British Heart Journal, 1977, **39**, 685–687

Infradiaphragmatic total anomalous pulmonary venous connection to portal vein
Diagnostic implications of echocardiography

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A case of total anomalous pulmonary venous connection to the portal vein is described. The diagnosis was suspected clinically, supported by the echocardiogram, and confirmed by cardiac catheterisation, angiocardiography, and contrast echocardiography. An echo-free space lying behind the left atrium initially was thought to represent the common pulmonary vein. However, contrast echocardiography showed that this space was not the anomalous vein but probably an artefact. This paper shows that the origins of intracardiac echoes cannot always be assumed from a simple comparison of echocardiography with angiocardiographic or necropsy findings. In some cases it is necessary to introduce a marker into the echocardiogram which unequivocally originates from, and, therefore, localises, the structure under examination. Contrast echocardiography provides such a marker.

Several echocardiographic features are considered of diagnostic importance in total anomalous pulmonary venous connection. These include right ventricular cavity enlargement and paradoxical septal movement, both suggesting right ventricular volume overload. In addition an echo-free space lying behind the heart has been taken to represent an anomalous common pulmonary vein (Paquet and Gutgesell, 1975).

In this report we present a case of total anomalous pulmonary venous connection to the portal vein in which the diagnosis was suspected on the basis of the clinical picture and the echocardiogram. This was subsequently proven by cardiac catheterisation, including portal vein catheterisation and angiocardiography (Tynan *et al.*, 1974), and the echocardiographic location of the common pulmonary vein was established using contrast echocardiography (Seward *et al.*, 1975).

Clinical information

The infant was born at 36 weeks' gestation (birthweight 2·4 kg). At the age of 14 days he was found to have a systolic murmur at the left sternal edge and was thought to be in cardiac failure. Despite digoxin and diuretics he did not improve and was admitted to this hospital at the age of 24 days.

On examination he was pale and slightly cyanosed with a heart rate of 140 beats a minute. The liver was palpable 3 cm below the right costal margin. On auscultation there was an ejection systolic murmur grade 3/6 at the left sternal edge and wide splitting of the second heart sound with a loud pulmonary component. The chest was clear to auscultation.

Chest x-ray film showed that the heart size was normal and that both lung fields had a faint reticular mottling particularly near the hila. The electrocardiogram showed sinus rhythm, a frontal QRS axis of $+170°$, and right ventricular hypertrophy.

ECHOCARDIOGRAPHIC FINDINGS

Single probe echocardiography (Fig.) showed a right ventricular end-diastolic dimension of 17 mm and a left ventricular end-diastolic dimension of 6 mm; the aortic root dimension was 8 mm; 4 valves were visualised. Mitral-aortic and septal-aortic continuity and paradoxical septal movement were shown. The left atrial cavity appeared to be of normal size (8 mm). However, behind the posterior aortic wall within the left atrial cavity a constant echo was seen moving parallel to the aorta. This was thought to be an artefact. Posterior to the left atrium there was an echo-free space which was assumed to indicate the presence of a common pulmonary vein.

