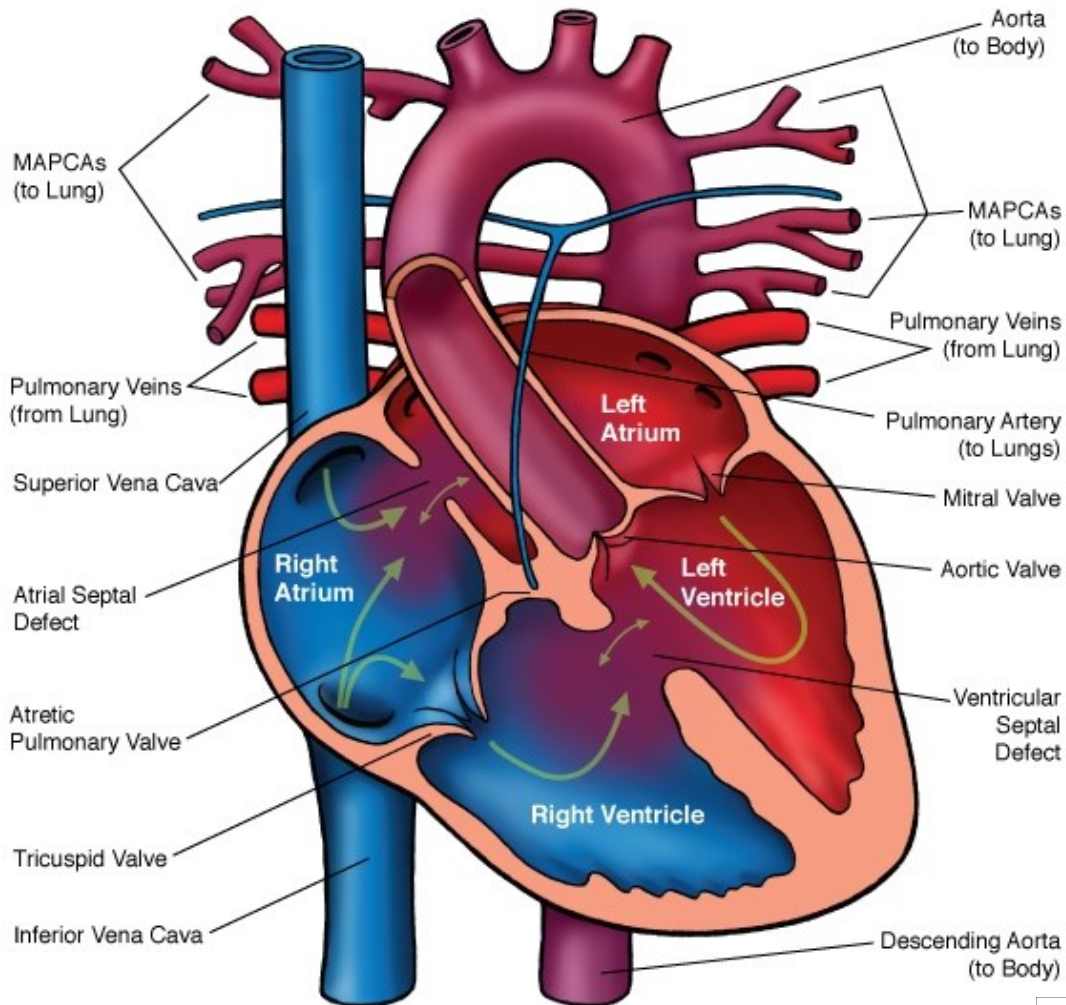


Pulmonary Atresia - MAPCAs

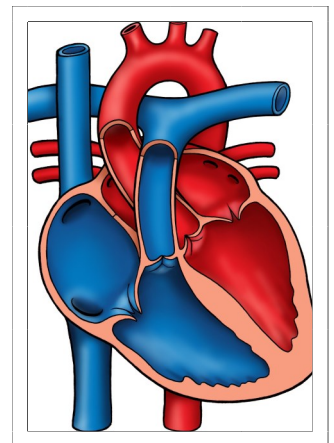
(Major Aorto-Pulmonary Collateral Arteries)



