

## CASE REPORT

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### CONGENITAL CYANOTIC HEART DISEASE WITH «SITUS SOLITUS», DEXTROVERSION, JUXTAPOSITION OF THE ATRIAL APPENDAGES AND ANATOMICAL MALPOSITION OF THE GREAT ARTERIES. REPORT OF A CASE WITH SURGICAL CORRECTION

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#### SUMMARY

A case of complex congenital cardiac malformation with successful corrective surgery is described. Comment on classification is made and the importance of accurate pre-operative diagnosis is stressed.

Associated congenital cardiac malformations with cyanosis are frequently observed and, in some cases, depending on the degree of anoxia and on the state of the pulmonary circulation, are compatible with a fairly long life span. Pre-operative diagnosis and accuracy in anatomical classification are essential for successful corrective surgery as shown in a case of cyanotic heart disease with situs solitus, dextroversion, juxtaposition of the atrial appendages, concordant atrio ventricular and ventriculo-arterial connections, atrial septal defect, multiple ventricular septal defects, valvar and subvalvar pulmonary stenosis and anatomical L malposition of the great arteries.

#### CASE REPORT

F. F. S., male, aged 23. Normal delivery. Cyanosis since birth, worse when crying and during frequent respiratory infections. Remembers having been told that his heart was on the right chest. Breathlessness on exertion which also increased cyanosis. No squatting. Aged 7, the patient was admitted to another Hospital where catheterization was carried out. Surgery was not indicated at the time. Worked in light jobs without any further deterioration until age 22, when he was admitted to the Department of Cardiology of the Lisbon City Hospitals. Right and left heart catheterization and angiograms were performed. He was, then, accepted for surgical correction of his congenital heart condition.

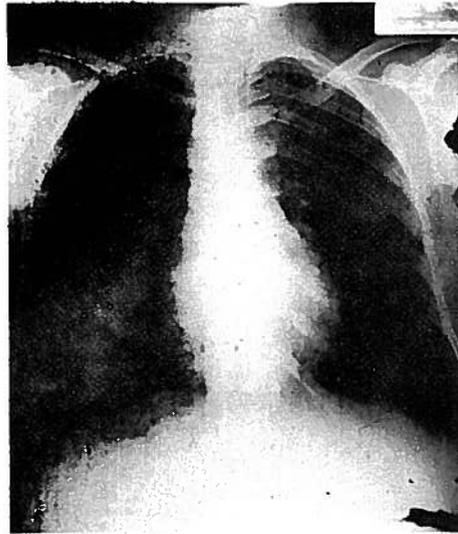


Fig. 1 — Chest X-ray: Centrally placed heart, behind the sternum

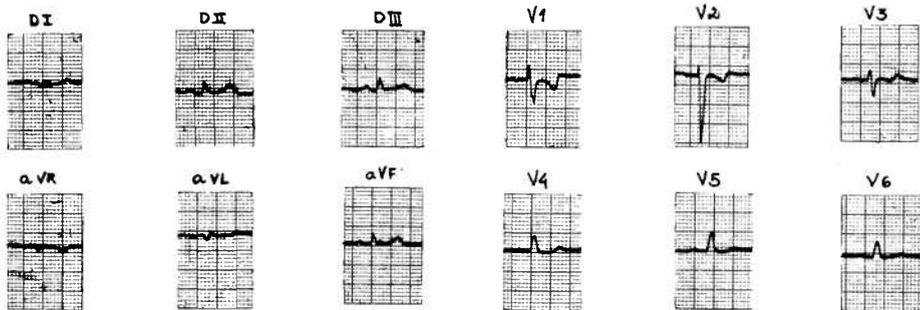


Fig. 2 — ECG: Sinus rhythm. Low voltage P wave and QRS. QRS 0.10 and slurred. Slow progression of R waves in precordial leads. ST negative in  $V_1$  and  $V_2$ .

