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# Pulmonary Hypertension Associated with Congenital Heart Disease

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# Pulmonary Hypertension Associated with Congenital Heart Disease

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## Introduction

- Pulmonary arterial hypertension (PAH) is a medical condition resulting in alterations in the small arteries of the lung.
- There are many causes of PAH including developmental abnormalities of the heart leading to increased pulmonary blood flow.
- After birth, in the normal heart, the blood flow is in series and does not "mix".
- Specific cardiac defects involve connections between the pulmonary and systemic circulation allowing blood to flow into the lower resistance system.
- As the resistance in the pulmonary circulation falls following birth and clamping of the umbilical cord, connections between the two systems allow oxygenated blood returning from the lungs to recirculate back to the lungs rather than being delivered to the body.
- Over time, pulmonary vasculature with high blood flow contributes to muscularization of the normally thin walled pulmonary arteries resulting in elevated pressure and increased vascular resistance – the hallmarks of PAH.
- Depending upon the cardiac lesion and the timing of repair, surgical closure of these connections may not improve PAH that is long-standing.

## Why Researched

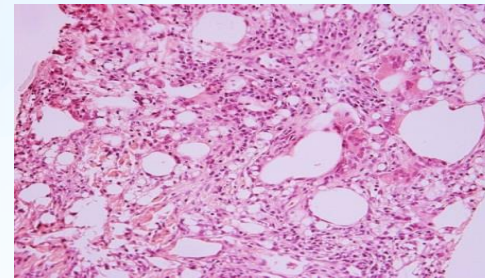
- This topic is extremely relevant to the field in which I work, as well as my future career as a cardiac nurse.
- I work as a Registered Nurse (RN) in the Heart Center Float Pool at Nationwide Children's Hospital.
- Children with CHD can in a matter of minutes have situations of life and death when they have pulmonary hypertension and develop a crisis.
- It is very important to be up to date on the education and information that is available for pulmonary hypertension in order to educate families on the disease.

## Presentation of Case

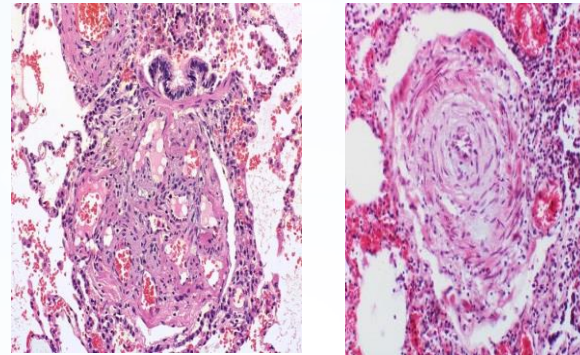
- Most cardiac defects can now be diagnosed during fetal life.
- After birth the defect is confirmed with an echocardiogram (ECHO).
- If the lesion is associated with the risk of increased pulmonary blood flow, monitoring for pulmonary hypertension begins.
- Children are monitored for both an increase 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 stuns the right ventricle causing it to fail. The failing RV cannot eject blood into the lungs to preload the left ventricle and with decreased systemic cardiac output, the child's blood pressure plummets.

## 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/s of right sided heart failure such as edema or hepatomegaly, murmur that is associated with pulmonary regurgitation, differential 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 dilated proximal pulmonary arteries (Park, p. 491)
- Cardiac Catheterization- directly measures the pressure within the RA and RV 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. El, 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.



Normal lung tissue



lung tissue with pulmonary hypertension

## Underlying Pathophysiology

- Prolonged exposure to high volume shunts prevents the normal remodeling of the pulmonary arteries that occurs 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. El 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.

## Significance of Pathophysiology

- Diagnosis of the disease is extremely important since changes in pathophysiology occur frequently and abruptly.
- Changes to the pulmonary bed and the regurgitation of the pulmonary valve cause right ventricular stress and ultimately failure.
- The RV adapts to the stress of PAH with hypertrophy or thickening of the ventricular wall leading to decreased compliance with each contraction and inadequate pumping.
- "Right ventricular contraction occurs in a peristalsis-like manner, beginning with contraction of the inlet portion and ending with contraction of the infundibulum" (Bronicki et. El, p 15)
- RV hypertrophy and pressure affect the left ventricle's ability to fill and eject normally.
- Over time, the RV severely compresses the LV leading to progressive decline and death.

## Implications for Nursing

- Avoid and limit strenuous exercise. When looking at this aspect for children it can be complicated since eating and playing are what they do. (Park, p. 494)
- Administer oxygen as needed (Park, p. 494)
- Administration of medications as prescribed, avoidance of abrupt discontinuation of medications (Park, p. 496)
- Ensure that when patient is on inhaled Nitric Oxide (iNO) that both the Respiratory therapist (RRT) and nurse collaborate for cares and administration of iNO.
- Sedation and analgesia for painful or stimulating procedures

## Conclusion

- Pulmonary hypertension is a rare complication of congenital heart disease in children.
- The clinical symptoms, evaluation, and prognosis varies greatly from adults.
- Understanding the pathophysiology and the available medical therapies and interventions allow us to anticipate and prevent crisis and stabilize or reverse the early stages of PAH.

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