Pulmonary Hypertension Associated with Congenital Heart Disease

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Most cardiac defects s can now be diagnosed during fetal life. 
After birth the defect is confirmed with an echocardiogram (ECHO).
If the lesion is associated with the risk of increased pulmonary blood flow, monitoring for pulmonary hypertension begins.
Children are monitored for both an increase in baseline pulmonary artery pressures as well as a propensity to have a pulmonary vascular bed that is capable of constricting suddenly and severely precipitating what is commonly called a pulmonary hypertensive crisis.
During a crisis, the abrupt elevation in pulmonary artery pressure abruptly increases right ventricular stress to fail. The failing RV cannot eject blood into the lungs to preclude the left ventricle and with decreased systemic cardiac output, the child’s blood pressure plummet.

Signs and Symptoms
- History of heart defects, underlying lung disease, syncopal episodes with activity or stress (Park, p. 491)
- Physical assessment: increased right ventricular impulse, loud second heart sound, cyanosis, s/o of right sided heart failure such as edema or hepatomegaly, murmurs that is associated with pulmonary hypertension, diaphragmatic clubbing (Park, p. 491)
- Children with pulmonary hypertension may have dyspnea , fatigue, syncopal episodes (Park, p. 491)
- Radiology exams - x-ray studies show normal to slightly enlarged heart and pulmonary artery. EKG demonstrates right ventricular hypertrophy. ECHO may show a large RV and RA, thickened RV, pulmonary regurgitation and distal proximal pulmonary arteries (Park, p. 491)
- Cardiac Catheterization: directly measures the pressure within the RV and RA as well as the pulmonary bed “Right heart catheterization is still the gold standard for the diagnosis of PAH and hemodynamic evaluation for surgical correction and medical treatment of CHD patients” (Fang et. al, p. 1066)
- Clinical signs of a child experiencing a pulmonary hypertensive crisis include abrupt rise in RA and pulmonary artery pressures, decreased systemic blood pressures, decreased heart rate, ST segment changes, decreased cerebral saturations, and decreased pulse oximetry.

Underlying Pathophysiology
- Prolonged exposure to high volume shunts prevents the normal remodeling of the pulmonary arteries that occur after birth.
- Rather than thin walled vessels in the normal lung, arteries in PAH become progressively thicker making the lumen of the vessel progressively smaller.
- “Progressive endothelial dysfunction, medial hypertrophy with hyperplasia of smooth muscle cells, increased connective tissue and elastic fibers, intimal and adventitial thickening and rarefaction of the pulmonary arterial tree” (D’Alto et al. E4, p. 3). 
- When looking at any disease process, depending on when the disease is found and how quickly the process is evolving, determines the possibility of reversibility. At the end stage of the disease process following long-term exposure to high volume shunts, the PAH may be irreversible.

References
Otterbein University, Westerville, Ohio.