The Newborn with Obstructed Pulmonary Veins is Coming to the Cath Lab Now!

**Moderators:**  
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**Objectives:**  
1. Discuss the use of ECMO and interventional catheterization lab procedures in newborns with obstructed total anomalous pulmonary venous connection (TAPVC)
2. Review the anesthetic management and resuscitation of newborns with obstructed TAPVC presenting emergently to the interventional catheterization lab
3. Discuss the management of pulmonary hypertensive crises in newborns
4. Discuss the risk factors for morbidity and mortality in newborns with obstructed TAPVC
5. Review the post-operative complications associated with surgical repair of TAPVC

**Case History**

As the pediatric anesthesiologist on call today, you have just been notified that a 40+ week EGA male infant born at an outside hospital has just been transferred to your facility. He was born this afternoon via Cesarean section for failure of progression of labor. A post-natal echo at the outside hospital revealed possible obstructed TAPVC and aortic coarctation. No prenatal care had been received. A repeat echo at your hospital confirms obstructed infradiaphragmatic TAPVC with a mean gradient of 7 mm Hg, atrial septal defect (ASD) with non-restrictive R⇒L flow, large patent ductus arteriosus (PDA) with R⇒L flow and situs ambiguous. Cardiology would like to bring the infant down to the cath lab now for possible ductus venosus stenting.

**Questions:**

What are the types of TAPVC? What concerns are present with obstruction? What causes obstruction? What clinical findings are present with obstruction? What would you expect to see on chest x-ray? Does the finding of situs ambiguous play a role?
Apgars at the time of birth were 3 and 7. The infant was intubated and placed on PGE infusion at 0.05 mcg/kg/minute. Both UAC and UVC lines are in place. Current vital signs are: T 37.5, HR 150, BP 61/36/46, O2 sat 84% on FiO2 100%. Weight is 3.36 kg.

Questions:

Who would be taking care of this infant at your institution—general peds vs cardiac? Do you think ECMO should be on stand-by? Does the cardiac surgeon need to be in house during the procedure? Why isn’t this infant just going for surgical repair instead?

The infant arrives to the cath lab. HR and BP are stable as given in report, however oxygen saturations have fallen to the 60’s.

Questions:

The cardiologist says he is getting femoral access. Any other lines you would like to place? What is your anesthetic plan? Is there an ideal recommended anesthetic strategy? Would you recommend TEE use during the procedure? Was the PGE infusion the right thing to do? How is ductus venosus stenting accomplished? What are the possible risks/complications associated with cardiac catheterization and ductus venosus stenting?

The first ABG you draw is 7.30/46/31/23/-4 with O2 saturation=54% as the cardiologist begins his procedure.

Questions:

How do you treat the low PaO2? What are other causes of hypoxia in this infant? Would you transfuse blood? Would you consider nitric oxide? Would start inotropic support at this time?

Two hours later, the cardiologist informs you he has a wire from the umbilical vein into the ductus venosus and across to the right atrium. He is ready to balloon and deploy the stent across the ductus venosus. Just after deployment, the oxygen saturation rises to 80%, however the saturations rapidly fall down into the 20’s and stop reading. HR remains in the 120’s. You rapidly draw an ABG that is 7.27/29/22/-12. Fluoroscopy of the chest shows worsening pulmonary congestion.

Questions:

What just happened? Will you give bicarbonate? What is the definition of pulmonary hypertension in pediatric patients and what are normal values for pulmonary pressures and
pulmonary vascular resistance (PVR)? What is a pulmonary hypertensive crisis? What triggers an increase in pulmonary vascular resistance? How is it treated?

Oxygen saturations are not improving despite all maneuvers. Systemic blood pressure is now starting to fall as well. The ECMO team is called for, but is currently unavailable as they are upstairs putting another patient emergently on ECMO. The decision is made to bring the child to the OR for surgical repair.

Questions:
What are the risk factors for post-operative morbidity and mortality following repair of obstructed TAPVC? What are the most common post-operative complications seen following surgical repairs? How is long-term outcome following repair?

Discussion:
Total anomalous pulmonary venous connection (TAPVC) accounts for 1-3% of all congenital heart defects. There is no connection between the pulmonary veins and the left atrium; rather the pulmonary veins drain to the right side of the heart. Survival is dependent on R⇒L shunting. The four types of TAPVC are classified according to the site where the connection occurs:

1. **Supracardiac** which accounts for approximately 50% of all TAPVC involves a connection to the right superior vena cava, azygous system, persistent left superior vena or left innominate vein
2. **Cardiac** which involves a connection to the coronary sinus or right atrium
3. **Infracardiac** which involves a connection below the diaphragm to the portal veins, ductus venosus or inferior vena cava
4. **Mixed-type** involves a combination of any of the above

