Masked Cor Triatriatum

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When cor triatriatum is the only abnormality in the heart, the features of left atrial obstruction are usually obvious (Belcher and Somerville, 1959). However, when it is complicated by other anomalies such as atrial septal defect or anomalous pulmonary venous drainage, the signs of left atrial obstruction may be masked. It is imperative to recognize the cor triatriatum in such patients, and it has been considered worth while to report two illustrative cases in which the correct diagnosis was made in life, and surgical treatment was planned accordingly.

CASE REPORTS

Case 1. N.C., a girl aged 22 months, attended Dr. Bonham Carter in December 1962. She was the seventh child and a full-term normal delivery from a normal pregnancy. There was no family history of congenital heart disease. All her life she had had recurrent respiratory infections and at 16 months was noted to be breathless after the slightest effort. Breathlessness rapidly increased, with the onset of anorexia, malaise, and sweating. She was admitted to Crumlin Hospital, Dublin, under the care of Dr. Conor Ward who found that she was in heart failure and had signs that suggested the presence of atrial septal defect. Following investigations she was transferred to Great Ormond Street for possible surgical treatment.

On examination at 22 months she was grossly undersized, weight 17 lb. (7.7 kg.).

Peripheral cyanosis and malar flush were present and there was prominent symmetrical bulging of the anterior chest wall (Fig. 1A). The peripheral pulses were minute, the jugular venous pressure raised, and the liver enlarged. The right ventricle was greatly enlarged and the pulmonary artery was palpable. On auscultation a pulmonary ejection click was noted and the second sound was constantly split. A delayed diastolic murmur was audible at the lower left sternal edge, but there was no continuous murmur heard in any part of the chest. The electrocardiogram showed right axis deviation and severe right atrial and right ventricular hypertrophy with T wave inversion from V1 to V5. The

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postero-anterior chest radiograph (Fig. 1B) confirmed the presence of cardiomegaly and pulmonary plethora. Pulmonary oedema was also suggested by the fuzzy appearance in the hilum and lines of congestion in the anterior mediastinum on the lateral chest radiograph. On the penetrated view the carina was splayed, the left bronchus was raised, and the right lower bronchus pushed outwards by what was presumed to be an enlarged left atrium.

TABLE I
RESULTS OF CARDIAC CATHETERIZATION IN CASE 1

	Oxygen saturation (per cent)	Pressure (mm. Hg)
Pulmonary artery ..	78	85/70
Right ventricle ..	69, 71	90/3
Right atrium (mid)	81	a = 8, v = 7, x = 2
Superior vena cava	38	
Inferior vena cava	61	
Pulmonary vein ..	92	
Left atrium ..	90	a = 13, v = 11, x = y = 8

Cardiac catheterization demonstrated a left-to-right shunt at atrial level and severe pulmonary hypertension (Table I). An obstructive lesion on the left side of the heart was suspected and pulmonary artery angiography was performed. This showed (Fig. 2A and 2B) that the pulmonary veins drained normally into the left atrium, which was enlarged, densely filled by opaque media for several frames, and appeared to be communicating with the right atrium. The appearances strongly suggested the presence of a cor triatriatum, so a second injection of contrast media was made into the left atrium. The catheter was passed into the upper part of the left atrium and then entered a part of the left atrium where the pressure was only 2-4 mm. Hg with a poor wave form. Contrast media was injected into this lower left atrial chamber and outlined the small left atrial appendage, left ventricle, and aorta (Fig. 3A and 3B).

These investigations confirmed the presence of a cor triatriatum dividing the left atrium. The child's general condition deteriorated and it was decided to operate as an emergency.

FIG. 1.—Chest radiographs from Case 1. (A) Right lateral view shows pronounced forward bulging of the sternum and lines due to venous congestion in the anterior part of the lung. (B) Postero-anterior view shows cardiomegaly, and prominence of the main pulmonary artery. The lung fields show pulmonary plethora and pulmonary oedema.

Operation (January 1, 1962). Using cardiopulmonary bypass, Mr. D. Waterston opened the right atrium through a bilateral transverse thoracotomy. A 1.5×1.5 cm.² atrial septal defect was present in the fossa ovalis, and below this was another minute atrial septal defect. The upper atrial septal defect opened into the upper part of the left atrium which received all the pulmonary veins. The defect was enlarged and the upper part of the left atrium was explored and found to be separated from the lower part of the left and lower atrium, which contained the mitral valve and appendage, by a septum in which there was a hole approximately 0.5 cm. in diameter. The anomalous septum was widely divided and the atrial septal defect was closed. After operation she developed nodal arrhythmias with periods of complete heart block but otherwise made an uneventful recovery and was discharged without anti-failure therapy. One year later she had no symptoms and had developed normally. There were signs suggesting residual pulmonary hypertension.

