

**Pathway Purpose:** To provide a comprehensive care plan that standardizes essential steps in the management of patients with single ventricle physiology undergoing the Norwood operation to optimize outcomes according to the milestones in the table below. Comprised of 5 phases of care, the pathway describes the expected clinical course and sets targets for key milestones.

**Inclusion Criteria:**  
Any infant undergoing Norwood surgery

**High-risk Criteria (consider hybrid strategy):**

- Other major system abnormalities (CDH, GI, neuro)
- Restrictive atrial septum (needing cath intervention)
- $\geq$  Moderate dysfunction
- $\geq$  Moderate valve regurgitation
- Additional major cardiac anatomic anomalies (ie, TAPVR)
- Prematurity (<36 weeks GA)
- Weight <2.5 kg
- Suspected significant genetic anomaly
- Or major pre-op co-morbidities (lung disease, sepsis)

## 5 Phases of Care with Key Milestones

Phases of Care	Key Milestones	Targets
<u>Pre-Operative Phase (NICU/CVICU)</u>	Time to OR	3-5 days of life
<u>Intra-operative Phase (OR)</u>	Induction to Incision	90 minutes
<u>Early Post-Operative Phase (CVICU) – To extubation</u>	Sternal Closure	POD 3
	Extubation	POD 7
<u>Late Post-Operative Phase (CVICU/ACCU) – Extubation to ACCU</u>	ACCU Transfer	POD 20
<u>Acute Care Phase – ACCU to discharge</u>	Discharge	POD 36

**Abbreviations**

**CRT:** Capillary Refill Time  
**HMP:** Home Monitoring Program  
**HRIF:** High risk infant follow up  
**LCOS:** Low Cardiac Output syndrome  
**MBM:** Maternal Breast Milk

**NIRS:** Near Infrared Spectroscopy  
**DLRA:** Double Lumen right atrial line  
**RA:** Broviac: Right atrial Broviac line  
**SVP:** Single ventricle program  
**TAG:** Team Assessment Group

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**Associated Order Set:** HLHS pre-op order set

**Associated Policies:** n/a



Feedback?

## Pre-Operative Phase (NICU/CVICU)

Milestone: Time to OR Target 3-5 days of life 

- Use **HLHS pre-op order set** for monitoring, labs, medications and studies
- Order consults for SVP, cardiogenomics, cardiac neurodevelopment, lactation, and palliative care
- Place HLHS Education Binder (for care team) at bedside (only for HLHS)
- Confirm surgical plan in surgical conference or TAG
- Arrange for PRE-OP HUDDLE with NORWOOD FLIGHT PLAN once OR time set

Is there  
pulmonary over-circulation (O<sub>2</sub> sat >85, tachypnea, increased WOB, or pulmonary edema on CXR)  
WITH  
systemic compromise (tachycardia, lactate >3, decreased perfusion, hypotension, decreased NIRS and/or end organ dysfunction)?

No

Yes

- **Avoid intubation & supplemental O<sub>2</sub>**
- Start caffeine
- Consider diuretic
- Initiate MBM buccal swabs and BF ad lib
- Frequent re-assessment
- Transfer to CVICU by DOL 2

### Communication:

- Update primary cardiologist & PCP
- Offer SV parent mentor & resiliency resources to family



- NPO
- Optimize HCT (40-45%)
- Begin vasoactive support (milrinone 0.25-0.5 mcg/kg/min or epinephrine 0.02-0.05 mcg/kg/min)
- Initiate diuretic
- Start caffeine
- Initiate HFNC or CPAP (21%)
- If intubation needed, use cuffed ETT and plan for urgent OR
- Frequent re-assessment
- Transfer to CVICU on DOL 1

## Intra-operative Phase (OR)

Milestone: Induction to Incision 90 minutes 

### LINES

- UAC
- Radial or axillary art line
- DL RA vs RA Broviac
- PD drain, if indicated

### RESPIRATORY

- If transitioning off CPB on 100% FiO<sub>2</sub>, wean promptly to goal targets and ideally to 21-30%
- Transfer from bypass to ICU ventilator for bypass separation

### SURGICAL

- RV to PA conduit (valved) unless systemic ventricle is single dominant LV
- Open sternum

### GOALS

PaO <sub>2</sub>	35-45
PCO <sub>2</sub>	40-50
O <sub>2</sub> sat	75-85%
Hct	40-45%
MAP	40-50 mmHg
Temp	36-37

### MEDICATIONS

- Vasoactives: Infusions in D5W
  - Milrinone 0.25-0.5 mcg/kg/min
  - Epinephrine 0.02-0.05 mcg/kg/min
  - +/- Dopamine 3-5 mcg/kg/min
  - Calcium gtt
  - Nitroprusside if needed for HTN
- Pain/Sedation:
  - At least 30 min prior to leaving OR, transition to solely fentanyl +/- vecuronium infusions
- Antibiotics:
  - Cefazolin
  - If MRSA/MRSE, use cefazolin + vanco
- Coagulation therapy per protocol
- Initial chaser rate max 10ml/hr

