Definition: First described in 1971 as a surgical intervention for patients with tricuspid atresia, the Fontan operation has become the most common operation performed for patients with hypoplastic left heart syndrome or single ventricle. Refinements in operative technique and perioperative management have resulted in improved outcome in this high-risk group of patients. The Fontan is usually the final planned operation in the staged single ventricle palliation pathway. The procedure involves the placement of a conduit from the right atrium to the main pulmonary artery which bypasses the hypoplastic right ventricle and results in a total cavopulmonary connection. The Fontan operation is typically performed between two and five years of age, depending on the specific clinical presentation and surgical history. Following the Fontan, appropriate postoperative management is essential for preventing postoperative mortality, reducing postoperative complications and length of stay, and promotion of optimal long term outcomes.

Inclusion Criteria
- Patients 18 years and younger
- Single ventricle

Exclusion Criteria
- Patients over the age of 18 years
- Fontan conversion procedures
- Fontan with concomitant procedures
- Preoperative renal insufficiency/failure
- Failing Fontan

Differential Diagnosis
Other left-sided obstructive lesions where the systemic circulation is dependent on ductal flow

Diagnostic Evaluation
History: Assess for
- A thorough clinical assessment
- Oximetry at rest
- ECG to assess for sinus rhythm
- Chest x-ray

Physical Examination
- Cardiac catheterization (ideal measurements):
  - Common atrial pressure: <12
  - PVR: <4 Woods units
  - TPG: <8
  - BDG/PA: ≤15
  - Oxygen saturations: >75%
- Echocardiography:
  - ≤ Mild atrioventricular valve regurgitation
  - ≤ Mild ventricular dysfunction or systemic EF >45%
  - ≤ Mild aortic insufficiency
- 24-hour Holter:
  - No clinically relevant dysrhythmia

Laboratory Tests
- Serum protein and albumin measurement. If low, increased 1 anti-trypsin clearance in the stool documents the presence of PLE (protein losing enteropathy).

Critical Points of Evidence*

Evidence Supports
- Administration of Lasix per the following postoperative regimen: 24 hours: IV Lasix 1 mg/kg every 12 hours (initial dose not to exceed 20 mg/dose); 48 hours: IV Lasix 1 mg/kg every 8 hours; 72 hours: PO Lasix 1.5 mg/kg every 8 hours (do not exceed 20 mg/dose). (2-6) – Strong recommendation, low quality evidence
- Initiation of ACE inhibitor for patients with significant hypertension or AV valve regurgitation once taking PO if patient does not have concern for renal injury. (2,3,4-6) – Strong recommendation, low quality evidence
- Administration of acetylsalicylic acid 81 mg for antithrombotic therapy beginning on postoperative day 2. (4,8-10) – Strong recommendation, low quality evidence
- Initiation of fluid restriction at 50% maintenance then up to a maximum of 80% maintenance once the patient begins taking liquids by mouth. (5,6,9) – Strong recommendation, low quality evidence
- Administration of supplemental oxygen for all patients at a minimum of 0.5 L/min via nasal cannula through postoperative day 3 when patient is asleep or in bed (OK to discontinue with ambulation: DO NOT limit out of bed or walking). Continue after postoperative day 3 if oxygen saturations are <94%. For patients with fenestrated Fontans, documented AV malformations and/or veno-venous collaterals, patient care should be individualized. (6,5,8) – Strong recommendation, low quality evidence
- If chylous drainage is suspected or confirmed (≥5 cc/kg drainage), test for the presence of triglycerides in drainage by 48 to 72 hours. If drainage is positive for fluid triglycerides (pleural fluid triglycerides >110 + lymphocyte count = chylous effusion), initiate minimal fat diet. (5,11,12) – Strong recommendation, low quality evidence
- Management of postoperative pain utilizing multimodal analgesia. Patients should be on a morphine PCA on admission to the CVICU and for 48 hours and around-the-clock acetaminophen (IV/PO) for 48 hrs. The day following surgery patients should be started on ketorolac, 6-8 doses, as long as their platelets are >100, creatinine <0.6, and they don’t have bloody chest tube output. Patients should initially be on dexametomidine for approximately 24 hours unless they are paced or their HR is <60. When being transferred to the floor, the PCA Morphine can transition to PRN morphine. When taking PO, they should be transition from PCA morphine or scheduled morphine to scheduled Hycet (with PRN morphine). (13-18) – Strong recommendation, low quality evidence
- Encourage highest degree of mobility within 18 hours postoperatively or the morning after surgery; order physical therapy consult on admission; encourage highest degree of mobility within 18 hours or morning of surgery, with an ambulation goal of a minimum of three times a day; to initiate respiratory physiotherapy using age-appropriate device (e.g., incentive spirometry, bubbles, pinwheel) within 18 hours of surgery or morning after procedure, every hour while awake, starting on postoperative day 1. (19-22) – Strong recommendation, low quality evidence

