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An Exploratory on Partial Anomalous Pulmonary Venous Return (PAPVR)

Gabriella Pitingaro
Grand Valley State University

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Grand Valley State University
Gabriella Pitingaro
HNR 499
Dr. Strickler

An Exploratory on Partial Anomalous Pulmonary Venous Return (PAPVR)

INTRODUCTION

Partial anomalous pulmonary venous return (PAPVR) is a congenital heart defect where instead of one or two pulmonary veins returning blood back to the left atrium from each lung, some blood is instead returned back to the right atrium (University of Wisconsin, 2015). This change in directionality causes oxygen-rich and oxygen-poor blood to mix before ultimately draining into the right ventricle. PAPVR is very rare, affecting only an estimated 0.4-0.7% of the population (Berman, 2020). This is likely to be an underestimated value due to the fact that the majority of PAPVR cases are found at autopsy. 80-90% of PAPVR patients also suffer from atrial septal defect (ASD), a hole in the wall/ septum between the atria of the heart (El-Kersh, 2019). PAPVR has a higher prevalence in the female population and there is seemingly no genetic predisposition. PAPVR also affects the vessels from the right lung more than the left lung with the most common defect being an upper pulmonary vein, carrying oxygenated blood, connecting to the SVC (Superior Vena Cava) or right atrium. In a normal system, oxygen-depleted blood is brought to the right atrium via the IVC (Inferior Vena Cava) and SVC. The blood then flows through the tricuspid valve, draining into the right ventricle, out into the pulmonary arteries, and ultimately into the lungs to be oxygenated. This oxygenated blood normally flows back into the left atrium of the heart, then through the bicuspid valve to the left ventricle and then out the aorta to the body. The altered course of blood flow in PAPVR causes viable oxygen-rich blood to flow back into the lungs rather than to the left atrium and then to the rest of the body. The flow of oxygenated blood back to the lungs is inefficient. If this condition is not addressed, the constant higher-pressure flow of oxygenated blood between the lungs and right atrium can cause dilation of the right atrium, possibly causing an abnormal heartbeat (arrhythmia) or increased pressure in the blood vessels of the lungs (pulmonary hypertension).

PAPVR has some forgiveness in that if only one of the pulmonary veins is affected, no symptoms are shown, but if both are compromised, that is a different story (El-Kersh, 2019).

EMBRYOLOGY

From an embryological standpoint, the primordial lung bud drains blood into the systemic veins during days 27-29 of gestation. Once days 32-33 arise the left atrium gives off the common pulmonary vein, which connects to the pulmonary vascular bed. This connection then leads to the systemic drainage regressing and eventually disappearing around day 40 of gestation. At this point, failure of the pulmonary vein or veins to completely connect with the common pulmonary vein leads to the onset of PAPVR. In a nutshell, PAPVR arises when the primitive lung drainage does not regress correctly and connect with the common pulmonary vein (Sears, 2012).

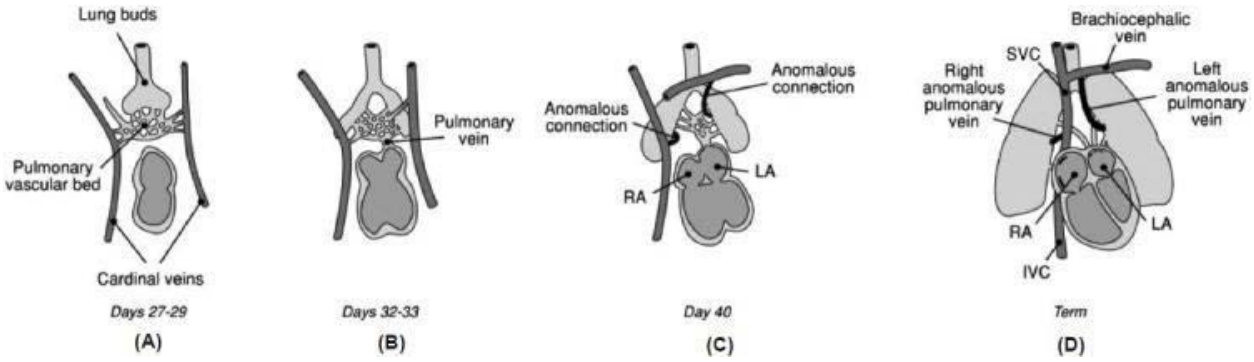

Fig 1. Development of the Pulmonary Veins. 2012. Partial Anomalous Pulmonary Venous Return Presenting with Adult-Onset pulmonary Hypertension. *NCBI*

CASE STUDY-AMY GRANT

In Amy's situation, she had no idea that she had PAPVR. The discovery of her condition only occurred when she attended a heart checkup with Dr. John Bright Cage in December of 2019 due to her family history of cardiac issues (Berk, 2020). The main reason Amy went in beside family history was an irregular heartbeat she had for the past 10 years. It began to affect her singing career and created a tight/suffocating feeling in her chest. Grant described the sensation as

"everything kind of tightening up as I was trying to sing. I remember a couple times telling Vince, 'I feel like I'm suffocating.' It's the weirdest thing, I'm breathing as deep as I can, but in my mind none of that had to do with my heart."

