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Case Study: Partial Anomalous Pulmonary Venous Return Of The Right Upper Pulmonary Vein Into The Cephalic Portion Of The SVC Causing A Left-To-Right Shunt In 56-Year-Old Asian Female

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Abstract

Partial Anomalous Pulmonary Venous Return (PAPVR) is a disease that is often asymptomatic in children and may not present until later in adulthood either symptomatically, or as an incidental finding on radiology or autopsy. When considering the configuration of anatomic congenital anomalies, it is important to appreciate the embryological development in both cardiology and pulmonology. The most common presentation of PAPVR involves the right upper pulmonary vein as it drains into the junction of the superior vena cava (SVC) and right atrium creating a left-to-right shunt which results in freshly oxygenated blood from the lungs being returned to, and mixed with, deoxygenated systemic blood in the right atrium. In addition, an atrial septal defect (ASD) is frequently seen in patients with PAPVR.

Case report

In the case presented, a 56-year-old Asian female was found to have a rare configuration of PAPVR of the Right Upper Pulmonary Vein that drained into the cephalic portion of the SVC instead of the more common right atrium-SVC junction and was not associated with an ASD as previously described. This patient required a modified Warden's procedure for successful surgical correction. While most cases are asymptomatic and the result of an incidental finding, the patient presented to the Emergency Department with a life-threatening right atrial pressure of 80 mmHG, which is clinically significant for pulmonary arterial hypertension (PAH) and caused the patient to go into Right-Sided Heart Failure. The patient also presented with significant right ventricular hypertrophy, as the result of consistent PAH.

Introduction

Embryology

Partial Anomalous Pulmonary Venous Return (PAPVR) is a highly variable disease that is often asymptomatic in children and may not present until later in adulthood either symptomatically, or as an incidental finding on radiology or autopsy. In most cases of PAPVR, the right upper pulmonary vein abnormally drains into the junction of the superior vena cava (SVC) and right atrium, creating a left-to-right shunt. There is almost always an associated Atrial Septal Defect (ASD) present in PAPVR **[1]**. Therefore, an understanding of the normal development of both the pulmonary and cardiovascular systems becomes essential to appreciate the functional consequences of PAPVR. Lung development begins at approximately 26 days of gestation, starting with an out-pouching of the primordial pharynx that gives rise to a bud-like structure called the laryngotracheal diverticulum. The laryngotracheal diverticulum eventually goes on to form the respiratory bud which will develop into the left and right lung buds. Successive branching of the lung buds eventually leads to the lobar shape seen in the adult lungs **[1-2]**. The communicating cardinal and umbilicovitelline veins form the initial route of pulmonary venous drainage. At approximately 27 to 29 days of gestation, an outgrowth from the posterosuperior aspect of the dorsal wall of the primitive left atrium gives rise to the primordial pulmonary vein. This primordial pulmonary vein will begin to communicate with the pulmonary venous portion of the splanchnic plexus, while the systemic veins simultaneously involute and degenerate. As the atrium expands, the primordial pulmonary vein and its main branches are incorporated into the left atrium giving rise to a smooth-walled left

atrium and four separate pulmonary veins **[1-2]**. In a normally developing embryo, this complete dissociation will render four separate pulmonary veins, all of which drain into the left atrium and establish optimal drainage of oxygenated blood from the developed lungs into the adult heart for redistribution to the head, and neck, heart, and rest of the body. In the occurrence of Partial Anomalous Pulmonary Venous Return (PAPVR), a failure of this separation process results in one or multiple pulmonary veins draining into an inappropriate structure. An overview of the embryologic development process can be seen below in (**Figure 1)**. The most common presentation of PAPVR involves the right upper pulmonary vein and drains into the junction of the superior vena cava (SVC) and right atrium, creating a left-to-right shunt, and there is almost always an associated Atrial Septal Defect (ASD) present **[1]**. The estimated overall incidence of PAPVR is 0.5 % **[3]**, while a slightly higher incidence of 0.7 % **[4]** is reported by autopsies. This discrepancy in statistics can most likely be explained by the asymptomatic nature of most PAPVR cases until later in adulthood. To further understand the etiology of PAPVR, the embryological development of the SVC must also be understood. The SVC is derived from the proximal portion of the right anterior cardinal vein and the right common cardinal vein at a point that is caudal to the transverse anastomosis within the embryo. At approximately the 8th week of gestation, a large anastomosis formed between the thymic veins and thyroid veins shunts blood from the left anterior cardinal veins towards the right side. The anterior cardinal veins are responsible for draining the cephalic portion of the embryo, while the posterior cardinal veins drain the remaining caudal portion. The anterior and posterior cardinal veins merge before entering the sinus venosus, giving rise to the common cardinal veins. The left anterior and posterior cardinal veins will degenerate and involute, while the right anterior and posterior cardinal veins remain. The inferior portion of the anterior cardinal vein will fuse with the right common cardinal vein to form the adult SVC that ultimately drains into the Right Atrium **[5-6]**.

