# PARTIAL ANOMALOUS PULMONARY VENOUS DRAINAGE FROM THE UPPER LOBE OF THE LEFT LUNG*

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In partial anomalous pulmonary venous drainage (PAPVD) some of the pulmonary veins do not drain into the left atrium but into the right atrium, coronary sinus or systemic veins. PAPVD from the right lung is fairly common,[1-3] but it is rare from the left lung only. D'Cruz and Arcilla could only find 54 reported cases of anomalous left lung drainage.[4] PAPVD from the left upper lobe alone is even more uncommon: only 18 well-documented cases have been reported in the British and American literature although other isolated cases may have been seen.[5-11] Cases of PAPVD were diagnosed only at autopsy until 1947, when Brantigan recognized the abnormality at operation,[12] and the first clinical diagnosis is attributed to Dotter *et al.*[13]

This report describes an unusual case of PAPVD from the upper lobe of the left lung to the left superior vena cava draining into the innominate vein. The other pulmonary veins entered the left atrium in a normal position. Additional pulmonary valve stenosis and a patent foramen ovale were present.

## CASE REPORT

L.McC. weighed 6½ lb. at birth. This followed an uncomplicated normal pregnancy. She was well during infancy but at the ages of 1½ and 2½ years was admitted to hospital with bronchopneumonia. She was then referred for cardiac consultation.

Clinically she was well developed and well nourished without features of respiratory distress or cardiac failure. A faint systolic thrill was felt along the right sternal border. The first heart sound was normal. The second heart sound was normally split with a loud pulmonary component. A grade 4/6 ejection systolic murmur and early systolic click were heard at the upper left sternal border (Fig 1). An electrocardiogram revealed right axis deviation with right ventricular hypertrophy; an incomplete right bundle-branch block pattern was evident (Fig. 2). The heart was enlarged on the chest X-ray (Fig. 3), with pulmonary plethora and a large mediastinal shadow.

Cardiac catheterization was performed to establish a definitive diagnosis. Routine left and right heart catheterization was undertaken from the right groin and was followed by selective angiocardiography. The intracardiac pressures and blood oxygen saturation are shown in Table I. A high oxygen saturation was present in the superior vena cava. A calculated left-to-right shunt of 30% was present with a pulmonary to systemic blood-flow ratio of 1·5 : 1. A peak systolic gradient of 70 mm.Hg was present

*Date received: 24 January 1969.

TABLE I. CARDIAC CATHETERIZATION FINDINGS

| Site | Oxygen saturation % | Pressure mm.Hg |
|---|---|---|
| Superior vena cava (high) | 82 | |
| Superior vena cava (low) | 82 | |
| Right atrium (middle) | 82 | 1·25 mean |
| Right atrium (low) | 63 | |
| Inferior vena cava (high) | 66 | |
| Right ventricle | 76 | 100/0 - 4 |
| Main pulmonary artery | 76 | 28/12·5 |
| Left pulmonary artery | 74 | 28/12·5 |
| Left atrium | | 5·0 mean |
| Left ventricle | 98 | 120/0 - 3 |
| Aorta | 98 | 120/75 |

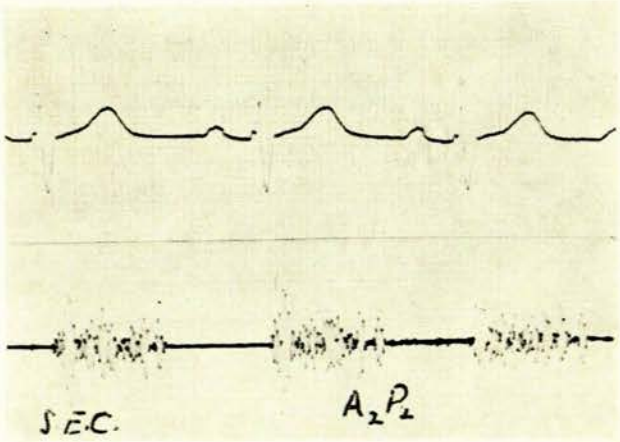


*Fig. 1.* Phonocardiogram showing ejection systolic click (S.E.C.), long systolic murmur and persistent delay of the second heart sound. (Paper speed—50 mm./sec.).

