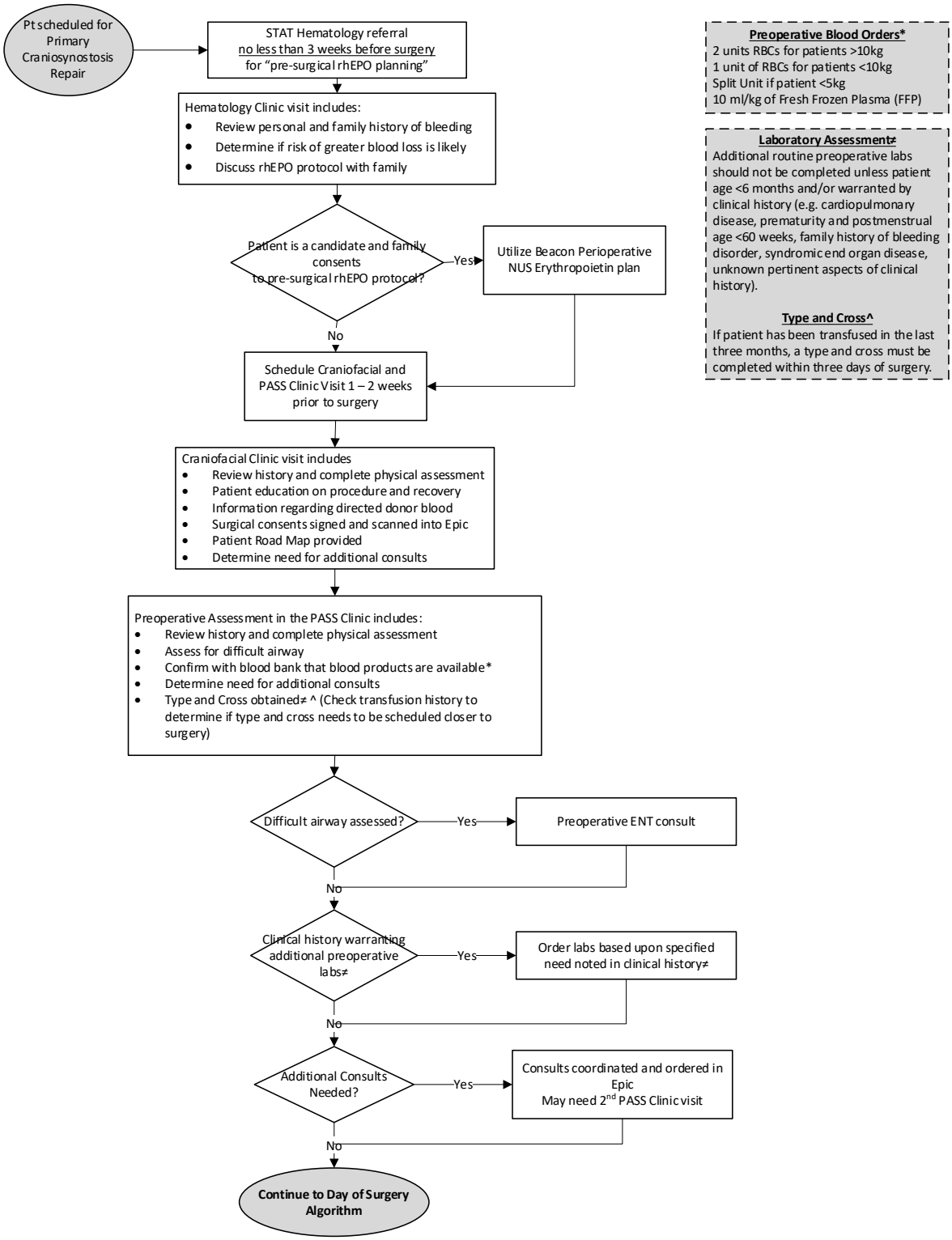
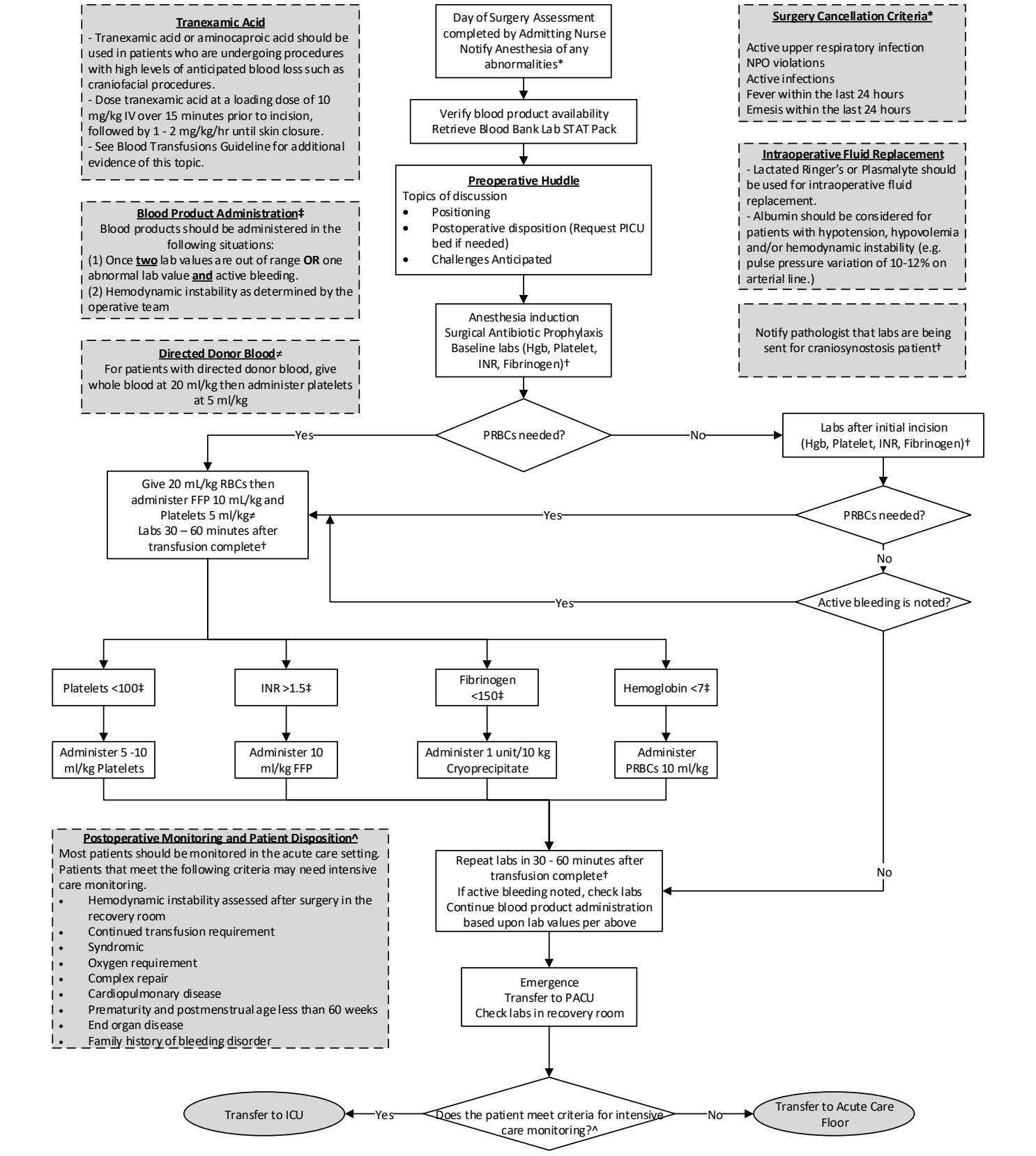
TEXAS CHILDREN'S HOSPITAL
EVIDENCE-BASED OUTCOMES CENTER
Preoperative Management of Patients Receiving Primary Craniosynostosis Repair
Evidence-Informed Pathway



