Interstage Monitoring for the Infant with Hypoplastic Left Heart Syndrome
What the Direct Care Nurse Needs to Know

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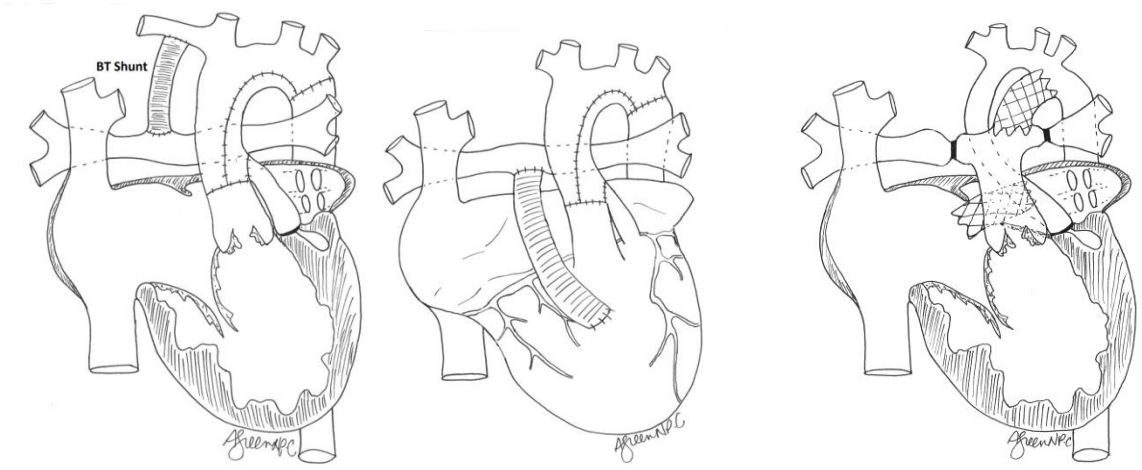
Introduction:

Children born with hypoplastic left heart syndrome (HLHS) are at high risk for serious morbidity, growth failure and mortality during the time from discharge home after first stage HLHS palliation, until admission for the second stage surgical intervention. To improve outcomes and survival during this time, referred to as the *"interstage period"*, multiple strategies have been successfully implemented resulting in improved interstage survival. A critical element of interstage care is family education and training. Variations in practice and interstage home surveillance do exist but general guidelines are described below.

Definitions:

Possible newborn interventions for HLHS

Figure 1: First stage surgical palliation procedures for hyoplastic left heart syndrome



A. Norwood Procedure with BT shunt

B. Norwood Procedure with RV to PA shunt ("Sano")

C. Hybrid Stage I Procedure

Surgical:

- Norwood /Modified Blalock Taussig (BT) shunt procedure – the Norwood aortic reconstruction of the ascending aorta and arch consists of enlarging the ascending aorta by using part of the proximal native main pulmonary artery (PA) with other material (Gore-Tex or homograft material) to connect the PA to the aorta. Pulmonary blood flow is provided by a controlled shunt (BT shunt) usually made from Gore-Tex and connects the right subclavian artery to the right pulmonary artery. The PDA is ligated. Following a

Norwood procedure, the right ventricle provides the entire cardiac output through the reconstructed aorta. Part of the cardiac output flows into the aorta and part will go thru the BT shunt, providing pulmonary blood flow flows to both right and left pulmonary artery via. See diagram 1A

- Norwood / Sano procedure – the Norwood aortic reconstruction of the ascending aorta and arch is completed. In the Sano modification, the source for pulmonary blood flow is a conduit placed from the right ventricle to the PA, supplying the left and right PA branches. See diagram 1B

Cardiac Catheterization, Interventional:

- Hybrid stage I procedure – in this procedure for HLHS the surgical Norwood arch reconstruction is not done as an initial surgery. Interventional cardiac catheterization is done to place a stent in the ductus arteriosus. Systemic blood flow is right-to-left from the right ventricle to the ductus arteriosus and into the aorta that remains patent. Surgeons then place PA bands around each pulmonary artery branch to control the pulmonary blood flow. In this case, the aortic reconstruction is completed at the surgery for the Bidirectional Glenn. See diagram 1C

Interstage monitoring – daily home surveillance monitoring of oxygen saturations, enteral intake and weight during the interstage period. The home monitoring begins at the time of discharge home and ends when the second stage Bidirectional Glenn surgery is done.

Critical Thinking Points:

- Arterial oxygen saturations following each stage I palliation is expected to be in the range of 75-85% on room air.
- All interstage Norwood/BT shunt patients are shunt dependent with their only source of pulmonary blood flow being through the shunt. The shunt can gradually or suddenly narrow or occlude, causing hypoxemia and cyanosis. This can be life-threatening. Always assess for the trends in oxygen saturations and the presence of a shunt murmur.
- These children have minimal cardiac reserve and can quickly develop hemodynamic compromise from simple childhood illnesses such as gastroenteritis, vomiting, poor feeding or respiratory infection.
- Unexpected death is usually preceded by a prodrome which can include poor oral intake, vomiting, fussiness, diarrhea, cyanosis, fever or increased work of breathing. A high level of vigilance and prompt intervention is imperative.
- The focus needs to be on *early* detection of signs that could lead to cardiopulmonary decompensation with prompt observation, family notification of the patient care team and intervention by the medical team.

