Anomalous pulmonary venous connexions of left lung associated with valvular heart disease

Report of two cases

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Various intracardiac defects may be associated with anomalous pulmonary venous drainage, atrial septal defect being the most common. However, reports of coexisting valvular heart disease are sparse. Two such cases are presented. In one patient the combination of anomalous venous connexion of the left pulmonary veins and atrial septal defect was associated with congenital aortic stenosis. The other patient had anomalous pulmonary venous connexion of the left lung and calcific mitral regurgitation. Both patients presented for operation during their adult lives. Diagnostic procedures and operative treatment are discussed.

The association of a variety of intracardiac defects with anomalous pulmonary venous connexion has been described, the most common being atrial septal defect (Keith, Rowe, and Vlad, 1967). Other complex anomalies involving association with conditions such as tetralogy of Fallot, tricuspid or mitral atresia, single ventricle, and ventricular septal defects have also been noted (Brody, 1942; Neirotti et al., 1968; Keith et al., 1967). The coexistence of anomalous pulmonary venous drainage with valvular heart disease, however, has been sparsely reported (Adler et al., 1964; Aldridge and Wigle, 1965; Bruschke and Bloch, 1969; Hunt et al., 1970; Maramba et al., 1970; Nichols, Woldow, and Goldberg, 1956).

Recently we have operated on two patients in whom the partial anomalous pulmonary venous drainage was associated with congenital aortic stenosis in one and mitral regurgitation in the other. The report of these cases, with emphasis on the mode of diagnosis and surgical treatment, is the purpose of this communication.

Report of cases

Case I A 28-year-old woman was admitted to the National Heart Hospital on 3 December 1969 for assessment with a view to operation. She had been found to have a heart murmur at the age of 2 and, though her activities were restricted, she became symptomatic in the latter part of her teens. She complained of breathlessness and tiredness, both related to exertion.

On examination she had a regular, slow-rising pulse and a blood pressure of 120/90 mmHg. There was a conspicuous left ventricular impulse and a systolic thrill was present at the left sternal border. On auscultation the first heart sound was followed by an ejection click and there was an ejection systolic murmur heard best at the second intercostal space within the left sternal border. In addition, there was a continuous murmur heard best just below the sternal head of the left clavicle. The diastolic component of this murmur was radiated down the left sternal edge.

The electrocardiogram revealed sinus rhythm, right bundle-branch block, and right axis deviation. The chest x-ray showed plethoric lungs, with prominence of the left upper border of the cardiovascular shadow (Fig. 1).

Cardiac catheterization disclosed a systolic gradient of 140 mmHg across the aortic valve. The left atrium was easily entered from the right side through an atrial septal defect, and a conspicuous pressure gradient was found between the left and right atria. There was a large (5:1) left-to-right shunt at the point where the innominate vein joined the superior vena cava and also a gradient of 10 to 16 mmHg across the junction of the two veins.

Differential angiography showed a normal pulmonary venous drainage of the right lung. The left lung drained abnormally into a vertical vein which, in turn, drained into the innominate vein producing opacification of this vessel. Left ventricular angiogram showed a domed, thickened aortic valve and no evidence of subvalvar stenosis.
On 18 January 1971 the patient was operated upon through a midternal incision. The pulmonary artery was larger than the aorta. There was, however, a thrill in the small aortic root consistent with aortic stenosis. The right pulmonary veins were seen to enter the left atrium, but there was no evidence of any left pulmonary vein entering the pericardium. A finger in the right atrium confirmed the presence of a secundum atrial septal defect.

Normothermic cardiopulmonary bypass was instituted after cannulating the ascending aorta and both venae cavae; the right atrium was opened by a vertical incision. The atrial septal defect was closed with direct sutures.

Dissection was then carried out in the region of the left side of the mediastinum along the left innominate vein. A large venous trunk was discovered similar in size to the innominate vein and entering it opposite the jugular subclavian junction (Fig. 2).

The venous trunk was divided and the innominate end was oversewn. An attempt was made to find a point of contact between this vessel and the posterior wall of the left atrium, but it did not seem feasible because of the distance between them. Consequently, the divided vein was anastomosed to the amputated end of the left atrial appendage.

Attention was then directed to the aorta which was opened by an oblique incision. This revealed a small aortic root with a stenotic calcified bicuspid valve which was excised. The aortic root would not accept more than a 1·75 cm bougie. The aortotomy was extended down through the valve ring into the outflow tract and a gusset of woven Dacron was inserted to enlarge the ring. Inspection of the subvalvar region showed subvalvar hypertrophy and a myotomy was carried out below the intercoronary commissure.

A 21 mm Bjork type of prosthesis was inserted into the aortic annulus with multiple interrupted sutures and attached to the gusset over the non-coronary sinus.

The patient's postoperative course was uneventful. A lung scan showed the left lung less well perfused than the right, but without evidence of a localized area of absent flow.

**Case 2** A 30-year-old Greek was admitted to Guy's Hospital on 2 May 1971 with shortness of breath and ankle oedema.

On examination, he was in atrial fibrillation with a blood pressure of 125/80 mmHg. The pericardium was hyperdynamic and a systolic thrill was felt at the apex. On auscultation there was a pansystolic murmur at the apex, which radiated to the axilla. A soft diastolic murmur was also heard in the same area. Just below the left clavicle a distinct, soft, continuous murmur could be heard. The electrocardiogram showed atrial fibrillation with combined left and right ventricular hypertrophy. The chest x-ray revealed cardiomegaly and a prominence of the left superior border of the cardiac shadow (Fig. 3).

