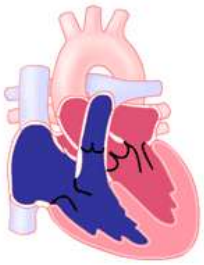
Single Ventricle and Pulmonary Vascular Disease: A Guide for Patients and Families

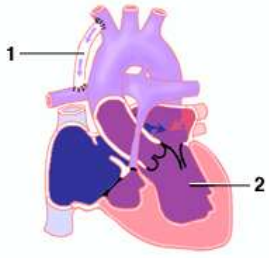
By: Melissa Magness, NP, Kerri Cram, NP, Claire Dlugosz RN, Michelle Ogawa NP, Tisha Kivett RN, Julie Breaux RN, Claire McCracken RN on behalf of the PPHNet APP and Nursing Committee

What is a Single Ventricle Heart?

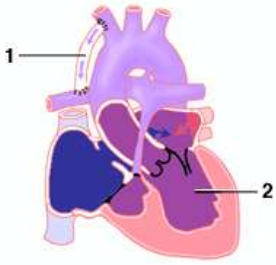
- In a typical heart, there are two pumping chambers called ventricles. The right ventricle sends blood to the lungs to get oxygen, and the left ventricle sends blood to the rest of the body.
- Your child was born with a heart problem that makes one of these ventricles too small or not fully developed. The other ventricle (or the single ventricle) must do all the work of pumping blood to the lungs and the body.
- Your child will likely have a series of surgeries to help the blood flow directly from the body to the lungs without needing a pumping chamber to get it there. For this system to work, the blood pressure in the lungs needs to be low. If the blood pressure in the lungs is too high, the blood will not flow easily into the lungs.



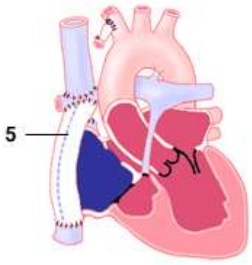
Typical Heart



BTT shunt



Bidirectional Glenn



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What is Pulmonary Vascular Resistance (PVR)?

- PVR stands for pulmonary vascular resistance. This is a measure of how tight the blood vessels are in the lungs. For blood to flow easily from the body to the lungs, these blood vessels need to have low resistance, or low PVR. If the PVR is even a little high, the blood pressure in the lungs will also be high. This makes it harder for blood to move from the body to the lungs.

What is Pulmonary Vascular Disease (PVD)?

- Pulmonary vascular disease (PVD) is a type of illness that affects the blood vessels in the lungs. It can make the PVR or tightness of the blood vessels higher than normal. For children with just a single ventricle, even a small increase in PVR can cause problems. In some cases, it can prevent a child from going to the next stage of surgery.
- It is not clear why a child with a single ventricle might have PVD. It might be because the blood flow from the body to the lungs over time causes changes in the lung's blood vessels, making them tighter. Other problems that can damage the lung's blood vessels, leading to increased PVR, include obstructive sleep apnea (OSA), aspiration, and lung infections.

How do we treat pulmonary vascular disease?

- Your child's provider may prescribe a medicine called a pulmonary vasodilator to treat PVD. This medicine helps relax the blood vessels in the lungs, lowering the blood pressure in the lungs and making it easier for blood to flow to the lungs.
- Doctors who are experts in using these medicines will decide if they are right for your child. Before starting the medicine, they will carefully check to make sure it will not make things worse. If the medicine is given, your child will need to be closely watched.
- Some studies have shown that these medicines can help improve oxygen saturation levels, lower blood pressure in the lungs, and make it easier for children to be active. Pulmonary vasodilators may also improve candidacy for staged single ventricle repair if your child has PVD.

Considerations for Pulmonary Vasodilator Therapy in Single Ventricle Population

Below is a list of potential conditions or complications that may occur due to PVD in single ventricle hearts. Patients that experience these complications may benefit from pulmonary vasodilator treatment.

Condition or Complication	Associated Symptoms
Pulmonary Vascular Disease (PVD)	• Exercise intolerance or decreased activity • Low oxygen saturation • Shortness of breath • Abdominal distention
Protein Losing Enteropathy (PLE)	• Diarrhea • Feeding intolerance (unable to eat or digest food without getting sick) • Swelling of the abdomen or bloating • Swelling of the legs • Low albumin levels
Fontan-Associated Liver Disease (FALD)	• Swelling of the abdomen or bloating • Large Spleen • Low platelet count • GI bleeding • Jaundice (yellow skin color) • Poor growth
Plastic Bronchitis	• Difficulty breathing • Cough • Low oxygen saturation • Coughing up casts

Evaluation and Testing

How do we evaluate a child with single ventricle heart and pvd?

The following tests or procedures may be ordered by your child's medical team to diagnose and evaluate PVD. There are many things that may cause PVD in children with single ventricle hearts. When these things are identified and treated, the PVD often improves

LABS	
Natriuretic peptide tests (NT-proBNP or BNP)	A blood test that shows the heart's level of stress. It can be abnormally high because of PVD.

