Case reports

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Cor triatriatum Diagnosis by echocardiography

D. G. Gibson, M. Honey, and S. C. Lennox

From the Cardiac Department, Brompton Hospital, London

A case of cor triatriatum is reported in which the left atrial diaphragm was shown by a preoperative echocardiogram, but not by angiocardiology. 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 electrocardiographic signs of mitral stenosis may be absent, the distinction is usually made preoperatively by angiocardiology, 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 angiocardiology 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 Cardiac catheterization results

Site	Pressure (mmHg above midthorax)	Oxygen saturation (%)
Right atrium	a 8 x 5	
Right ventricle	90/2-8	
Pulmonary artery	90/42 mean 58	55
Pulmonary artery wedge	v 44 y 39 mean 40	
Left ventricle	84/0-6	
Aorta	90/70	86
Cardiac index* 3.1 l./min per m ² .		
Pulmonary arteriolar resistance 5.8 units x m ² .		
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

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

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.

Echocardiogram¹ (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

¹Ekoline 20 Ultrasonoscope

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 supralvalvar 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 (Lacquet *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 angiocardiology 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. Cineangiocardiology 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 angiocardiology, 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 silent mitral stenosis, left atrial myxoma, cor triatriatum, supraventricular stenosing ring, intra-pulmonary 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 angiocardiology was against a diagnosis of atrial myxoma. The angiocardiology 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 Feigenbaum, 1968). In myxoma, a characteristic cloud of echoes appears on the atrial side of the valve (Popp and Harrison, 1969; Wolfe, Popp, and Feigenbaum, 1969; Finegan and Harrison, 1970; Nicholls, Goodman, and Pridie, 1971). Lundstrom (1972) has recently described echocardiograms from infants with supraventricular 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 successful 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 supraventricular stenosing ring. The technique is non-invasive and therefore safer than transseptal left atrial puncture or angiocardiology, 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, angiocardiology and left atrial puncture may need to be employed only when the information obtained by echocardiography is equivocal.

