

Partial anomalous pulmonary venous return with intact atrial septum: report of four cases

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ABSTRACT Four patients are reported who underwent repair of partial anomalous pulmonary venous drainage with intact atrial septum. One patient also had azygos continuation of the inferior vena cava and two patients had associated mitral stenosis. Diagnostic considerations and guidelines for operative repair are presented.

Partial anomalous pulmonary venous drainage associated with a defect in the interatrial septum is not an unusual cardiac malformation. About 10% of patients with atrial septal defect have one or more abnormally draining pulmonary veins.^{1,2} In the absence of an interatrial communication, however, the condition is uncommon. From 1970 to 1981 four patients with partial anomalous venous drainage and an intact atrial septum had operative correction at the Johns Hopkins Hospital. These are described and the pertinent diagnostic, physiological, and surgical aspects of the abnormality are reviewed.

Case reports

CASE 1

A 28 year old man³ presented with a continuous murmur and congestive heart failure. At cardiac catheterisation there was a step up in oxygen saturation at the level of the superior vena cava and a calculated 2:1 left to right shunt at the atrial level. The atrial septum could not be crossed with the catheter and transseptal left atrial catheterisation was performed. A 13 mm Hg mitral valve gradient was measured. Angiography confirmed the presence of a non-calcified and mobile but thickened mitral valve. Through a left posterolateral thoracotomy a large, tense left superior pulmonary vein was found that bifurcated at the hilum. The inferior branch entered the left atrium and the superior branch drained into

the innominate vein. The atrial septum was intact to palpation. A mitral commissurotomy was carried out with a transventricular dilator and the anomalous superior pulmonary venous branch was doubly ligated and divided. After operation the continuous murmur was absent and the patient recovered uneventfully.

CASE 2

A 38 year old man presented with symptoms and signs of mild congestive heart failure. He had atrial fibrillation. Catheterisation showed azygos continuation of the inferior vena cava and a calculated 3:1 left to right shunt was present at the atrial level. A diagnosis of ostium secundum atrial septal defect with azygos continuation of the inferior vena cava was made and the patient was referred for operation. Through a median sternotomy the superior vena cava was noted to be considerably enlarged. Cardiopulmonary bypass was established through a single superior vena cava cannula (fig 1). The atrial septum was intact and the coronary sinus was normal. The right superior and inferior pulmonary veins entered the right atrium and were of normal calibre. An atrial septal defect about 3 cm in diameter was made immediately adjacent to the anomalous veins, beginning at the fossa ovalis and extending posteriorly and superiorly to the base of septum at its insertion in the posterior atrial wall. A Dacron patch was used to divert the right pulmonary venous blood into the left atrium and close the septal defect (fig 2). The patient was symptom free with persistent cardiomegaly and normal heart sounds seven years after operation.

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Accepted 6 July 1983

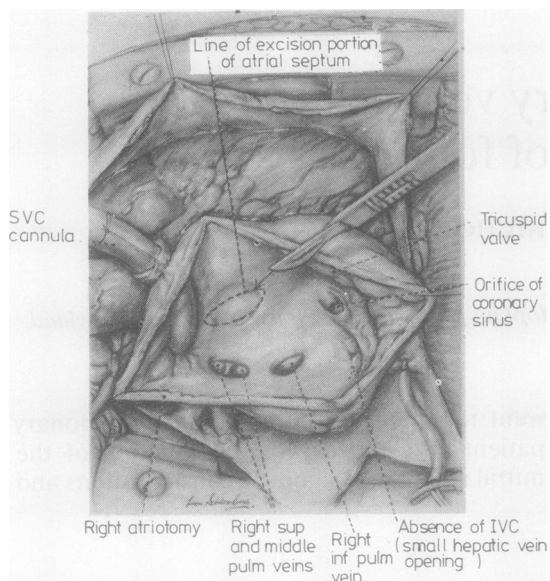


Fig 1 Intraoperative view of the most common variety of partial anomalous pulmonary venous drainage with intact atrial septum, in addition to azygos continuation of the inferior vena cava (IVC). Atrial septectomy is shown. SVC—superior vena cava; sup—superior; inf—inferior; pulm—pulmonary.