TABLE I  
Heart catheterization

	RA a wave	LA a wave	RV	LV	Fem. art.
Pressures	7,5 mm Hg	7,5 mm Hg	87 mm Hg	87 mm Hg	—
$O_2$	66 %	95 %	67 %	95 %	88 %

$O_2$  Capacity 14,5 mm Hg — 19,2 %

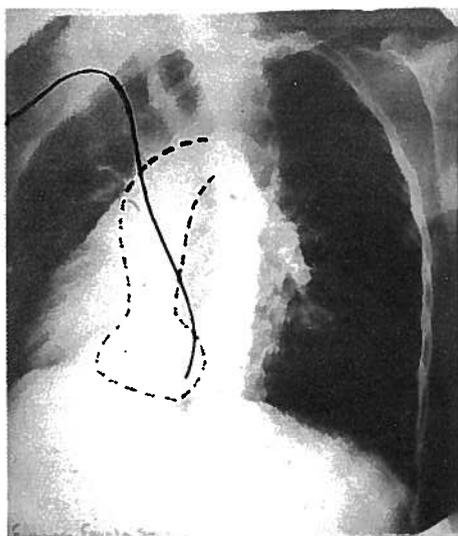


Fig. 3 — Right (anterior) ventricle injection showing little, if any, trabeculation, pulmonary stenosis and VSD

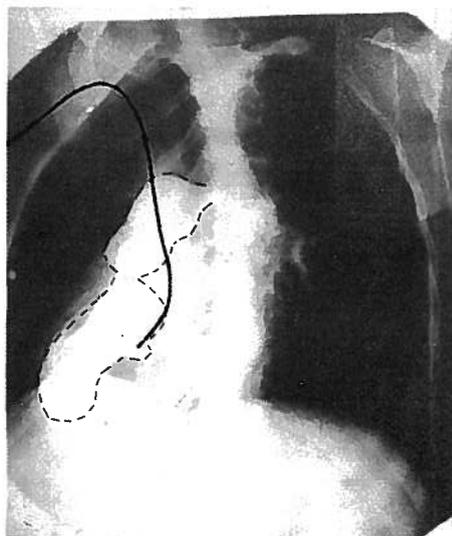


Fig. 4 — Left (posterior) ventricle injection. Aorta to the left of pulmonary artery

Thin, short, very cyanotic young man, with clubbed fingers and toes. Lungs: nothing abnormal detected. Heart: Harsh systolic murmur (4/6) on the right of the sternum more audible over the right 3rd and 4th inter costal spaces. Softer systolic murmur (2/6) over the 4th left inter costal space near the sternum. Blood pressure 120/70. No more abnormalities detected in the rest of the examination. Blood count: Red Cells 7.4 m. Haemoglobin 24.5 g. Haematocrit 66 %. Chest X Ray (Fig. 1). Heart placed behind the sternum in slight dextroversion. EKG (Fig. 2): Sinus rhythm, 72/min. Low voltage P wave and QRS. QRS — 0.10 and slurred. Slow progression of R waves in precordial leads. ST negative in  $V_1$  and  $V_2$ . Cardiac catheterization: Table I. Right and left angiograms (Figs. 3 and 4). Injection in the anteriorly situated right ventricle with less than normal trabeculation, filling a good sized pulmonary artery although there was a marked sub valvar stenosis and perhaps valvar stenosis. Through the atrial septal defect the catheter progressed to the posteriorly situated left ventricle where injection filled the aorta in L malposition. Juxtaposition of the atrial appendages was not suspected.