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Notes:

Normal Heart



Pulmonary Atresia, Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries (PA/VSD/MAPCAs)

Pulmonary atresia (PA), ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs) is a rare type of congenital heart defect, also referred to as Tetralogy of Fallot with PA/MAPCAs. Tetralogy of Fallot (TOF) is the most common cyanotic heart defect and occurs in 5-10% of all children with congenital heart disease. The classic description of TOF includes four cardiac abnormalities: overriding aorta, right ventricular hypertrophy (RVH), large perimembranous ventricular septal defect (VSD), and right ventricular outflow tract obstruction (RVOTO). About 20% of patients with TOF have PA at the infundibular or valvar level, which results in severe right ventricular outflow tract obstruction. PA means that the pulmonary valve is closed and not developed. When PA occurs, blood can not flow through the pulmonary arteries to the lungs. Instead, the child is dependent on a patent ductus arteriosus (PDA) or multiple systemic collateral vessels (MAPCAs) to deliver blood to the lungs for oxygenation. These MAPCAs usually arise from the descending aorta and subclavian arteries. Commonly, the pulmonary arteries are abnormal, with hypoplastic (small and underdeveloped) central and branch pulmonary arteries and/ or non-confluent central pulmonary arteries.

Physical Exam/Symptoms:

- There is usually no audible murmur and a single, loud S2.
- Cyanosis (blue color) and hypoxemia is present at birth. Cyanosis may be less severe in neonates with a PDA and/or extensive MAPCAs.
- Rarely, children with significant MAPCAs and excessive pulmonary blood flow develop symptoms of congestive heart failure (CHF), such as tachypnea (fast breathing), tachycardia (fast heart rate), poor feeding and growth.

Diagnostics:

- Chest X-ray: The heart is normal in size and is often boot-shaped. Pulmonary vasculature is usually diminished.
- EKG: Right atrial dilation and right ventricular hypertrophy are present due to right ventricular outflow tract obstruction.
- Echocardiogram: Diagnostic.
- Cardiac Catheterization or Computed Tomography Angiogram (CTA): Useful in determining the origin of each MAPCA and which part of the lung it supplies with blood.

Medical Management/Treatment:

- For infants that are diagnosed with PA/VSD/MAPCAs in utero, it is recommended that delivery take place in a tertiary care hospital with transfer to the Intensive Care Unit as soon as possible to initiate cardiology evaluation and medical interventions.
- Prostaglandin E infusion is started as soon as possible after birth or diagnosis to keep the ductus arteriosus open prior to surgery.
- Cardiac catheterization or CTA is performed soon after birth to determine the anatomy of the pulmonary arteries and MAPCAs.
- Surgical repair in the first week of life is needed to establish reliable pulmonary blood flow. Surgical options and timing will be discussed with you by your child's cardiologist. Most children with PA/VSD/MAPCAs require several staged surgeries early in infancy/childhood.
- Children with a right ventricle to pulmonary artery conduit may require surgical replacement of the conduit during childhood, adolescence, and/or adulthood.
- Bacterial endocarditis antibiotic prophylaxis is required prior to any dental procedures.
- Some degree of activity restriction is often required due to exercise intolerance. Your child's cardiologist will discuss this with you.
- Life-long cardiology follow up is needed.

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Long-Term Outcomes:

- Infants without adequate pulmonary blood flow who do not have surgery usually do not survive.
- Children with significant MAPCAs may develop hemoptysis (coughing up blood) in late childhood/adolescence.
- Growth and developmental outcomes vary depending on severity of disease and the presence or absence of other co-morbidities. Follow up with Neurodevelopmental specialists during infancy and childhood is recommended.