"Teach Back" is the Recommended Method for Discharge Instructions:

- It's important to assess for thorough understanding and retention of family information. This shouldn't be done by simply asking, "Do you understand?" The family may indicate yes to please the caregiver, not seem ignorant or to avoid embarrassment.
- When providing the education, use plain language, ask open-ended questions that cannot be answered with a simple yes or no, and provide reader-friendly print materials to support learning.

- "Teach Back" is a technique to make sure you, the health care provider, has explained information clearly. It is a way to check for care-giver understanding and, if needed, re-explanation of content.
- Tell the parent there is a lot of information that can be overwhelming and/or confusing so you want to make sure you have explained it clearly. Ask them to explain **in their own words** with a demonstration of what was taught to them. The parent will "teach back" the information to you as if they were the teacher.
    - The family can be prompted, for example, to show you how they would teach grandma to prepare the Aspirin and give it to the baby.
- Using a "Teach Back" approach, the family will *describe* their understanding, *explain* how they would teach this skill to a different family member, as well as *perform* the task.
- Teach Back is a research-based health literacy intervention that improves patient-provider communication and patient health outcomes (Schillinger, 2003).

Red Flags / When to Call:

During the interstage it is important to have early identification of potentially dangerous symptoms in the infant which can signal developing cardiopulmonary deterioration. Caregivers and the collaborating physician teams are counseled to detect and promptly report these "Red Flag Symptoms" of danger:

- Failure to gain 0.01-0.02 kg (10-20 grams) over a 3 day span
- Child loses 0.03 kg (30 grams) or more
- Oxygen saturations drop consistently below 75%, saturations above 90% or a change from baseline values
- Feeding problems, such as vomiting or decreased intake
- Increased work of breathing
- Unusual, increased irritability
- Something "just doesn't seem quite right"…when in doubt, call!
- Example of "Red Flag" Teaching sheet from Miami Children's Hospital Pediatric and Adult Congenital Heart Disease Program - Appendix A

Preparing for Discharge Home / Discharge Checklist:

- Start the discharge teaching process **early** as the baby is recovering in the ICU
- Nurses to develop a "Discharge Teaching Checklist" to identify required areas
- Families can receive a checklist identifying what they need to learn prior to discharge home ("Journey Board")
- Include a second caregiver in the discharge education
- Instruction regarding interstage monitoring and logbook ("binder") record keeping
    - How/when to use the pulse oximeter
    - How to use the baby scale and calculate changes in weight
    - Use of care binder
    - Families should actively practice recording of feedings, weights, saturations and heart rate on the log sheets. Programs may elect to use a digital version of the daily logs.

    - Families will typically receive communications, or phone calls from the single ventricle team member between clinic visits to provide reassessment and support
- Nutrition and Feeding
    - How to feed your baby – oral, tube fed, both?
    - Positioning
    - Feeding schedule
        - Family to receive feeding plan, volume per feed, total ounces per day prior to discharge
        - Feeding plan to be reviewed and updated as out-patient
    - Formula preparation
        - Calorically enhanced formula recipe is given to family
        - 24, 27 or 30 calorie per ounce formula
        - Family must prepare recipe with "Teach back"
- Assessing adequate intake and output
    - Medications
    - Call their pharmacy to make sure compounding available
    - Medication teaching with teach back
    - Provide syringes and dosing schedule
    - Have medication picked up and ready for rooming in
    - Commonly prescribed medications at the time of discharge home:
        - Diuretic, ace inhibitor, anti-platelet or anticoagulant, anti-reflux
- Rooming In for 24-48 hours should be mandatory prior to dismissal
    - Simulate home environment with parent independently caring for baby
    - Involve two caregivers
    - All discharge teaching should be done **prior** to rooming in
- Plan for immunizations and Synagis
    - There is no known association with adverse events or sudden death following immunizations
    - Some centers elect to withhold or delay immunizations in fragile interstage infants
    - All interstage infants should receive a monthly injection of Synagis (palivizumab) during RSV season
- Home equipment & supplies - Teach Back required for:
    - Digital baby scale
    - Pulse oximeter
    - Feeding tube (NG or GT) supplies
- Completion of newborn care (BAER, newborn screen)
- Infant CPR training
- Follow-up in the Out-Patient setting
    - The infant is typically seen every 1-2 weeks after discharge with phone contact in between visits.
    - Appointments for the first month should be made prior to dismissal
        - PCP, cardiologist, CT surgery
        - Interstage monitoring requires close, frequent follow-up. Often these Cardiology visits are done in a specialty "high risk" cardiac clinics.
- Discharge coordination with PCP / referring cardiologist

