - Guidelines and recommendations from the American Society of Anesthesiologists
   - Task Force on Preanesthesia Evaluation, Practice Advisory for Preanesthesia Evaluation, Preoperative tests (update)
   - Routine preoperative tests for elective surgery, National Institute for Health and Care Excellence; Benefits and Harms of Routine Preoperative Testing: Comparative Effectiveness, Agency of Healthcare Research and Quality
   - Blood Transfusions, National Institute of Clinical Excellence
   - National Blood Authority of Australia; Platelet Transfusion: A Clinical Guideline from the AABB
3. Literature Review of Relevant Evidence
   - Search: PubMed, Cochrane Collaboration, CINAHL, Google Scholar
   - 4. Critically Analyze the Evidence
   - 10 randomized controlled trials, and 22 nonrandomized studies, as applicable
5. Summarize the Evidence
   - Materials used in the development of the guideline, evidence summary, and order sets are maintained in a craniosynostosis repair evidence-based review manual within EBOC.

Evaluating the Quality of the Evidence
Published clinical guidelines were evaluated for this review using the AGREE II criteria. The summary of these guidelines are included in the literature appraisal. AGREE II criteria evaluate Guideline Scope and Purpose, Stakeholder Involvement, Rigor of Development, Clarity and Presentation, Applicability, and Editorial Independence using a 4-point Likert scale. The higher the score, the more comprehensive the guideline.

This clinical standard specifically summarizes the evidence in support of or against specific interventions and identifies where evidence is lacking/inconclusive. The following categories describe how research findings provide support for treatment interventions. “Evidence Supports” provides clear evidence that the benefits of the intervention exceed harm.

“Evidence Against” provides evidence that the intervention is likely to be ineffective or that it is harmful.

“Evidence Lacking/Inconclusive” indicates there is currently insufficient data to support or refute a specific intervention.

The GRADE criteria were utilized to evaluate the body of evidence used to make practice recommendations. The table below defines how the quality of the evidence is rated and how a strong versus weak recommendation is established. The literature appraisal reflects the critical points of evidence.

<table>
<thead>
<tr>
<th>Quality</th>
<th>Type of Evidence</th>
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<tbody>
<tr>
<td>High</td>
<td>Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Moderate</td>
<td>Evidence from RCTs with important limitations (e.g., inconsistent results, methodological flaws, indirect evidence, or imprecise results) or unusually strong evidence from unbiased observational studies</td>
</tr>
<tr>
<td>Low</td>
<td>Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence</td>
</tr>
<tr>
<td>Very Low</td>
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Recommendations
Practice recommendations were directed by the existing evidence and consensus amongst the content experts. Patient and family preferences were included when possible. The Content Expert Team and EBOC team remain aware of the controversies craniosynostosis repair for children. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

Approval Process
Clinical standards are reviewed and approved by hospital committees as deemed appropriate for its intended use. Clinical standards are reviewed as necessary within EBOC at Texas Children’s Hospital. Content Expert Teams are involved with every review and update.

Disclaimer
Practice recommendations are based upon the evidence available at the time the guideline was developed. Clinical standards (guidelines, summaries, or pathways) do not set out the standard of care, and are not intended to be used to dictate a course of care. Each physician/practitioner must use his or her independent judgment in the management of any specific patient and is responsible, in consultation with the patient and/or the patient family, to make the ultimate judgment regarding care.

Version History
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<th>Date</th>
<th>Action</th>
<th>Comments</th>
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<tr>
<td>Dec 2017</td>
<td>Pathway Development</td>
<td>TXA Dosing Change</td>
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<tr>
<td>Feb 2018</td>
<td>Revision</td>
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We appreciate the feedback from the EBOC Team and EBOC team remain aware of the controversies craniosynostosis repair for children. When evidence is lacking, options in care are provided in the clinical standard and the accompanying order sets (if applicable).

Evaluation of the Quality of the Evidence
- Evidence is evaluated using the AGREE II criteria, which assess the scope and purpose, stakeholder involvement, rigor of development, clarity and presentation, applicability, and editorial independence.

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