

## Partial Anomalous Pulmonary Venous Connection in Adult: A Challenge in Diagnosis

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### Abstract

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital cardiac disease. Affected individuals may present with a spectrum of clinical signs and symptoms depending on the severity of the anomaly, making the diagnosis specifically challenging. Herein, we describe a clinical case of a 25-year old lady who had an uneventful childhood but began experiencing episodic syncopal attack since her late adolescent years throughout her early adulthood. Subsequent referral to the tertiary cardiac centre resulted in series of advanced diagnostic modalities, affirming the diagnosis of PAPVC that was complicated with pulmonary hypertension. She had successfully undergone open heart surgery for pulmonary vein repositioning with excellent post-operative results.

**Keywords:** Hypertension; Pulmonary; Heart Defects; Congenital; Syncope; Pulmonary Veins

### Introduction

Congenital cardiac anomaly amasses a spectrum of clinical presentation. Early detection of cyanotic congenital heart disease allows prompt management and referral to the tertiary cardiac centre with definitive cardiac care. Unfortunately, certain acyanotic congenital cardiac diseases may go undetected during early life and may only manifest at a later stage, often with complications.

Partial anomalous pulmonary venous connection is a rare cardiac defect that describes the blood flow, from usually, a single pulmonary vein, that returns to the right atrium, instead of the left atrium [1]. The worldwide incidence was only at 0.7% of the population [2].

Herein we describe a clinical case of a young adult female presented with recurrent syncopal attack since her late adolescent years, eventually diagnosed with partial anomalous pulmonary venous connection.

### Case Report

A 25 years old lady with recurrent history of syncopal attack since the age of 18 years old was referred to our centre for evaluation. Upon further history, she had been suffering from episodic fainting spells, particularly when she was under physical or emotional stress. The fainting episodes were brief with rapid recovery. They were associated with photophobia and dizziness prior to black-out. Electroencephalogram (EEG) was normal. Her haemoglobin was within acceptable limit. There was no similar history in her family members.

Her weight was appropriate to her height. She was normotensive with normal oxygen saturation. End systolic murmur grade 3/6 was heard best at the upper left sternal edge, with no loud P2.

Her symptoms worsen with progressive reduced in her effort tolerance and frequent episodes of palpitations, affecting her daily activities. Echocardiogram performed revealed a small patent foramen ovale (PFO) at 11 x 11 mm and an anomalous pulmonary vein was noted on the right side with dilated right atrial and right ventricular walls. LVEF was at 60 to 65% (Figure 1).