**Evidence Against**
- Postoperative use of ACE inhibitors for all Fontan patients. (2,3,4-6) – Strong recommendation, low quality evidence

**Evidence Lacking/Inconclusive**
- Postoperative use of ace inhibitors for all Fontan patients. (2,3,4-6) –Unable to make a recommendation
- The use of vasopressin for the reduction of chest drainage following the Fontan procedure. –Unable to make a recommendation

*NOTE: The references cited represent the entire body of evidence reviewed to make each recommendation.

### Condition-Specific Elements of Clinical Management

<table>
<thead>
<tr>
<th>Discharge Criteria</th>
<th>Measures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Drains: chest tubes removed</td>
<td>Bed availability</td>
</tr>
<tr>
<td>Diuretics: 1) All enteral, 2) Lasix ≤3 doses/day, 3) Diuril ≤2 doses/day</td>
<td>Process</td>
</tr>
<tr>
<td>Mobilization: adequate mobilization</td>
<td>Postoperative additional procedures</td>
</tr>
<tr>
<td>Pain: pain well controlled by PO medicines (PRN or scheduled morphine or Dilaudid no longer needed)</td>
<td>Outcome</td>
</tr>
<tr>
<td>Feeds: tolerating regular diet</td>
<td>Mortality</td>
</tr>
</tbody>
</table>
| Elimination: normal bowel movements | Morbidity (infection, thromboembolic events, etc…)
| Discharge teaching complete | Length of stay |
| Discharge ECHO, EKG and CXR performed | Length of stay in CVICU |
| Follow up appointments made with 1) Cardiology and 2) CV surgery | Duration of chest tubes |
| Follow-Up Care | Total chest tube drainage |
| Follow-up clinic date and time confirmed with surgical PA prior to discharge | Number of days chest tube drainage |
| Families instructed to call if new: redness, swelling, pain, discharge around drain, edema, abrupt change in amount or character of drainage, cough, fever, cyanosis, and/or SOB | Readmissions |
| Patient/family satisfaction | Quality of life |
| Functional health status | Cost |
| Quality of life | |

