

Surgical treatment of cor triatriatum in a 4½-month-old infant¹

H. OELERT, I. M. BRECKENRIDGE, G. ROSLAND,
and J. STARK²

*The Thoracic Unit, Hospital for Sick Children, Great Ormond Street, London,
and The Haukeland Sykehus, Bergen, Norway*

Successful correction of cor triatriatum in a 4½-month-old infant is presented. Altogether six patients with cor triatriatum underwent surgical correction during the last eight years. All but one survived the operation. The clinical features and the surgical treatment are discussed.

Cor triatriatum is an uncommon congenital malformation which may cause severe symptoms and often death early in life (Ahn, Hosier, and Sirak, 1968; Jorgensen *et al.*, 1967). There have been some 24 reports of successful surgical correction (Ahn *et al.*, 1968; Jorgensen *et al.*, 1967; Jimenez Martinez *et al.*, 1969; Perry, Scott and McClenathan, 1967; Wolfe, Ruttenberg, Desilets, and Mulder, 1968), the youngest patient being 3½ months old (Wolfe *et al.*, 1968).

The classical form of triatrial heart is characterized by a fibromuscular septum subdividing the left atrium into an upper compartment which receives the pulmonary veins and a lower compartment, incorporating the left atrial appendage, which leads into the left ventricle through the mitral valve. Obstruction to pulmonary venous return may occur at the opening or openings in the intra-atrial septum or membrane, and the haemodynamic features then resemble those of mitral stenosis. As demonstrated by van Praagh and Corsini (1969), this type of cor triatriatum results from failure of the common pulmonary vein to be incorporated into the true left atrium during the fifth embryonic week.

Ahn and colleagues (1968) reviewed a series of 18 reported cases in whom the correct diagnosis was made at or before operation, with only one death. Successful operation has subsequently been reported in two infants under 1 year of age (Perry *et al.*, 1967; Wolfe *et al.*, 1968). In view of the

unsatisfactory natural history and the possibility of successful surgical treatment of triatrial heart at all ages, early operation is advised. In this paper we report the surgical correction of triatrial heart in a critically ill 4½-month-old infant.

CASE REPORT

The patient (R. F.), a Norwegian boy, was born after a normal pregnancy and delivery with a birth weight of 2.95 kg. He was dyspnoeic and slow to feed, but never cyanosed. In hospital (Bergen), cardiac failure was treated with digoxin and diuretics, and cardiac catheterization and angiography disclosed pulmonary hypertension due to an obstructing membrane in the left atrium. A mild coarctation of the aorta was noted at the same time. He was admitted for operation to the Hospital for Sick Children, Great Ormond Street, London, on 6 May 1971, at the age of 4 months, weighing 4.65 kg. Pallor and dyspnoea were noted but no cyanosis. All pulses were easily palpable, while systolic blood pressure was 130 mmHg in the right arm and 80 mmHg in the right leg. The second

TABLE I
CARDIAC CATHETERIZATION DATA OF 6 PATIENTS WITH TRIATRIAL HEART BEFORE OPERATION

Patient	RA	RV	MPA	PA wedge	LA	LV	Systemic	Qp:Qs
1 N.C.	3	90/3	85/45		10			> 3:1
2 M.A.	8	55/0	40/20	25/			130/90	> 3:1
3 L.E.		90/-8	90/40				95/55	
4 G.F.	7	80/8			̄ 100/6			
5 P.B.	̄	50/6	54/30	15/5	̄ 116/6	120/70		2.5:1
6 R.F.	±	80/10	80/40	30/20	100/4-8		Asc. aorta: 110/60 (m = 80) Desc. aorta: 80-85/60 (m = 70)	

(Pressures in mmHg) RA = right atrium; RV = right ventricle; MPA = main pulmonary artery; LA = left atrium; LV = left ventricle; ̄ = mean pressure of 2 mmHg; Qp:Qs = pulmonary to systemic flow ratio.

