Inclusion Criteria
- Neonates with ductal dependent systemic blood flow

Exclusion Criteria
- Isolated coarctation of the aorta
- Obstructive pulmonary venous return
- Cardiogenic shock
- Prematurity (< 35 weeks postmenstrual age)
- Other significant comorbidities
- Sepsis
- Withdrawal of life sustaining therapy

Pre-hospital Care
- IVF: D10W @ 60 mL/kg/day
- NPO
- PGE infusion: consider decreasing if significant apnea
- IV access: in order of preference, not to delay transfer
  a. High UVC above diaphragm
  b. PIV is adequate (consider 2 PIVs for long-term)
  c. Low UVC below diaphragm
- Phone call to operating institution on DOB
- For transport from distance: intubation per ALNW standard
- Transfer to SCH as soon as possible

Assessment
- Admission History and Physical, vitals, NIRS
- Assessment of Qp:Qs
- Admission labs: lytes, CBC (if not already drawn), +/- blood gas and lactate (if in distress). Type and screen on admission
- Chest X-ray
- ECG
- Echo
- Renal ultrasound
- Assessment for operative readiness

Therapy
- Routine neonatal care
- FEN: refer to Feeding Protocol, IVF’s, begin TPN hospital day 2, total fluids: 80-100ml/kg/d
- Resp: if apnea refractory to decreasing PGE start HFNC→CPAP→intubation
- CV: PGE
- Heme: if perfusion is borderline, maintain hct > 40%
- ID: initiate nasal mupirocin on admission
- Care coordination:
  - Discuss with family
  - Discuss with referring cardiologist
  - Discuss with referring neonatologist
  - Conference presentation
  - Pre-operative teaching with ARNP: provide family with PE1545

IV Access
- Prehospital: request that referring hospital place a UVC, but should not delay transport
- Inpatient: arrange for lower extremity PICC line
- If unsuccessful, request lower extremity PICC line placement by IR
- Remove UVC after PICC placement
- Note: in most cases, a UAC is not necessary, but can be placed if indicated

Consultation
- Neonatology (if indicated)
- Cardiology
- Social Work
Ongoing Assessment of Qp:Qs

- Well balanced: SaO2 75-80%, no/mild tachypnea, normal perfusion
- Elevated: SaO2 80-90%, tachypnea, tachycardia, +/- decreased systemic perfusion, +/- WOB
- Elevated with signs of hypoperfusion: increased WOB, decreased systemic perfusion, decreased urine output, lactic acidemia

**Well balanced**
- Continue current management
  - Cerebral/renal NIRS
  - Continue monitoring cardiac output
  - Notify CV surgery

**Elevated**
- Consider intubating patient
  - Control ventilation with muscle relaxation
  - To OR

**Elevated with signs of hypoperfusion**
- Step 1
  - Initiate furosemide - titrate to effect
  - Consider decrease of PGE
- Step 2
  - Non-invasive positive pressure ventilation
  - Consider lactate/ABG
- Step 3
  - Initiate milrinone
  - Consider ECHO
  - Assure CV surgery service is aware of patient status
Clinical Standard Work Pathway
Pre-operative Management of Patients with Ductal Dependent Systemic Blood Flow

July 2016
Prepared by: Kihan Kim and CE Team
Department: Clinical Effectiveness
Sponsors: Mark Del Beccaro MD, Bob Sawin MD, Madlyn Murrey RN MN

Course Goal

• This training module is the result of many hours of work by your colleagues at Seattle Children’s Hospital. Our goal was to create an evidence based guideline that addressed the diagnosis, care, and treatment for children with ductal dependent systemic blood flow.

• The foundation for this work began with a comprehensive review of the literature. From this, specific treatment recommendations were created using the best available evidence. We believe that this pathway represents “state of the art” care.
Introduction to Clinical Standard Work

• Improvement in any process begins with standardization. This module is part of a series whose aim is to standardize the way we care for patients.
• Standardization affords us the ability to monitor outcomes and adjust our processes so that all future patients may benefit.
• As you progress through this module, we will illustrate for you the rationale behind various decision points. More importantly, you will learn where there are “gaps” in the literature. To this end, each pathway is revised whenever compelling new evidence arises.

Learning Objectives

Upon completion of this module, participants will be better able to:

1. Identify patients appropriate for this pathway.
2. Improve clinical assessment of infants with ductal dependent systemic blood flow.
3. Recognize symptoms suggesting pulmonary overcirculation and reduced systemic blood flow.
4. Initiate therapy to modulate pulmonary vascular resistance and enhance systemic blood flow.
Infants with ductal dependent systemic blood flow have left ventricular outflow obstruction:

- Hypoplastic left heart syndrome (HLHS)
- Interrupted aortic arch
- Critical aortic stenosis

Patients with HLHS, and other lesions with similar physiology, are at the highest risk, and are the costliest group among those with congenital heart disease.

