CASE REPORTS

COR TRIATRIATUM

A CASE SUCCESSFULLY CORRECTED BY SURGERY

BY

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Cor triatriatum is an uncommon congenital anomaly. Abbott (1946) reported only seven cases in 1000 necropsies of congenital heart disease and there were only four examples in 3740 autopsies performed at the Buffalo Children's Hospital from 1936 to 1948. It has been estimated that 75 per cent of cases die in infancy (Keith *et al.*, 1958) and eighteen of the thirty-nine cases so far reported have been under sixteen months of age (Niwayama, 1960). Therefore, the report of an adult patient successfully corrected by operation is of interest.

Case Report

A married woman aged 33 years had suffered from tuberculous cervical adenitis in childhood but she gave no history of rheumatic fever. She had always been somewhat short of breath on exertion and a heart murmur had been noted at a routine medical examination when she was 21 years old. She had two normal full-term pregnancies without difficulty, but noticed increasing shortness of breath on exertion following the birth of her second child two and a half years before coming into hospital. Eight months before admission

FIG. 1.—Chest X-ray, P.A. view showing high aortic arch and some enlargement of the left atrium.

FIG. 2.—Barium swallow showing minimal posterior enlargement of the left atrium.

FIG. 3.—Pressure curves showing no gradient between the lower chamber of the left atrium (L.A.) and the left ventricle (L.V.).

she complained of palpitations and severe dyspnoea and was found to have atrial fibrillation. In spite of treatment with digitalis, the dyspnoea increased and she also developed winter bronchitis. At the time of admission she was very breathless on climbing twelve stairs or walking 500 yards on the level.

On examination she had a malar flush and her hands and feet were cold and blue. Her pulse was of small volume and there was well controlled atrial fibrillation. The venous pressure was normal and the blood pressure was 110/65. The cardiac impulse was of right ventricular type. The second heart sound in the pulmonary area was accentuated. At the apex there was a diastolic murmur of moderate length and slight intensity. This murmur could be better heard after exercise and turning the patient on to the left side.

The electrocardiogram showed atrial fibrillation and slight right ventricular hypertrophy. A chest X-ray (Fig. 1) showed slight cardiac enlargement and a high, prominent, aortic arch. This view suggested enlargement of the left atrium but a barium swallow (Fig. 2) did not show any oesophageal displacement. No valve calcification was seen on screening.

A diagnosis of mitral stenosis was made and operation was performed (O.S.T.). The left lung appeared normal and no thrills were palpable over the surface of the heart. With needles in the left atrial appendage and in the left ventricle no pressure gradient was demonstrated across the mitral valve (Fig. 3). When examined with the index finger inserted through the left atrial appendage, the mitral valve was normal. However, a circular foramen about 2 cm. in diameter was felt just above the medial commissure of the mitral valve. This opening lay in a diaphragm stretching obliquely across the interior of the left atrium. No orifices of pulmonary veins could be felt in the lower chamber, but it was possible to push the finger through the foramen and to demonstrate that the left pulmonary vein entered the upper chamber. Further pressure measurements

FIG. 4.—Pressure tracings showing no gradient between lower chamber of left atrium and the left ventricle, but a gradient of 5 mm. Hg between the upper atrial chamber and the left ventricle.

(Fig. 4) showed a gradient of 5 mm. Hg between the upper chamber and the left ventricle. A Brock mitral knife, applied to the left index finger, was introduced into the lower atrial chamber and the velum was incised in a lateral direction. An opening at least 4 cm. across was obtained. Pressure measurements then showed no gradient between the two atrial chambers or across the mitral valve. Examination was made for the presence of an atrial septal defect but none was found.

The patient made an uninterrupted recovery. During the third week after operation an effort was made to restore sinus rhythm with quinidine, but this was unsuccessful despite heavy dosage. Two months after leaving hospital she was doing all her housework and could hurry up two flights of stairs. The heart was still in atrial fibrillation. No murmurs were heard. A malar flush was still present.

Discussion

Survival would appear to depend on the size of the opening in the diaphragm. Keith *et al.* (1958) found that adult patients always had an opening over 7 mm. in diameter. With larger openings long survival is possible and Loeffler (1949) has reported the condition in a woman of seventy years. In the case reported here, severe symptoms only occurred with the onset of atrial fibrillation. Improvement did not follow digitalization, so, presumably, atrial contraction had been important in securing left ventricular filling.

