Case Reports

Origin of Both Great Vessels from the Right Ventricle

Report of a Case Without Pulmonary Stenosis

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A rare, however, surgically important condition in which both great vessels arise from the right ventricle has been recognized for many years. Witham,¹⁾ applying the term of "double outlet right ventricle" to the anomaly, divided it into two groups clinically at the first time in 1957: the "Eisenmenger type" without pulmonary stenosis and the "Fallot type" with pulmonary stenosis. The term "origin of both great vessels from the right ventricle" has also been used to define the entity. Since then McGoon²⁾ reported the first case of successful repair in 1961 and was followed by the case of Redo et al.³⁾ which was diagnosed preoperatively and operated upon in May of 1961.

The development of cardiology and cardiac surgery, and an increasing experiences in cases of tetralogy of Fallot and advanced ventricular septal defect will give a chance to meet the anomaly, even though their differentiation will be occasionally quite difficult. And from surgical point of view, the recognition of this disease is very important to have it repair correctly even in surgery.

This is an additional case which was clinically very similar to ventricular septal defect with moderate pulmonary hypertension and was found to be the anomaly at surgery.

CASE REPORT

A 5-year-old Japanese boy was first seen at the Outpatient Clinic of our Department in July, 1966, because of heart murmur and repeated infections of respiratory tract. The patient was the product of a normal pregnancy and delivery. He had been as usual until the first 2 months of life when he suffered from bronchitis and was found to have heart murmur accidentally. Since then he had had frequent

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upper respiratory infections including 2 episodes of pneumonia. At the age of 2 he was told to have pulmonary tuberculosis, however, repeated bacteriological examinations of sputum were negative for A.F.B. Respiratory infection continued intermittently until the age of 4. Then he improved gradually and had been symptomfree for several months before admission.

Physical examination on admission revealed a well nourished and alert, but somewhat small boy for his age without cyanosis and clubbing. Pulse rate was 86 and regular, respiration 26. The blood pressure was 80 systolic and 60 diastolic mm. Hg in both arms. RBC was 483×10^4 , Hb 12.2 Gm./100 ml. and Ht 37.7%. The shape of the chest showed chicken breast, however, it was clear to percussion and auscultation. The heart was slightly enlarged to both sides and P.M.I. was felt in the fifth left intercostal space just lateral to the mid-clavicular line. A systolic thrill was palpable over the third and fourth intercostal space to the left of the sternal border. A grade 5, harsh, holosystolic murmur was heard over the whole precordium with the point of maximum in the fourth intercostal space along the left sternal border. A grade 2 diastolic murmur was audible at the apex. The second sound was slightly accentuated over the pulmonic area. The P-A roentgenogram showed a slightly enlarged transverse diameter of the heart with a cardiothoracic ratio of 0.58. The left lower cardiac contour might be slightly elongated. The right cardiac shadow also extended a little far to the right of the sternal border. Hilar and pulmonary vascular markings were more prominent than normal (Fig. 1 A and B). Electrocardiogram was suggestive of biventricular enlargement: frontal QRS

Fig. 1. A. P-A roentgenogram showing cardiomegaly, fairly wide supracardiac shadow and increased pulmonary vascular markings. B. Lateral projection.

axis $+70^\circ$, rather tall R wave and deep S wave in V_1 , and tall R wave in $V_{5,6}$ (Fig. 2). These findings were indicative of much possibility of ventricular septal defect with moderate pulmonary hypertension. No further examinations were performed.

Operation was done on October 25, 1966, with cardiopulmonary bypass. The heart and great vessels were exposed through a median-sternotomy. The aorta was noted lying rather anteriorly, almost parallel to the pulmonary artery. The right

Fig. 2. Electrocardiogram disclosing frontal QRS axis $+70^\circ$, rather tall R wave and deep S wave in V_1 , and tall R in $V_{5,6}$.

