

Pulmonary vein abnormalities into the human left atrium

Sharleen Yuan, Natalie Mushitz, Stuart D. Inglis

ABSTRACT

Introduction: Abnormalities in the number and size of pulmonary veins are not common. However, incidences that do arise often are the result of congenital or acquired disorders. **Case Report:** We present a case report on abnormal pulmonary veins draining into the left atrium, which was uncovered during dissections in gross anatomy. **Conclusion:** During a single anatomy dissection class, three separate cadavers demonstrated anomalies in pulmonary vein number. In the first instance, an 84-year-old female presented with a single, dilated pulmonary vein connecting the left atrium to the left lung. A second cadaver of an 85-year-old female had a similar manifestation of a single left pulmonary vein. In the third case, a 70-year-old male presented with three pulmonary veins draining into the left atrium.

Keywords: Acquired defects, Congenital defects, Pulmonary vein abnormalities, Pulmonary vein

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INTRODUCTION

One of the principle benefits of anatomical dissection for medical students is the development of appreciation for anatomical variability and for common and unusual deviations from standard textbook depictions of anatomical structure. In the course of a single term, deviations in left pulmonary vein number were observed in 3 of 17 cadavers dissected by pre-clinical medical students at the Sanford School of Medicine. In the body of an 84-year-old female, a single dilated pulmonary vein was found connecting the left lung to the left atrium (Figure 1). Two normal pulmonary veins returned to the left atrium from the right lung. No other deviation was detected in regards to the pulmonary or cardiac anatomy. Gross anatomical dissection of an 85-year-old female revealed an identical deviation in pulmonary vein structure, with a single dilated left pulmonary vein and otherwise normal cardiovascular anatomy (Figure 2). In a third cardiovascular dissection of a 70-year-old male, three left pulmonary veins were observed (Figure 3A–B), with no additional pulmonary or cardiac abnormalities being detected. In all three instances, these abnormalities in pulmonary vein numbers appear to be congenital.

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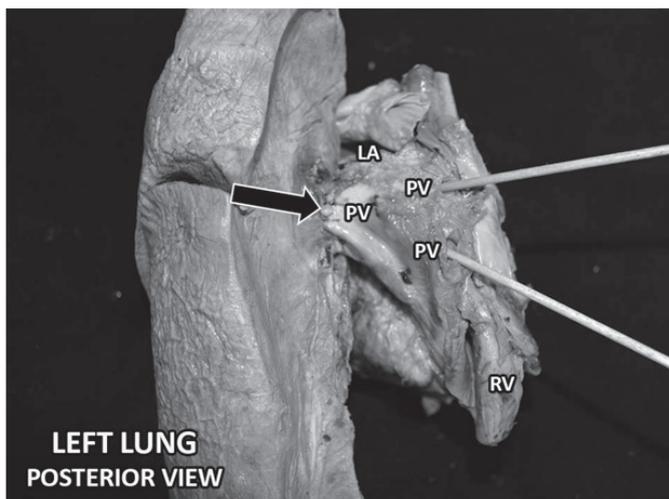


Figure 1: Case 1 Posterior view of left lung and heart of an 84-year-old female. An abnormal, single pulmonary vein connecting the left lung to the left atrium (black arrow). The pulmonary veins that connect to the right lung (indicated with the wooden sticks) are shown and appear to be normal. LA = left atrium, LT PV = left pulmonary vein, RT PV = right pulmonary vein, RV = right ventricle.

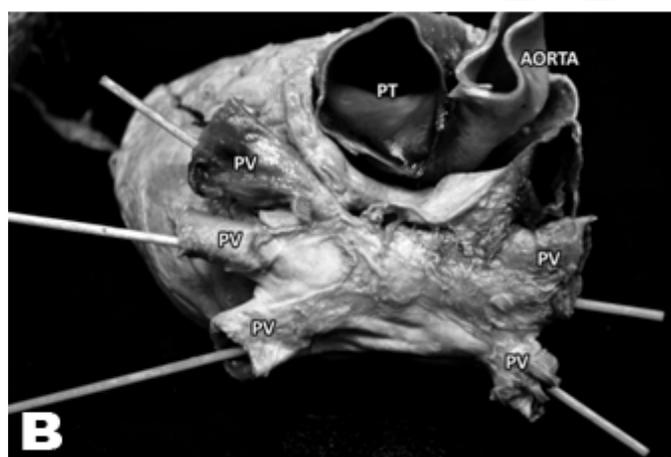
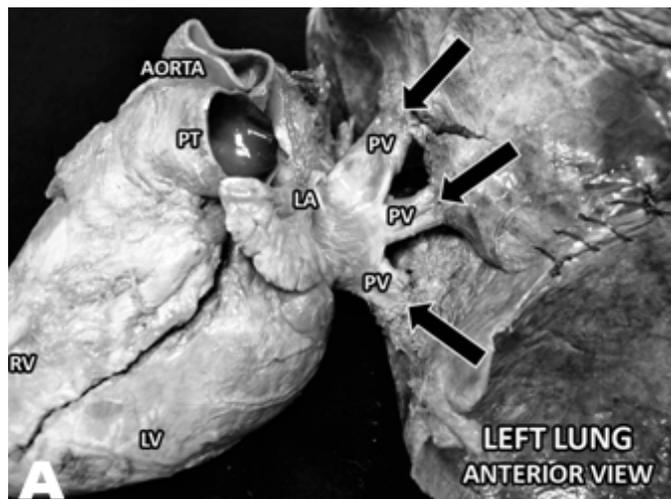