### Communication:

- OR to update family throughout case
- CVICU attending to go to OR when closing



## Early Post-Operative Phase (CVICU) – To extubation

Milestone: Sternal closure by POD 3 & extubation by POD 7 

- Use **HLHS post-op order set** for monitoring, labs, medications and studies
- Use bedside QpQs and AVO<sub>2</sub> monitoring chart (see Appendix A)
- Initiate TPN on POD 1
- Remove UAC/UVC within 48 hrs of sternal closure
- Initiate feeds per Heart Center feeding protocol

### RESPIRATORY

- Wean FiO<sub>2</sub> to 21-25%
- If O<sub>2</sub> sat below goal, optimize cardiac output before increasing FiO<sub>2</sub>

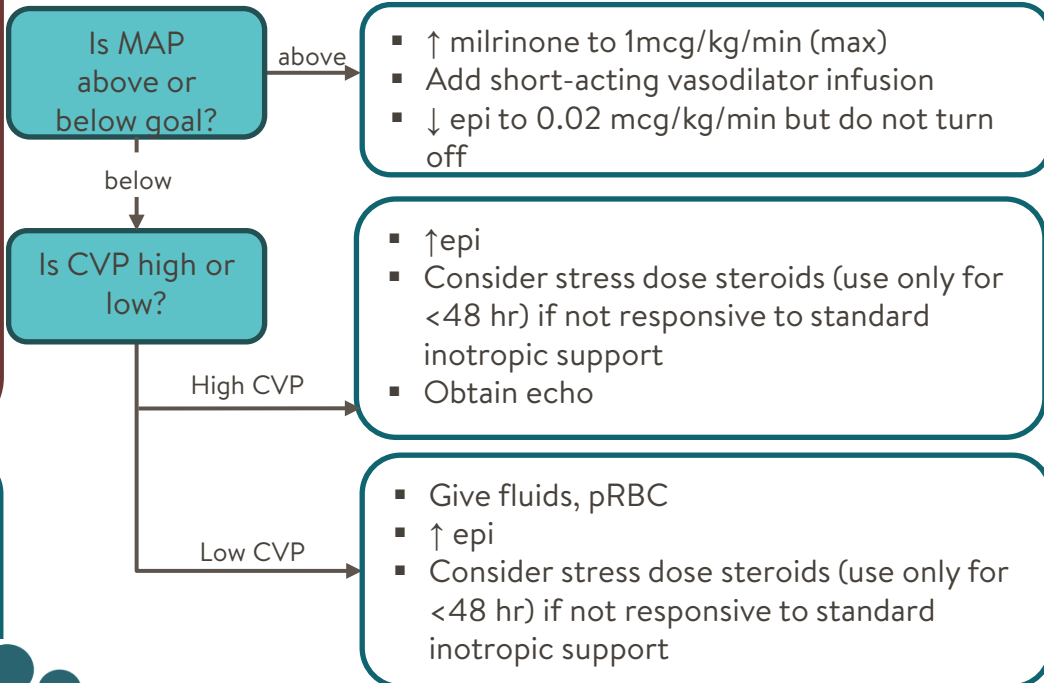
### MEDICATIONS

- Continue milrinone, epinephrine & calcium through chest closure
- Avoid neuromuscular blockade, dexmedetomidine and midazolam infusions
- Antibiotics: Continue through 24 hrs after chest closure
- Diuretics: Initiate furosemide gtt by POD 2
- Anticoagulation:
  - RV-PA conduit = ASA (5-10mg/kg) daily on POD 1
  - mBTTS = rectal ASA in OR or <6 hrs post, then heparin (10 units/kg/hr)

### GOALS

PaO <sub>2</sub>	35-45
PCO <sub>2</sub>	40-50
O <sub>2</sub> sat	75-85%
Hct	40-45%
MAP	40-50 mmHg
Temp	36-37

**Communication:**  
Update primary cardiologist  
Identify primary CVICU cardiologist



If persistent LCOS, hypoxemia, or significant end-organ dysfunction, plan for sternal exploration +/- ECMO

## Late Post-Operative Phase (CVICU/ACCU) – Extubation to ACCU

Milestone: ACCU transfer by POD 20 

### MONITORING

- At least weekly ECHO
- Daily upper/lower BP
- At least monthly EKG
- Daily weights, weekly length and HC
- Check Hct weekly, transfuse to keep Hct 40-45%

### NUTRITION

- Advance NG/NJ feeds to goal 120-140 kcal/kg/day
- Consider ENT evaluation if vocal cord/swallow concern

### RESPIRATORY

- If requires continued respiratory support, wean FiO<sub>2</sub> to 21-25%

### GOALS

PaO <sub>2</sub>	35-45
PCO <sub>2</sub>	40-50
O <sub>2</sub> sat	75-85%
Hct	40-45%
MAP	40-50 mmHg