ventricle looked rather roundish than usual in contour. A systolic thrill was palpable over the outflow tract of the right ventricle and pulmonary artery. During preparation for bypass encircling the tapes around the superior vena cava, aorta and pulmonary artery, accidentally ventricular fibrillation took place. At that time body temperature was 39.5°C . The heart beats returned after electric counter-shock on 2 occasions and cardiopulmonary bypass was quickly instituted. Following reduction of temperature per rectum to 31°C , right ventriculotomy was placed. Right ventricle was moderately hypertrophied and ventricular septal defect, 1.5 cm.^2 in size, was found. Besides, the aortic valve was far displaced to the right, taking off from the right ventricle in the same plane to the pulmonary artery. The leaflets of the aortic and pulmonic valves were normal. No stenosis was demonstrable in the infundibulum and valvular areas of the pulmonary artery (Fig. 3). In the begin-

Fig. 3. Drawing shows the position of the aortic and pulmonic valves, and ventricular septal defect in relation to the crista supraventricularis in our case.

ning of the repair, ventricular septal defect was enlarged to the size of 2.6 cm.² by excising its anterior muscular portion, and then the doubled pericardial patch was secured with 3-0 black silk interrupted sutures commencing from the posteroinferior margin of the septal defect to the annulus of the aortic valves superiorly and down to between the roots of both great vessels, as well as to the lower edge of the ventricular septal defect. To avoid the damage to the leaflets of the aortic valve, sutures were placed rather to the pulmonic valve leaflets (Fig. 4 A and B). After completion of the procedure, the left ventricular pressure was 80/0, and pressures in the right ventricle and pulmonary artery were 70/8 and 28/8 mm. Hg respectively. Systemic pressure at this time was 95 mm. Hg (Fig. 5).

Fig. 4. A. Photograph is just before the construction of a conduit. Interrupted mattress sutures with white pledgets around VSD and the aortic valves. B. After the construction of the conduit. A whitish pericardial patch protrudes to the right ventricle.

Fig. 5. Drawing of operative method done in our case.

Total bypass time was 1 hour and 25 min., during which time periodic anoxic arrest was applied on 7 occasions. The patient tolerated the operation and was uneventful postoperatively.

On postoperative examination of the heart, a systolic thrill was palpable in the third intercostal space along the left sternal border with a grade 5 systolic murmur in the same place. In postoperative angiocardiology as shown in Fig. 6 A and B, the aortic valve lying higher than normal was demonstrable at the same level as the pulmonic valve and the dilatation of the pulmonary artery. Simultaneous measurement of intracardiac pressures disclosed 80/0 mm. Hg in the right ventricle. No shunt was proved in dye dilution curve.

Fig. 6. Postoperative selective angiocardiology. The level of the aortic and pulmonic valves is noticeable in the same plane. A. The aortic valve. B. The pulmonic valve.

DISCUSSION

From phylogenetical and embryological point of view, Spitzer,⁴⁾ in 1923, assumed the development of the malformation in association with "pathologische Detorsion" and "rechtskammerige Aorta" based on some reptilian embryos. Shaner^{5), 6)} showed in his interesting investigation in pig embryos that the conus and truncus arteriosus consist of a simple tube, taking its origin entirely from the right ventricle, and in a later stage the truncus arteriosus becomes divided into the aorta and pulmonary artery, migrating the vessel orifices to the left. However, the migration of the aorta will be completely arrested in the case, in which complete disappearance of the conoventricular ridge and the fusion of the ventral cushion to the dorsal are disturbed, resulting in overriding or transposition of the aortic orifice to the right ventricle. This conception is supported by Doerr⁷⁾ in man.

Neufeld et al.^{8),10)} pointed out the common features in the anomaly: a normal relationship between the ascending aorta and the pulmonary artery from the exterior, a communication of the aortic orifice only with the right ventricle and its location to the right of the pulmonary artery, and a level of the aortic and pulmonic valves in the same coronal plane. Besides, the presence or absence of significant pulmonary stenosis described by Witham¹¹⁾ and the continuity between the aortic and mitral valvular tissue revealed by Neufeld et al.¹⁰⁾ are necessary to classify cases of this anomaly. According to Van Mierop and Wiglesworth⁹⁾ the type with continuity between the aortic and mitral valves is considered as merely an extreme form of the Eisenmenger complex, or of the tetralogy of Fallot if significant pulmonary stenosis present, however, that without their continuity because of persistent conoventricular flange differs from the former anatomically. Most cases of the malformation have been included in the latter.