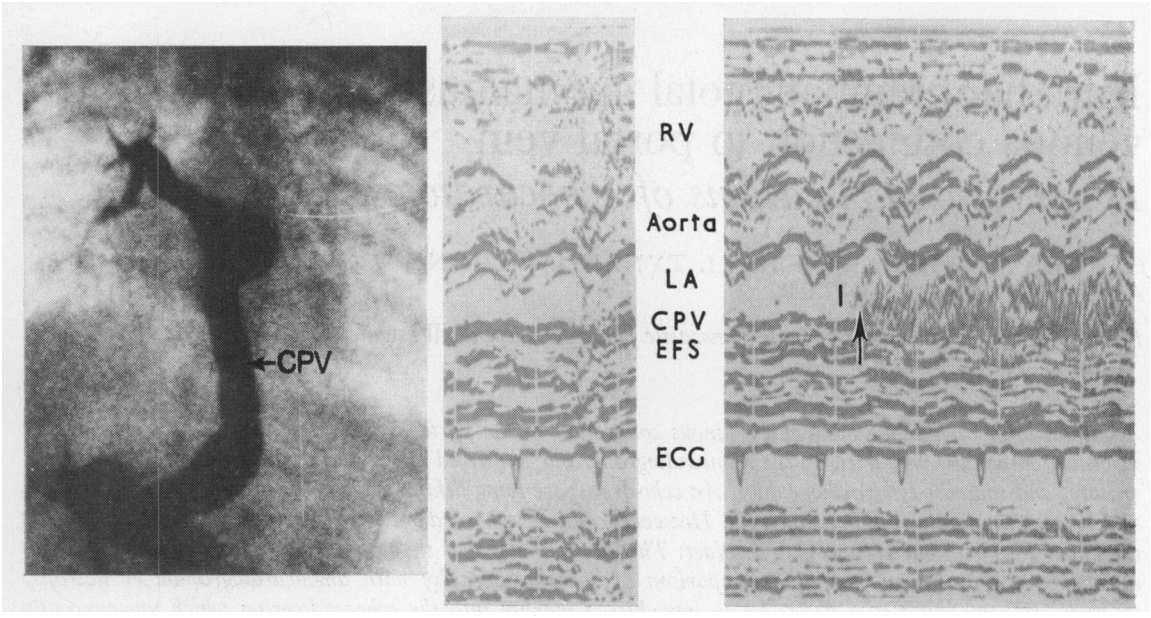

Fig. *Angiogram into the common pulmonary vein (left) with initial echocardiogram (centre) and contrast echocardiogram (right). RV, right ventricle; LA, left atrium; CPV, common pulmonary vein; EFS, echo free space; I, injection of dextrose.*

A presumptive diagnosis of infradiaphragmatic total anomalous pulmonary venous connection was made and the baby underwent cardiac catheterisation.

CARDIAC CATHETERISATION; ANGIOCARDIOGRAPHY AND CONTRAST ECHOCARDIOGRAPHY

Cardiac catheterisation was performed from the right long spahenous vein and the umbilical vein. The findings are summarised in the Table. The common pulmonary vein was entered from the umbilical vein using a Muller guide system. Before angiography a contrast echocardiogram was performed by injecting 2 ml 5 per cent dextrose into the common pulmonary vein while the echo probe was positioned to visualise the aorta, the left atrium, and the previously noted echo-free space. The injection of dextrose opacified the lumen of the common pulmonary vein showing that it occupied most of what had previously been thought to be the left atrium (Fig.) and that the true dimension of the left atrium was only 3 mm. The echo free space lying posteriorly to the heart did not appear to be part of the pulmonary venous system. Cineangiography into the common pulmonary vein (Fig.) established the diagnosis and showed that the contrast echocardiogram had been performed in the common pulmonary vein.

Discussion

Echocardiography is of considerable diagnostic value in infants with congenital heart disease (Godman *et al.*, 1974). There are, however, limitations to the technique which are related to the variability in the anatomy and haemodynamics of the malformations encountered in the newborn period. Thus, a normal echocardiogram has been recorded in a case of total anomalous pulmonary

Table

	Pressures (mmHg)	*Oxygen saturation (%)*
Superior vena cava		23
Inferior vena cava		
suprahepatic		65
infrahepatic		34
Right atrium	mean=7	85
Right ventricle	70/9	86
Main pulmonary artery	60/25	78
Left atrium	mean=7	86
Left ventricle	50/6	84
Descending aorta	47/25	93
Right pulmonary vein	20/16	
Left pulmonary vein	20/16	
Common pulmonary vein	20/16	97
Portal vein	18/14	
Umbilical vein	6/0	93

venous connection to the portal vein where the confluence of the pulmonary veins was close to the diaphragm (Hagler, 1976). Because of this variability, it is essential, in the present state of our knowledge, to validate wherever possible the origin of echoes related to the heart. This report shows that contrast echocardiography performed at catheterisation has a valuable role to play in this process.

An echo-free space bounded by thick continuous echoes lying posteriorly to the left atrium is a common finding in infants and small children. In this case the clinical profile suggested that the diagnosis was obstructed total anomalous pulmonary venous connection. We, therefore, misinterpreted the significance of these echoes and took the space to represent the common pulmonary vein. However, contrast echocardiography showed that these echoes did not originate from the common pulmonary vein. Their origin remains speculative. They may represent part of the descending aorta or left pulmonary artery or they may be artefacts. In retrospect a normal-sized left atrial cavity is an unlikely finding in infradiaphragmatic total anomalous pulmonary venous connection (Glaser *et al.*, 1972) and contrast echocardiograms showed that the true left atrial dimension in this case was very small.

The clinical and echocardiographic appearances may be similar in cortriatriatum and at present the differentiation of this condition from obstructed total anomalous pulmonary venous connection may not be possible without cardiac catheterisation. If either of these conditions are suspected clinically the abnormal echoes arising from the anterior wall of the common pulmonary vein or the septum separating the triatrial chamber from the left atrium should be sought within the apparent left atrial chamber and not necessarily posterior to it.

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