CASE REPORTS

Double Outlet Right Ventricle with Pulmonary Stenosis

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Although transposition of the great vessels is a well-known clinical and pathological entity, the partial transposition complex, resulting in the origin of both the aorta and the pulmonary trunk from the right ventricle, is rare. The association of this malformation with pulmonary stenosis has been described previously (Braun et al., 1952; Witham, 1957; Neufeld, DuShane, and Edwards, 1961; Morgan et al., 1962; Levy et al., 1962 and others). In view of the importance of this malformation in the differential diagnosis of Fallot's tetralogy the following case is reported.

Case Report

A boy, aged 5 years when first seen in 1953, presented with a history of frequent squatting and persistent cyanosis. The physical findings were central cyanosis, clubbing of the fingers and toes, a systolic thrill and murmur over the pulmonary area, and a single second heart sound. The heart was not enlarged clinically. The chest radiograph showed right ventricular enlargement and pulmonary oligemia. The electrocardiogram showed right ventricular hypertrophy. An intravenous angiocardiogram showed simultaneous filling of the aorta and pulmonary trunk and evidence of pulmonary stenosis. Even though a ventricular septal defect was not clearly visualized a diagnosis of Fallot's tetralogy was made. In view of worsening of his symptoms a Blalock's anastomosis was performed, with considerable improvement of his breathlessness and cyanosis.

Ten years after the operation symptoms began again, and within a year the patient was severely handicapped by breathlessness. He was readmitted to hospital in 1964 for reassessment with a view to total correction.

On admission he was dyspneic and deeply cyanosed with marked clubbing. The pulse rate was 100 a minute and the blood pressure 110/70 mm. Hg. The jugular venous pressure was raised to 4 cm. above the sternal angle with a distinct "a" wave. The apex beat was in the sixth left intercostal space and was very forceful and localized. There was a right ventricular heave. There was no thrill. A grade 2/4 pansystolic murmur was heard in the third left intercostal space and under the clavicle a continuous murmur was audible. The second heart sound was single. A loud atrial gallop was heard in the mitral area.

The chest radiograph (Fig. 1) showed a cardiothoracic ratio of 57 per cent with right and left ventricular enlargement. The aortic shadow was not prominent and the main pulmonary artery was small. The lungs were underfilled. The electrocardiogram (Fig. 2) showed sinus rhythm. The mean QRS axis was +135°; there was a tall peaked P wave in lead II and a biphasic P in V1, suggesting both right and left atrial hypertrophy. A tall R wave was present in V1 and very deep S waves in V2, V3, and V4. The T waves over V5 and V6 were deeply inverted. The findings suggested biventricular hypertrophy. A phonocardiogram confirmed the clinical auscultatory findings. The fourth heart sound was clearly shown over the mitral area in the medium frequency tracing.

Although the patient had previously been diagnosed as having Fallot's tetralogy, these findings made the diagnosis untenable. The most formidable objection was provided by the electrocardiographic evidence of left ventricular hypertrophy, which was conspicuous, and the occurrence of a left atrial gallop suggesting additional left-sided disease.

An angiocardiogram was planned, but on the day of the proposed investigation the patient suddenly collapsed and died, in spite of attempts at resuscitation.

At necropsy the heart was very large, weighing 573 g. Both ventricles were greatly hypertrophied. The aorta and pulmonary trunk were normally related externally, but further examination showed that the aorta originated entirely from the right ventricle lying to the right of a normally situated pulmonary trunk and separated from the atrio-ventricular valvular tissue by a horizontal limb of the crista supraventricularis (Fig. 3 A, B). The aortic and pulmonary valves lay approximately in the...
same coronal and horizontal planes. The aortic valve and aorta were otherwise normal. The outflow tracts of the great vessels were separated by a prominent vertical limb of the crista supraventricularis. There was a tight infundibular pulmonary stenosis measuring 7 mm. in diameter at the narrowest point. Apart from moderate hypoplasia, the pulmonary valve and trunk were normal. The only outlet from the left ventricle was a small calcified ventricular septal defect, measuring 4 mm. in diameter, which opened into the right ventricle below the horizontal limb of the crista supraventricularis so that the direction of blood-stream from the left ventricle appeared to proceed predominantly to the aorta. The right and left atria were hypertrophied and the foramen ovale was patent to a probe 2 mm. in diameter. The mitral and tricuspid valves were normal and the coronary arteries were normally distributed. The Blalock's anastomosis was patent, 4 mm. in diameter.

The remainder of the necropsy findings were unremarkable apart from vascular congestion of the liver and spleen.

**Discussion**

Witham (1957) divided cases of double outlet right ventricle into two groups according to whether or not there was associated pulmonary stenosis. Those without pulmonary stenosis were called of the “Eisenmenger type”, and those with pulmonary stenosis were called of the “Fallot type”.

The differentiation of these patients with Fallot type double outlet right ventricle from Fallot’s tetralogy is important, because of the different surgical approaches required for correction of these malformations. Closure of the ventricular septal defect, part of the operation of total correction in Fallot’s tetralogy, would be disastrous in double outlet right ventricle. Levy et al. (1962) report
successful correction of a case of double outlet right ventricle with pulmonary stenosis.

The diagnosis of Fallot’s tetralogy in this patient became suspect at his second admission because of the left atrial gallop and electrocardiographic evidence of left ventricular hypertrophy. Mirowski, Mehrizi, and Taussig (1963) have reviewed the electrocardiographic findings in double outlet right ventricle with stenosis. The small size of the ventricular septal defect and the previous Blalock’s anastomosis in this patient caused more marked electrocardiographic changes of left ventricular hypertrophy than in any of the patients described in that paper.

A review of the 1953 intravenous angiocardiogram on this patient showed that there was considerable superimposition of the atrial and ventricular shadows, and the origins of both great vessels were obscured. However, with biplane selective right ventricular angiocardiography this problem does not arise; both great vessels could be shown to arise from the right ventricle and the anterior position of the aorta could be appreciated.

Summary

A patient with double outlet right ventricle with pulmonary stenosis is reported. Clinical, electrocardiographic, and necropsy findings are described. The diagnosis is discussed.

References


Double outlet right ventricle with pulmonary stenosis.

A V Shetty and R Martin

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