CASE 3

A 57 year old man was admitted for evaluation of congestive heart failure. Cardiac catheterisation showed a mitral valve gradient of 12 mm Hg and pulmonary hypertension. The mitral leaflets moved poorly and there was calcium in the mitral annulus. There was a left to right shunt at the atrial level and angiography showed pulmonary venous return to the superior vena cava. Through a median sternotomy, the right heart was found to be notably enlarged, and the right pulmonary veins entered the right atrium. The inferior vena cava was cannulated as usual, but the superior vena cava cannula was placed above the entrance of the anomalous pulmonary veins. An atrial septal defect was created, as in the previous case, adjacent to the pulmonary veins in the posterior and superior atrial septum, and a Dacron patch used to divert right pulmonary venous flow to the left atrium. The mitral valve was replaced with a Björk-Shiley mitral prosthesis. Prolonged cardiopulmonary bypass was required to stop suture line bleeding and the patient had a low cardiac output after operation. Despite the use of an intra-aortic balloon and inotropic support he died after a few hours in the intensive care unit.

CASE 4

A 26 year old symptomless man was referred for

Dacron patch to divert blood from right pulm veins into left atrium

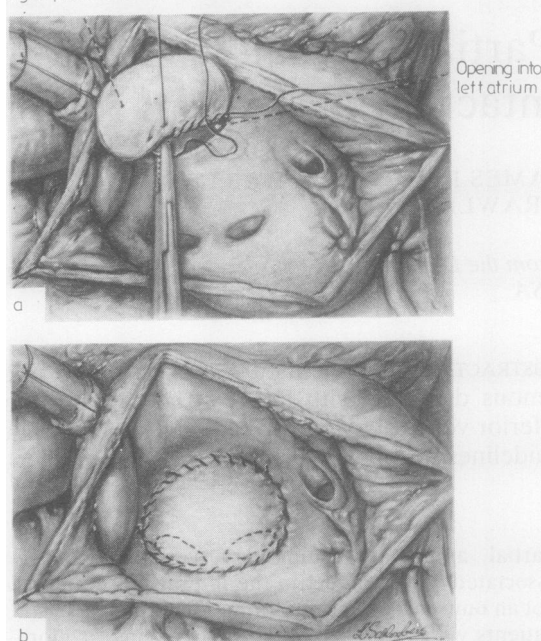


Fig 2 A Dacron patch sewn to (a) the anterior edge of the surgically created atrial septal defect and (b) continued to include the orifices of the anomalous pulmonary veins, diverting blood from the pulmonary veins into the left atrium.

investigation of a soft systolic murmur noted during an employment examination. An atrial septal defect was suspected clinically and cardiac catheterisation showed a step up in oxygen saturation at the right atrial level. At operation the atrial septum was found to be intact. All left pulmonary veins drained into an ascending vein connected to the innominate vein. The side of the anomalous ascending vein was anastomosed to the left atrial appendage before ligation of the anomalous vein. After operation the patient required prolonged ventilatory support and developed unilateral left pulmonary oedema. Pulmonary angiography showed obstruction to left pulmonary venous drainage at the anastomosis. Immediate reoperation was considered but not performed as the patient's general condition improved steadily. Eighteen months after operation the patient was symptom free and pulmonary scans showed good perfusion of the left lung.

Discussion

One hundred and one cases of partial anomalous pulmonary venous drainage with intact atrial septum

have been reported (excluding the Scimitar syndrome).^{4-13 14-27} A single anomalous pulmonary vein in the presence of an intact atrial septum is probably clinically unimportant and may account for the rarity of reported cases. Patients may not have symptoms unless there are multiple anomalous veins or pulmonary hypertension develops.

