Preoperative Management of Patients Receiving Primary Craniosynostosis Repair
Evidence-Informed Pathway

**Pt scheduled for Primary Craniosynostosis Repair**

STAT Hematology referral no less than 3 weeks before surgery for "pre-surgical rhEPO planning"

**Hematology Clinic visit includes:**
- Review personal and family history of bleeding
- Determine if risk of greater blood loss is likely
- Discuss rhEPO protocol with family

Patient is a candidate and family consents to pre-surgical rhEPO protocol?

Yes

Utilize Beacon Perioperative NUS Erythropoietin plan

No

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

**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*
- Type and Cross obtained* (*Check transfusion history to determine if type and cross needs to be scheduled closer to surgery)

**Preoperative Blood Orders**
- 2 units RBCs for patients >10kg
- 1 unit of RBCs for patients <10kg
- Split Unit if patient <5kg
- 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 warrants additional preoperative labs?

Yes → Order labs based upon specified need noted in clinical history

No

Additional Consults Needed?

Yes → Consults coordinated and ordered in Epic
May need 2nd PASS Clinic visit

No

Continue to Day of Surgery Algorithm

© Evidence-Based Outcomes Center
Quality and Outcomes Management, Texas Children’s Hospital
**Day of Surgery Management of Patients Receiving Primary Craniosynostosis Repair**

**Evidence-Informed Pathway**

**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 and administer platelets at 5 ml/kg.

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

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

**Intraoperative 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 10-20% on arterial line).

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

**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
  - Syndromic
  - Oxygen requirement
  - Complex repair
  - Cardiopulmonary disease
  - Prematurity and postmenstrual age less than 60 weeks
  - End organ disease
  - Family history of bleeding disorder

**Transfer to ICU**
- Does the patient meet criteria for intensive care monitoring?

**Transfer to Acute Care Floor**

---

© Evidence-Based Outcomes Center
Quality and Outcomes Management, Texas Children’s Hospital
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.\(^{14}\) – 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
- Acute normovolemic hemodilution should not be routinely used to decrease the rate of transfusion in perioperative craniosynostosis patients.\(^{36,51}\) – Strong recommendation, Low quality evidence
- Controlled hypotension should not be used to decrease the rate of transfusion in perioperative patients receiving craniosynostosis patients.\(^{36,51}\) – Strong recommendation, moderate quality evidence
- Preoperative vitamin K should not be used to decrease the rate of transfusion in perioperative craniosynostosis patients.\(^{36,51}\) – 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
- 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.\(^{36,37,39-43}\) – Unable to make a recommendation.
  - Recombinant human erythropoietin (rhEPO) has been used to reduce the need for blood transfusion in pediatric open heart surgery, spinal surgery and craniofacial surgeries. Studies of children undergoing craniosynostosis surgery have successfully demonstrated that it reduces the rate of transfusion in these patients.\(^{43-49}\)

Evidence Against

- Preoperative vitamin K should not be used to decrease the rate of transfusion in perioperative craniosynostosis patients.\(^{50}\) – Strong recommendation, low quality evidence
- Controlled hypotension should not be used to decrease the rate of transfusion in perioperative patients receiving craniosynostosis patients.\(^{36,51}\) – 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.\(^{36,52}\) – Strong recommendation, low quality evidence
References

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
Rahul Baijal, MD, Anesthesia
David Bauer, MD, Neurosurgery
Anthony Bodnar, RN, Acute Care
Robert Dempsey, MD, Plastic Surgery
Rebecca Hanson, RN, Perioperative Services
Lisa Hensch, MD, Transfusion Medicine
Helena Karlberg, MD, Anesthesia
YiChen Lai, MD, Critical Care
Valentina Briceno Marmol, RN, Surgery
Laura Monson, MD, Plastic Surgery
JoWinsyl Montejo, RN, Operating Room
Julie Nicholson, RN, Anesthesia
Olutoyin Olutoye, MD, Anesthesia
Sandra Taylor, RN, Acute Care
Jun Teruya, MD, Transfusion Medicine
Alexander Wasserman, Anesthesia

EBOC Team
Andrea Jackson, MBA, RN, Evidence Based Practice Specialist
Binita Patel, MD, Chief Medical Quality Officer

Additional EBOC Support
Betsy Lewis, MSN, RN, CNL, Evidence-Based Practice Specialist
Sheesha Porter, MSN, RN, Evidence-Based Practice Specialist
Anne Dykes, MSN, RN, ACNS-BC, Manager

No relevant financial or intellectual conflicts to report.

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
3. Literature Review of Relevant Evidence
   - Searched: 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 clear evidence that the intervention is likely to be ineffective or that it is harmful.
“Evidence Lacking/Inconclusive” indicates there is currently insufficient data or inadequate 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>Recommendation</th>
<th>Quality</th>
<th>Type of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>STRONG</td>
<td>Desirable effects clearly outweigh undesirable effects or vice versa</td>
<td></td>
</tr>
<tr>
<td>WEAK</td>
<td>Desirable effects closely balanced with undesirable effects</td>
<td></td>
</tr>
<tr>
<td>High</td>
<td>Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies</td>
<td></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>
<td></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>
<td></td>
</tr>
<tr>
<td>Very Low</td>
<td>Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence</td>
<td></td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dec 2017</td>
<td>Pathway Development</td>
<td></td>
</tr>
<tr>
<td>Feb 2018</td>
<td>Revision</td>
<td>TXA Dosing Change</td>
</tr>
<tr>
<td>Mar 2022</td>
<td>Update</td>
<td>Pre-op algorithm change and rhEPO update</td>
</tr>
</tbody>
</table>

Related Documents

Craniosynostosis Literature Appraisal Document