CARDIAC PATHWAY

Aortic Arch Watch Pathway

Pathway Purpose: To provide a standardized management pathway for infants at risk for developing coarctation of the aorta with constriction of the ductus arteriosus. In infants on the pathway who do not develop surgical coarctation of the aorta and have no other major comorbidities, we aim to reduce umbilical line placement and NPO orders to less than 10%.

Inclusion Criteria: Patients with fetal echocardiogram performed at ≥30 weeks gestational age raising concern for coarctation of the aorta.

Prenatal Phase

Assess risk based on prenatal echocardiogram considerations

Slightly Increased Risk Criteria

Findings concerning for coarctation (e.g. isthmus z-score is < -2)

Moderate Risk Criteria

1. Moderately hypoplastic aortic arch/isthmus (z < -3), **OR**
2. Mildly hypoplastic aortic arch/isthmus (z<-2 or isthmus/ductus ratio < 0.7)
   AND at least one of the following:
   - Mitral and aortic valve hypoplasia (combined z-score < -4)
   - Another sign of coarctation, including
     - Presence of posterior shelf
     - Increased MPA/Ao or RV/LV ratio (above 2.0)
     - Increased length of distal transverse arch (carotid subclavian index < 1.0)
     - Disease which increases the risk of coarctation: large ventricular septal defect, left superior vena cava, atrioventricular septal defect, double outlet right ventricle, Turner's syndrome

High Risk Criteria

Moderately hypoplastic aortic arch **AND** otherwise unexplained left to right flow at the PFO or reversed aortic arch flow

Determine preliminary perinatal plan according to risk identified on prenatal echocardiogram
See page 2 for plan based on results of postnatal echocardiogram

Slightly Increased Risk

- No prenatal consultation with a surgeon necessary
- No need for change in delivery location to a hospital with a congenital heart surgical program
- Plan to admit to well-baby nursery
- Plan to initiate standard postnatal care

Moderate Risk or High Risk

- Consult: fetal center neonatology +/- surgical consult (at fetal cardiologist/parental preference)
- Plan for delivery at hospital with a congenital heart surgical program (e.g. LPCH**; may require change in planned delivery location**)
- Plan to admit to neonatal intensive care unit (NICU)
- Obtain echocardiogram upon arrival to NICU to guide further management
- Need for UAC/UVC placement and Prostaglandin E1 (PGE1) to be determined by postnatal echocardiogram (see page 2)

Feedback?

Owner: Shiraz Maskatia, MD
Team: David Kwiatkowski, MD; Alexis Davis, MD
Physician Liaison: Claudia Algaze, MD
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Associated Order Set: Neonatal Aortic Coarctation Evaluation and Management
Associated Policies: None

Packard Clinical Pathway Program

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Postnatal Phase

Did the prenatal echocardiogram indicate moderate or high risk for coarctation?

NO

Obtain echocardiogram between 1 and 7 days after birth; may be done as inpatient or outpatient

YES

Obtain echocardiogram immediately upon arrival to NICU.
If **high** risk, results should be directly relayed to ICU attending by echo attending within 4 hours of birth

What is the updated diagnosis or risk based on the postnatal echocardiogram?

At any time, notify cardiology consultant if any of the following noted: **(1)** systolic upper limb BP is > 15 mmHg greater than lower lower limb BP, **(2)** post ductal 02 saturation is lower than pre ductal by ≥ 10 %, **(3)** post ductal saturation does not read, **(4)** lactate > 2 mg/dL

Mild risk or no coarctation

Management Considerations:

- Referral to cardiology if mild coarctation and no PDA or aortic arch clearly normal despite presence of PDA
- No referral to cardiology if there is no coarctation and no PDA

Confirmed mild or no coarctation

Cannot Rule Out Severe Coarctation

If postnatal echocardiogram performed as outpatient, admit patient to CVICU/NICU

Perform echocardiogram q48-72h until establishing mild risk or no coarctation or confirmed severe coarctation

Ok to repeat echocardiogram earlier if clinical change

- PIV; no UAC/UVC in majority of cases
- Case-by-case central line placement for complex or particularly high-risk cases
- 4-limb blood pressures (BP) and perfusion/pulse checks q6h
- Continuous NIRS, pre/post ductal 0_2 sat
- PO ad lib if no other contraindication; no nasogastric feeds

Confirmed Severe Coarctation

Management Considerations:

- PGE1, UAC, UVC
- 4-limb BP q6h
- Perfusion/pulse check q3h
- ABG with lactate q12h
- Continuous NIRS and pre/post ductal 0_2 sat
- May initiate PO trophic feeds and advance as tolerated
- No nasogastric feeds
- Notify CV surgery team and schedulers
- Repeat echocardiogram if clinical change

Confirmed severe coarctation

References:

Maskatia et al, “A Fetal Risk Stratification Pathway for Neonatal Aortic Coarctation Reduces Medical Exposure” J Pediatr. 2021 Oct:237:102-108

Villalaín et al, “Diagnostic accuracy of prenatal ultrasound in coarctation of aorta: systematic review and individual participant data meta-analysis” Ultrasound Obstet Gynecol. 2024 Apr;63(4):446-456

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