Double-Outlet Right Ventricle with Intact Interventricular Septum

By H. Edward MacMahon, M.D., AND Michael Lipa, M.D.

WITHAM in 1957 introduced the term "double outlet right ventricle" to designate an unusual anomaly of the heart and great vessels in which both the aorta and the pulmonary artery arise separately from the right ventricle. He described four of his own cases and several others that he had collected from the literature. In all of these there was a patent interventricular septum. Neufeld et al. in 1961 and again, a year later, published two small series, totaling 17 cases in all, of the same anomaly, and in each of these there was also a patent interventricular septum. This combination of total transposition of the aorta into the right ventricle and a patent interventricular septum, has been so consistently found together, that Neufeld has simply referred to the latter as an anomaly that is universally present in this type of double outlet. The purpose of this report is to describe a pure case of "double outlet right ventricle" in which there was neither a septal defect nor any other major cardiovascular anomaly.

Report of a Case

Clinical Summary

The prenatal and obstetrical histories were unremarkable and for the first 6 weeks the infant developed normally. At about this time, cyanosis was first noticed. By x-ray, the heart was enlarged but the lungs were clear. At 10 weeks of age, cyanosis was more marked, tachycardia was noted, and by x-ray there was prominence of the right side of the heart and selective enlargement of the left atrium. An electrocardiogram at this time was interpreted as showing combined atrial enlargement, right ventricular hypertrophy, and possibly a single ventricle. An apical impulse was felt medial to the left midclavicular line with a pansystolic ejection murmur at the left sternal border in the third and fourth intercostal spaces. The child became increasingly ill and during the last 2 weeks tachypnea, tachycardia, irritability, and cyanosis increased progressively. At the end of the twelfth week, the infant died suddenly immediately after a feeding.

Autopsy Summary

With the exception of cyanosis of the lips and nails, the external appearance of the infant was quite unremarkable. The pericardium was distended by a large cyanotic heart lying transversely across the diaphragm (fig. 1). The anterior projection was represented by a dilated right atrium and atrial appendage; an enlarged right ventricle formed the visible apex of the heart. The left atrium was moderately dilated and projected beyond the left margin of the heart. The left ventricle was represented by a relatively small, very firm conical mass lying posteriorly. This was separated from the large right ventricle by a distinct indentation or groove. The pulmonary artery, slightly larger than the aorta, was in its normal position. The aorta lay to the right and almost parallel to the pulmonary artery. When viewed in situ, both major arteries

Figure 1

The heart as it was seen at the autopsy table after the pericardium had been removed. Note its transverse position with the large right atrium and right ventricle lying on the diaphragm. A small tab of left atrial appendage projects to the left beyond the margin of the right ventricle. The right ventricle forms the visible apex of the heart. The left ventricle is not visible. The distended innominate veins and the superior vena cava lie immediately above the right atrium. Only a very small portion of the pulmonary artery and aorta are visible in this field.

From the Department of Pathology, Tufts-New England Medical Center, Boston, Massachusetts.

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appeared, on superficial examination, to arise normally. The inferior and superior venae cavae and the four pulmonary veins were dilated but could easily be followed to their respective atria. The heart weighed 65 Gm. which is almost three times normal. When opened, the aorta and pulmonary artery were found emerging vertically side by side from the right ventricle (fig. 2). The valves were well formed and clearly separated from one another, and lay in the same cross-sectional and coronal planes. The wall of the pulmonary artery was slightly thickened and the lumen slightly larger than that of the aorta. The right atrium and atrial appendage were dilated. There was a small opening 2 to 3 mm. in diameter in the fossa ovalis, providing a very limited but free communication between the two atria. The tricuspid valve and the right ventricular cavity were dilated and the wall of the right ventricle was both hypertrophied and stretched. The cavity of the left atrium was large and offered a striking contrast to the mitral ring and mitral valve, which were extremely small. The entire left ventricle was represented by an almost solid mass of muscle showing in cross section a small contracted centrally placed empty ventricular cavity. The endocardium of the left ventricle was represented by a thick blanket of gray fibrous tissue that reached out into the myocardium along the Thebesian sinuses. There was no communication between the two ventricles, and there was nothing to suggest a pre-existing defect that might have undergone atresia. The ductus arteriosus was closed. One very interesting finding that we had overlooked, and one to which Edwards4, 5 drew attention as a possible basis for murmurs, was a minute "jet lesion" or endocardial pocket, directly opposite the interatrial septal defect on the anterior wall of the right atrium. The lungs were large, rubbery, congested, edematous, cyanotic, and heavy. Each pleural cavity, and also the peritoneal cavity contained little clear watery fluid. All other organs were congested and cyanotic. The only other gross anomaly in the entire body was an anterior displacement of the ascending and transverse colon. Lastly, it should be mentioned that the trachea and major bronchi contained recently aspirated stomach contents.