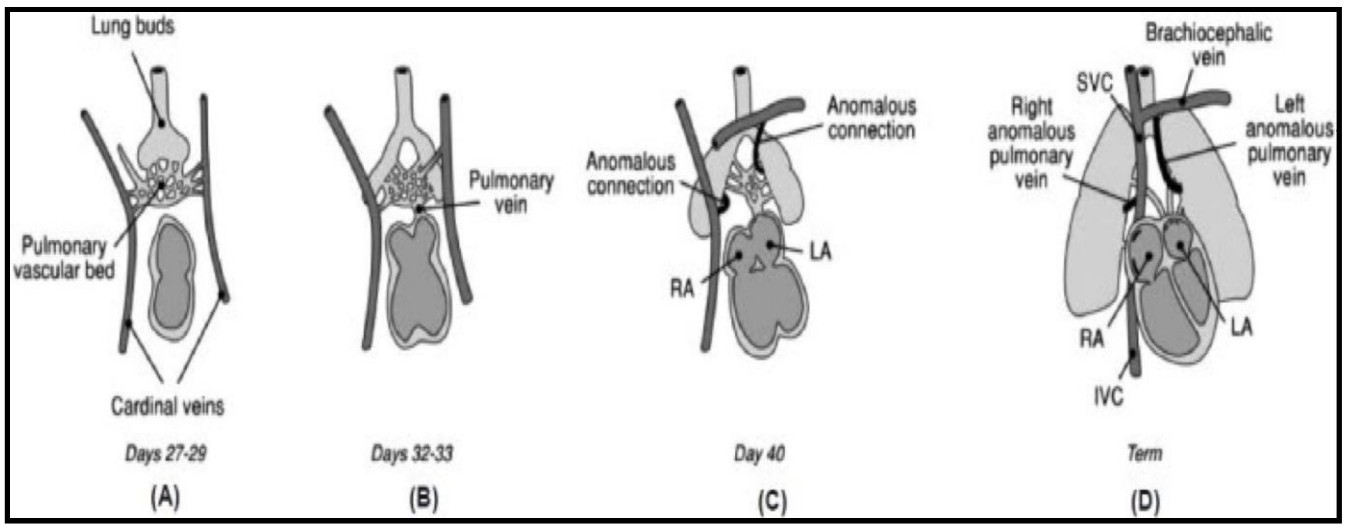

Figure 1 (above): (A) At post-conceptional Days 27–29, the primordial lung buds drain through a vascular bed to the cardinal veins, which will develop into systemic veins. (B) By Days 32–33, the common pulmonary vein forms from the left atrium and establishes a connection with the pulmonary venous circulation. Pulmonary venous connections to systemic veins begin to regress and pulmonary venous blood drains into the common pulmonary vein. (C) By Day 40, the primitive connections from the pulmonary vascular bed to the cardinal veins should have regressed, but in PAPVR anomalous connections persist. (D) At term, the anomalous connection will have developed into anomalous pulmonary veins draining most commonly into the SVC on the right, or the brachiocephalic vein on the left **[7].**