Pulmonary hypertension can be a late manifestation of isolated partial anomalous pulmonary venous drainage and is thought to result from increased pulmonary blood flow, reflex pulmonary vasoconstriction, and eventually pulmonary vascular obstructive disease.^{6 14} On the other hand, in the presence of mitral stenosis the anomalous vein may moderate left atrial hypertension by shunting pulmonary blood into the systemic venous system.¹⁵ Pathophysiologically, partial anomalous pulmonary venous drainage with intact atrial septum differs little from ostium secundum atrial septal defect.

There are no unique clinical features to distinguish partial anomalous pulmonary venous drainage with intact atrial septum from atrial septal defect. Dyspnoea and fatigue with exercise are the most common symptoms but many patients are symptomless, especially early in life. On physical examination a soft systolic murmur due to increased flow across the pulmonary valve is common, as is fixed splitting of the second heart sound during respiration.¹⁷ A continuous murmur was present in one of our patients, who had co-existing mitral stenosis.³ Typically the electrocardiogram resembles that found with atrial septal defect and shows incomplete right bundle branch block. Cardiac enlargement and increased pulmonary vascular markings on chest radiographs are common with both atrial septal defect and partial anomalous pulmonary venous drainage with intact atrial septum. Two dimensional echocardiography may be helpful in establishing the latter diagnosis, but this technique was not employed in any of the patients we report here.

A diagnosis of partial anomalous pulmonary venous drainage with intact atrial septum is not easily made at cardiac catheterisation. The defect must be specifically considered and several techniques can be used to make the diagnosis. As suggested by Helseth and Peterson,¹⁴ a diagnosis of partial anomalous pulmonary venous drainage of the right upper and middle lobes to the superior vena cava can be made by observing differences in right and left pulmonary artery wedge pressures and by using selective dye dilution curves. The most specific diagnostic technique remains selective cardiac catheterisation with angiography. Anomalous pulmonary veins are recognised by direct entry of the cardiac catheter into a pulmonary vein from the superior vena cava or when there is an oxygen step up in the superior

vena cava. Unrecognised catheter traversal of an atrial septal defect may give similar findings and detailed pulmonary venous anatomy is difficult to delineate. In two of the four patients reported here the diagnosis was not made at catheterisation.

Partial anomalous pulmonary venous drainage with intact atrial septum in combination with valvular heart disease was present in two of the four patients in the present report and this association has been noted by others.^{7 9 14 15} Associated cardiac defects, including dextrocardia,^{7 12} azygos continuation of the inferior vena cava, congenital mitral stenosis or atresia,^{5 25} double outlet right ventricle, ventricular septal defect, tetralogy of Fallot,²⁵ pulmonary valvular stenosis,²⁰ coarctation of the aorta,¹⁴ patent ductus arteriosus,²⁰ mild aortic stenosis,¹² and hypoplasia of the aorta,^{12 21} have also been reported.

Partial anomalous pulmonary venous drainage of three varieties must be suspected when operation for atrial septal defect is undertaken and the atrial septum found intact. Anomalous drainage of the right upper and middle lobe pulmonary veins to the right atrium or superior vena cava is the most common variety (about 90%). A patch covering the orifices of the anomalous veins and a surgically created atrial septal defect will redirect pulmonary venous blood to the left atrium and accomplish repair (cases 2 and 3, fig 1 and 2).

Drainage of the left pulmonary veins to the innominate vein via an abnormal venous channel is the next most common variety. This defect can be repaired by anastomosis of the ascending vein to the left atrium. In our case 4 the side of the ascending vein was anastomosed to the left atrial appendage. In this patient anastomotic occlusion was probably secondary to excessive tension on the vein and anastomosis. Occlusion might have been avoided if the ascending vein had been transected and an end to side rather than a side to side anastomosis made.

Rarely a patient with drainage of the left pulmonary veins into the coronary sinus may be encountered. This anomaly can be corrected by unroofing the coronary sinus through an atrial septostomy with patch closure of the septostomy directing pulmonary and coronary venous flow into the left atrium. Correction of partial anomalous pulmonary venous drainage with intact atrial septum should be successful in most patients, with minimal morbidity and mortality.

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