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²Correspondence to: Mr. J. Stark, Thoracic Unit, The Hospital for Sick Children, Great Ormond Street, London W.C.1

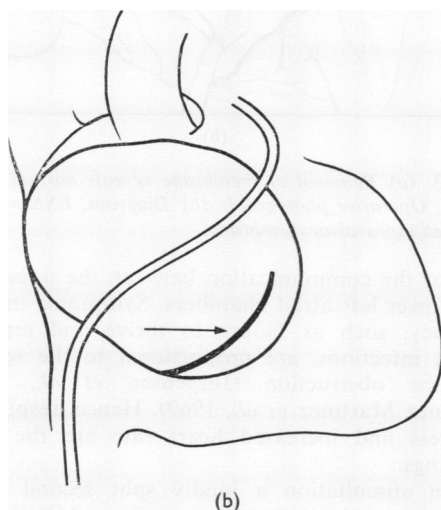
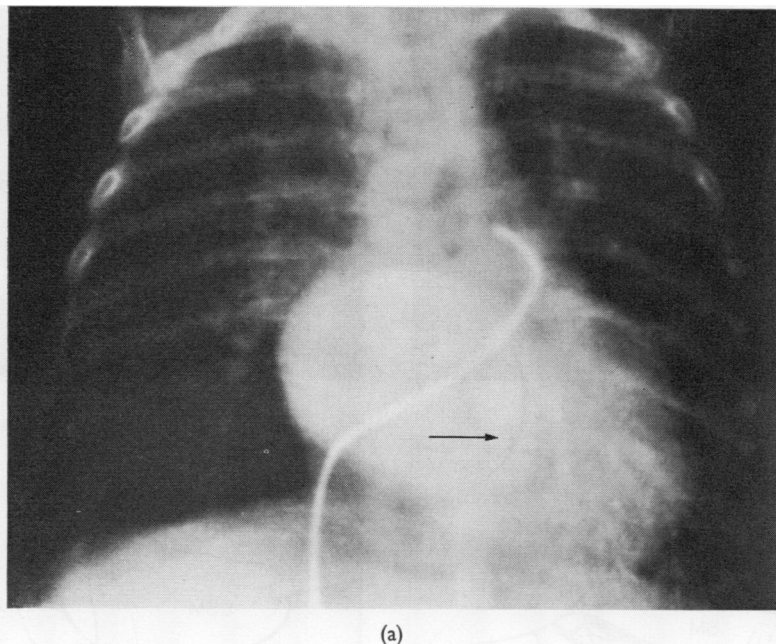


FIG. 1. (a) Angiogram showing shelf-like opacity (arrowed) in large left atrium. (b) Diagram of shelf-like opacity.

sound was split and a third sound was audible at the left sternal edge and at the apex. A grade 2/6 pansystolic murmur could be heard at the mitral area and between the scapulae. The liver edge was palpable 2 cm below the right costal margin.

Haemoglobin was 8.5 g/100 ml (PCV 33%); serum electrolytes and blood urea were normal. An electro-

cardiogram showed normal sinus rhythm with biventricular hypertrophy, while a chest film revealed some increase in heart size, particularly the left atrium, with pulmonary plethora.

Cardiac catheterization data are shown in Table I. Dye injection into the main pulmonary artery outlined a large left atrium which emptied very slowly. A shelf-like opacity just upstream to the mitral annulus obstructed the flow of contrast medium (Fig. 1a and b). The left ventricle contracted normally with no mitral regurgitation. The coarctation was seen and a systolic gradient of 25–30 mmHg was measured between the ascending and descending aorta.

OPERATION (20 May 1971) Through a right thoracotomy, the left atrium was found to be bulging and tense. The patient was connected to the heart-lung machine using superior vena cava and inferior vena cava cannulation for the venous drainage and aortic cannulation for the arterial return. On partial bypass a left ventricular vent was inserted through the apex of the heart, and when full flow was achieved (2.4 l./m²/min) total bypass was instituted. The aorta was cross-clamped at 30° C and the left atrium was incised parallel to and behind the interatrial groove. A thick membrane stretched across the chamber above the mitral valve annulus (Fig. 2a and b). It was perforated by a lateral slit some 6 mm in length which led into the dorsal chamber whereas the left atrial appendage opened just below the membrane into the ventral chamber. The membrane, 2.5 × 1.5