### MEDICATIONS

- Vasoactives: Transition off infusions, target SBP 60-80, use captopril if needed
- Pain/Sedation: Transition to enteral regimen with wean plan
- Once on FULL feeds, start digoxin (5 mcg/kg/dose BID) and ferrous sulfate (5 mg/kg/day)

### Criteria for transfer to acute care

- Off infusions
- No change in afterload agents, diuretics or anti-arrhythmics for >24 hrs
- No change in respiratory support for >24 hours
- Nutrition regimen (enteral +/- TPN) with stable weight trend (no active weight loss)
- Secure venous access
- Establish long-term plan for lines (ie timing for Broviac removal) with surgeons

### Communication:

- Start HMP teaching once extubated
- Update primary cardiologist
- Offer SV parent mentor & mental health resources
- Weekly family care conferences



## Management Suggestions for Excessive Pulmonary Blood Flow

### Signs/symptoms of excessive PBF with **ADEQUATE** systemic perfusion

- HR 120-170's
- Increased WOB RR 60-80's
- Warm extremities
- 2+ peripheral pulses
- Capillary refill time < 3 sec
- SaO<sub>2</sub> to MvO<sub>2</sub> difference < 30
- NIRS > 40 or at baseline
- MAP appropriate for age
- Lactate < 3
- Base deficit no more than -2
- Stable Creatinine
- UOP > 1ml/kg/h
- CXR with pulmonary edema

Yes

- Start HFNC FiO<sub>2</sub> 21%
- Diurese to BUN ~ 20-30
- Increase enteral afterload meds to goal SBP
- Advance feeds as tolerated

Adequate systemic perfusion?

No

Yes

### Monitor

- ABG lactate
- Trend HR, RR, Sat
- Follow UO closely
- Follow renal function
- Follow CXR

Yes

Adequate systemic perfusion?

No

Consider further imaging (MRI),  
catheterization, surgical intervention, or  
mechanical support

### Signs/symptoms of excessive PBF with **INADEQUATE** systemic perfusion

- HR >170 sustained
- Increased WOB RR > 80
- Cool extremities
- Pulses < 1+
- Delayed capillary refill time > 3 sec
- Lactate > 3 persistently
- SaO<sub>2</sub> to MvO<sub>2</sub> > 30 persistently
- Need for > 2 bicarb replacements
- Metabolic acidosis
- Decreased NIRS > 10 points from baseline
- Increased Creatinine
- UOP < 1ml/kg/hr
- Elevated inflammatory markers non-infectious
- CXR with pulmonary edema
- Increased Cystatin C
- Elevated pro BNP

Yes

- IV afterload reduction to goal MAP
- Consider low dose Epi
- Transfuse to Hct > 40
- HFNC 6-8L 21%
- Diurese to BUN ~ 20-30
- NPO, TPN with optimal kcals
- Consider intubation (with permissive hypercapnia)
- Minimize metabolic demands
- Daily CXR
- ECHO

## Post-operative Phase – ACCU to discharge

**TARGET TIMES: Discharge by POD 36** 

### MEDICATIONS

- Digoxin (5 mcg/kg/dose BID), unless contraindicated
- Ferrous sulfate (5 mg/kg/day) when on full feeds
- Wean to enteral diuretics, max TID frequency; add spironolactone if TID
- Wean to enteral sedative & opioids, max TID frequency
- GI prophylaxis if GERD or NG
- ASA prophylaxis
- ACE inhibitor, unless contraindicated

### MONITORING

- EKG monthly
- ECHO q2 weeks
- CXR weekly
- Labs:
  - Chem/CBC prn & before discharge
  - Thyroid function tests (once >6 weeks of age)
  - Cystatin C upon transfer and before discharge
- Weight: daily
- HC & length: weekly
- If persistent need for O<sub>2</sub>, consider sleep study

### GOALS

O2 sat	75-85%
Weight Gain	20-30 grams/day
PO or bolus feeds	120-150 kcal/kg/day
Hct	40-45%

Transfuse if needed before discharge

### CRITERIA FOR DISCHARGE

- HMP teaching complete
- No changes in cardiac meds/diuretics x 2 days
- No oxygen supplementation x 3 days
- Stable nutrition (no changes in formula or caloric density) x 3 days
- Home formula ordered and available
- Nutrition teaching with nutrition bundle complete
- PMD identified & appointment made within 2 weeks
- Primary cardiology appointment made within 2 weeks
- All subspecialists appointment requests sent
- Pulse ox and durable medical equipment delivered
- >24 hr rooming in completed
- Medications pickup and verified
- OT/PT Rx signed and given to parents
- Referral to HRIF/Early Start
- Health care maintenance completed (CPR/car seat test/ALGO)
- RSV prophylaxis arranged (seasonal)
- Car seat and safe sleeping space confirmed

### Communication:

- Ongoing HMP teaching
- Update primary cardiologist
- Sign out to PMD