The checkup, however, found more than just an irregular heartbeat. Instead. Dr. Cage found Partial Anomalous Pulmonary Venous Return (PAPVR). Being 59 at the time of her diagnosis, her team pushed her to get surgery to fix the birth defect. With her approval, a surgery date of June 3, 2020 was scheduled. During the surgery, the sternum of the patient, Grant, was cut open to expose the heart, while the blood was redirected by bypass. Cardiopulmonary bypass (CPB) is a procedure where a machine takes over the function of the cardiovascular and pulmonary systems during surgery to make sure oxygen and blood still get to circulate. The anomalous venous connections were removed, Amy made a complete and full recovery and is now continuing on with her career (Berman, 2020).

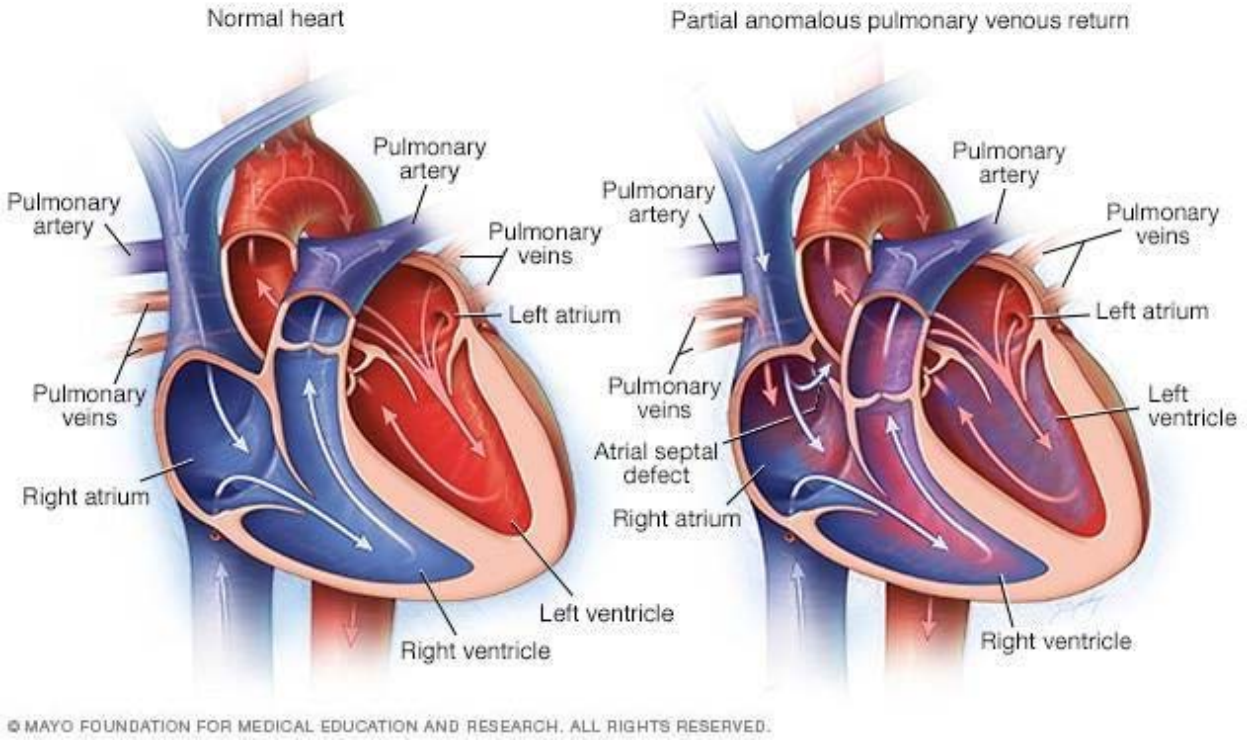

Fig 2. Normal Heart Versus PAPVR. 2020. Congenital Heart Disease in Adults. *Mayo Clinic.*

In this image the differences between a normally developed heart and a heart with PAPVR are shown. The mixing of oxygen-rich with oxygen-poor blood occurs prior to draining to the right atrium. The blue shading in the left ventricle of the PAPVR heart does not indicate the presence of deoxygenated blood, but is meant to show that oxygenated flow from the right lung is decreased because of PAPVR.

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