This patient was operated upon on July 7th 1977 with the diagnosis of situs solitus, dextroversion, with concordant connections between atria, ventricle and great arteries, anatomical malposition of the great arteries, ASD, VSD, subvalvar and probable valvar pulmonary stenosis. Extra corporeal circulation. Sarns 5000 heart lung machine. Temptrol oxygenator. Middle sternotomy. Heart not increased in size, just behind the sternum with anterior right ventricle pointing to the right. Both vena cavae in the midline to the right of the right atrium which was difficult to examine and slightly anterior to the left atrium. Left sided juxtaposition of atrial appendages, the right lying superior or higher than the left. Right ventricle occupying most of the anterior surface of the heart and left ventricle to the left and posterior. The pattern of the coronary arteries was abnormal. A right, anterior, coronary artery was dominant giving the left anterior

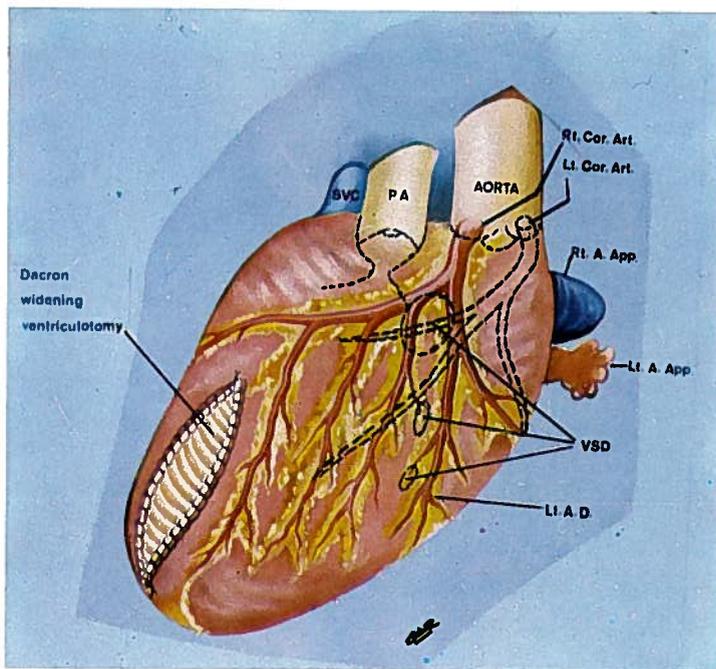


Fig. 5—Heart in dextroversion. Juxtaposition of atrial appendages. L malposition of great arteries. Multiple ventricular septal defects (VSD). Valvar and subvalvar pulmonary stenosis. Abnormal coronary arteries. Dacron widening ventriculotomy

descending and three wide branches to the right ventricle. Only a small area of the right border of the right ventricle, near the apex, was muscular and free of vasculature. The left coronary artery arose from behind the aorta and only had two small branches: circumflex and posterior descending. The inter ventricular septum appeared to be in the sagittal plane. Left sided and anterior aorta following the left border of the heart. Its diameter was 2,5 centimeters. The pulmonary artery arose from the right ventricle, had a diameter of 2 centimeters and was posterior to the aorta. A harsh thrill was felt over the right ventricle just below and over the pulmonary valve (Fig. 5). Intra cardiac pressures: Right atrium 14/7. Left atrium 11/8. Right ventricle 78/0 (37). Left ventricle 83/0 (37).

To establish extra corporeal circulation and after cannulation of the aorta, the SVC was cannulated through the wall of the right atrium. Perfusion was started and, then, it was easy to cannulate the IVC through the posterior wall of the right atrium. Left heart decompression through left atrial appendage. Hypothermia to 26°. Perfusion time 3 h 10 m. Aorta clamped for 2 h 12 m. in 4 periods respectively of 45 min., 18 min., 9 min. and 60 min.. During aortic clamping the heart was locally cooled with Ringer Lactate at 4°C. Right atrial appendage was opened (from the left side) and a large, low atrial septal defect was closed with a running suture of Prolene 4/0. Ventriculotomy in the only, already described, muscular area of the right ventricle near and paralell to the right border just above the apex. Normal tricuspid valve obstructing

