

Cor triatriatum with pericardial agenesis

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A case of cor triatriatum associated with pericardial agenesis, which was successfully treated, is reported. Details of the clinical picture and surgical treatment are given, with emphasis on the good prognosis which results when the diagnosis is made before attempting surgical correction.

Cor triatriatum is a rather uncommon congenital malformation; it was first described by Church in 1868. Since then 72 cases have been reported in the literature, of which only 22 have been correctly diagnosed pre-operatively or at operation (Ahn, Hosier, and Sirak, 1968; Enjalbert, Calazel, Bounhoure, Vadhat, Puel, and Meriel, 1966; Lasalle, Ethier, Stanley, and Davignon, 1963; McGuire, Nolan, Reeve, and Dammann, 1965; Neufeld, Pautner, Gueron, Deutsch, and Cohen, 1965; Soulié, Vernant, Corone, Galey, Bouchard, Poisson, and Guérin, 1965).

The association of cor triatriatum with pericardial agenesis has not been reported previously. In the present case the diagnosis of cor triatriatum was made before attempting surgical repair.

CASE REPORT

R. H. J., a 10-year-old boy, was first seen in September 1965 because of coughing episodes. A heart murmur had been heard six months previously and there was a history of repeated upper respiratory infections, exertional dyspnoea, and fatigue.

On physical examination the child was poorly developed, with no evidence of congestive heart failure. The chest was deformed with a prominent precordium. The maximum cardiac impulse was visible and palpable at the sixth left intercostal space over the anterior axillary line. A thrill was felt at the apex and along the sternal border. A grade 2 to 3/4 systolic murmur was heard at the apex. The second heart sound was split and the pulmonary component accentuated. Peripheral pulses were found to be normal with a blood pressure of 120/60 mm. Hg.

Routine laboratory tests were within normal limits.

A chest radiograph showed cardiac enlargement and the heart shadow was displaced to the left, the pul-

monary artery was prominent, and the broncho-vascular markings were increased (Fig. 1).

The electrocardiogram was consistent with right ventricular hypertrophy and enlargement of the left and right atria (Fig. 2).

Cardiac catheterization showed (Table) hypertension of the right ventricle and pulmonary artery. The pulmonary artery wedge pressure was 23 mm. Hg. No shunts were demonstrated. Trans-septal catheterization was unsuccessful.

A selective pulmonary artery angiogram demonstrated a dilated pulmonary artery and veins and delayed filling of the left atrium with a small left ventricle (Fig. 3).

The patient was operated upon with a diagnosis of cor triatriatum on 6 November 1965 through a mid-line sternotomy. The pericardium was absent and the heart was displaced to the left costophrenic angle (Fig. 4). The superior and inferior venae cavae were cannulated as was the ascending aorta by means of an Allen-Jiménez cannula (Jiménez Martínez, Pérez Alvarez, and Argüero Sánchez, 1968).

Total body perfusion was started by means of a disc oxygenator. The right atrium was opened, an intact auricular septum was identified, and three puncture holes of the attempted trans-septal catheterization were seen. An incision was made on the upper portion of the septum opening the accessory chamber. A white greyish membrane with a 6-mm. orifice dividing both left atrial chambers was found; this membranous diaphragm was completely excised (Fig. 5). The mitral valve was normal. The interatrial septum and right atrium were closed. Total perfusion lasted 27 minutes and the blood pressure returned to normal after the procedure.

The post-operative course was uneventful and the patient was discharged from hospital on the 14th post-operative day. The patient has been controlled in the out-patient clinic during the last three years and is asymptomatic. Physical development is now normal. There are no murmurs and the second sound remains split.