Obstruction to pulmonary venous return occurs in approximately 25% of all TAPVC cases, with infradiaphragmatic connections having the highest association with obstruction. Obstruction is related to either intrinsic abnormality of the vessel wall, extrinsic compression by adjacent structures, or narrowing at the level of the diaphragm or ductus venosus. Obstruction constitutes a true surgical emergency. Neonates with obstruction to pulmonary venous return present with pulmonary venous hypertension, pulmonary edema and increased pulmonary pressures. These infants have refractory hypoxemia and marked acidosis placing them at extremely high risk for morbidity and mortality related to emergency surgery requiring deep hypothermic circulatory arrest. Intervention in the catheterization lab such as stenting of the ductus venosus or vertical vein is a temporizing alternative, relieving obstruction of the pulmonary venous return allowing the neonate to clinically stabilize and allow for surgical repair at a later date when the PVR has fallen.
Intervention such as this may help to improve outcomes as emergency surgical mortality is reported to be as high as 10-30%. A second alternative is the use of extracorporeal membrane oxygenation (ECMO). ECMO is the most common form of mechanical circulatory support for pediatric patients, especially in those with pulmonary hypertension.  There are essentially no patient size limitations in comparison to the use of ventricular assist devices, and ECMO provides complete cardiopulmonary support from either central or peripheral cannulation. The disadvantages include a higher associated incidence of neurologic morbidity, non-pulsatile flow, and a high risk for thromboembolic events necessitating systemic anticoagulation that unfortunately increases the risk of bleeding. The presence of congenital heart disease has been shown to be a predictor of in-hospital death with mortality reaching 66%.vi

Pulmonary arterial hypertension is defined as a mean pulmonary artery pressure > 25 mm Hg at rest or > 30 mm Hg with exercise, an associated pulmonary capillary wedge pressure ≤15 mm Hg and PVR >3 Wood units (or >300 dyne/s/cm²). Normal PVR is 1.1-1.4 Wood units or 90-120 dyne/s/cm². Pulmonary hypertension in pediatric patients is more commonly defined as systolic pulmonary artery pressure > 1/2 of systolic systemic blood pressure. Over half of the neonates with TAPVC were shown to have pulmonary hypertension when cardiac cath was performed.ivi Patients with pulmonary hypertension are at a significantly higher risk for major complications such as pulmonary hypertensive crises, arrhythmias, cardiac arrest and post-operative death.viii Pulmonary hypertensive crisis is defined as an acute increase in PVR with resultant pulmonary artery pressures exceeding systemic blood pressure. This can lead to decreased right ventricular function with subsequent acute right heart failure resulting in decreased pulmonary blood flow and hypoxia, decreased cardiac output and eventual biventricular failure. Triggering factors that can increase PVR include: hypercarbia, hypoxia, acidosis, hypothermia (shivering), noxious stimuli such as pain or airway management, and catheter stimulation of the heart or pulmonary vasculature. Treatment ix involves:

1. Airway management
   a. Check patency of endotracheal tube
   b. Administration of 100% oxygen
   c. Hyperventilation
   d. Administration of nitric oxide (20 ppm)

2. Avoid myocardial depressants
   a. Limitation of inhalational agents
   b. Propofol and Thiopental decrease SVR which can increase R⇒L shunting

3. Treatment of metabolic acidosis
   a. Beware of volume overload on an impaired right ventricle
   b. Excessive preload may worsen right ventricular dilatation shifting the septum towards the left impeding left ventricular filling thereby decreasing cardiac output
4. Support cardiac output
   a. Inotropic support with dopamine, epinephrine or milrinone
   b. Would like to avoid decrease in SVR which decreases systemic blood
class pressure impairing coronary blood flow (may be seen with isoproterenol or
dobutamine—consider using vasopressin)
   c. Maintain sinus rhythm
5. Attenuate noxious stimuli
   a. Adequate anesthesia and analgesia
   b. Neuromuscular paralysis

Risk factors associated with surgical mortality (death within 30 days of operation) include:
infracardiac anatomy type, obstruction at presentation, pre-operative renal or heart failure,
female gender, single-ventricle physiology, heterotaxy syndrome, cardiac catheterization prior
to repair, and post-operative pulmonary hypertensive crises. Neonates with infracardiac
anatomy tend to be smaller in size and more critically ill at time of presentation. Those who
underwent cardiac catheterization prior to repair were shown to have significantly higher
pulmonary artery pressures and pulmonary vascular resistance in comparison to the other
anatomic types. Understandably, there is a significantly lower probability of survival for this
particular group. Post-operative causes of death include: low cardiac output syndrome,
recurrent pulmonary hypertensive crises, supraventricular arrhythmias, acute renal failure and
heart failure. The post-operative complication of recurrent pulmonary venous obstruction
which occurs in approximately 15% of cases is associated with male gender, cardiopulmonary
and aortic cross clamp times, mean transesophageal gradient >2 mm Hg at the pulmonary vein
confluence anastomosis to the left atrium, single-ventricle physiology, and heterotaxy
syndrome. Late neurodevelopmental outcomes show that 58% of the patients with
TAPVC were impaired in one or more of the following areas: intelligence, academic
achievement, attention, or motor function.

High-risk catheterization interventions and hybrid procedures will continue to become more
commonly performed in the cath lab. Many catheterization labs cannot provide the same level
of care and monitoring available in the operating room, despite the fact that it has been shown
that a higher incidence of complications occur in the catheterization lab compared to the
operating room, especially in pediatric patients <1 year of age. Anesthesiologists who
provide care in the catheterization lab must have a working knowledge of congenital heart
disease and their associated sequelae to be able to recognize and treat major events such as a
pulmonary hypertensive crisis.
References:


