Spontaneous Closure of Ventricular Septal Defect

Anatomic Proof in an Adult with Tricuspid Atresia

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Spontaneous closure of ventricular septal defect has been suspected on the basis of clinical examinations by physicians caring for children with congenital cardiac disease. Confirmative clinical and hemodynamic evidence documenting spontaneous closure of such lesions also have been presented in several recent reports.1-6 In only one patient, however, has anatomic proof of spontaneous closure of a ventricular septal defect been recorded.7

We recently studied an adult patient in whom the diagnosis of tricuspid atresia was established and who died after operation. At autopsy, there was unequivocal evidence that a functional ventricular septal defect had been present and had subsequently closed. The clinical and pathologic observations leading to this conclusion are summarized in this report.

Clinical Summary

A. H. (No. 03-87-02), a 27-year-old man, had had cyanosis, clubbing, and a precordial murmur since infancy. During childhood and adolescence, fatigue, dyspnea, and repeated upper respiratory infections prevented him from attending school. At the age of 17 a left subclavian-pulmonary arterial anastomosis was performed at another hospital. The cyanosis and dyspnea, however, were only transiently improved, and his physical activity became progressively limited.

On examination he was cyanotic, and there was marked clubbing of the fingers and toes. The heart was enlarged, and a left ventricular thrust was palpable. The second sound at the base was single, and a grade II/VI ejection-type systolic murmur and a faint continuous murmur were heard at the upper left sternal border. The electrocardiogram revealed left ventricular hypertrophy, left axis deviation, left atrial enlargement, and abnormal initial forces indicative of an old antero-septal myocardial infarct. Fluoroscopic and radiographic examinations disclosed enlargement of the left ventricle and hypoplasia of the pulmonary arterial segment. The hematocrit value was 82 per cent.

At right heart catheterization the catheter passed across an interatrial communication into the left atrium and then into a ventricular chamber, where a pressure of 116/12 mm. Hg and an oxygen saturation of 83 per cent were recorded. Simultaneously, the systemic arterial pressure was 112/66 mm. Hg and systemic arterial oxygen saturation was 88 per cent. Neither the pulmonary artery nor the right ventricle was entered by the catheter. Indicator-dilution curves indicated a large right-to-left shunt at the atrial level. A selective angiocardogram with right atrial injection confirmed the clinical diagnosis of tricuspid atresia.

At operation an anastomosis was created between the distal end of the right pulmonary artery and the proximal end of the superior vena cava. The procedure was complicated by the presence of an extensive collateral circulation between the lung and chest wall, and the patient died in the early postoperative period of massive and uncontrollable bleeding into the pleural space.

Pathologic Findings

The pertinent patho-anatomic features of the heart are summarized in figure 1 and illustrated in figures 2 through 4. A closed defect was present in the basal portion of the muscular ventricular septum. The gross and microscopic appearance of this lesion is shown in figure 3.

Discussion

In this patient the evidence provided by both gross and microscopic study furnishes proof not only that a ventricular septal defect had been present but that prior to its spontaneous closure it had been of functional significance. This is indicated by the promi-
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Diagram summarizing the multiple cardiac anomalies in the patient herein described. There is atresia (agenesis) of the tricuspid valve, a large atrial septal defect (A.S.D.), a large left ventricle (L.V.) (functional single ventricle), a ventricular septal defect (V.S.D.) which has closed, and a hypoplastic right ventricle. The pulmonic valve is bicuspid. S.V.C., superior vena cava; I.V.C., inferior vena cava; R.A., right atrium; L.A., left atrium; P.V., pulmonary vein; and P.T., pulmonary trunk.

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nent jet lesion still evident in the right ventricle and also by the size of the right ventricle. For some time prior to the terminal operation and death, pulmonary blood flow was supplied entirely by systemic collateral vessels and by the subclavian-pulmonary arterial anastomosis. The right ventricle was functionless and received no blood except that minute amount which may have been returned to it from Thebesian vessels or retrograde through the pulmonic valve. Had this situation been present throughout the patient’s life, the right ventricle would probably have been atretic. Instead, its cavity, although small, approximated the size of the pulmonary trunk, which was essentially normal. It seems clear, therefore, that the right ventricle attained its size as a result of ejection of blood that was shunted into it when the defect was patent.

