JOHNS HOPKINS ALL CHILDREN'S HOSPITAL HEART INSTITUTE

High Risk Neonatal Cardiac Deliveries

JOHNS HOPKINS
All Children's Hospital

Johns Hopkins All Children's Hospital

High Risk Neonatal Cardiac Deliveries Clinical Pathway

Table of Contents

Updated: November 2023

Owners: Zachary Coffman MD; Joy Johnson MD; Noura Nickel MD

Johns Hopkins All Children's Hospital

High Risk Neonatal Cardiac Deliveries

Rationale:

This clinical pathway was developed by a consensus group of JHACH physicians, advanced practice providers, nurses and pharmacists to standardize the management of neonatal deliveries for patients with high-risk cardiac disease.

Background:

Improvements in fetal echocardiography have increased recognition of fetuses with congenital heart disease that require specialized delivery room care.[1] With this increase in fetal detection, the clinical course at delivery can be predicted, and postnatal instability can be mitigated. Despite these advances, certain populations of patients with congenital heart disease have high rates of hemodynamic compromise that can begin as early as the delivery room, even if diagnosed prenatally. For these infants, subspecialty and multidisciplinary care must begin in the delivery room to optimize their outcomes.[2,3] Prior literature has described the efficacy and importance of risk stratifying infants based on their expected postnatal physiology and risk for clinical decompensation at that time of delivery.[2] In addition, the literature has described the benefits of beginning multi-disciplinary planning prior to delivery, including a multidisciplinary, detailed plan for delivery room management in the most high risk infants.[1,3]

To optimize the birth and transition of the infants we care for, we sought to create a comprehensive, nuanced, multidisciplinary risk stratification for infants prenatally diagnosed with congenital heart disease. We also sought to create a process for planning for their birth, beginning with maternal care prior to delivery, and culminating in a well-organized and communicated delivery room and transfer plan of care. This pathway serves as a guideline for risk stratifying fetal cardiac deliveries and a framework for planning delivery room management for those who are high risk for neonatal hemodynamic compromise.

Impacted Patient Population:

This guideline applies to fetal patients with prenatally detected congenital heart disease. Patients with clinically significant heart disease will be risk stratified into risk categories to guide need for potential postnatal interventions in those with concern for hemodynamic instability.

Risk Stratification of Fetal Patients with Cardiac Disease

Patient anatomy and expected postnatal physiology is delineated in as much detail as possible during the patient's fetal echocardiogram. Additional echocardiograms may be required to adequately detail the patient's anatomy and potential postnatal physiology throughout pregnancy.

Due to the variability in fetal imaging, the initial fetal echocardiogram may not identify all aspects of a patient's cardiac disease. Additional imaging throughout pregnancy can help monitor fetal progression and evolution of cardiac defects. Despite close monitoring, postnatal physiology may still vary and the plan of care may need to be adjusted to optimally care for the patient. Significant extracardiac malformations or defects may require deviation from this pathway as

well. Discussion with NICU, CVICU, cardiology, palliative care, and other consulting services should be completed prenatally to determine the most appropriate direction of care for both patient and family if these malformations are believed to significantly impact survivability.

Once the anatomy and expected physiology are clarified, the fetal cardiologist will place a patient in an appropriate Level of Complexity (LOC) based on a number of factors including but not limited to:

- Dependence on a patent ductus arteriosus for either pulmonary or systemic circulation
- Presence and severity of circular shunts
- Arrhythmias
- Need for urgent postnatal cardiac intervention
- Concern for hemodynamic or respiratory compromise upon disconnection of the placenta
- Evidence of prenatal hydrops or poor ventricular function

Level of Care (LOC) Categorization:

Table 1: Neonatal Cardiac Delivery Risk Stratification Table					
LOC	**Complexity and Concerns**	**Example CHD**	**Prenatal Planning**	**Delivery Planning**	**Post-delivery Recommendations**
1	Mild complexity CHD. No physiologic instability in first weeks of life.	1. Septal defects (e.g. ASD, VSD, AVSD) 2. Benign arrhythmias	No specific planning Phone call from fetal cardiology x 1 in pregnancy	Spontaneous vaginal delivery	Nursery or NICU (if extracardiac concerns) Routine perinatal management. Postnatal cardiac testing as per fetal cardiology to confirm diagnosis.
2	Moderate complexity CHD. Possible physiologic instability in first weeks of life. May require early intervention.	1. Possible ductal dependency (e.g. "pink" TOF, possible coarctation of aorta) 2. Non-sustained or controlled tachyarrhythmia.	Prenatal evaluation at JHACH Fetal Care Program. Delivery at location with pediatric cardiology support available.	Spontaneous vaginal delivery.	CVICU/NICU. Early postnatal cardiology consult. Prostaglandins readily available, but do not start without discussing with cardiology first.
3	Severe complexity CHD with physiologic stability. Requires postnatal intervention/surgery before discharge.	1. Ductal-dependent lesions or lesions with complex physiology (e.g. HLHS, PA/IVS, "blue" TOF, truncus arteriosus, unbalanced AVSD).	Prenatal evaluation at JHACH Fetal Care Program. Delivery at BFBP.	Spontaneous vaginal delivery vs. induction at 39 wks. gestation	CVICU admission. Umbilical catheters. Prostaglandins for ductal dependent lesions.
4	Severe complexity CHD with expected instability. Requires postnatal intervention/surgery before discharge, and possibly urgently	1. Severe Ebstein's anomaly or TOF/APV 2. D-TGA, HLHS with restrictive PFO/Intact Atrial septum 3. Palliative care: Possible instability leading to death shortly after delivery or may be stable for several hours to several days before death. Family has chosen comfort care. 4. Significant cardiomyopathy 5. Hydrops	Prenatal evaluation at JHACH Fetal Care Program. Delivery at BFBP. Pre-delivery team meeting with CVICU, cardiology, neonatology, and MFM. Interventional/surgical team on standby.	Planned induction at 39 wks. with daytime delivery. NICU contacts CVICU and cardiology when mother arrives for delivery to arrange pre-delivery huddle. Consideration of planned c-section to ensure coordinated daytime delivery in the most severe cases	CVICU admission <1 hour of age. Umbilical catheters. Prostaglandins for ductal dependent lesions. Stabilizing medications/equipment predetermined by care plan. HIGH ACUITY RAPID TRANSPORT PROTOCOL (Code HART)

P	Palliative care: Possible instability leading to death shortly after delivery or may be stable for several hours to days before death. Family has chosen comfort care	Trisomy 13, trisomy 18, severe complex CHD with other congenital anomalies	Palliative care consult with delivery plan in chart including parental wishes. Delivery can be supported at any location with advanced planning regarding the palliative plan with the delivery team.	As per palliative care delivery plan, per parent wishes	Plan for post-delivery interventions as per parent wishes
Perinatal Palliative Care via the Fetal Care Team to meet with all single ventricles, anyone with multiple congenital anomalies expected to affect survival, any baby with hydrops, plus others at the discretion of the fetal cardiology team.					

Prenatal preparation:

- Consults:
    - Fetal Cardiology Team
    - Maternal-Fetal Medicine Team
    - Social Work
    - NICU via Fetal Care Program
    - Palliative Care Team if appropriate and available (Patients with single ventricle anatomy/physiology, multiple congenital anomalies, fetal hydrops, or others at the discretion of the fetal cardiology team)
- For all pregnancies with a congenital heart defect at a LOC 3 or 4, a "Cardiac HART (High Acuity Rapid Transport) Checklist" documenting the postnatal plans will be completed after the fetal care program meeting. This huddle sheet will include:
    - Mother and fetus identification
    - Cardiac Diagnosis
    - Expected postnatal physiology
    - Delivery room plan including:
        - Respiratory support
        - IV access
        - Medications (PGEs, vasoactive medications, antiarrhythmics, etc.)
        - Anticipated saturations
    - Infant disposition
    - Phone tree
    - Special considerations
- For those with a LOC 4, a multidisciplinary provider huddle will be completed 1-2 weeks prior to planned delivery including all expected involved teams (CV surgery, CV anesthesia, Interventional cardiology, echocardiography, CVICU, Perfusion if needed) with a delineation of delivery room plans, postnatal disposition, and phone tree for notification. This huddle will result in a completed checklist which will be attached to the delivery invite and distributed to the Stork team.
    - CVICU Preadmit Huddle Plan will also be completed the morning of delivery during or immediately following the CVICU morning huddle at 08:30.