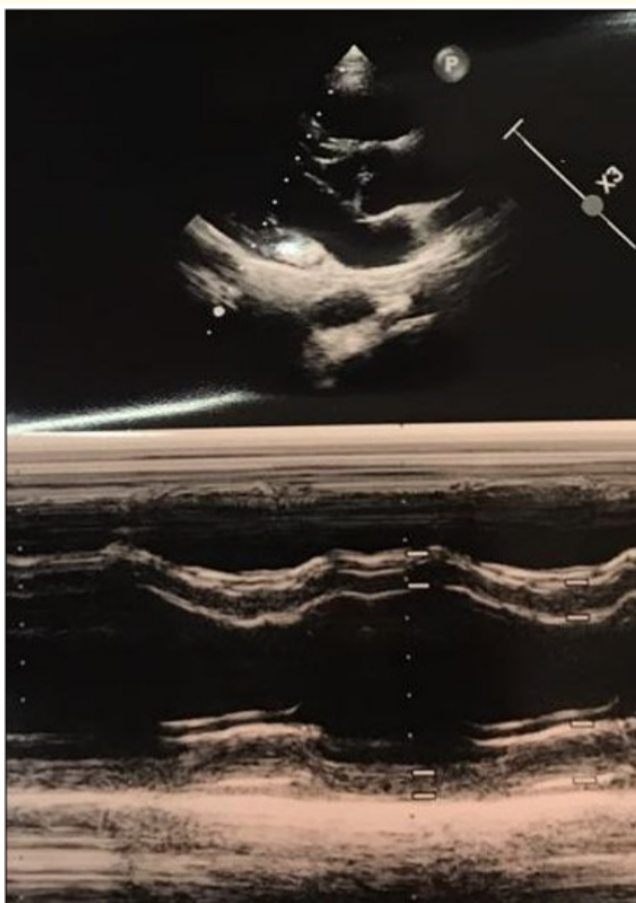


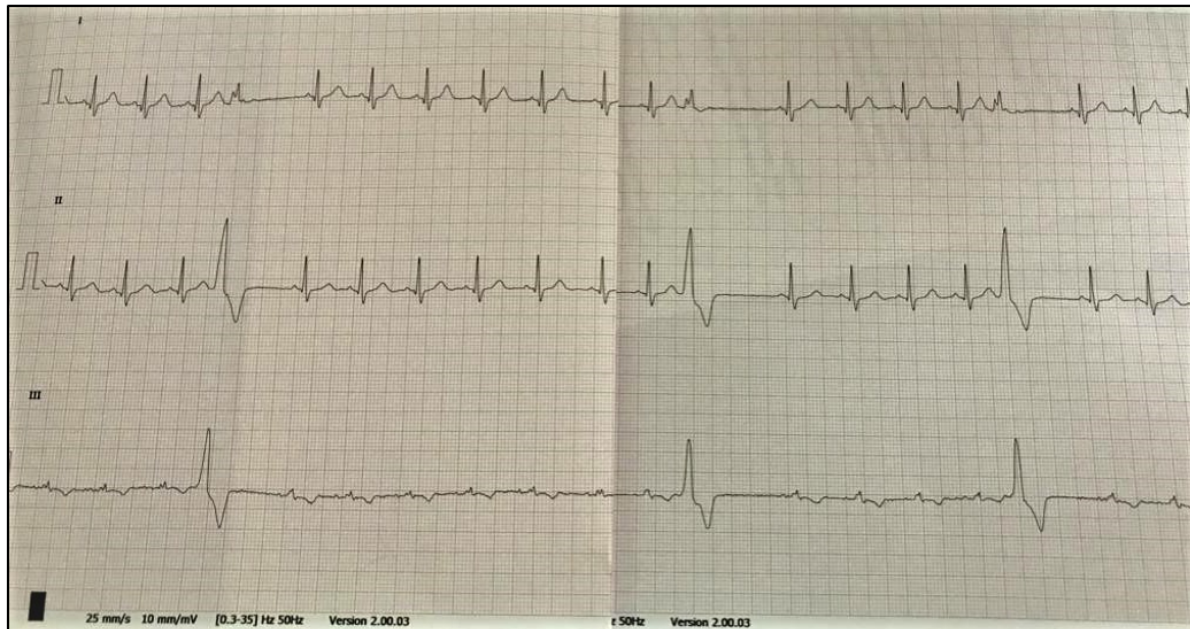
Figure 1: Echocardiogram to assess the dynamic ventricular function.

Computed tomography of the pulmonary artery (CTPA) confirmed an anomalous, single right pulmonary venous draining into infra-diaphragmatic systemic venous circulation, complicated with pulmonary hypertension and mild cardiomegaly (Figure 2).



Figure 2: Chest/abdominal X-ray showing mild cardiomegaly.

Electrocardiogram (ECG) (Figure 3) and Holter was also done revealing multiple premature ventricular complexes (PVCs).



**Figure 3:** ECG showing multiple premature ventricular complexes (PVCs).

She was subsequently referred for pulmonary vein repositioning at a specialized cardiac centre. An open cardiac surgery was performed with excellent outcomes. Post-operatively, she continued to be in cardiac rehabilitative programme and is currently recuperating well. She neither report further episode of palpitation nor black-out episodes.

### Discussion and Conclusion

Partial anomalous pulmonary venous connection is a rare congenital acyanotic heart disease. Despite the growing diagnostic modalities [3], the clinical diagnosis is often masked by other illnesses as the presentation spectrum is so diverse, depending on the number of pulmonary veins involved, the site of anomalous connection, presence of pulmonary venous obstruction and presence of additional cardiac defects [4]. These often complicates clinical scenario resulting in delayed diagnosis, hence treatment. Therefore, it is pertinent to consider congenital anomaly even in adults, even though the disease is commonly attributable to children.

The sensitivity of the echocardiogram may be limited by both the operator skill and experience, and the patient factors, such as the body habitus and the patient's cooperation during the procedure. Computed tomography of the pulmonary artery (CTPA) has emerged as the prime diagnostic modality [5] due to its high sensitivity and its tolerability among patients. Our patient has undergone various diagnostic modalities to evaluate her cardiac status prior to her definitive treatment, which serves to assess her cardiac status prior to the surgical intervention.

Our patient has successfully undergone open heart surgery for pulmonary vein repositioning and is currently recuperating well. Hence, an open-heart surgery is the gold standard in correcting congenital aberrant structural variant of the heart, with excellent outcomes [6].

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