Dextrocardia with Pulmonary Stenosis and Functionally Single Right Ventricle

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An unusual case of congenital heart disease is reported which, despite numerous circulatory abnormalities, permitted a life span of 34 years.

This is an unusual case of congenital heart disease in a patient with dextrocardia and situs inversus in which both the aorta and stenosed pulmonary artery originated from the right ventricle. Despite numerous circulatory abnormalities, this patient lived 34 years.

Case Report

A 34-year-old Puerto Rican was admitted to Lincoln Hospital February 10, 1956, complaining of chest pains, vomiting, marked dyspnea, and ankle edema.

According to the mother, the child was normal at birth and developed normally to about the age of 8. Then he began to develop a deformity of his back and to show cyanosis. Dyspnea on exertion, cyanosis, and deformity of the back became progressively worse, but he was not under any continuous medical care.

Examination at the time of admission revealed an acutely ill, markedly dyspneic, and cyanotic man. There was marked deformity of the thoracic cavity due to kyphoscoliosis of the dorsal spine with a convexity to the left. The blood pressure was 100/60. The heart rate was very rapid and irregular. There was a grade IV, rough systolic murmur and a long blowing diastolic murmur at the fourth left interspace, and a grade IV systolic murmur over the second left interspace. Moist rales were heard throughout both lung fields. There was heaving over the entire precordial area and a point of maximum cardiac impulse could not be determined. The thrust to the right of the sternum appeared to be greater than to the left. The liver was enlarged 4 fingers below the left costal margin. There was 3+ pitting edema of the extremities.

Fluoroscopy and x-ray of the heart were confusing because of the kyphoscoliosis. The aorta was to the right of the esophagus and the transverse diameter of the heart was increased. The stomach was in the right upper quadrant (fig. 1).

The electrocardiogram (fig. 2) was most difficult to interpret. Atrial fibrillation with a very rapid ventricular rate and digitalis effect were definite. There was no definite evidence of dextrocardia. The hemoglobin was 15 Gm. per cent and the white count was 6,400, with a normal differential. The urea nitrogen was 28 mg. per cent. Other tests were within normal limits.

The patient was treated with digitalis, mercurials, quinidine, and a salt-free diet, and the decompensation improved. However, on April 14, he complained of pains in the chest and marked dyspnea, went into shock, and died.

Autopsy Findings

At autopsy, the heart was predominantly in the right chest (fig. 3). The ascending aorta was on the left with the descending portion on the right side of the esophagus. There was marked dilatation and hypertrophy of the right* atrium and right ventricle, which were on the left side of the heart. The left atrium and left ventricle were very small and were posterior and on the right side. In the heart, both the aorta and stenosed pulmonary artery arose from the markedly enlarged right ventricle. The aortic valves appeared normal, except for fenestrations of the cusps. The narrowed opening of the pulmonary artery (which was about 0.5 cm. in diameter) was below and to the right of the aortic opening (fig. 4). The pulmonary valve had only 2 cusps. The pulmonary artery was directed posterior to the aorta, and from right to left. There was an interventricular septal defect measuring about 2 cm. (fig. 5) in diameter in the muscular portion of the septum and below the crista supraventricularis. It did not involve the openings of either the pulmonary artery or aorta. The myocardium appeared normal in color and consistency. One coronary artery arose from the aorta and one from the pulmonary artery. The right pulmonary veins emptied into the right atrium. A patent ductus arteriosus was not found. The liver weighed 1250 Gm. There was situs inversus of all the organs, including the lungs, except the cecum and appendix, which were on the right side.

* To avoid misunderstanding, right atrium and right ventricle refer in this article to the atrium and ventricle supplying the pulmonic circulation although they were on the left side of the heart. The left atrium and left ventricle refer to the systemic circulation and were on the right side.
Dextrocardia with Pulmonary Stenosis

Fig. 1 Posteroanterior x-ray of chest showing marked deviation of the esophagus to the left in the upper portion. As the esophagus descends, it deviates to the right to empty into the stomach in the right upper quadrant.

Fig. 2. Electrocardiogram showing atrial fibrillation and digitalis effect.

Discussion

The diagnosis of dextrocardia was clinically difficult to establish for the following reasons. Because of the large right ventricle, the electrocardiogram was not diagnostic of dextrocardia. Since there was atrial fibrillation, the direction of the P waves could not be determined. The marked scoliosis and kyphosis caused marked distortion of the thoracic cavity with rotation of the heart, so that chamber size could not be determined on either fluoroscopy or x-ray examination. Fluoroscopy revealed a right-sided aorta, equal pulsations on both sides of the cardiac shadow in the posteroanterior view, and situs inversus. Physical examination revealed a heaving over the anterior chest, so that a point of maximum cardiac impulse could not be determined. Neither angiocardiology nor cardiac catheterization could be performed.