References

- Ahn, C., Hosier, D. M., and Sirak, H. D. (1968). Cor triatriatum: a case report and review of other operative cases. *Journal of Thoracic and Cardiovascular Surgery*, **56**, 177.
- Al Abdulla, H. M., Demany, M. A., and Zimmerman, H. A. (1970). Cor triatriatum. Preoperative diagnosis in an adult patient. *American Journal of Cardiology*, **26**, 310.
- Arkoff, R. S., De Vries, P. A., Leonards, R., and Stevens, L. E. (1966). Cor triatriatum. A case report. *Radiologia Clinica et Biologica*, **35**, 274.
- Barrillon, A., Blondeau, P., Cachin, J. C., Ourbak, P., and Lenegre, J. (1968). Le coeur triatrial de l'adulte. *Archives des Maladies du Cœur et des Vaisseaux*, **61**, 1306.
- Brickman, R. D., Wilson, L., Zuberbuhler, J. R., and Banhsen, H. T. (1970). Cor triatriatum. Clinical presentation and operative treatment. *Journal of Thoracic and Cardiovascular Surgery*, **60**, 523.
- Edler, I. (1956). Ultrasound-cardiogram in mitral valve disease. *Acta Chirurgica Scandinavica*, **111**, 230.
- Ellis, K., Griffiths, S. P., Jesse, M. J., and Jameson, A. G. (1964). Cor triatriatum. Angiographic demonstration of the obstructing left atrial membrane. *American Journal of Roentgenology*, **92**, 669.
- Finegan, R. E., and Harrison, D. C. (1970). Diagnosis of left atrial myxoma by echocardiography. *New England Journal of Medicine*, **282**, 1022.
- Girod, D. A., and Kurlander, G. J. (1966). Diagnosis of cor triatriatum by left ventricular angiography. *Diseases of the Chest*, **50**, 535.
- Heath, D., Segel, N., and Bishop, J. (1966). Pulmonary veno-occlusive disease. *Circulation*, **34**, 242.
- Joyner, C. R., Reid, J. M., and Bond, J. P. (1963). Reflected ultrasound in the assessment of mitral valve disease. *Circulation*, **27**, 503.
- Lacquet, L. K., Los, J. A., Nauta, J., and Brom, A. G. (1966). Cor triatriatum. *Thorax*, **21**, 175.
- Lassalle, R., Ethier, M., Stanley, P., and Davignon, A. (1963). Cor triatriatum. Report of a case with emphasis on cine-angiography. *Canadian Medical Association Journal*, **89**, 616.
- Lundstrom, N.-R. (1972). Ultrasoundcardiographic studies of the mitral valve region in young infants with mitral atresia, mitral stenosis, hypoplasia of the left ventricle, and cor triatriatum. *Circulation*, **45**, 324.
- McGuire, L. B., Nolan, T. B., Reeve, R., and Dammann, J. F. (1965). Cor triatriatum as a problem of adult heart disease. *Circulation*, **31**, 263.
- Miller, G. A. H., Ongley, P. A., Anderson, M. W., Kincaid, O. W., and Swan, H. J. C. (1964). Cor triatriatum. Hemodynamic and angiocardigraphic diagnosis. *American Heart Journal*, **68**, 298.
- Neufeld, H. N., Pautzner, Y., Gueron, M., Deutsch, V., and Cohen, B. (1965). Cor triatriatum. Report of a case successfully operated with special reference to angiocardigraphic diagnosis. *Israel Journal of Medical Sciences*, **1**, 71.
- Nicholls, A. J. S., Goodman, M. A., and Pridie, R. B. (1971). Left atrial myxoma in a child diagnosed by ultra-sound. *Postgraduate Medical Journal*, **47**, 804.
- Niwayama, G. (1960). Cor triatriatum. *American Heart Journal*, **59**, 291.
- Park, M. K., Ricketts, H. J., and Guntheroth, W. G. (1972). Cor triatriatum, an operable form of pulmonary hypertension. *American Journal of Diseases of Children*, **123**, 500.
- Perry, L. W., Scott, L. P., and McClenathan, J. E. (1967). Cor triatriatum. Preoperative diagnosis and successful surgical repair in a small infant. *Journals of Pediatrics*, **71**, 840.
- Popp, R. L., and Harrison, D. C. (1969). Ultrasound for the diagnosis of atrial tumour. *Annals of Internal Medicine*, **71**, 785.
- Segal, B. L., Likoff, W., and Kingsley, B. (1966). Echocardiography. Clinical application in mitral stenosis. *Journal of the American Medical Association*, **195**, 161.
- Shone, J. D., Amplatz, K., Anderson, R. C., Adams, P., and Edwards, J. E. (1962). Congenital stenosis of individual pulmonary veins. *Circulation*, **26**, 574.
- Tesler, U. F., Hallman, G. L., McNamara, D. G., and Cooley, D. A. (1971). Il trattamento chirurgico del cor triatriatum. *Minerva Cardioangiologica*, **19**, 397.
- Wolfe, R. R., Ruttenberg, H. D., Desilets, D. T., and Mulder, D. E. (1968). Cor triatriatum. Total correction in an infant. *Journal of Thoracic and Cardiovascular Surgery*, **6**, 114.
- Wolfe, S. B., Popp, R. L., and Feigenbaum, H. (1969). Diagnosis of atrial tumours by ultrasound. *Circulation*, **39**, 615.
- Zaky, A., Nasser, W. K., and Feigenbaum, H. (1968). A study of mitral valve action recorded by reflected ultrasound and its application in the diagnosis of mitral stenosis. *Circulation*, **37**, 789.

Requests for reprints to Dr. Michael Honey, Cardiac Department, Brompton Hospital, Fulham Road, London SW3 6HP.