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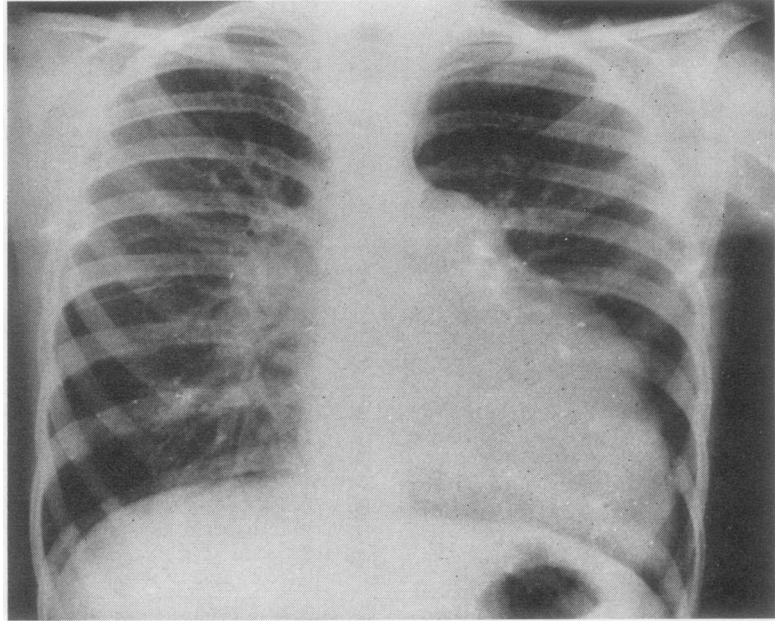


FIG. 1. Postero-anterior chest radiograph showing cardiomegaly with the heart shadow shifted toward the left. There is a flattened left heart border and a well-defined pulmonary artery segment.

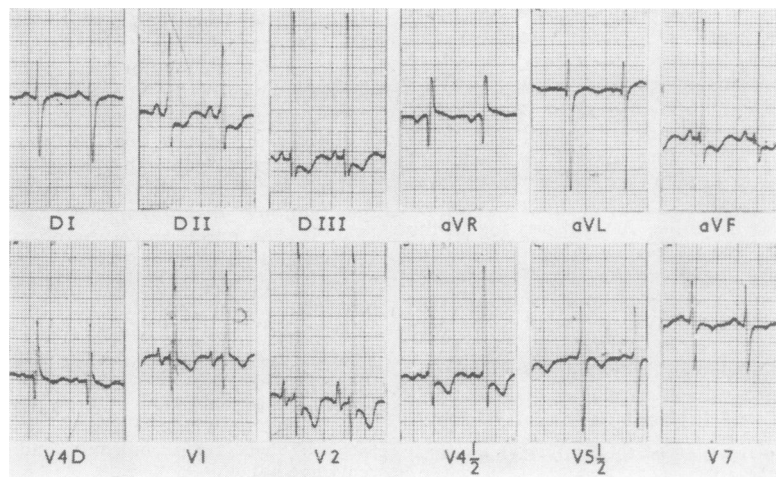


FIG. 2. Electrocardiogram.

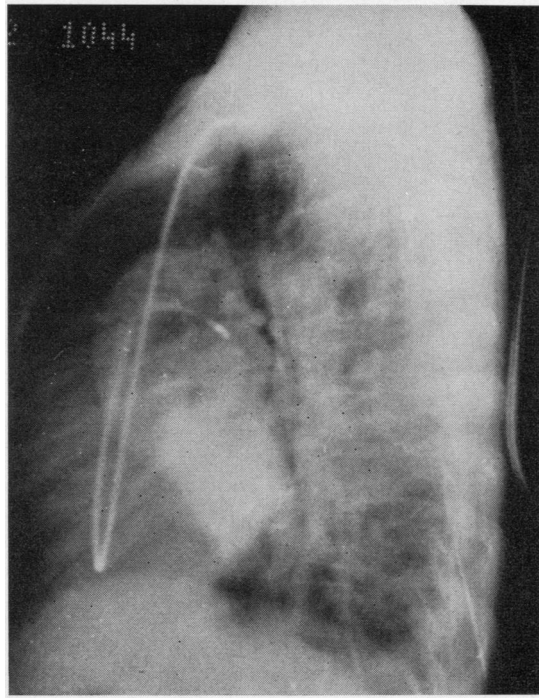
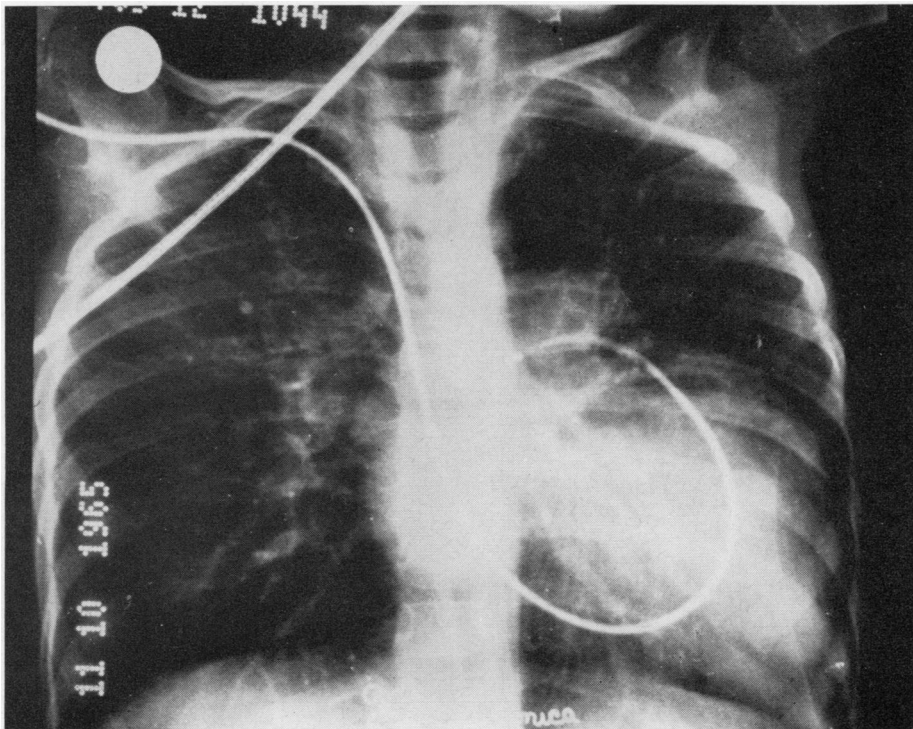


