Case reports


Cor triatriatum

Diagnosis by echocardiography

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A case of cor triatriatum is reported in which the left atrial diaphragm was shown by a preoperative echocardiogram, but not by angiocardiography. Echocardiography may be a useful diagnostic procedure in this uncommon form of congenital heart disease.

Cor triatriatum can be readily corrected surgically. Adult patients present with signs of pulmonary venous and arterial hypertension, and the condition must be distinguished from mitral valve disease which it closely resembles and from less common left-sided obstructive lesions. Though some of the characteristic clinical, radiological, and angiocardiographic signs of mitral stenosis may be absent, the distinction is usually made preoperatively by angiocardiography, or occasionally by recording a low left atrial pressure distal to the obstructing diaphragm.

In the case reported here echocardiography showed a normal mitral valve echo together with an abnormal echo from the left atrial diaphragm, though the angiocardiographic appearances were not diagnostic.

Case report

The patient was a 23-year-old Palestinian Arab bank clerk from Qatar. He complained of increasing breathlessness for two years; though he had been able to play football until nine months previously, at the time of admission he was severely disabled. He reported recurrent cough and occasional haemoptysis. There was no rheumatic history.

On examination, the arterial pulse was very small but regular. There were no signs of right heart failure. There was clinical evidence of severe pulmonary arterial hypertension, with obvious right ventricular hypertrophy and a very loud pulmonary closure sound. He had a soft pansystolic murmur increasing with inspiration and a third sound at the left sternal border, but at no time was any apical murmur heard; the first sound was normal and there was no opening snap. The blood pressure was 95/70 mmHg. Other systems were normal.

The chest radiograph showed moderate cardiac enlargement, enlargement of the pulmonary trunk, and severe pulmonary venous congestion, with Kerley B lines and early pulmonary oedema; there was no definite left atrial enlargement and no mitral calcification. The electrocardiogram showed sinus rhythm with normal P waves, and moderate right ventricular hypertrophy.

Cardiac catheterization (Table)

This showed severe pulmonary venous and arterial hypertension with a moderate rise in pulmonary arteriolar resistance. The mean diastolic pressure gradient between the pulmonary artery wedge position and the left ventricle was 38 mmHg.

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mmHg above midthorax)</th>
<th>Oxygen saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td>a 8 x 5</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>90/2-8</td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>90/42 mean 58</td>
<td>55</td>
</tr>
<tr>
<td>Pulmonary artery wedge</td>
<td>44 y 39 mean 40</td>
<td></td>
</tr>
<tr>
<td>Left ventricle</td>
<td>84/0-6</td>
<td></td>
</tr>
<tr>
<td>Aorta</td>
<td>90/70</td>
<td>86</td>
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Cardiac index* 3 x 1.1/min per m². Pulmonary arteriolar resistance 5.8 units x m². Pulmonary artery wedge – left ventricle mean gradient 38 mmHg

*Assumed basal oxygen uptake.
Angiocardiogram (Fig. 1)
After injection into the pulmonary trunk, there was delayed emptying of the apparent left atrium. The pulmonary veins were dilated, and there was a V-shaped confluence of the right and left upper lobe pulmonary veins; the usual convex upper border of the left atrium was not seen, and the atrial appendage did not opacify. The lower left border of the chamber had a crescentic contour, but the appearance was not diagnostic of cor triatriatum.

Echocardiogram\(^1\) (Fig. 2A, B)
This showed normal movement of the anterior cusp of the mitral valve with increased diastolic closure rate. An abnormal echo was recorded on the left atrial aspect of the mitral valve. This was superimposed on the cusp echo during ventricular systole, and moved slightly in an anterior direction with mitral valve opening. There was no evidence of left atrial myxoma. The right ventricular cavity was enlarged, and the tricuspid valve echo was normal.

At operation (S.C.L.) on 25 February 1972, under cardiopulmonary bypass, the grossly dilated pulmonary veins were seen to enter a chamber in which the mean pressure was 40 mmHg. When this chamber was opened, the mitral valve was obscured by a diaphragm with a 2 mm orifice which opened into a small distal chamber. The septum was excised revealing a normal mitral valve. The mean left atrial pressure was now 8 mmHg.

The postoperative course was smooth. Serial chest radiographs showed clearing of the pulmonary congestive changes. A loud pulmonary closure sound persisted but there were no other abnormal auscultatory signs.

The echocardiogram (Fig. 2C) again showed normal

\(^1\)Ekoline 20 Ultrasonoscope

FIG. 1 Angiocardiogram after injection of contrast into pulmonary trunk. Crescentic lower border of accessory chamber is indicated, but no linear translucency is seen.

FIG. 2 A) Preoperative echocardiogram of the mitral valve. The diastolic closure rate is increased at 280 mm/sec. The thickness of the echo and the amplitude of its movement are normal. B) Echocardiogram showing echo thought to originate from the supravalvar diaphragm, which was not separable from the anterior cusp echo, and considerably thicker than that originating from a normal mitral valve ring. C) Postoperative echocardiogram of the anterior cusp of the mitral valve. The diastolic closure rate is normal at 120 mm/sec. The echo thought to have originated from the diaphragm is no longer apparent.
movement of the anterior cusp of the mitral valve. No abnormal echo could be identified on the atrial side of the valve.