No congenital heart defect has undergone a more dramatic change in diagnostic approach, management, and outcomes than hypoplastic left heart syndrome (HLHS).

Outcome data is highly regarded and often synonymous with overall programmatic success.

Tend to have long lengths of stay

- Provides a metric to make measurable improvements that are significant (days not hours)
- Potential for increased risk of iatrogenic harm
**Inclusion Criteria**

Neonates with ductal dependent systemic blood flow

**Exclusion Criteria**

- Isolated Coarctation of the aorta
- Obstructed pulmonary venous return
- Cardiogenic shock
- Prematurity (< 35 weeks post menstrual age)
- Other significant comorbidities
- Sepsis
- Withdrawal of life-sustaining therapy
Pre-operative Management

The goals of pre-operative management include:

- Clinical stabilization
- Complete definition of cardiac anatomy
- Recognition of noncardiac diagnoses
- Preparation for surgical palliation
- Family education

Pre-hospital Care

- Neonates with ductal dependent systemic blood flow require intravenous access for continuous intravenous infusion of prostaglandin E1 (PGE) to maintain ductal patency for adequate systemic blood flow.
- Intravenous access in order of preference:
  - High umbilical venous catheter above diaphragm is preferred
  - Two peripheral intravenous catheters
  - Low umbilical venous catheter
- Prostaglandin E infusion
  - Consider decreasing if significant apnea
- Intravenous fluids
  - D10W @ 60 ml/kg/day
Pre-hospital Care Recommendations

Transfer neonates with complex congenital heart disease to tertiary/quaternary centers as soon as possible. **Expert opinion**

Initial Care at SCH

- Admission history and physical
- Laboratory assessment
  - Electrolytes and CBC (if not already done)
  - Arterial blood gas and lactate (if in distress)
  - Type and screen
- Diagnostic studies
  - Chest X ray
  - Electrocardiogram / Echocardiogram
  - Renal ultrasound – (see next slide)
- Consultation
  - Cardiology, Neonatology if indicated, Social Work
Initial Therapy

- Fluids and nutrition
  - IV fluids or TPN: 80-100 ml/kg/day
  - Refer to Feeding Protocol
- Cardiovascular:
  - PGE 0.01-0.03 mcg/kg/min
- Respiratory:
  - If apneic, decrease PGE
  - If refractory → HFNC → CPAP → Intubation

Initial Therapy (Cont’d)

- Infectious Disease:
  - Initiate nasal mupirocin 3 days prior to anticipated OR date
- Heme:
  - Enroll in infant protocol
  - HCT > 35% is usually adequate
  - If poorly perfused maintain HCT > 40%
- Occupational therapy:
  - Non nutritive oral therapy
Recommendation: Renal Ultrasound

Clinical Question: What is the evidence for or against cranial and/or renal ultrasounds prior to surgery?

Do not perform routine HUS in pre-operative neonates unless indicated. Patients with ductal dependent systemic blood flow should have routine renal ultrasounds prior to surgical intervention.

[่อ่อ่อ Very low quality evidence]
**Intravenous Access**

- Insert high umbilical venous catheter (see pre-hospital care).
- Arrange for lower extremity PICC line.
- If unsuccessful, request lower extremity PICC line placement by IR.
- Remove UVC after PICC insertion.

**Recommendation: Timing of Surgery**

**Clinical Question:** *What is the impact of gestational age on outcomes of Norwood or other congenital heart surgeries?*

Infants beyond 39 weeks post menstrual age are candidates for the Norwood procedure. In special circumstances, infants between 36 and 39 weeks post menstrual age may be considered.

[☆☆☆☆ Low quality] (Jacobs, 2008; Nilsson, 2006)
Assessment of Pulmonary to Systemic Blood Flow Ratio (Qp:Qs)

Neonates with ductal dependent systemic blood flow require continuous intravenous infusion of prostaglandin E1 (PGE) to maintain ductal patency for adequate systemic blood flow.

Pulmonary vascular resistance (PVR) falls following birth and may result in a decrease in systemic perfusion as pulmonary blood flow increases.