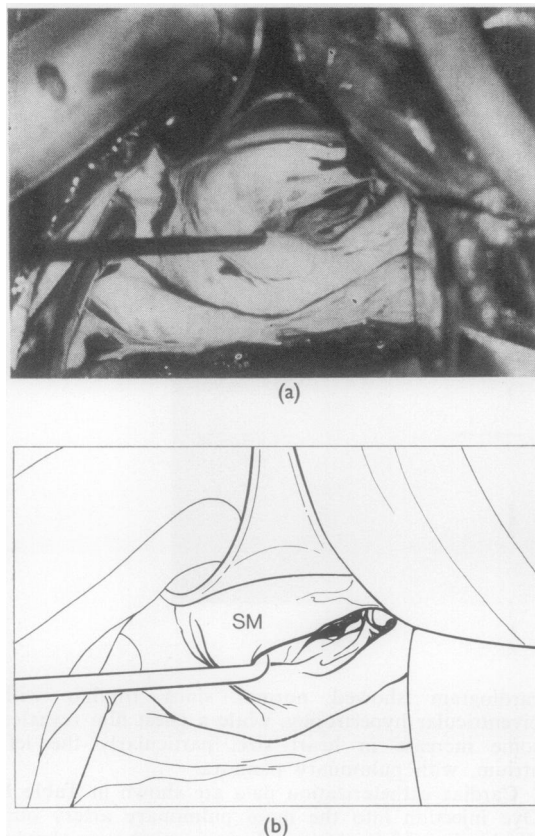


FIG. 2. (a) Operative view of membrane in left atrium with mitral valve seen through slit. (b) Diagram of operative view of membrane. SM=supra-valvar membrane.

cm in size, was excised (Fig. 3a and b), connecting the two atrial compartments into one single cavity and revealing a competent mitral valve. Rewarming was started, the left atrial incision closed, and bypass discontinued at normal temperature. After bypass cardiac output was satisfactory, with an arterial blood pressure of 100/70 mmHg and a mean left atrial pressure of 13 mmHg.

Microscopic examination of the membrane showed loose connective tissue covered by attenuated endocardium.

The postoperative course was uneventful and no further medication was required. The child left the hospital 13 days after operation. Follow-up examination six months later showed that the coarctation had become more significant and will require resection.

DISCUSSION

Pulmonary congestion and low cardiac output in cor triatriatum depend upon the degree of narrow-

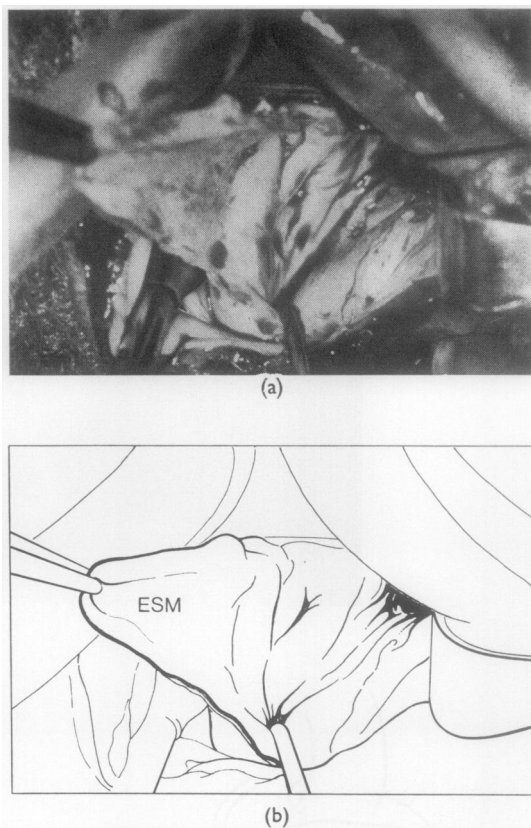


FIG. 3. (a) Removal of membrane reveals normal mitral valve. Operative photograph. (b) Diagram. ESM=partly excised supra-valvar membrane.

ing of the communication between the upper and the lower left atrial chambers. Symptoms in early infancy, such as failure to thrive and repeated chest infections, are proportional to the severity of the obstruction (Jorgensen *et al.*, 1967; Jiminez Martinez *et al.*, 1969). Hence, respiratory distress and increased heart rate are the usual findings.

On auscultation a loudly split second sound indicates pulmonary hypertension. Mitral diastolic murmurs are seldom heard, presumably because cardiac output is low.

The electrocardiogram consistently shows right ventricular hypertrophy. A coarctation in the present case caused left ventricular hypertrophy as well. On chest radiography right ventricular enlargement and signs of pulmonary congestion are seen (Jiminez Martinez *et al.*, 1969; Somerville, 1966; Wolfe *et al.*, 1968). Cor triatriatum is an uncommon cause of pulmonary oedema in

infancy. Pulmonary congestion, giving rise to pulmonary hypertension, is associated with a variety of obstructing lesions of the left heart, such as stenosis of a common pulmonary vein, mitral valve stenosis, aortic stenosis, coarctation of the aorta, and hypoplastic left heart syndrome.

The correct diagnosis of cor triatriatum is made only by cardiac catheterization and angiocardiology (Ahn *et al.*, 1968; Jorgensen *et al.*, 1967; Lam, Green, and Drake, 1962) when pulmonary arterial and venous hypertension without detectable shunts are found. If the left atrium can be entered, the finding of a low left atrial pressure in the presence of elevated pulmonary capillary wedge pressure localizes the obstruction to the pulmonary veins or upper left atrium (Perry *et al.*, 1967; Wolfe *et al.*, 1968). Low systemic venous saturations indicate low cardiac output. Angiocardiology, which outlines the left-sided chambers of the heart, is necessary to reveal the anatomy of the obstruction. In our case, the foramen ovale was no longer patent and the true left atrium was therefore not entered. Hence, a pulmonary artery injection was used to opacify the pulmonary veins. With the obstructing lesion in the left atrium, their drainage into an upper pulmonary venous chamber was easily detected. This compartment remained well opacified throughout a long period while the left ventricle filled poorly. A shelf-like shadow was localized just above the mitral annulus, demonstrating the obstructing membrane, subdividing the left atrium into two cavities.

If the correct diagnosis is made, the operative mortality is low (Ahn *et al.*, 1968), whereas the prognosis without operation is extremely poor even with mild symptoms in infancy (Ahn *et al.*, 1968; Jorgensen *et al.*, 1967). As soon as the diagnosis of triatrial heart is established, removal of the obstructing membrane is therefore indicated (Jorgensen *et al.*, 1967; Jiminez Martinez *et al.*, 1969; Wolfe *et al.*, 1968). Several attempts to relieve the obstruction by closed procedures have failed (Jorgensen *et al.*, 1967; Wolfe *et al.*, 1968). Total cardiopulmonary bypass with complete excision of the subdividing membrane is the procedure of choice. The heart may be exposed through a median sternotomy but we prefer a right thoracotomy. Triatrial heart with associated lesions is probably best approached by a transverse bi-atrial incision, opening both the right and the upper left atrium at the same time. Good access to an isolated supramitral valve membrane, however, is obtained through the left atrium by an incision parallel to and behind the interatrial groove. After excision of the obstructing

membrane, the mitral valve can be examined.

Once the left atrial obstruction is excised, excellent symptomatic relief follows (Ahn *et al.*, 1968; Wolfe *et al.*, 1968). Pre- and postoperative investigations have shown that pulmonary capillary wedge and pulmonary arterial pressures return to near normal values in 9 to 10 months following operation (Ahn *et al.*, 1968; Gialloreto and Vineberg, 1962; Jorgensen *et al.*, 1967; Miller *et al.*, 1964). Surgical correction in early infancy may prevent the persistent elevation of pulmonary arteriolar resistance reported in older patients (Jorgensen *et al.*, 1967; Miller *et al.*, 1964).

Six patients with triatrial heart were operated on between 1962 and 1971 at the Hospital for Sick Children, Great Ormond Street, London. All had associated cardiac lesions, the commonest being an ASD (Table II). The haemodynamic data

TABLE II

TRIATRIAL HEART: ANALYSIS OF 6 PATIENTS TREATED SURGICALLY AT THE HOSPITAL FOR SICK CHILDREN, GREAT ORMOND STREET, LONDON BETWEEN 1962 AND 1971

Patient	Age	Sex	Additional Lesion	Date of Operation	Result
1 N.C.	22 mth	F	ASD secundum	Nov. 8, 62	Alive
2 M.A.	13 yr	F	" "	May 23, 63	Died after second operation (see text)
3 L.E.	4 yr	F	Left PAPVD to left SVC	Feb. 18, 64	Alive
4 G.F.	11½ yr	F	AV canal	June 30, 66	Alive
5 P.B.	7½ yr	F	ASD secundum	May 8, 67	Alive
6 R.F.	4½ mth	M	Coarctation of aorta	May 20, 71	Alive

ASD = atrial septal defect; AV canal = atrioventricular canal; SVC = superior vena cava; PAPVD = partial anomalous pulmonary venous drainage.

of these six patients are summarized in Table I. Their ages ranged from 4½ months to 13 years. In all of them the obstructing membrane was removed using cardiopulmonary bypass. One patient died following a second operation for recurrent pulmonary venous obstruction. Two of the five survivors have been reported earlier and the difficulties in establishing the diagnosis when triatrial heart is complicated by other anomalies emphasized (Somerville, 1966). The patient described here is the second infant under 6 months of age (Wolfe *et al.*, 1968) known to have successfully undergone total correction of cor triatriatum.

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