Case reports

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Spontaneous closure of ventricular septal defect in a case of double outlet right ventricle

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SUMMARY A 5 year old child, previously diagnosed as having tetralogy of Fallot, was admitted to hospital in severe congestive heart failure. The electrocardiogram showed left anterior hemiblock and incomplete right bundle-branch block, neither of which was previously present. The child died in intractable congestive heart failure and the necropsy showed a double outlet right ventricle with complete spontaneous closure of the subaortic ventricular septal defect by fibrous tissue. The possible mechanism involved in the production of this unusual complication of double outlet right ventricle is discussed, together with an explanation for the electrocardiographic changes.

It is well known that an isolated ventricular septal defect can reduce in size spontaneously, and even close completely.¹⁻³ Occasionally, this development has been described in patients with complex congenital heart disease, in whom the ventricular septal defect was physiologically advantageous such as the "classic" double outlet right ventricle, that is with bilateral conus,⁴⁻⁶ or tricuspid atresia.^{7,8} Conversely, complete spontaneous closure of a ventricular septal defect has never been described, to our knowledge, in patients with tetralogy of Fallot or with double outlet right ventricle and infundibular features of Fallot's tetralogy.

We report the clinical and anatomical findings of a case of double outlet right ventricle with some infundibular features of Fallot's tetralogy, in which there was spontaneous closure of the ventricular septal defect.

Case report

A 5 year old girl was born with a cyanotic cardiac defect. She was admitted to our hospital in congestive heart failure.

A heart murmur had been heard since birth. Cyanosis appeared when she was 5 months old. At 3 years of age she underwent a right heart catheterisation in another institution which disclosed tetralogy of Fallot. The electrocardiogram showed right ventricular hypertrophy including QRS right axis deviation (Fig. 1A). No surgical procedure was performed at that time. On admission to our unit, neither the

catheterisation data nor the angiogram were available.

Physical examination showed a very ill child with blood pressure of 90/40 mmHg, pulse rate of 130 beats/min, and respiratory rate of 35/min. She weighed 14 kg and was 105 cm tall. Mild cyanosis and dyspnoea were noted. The neck veins were distended. There was oedema at the ankles, hepatomegaly, ascites, and bilateral pleural effusions. The precordium was hyperactive. The first heart sound was normal and the second sound was single. A 3/6 ejection systolic murmur and a third heart sound were heard in the third left intercostal space on the sternal border.

The chest x-ray film showed cardiac enlargement, pulmonary venous congestion, bilateral pleural effusions, and a left aortic arch.

The electrocardiogram recorded on admission (Fig. 1B) showed normal sinus rhythm, PR interval 0.18 s, left atrial enlargement, QRS width 0.08 s, and QRS mean frontal axis -60° . The QRS pattern indicated left anterior hemiblock; QRS morphology was QrS, QS, and rS in V1, V2, and V6, respectively, indicating a posterior, rightward, and counterclockwise inscription of the QRS forces in the transverse plane.

Another electrocardiogram was recorded four days later (Fig. 1C) and disclosed an incomplete right ventricular conduction defect (qR morphology in V1 with tall R wave and mild increase in QRS width). A more pronounced superior deviation of the QRS axis was also evident.

The M-mode echocardiogram disclosed the presence of two enlarged ventricles with two atrioventricular

Fig. 1 (A) Electrocardiogram recorded at age of 3 years, showing right ventricular hypertrophy and QRS right axis deviation; (B) electrocardiogram recorded on admission to hospital, showing left atrial enlargement and pronounced QRS left axis deviation, suggesting left anterior hemiblock; (C) electrocardiogram recorded four days later, showing an incomplete right ventricular conduction defect and more pronounced QRS axis superior deviation.

lar and semilunar valves. Aortic overriding was present.

Because of the poor clinical state of the child, heart catheterisation was not performed. On the fifth day after admission she died because of pulmonary oedema.

ANATOMICAL FINDINGS

The heart was in situs solitus of the atria, with laevocardia and normal systemic and pulmonary venous drainages. The atrioventricular connection was concordant and the atrioventricular valves were normal. The left atrioventricular valve was between the left atrium and left ventricle and it had no communication with the right ventricle. The two ventricles were enlarged. Hypertrophy of both left and right ventricular walls was present. Mural thrombi were found in the left ventricular cavity.

The type of ventriculoarterial connection was a double outlet right ventricle, the aorta originating entirely from the right ventricle with a muscular rim separating the tricuspid and aortic valves (Fig. 2B).