• Ensuring adequate systemic perfusion (i.e., balancing the systemic and pulmonary circulations) becomes crucial.
Clinical Assessment of Qp:Qs

Balanced Qp:Qs
- O2 saturation 75-80%
- No or only mild tachypnea
- Normal perfusion

Clinical Assessment of Qp:Qs Balance

Elevated Qp:Qs
(increasing PBF at expense of SBF)
- O2 saturation 80-90%
- Tachypnea +/- work of breathing
- Tachycardia
- Normal systemic perfusion
Management of Qp:Qs Pertubations

Management options include:

- Increase PVR
  - Intubation with hypoventilation
- Increase cardiac output
  - Begin inotropic support (e.g., milrinone)
- Pursue early surgical intervention
Specific Management Considerations

Well balanced Qp : Qs

- Continue current management

Elevated Qp:Qs (without signs of hypoperfusion)

1) Initiate furosemide 1 mg/kg IV q 12 hours
   - Consider decreasing PGE to 0.01 mcg/kg/min

2) Non-invasive positive pressure ventilation
   - Consider frequent surveillance Lactate/ABG

3) Milrinone infusion 0.25 microgram/kg/min
   - Consider echocardiogram and assure CV surgery aware of patient status

Elevated Qp:Qs with signs of hypoperfusion

- Intubate and begin mechanical ventilation.
- Controlling ventilation with muscle relaxation.
- Continue Milrinone infusion.
- To OR.
CICU Neonates with Congenital Heart Disease
PowerPlan

Two phase PowerPlan to help standardize the care of patients admitted to the CICU

1. **Admit phase** – for all patients admitted to CICU
2. **Balancing Qp:Qs Ductal Dependent** – for management of Qp:Qs balance
We used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial, or observational studies. The rating is then adjusted in the following manner:

**Quality ratings are downgraded** if studies:
- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings can be **upgraded** if it is felt that:
- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

**Quality of Evidence:**
- 🌟🌟🌟🌟 High quality
- 🌟🌟🌟 Moderate quality
- 🌟🌟 Low quality
- 🌟🌟🌟 Very low quality

Expert Opinion (E)
Summary

Now that this module has been completed, participants should be better able to:

1. Identify patients appropriate for this pathway.
2. Improve clinical assessment of infants with ductal dependent systemic blood flow.
3. Recognize symptoms suggesting pulmonary overcirculation and reduced systemic blood flow.
4. Initiate therapy to modulate pulmonary vascular resistance and enhance systemic blood flow.

Contact Information

We want your opinions regarding the content of this pathway!

If you have questions or comments please contact ductaldependentsystemicbloodflow@seattlechildrens.org
# Value Analysis

## Value Analysis Tool

<table>
<thead>
<tr>
<th>DIMENSION</th>
<th>CARE OPTION A</th>
<th>CARE OPTION B</th>
<th>PREFERRED OPTION</th>
<th>ASSUMPTIONS MADE</th>
</tr>
</thead>
<tbody>
<tr>
<td>DESCRIPTION OF CARE TREATMENT OPTION</td>
<td>Cranial US on all pre-operative patients</td>
<td>Cranial US only as clinically indicated</td>
<td></td>
<td></td>
</tr>
<tr>
<td>OPERATIONAL FACTORS</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Percent adherence to care (goal 80%)</td>
<td>100%</td>
<td>20%</td>
<td>OPTION B</td>
<td>Team would prefer less tests and disruption to patient care</td>
</tr>
<tr>
<td>Care delivery team effects</td>
<td>More staff resourcing required</td>
<td>Less staff resourcing required</td>
<td>OPTION B</td>
<td></td>
</tr>
<tr>
<td>BENEFITS / HARMs (QUALITY/OUTCOME)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Degree of recovery at discharge</td>
<td>No difference</td>
<td>No difference</td>
<td>NEUTRAL</td>
<td></td>
</tr>
<tr>
<td>Effects on natural history of the disease over equivalent time</td>
<td>No difference</td>
<td>No difference</td>
<td>NEUTRAL</td>
<td></td>
</tr>
<tr>
<td>Potential to cause harm</td>
<td>No difference</td>
<td>No difference</td>
<td>NEUTRAL</td>
<td></td>
</tr>
<tr>
<td>Palatability to patient/family</td>
<td>Requires more interventions to patient</td>
<td>Requires less interventions to patient</td>
<td>OPTION B</td>
<td>Families would prefer less interventions</td>
</tr>
<tr>
<td>Population-related benefits</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td></td>
</tr>
</tbody>
</table>

**COST (Arising from Options A or B)**: Average Direct Cost Per Day: this represents supply + labor costs, not charge to patient or actual cost of an item

| ROOM RATE ($)                          | No difference                                      | No difference                                      | NEUTRAL           | N/A               |
| Diagnostic and Treatment Costs ($)     | Average annual cost over 3 years= $5,554             | Predicted cost for 5 studies= $1,140                | Estimate annual savings= $4,414. |