FIG. 3. *Angiocardiogram, postero-anterior and lateral views, shows the third chamber filled and an empty space in between this and the left ventricle.*



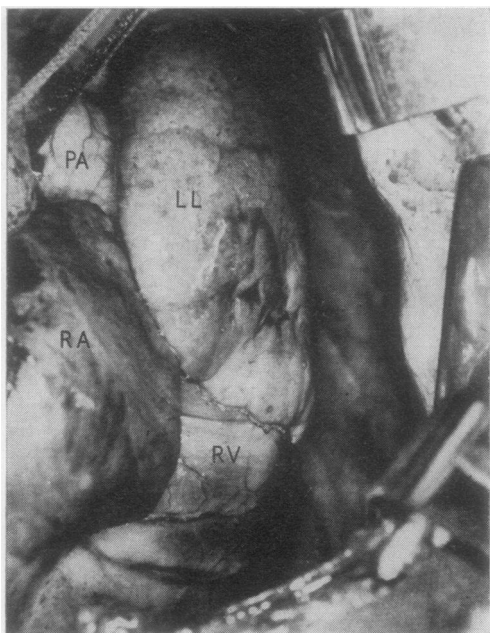


FIG. 4. Photograph taken during operation shows the heart displaced to the left and absence of the pericardium. Pulmonary artery (PA), right atrium (RA), right ventricle (RV), and left lung (LL).

FIG. 5. Shows the excision of the diaphragm (D) with a 6-mm. hole communicating the accessory atrial chamber (AACH) with the true left atrium (LA).