TEXAS CHILDREN'S HOSPITAL
EVIDENCE-BASED OUTCOMES CENTER
Day of Surgery Management of Patients Receiving Primary Craniosynostosis Repair
Evidence-Informed Pathway



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).[4,23-31] – 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.[36,37,39-43] – 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.
- 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]
  Remarks: See S-20210039 Perioperative Erythropoietin & Iron Supplementation for Craniosynostosis Repair TXCH Best Current Practice Standard

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

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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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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
   - Parameters of Care for Craniosynostosis, National Foundation of Facial Reconstruction; Guideline for Care of Patients with the Diagnosis of Craniosynostosis, Working Group of Craniosynostosis; Craniosynostosis, Seattle Children's Hospital; An Updated Report by 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; Patient Blood Management Guidelines: Neonatal & Pediatrics, National Blood Authority of Australia; Platelet Transfusion: A Clinical Guideline from the AABB, AABB
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.

Recommendation	
STRONG	Desirable effects clearly outweigh undesirable effects or vice versa
WEAK	Desirable effects closely balanced with undesirable effects
Quality	**Type of Evidence**
High	Consistent evidence from well-performed RCTs or exceptionally strong evidence from unbiased observational studies
Moderate	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
Low	Evidence for at least 1 critical outcome from observational studies, RCTs with serious flaws or indirect evidence
Very Low	Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence

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

Date	Action	Comments
Dec 2017	Pathway Development	
Feb 2018	Revision	TXA Dosing Change
Mar 2022	Update	Pre-op algorithm change and rhEPO update

Related Documents

Craniosynostosis Literature Appraisal Document