**COST (Arising from Options A or B)**: Average Direct Cost Per Day: this represents supply + labor costs, not charge to patient or actual cost of an item

| ROOM RATE ($)                          |                                                | If possible, estimate probability of complication |
| Diagnostic and Treatment Costs ($)     |                                                | |

## Value Analysis Grid

<table>
<thead>
<tr>
<th>COST</th>
<th>A &gt; B</th>
<th>A = B</th>
<th>A &lt; B</th>
<th>Unclear</th>
</tr>
</thead>
<tbody>
<tr>
<td>A costs more than B</td>
<td>Make value judgement</td>
<td></td>
<td></td>
<td>Do B and PDSA in 1 year</td>
</tr>
<tr>
<td>A and B costs are the same</td>
<td>A</td>
<td>A or B, operational factors may influence choice</td>
<td>B</td>
<td>A or B, operational factors may influence choice, PDSA in 1 year</td>
</tr>
<tr>
<td>B costs more than A</td>
<td>A</td>
<td></td>
<td>Make value judgement</td>
<td>Do A and PDSA in 1 year</td>
</tr>
</tbody>
</table>

## Value Statement

**FINAL CSW VALUE STATEMENT**

Option B is preferred because it has the cost advantage in the absence of clear benefit with Option A. Key assumptions include: 1) Team would prefer less tests; 2) Less tests would lead to less disruption to patient care; 3) Families would prefer less tests and interventions when deemed not routinely necessary. This recommendation is based on very low quality evidence. A cost-benefit analysis was applied. Estimated yearly cost savings is $11057.*

## Cost Savings Hypothesis Narrative

**COST SAVINGS HYPOTHESIS NARRATIVE**

Total costs for cranial US over the past 3 years= $16,611 with a total of 73 studies. Average annual basis equals $5,554, with 24 studies per year at an average cost of $228/test.
Approved by the CSW Ductal Dependent Systemic Blood Flow Pre-Op Pathway Team for July 13, 2016 Go-Live.

CSW  Ductal Dependent Systemic Blood Flow Pre-Op Pathway Team:

Cardiac Intensive Care Unit, Owner: Kihan Kim, MD
Cardiac Intensive Care Unit: Rob Mazor, MD
Cardiac Intensive Care Unit: Aimee Jennings, ARNP
Cardiac Intensive Care Unit: Jennie Choe, ARNP
Cardiac Intensive Care Unit: Karen Corlett, ARNP
CICU Clinical Nurse Specialist: Christy Cain, RN
CICU Clinical Pharmacy: Christa Jefferis Kirk, PharmD
Heart Center: Barbara Miller, ARNP
Pharmacy Informatics: Rebecca Ford, PharmD

Clinical Effectiveness Team:

Consultant: Jeff Foti, MD
Project Manager: Gioia Gonzalez, MSW, LICSW
CE Analyst: James Johnson
CIS Informatician: Carlos Villavicencio, MD, MMI
CIS Analyst: Heather Marshall
Librarian: Susan Klawansky, MLIS
Project Manager Associate: Asa Herrman

Executive Approval:

Sr. VP, Chief Medical Officer: Mark Del Beccaro, MD
Sr. VP, Chief Nursing Officer: Madlyn Murrey, RN, MN
Surgeon-in-Chief: Bob Sawin, MD


Please cite as:
We used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial, or observational studies. The rating is then adjusted in the following manner:

Quality ratings are *downgraded* if studies:
• Have serious limitations
• Have inconsistent results
• If evidence does not directly address clinical questions
• If estimates are imprecise OR
• If it is felt that there is substantial publication bias

Quality ratings can be *upgraded* if it is felt that:
• The effect size is large
• If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
• If a dose-response gradient is evident

**Quality of Evidence:**

- ☉☉☉☉ High quality
- ☉☉☉ Moderate quality
- ☉☉ Low quality
- ☉☉☉ Very low quality

Expert Opinion (E)

Summary of Version Changes

- **Version 1.0 (08/15/2012):** Go live
- **Version 2.0 (7/13/2016):** Periodic Review
- **Version 3.0 (2/6/2017):** CSW Value Analysis completed, changes include removing cranial ultrasound unless clinically indicated.
Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children’s Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.
629 records identified through database searching

2 additional records identified through other sources

631 records after duplicates removed

631 records screened

605 records excluded

13 full-text articles excluded, 11 did not answer clinical question
2 did not meet quality threshold
0 outdated relative to other included study

26 records assessed for eligibility

13 studies included in pathway

Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535