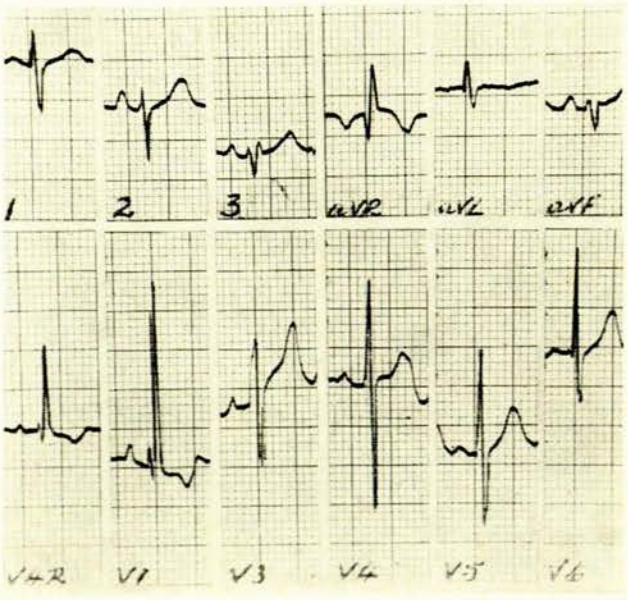


*Fig. 2.* The electrocardiogram shows extreme right axis deviation, delayed intrinsicoid deflection of the right ventricle and right ventricular hypertrophy.

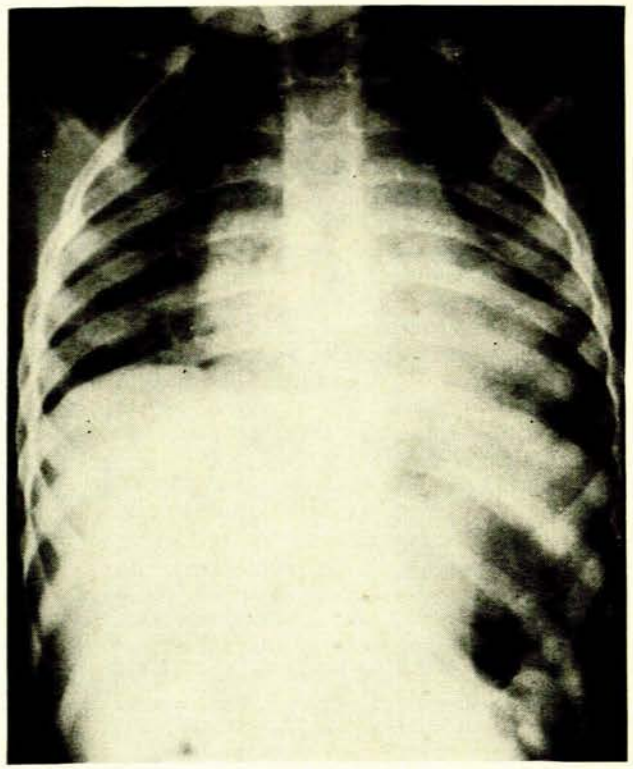

*Fig. 3.* Chest X-ray. Note the wide superior mediastinum due to the ascending left superior vena cava, large superior vena cava, dilated main pulmonary artery and increased pulmonary vascular markings.

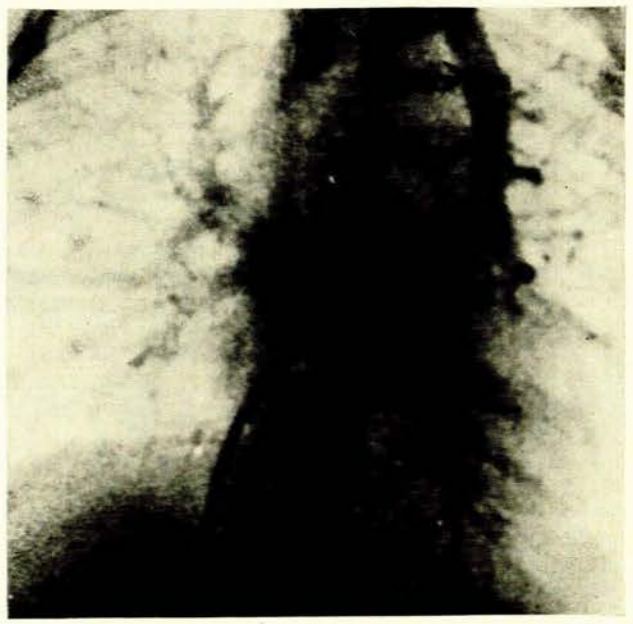

*Fig. 4.* Selected frame from 35-mm. cine-angiocardiogram in the AP position during the pulmonary venous phase, showing 3 pulmonary veins draining normally into the left atrium while the upper left pulmonary vein drains anomalously (arrow).

at pulmonary valve level. The pulmonary vascular resistance was normal.