To correlate the anatomic findings with the clinical and hemodynamic data, Neufeld et al.¹⁰⁾ divided the anomaly without pulmonary stenosis into Type I and II, and Type II into two subgroups, A and B, depending on the position of ventricular septal defect to the crista supraventricularis, and to the aortic and pulmonic valves. Sakakibara et al.¹¹⁾ reviewing their own 10 cases, classified the anomaly into retro-cristal and antero-cristal type, and subdivided them into retro-supra-cristal, retro-cristal intermediate and retro-infracristal, and antero-supracristal, antero-cristal intermediate and antero-infracristal type respectively with almost the same intention as Neufeld did. In Type I, in which ventricular septal defect is below the crista supraventricularis, as well as retro-cristal type of Sakakibara et al., blood from the left ventricle easily ejects into the aorta unless significant pulmonary stenosis or pulmonary vascular disease is present and venous blood from the right ventricle into the pulmonary artery simultaneously, and arterial saturation is often relatively within normal. On the other hand, Type II A and B in which septal defect is subpulmonary or overriding between the pulmonary artery and aortic orifice above the crista supraventricularis, and antero-cristal type of Sakakibara et al. can have more anatomical predilection to send unsaturated blood into the aorta producing lower oxygen saturation of peripheral arterial blood. In the case complicated by pulmonary stenosis hemodynamic situation can be much influenced by the constriction of the pulmonary artery.

The diagnosis can not be made from the clinical history and physical examination alone. In our case, the patient was found to have heart murmur 2 months after birth when he suffered from respiratory infection, probably secondary to pulmonary congestion. In other reports, most cases without pulmonary stenosis have almost similar trend. The murmur suggested

the presence of ventricular septal defect with pulmonary hypertension, and its origins are likely due to an obstruction to outflow from the left ventricle provided by the septal defect and to an excessive pulmonary flow. Another type with pulmonary stenosis shows an identical pattern of tetralogy of Fallot clinically and physically. And as already stated, the presence of cyanosis is dependent on the anatomic relationship of the septal defect to the crista supraventricularis and to the aortic and pulmonic valves, and on complication by severe pulmonary stenosis or pulmonary vascular obstruction.

Roentgenographic findings of our case were characteristic to those of ventricular septal defect with pulmonary hypertension except for fairly wide cardiac base shadow in the P-A view which will often give a clue to distinguish the malformation from advanced ventricular septal defect as reported by Witham.¹⁾ Most of cases with pulmonary stenosis show the same roentgenographic configuration as those of tetralogy of Fallot, although biventricular hypertrophy of Witham's case and left ventricular hypertrophy of Taussig's case¹²⁾ were noticeable in the literature.

Electrocardiogram and data from cardiac catheterization will be more suggestive of the diagnosis. Neufeld et al.⁸⁾ described an electrocardiographic pattern similar to that of endocardial cushion defect in the case without pulmonary stenosis, and later related it to a pathoanatomic condition classified as Type I, in which ventricular septal defect lay below the crista supraventricularis. However, right ventricular hypertrophy with left axis deviation concomitant with counterclockwise direction of the QRS vector loop in the frontal plane is characteristic not only to endocardial cushion defect but also to some of ventricular septal defect. The findings accordingly do not disclose constant pattern but do not negate the value as a diagnostic clue in the case with a clinical impression of ventricular septal defect with pulmonary hypertension. On the other hand, Mirowski¹³⁾ reported left ventricular hypertrophy, complete right bundle branch block and first degree heart block in 75, 63 and 60 per cent of his series, respectively, as points to distinguish these patients accompanied by pulmonary stenosis from these with tetralogy of Fallot in the cardiogram. Most cases in the literature^{1), 9), 14), 15)} have no specific electrocardiographic findings to define the anomaly, however, similarly the case with clinical features of tetralogy of Fallot or ventricular septal defect with pulmonary stenosis and an electrocardiographic pattern described by Mirowski is much suggestive of the diagnosis of the lesion. The findings at cardiac catheterization rely on the presence or absence of pulmonary stenosis, or pathoanatomic relationships inside the heart as Type I and II A and B or retro-cristal and anterocristal VSD type, but "patient with ventricular septal defect, and right ventricular pressure near systemic should be suspected of

having aortic transposition" as stated by Engle et al.¹⁶⁾ is the important data in either case. In our case, no intracardiac pressures were recorded because of a severe complication which occurred in the early stage of the operation.