Although both levocardia in situs inversus and isolated dextrocardia are nearly always associated with congenital cardiac defects, the occurrence of congenital cardiac defects in dextrocardia with situs inversus is more unusual. In Maude Abbott’s 1,000 cases, there were 11 cases of dextrocardia, situs inversus, and cardiac defects. The incidence of a patent right ventricular aorta is rare. Edwards mentioned 8 cases of this abnormality. Also, in Abbott’s series, there were 3 cases of aorta originating from the right ventricle. Neither Edwards’ nor Abbott’s cases showed dextrocardia with situs inversus.

It is difficult to classify this type of abnormality. Although this patient had the 4 charac-
Fig. 4. Top. Arrow no. 1 points to stenosed pulmonary artery. Arrow no. 2 points to opening of aorta.

Fig. 5. Bottom. Shows tricuspid valve and relationship of interventricular septal defect to openings of aorta and stenosed pulmonary artery. Note that there is no overriding of either the aorta or the pulmonary artery.
Dextrocardia with Pulmonary Stenosis

269

teristic defects usually classified as tetralogy of Fallot, namely right-sided aorta, pulmonary stenosis, interventricular septal defect, and right ventricular hypertrophy, there was no overriding of the aorta over the septal defect. Also, there was no overriding of the stenotic pulmonary artery. The ventricular septal defect was below the crista supraventricularis, in the muscular part of the septum, and the course of the pulmonary artery was directed posterior to the aorta from right to left. In Taussig's description of the types of tetralogy, there is always some degree of overriding of the aorta. Even if one does not accept this concept of overriding of the aorta, the location of the defect and the relationship of the pulmonary artery and the aorta are not those found in the tetralogy of Fallot. This heart would fit into the type II classification of Spitzer. According to his theory there is a greater degree of distortion than that found in his type I classification (in which the tetralogy would be placed). As a result, the ascending aorta is to the right (in this case, to the left, because of the dextrocardia) and anterior to the pulmonary artery. Also, Spitzer maintained that the aorta is a right ventricular aortic conus that has reopened and is connected with the common ascending aorta, the left ventricular aortic conus having been obliterated. As a result, there is no direct outflow ostium from the left ventricle. The only means of exit of blood from the left ventricle is through the septal defect into the right ventricle.

Spitzer also stated that the pulmonic valve usually has only 2 cusps and 1 of the coronary arteries is transposed so that it arises from the pulmonary artery. These abnormalities were also present in this case.

The electrocardiogram was most unusual. In spite of the tremendous right ventricle, characteristic right ventricular complexes (R or Rs) were not seen on either the right or left side of the heart. Only in V1, V6R, and V6S were RS complexes seen. The unipolar limb leads did not definitely indicate dextroposition. As mentioned previously, because of the atrial fibrillation, the direction of the P waves was of no resistance. In retrospect, one can surmise right ventricular hypertrophy in the dextrocardia because of the late intrinsicoid deflection of the R wave on the left side of the heart (V2-R) and aVL with progressive formation of an rS complex as one reads from V1 to V6R (from right sternal border to right axilla) and the formation of an R complex in V2R and V3R, suggesting a left ventricular complex in the right posterior chest.

It is obvious that there must have been some fairly competent pulmonary collateral circulation to allow this man to live 34 years. No patent ductus arteriosus was found. Ghoreyeb and Karsner as early as 1913 showed that when the pressure in the pulmonary radicles is markedly reduced the bronchial circulation is brought into service to compensate for the decreased pulmonary flow. The earlier in life that this occurs, the more readily the bronchial arteries enlarge. This pattern of collateral circulation certainly must have occurred in this case, aided undoubtedly by the anterior and posterior mediastinal and esophageal arteries. It is only through development of these collateral vessels that sufficient pulmonary circulation could be maintained, as the markedly stenosed pulmonary valve certainly could not permit sufficient blood to get to the lungs.

Summary

An unusual case is presented of a 34-year-old man with dextrocardia and situs inversus in which the aorta and stenosed pulmonary artery arose from a markedly hypertrophied and dilated right ventricle. There were numerous other congenital abnormalities, namely, interventricular septal defect, 1 coronary artery arising from the pulmonary artery, bicuspid pulmonary valve, the right pulmonary vein emptying into the right atrium, and fenestration of the cusps of the aortic valve. This combination of congenital cardiac defects best fits in with Spitzer's type II classification of congenital malformations of the heart. There must have been a well-developed collateral pulmonary circulation to allow this man to live 34 years.

Summario in Interlingua

Es presente e caso inusual de un homine de 34 annos de etate, con dextrocardia e sito inverse in que tanto le aorta como etiam le stenotic arteria pulmonar partiva ab un marca-
FISHER AND SUH

temente hypertrophic e dilatate ventriculo dextere. Le caso se distinguева per numerose altere anormalitates congenite, i.e. defecto de septo interventricular, 1 arteria coronari a origine in le arteria pulmonar, bicuspide valvula pulmonar, drainage del vena dextra-pulmonar a in le atrio dextere, e fenestration del cuspides del valvula aortic. Iste combinatio de congenite defectos cardiac se accorda le melio con le typo II del classification de congenite malformationes del corde secundo Spitzer. Viste que iste homine viveva 34 annos, il es necessari supponer que ille habeva un ben-disveloppate circulation pulmonar collateral.

REFERENCES
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