IMAGING	
Chest X-ray	Imaging study of the heart, lungs, airways, blood vessels, and bones of the spine and chest
Chest CT Angiogram	Imaging study of the lungs' airways, blood vessels, and tissue. It is more detailed than a chest x-ray
Echocardioram	Ultrasound that shows how well the heart valves are working as well as the size and function of the heart chambers
Cardiac MRI	Imaging study to evaluate the structure, size and function of the heart chambers, heart valves and flow through the major blood vessels

EXERCISE TESTING	
Six-minute Walk Test	Test that measures how far your child can walk in 6 minutes while measuring oxygen levels and monitoring symptoms
Cardiopulmonary Exercise Test (CPET)	Test that measures how your child's heart and lungs respond to physical activity

OTHER TESTING	
Cardiac Catheterization	Procedure that provides oxygen saturation and pressure measurements in the heart and major blood vessels connected to the heart, including the pulmonary arteries. The pulmonary vascular resistance (PVR) can be calculated with the information from this procedure
Swallow Study	Test to see how your child is swallowing and if he/she is aspirating (when food goes "down the wrong way" in the airway and enters the lungs)
Electrocardiogram (EKG/ECG)	Test that shows the electrical activity of your child's heart

Medication Options

Drug Class	Examples of Medications	Routes of Administration	Common Side Effects	Special Considerations and Information
Phosphodiesterase Type 5 Inhibitors	Sildenafil (Revatio®)	Orally 3 times per day OR IV (in hospital setting only)	• Headache or irritability • Increased spit up/reflux • Low BP in the body • Mismatch between air/blood interface in the lungs • Erections in males	Available in liquid form (suspension) or tablets in the outpatient setting
	Tadalafil (Adcirca®)	Orally once per day		
Endothelin Receptor Antagonists	Bosentan (Tracleer®)	Orally twice per day	• Low BP in the body • Reversible liver injury • Nasal congestion • Potential harm to fetus – female who is pregnant or who may be pregnant must wear face mask and gloves when administering the medication.	• Child must be enrolled in FDA-required monitoring program due to possible liver concerns • Lab work to check liver function monthly and for anemia every month
	Ambrisentan (Letairis®)	Orally once per day		Lab work to check liver function and anemia (frequency will depend on provider)
	Macitentan (Opsumit®)	Orally once per day		

Abbreviations: IV - Intravenous, BP - blood pressure

ALTHOUGH NOT FDA APPROVED FOR USE IN THIS AGE GROUP OR TYPE OF PH, THESE THREE CLASSES OF DRUGS HAVE BEEN USED OFF-LABEL IN SINGLE VENTRICLE PATIENTS WITH PVD

Medication Options

Drug Class	Examples of Medications	Routes of Administration	Common Side Effects	Special Considerations and Information
Prostacyclins	Epoprostenol (Veletri®)	Continuous IV infusion OR inhaled (in hospital setting only)	• IV and SQ - May cause low BP in the body, headache, nausea, vomiting, diarrhea • SQ may cause pain redness or swelling at the site of infusion • Inhaled has the potential to be irritating to the lungs	IV and SQ options require extensive training on how to prepare and deliver medications at home. These medications require a pump to infuse 24 hours a day, 7 days a week.
	Treprostinil (Remodulin®)	Continuous IV or SQ infusion OR inhaled 4 times a day		
Prostacyclin receptor agonist	Selexipag (Uptravi®)	Oral twice a day	May cause low BP in the body, headache, nausea, vomiting, diarrhea	Taking with food may decrease side effects.

Abbreviations: SQ - Subcutaneous (through the skin), IV - Intravenous, BP - Blood pressure

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To help keep your child stable between and after planned surgery stages

Stay up to date on childhood immunizations, including annual vaccinations and boosters for influenza (flu shot) and COVID-19, as well as Pneumovax 23 after 2 years old. Caregivers and family members are also encouraged to be vaccinated against viral illnesses, such as COVID-19 and the flu.

Receive respiratory syncytial virus (RSV) prophylaxis if your child qualifies. For infants, RSV prophylaxis is recommended with nirsevimab-alip (Beyfortus®) or palivizumab (Synagis®). Please discuss this further with your child's pediatrician and cardiologist.

Avoid secondhand smoke in your home or other places where your child spends time. This includes vaping in addition to cigarette smoke

Stay inside and wear masks if the air quality is poor; for example, due to wildfires or pollution

Take extra caution to minimize exposures to viral illnesses. This includes avoiding individuals who are sick, practicing good hand hygiene, and social distancing during the viral season.

If your child is on daily medications, make sure they are taken regularly and as prescribed. Making changes to your child's medications should be done under the guidance of your specialty team.

Should your child require oxygen, adjustments or weaning of the oxygen should be directed by your specialty team.

Routine exercise and a healthy diet can improve the long-term outcome of patients with single ventricle heart disease

Close follow up with your child's pediatrician, cardiologist, and other specialty teams to follow growth, respiratory status, and developmental progress.

What is long-term care and follow up?

- Children with single ventricle heart disease need to see a heart doctor (cardiologist) for their whole life. They will have many check-ups when they are babies and young children, and they will need to keep seeing the doctor as they grow up. These visits help find and treat problems early.

- Children with single ventricle heart disease often have long-term problems. Your child might need treatments like medications, additional surgeries, or other procedures. In some cases, children might need to be considered for a heart transplant.