CHOA Delivery Room Protocol
Severe Ebstein's Anomaly (EA)

Children's Healthcare of Atlanta

Ebstein's Anomaly (EA) is a congenital dysplasia of the tricuspid valve secondary to failure of delamination of the septal leaflet from the ventricular myocardium. This often leads to a coaptation defect resulting in tricuspid regurgitation (TR). EA represents a spectrum of disease ranging from mild displacement of the tricuspid valve leaflets and minimal TR to severe cardiomegaly associated with severe TR. Other associated features of EA include right ventricular outflow tract (RVOT) obstruction, atrial arrhythmias, and biventricular dysfunction. Severe TR can lead to cardiomegaly and compression of the lung fields resulting in pulmonary hypoplasia. Newborns at the highest risk of hemodynamic instability have evidence of significant pulmonary regurgitation in addition to tricuspid regurgitation, which can lead to a circular shunt in-utero and postnatally. A circular shunt results in blood flowing in a loop that bypasses the pulmonary circulation, and severely compromises the systemic circulation and oxygen delivery ("systemic steal"). Postnatally, newborns with severe EA can have pronounced hypoxia and poor perfusion in the delivery room. These newborns can present with hypoxia, inadequate systemic perfusion, respiratory distress or arrhythmias.

Prenatal clinical checklist:

1. Is there evidence of flow reversal in the ductus arteriosus?
2. Is anatomic or function pulmonary valve atresia suspected?
3. Is there either right or left ventricular dysfunction (>mild)?
4. Is there evidence of lung hypoplasia seen by fetal MRI?
5. Is there pulmonary regurgitation (>mild) seen on the last fetal echocardiogram?
6. Are there fetal tachyarrhythmias (i.e. supraventricular tachycardia SVT)?
7. Is there hydrops or other signs of fetal distress?

Goals for delivery room and transport:

Assessment of degree of hypoxia and respiratory distress. Hypoxia can occur due to a poorly functioning right ventricle as well as compression of the lungs due to right atrial enlargement. Hemodynamic compromise can occur due to decreased cardiac output from the left ventricle due to circular shunting, LV compression, and/or myocardial dysfunction.

Clinical goals in the DR/during transport include:

- Secure airway for adequate ventilation and oxygenation
- Insure stable source of pulmonary blood flow
- Minimize oxygen consumption with sedation/paralysis
- Support systemic perfusion
- Treat tachyarrhythmias and maintain sinus rhythm

Access plan:

1. Low-lying single lumen 5 French UVC without XR confirmation
2. Peripheral IV if able to place in timely fashion
3. UAC to monitor hemodynamics
4. Telemetry to assess rhythm (EKG not necessary)

Medication plan:

Have the following available in the delivery room:

1. D10W at 60 ml/kg/day (80 ml/kg/day: maternal DM, IUGR, preterm, or low DS)
2. Medications to aid intubation OR for sedation and reduction in metabolic demands:
   a. Fentanyl IV 1 mcg/kg/dose (fentanyl may be given rapidly if immediately followed by vecuronium)
   b. Vecuronium IV 0.1 mg/kg/dose or Rocuronium 1 mg/kg dose for neuromuscular blockade
3. Start prostaglandin (PGE1) at 0.01 mcg/kg/min on all patients
4. Inotropic support:
   a. Epinephrine 0.05-0.01 mcg/kg/min IV infusion
5. Rescue medications to have available:
   a. IV atropine (0.02 mg/kg/dose) for bradycardia
   b. IV epinephrine (0.02 mg/kg/dose) for circulatory collapse
   c. Adenosine (0.1-0.2 mg/kg/dose) for the presence of sustained SVT
   d. Procainamide 5mg/kg in case of ongoing SVT

Equipment/supplies:

1. Recommend cuffed endotracheal tube for term infants
2. Blood pressure cuff
3. Pulse-oximetry monitoring
4. Cardiac monitoring to evaluate rhythm strip

Delivery room management plan for High-Risk:

High-Risk indications:

- Severe cardiomegaly (cardiothoracic ratio >0.5)
- Retrograde or bidirectional shunting at the ductal level
- Pulmonary regurgitation that is greater than mild
- Left ventricular dysfunction
- Evidence of fetal hydrops

1. Delayed cord clamping per institutional protocol. This may not be feasible in the setting of fetal distress.
2. Follow NRP guidelines for initial resuscitation.
3. Pre-ductal saturations should be >75% by 10 min.
4. If hypoxemia is not improved or respiratory distress is not stabilized, proceed to intubation:
   a. If intubating, recommend providing sedation and paralysis to both facilitate intubation and decrease metabolic demand post intubation
5. Start PGE at 0.01 mcg/kg/min for all high-risk patients
6. Consider pulmonary vasodilators (i.e. nitric oxide) with persistent hypoxemia (will limit patient to ground transport).
7. If there is evidence of poor perfusion, consider initiating inotropic support:
   a. Recommend epinephrine as first line, 0.02 mcg/kg/min (avoid if there is evidence of sustained tachyarrhythmia)
8. If there is evidence of supraventricular tachycardia (SVT), administer IV adenosine (0.1 mg/kg (max 6 mg) as a rapid bolus. Can give a second IV adenosine dose of 0.2 mg/kg.
   a. Consider synchronized cardioversion if the patient has hemodynamic instability, adenosine is ineffective, or difficult IV access.
   b. Consider Procainamide bolus if SVT persists
9. Plan for transport to AMBH

Delivery room management plan for Standard-Risk:

Standard-Risk indications:

- No evidence of cardiomegaly
- Antegrade flow seen at the ductus arteriosus
- No significant pulmonary regurgitation
- No ventricular dysfunction
- No fetal hydrops

1. Delayed cord clamping per institutional protocol. This may not be feasible in the setting of fetal distress.
2. Follow NRP guidelines for initial resuscitation. Supplemental oxygen may be used.
3. Obtain blood gas and address any metabolic abnormalities.
4. Consider echocardiogram for assessment of anatomy and postnatal physiology with cardiology consult.
5. Consult with cardiologist-on-call regarding disposition (i.e. need for CICU versus NICU versus nursery)

Post delivery room strategies:

Ongoing ventilatory strategies:

- Higher PEEP (target of 8 cmH2O) to overcome effect of cardiomegaly on lung inflation
- FiO2 of 1.0, iNO, and PGE to promote pulmonary blood flow
- Sedation with neuromuscular blockade while mechanically ventilated

Ongoing cardiac monitoring:

- Monitor cardiac rhythm while on inotropic support to assess for tachyarrhythmias

Developed through the efforts of Children's Healthcare of Atlanta and physicians on Children's medical staff in the interest of advancing pediatric healthcare. This is a general guideline and does not represent a professional care standard governing providers' obligation to patients. Ultimately the patient's physician must determine the most appropriate care.
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