The possibility of the lesion arises in patients with apparent ventricular septal defect plus suggestive electrocardiogram and right ventricular hypertension of systemic level, and in patients who appear to have the tetralogy of Fallot with extreme dextroposition of the aorta, right ventricular hypertension and analogous electrocardiogram of Mirowski's description.

And confirmation of the diagnosis can usually be made with selective or cineangiocardiology. The demonstration of the relationship of the great vessels to the right ventricle, of the level of both semilunar valves in the same coronal and cross-sectioned body planes, and of anterior displacement of the aorta to the plane of the ventricular septum above the right ventricle in the lateral view is necessary with careful simultaneous consideration on dilution effects due to the degree of pulmonary stenosis or the position of the pulmonary artery to ventricular septal defect.

Surgical therapy^{2), 3), 17)–19)} for this lesion is dependent on hemodynamic condition based on pathoanatomic situations. In the case without pulmonary stenosis, inspection of the heart and great vessels is required to see the displacement of the aortic root to the right and its relationship to the pulmonary artery in the same plane on exposure of the heart. After an incision into the right ventricle through right ventricular cardiomy or transatrial route, the position of the aortic valve and its individual leaflets to ventricular septal defect should be explored, especially in any case suspected.

The basic objective of the repair is the construction of a conduit on the floor of the outflow tract of the right ventricle with a prosthesis to eject blood from the left ventricle via ventricular septal defect, separating two currents of blood passing through the outflow tract of the right ventricle en route to the aorta and pulmonary artery. Prior to constituting the prosthesis, the size of ventricular septal defect should be estimated and enlarged to that of the aortic annulus at least in the case with significantly smaller defect than the aortic orifice by excising the septum ventral to the defect as our case did, and cares to be taken for all suturings in the region is to avoid the damage to the bundle of His and the leaflets of the aortic valve near the aortic root. The shape of the prosthesis should be suitable for the requirement to be its anterior, or ventral surface much longer than the dorsal. McGoon²⁾ used somewhat diamond-shaped and Redo et al.³⁾ hammock-like prosthesis. A rectangular piece of the pericardium was utilized in our case. In another case with pulmonary stenosis, a successful repair was reported first by Levy et al.,¹⁶⁾ in 1962, placing Teflon felt patch to exclude the aorta from the right ventricle and

utilized a tear-drop Teflon patch as an outflow roof over outflow tract to the pulmonary artery annulus after excision of muscle and fibrotic endocardium from the infundibular area. Blalock-Taussig or Potts anastomosis as a non-corrective method may be unsatisfactory in this case because of subsequent left ventricular overload and resulting heart failure as pointed out by Taussig.¹²⁾

Postoperatively residual systolic murmur in the third intercostal space along the left sternal border on physical examination, the dilatation of the pulmonary artery in angiocardigraphy and elevated right ventricular pressure are evident in our case. These findings will exhibit the presence of possible pulmonary stenosis and probably be caused by either the protrusion of the pericardial patch into the right ventricle or well-developed crista supraventricularis obstructing the outflow tract during systole. In the description of Kirklin et al.,¹⁸⁾ about 80 mm.Hg of systolic right ventricular pressure was also obtainable in 2 cases of their series which were in good health 22 and 24 months postoperatively.

SUMMARY

A 5-year-old boy who had a clinical impression of having ventricular septal defect with moderate pulmonary hypertension was ascertained to be this lesion at the time of surgery. The operation was performed successfully constructing a conduit with a rectangular piece of the pericardium. Problems on this lesion are discussed with a brief review of literature.

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