**Discussion**

Cor triatriatum is usually found in early childhood but some patients survive to adult life. The clinical and radiological signs are those of pulmonary arterial and venous hypertension. Apical systolic, diastolic, and continuous (McGuire et al., 1965; Neufeld et al., 1965) murmurs have been reported, but typical mitral diastolic and presystolic murmurs are unusual, and there is no opening snap. Murmurs of pulmonary regurgitation or tricuspid regurgitation may be present. Other features suggesting cor triatriatum are normal left atrial size and normal (or right atrial) P waves, though there is sometimes radiological evidence of left atrial enlargement and P mitrale may occur; atrial fibrillation is rare.

Cor triatriatum is confirmed when a normal left atrial pressure can be recorded from the distal chamber by transseptal puncture (McGuire et al., 1965), by passage of a catheter across a patent foramen ovale (Lacque et al., 1966; Perry, Scott, and McClenathan, 1967; Wolfe et al., 1968; Brickman et al., 1970), though this sometimes communicates with the proximal high-pressure chamber (Niwayama, 1960), or retrogradely into the true left atrium (Brickman et al., 1970). The diagnostic value of angiography has been stressed by many authors (Lassalle et al., 1963; Ellis et al., 1964; Miller et al., 1964; McGuire et al., 1965; Neufeld et al., 1965; Arkoff et al., 1966; Ahn, Hosier, and Sirak, 1968; Wolfe et al., 1968; Barrillon et al., 1968; Al Abdulla, Demany, and Zimmerman, 1970; Brickman et al., 1970; Tesler et al., 1971; Park, Ricketts, and Guntheroth, 1972). Pulmonary arteriography shows prolonged opacification of the proximal accessory chamber, with delayed emptying into the true left atrium and atrial appendage; the obstructing diaphragm may be seen in profile as an oblique linear translucency. Cineangiography shows the distal chamber to contract vigorously during atrial systole, but the proximal accessory chamber contracts poorly; the membrane moves towards the mitral valve during ventricular diastole, but straightens after closure of the mitral valve (Ellis et al., 1964). Injection directly into the true left atrium opacifies this chamber, which is small and outlined above by the straight contour of the membrane (Perry et al., 1967; Wolfe et al., 1968; Brickman et al., 1970), and the atrial appendage. A similar appearance may be seen during left ventricular angiocardiography, if ectopic beats result in mitral regurgitation (Miller et al., 1964; Girod and Kurlander, 1966).

The young man reported here had clinical and radiological signs of pulmonary arterial and venous hypertension, confirmed by catheterization. The systolic murmur was attributed to tricuspid regurgitation, and no auscultatory signs of mitral stenosis were detected. The differential diagnosis included slight mitral stenosis, left atrial myxoma, cor triatriatum, supravalvar stenosing ring, intrapulmonary veno-occlusive disease (Heath, Segel, and Bishop, 1966), and stenosis of all four individual pulmonary veins (Shone et al., 1962). Enlargement of the main pulmonary veins excluded pulmonary veno-occlusive disease and the absence of a filling defect in the left atrium on the angiocardiogram was against a diagnosis of atrial myxoma. The angiocardiogram also failed to establish the diagnosis of cor triatriatum.

Echocardiography in mitral stenosis shows a reduced diastolic closure rate and also increased thickness of the anterior cusp echo (Edler, 1956; Joyner, Reid, and Bond, 1963; Segal, Likoff, and Kingsley, 1966; Zaky, Nassir, and Feiggenbaum, 1968). In myxoma, a characteristic cloud of echoes appears on the atrial side of the valve (Popp and Harrison, 1969; Wolfe, Popp, and Feiggenbaum, 1969; Finegan and Harrison, 1970; Nicholls, Goodman, and Pride, 1971). Lundstrom (1972) has recently described echocardiograms from infants with supravalvar stenosing ring and cor triatriatum. His illustration of the abnormal echo in one infant with cor triatriatum shows a greater separation from the mitral valve and larger amplitude of movement than in our case; the diagnosis was confirmed at necropsy. In our case, the origin of the echo from the left atrial diaphragm was confirmed by its disappearance after surgical excision.

The value of echocardiography in the differential diagnosis of lesions causing pulmonary venous hypertension is clear. A normal mitral valve echo excludes haemodynamically significant mitral stenosis, and abnormal echoes may indicate the presence of left atrial myxoma, cor triatriatum, or supravalvar stenosing ring. The technique is non-invasive and therefore safer than transseptal left atrial puncture or angiocardiography, particularly in the presence of severe pulmonary arterial hypertension. Furthermore transseptal puncture or passage of a catheter through a patent foramen ovale may fail to provide diagnostic information if the high-pressure accessory chamber is entered by the needle or catheter. If experience confirms that an abnormal echo can be recorded in other cases of cor triatriatum, angiocardiography and left atrial puncture may need to be employed only when the information obtained by echocardiography is equivocal.
References


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