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MANAGEMENT OF CONGENITAL RIGHT CORONARY ARTERY TO RIGHT ATRIAL FISTULA

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INTRODUCTION:

CONGENITAL CORONARY ARTERY to cardiac chamber fistula is a rare condition. Krause initially reported a fistulous communication between a coronary artery and a cardiac chamber in 1865. The first surgical correction of this condition was performed by Bjork and Crafoord (1947) when they successfully ligated a fistula between a branch of the left coronary artery and the pulmonary artery. The need for surgical closure of the fistula is clear in symptomatic patients. When the lesion is discovered in infancy in symptomless patients, it is generally accepted that the operation should be performed electively during childhood as the mortality and morbidity following closure of the fistula has been shown to be low (Rittenhouse *et al.*, 1975).

We wish to report on a patient with a right coronary artery to right atrial fistula successfully operated on at the University Hospital, Kuala Lumpur.

CASE REPORT:

N.E.O. is a four year old male child who was initially discovered to have a heart murmur at the age of six months when he was seen for a respiratory tract infection. Apart from frequent respiratory infections he was asymptomatic when initially assessed at the age of 8 months.

Physical examination revealed that he was an active child. The peripheral pulses were of a large volume and the arm blood pressure was 90/40 mm of Hg. There was cardiomegaly associated with a forceful apex beat. A thrill was palpable in the second right intercostal space parasternally. Auscultation revealed a grade 3/6 continuous murmur maximally heard over this same area. Chest radiographs showed that the heart was enlarged and there was pulmonary plethora. The electrocardiogram was within normal limits.

The child was followed up and when he was reassessed at the age of 3 years he was still asymptomatic apart from an increase in respiratory tract infections. With the exception of obviously collapsing pulses and clinical left ventricular hypertrophy confirmed on electrocardiography, the physical signs remained essentially unchanged.

In view of these progressive changes clinically and on the electrocardiogram, a right heart catheterization was performed on the 30th July, 1976. This revealed a step up of oxygen saturation at right atrial level with a pulmonary-to-systemic flow ratio of 2: 1. The pulmonary arterial wedge pressure and pulmonary artery pressure were 20 mmHg and 52/25 mmHg (mean 38 mmHg) respectively, while the pulmonary vascular resistance was 192 dyne-sec-cm^5. On the 23rd November 1976 a left heart catheterization was performed. The left ventricular and aortic pressures were 105/0 mmHg and 105/52 mmHg respectively. There was no systemic arterial desaturation. Aortography revealed a large right coronary artery (RCA) to right atrial fistula (Fig. 1). The proximal RCA and its sino-atrial node branch which emptied into the upper right atrium were noted to be grossly dilated. The distal RCA and the left coronary artery appeared normal. Left ventricular angiography excluded the presence of a ventricular septal defect.