Figure 3: (A) Case 3 Anterior view of the left lung and heart of a 70-year-old male. Three abnormal pulmonary veins are observed connecting the left lung to the left atrium (black arrows). Two normal pulmonary veins connected the right lung to the left atrium (not shown). PT = pulmonary trunk, RV = right ventricle, LV = left ventricle, PV = pulmonary vein, LA = left atrium, (B) Case 3 View of the heart alone of a 70-year-old male. Three pulmonary veins connecting the left lung and two pulmonary veins connecting the right lung are shown anastomosing with each other and draining into the left atrium. PV = pulmonary vein, PT = pulmonary trunk.

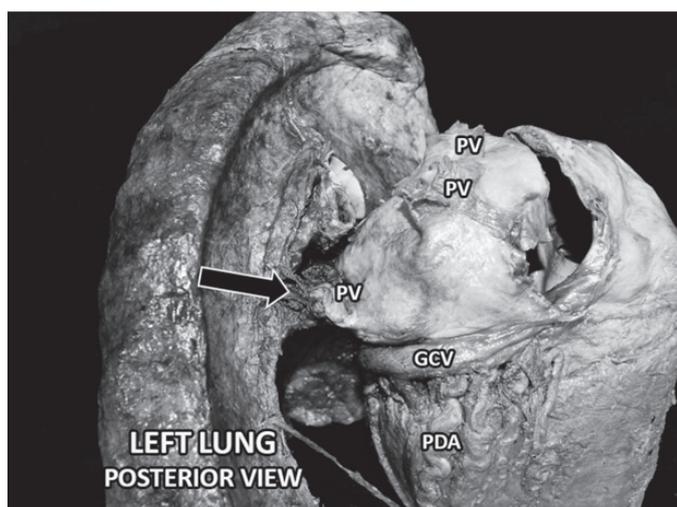


Figure 2: Case 2 Posterior view of the left lung and heart of an 85-year-old female. An abnormal, inferior, and single pulmonary vein is observed to connect the left lung to the left atrium (black arrow). Superiorly, two pulmonary veins can be observed that connected the right lung to the left atrium. PV = pulmonary vein, GCV = great cardiac vein, PDA= posterior descending branch of the right coronary artery.

CASE REPORT

Normal Pulmonary Vein Anatomy

Research into the precise development of the pulmonary veins is ongoing. Recent reports suggest that the pulmonary vein initially forms as a distinct channel

within the posterior mediastinum between the left and right pulmonary ridges [1, 2]. During atrial septation in the fourth week, the primordial pulmonary vein dilates and fuses with the posterior wall of the newly differentiated left atrium [2], although recent literature proposes an initial connection to the primitive sinus venosus of the developing heart tube [1]. As the vein increases in length, it starts to branch. The first bifurcation yields left and right branches, which begin to project towards the respective developing lung buds. The second bifurcation, seen in both left and right branches, yields superior and inferior branches which later become the four pulmonary veins. The inferior branches serve to drain the lower lobes on both the left and right sides. The right superior branch

drains the right superior lobe while the left superior branch divides further to drain the superior and middle lobe [2]. Ultimately these projections anastomose with the pulmonary venous plexus to establish pulmonary venous return with the left atrium. The 5th week of gestation is marked by a period of intussusception, in which left ventricular expansion favors the posterior wall over the constricted anterior wall. As a result of this expansion, the primordial pulmonary vein expands and contributes to the posterior wall of the left atrium [2]. Intussusception progresses with complete incorporation of the primordial pulmonary vein and subsequently the initial left and right branches, terminating with partial incorporation of the left and right superior and inferior branches. The result is a smooth posterior atrial wall derived from the initial segments of the primordial pulmonary vein and four orifices emptying into the left atria, consistent with the two superior and two inferior branches [1]. In most cases, the unitary pulmonary vein develops on the posterior atrial wall and its branches are fused into the left atrium. In embryogenesis, the degree of integration of the common pulmonary vein branches may be due to variations of pulmonary vein opening into the left atrium [9].