Delivery Room Management:

- Postnatal consults
    - Neonatal Intensive Care Unit

        - Any patient in the CVICU younger than 28 days of age should have at minimum a one-time NICU consult for evaluation and management of routine newborn issues.
        - If there are ongoing issues of prematurity, extracardiac anomalies, or other neonatal concerns, the NICU team will remain available for continued participation and assistance with management of the patient.
- The CVICU is the optimal postnatal location for patients with ductal dependent cardiac physiology or concern for developing clinical instability secondary to congenital heart disease with the following characteristics:
    - CHD without significant extracardiac congenital anomalies who are anticipated to require cardiac intervention during the newborn hospitalization.
    - Gestational age >34 weeks **AND** birth weight >1500 grams with anticipated cardiac intervention during the first 2 weeks of life.

Admission considerations based on LOC:

- **LOC 1: Nursery or NICU**
    - If the infant qualifies for admission to the newborn nursery, the infant can be delivered at an outside hospital with postnatal cardiology clinic follow up within the first few days of life.
- **LOC 2: CVICU/NICU**
    - If the infant qualifies for a CVICU/NICU admission, the infant needs to be delivered at Bayfront Babyplace given very limited echocardiographic capabilities at outside facilities. Ability to rapidly escalate care to the CVICU may be needed for these patients and possible CVICU admission should be discussed with cardiology.
    - Babies who are in the NICU and develop a requirement for cardiac intervention will be transferred to the CVICU 48 hours prior to intervention if clinically feasible, or as soon as medically possible if decision is made within < 48 hours.
- **LOC 3: CVICU admission**
- **LOC 4: CVICU admission <1 hour of age**
    - Consider transportation to the interventional suite based on discussion with cardiology about patient's postnatal physiology and potential need for more urgent intervention.
    - Stabilizing medications and equipment will be determined prior to delivery at the high-risk cardiac delivery team discussion and detailed in the Cardiac HART Checklist.
    - If deemed necessary based on postnatal physiology, initiate Urgent Transport Protocol **(Code HART).**

Johns Hopkins All Children's Hospital

Inpatient High Risk Neonatal Cardiac Deliveries Clinical Pathway

Pregnant patient identified and evaluated in Fetal Care Center with concern for fetal congenital heart disease (CHD).

Fetal Cardiology identifies Level of Complexity (LOC) based on the Neonatal Cardiac Delivery Risk Stratification Table (Table 1)

LOC 1-2

Neonatology and Fetal Cardiology plan for routine delivery based on patient's and mother's additional risk factors.

LOC 3-4

Fetal Cardiology, CVICU, Neonatology, and Maternal-fetal Medicine meet 1-2 weeks prior to delivery to discuss patient's anticipated postnatal course.

Discussion guided by Cardiac HART (High Acuity Rapid Transport) checklist. Checklist will be distributed to all delivery personnel prior to delivery. (Appendix A)

Comfort Care

Significant congenital malformations or cardiorespiratory instability leading to death shortly after delivery or within several hours to days before death. Family has chosen comfort care.

Palliative care consult with delivery plan in chart including parental wishes. Delivery can be supported at any location with advanced planning regarding the palliative plan with the delivery team.

LOC 3

Severe complexity CHD with physiologic stability. Requires postnatal intervention/surgery before discharge.

- Prenatal evaluation at JHACH FetalCare Program.
- Delivery at BFBP. Pre-delivery Team meeting with CVICU, cardiology, neonatology, and MFM.
- Spontaneous vaginal delivery vs. induction at 39 wks. gestation

LOC 4

Severe complexity CHD with expected instability. Requires postnatal intervention/surgery before discharge, and possibly urgently.