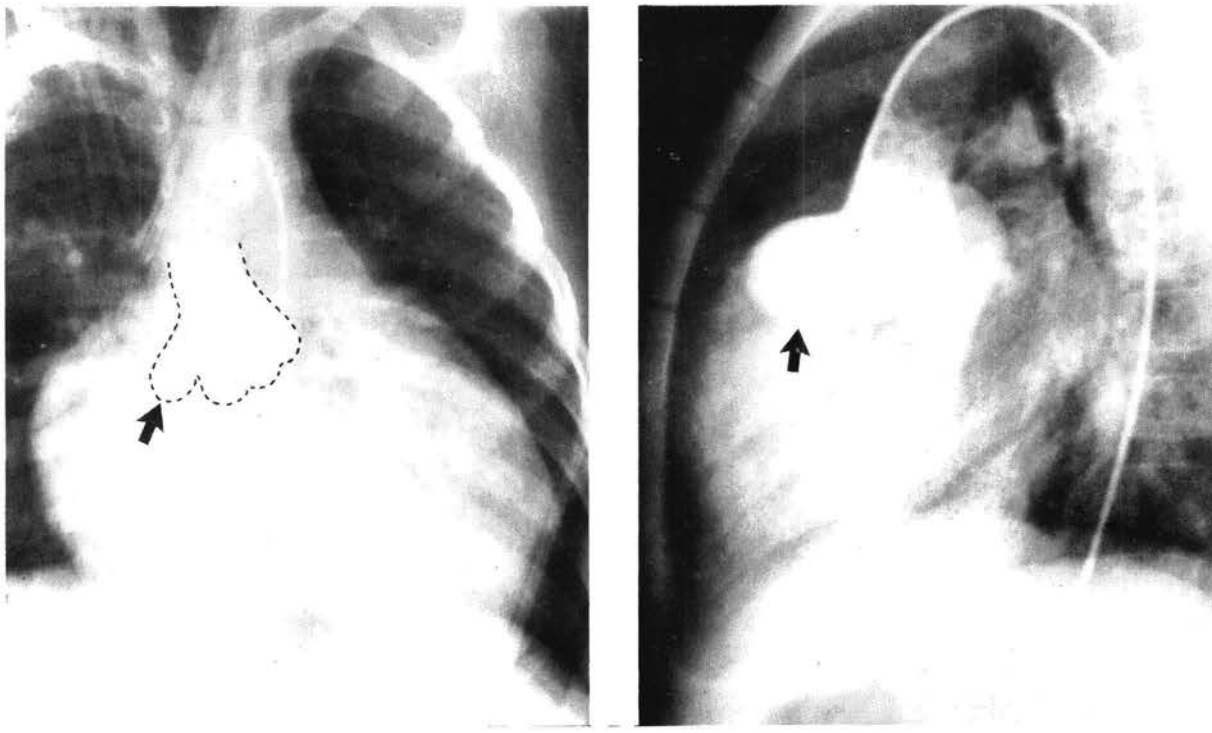

Figure 1: Aortogram showing the dilated proximal right coronary and sinoatrial node arteries (arrowed). (A) P.A. view. (B) lateral view.

In view of the presence of marked elevation of the pulmonary arterial wedge pressure, raised pulmonary arterial pressure, a large pulmonary to systemic flow ratio and progressive changes on electrocardiography the patient was digitalised. At thoracotomy through a midsternotomy incision on the 7th of December 1977, the proximal RCA and its sinoatrial node branch were both noted to be dilated (7mm. diameter). The distal RCA was of normal calibre (Fig. 2). The sinoatrial node artery was mobilised and ligated with multiple heavy silk ligatures so as to obliterate it throughout its entire length. The child tolerated this procedure well and had an uneventful postoperative course.

DISCUSSION:

In a collected series of 163 patients and 8 personal cases of congenital coronary artery to cardiac chamber fistula, Rittenhouse and associates (1975) noted that 89 fistulae originated from the RCA, 66 from the left coronary artery and 8 from both coronary arteries. The right ventricle was also found to be the commonest site of termination (44%), while a right atrial termination was the next in order of frequency (22%). Other sites of termination include the pulmonary artery (17%), the coronary sinus (8%), the left atrium (4%), the superior vena cava (2%), the left ventricle (2%) and the pulmonary vein (1%).

This case illustrates the clinical course in the majority of cases. Symptoms in childhood are exceptional. In contrast, they may arise later in life in untreated patients when the shunt volume is large. In the collected series reviewed by Rittenhouse *et al.* (1975), 59% of patients were symptomless. Dyspnoea at rest or effort, fatigue, features of heart failure, angina, palpitations, bacterial endocarditis, pulmonary hypertension and frequent respiratory infections formed the presenting features or conditions in symptomatic patients. The most frequent and diagnostically useful physical sign is the presence of a continuous murmur which differed from that of a patent ductus arteriosus (P.D.A.) in that it is best heard in the third or fourth intercostal space at the left or right parasternal edges. Other than a P.D.A., the differential diagnosis entertained should include aorto-pulmonary window, aortic