Case 2. L.E., a girl aged 4 years, was a full-term normal delivery. She was the product of a normal pregnancy, and there was no history of congenital abnormalities in the family. Apart from her small size nothing abnormal had been noticed until she was aged 3 years, when she developed cough and paroxysmal dyspnoea. This rapidly increased over the following month and she was found to be in congestive heart failure when she attended Dr. Bonham Carter's clinic. On examination she had signs of severe pulmonary hypertension, with small peripheral pulse, raised jugular venous pressure and high A and V waves, hepatomegaly, large overactive

right ventricle, and a loud pulmonary ejection click. The second sound was closely split; the pulmonary element was loud and preceded an inconstant Graham Steell murmur. A variable pansystolic murmur, audible at the left sternal edge, was attributed to tricuspid regurgitation. The electrocardiogram showed severe right atrial and right ventricular hypertrophy with right axis deviation. The chest radiograph (Fig. 4) indicated cardiomegaly, pulmonary oedema with probable plethora in the left lung, and the pulmonary veins in the upper lobes were more prominent than in the lower lobes.

Cardiac catheterization showed a left-to-right shunt entering the left innominate vein and considerable rise in the pulmonary vascular resistance. No pulmonary venous wedge pressure was obtained and the left atrium could not be entered (Table II).

Pulmonary artery angiography demonstrated hemi-anomalous drainage of the pulmonary veins from the left lung into left innominate (Fig. 5) and showed a difference in blood flow through the lungs, which was greater on the left. The diagnosis was considered to be severe pulmonary disease complicating hemi-anomalous pulmonary venous drainage. Two months later she was readmitted with severe pulmonary oedema, orthopnoea, and right heart failure which responded to neptal and rest, but recurred within a few days. When the previous investigations were reviewed, it was noted that the main bronchi were splayed by the left atrium, and on the angiocardigram the left atrium appeared as a dense golf ball and remained opacified on many frames as if it was unable to empty normally due to obstruction (Fig. 5). Another pulmonary artery angiocardigram was performed in order to outline the obstruction more

Operation (February 18, 1964). Mr. D. Waterston explored the heart through a median sternotomy. The presence of hemi-anomalous left pulmonary veins entering the left innominate vein was confirmed. Using total cardiopulmonary bypass, the right atrium was opened, and the fossa ovalis was found to be closed, and

FIG. 2.—Angiocardiogram from Case 1 (right ventricular outflow tract injection). (A) Antero-posterior view shows pulmonary veins draining normally into the left atrium which is densely opacified and enlarged. (B) Lateral projection delineates the enlarged upper left atrial chamber lying posteriorly.

clearly. However, the difference in the blood flow of the two lungs was more marked than previously (Fig. 6A), with further reduction in flow through the right pulmonary vessels which resulted in poor opacification of the left atrium (Fig. 6B).

In view of the child's continued deterioration, it was decided to operate on her for left atrial obstruction.

the atrial septum was incised. The upper part of the left atrium, into which the right pulmonary veins drained, was obstructed by a membrane in which there were two small holes, 2 mm. in diameter. The septum was excised, the mitral valve was seen to be normal, and the atrial septum and atrial wall were closed. It was decided not to correct the anomalous drainage on this occasion

as the left atrial appendage was so small. The pulmonary artery pressure fell from 85 to 40 mm. Hg. during the operation.

Recovery was uneventful and one year later there was no heart failure and no dyspnoea. Section of the re-

FIG. 3.—Angiocardiogram from Case 1 (injection into the lower left atrial chamber). (A) Antero-posterior and (B) lateral views show opacification of the lower left atrial chamber with its appendix, communicating with the left ventricle.

TABLE II
RESULTS OF CARDIAC CATHETERIZATION IN CASE 2

	Oxygen saturation (per cent)	Pressure (mm. Hg)
Pulmonary artery ..	65, 64,	90/40 mean 50
Right ventricle ..	63, 65	90/-8
Right atrium ..	65, 59, 54	a = 7
Inferior vena cava ..	67	
Left innominate ..	69, 71	
Subclavian vein ..	48	
Femoral artery ..	98	95/55 mean 65

moved triatrial septum showed in one part that there was a narrow central band of cardiac muscle separating two layers of endocardium and in another part there was fibro-elastic tissue in which there was patchy mucoid degeneration.