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## FONTAN MANAGEMENT TABLE

<table>
<thead>
<tr>
<th>POD</th>
<th>CV Meds</th>
<th>Diuretics</th>
<th>Fluid</th>
<th>Nutrition</th>
<th>Respiratory</th>
<th>Chest Tubes</th>
<th>Mobilization</th>
<th>Pain Mx</th>
<th>Disposition</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Inotropes</td>
<td>None</td>
<td>Crystalloid / blood product replacement</td>
<td>NPO then clears if stable</td>
<td>OR extubation vs. &lt;6hrs ICU&lt;sup&gt;5&lt;/sup&gt;</td>
<td>Head of bed to 30 degrees</td>
<td>• PCA</td>
<td>• Ofirmev</td>
<td>CVICU</td>
</tr>
<tr>
<td>1</td>
<td>Wean Inotropes</td>
<td>• IV furosemide&lt;sup&gt;3&lt;/sup&gt; (1 mg/kg q12 max 20 mg/dose)</td>
<td>Start enteral intake</td>
<td>Min: 0.5L/min&lt;sup&gt;6&lt;/sup&gt;</td>
<td>Mediastinal out&lt;sup&gt;7&lt;/sup&gt;</td>
<td>Encourage highest degree of mobilization</td>
<td>• PCA</td>
<td>• IV acetaminophen</td>
<td>• Consider Toradol&lt;sup&gt;8&lt;/sup&gt;</td>
</tr>
<tr>
<td>2</td>
<td>• Initiate ASA • Consider resuming home meds</td>
<td>• IV furosemide&lt;sup&gt;3&lt;/sup&gt; (1 mg/kg q8 max 20 mg/dose) • PO Diuril</td>
<td>75-80% maintenance</td>
<td>PO encouraged</td>
<td>Min: 0.5L/min&lt;sup&gt;6&lt;/sup&gt;</td>
<td>Mediastinal out&lt;sup&gt;7&lt;/sup&gt; Pleural to bulb</td>
<td>Walk in ICU at least once per shift</td>
<td>• PO opioid +acetaminophen • Consider ketoralac&lt;sup&gt;8&lt;/sup&gt;</td>
<td>Transfer to 15T</td>
</tr>
<tr>
<td>3</td>
<td>• ASA • Consider resuming home meds</td>
<td>• PO furosemide&lt;sup&gt;3&lt;/sup&gt; (1.5 mg/kg q8 max 20 mg/dose) • PO Diuril</td>
<td>75-80% maintenance</td>
<td>Full PO</td>
<td>Min: 0.5L/min&lt;sup&gt;6&lt;/sup&gt;</td>
<td>Pleural to bulb</td>
<td>Walk x3</td>
<td>• PO opioid +acetaminophen • PO NSAID</td>
<td>Transfer to 15T</td>
</tr>
<tr>
<td>4</td>
<td>• ASA</td>
<td>Optimize PO diuretics</td>
<td>75-80% maintenance</td>
<td>Full PO</td>
<td>Stop O&lt;sub&gt;2&lt;/sub&gt; if Sats &gt;94%</td>
<td>Remove CTs if &lt;2cc/kg/d each</td>
<td>Full mobilization as at home (minimum 3x)</td>
<td>• PO NSAID • PRN PO opioid</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>• ASA</td>
<td>Decrease or maintain</td>
<td>75-80% maintenance</td>
<td>Full PO</td>
<td>Stop O&lt;sub&gt;2&lt;/sub&gt; if Sats &gt;94%</td>
<td>Remove CTs if &lt;2cc/kg/d each</td>
<td>Full mobilization as at home (minimum 3x)</td>
<td>• PO NSAID • PRN PO opioid</td>
<td>d/c criteria met?</td>
</tr>
<tr>
<td>6</td>
<td>• ASA</td>
<td>Decrease or maintain</td>
<td>75-80% maintenance</td>
<td>Full PO</td>
<td>Stop O&lt;sub&gt;2&lt;/sub&gt; if Sats &gt;94%</td>
<td>Remove CTs if &lt;2cc/kg/day each</td>
<td>Full mobilization as at home (minimum 3x)</td>
<td>• PO NSAID • PRN PO opioid</td>
<td>d/c criteria met?</td>
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</table>
References


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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.

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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 External Guidelines
   - CVS: ICU Management of HLHS and Single Ventricle Physiology; Management of Postoperative Pain: A Clinical Practice Guideline from the American Pain Society, the American Society of Regional Anesthesia and Pain Medicine, and the American Society of Anesthesiologists’ Committee on Regional Anesthesia, Executive Committee, and Administrative Council
3. Literature Review of Relevant Evidence
   - Searched: PubMed, Cochrane Library, EMBASE
4. Critically Analyze the Evidence
   - 4 meta-analyses, 5 randomized controlled trials, and 12 nonrandomized studies
5. Summarize the Evidence
   - Materials used in the development of the clinical standard, literature appraisal, and any order sets are maintained in a postoperative management of the Fontan patient 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 evidence to support an intervention “Evidence Against” provides evidence against an intervention. “Evidence Lacking/Inconclusive” indicates there is insufficient evidence to support or refute an intervention and no conclusion can be drawn from the evidence. 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>
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<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>
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</table>

<table>
<thead>
<tr>
<th>Quality</th>
<th>Type of Evidence</th>
</tr>
</thead>
<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>
<td>Evidence for at least 1 critical outcome from unsystematic clinical observations or very indirect evidence</td>
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</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 in the postoperative management of children following the Fontan Procedure. 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’s family, to make the ultimate judgment regarding.

Version History
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