Spontaneous closure of a ventricular septal defect is probably a relatively unusual occurrence, and it would appear likely that only those defects whose margins are entirely muscular can do so. Edwards has suggested that the closure of defects of this type, which occur relatively infrequently, may be related to progressive elongation of the myocardial fibers bordering them. Initially the defect may be round or oval but with growth it becomes slit-like and finally its margins approximate each other as the myocardium hypertrophies and stretches. In the elderly patient reported by Edwards closure was apparently effected entirely by apposition of muscle. In the present patient this process also was operative but actual closure resulted from endocardial proliferation, probably stimulated by turbulent blood flow through the defect. In the usual type of ventricular septal defect, involving principally the membranous septum, muscle approximation is impossible and it would seem unlikely that closure of a defect in this location could occur without the superimposition of an active inflammatory process, such as bacterial endocarditis. In this regard, it should be noted that the patient described gave no history suggestive of endocarditis and there were no lesions in the myocardium bordering the closed defect that suggested previous inflammation.

Of additional interest is the prolonged survival of the present patient. Recently Fontana and Edwards reported 125 cases of tricuspid atresia confirmed at autopsy, 119 of which were collected from the literature. Two thirds of these patients died within the first year of life and only eight lived for more than 10 years. Probably the main factor allowing such a long survival in our patient was the extensive bronchial collateral circulation. The left subclavian-pulmonary arterial anastomosis, which was performed when the patient was 17 years old, further augmented the collateral blood flow to the lungs. The decrease in pulmonary blood flow during his latter years, as evidenced by increasing cyanosis and disability, no doubt was caused by pro-
Photographs showing the interior of the cardiac chambers. Upper left: The right atrium. No remnant of the tricuspid valve is present. The ostium of the coronary sinus is also atretic. The atrial septal defect (D.), which measures 3.5 by 2.0 cm., is of the foramen ovale type. The superior (S.V.C.) and inferior (I.V.C.) venae cavae are connected normally to the right atrium. Upper right: The left atrium (L.A.), mitral valve and left ventricle (L.V.). The valve guarding the foramen ovale is totally incompetent resulting in the large atrial septal defect (D.). The dashed circle depicts the communication between the coronary sinus (C.S.) and the left atrium. The left ventricular chamber is considerably dilated and its wall thickened. Lower Left: The ascending aorta (Ao.), aortic valve (A.V.), and septal wall of the left ventricle (L.V.) are shown. The ventricular septal defect (V.S.D.), which has closed, is located immediately below the aortic valve. This view also illustrates the normal continuity between the anterior leaflet of the mitral valve (A.M.L.) and the aortic valve. The ostia of the coronary arteries are apparent. These vessels were widely patent and normally distributed. Lower right: The anterior wall of the hypoplastic right ventricle (R.V.) has been removed, exposing the site of the former defect (V.S.D.) in the muscular ventricular septum. Note the jet lesions on the endocardium of the right ventricle adjacent to the site of the former opening in the ventricular septum. The pulmonic valve (P.V.) and pulmonary trunk (P.T.) are only slightly smaller than normal. Note that the left ventricle (L.V.) accounts for most of the mass of the heart. The inset is the bicuspid pulmonic valve as seen from above. (R.A.) right atrium.

Figure 2
Photographs demonstrating the gross and histologic appearance of the closed ventricular septal defect. Upper left: The closed defect from the left ventricular (L.V.) aspect. The site of the former defect (designated by the arrows) is a linear indentation 1 cm. below the aortic valve (A.V.). The endocardium adjacent to the indentation is elevated, smooth, and pearly white. The endocardial thickening is probably the result of turbulent flow of blood in this area. Lower left: The site of the former defect as viewed from the right ventricular aspect. The anterior wall of the hypoplastic right ventricle has been removed. The depression between the muscle bands is the site of the former defect. The arrow points to the pearly white endocardial thickening, clearly the result of a jet lesion, on the lateral and superior walls of this chamber opposite the depression. (P.V.), pulmonic valve. Upper right: Photomicrograph of a section through the closed defect in the muscular ventricular septum. The entire area of the former defect was blocked, embedded in paraffin, and serially sectioned at intervals of 6 micra. In none of the sections was a residual opening apparent. A representative section is shown here. Note that the actual closure of the defect is produced by fibrous proliferation (jet lesion), and not by direct apposition of the myocardium. No lesions were found in the adjacent myocardium. (R.V.), right ventricle; (L.V.), left ventricle. Verhoeff-Van Gieson elastic tissue stain: original magnification, × 8. Lower right: Photomicrograph of the jet lesion on the laterosuperior aspect of the right ventricle. There is marked fibroelastic thickening of the endocardium. Verhoeff-Van Gieson elastic tissue stain: original magnification, × 21.
gressive closing of the ventricular septal defect.

References

chial collateral blood flow which was further augmented by the subclavian-pulmonary arterial anastomosis. Upper: The descending thoracic aorta is opened. The dilated ostia of the bronchial arteries are designated (arrows). Lower: Photomicrograph of a section of lung demonstrating the dilated and thick-walled bronchial arteries (arrows). The bronchial cartilage is on the right. Verhoeff-Van Gieson elastic tissue stain: original magnification, × 16.
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