By the end of embryogenesis, the adult pulmonary circulation system is fully formed. The main pulmonary artery continues out of the right ventricle with the pulmonary valve separating the two structures. The main pulmonary artery, also known as the pulmonary trunk, then splits into right and left pulmonary arteries [3, 4]. The right pulmonary artery branches into the upper, middle, and lower lobe arteries, corresponding to the three lobes of the right lung. These arteries further divide to supply blood to each segment of the right lung. The left pulmonary artery branches into the upper and lower lobe arteries, again corresponding to the two lobes of the left lung [4].

The pulmonary veins follow a course similar to the pulmonary arteries as they return oxygenated blood from the lungs to the left atrium. Their course varies from that of the pulmonary arteries when the right upper pulmonary vein is formed from the joining of the right upper and right middle veins before draining its blood supply into the left atrium. The right lower pulmonary vein continues directly from the lower lobe vein to drain into the left atrium. The left upper pulmonary vein is produced from the merger of the superior segment and lingual veins before draining into the left atrium. The left lower pulmonary vein drains the left lower lobe of the lung into the left atrium to form the fourth principle vein of pulmonary circulation [3].

Congenital Abnormalities with Pulmonary Veins

In many instances, pulmonary vein abnormalities are typically congenital in nature. However, they can

be classified as congenital or acquired conditions. In congenital pulmonary vein abnormalities, most include anomalies in the overall diameter and number of veins. However, irregular flow due to aberrant connections is not uncommon in congenital defects [3]. Acquired defects are typically seen due to obstruction or stenosis, many times due to secondary issues such as hypertension [3]. In most cases, these pulmonary vein abnormalities go unnoticed until discovered in adulthood, usually as an incidental diagnosis [3]. The present cases represent abnormal pulmonary vein numbers, suggesting that the defects were all congenital in nature.

Although rare, previous research has observed anomalous unilateral single pulmonary vein in the human heart. Known as a meandering vein, the single pulmonary vein attaches to the left atrium and unites the pulmonary veins from one lung [5]. This abnormality is connected with other pulmonary disorders such as partial anomalous pulmonary venous return [3]. However, in most cases, very little treatment is necessary for an anomalous unilateral single pulmonary vein.

Another congenital abnormality is the congenital unilateral pulmonary vein stenosis or atresia. In this condition, a complete or partial obliteration of the pulmonary veins are observed unilaterally [6], indicating a failed connection of a pulmonary vein into the left atrium [3]. Congenital unilateral pulmonary vein atresia can result in congenital heart disease or abnormal pulmonary flow in over 50% of the individuals that present with this disorder [6]. Therefore in many cases, the symptoms for congenital unilateral pulmonary vein atresia include chronic pulmonary infection, hemoptysis, and problems during exertion. These congenital disorders could have very easily affected the cadavers presented with the abnormal pulmonary veins. In fact, the cause of death from the 84-year-old heart was myocardial infarction. She presented with a single, dilated pulmonary vein attached to the left lung. Incidentally, it is possible that the increased in diameter of this single pulmonary vein was a compensatory mechanism, resulting in an acquired abnormality such as a pulmonary vein varix. A pulmonary vein varix is commonly seen as a dilation of a pulmonary vein, but does not have an arterial connection [7, 8], with the dilation near the left atrium. A pulmonary vein varix can also be observed as a secondary manifestation, acquired due to pulmonary disease, such as hypertension or valve issues, although most patients can be asymptomatic [3, 7, 8].

In previous research, increased pulmonary vein numbers was more commonly observed on the right side [9]. However, in this case, three pulmonary veins were observed on the left side in the 70-year-old male. This anatomy suggests a variety of clinical implications such as atrial fibrillation by ectopic beats [10]. Atrial fibrillation is one of the most common cardiac arrhythmias, which increases the risk for stroke. Interestingly, a majority of atrial ectopic beats that lead to atrial fibrillation arise in

the pulmonary veins. Individuals with increased number of pulmonary veins could develop an elevated risk of atrial fibrillation-induced stroke because pulmonary veins are identified as a source of these atrial ectopic beats.

CONCLUSION

In three different cadavers, abnormalities were observed in the left pulmonary veins draining into the left atrium. These anomalies included two individuals with a single, unilateral left pulmonary vein and one individual with three pulmonary veins. Unilateral, single pulmonary veins could implicate pathologies such as pulmonary vein stenosis or meandering vein, which increases the risk of pulmonary diseases. In terms of an increased number of pulmonary veins, there is an association of ectopic atrial fibrillation, leading to an elevated risk of stroke.

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Author Contributions

Sharleen Yuan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Natalie Mushitz – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Stuart D. Inglis – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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