Cases in infancy usually present with respiratory infections and congestive cardiac failure (Barnes and Finlay, 1952; Doxiadis and Emery, 1953; Parsons, 1950). In the older age groups there is a close resemblance to mitral stenosis both in symptoms and in physical signs. Hæmoptysis has often been reported (Barrett and Hickie, 1957; Hartmann, 1955; McLester *et al.*, 1940). Peripheral cyanosis occurs (Hartmann, 1955; Vineberg and Gialloretto, 1946), also winter bronchitis, as in the present case. The pulmonary second heart sound has usually been noted as accentuated (Keith *et al.*, 1958; Pedersen and Therkelsen, 1954; Sawyer *et al.*, 1957). Children have usually had a systolic murmur but an apical diastolic murmur occurs when the opening in the diaphragm is over 5 mm. across and where there is no interatrial communication (Niwayama, 1960). As in the present case, such diastolic murmurs have been of low intensity and short duration and a mitral opening snap has not been heard.

In children the chest X-ray usually shows an enlarged globular heart (Barnes *et al.*, 1952; Nash and MacKinnan, 1956). In adults enlargement of the pulmonary artery and pulmonary venous congestion without left atrial enlargement has been noted (Belcher and Somerville, 1959; Pedersen *et al.*, 1954; Vineberg *et al.*, 1956). Our patient did not have the usual pear-shaped mitral heart outline (Fig. 2) and, although there does not seem to be a typical X-ray appearance for this condition, an unusual cardiac silhouette may be helpful in diagnosis.

Anderson and Varco (1961) have reported the correct pre-operative diagnosis and successful treatment of a child of three years using cardio-pulmonary by-pass. There are reports of five adult patients treated successfully (Lewis *et al.*, 1956; Vineberg *et al.*, 1956; Barrett *et al.*, 1957; Lam, 1958; Belcher *et al.*, 1959), but in none of these was a correct pre-operative diagnosis made. Indeed the condition may easily be missed at an exploratory cardiectomy, as reported by Pedersen *et al.* (1954) and by Darke *et al.* (1961).

The surgical methods used have varied. Lewis (1956) and Lam (1958) both used open operation under hypothermia, as the pre-operative diagnosis had been atrial septal defect. The other cases were operated upon with a presumptive diagnosis of mitral stenosis. Vineberg (1956) fractured the partially calcified membrane with finger pressure. Barrett (1957) and Belcher (1959) both used a finger knife as in the present case. During this operation it was found that once an initial incision had been made the membrane became lax and difficult to cut. It is, therefore, suggested (O.S.T.) that an alternative method might be to introduce a bistoury through a stab incision in the upper chamber, which could then be used to cut the membrane under guidance of the left index finger, introduced into the lower chamber.

Summary

A case of cor triatriatum is reported. The raised pressure in the upper chamber of the left atrium was abolished by surgical section of the velum separating the two chambers, with subsequent symptomatic relief.

We wish to thank Dr. G. W. Hayward for kindly referring the case for treatment.

References

- Abbott, M. E. (1946). *Nelson Loose-Leaf Medicine Vol. 4*, p. 207. Nelson, New York.
- Anderson, R. C., and Varco, R. L. (1961). *Cardiology*, **7**, 436.
- Barnes, C. G., and Finlay, H. V. L. (1952). *Brit. Heart J.*, **14**, 283.
- Barrett, N. R., and Hickie, J. B. (1957). *Thorax*, **12**, 24.
- Belcher, J. B., Somerville, W. (1959). *Brit. med. J.*, **1**, 1280.
- Darke, C. S., Emery, J. L., and Lorber, J. (1961). *Brit. Heart J.*, **23**, 329.
- Doxiadis, S. A., and Emery, J. L. (1953). *J. Pediatrics*, **42**, 87.
- Hartmann, H. A. (1955). *A.M.A. Arch. Path.*, **59**, 706.
- Keith, J., Rowe, R., and Vlad, P. (1958). *Heart Disease in Infancy and Childhood*, p. 352. Macmillan, New York.
- Lam, C. R. (1958). *Modern Medicine*, **26**, 134.
- Lewis, J. J., Varco, R. L., Taufic, M., and Niazi, S. A. (1956). *Surg. Gynec. Obstet.*, **102**, 713.
- Loeffler, E. (1949). *A.M.A. Arch. Path.*, **48**, 371.
- Nash, F. W., and MacKinnon, D. (1956). *Arch. Dis. Childhood*, **31**, 222.
- Niwayama, G. (1960). *Amer. Heart J.*, **59**, 291.
- McLester, J. B., Bush, J. D., and DuBois, I. S. (1940). *Amer. Heart J.*, **19**, 492.
- Parsons, C. G. (1950). *Brit. Heart J.*, **12**, 327.
- Pedersen, A., and Therkelsen, F. (1954). *Amer. Heart J.*, **47**, 676.
- Sawyer, C. G., Pool, R. S., Beck, W. C., and David, L. B. (1957). *Amer. J. med.*, **23**, 798.
- Vineberg, A., and Gialloretto, O. (1956). *Canad. med. Ass. J.*, **74**, 719.