DISCUSSION

It is exceptional for patients with partial anomalous pulmonary venous drainage or secundum atrial septal defect, or both, to present in infancy or early

FIG. 4.—Postero-anterior chest radiography from Case 2. This shows cardiomegaly, caused by dilatation of the right atrium and right ventricle, pulmonary oedema, and prominent upper lobe veins.

childhood with severe disability and a high pulmonary vascular resistance. When they do, a search should be made for an additional lesion causing pulmonary venous obstruction, such as

FIG. 5.—Angiocardiogram from Case 2 (injection made into right ventricle). Late picture (film 14 taken at 6 per second) shows dense opacification of the upper part of the left atrium. In addition, the innominate vein is opacified due to anomalous venous return from the left lung.

stenosis of a common pulmonary vein, cor triatriatum, mitral stenosis, aortic stenosis, or aortic coarctation.

In both patients now described, scrutiny of the plain chest radiograph showed evidence of left atrial obstruction, namely distortion of the bronchi due to left atrial enlargement and signs of pulmonary oedema, including fuzziness around the hilum, fluid in the fissures, and lines of congestion in the anterior portion of the lateral view of the chest radiograph (J. Sutcliffe, 1963, personal observations). This view has been found to be useful for recognizing pulmonary oedema in infants and young children. In both patients left atrial enlargement was slight though bronchial distortion was obvious.

When the presence of left atrial obstruction has been recognized, angiocardiology is necessary to elucidate the cause. The angiocardiological appearances associated with cor triatriatum may be characteristic (Ellis *et al.*, 1964), as in these reported cases, but it may be difficult to distinguish this condition from mitral valve stenosis.

If the left atrial appendage is large and attached to the obstructed chamber, mitral stenosis is the cause of the obstruction, but if it is not seen, the presence of cor triatriatum must be suspected since in this condition the appendage is attached to the lower small unobstructed left atrial chamber (Fig. 3A and 3B). It is rarely possible to recognize the position of the left atrial appendage in patients under 1 year, and thus it is difficult to make this distinction in infancy.

An additional radiological sign in the patient with hemi-anomalous pulmonary venous drainage was

FIG. 6.—Angiocardiogram from Case 2 (pulmonary artery injection). (A) Early picture from antero-posterior series shows difference in blood flow in the lungs, which is increased on the left side. (B) Later film in the same series shows opacification of the left innominate vein due to anomalously draining pulmonary veins from the left lung. The right pulmonary veins and left atrium have not opacified.

the obvious difference in blood flow in the two lungs. The reduction in flow in the right lung presumably resulted from the high right pulmonary venous pressure from the obstruction which caused intense reactive pulmonary arteriolar vasoconstriction on the right side. This appearance was confirmed by pulmonary artery angiocardiography and was more striking on the second angiocardiogram

(Fig. 6A) which showed even poorer filling of the right pulmonary vessels and led to inadequate opacification of the obstructed left atrium. In these circumstances, the fundamental part played by the cor triatriatum may easily be overlooked.

Pulmonary artery angiography should be done in this type of case and where possible should be followed by left atrial angiography if the left atrium can be entered. When an obstructive lesion in the left atrium which, though it may not be much enlarged, remains well opacified for a long time, it may be difficult to distinguish this appearance from total anomalous pulmonary venous drainage into the coronary sinus. Usually in this condition, the dilated coronary sinus lies lower in the cardiac silhouette and does not distort the carina, though it may distort the lower lobe bronchi.

When cor triatriatum coexists with two atrial septal defects, patients may present with the signs of a large atrial septal defect and central cyanosis (Somerville, 1962). However, if there is either one atrial septal defect, or two very small atrial septal defects, left atrial obstruction results, as in Case 1. A continuous murmur was not heard in either patient, though theoretically it should have been. Its absence might have been due to the low systemic output which was further reduced with the onset of heart failure.

The dramatic benefits of removing the left atrial obstruction were shown in both patients despite the presence, in one, of unrelieved hemi-anomalous pulmonary venous drainage. The best approach to treatment of the obstructing membrane in the left atrium is through the right atrium and atrial septum, using cardiopulmonary bypass.

SUMMARY

Two children, aged 22 months and 4 years with cor triatriatum, have been described. Both had, in addition, pulmonary hypertension and one had hemi-anomalous drainage of the left pulmonary veins into the left innominate vein. The accessory membrane in the left atrium was excised under direct vision in both patients. The importance and difficulties of recognizing the left atrial obstruction in young children who present as atrial septal defect with pulmonary hypertension have been emphasized. Diagnostic features on the plain chest radiograph and angiocardiogram have been illustrated.

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ADDENDUM

In the last year, the author has seen two infants, aged 1 month and 5 months respectively, who had hemi-anomalous venous drainage from the left lung into the left innominate vein and severe mitral valve stenosis. Both were under the care of Dr. A. Leatham and Dr. C. Pinckney, and presented a similar clinical picture to the patients described here.

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