    - A pre- discharge conference call is done at many centers. This usually takes place 1-2 days prior to dismissal to discuss the patient's medical history, home surveillance monitoring, and plans for outpatient care.
    - The importance of a high level of vigilance and low threshold for readmission with occurrence of red flags needs to be conveyed.
    - The meeting should include parents, PCP, cardiologist, CT surgery NP, discharge coordinator and others as needed (case management, OT/PT, dietitian, social work)
- Assess the need for home health nursing visits, OT/PT, other specialists
- Enroll in a neurodevelopmental clinic, if available
- Provide parent with contact information for questions/problems that can be used 24/7
    - Families must demonstrate how they will contact the team 24/7
- Provide parent with written instructions of all discharge information for future reference
    - Binder will include all "Red Flag" symptoms lists, contact numbers, nutrition, immunization, daily logs, developmental information

Outpatient Clinic Assessment (elements to include):

- Weight, weight-for-age percentile, average daily weight gain
- Length, weight-for-length percentile
- Oxygen saturation
- Current medications
- Nutrition – route, type of formula, caloric density, total # feeds/day and volume/feeding
- Calculate actual cal/kg/day and identify goal cal/kg/day
- Review red flags, plan for immunizations/Synagis, interim issues or ER visits
- Review plan/date for next visit
- Example of Follow-Up sheets from Miami Children's Hospital Pediatric and Adult Congenital Heart Disease Program – Appendix B

References:

Nieves, J., Uzark, K., Rudd, N., et al. (Publication pending 2015). Interstage home monitoring following newborn first stage palliation for hypoplastic left heart syndrome: The process and family education strategies. *Critical Care Nursing*

Rudd, N., Frommelt, M. A., Tweddell, J. S., et al. (2014). Improving interstage survival after Norwood operation: Outcomes from 10 years of home monitoring. *J Thorac Cardiovasc Surg, 3*, 1-8.

Helpful sites for additional information:

National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC):
Multicenter collaborative looking at quality improvement during the interstage period for infants with HLHS.
https://jcchdqi.org

Sisters by Heart: Online parent support for those who have a child with HLHS
http://wwwsisters-by-heart.org

Society of Pediatric Cardiovascular Nurses (SPCN): Professional organization with links to clinical practice guidelines and other reputable websites
http://wwwspcnonline.com

Appendix A

THE HEART PROGRAM

MIAMI CHILDREN'S HOSPITAL

HIGH RISK CARDIAC SURGICAL CLINIC

"RED FLAGS" DANGER SYMPTOMS LIST

Your baby has *Hypoplastic Left Heart Syndrome. A Norwood* surgery was done. This includes placement of **a GORETEX SHUNT** to provide blood flow to the lungs. If the shunt becomes narrow, the baby will have lower oxygen levels in the blood & be more blue.

****Expected oxygen saturation levels will be: 75% - 90%**

CALL your Cardiologist or the Miami Children's Hospital Cardiac Care Center if your baby has any of the "Red Flag" symptoms:

- Poor feeding
- Weight loss or failure to gain weight for 3 days
- Breaths fast or hard
- Oxygen levels are less than 75% or more than 90%
- Appears blue or pale in the face, lips or hands while at rest
- Diarrhea or vomiting, sweating
- Irritable, fussy
- If it necessary to hospitalize your baby

Primary Cardiologist: Dr. xxxxxx (305) xxx-xxxx

24 HOURS/Day CONTACT: Miami Children's Hospital Cardiac ICU –

305 669 6500 or **1 800 666 4278**

Miami Children's Hospital Cardiology 786 624 4344 (Jo Ann Nieves ARNP) or cell phone 786-414-0751

If you feel you have an Emergency Call 9 1 1

Appendix B

Miami Children's Hospital Adult and Pediatric Congenital Heart Program

Sample Home Care Binder Log

Feedings, weights and vital signs

Oxygen Saturation & Weight Record:

WEEK # ____

DATE:	Oxygen level in blood (SATURATION)	Heart beats per minute	WEIGHT	Feeding: TOTAL ounces per 24 hour day
Monday				
Tuesday				
Wednesday				
Thursday				
Friday				
Saturday				
Sunday				

Feeding GOAL AT LEAST ____ oz/day

½ ounce = 15 ml
1 ounce = 30 ml
1 ½ ounce= 45 ml

Formula:
_____ ___ Calories/oz.

Comments:
_____
_____

Please call your Cardiologist if: ** If the SATURATION is 75% OR LESS
- ** If your baby **loses weight** (1 ounce) or falls to have a weight gain over 3 days.
- Note: We **expect** your baby to **gain weight of ½ to one ounce every day**