Double Outlet Right Ventricle With Laevomalposition Of Aorta

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Summary:

A rare congenital anomaly - Double Outlet Right Ventricle DORV (SDL) with laevomalposition of aorta, is reported with its clinical, echocardiographic and angiocardiographic characteristics.

Introduction:

Double outlet right ventricle (DORV) is an uncommon congenital anomaly having several subtypes[1]. One rare subtype described is its association with laevomalposition of the aorta. We describe one such case encountered.

Case Report:

A 18-year old boy of average built reported with dyspnoea and palpitation of NYHA II since childhood. His infancy had been unremarkable and there was no history of squatting, anoxic spell or syncope. Examination revealed symmetrical central cyanosis and clubbing, normal vitals with a normal JVP. The precordial examination revealed apex beat in left fifth intercostal space, medial to mid clavicular line, normal in force, character and duration, grade I/III left parasternal pulsations extending upto left second intercostal space; normal S_1, single S_2 and grade III/VI ESM at base. EKG showed NSR with rate of 90/min., axis +270°, clockwise loop in frontal plane and rS pattern in precordial leads from V_1-V_6. X-ray chest revealed no cardiomegaly and there was normal pulmonary vasculature and cardiac contour. 2DE and doppler studies showed visceral situs in solitus, atrioventricular concordance, bilateral coni with aorta anterior and left and pulmonary artery posterior and right, large subaortic VSD; MPA, RPA, LPA and pulmonary annulus normal and pulmonary velocity of 3 ms^{-1}. On cardiac catheterisation the right heart catheter passed up from femoral vein through IVC to RA and RV but could not be negotiated into PA. Left heart catheter passed up the femoral artery through aorta to LV and RV. The pressures and oxymetry of different chambers were as follows:

Chamber	SVC	RA	RV	LA	LV	AO	PA
Pressure (mm Hg)	11/12/10	11/12/10	98/2-6	7/25/8	98/0-8	98/70	-
Oxymetry (%)	46	52	54	100	79	66	-

Angiography of RV and LV were done (Fig. 1 and 2). It showed both aorta and PA filling simultaneously from RV injection and aortic and pulmonary valves at same level with bilateral coni. Aortic valve was anterior and to left of pulmonary artery and pulmonary valve was doming with infundibular stenosis and a large subaortic VSD of about 9 mm in size.

A diagnosis of DORV (SDL) laevomalposed aorta with subaortic VSD and pulmonary stenosis was made. Patient was given an option for surgical intervention, but refused.

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Discussion:

DORV (SDL) laevomalposition of the aorta is a rare variety of DORV, thought to result due to the ventricular loop folding normally to the right (d-loop), but the conotruncus twisting to the left (L-malposition). Both subaortic and subpulmonary coni persist and the subpulmonary conus remains poorly expanded resulting in infundibular stenosis. This type of malformation was first described by Birmingham (1893)[2] and since then many case reports have appeared in subsequent literature. In their series of DORV, Neufeld et al.[3], (8 cases), Vanables and Compbell[4], (16 cases), Lev et al.[5], (133 cases), Zamora et al.[6], (33 cases) and Cameron et al.[7], (27 cases) found no case of laevomalposition of aorta. Sridaromont et al.[8], (70 cases), Wilkinson et al.[9], (84 cases) and Wilcox et al.[10], (63 cases) found its incidence to range from 5-7% of all DORV patients. Van Praagh et al.[11] first published a series of DORV (SDL) in situs solitus and reported overall 23 cases from 6 centres.

Clinically this anomaly resembles tetrology of Fallot. However, inconsistent features are pulsations and palpable S_2 in the pulmonary area, absence of coeur en sabot contour and straight or

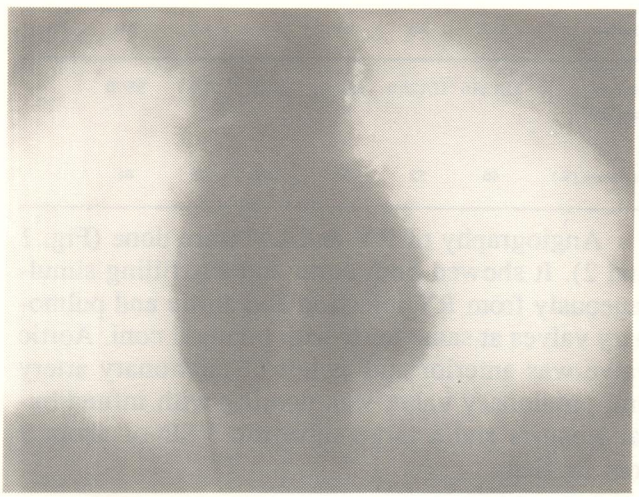

Figure 1.
RV angiography in PA view showing large VSD, dye filling in LV and simultaneous opacification of aorta and pulmonary artery.

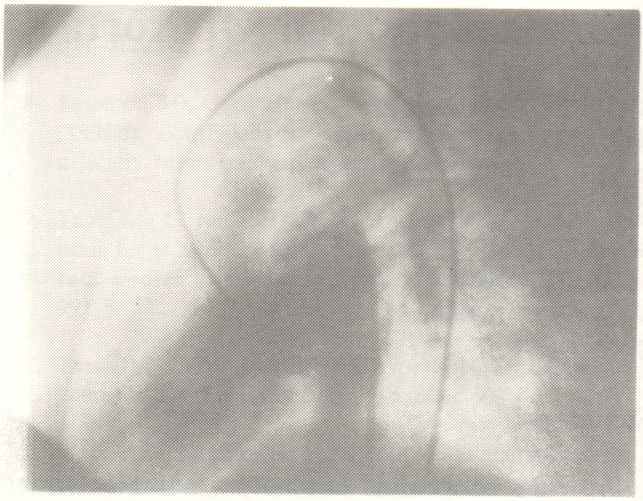

Figure 2.
LV angiography in left lateral view with pigtail cathetor in LV (posterior ventricle) and venous cathetor in RV (anterior ventricle). Dye is seen filling both ventricles, aorta and pulmonary artery. Bilateral coni with both semilunar valves can be seen.

even convex left heart border on frontal chest roentgenogram. 2DE is diagnostic and establishes the abnormal leftward position of aorta.

Surgical correction of this anomaly is readily possible by connecting the aorta to left ventricle by way of subaortic VSD utilising a tunnel prosthesis to reconstruct the left ventricular outflow[12,13]. Difficulties encountered may be with the normal right coronary artery that runs across the right ventricular outflow tract precluding a vertical incision. Since there are only case reports of surgery in such situations, long term results of such procedure are not known.

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