- Prenatal evaluation at JHACH Fetal Care Program.
- Delivery at BFBP. Pre-delivery Team meeting with CVICU, cardiology, neonatology, and MFM.
- Planned induction at 39 wks. with daytime delivery.
- CVICU admission <1 hour of age.
- Consider planned C-section to ensure coordinated daytime delivery in the most severe cases
- Interventional/surgical team on standby.
- URGENT TRANSPORT PROTOCOL (Code HART)

CVICU Preadmit Huddle Plan

1. Scheduled IOL/C section: Discuss admission during 08:30 huddle
2. Admission not anticipated at time of 08:30 huddle should prompt huddle once admission known including
   a. CVICU attending
   b. CVICU APP
   c. CVICU CN
   d. CVICU bedside RN
3. Phone call to NICU STORK Team (Yellow) to review admission plan (CVICU Newborn Admission Flow):
   a. Umbilical line placement
   b. Intubation
   c. Medications (Vasoactive medications, PGE).
   d. Need for urgent intervention
4. Discuss during prenatal meeting and with STORK Team if Pre-admission is warranted.
   a. Follow Pre-admission guidelines (Appendix B).

References

1. Donofrio MT, Levey RJ, Schuette JJ, et al. Specialized Delivery room planning for Fetuses with Critical Congenital Heart Disease. Am J Cardiol 2013:111:737-747
2. Donofrio MT, Skurow-Todd, K, Berger, JT, et al. Risk-Stratified Postnatal Care of Newborns with Congenital Heart Disease determined by Fetal Echocardiography. J am Soc Echocardiogr 2015; 28:1339-49
3. Sanapo L, Moon-Grady AJ, Donofrio MT. Perinatal and Delivery Management of Infants with Congenital Heart Disease. Clin Perinatol 43 (2016) 55-71

Outcome Measures:

- Postnatal mortality
- Unexpected interventions
- Time to disposition (admission to appropriate unit or interventional suite)

Clinical Pathway Team

High Risk Neonatal Cardiac Deliveries Clinical Pathway

Johns Hopkins All Children's Hospital

Owner(s): Zachary Coffman MD, Joy Johnson MD, Noura Nickel MD

Also Reviewed by: Heart Institute Clinical Practice Council

Pharmacists: Amy Kiskaddon, PharmD, MBA

Date Approved by JHACH Heart Institute Clinical Practice Council: May 14, 2023

Date Approved by JHACH Clinical Practice Council: Sept 20, 2023

Last Revised: 11/15/23

Live on Webpage: 11/16/2023

Disclaimer

Appendix A: Cardiac HART Checklist

Heart Institute
600 5th St. S, Suite 5200
St. Petersburg, FL 33701
727-767-3333 P
727-767-8990 F

JOHNS HOPKINS
MEDICINE
JOHNS HOPKINS
ALL CHILDREN'S HOSPITAL

Cardiac HART Checklist

Patient Name: _____ DOB: _____ MRN: _____

EDD: _____ Planned delivery date: _____ Planned delivery mode: _____

High risk cardiac diagnosis: _____ ☐ LOC3 ☐ LOC4

Expected postnatal physiology:

- ☐ Ductal dependent pulmonary blood flow (Cyanosis)
- ☐ Ductal dependent systemic blood flow (Poor perfusion)
- ☐ Ineffective pulmonary blood flow (TGA, TAPVR with obstruction)
- ☐ Cardiomyopathy
- ☐ Arrhythmia
- ☐ _____

Delivery room plan:

- ☐ Intubate _____
- ☐ Umbilical lines to be placed in ☐ DR ☐ CVICU
- ☐ PIV
- ☐ PGE at 0.01 mcg/kg/min
- ☐ Vasoactive Medications (med and rate) _____
- ☐ Antiarrhythmics _____
- ☐ Anticipated O_2 saturation_____
- ☐ _____

Infant disposition:

☐ CVICU ☐ NICU ☐ CV OR ☐ CV Cath lab

Phone tree: Who needs to be notified about impending delivery?