Clinical Presentation: In the case presented, a 56-year-old Asian female was found to have PAPVR of the right upper pulmonary vein that drained into the cephalic portion of the SVC. The MRI image demonstrates the aberrant pulmonary venous drainage **(Figure 2).** This type of PAPVR causes a left-to-right shunt, where freshly oxygenated blood from the lungs is returned to, and mixed with, deoxygenated systemic blood in the right atrium. While having a right upper pulmonary vein drain into the SVC is the most common PAPVR, this case is rare as it is not associated with an ASD as previously described **[8]**. The patient instead presented with a clinically insignificant Patent Foramen Ovale (PFO) which is not considered a true ASD as no true structural deficiency of the atrial septal tissue is present, but rather there is an incomplete fusion of the normally developed septum primum and septum secundum **[9]**. The abnormal site of attachment of the right pulmonary vein was noteworthy. While in most cases this vein will attach at the junction of the SVC and right atrium, in this case, the vein was draining into the cephalic portion of the SVC alone. This patient required a modified Warden's procedure for surgical correction. A classic Warden's Procedure will involve the transfer of the SVC to the Right Atrium (RA) appendage, while the aberrant pulmonary veins are redirected to the left atrium **[10]**. This patient did not have a VSD, but that would also be corrected during the procedure. Because of the left-to-right shunt that is established by PAPVR, a significant enough unrepaired shunt can result in pulmonary vascular remodelling that leads to the development of Pulmonary Artery Hypertension (PAH) **[11]**. The patient presented to the Emergency Department with a right atrial pressure of 80 mmHG, which is clinically significant for PAH and resulted in the patient going into Right Heart Failure. The patient also presented with significant right ventricular hypertrophy as the result of consistent pulmonary arterial hypertension. The cause of

PAH and right ventricular hypertrophy is the result of an increase in blood volume being introduced to the right side of the heart, resulting in right-sided volume overload and leading to PAH and ventricular hypertrophy **[8]**. The patient's surgery was successful, and they are now being managed with classes of drugs known as Beta Blockers and Phosphodiesterase Inhibitors to alleviate any residual RA and pulmonary hypertension.

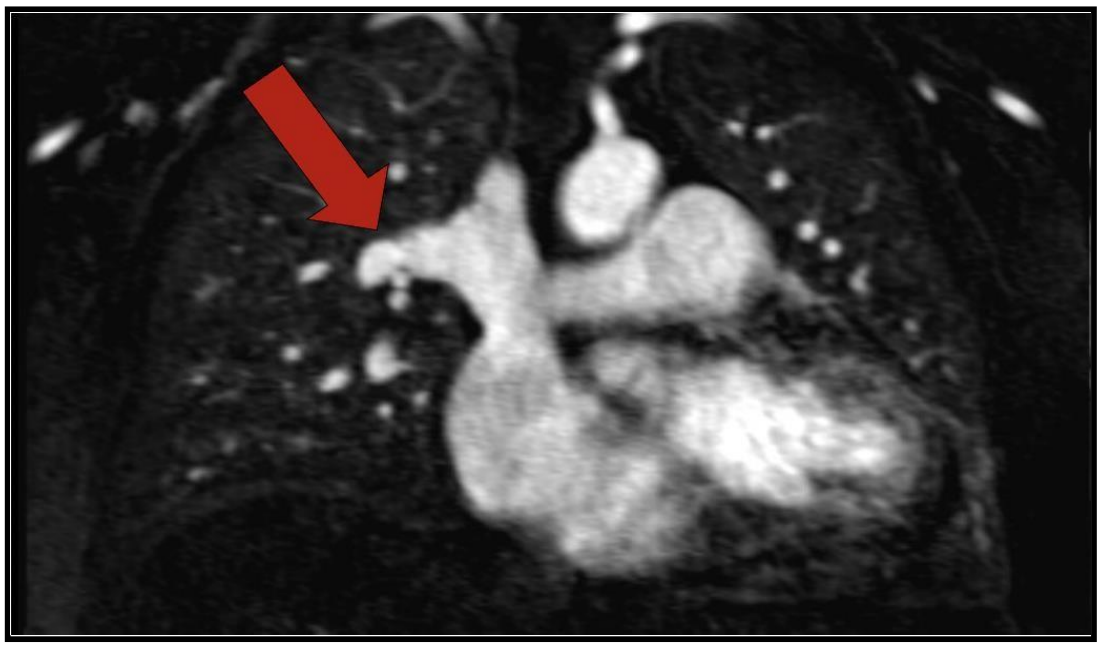

Figure 2 (Above, MRI coronal): Red arrow depicts aberrant right upper pulmonary vein draining into the SVC. Right sided PAPVR is more commonly diagnosed in children. PAPVR in this case-report is right sided and was not diagnosed until symptomatic onset later in adulthood. The thought is that children are more often symptomatic and will present earlier in life as a result. Also, secondary pulmonary hypertension is typically a rare complication, but was seen in this case resulting in Right Ventricular (RV) enlargement noted on MRI above, as well as the case-report symptoms (Shortness of breath, hypertension, etc.) Images courtesy of Dr. Shackles.

Author Contributions:

Wesley Banks, MS: First author and main writer of the article. Gathered resources and compiled the final applications into the text.

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