The right ventricular outflow tract showed an anterior and cephalad deviation of the infundibular septum. The malalignment of the infundibular septum with the rest of the muscular septum determined

an infundibular pulmonary stenosis, as observed in Fallot's tetralogy.

The left ventricular outflow tract (Fig. 2B) was atretic because of the presence of accessory endocardial fibrous tissue. The mitral valve was separated from the aortic valve by endocardial fibrous tissue without any muscular component.

There was no ventricular septal defect; instead the ventricular septum was intact because the accessory fibrous tissue extended from the posterior and left aortic cusps to the muscular septum (Fig. 2A).

The atrial septum showed a large patent foramen ovale and an aneurysm from the left to the right of the Vieusseux valve.

On macroscopical examination signs of acute pulmonary oedema were evident; histological features of chronic venous hypertension were observed.

Discussion

Our case was a double outlet right ventricle *de facto* with some infundibular features of tetralogy of Fallot, such as malalignment of the infundibular septum with the rest of the muscular septum and mitroaortic fibrous continuity. According to Anderson *et al.*,⁹ double outlet right ventricle as a type of ventriculoar-

Fig. 2 (A) Posterior outlet of the right ventricle (RV): the aorta (A) arises entirely from the right ventricle. A muscular rim (*) separates tricuspid (T) and aortic (A) valves. A fibrous membrane (arrow) is attached superiorly to the posterior and left aortic cusps and inferiorly to the muscular septum, probably obstructing a previous ventricular septal defect. (B) Left ventricular (LV) view; the LV outflow tract, encircled by symbols (*), is atretic because of the presence of endocardial fibrous tissue separating the aortic and mitral (M) valves.

terial connection can coexist with the infundibular morphology of Fallot's tetralogy.

In our case ventricular septal defect was absent, accessory fibrous tissue extending from the aortic cusps to the muscular septum. Direct evidence of the previous existence of ventricular septal defect in this patient was not available at the time of our observation. Circumstantial evidence, however, of a previous ventricular septal defect was the left ventricular dilatation and hypertrophy, since the left ventricle would be hypoplastic if it had not previously had an outlet. Furthermore, left ventricular wall hypertrophy might be produced by progressive left ventricular outflow obstruction, that is by progressive obstruction of a communication between the two ventricles.

The reduction in size of physiologically advantageous ventricular septal defect has been described in several cases of double outlet right ventricle with bilateral conus.⁴⁻⁶ In such cases, a muscular subaortic conus, separating mitral and aortic valves and closely related to the ventricular septal defect, was considered

to be the anatomical factor leading to the ventricular septal defect⁶ by fibrous and/or muscular tissue.

Spontaneous closure of ventricular septal defect has not been reported in patients with double outlet right ventricle without subaortic conus. This event is exceptional also in tetralogy of Fallot. An obstructive ventricular septal defect is rarely observed in Fallot's tetralogy: anomalous accessory tricuspid tissue^{10,11} or anomalous attachment of the mitral valve to the ventricular septum, as in endocardial cushion defects,¹² have been described as causes of ventricular septal defect obstruction in tetralogy of Fallot.

In our case, the closure of the ventricular septal defect was not caused by abnormalities of the atrioventricular valves. We suggest that the closure had been brought about by progressive growth of the accessory fibrous tissue extending from the aortic cusps to the muscular septum. In our opinion, the extreme dextroposition of the aorta and the presence of a muscular rim separating the tricuspid and aortic valves were predisposing factors to the closure of the

left side of the bulboventricular foramen in this patient.

The evolution of the electrocardiogram in this patient is interesting: that recorded at the age of 3 years suggested right ventricular enlargement. During our observation, the electrocardiogram evolved from a left anterior hemiblock to an incomplete right ventricular conduction defect, associated with a more pronounced superior QRS axis orientation. It is well known that spontaneous closure or reduction in size of an isolated ventricular septal defect may affect the conducting system, with eventual development of left anterior hemiblock.¹³ We suggest that the evolution of the electrocardiogram in our case could be the result of widespread progressive fibrosis in the area of the inferior rim of the ventricular septal defect, causing early electrical delay in anterolateral ramifications of the left bundle-branch and subsequently in the right bundle-branch. Late appearance of a right ventricular conduction defect associated with a higher degree of block in the left anterolateral ramifications seems to validate this opinion.

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Notice

British Cardiac Society

The Autumn Meeting will be held at Wembley on 21 and 22 November 1983, and the closing date for abstracts will be 26 July 1983.

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The Autumn Meeting 1984 will be held on 3 and 4 December 1984, and the closing date for receipt of abstracts will be 15 August 1984.