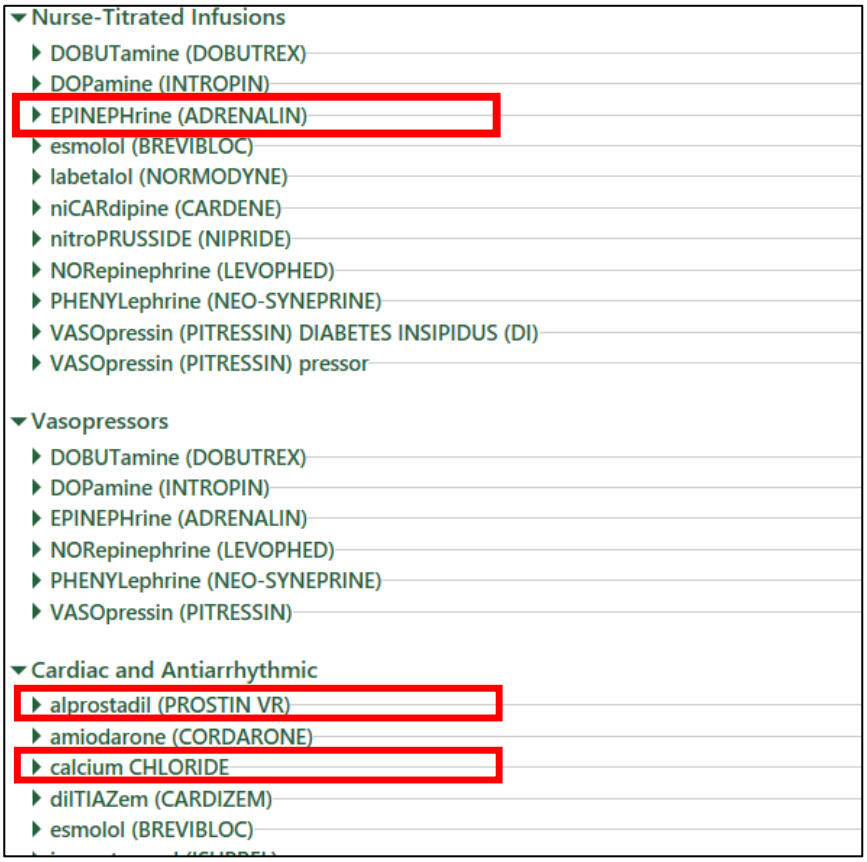
- ☐ CVICU attending and charge nurse
- ☐ Interventional cardiology
- ☐ CV surgery
- ☐ Electrophysiology

Special Considerations:

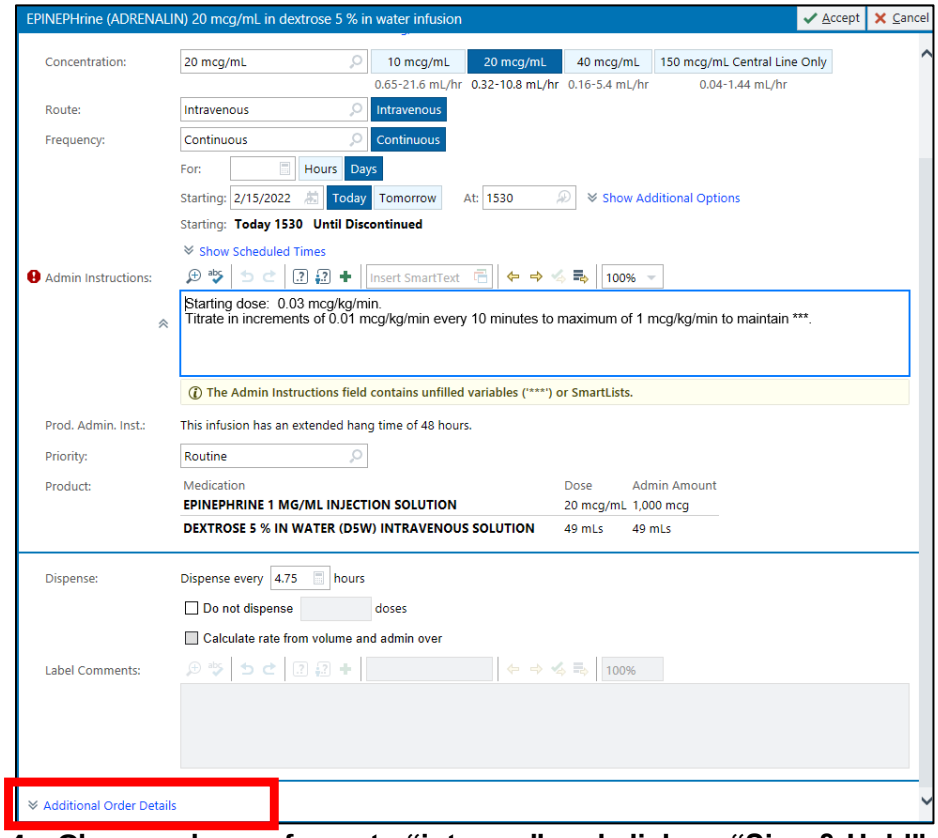
Appendix B: Pre-admission Orders in Epic for High Risk Patients