the view of a very large (2 cms in diameter) ventricular septal defect, related to the aorta. Subvalvar pulmonary stenosis, muscular and membranous, very much like a subvalvar, membranous, aortic stenosis. This was resected preserving the muscle in its medial aspect and resecting as much as possible in the outer border. Then, the stenosed pulmonary valve was inverted and a commissurotomy was easily carried out. The VSD was patched with interrupted stitches taking care not to injure the bundle which should be in the normal situation. The inner surface of the right ventricle was less trabeculated than expected but definitely not smooth. Closure of the ventriculotomy was carried out enlarging the gap with a generous patch of arterial Dacron to avoid resecting too much muscle in the outflow tract. Before the suture of the patch was completed the left atrial vent was clamped and a substantial amount of bright red blood had to be aspirated. Reexamination of the inter ventricular septum demonstrated that there was no leak through the patched VSD but that there were two other defects, muscular, lower down in the septum. These were closed with mattress stitches over strips of Dacron. Spontaneous excellent heart contractions at 33°C and perfusion stopped shortly afterwards, at 36°C.

Pressures after operation: RV 55 mm Hg systolic; LV 90 mm Hg systolic.

Pacemaker wires at the end of the operation. Uneventful recovery.

## COMMENT

The main interest of this case is the peculiar and rare anatomy with several associated defects. To us it was a definite challenge to establish a proper classification for this complex congenital cardiac malformation. Firstly, juxtaposition of the atrial appendages is a rare condition mentioned in a number of papers. The name of juxtaposition was proposed by Dixon in 1954. Since then more details of this malformation have been added on by Melhuish and Van Praagh (1968), Becker and Becker (1970), Charuzi et al (1973), Deutsch et al (1974) and more recently by Urban et al (1976). The existence of juxtaposed atrial appendages is a sign of complex congenital heart disease and its anatomy can be, essentially, of three forms, namely right sided, left sided and with bifid right atrial appendage (Charuzi et al 1973).

Juxtaposition of the atrial appendages, although rare (97 cases described up to 1976) are much more common on the left as compared to the right in a proportion of 6: 1.

Juxtaposition of the atrial appendages has not been a factor of more difficulty in performing corrective surgery.

Amongst the congenital heart malformations, transposition of the great arteries seems to be the most frequent in which juxtaposition of the atrial appendages is observed.

Isolated dextrocardia has been described by Charuzi et al (1978) in 4 of their 16 cases of juxtaposition.

According to the nomenclature mentioned by Anselmi et al (1972) dextroversion instead of dextrocardia should be preferred as dextrocardia is the normal position of the heart in situs inversus.

Our case, however, was much more complex as, besides dextroversion and juxtaposed left sided atrial appendages, there were other associated defects, namely membranous type ventricular septal defect, muscular ventricular septal defects, valvar and subvalvar pulmonary stenosis and very abnormal anatomy of the coronary arteries.

We believe that the correct nomenclature for this case is dextroversion in situs solitus with concordant atrio ventricular and ventriculo arterial connections, anatomical

L malposition of the great arteries, juxtaposition of the atrial appendages, atrial septal defect, multiple ventricular septal defects, valvar and subvalvar pulmonary stenosis and abnormal distribution of the coronary arteries. For surgical correction, the work of Van Praagh et al. and of Anderson et al., was essential as proper classification and knowledge of connections between heart chambers and great arteries gave us the information required to avoid AV block.

Successful corrective surgery of such a rare association of several cardiac defects made us believe it was worth reporting this case.

### RESUMO

Cardiopatias congénitas cianosantes com várias malformações associadas observam-se com frequência e, em muitos casos, dependendo do grau de anoxia e do estado da circulação pulmonar são compatíveis com uma vida relativamente longa. O diagnóstico pré-operatório e a perfeita classificação anatómica são essenciais para que se possa fazer um tratamento cirúrgico completo e correcto como se mostra num caso de doença cardíaca cianosante com situs solitus, dextroversão, juxtaposição dos apêndices auriculares, conexões atrio-ventricular e ventrículo-arterial concordantes, comunicação inter auricular, comunicações inter ventriculares múltiplas, estenose pulmonar valvular e infundibular e malposição L das grandes artérias.

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