Because the only significant histologic findings were confined to the heart and lungs, the microscopic description is limited to those organs. The muscle fibers of both right and left ventricles were hypertrophied, and in the myocardium of the left ventricle there were several small sharply defined areas of very recent and older infarction. A heavy blanket of fibroelastic tissue covered the endocardium of the left ventricle and extended out along the surfaces of the Thebesian sinuses into the myocardium. All these sinuses were narrowed by this tissue and some were occluded by superimposed recent and older organized and recanalized thrombi. Arterioluminal and arterio-sinusoidal vessels of the type described by Wearn 6, 7 and Williams, 8 which offer a communication between small arteries and venous sinuses of the myocardium on the one hand, and the cavity of the left ventricle on the other, were accentuated. In the case of the arterioluminal vessels one could readily trace the gradual transition of a Thebesian sinus into a small myocardial artery with its internal elastic lamina and well-formed muscular wall. As for the lungs, there was extreme congestion and edema of both alveolar spaces and walls.

**Discussion**

The pathophysiology of this case seems quite simple. With an aorta arising from the right ventricle and with no communication between the right and left ventricles and with only a very small opening in the interatrial septum, it is obvious that a very dangerous situation exists. If the interventricular septum had been patent in this infant, as is
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the “rule” in cases of double outlet from the right ventricle, oxygenated blood from the left ventricle could have reached the systemic circulation, but, in this case, there was only a small and very inadequate opening in the interatrial septum through which oxygenated blood could reach the right side. For the first 6 weeks one must assume that sufficient oxygenated blood to meet the needs of the child was being shunted from left to right through a patent foramen. Later, with narrowing of this foramen, with progressive pulmonary congestion and with the growing needs of the infant, this type of circulation became totally inadequate.

There is one case reported by Edwards⁹ that has many features in common with the case under discussion. An infant of about 4 months of age had a complete transposition of the aorta, with both great vessels arising from the right ventricle. A suspected interventricular septal defect was closed by adhesions between this defect and the anterior mitral leaflet. In any case there was no patency in the interventricular septum. There was one small opening in the interatrial septum and there was a second, but very inadequate, opening in the anterior mitral leaflet through which blood could pass directly from the left atrium into the right ventricle. There was one other vascular anomaly in that case, namely, a single coronary artery. Though these two cases differ anatomically, they were functionally very similar.

There were several other interesting histologic features in our case that may warrant brief comment, such as the fibroelastosis of the endocardium of the left ventricle, the mural thrombosis of the endocardium and Thebesian sinuses, and the signs of recurrent infarction of the thickened wall of the small left ventricle. These infarcts were apparently of little or no significance, since the left ventricle played little or no role in maintaining the systemic arterial circulation. There were two other histologic findings of interest, namely, the fibrous patch on the surface of the right atrium opposite the interatrial defect and the exaggerated arterioluminal communications between the sinuses of the endocardium of the left ventricle and small arteries within the adjacent myocardium. The former, which has been referred to as a “jet lesion” may be easily overlooked in the course of an autopsy, but the latter is probably nothing more than hypertrophy of the Thebesian vascular bed.

Summary

This case is one of a 3-month-old infant showing complete transposition of the aorta, with both great vessels arising from the right ventricle. Of particular interest was the absence of a patent interventricular septum. A single but quite inadequate opening in the interatrial septum was the only visible means by which oxygenated blood from the lungs could reach the systemic circulation. No identical case has been found in the literature.

Acknowledgment

We are extremely grateful to Dr. Jesse E. Edwards, to whom we sent this heart for examination, and for his own personal collection. He and his associates have been of the greatest assistance to us, and it is hoped that a complete clinicopathologic report, with Dr. Edwards and his associates participating, will follow.

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The Scientist's Perspective

Who does not recognize today that the impact of science on society is truly overwhelming in importance, and that the future welfare, if not the very existence, of human society will depend increasingly upon the public understanding of science—not so much of the facts or even the concepts of science as an understanding of what science really is and how it yields its result? Scientists deplore the popular image of science as a benevolent genie who will provide any gift the Master of the Lamp may demand, or the popular conception of scientific method as a sort of "intellectual machine that inevitably grinds out ultimate truth in a series of orderly, predictably sequential 'steps,' with complete accuracy and certainty" (H. K. Schilling, in a paper presented at a meeting of Section L of the AAAS in Atlanta.) Nevertheless, few scientists care to undertake the labor of explaining the real nature of science; in fact, but few of them take time to think the matter out for themselves. The philosophy of science and the history of science are glaringly neglected by the very practitioners of science itself.—Bentley Glass, Johns Hopkins University.
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