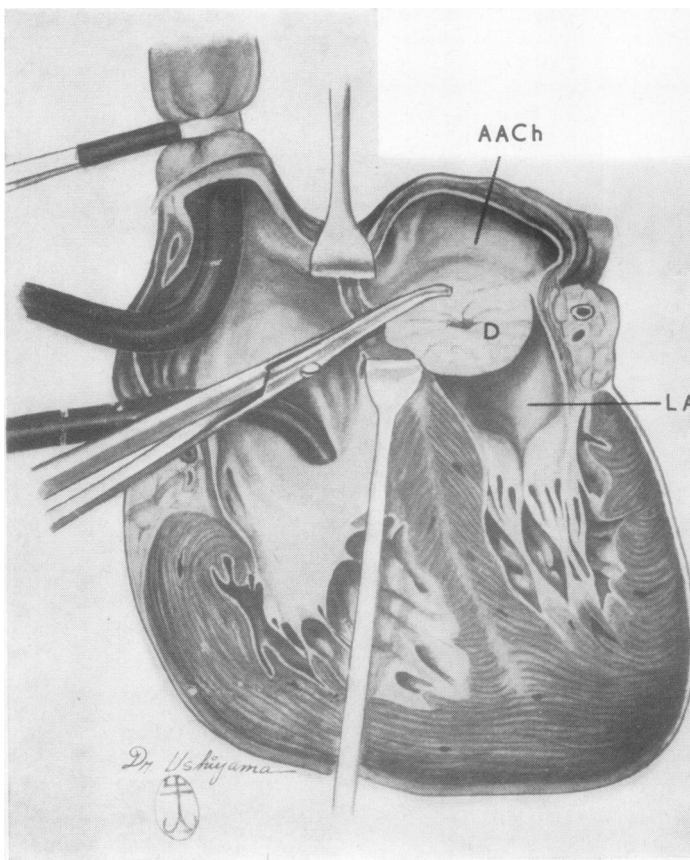


TABLE
CATHETERIZATION DATA

Cavity	O ₂ (vol.)	Satura- tion (%)	Pressure (mm.Hg)		
			Sys- tolic	Dias- tolic	Mean
Right atrium ..	10.7	65	—	—	5
Right ventricle ..	10.6	64	77	5.5	—
Main pulmonary artery	10.9	66	77	40	58
Right pulmonary artery ..	—	—	76	46	57
Wedge ..	—	—	—	—	23
Femoral artery ..	15.5	93	114	69	85
Haemoglobin ..	11.8	—	—	—	—

COMMENTS

Cor triatriatum is a congenital anomaly of the heart in which all the pulmonary veins drain into an accessory chamber which is incompletely communicated, due to failure of the common pulmonary vein to become incorporated with the left atrium in a normal manner (Edwards, DuShane, Alcott, and Burchell, 1951). In the present case it was also associated with pericardial agenesis resulting from incomplete development of the pleuro-pericardial folds (Ellis, Leeds, and Himmelstein, 1959).

Since this was first described by Church (1868), several authors (Ahn *et al.*, 1968; Lacquet, Los, Nauta, and Brom, 1966; Niwayama, 1960; Neufeld *et al.*, 1965) have reviewed the literature. The first successful operation was performed by Vineberg and Gialloretto in 1956; since then, 22 patients have been diagnosed at operation or pre-operatively (Ahn *et al.*, 1968; Enjalbert *et al.*, 1966; Lasalle *et al.*, 1963; McGuire *et al.*, 1965; Neufeld *et al.*, 1965; Soulié *et al.*, 1965).

Of this group of patients, only three (Lasalle *et al.*, 1963; McGuire *et al.*, 1965; Miller, Ongley, Anderson, Kincaid, and Swan, 1964) died in the immediate post-operative period. Seven additional patients, operated upon with an incorrect diagnosis, died during or following surgery (Abedrop, Espino Vela, and Correa, 1961; Beller, Childers, Eckner, Duchelle, Ranniger, and Rabinowitz, 1967; McGuire *et al.*, 1965). Thus a correct diagnosis is an indispensable step in the correction of this anomaly.

The clinical picture is that of mitral stenosis with variable clinical manifestations, depending on the degree of stenosis of the communication with the true left atrium.

Usually, dyspnoea on exertion, failure to thrive, and cyanosis are present, as in our patient, and the more common physical findings are apical murmurs, right ventricular hypertrophy, and accentua-

tion of the pulmonary closure sound. These findings are also described in other anomalies such as congenital mitral stenosis and stenosis of the pulmonary veins. Several types of heart murmur have been reported; but a systolic murmur at the left sternal border and at the apex has been described most often.

Chest radiographs may show varying degrees of cardiac enlargement, a prominent pulmonary artery, and evidence of pulmonary venous hypertension.

In our patient the typical picture of pericardial agenesis was added (Dimich, Grossman, Bowman, and Griffiths, 1965; Ellis *et al.*, 1959; Tubbs and Yacoub, 1968). The electrocardiographic findings usually show severe right ventricular hypertrophy. The catheterization studies showed an elevated right ventricle, pulmonary artery, and wedge pressures. During catheterization an elevated pressure in the pulmonary veins and accessory chamber is highly suggestive of cor triatriatum. A selective angiogram of the pulmonary artery is probably the best way to demonstrate the lesion. It usually shows a dilated pulmonary artery and veins and delayed emptying of the accessory chamber.

Cor triatriatum is easily corrected surgically with the aid of extracorporeal circulation.

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