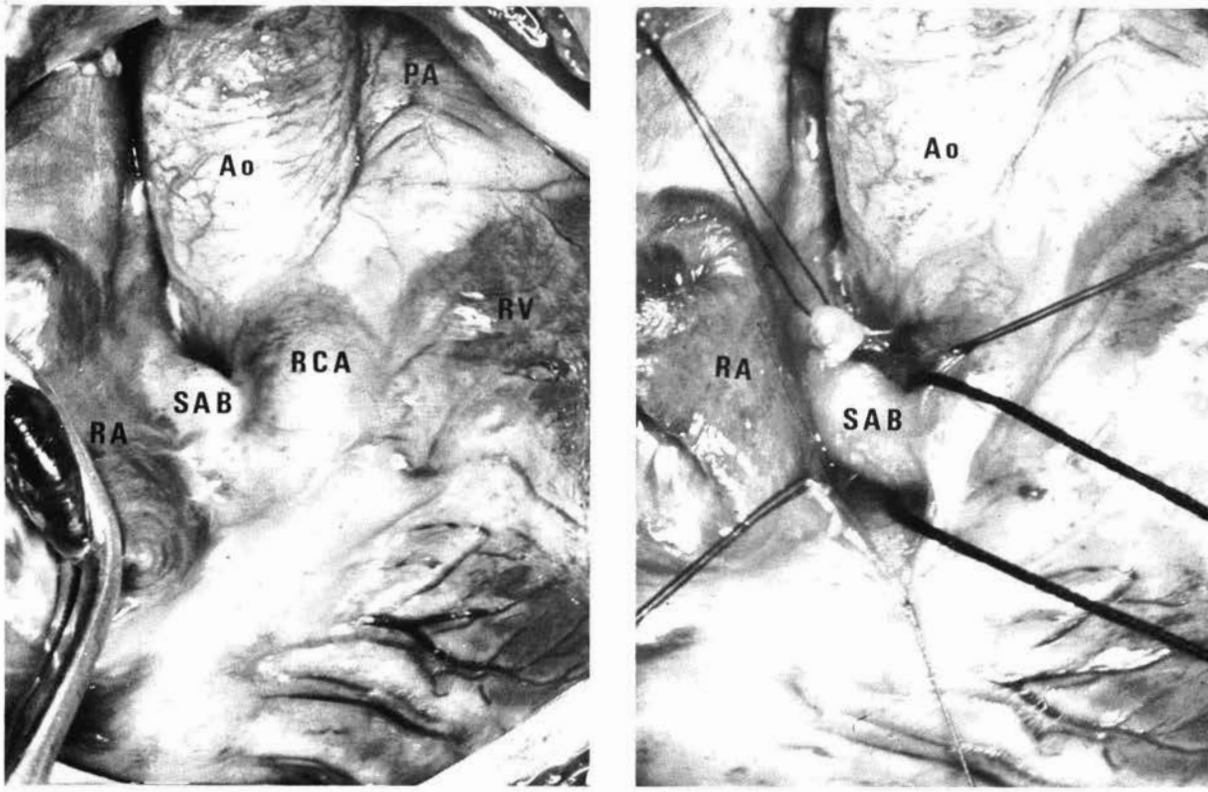

Figure 2: Operative Findings
(A) Before mobilization of the sinoatrial node artery.
(B) After freeing the artery.

(AO = Aorta, PA = Pulmonary Artery, RA = Right Atrium, RCA = Right Coronary Artery, RV = Right Ventricle, SAB = Sino-atrial Node Branch)

sinus of valsalva fistula and ventricular septal defect with aortic regurgitation. Newfeld *et al.* (1961) have noted that the site of maximum intensity of the murmur may vary according to the chamber in which the fistula terminates. Fistulae draining into the right ventricle may have the murmur best heard along the left sternal edge at the fourth or fifth intercostal space parasternally. For right atrial fistulae and fistulae terminating in the pulmonary artery and left atrium the murmur is best heard in the second right intercostal space parasternally as is borne out in our patient. Left ventricular fistulae usually have murmurs which are best heard along the right fourth or fifth intercostal space.

Pathologically, fistulae may occur at any point along the course of a coronary artery. In fistulae from the sinoatrial node artery, the artery may originate from the RCA and course between the aortic root and the right atrium as was the case in our patient (Fig. 3A). More infrequently, the anomalous sinoatrial artery may arise from the left circumflex coronary artery and follow a course through the transverse sinus. (Fig. 3B).

Aortography followed by surgical correction was advocated by Haller and Little (1963). It is presently generally accepted that aortography is essential for definitive diagnosis and identification of the arteries involved and for confirming the site of termination of the fistula.

In most cases simple ligation of the fistula will adequately deal with the problem; if an aneurysm of the coronary artery is present as a result of a long standing fistula, Cooley and Norman (1975) are of the opinion that it should be excised.

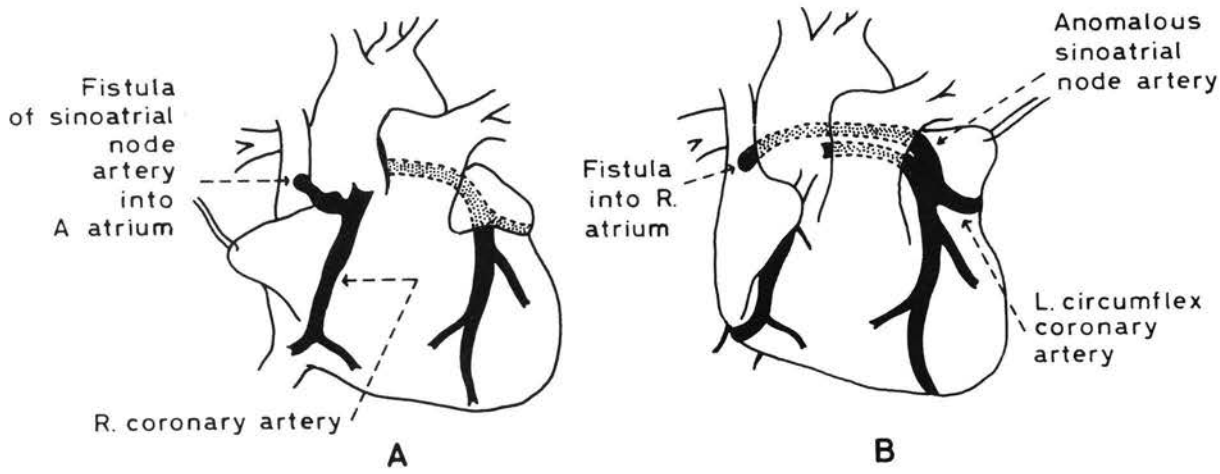

Figure 3: Illustration to show the course normally taken by the sinoatrial node artery.

SUMMARY:

The management of congenital right coronary artery to right atrial fistula in a 4 year old child initially diagnosed at 8 months of age is discussed. The effect of chronic circulatory overload from a large left to right shunt required surgical interruption of the fistula at the age of 4 years. The most consistent clinical clue is a continuous murmur at an atypical site. Cardiac catheterization and aortography are essential for a definitive diagnosis. Surgical correction can be successfully carried out with low risks in the majority of patients and hence should be performed in all symptomatic patients and electively in symptomless children to avoid circulatory problems associated with a long standing cardiac shunt.

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