Selective right ventriculography showed the stenotic pulmonary valve with a thin, central jet of contrast medium emerging through a narrow valve orifice and significant poststenotic dilatation of the pulmonary artery. Contrast medium passed through the lungs and outlined the pulmonary veins. All the right and the left lower lobe veins emptied normally, while the left upper pulmonary vein drained into a left ascending superior vena cava (ascending vertical vein) and then into the innominate vein (Fig. 4). The right atrium filled later from the right superior vena cava, excluding a left-to-right shunt across the atrial septum.

## DISCUSSION

The clinical features of PAPVD and atrial septal defect resemble each other, but the two defects often occur together. The differential diagnosis can be difficult, even after cardiac catheterization, unless each pulmonary vein is entered and its drainage is demonstrated angiographically.

This unusual case demonstrates several interesting clinical features. A tentative clinical diagnosis of pulmonary stenosis was based on the loud, long systolic murmur in the pulmonary area, the ejection click and mobile splitting of the second heart sound, although the phonocardiogram demonstrated persistent delay of a loud pulmonary component even on expiration. The X-ray was unusual and demonstrated a broad superior mediastinum and pulmonary plethora.

The increase in oxygen saturation in the superior vena cava suggested anomalous pulmonary venous drainage, but the exact anatomical diagnosis was dependent on good quality pulmonary venous angiography. In this patient, 3 features excluded a significant atrial septal defect and suggested a patent foramen ovale: the pressure in the left atrium was higher than the right atrium, there was no increase in oxygen saturation in the right atrium and this chamber was not seen on pulmonary angiography until the superior vena cava filled from the anomalous vein.

The clinical features can now be explained: the long systolic murmur and delayed closure of the pulmonary valve were due to valvular pulmonary stenosis of moderate degree accentuated by increased flow across the valve. The pulmonary component of the second sound was loud, suggesting an additional abnormality, but the variation in splitting of S2 excluded a large, non-restrictive atrial septal defect. The pressure-flow relationship in the two atria varied with different phases of respiration so that the second sound was split on expiration, but this increased further on deep inspiration. In the presence of a large atrial septal defect, the two atria are in communication and this respiratory variation is abolished: the delayed P2 is due to the large left-to-right shunt and prolonged right ventricular systole. In isolated PAPVD without an atrial septal defect, right ventricular systole is prolonged by the increase in pulmonary blood flow but respiratory variation in splitting of the second heart sound persists, since free communication is not present between the two atria.

*Embryology*

The embryology is interesting. The normal pulmonary veins develop from a bud which develops from the posterior wall of the common atrium, but which is deflected into the left atrium by the formation of the interatrial septum. The vein grows and divides into 2 and then 4 branches with a tributary from the upper and lower lobes of each lung. The main stem dilates, and is gradually absorbed into the back of the left atrium. The lungs develop from the enteric (alimentary) buds so that their initial efferent venous drainage is into the splanchnic plexus—the cardinal umbilical and vitelline veins. Anomalous pulmonary venous drainage into the systemic venous return persists if the efferent pulmonary veins do not fuse with the pulmonary venous buds derived from the left atrium. In this patient, the left upper zone pulmonary vein did not join the primitive pulmonary venous bud but retained its normal connection to the left anterior cardinal vein. Most of this vein disappeared as the innominate vein usurped the drainage of blood from the left anterior half of the body into the normal SVC. A small branch persisted, however, as the only route of drainage, to return blood from the left upper lobe pulmonary vein.

Corrective surgery in this patient would consist of pulmonary valvotomy and reimplantation of the left upper pulmonary vein into the left atrium after disconnecting it from the left ascending superior vena cava (vertical vein). We have decided to defer surgery in this patient because the child is well, the shunt is small and there is only a moderate gradient across the pulmonary valve.

## SUMMARY

This paper reports a 31-month-old child with isolated partial anomalous pulmonary venous drainage from the upper lobe of the left lung into a left ascending superior vena cava (vertical vein) with normal drainage of the other pulmonary veins. Additional valvular pulmonic stenosis with a peak systolic gradient of 70 mm.Hg and a patent foramen ovale were present. Exact anatomical diagnosis was confirmed by pulmonary angiography. The clinical diagnosis and embryology are discussed.

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