1. **After patient is pre-admitted, please order medication drips using the JHH-ACH PICU-CVICU-PCICU Infusions Focused Order Set**

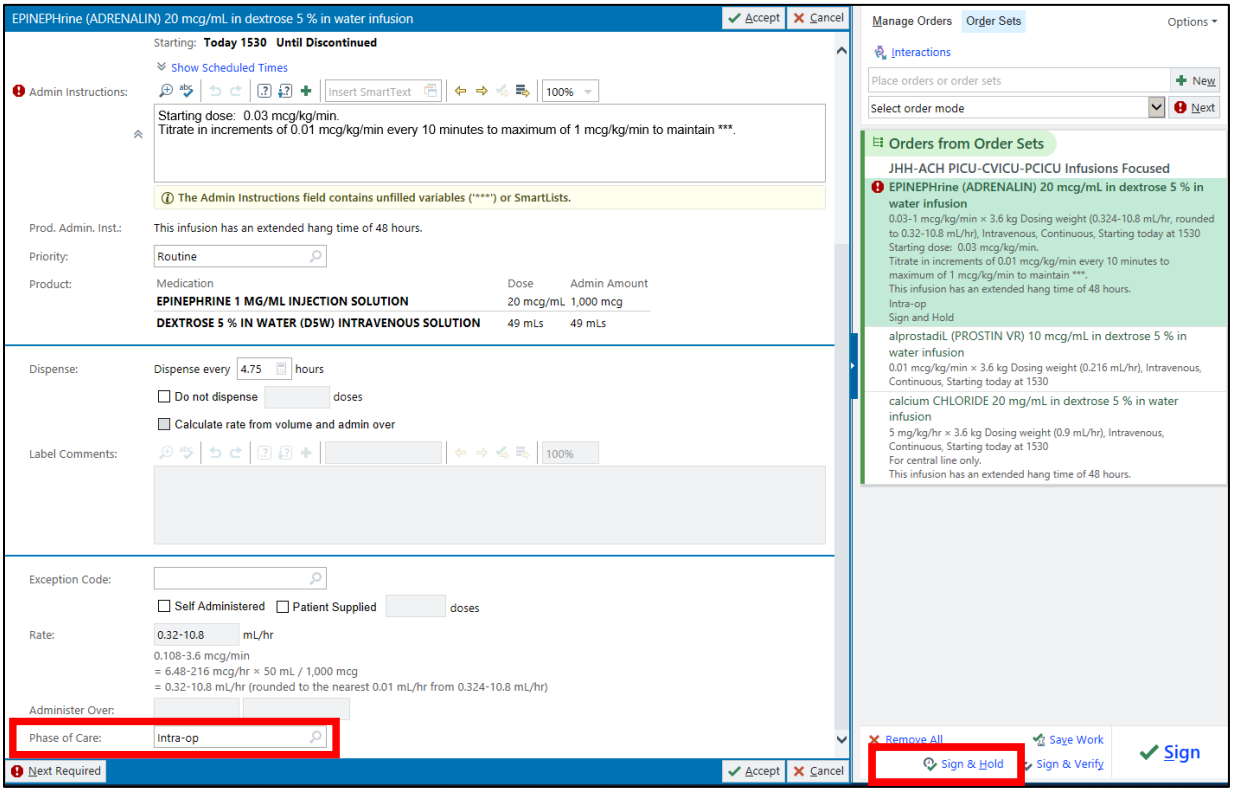
2. **Select medications to order**



3. After selecting dose and instructions, click on additional order details



4. Change phase of care to “intra-op” and click on “Sign & Hold”



5. **Once ordered, please call CVICU Pharmacist or Main Pharmacy to request drips be verified and tubed to CVICU tube station #352**
    a. Meds will be handed off to the STORK team prior to going to the delivery room to ensure appropriate and rapid administration if and when they are needed.