Cardiac catheterization disclosed a pulmonary artery pressure of 85/35 mmHg, with raised pulmonary vascular resistance (5 units) and a left-to-right shunt from anomalous venous drainage of the left lung with a pulmonary-systemic flow ratio of 2:1.
An angiogram was performed through the left subclavian vein; the injection showed a normal left subclavian vein with a very wide innominate vein and the turbulence within it suggested flow from the rounded opacity on the left heart border, into which the catheter entered. It appeared that an anomalous trunk drained from the left lung into the dilated left innominate vein and this was confirmed by pulmonary arteriogram, in which the right pulmonary vein could already be seen to enter the right atrium while on the left side the pulmonary vein drained into the innominate vein.

Left ventricular angiogram showed gross mitral regurgitation (grade 3/4) with extensive calcification of the valve.

On 13 May 1971 the patient underwent surgery through a midsternal incision. The external appearances of the heart were those of a large right atrium, right ventricle, and pulmonary artery, and a small aortic root. The left atrium was enlarged and tense. A finger in the right atrium disclosed the presence of a minor degree of tricuspid regurgitation from a dilated valve ring without atrial septal defect.

On raising the heart there was a large vessel entering the left atrium in the region of the pulmonary vein but this could not be defined.

Normal temperature bypass was instituted and the left atrium was opened by longitudinal incision. This presented some difficulty because there was a torrential blood flow into the left atrium from the aforementioned vessel and only when a clamp was applied to it could the mitral valve be seen. It was excised and replaced with a size 3 silastic ball Starr valve prosthesis fixed in place with a continuous 2/0 suture interrupted four times.

Attention was then directed to the anomalous drainage (Fig. 4). The pulmonary veins were dissected out and their orifices were identified from within the anomalous vessel draining into the left atrium. The common venous trunk was clamped above the lung hilum and below the subclavian jugular junction and then tied.

In the immediate postoperative period the patient was jaundiced and also developed a left lower lobe pneumonia, but both of these resolved rapidly and at the time of his discharge from the hospital, he was in good clinical condition.

**Discussion**

Two general varieties of anomalous pulmonary venous connexion are recognized, the partial and the total. In the partial form there is usually anomalous venous connexion of one lung, or a portion of the lung, from which the venous channel leads to a structure or chamber other than the left atrium.

Partial anomalous venous connexion involving the pulmonary veins on the left is much less common than those involving the veins on the right by a ratio of about 1:10 (Neirroti et al., 1968).

Atrial septal defect is the most frequently associated lesion. In one series it was found in 92 per cent of the cases of right pulmonary anomalous venous drainage, but in only 16 per cent of the cases with left-sided anomalous drainage. All the defects were of the secundum type. The anomalous veins may connect directly with the innominate vein, with a left
vertical vein joining the innominate vein or, in a small number of cases, with the coronary sinus (Neirotti et al., 1968).

The clinical, haemodynamic, and electrocardiographic findings of patients with isolated partial anomalous pulmonary venous drainage are similar to those seen in cases of isolated atrial septal defect, since the pulmonary blood draining into the right side of the heart constitutes a left-to-right shunt at the atrial level (Oropeza et al., 1970).

When anomalous pulmonary venous drainage coexists with other cardiac malformations, the clinical and haemodynamic features are usually modified or are chiefly determined by the complicated defect. Left-sided anomalies can be an incidental operative finding during a left thoracotomy for lung resection or closed mitral valvotomy (Acker et al., 1966; Adler et al., 1964). If the pulmonary venous connexion is into the left innominate vein and there is some constriction at the site of junction, a high frequency continuous murmur may be heard at the infraclavicular area (Gasul, Arcilla, and Lev, 1966). The chest x-ray may be useful in the differential diagnosis showing a prominence of the left upper border of the cardiovascular shadow and plethoric lungs. These features were present in the two cases reported here.

Differential angiography, with injection of the contrast substance into the pulmonary trunk, usually outlines the vertical vein and also produces opacification of the left innominate vein.

Cardiac catheterization is necessary to show the left-to-right shunt. A gradient across the junction of the innominate and superior vena cava can be found when there is some constriction at this point. Pulmonary venous obstruction at the point of entry of the vertical vein into the systemic vein has been shown in patients with total anomalous venous return (Gomes et al., 1970). Obstruction can also be produced by the left main bronchus and the pulmonary artery which compresses the anomalous vein (Elliott and Edwards, 1962; Warren, Benaron, and Sissman, 1968).

In our second case, the left-aortic-cardinal vein was of great functional importance because the left atrial pressure was raised due to the severe mitral regurgitation. Surgical correction can be accomplished by performing an anastomosis between the left vertical vein and the left atrium (Cooley and Mahaffey, 1955).

In reviewing 124 cases of anomalous pulmonary venous connexion, Snellen, Van Ingen, and Hoeftsmidt (1968) found that the aortic annulus and valve were affected in 6 cases and the mitral valve in 10 either as solitary occurrences or as part of more complex malformations.

In conclusion, the combination of anomalous pulmonary venous connexion and valvular heart disease is not common. The clinical picture presented by these patients makes diagnosis difficult, but the presence of the anomalous venous drainage should be considered when a continuous type of murmur below the left clavicle, a prominence of the left upper border of the cardiovascular shadow, and pulmonary plethora are present in patients who otherwise have evidence of valvular disease. Cardiac catheterization and pulmonary arteriography are of great importance in arriving at the correct diagnosis.

Surgical correction should be done at the same time as the operation for valvular disease by performing an anastomosis between the anomalous venous trunk and the posterior wall of the left atrium, when anatomically feasible, or with the left atrial appendage.

When there is a persistent laevo-atrial-cardinal vein, ligation of this vessel proximal to the innominate vein is recommended.

References
Brody, H. (1942). Drainage of the pulmonary veins into the right side of the heart. Archives of Pathology, 33, 221.
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