An Aortic-Right Ventricular Fistula of More than Eighteen Months' Duration

An Approach to Dynamics

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A 79-year-old man who developed evidence suggesting an intracardiac shunt survived at least 18 months after its onset. Intracardiac mechanisms that may protect the pulmonary circulation from excessive flows are suggested, based on anatomic findings at necropsy.

The establishment of a fistulous tract between the aorta and the right ventricle poses critical problems in cardiopulmonary dynamics. The higher aortic pressure drives an excessive flow of blood into the right ventricle and overloads it. The increased pulmonary arterial pressure and blood flow lead to pulmonary edema and right heart failure, thereby precipitating a fulminating course and early death in most patients who develop such a fistula.

We have observed a patient with such a fistula that had been present for at least 18 months. Anatomic findings in the heart suggest a possible mechanism for the prolonged survival.

Case Report

A 79-year-old white man was first admitted to Mount St. Mary's Hospital on November 21, 1954, complaining of severe shortness of breath. The patient had noted the onset of dyspnea about 4 years previously. Treatment with digitalis and Mercuhydrin gave definite improvement until 2 weeks prior to admission, when mental confusion, weakness, dyspnea, orthopnea, and ankle edema all became marked.

The patient did heavy manual labor all his life and drank heavily for many years. From age 70, he worked as a gardener (mowing lawns and doing other moderately hard work) without difficulty until 4 years before death. Three years before death, a heart murmur was noted.

Examination in May 1954 revealed minimal exertional dyspnea with neither orthopnea nor dependent edema. On hospitalization in November 1954 he was in acute distress. A rough thrill was palpable over the precordium and a machinery murmur was heard over the entire chest, loudest along the left sternal border. The maximal impulse was at the left anterior axillary line. Breath sounds were indistinct, but numerous rales and rhonchi were heard bilaterally. The blood pressure was 162/40. A double femoral arterial sound (Duroziez) was present. The electrocardiogram showed delay in ativoventricular conduction, right bundle-branch block, and digitalis effect with runs of bigeminy. The white cell count was 27,000 per mm$^3$ initially but fell later to 16,000. Four blood cultures were negative. X-ray and fluoroscopy revealed marked cardiac enlargement. The clinical picture was considered compatible with ventricular septal defect, superimposed on arteriosclerotic heart disease. The patient improved markedly on digitalis and Mercuhydrin, and was discharged home.

The patient was readmitted 1 year later complaining of intermittent severe chest pain of 3 weeks' duration, not necessarily related to exertion. His general physical condition was unchanged, except for some progression of the congestive heart failure. Blood pressure was 216/60. The hemogram showed a hematocrit of 35, hemoglobin of 74 per cent, and white blood cell count of 16,000 per mm$^3$. Chest films showed an increase in the heart size and a progression of hilar...
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Fig. 1 Top left. The septal surface of the right ventricle is shown. A wooden probe is inserted into the fibrous orifice of the anomalous fistula. Immediately above the orifice, the hypertrophied crista supraventricularis (CS) is evident as a broad band extending from the left upper corner of the photograph to the right margin of the exposed muscle. The dilated infundibulum (Inf.) is seen above the crista on both the right and left sides of the opened ventricle. The pulmonary artery and valve are above.

Fig. 2 Top right. The probe extends from the fistula in the right coronary sinus, immediately below the right coronary artery (C). Aorta (A) is above.

Fig. 3 Bottom left. Radiograph of the heart showing the fistula at arrow, filled with radioopaque medium. The free walls of the right and left ventricles are on the right and left of the septum respectively.

Fig. 4 Bottom right. Masson trichrome stain of a section of lung showing severe vascular congestion with normal pulmonary vessels (× 100).

congestion with a suggestion of an inflammatory process in the right lung. The electrocardiogram continued to show delay in atrioventricular conduction, right bundle-branch block, and bigeminy.

Again the patient improved markedly with digitalis and Mercuhydrin. He was discharged to a nursing home after 10 days. Except for gradually increasing anasarca, his condition remained essentially unchanged until death on April 11, 1956.

The principal pathologic findings were in the heart, which was markedly enlarged, weighing 625 Gm. The right ventricular wall at the conus ateriosus was 7 mm. thick, and the left ventricular wall was 20 mm. thick. The conus, evident as a saecular dilatation immediately below the pulmonary valve, was clearly differentiated from the sinus of the right ventricle by a markedly hypertrophied supraventricular crest on the septal wall. Immediately below the crest a completely fibrotic band less than 1 cm. in circumference was seen at what appeared to be the junction of 2 columnae carneae (fig. 1). This opening led through a tract to an ovoid opening with rolled fibrotic edges 1 cm. in circumference, near the lower attachment.
of the right coronary cusp (fig. 2). Marked calcification with some granular projections extended into the sinus of Valsalva at the attachment of the aortic cusp. The tract was funnel-shaped with its greatest circumference at the sinus of Valsalva. When it was filled with radiopaque material and x-rayed (fig. 3), a narrow diverticulum was demonstrated in the midportion of the tract extending somewhat beneath the muscle of the crista supraventricularis and ending blindly. The right coronary orifice was immediately above the opening of the tract but had no connection with it.

Microscopically, the fistulous tract was lined by flattened endothelial cells and encircled by a thick band of fibrosis, which in places was relatively hyalinized. In other regions, numerous elongated spindle cells with narrow elongated nuclei were present in parallel arrangement. Many elastic fibers were mixed with the fibrous tissue covering the tract. The right coronary artery and the circumflex branch of the left were patent, but showed considerable atheromatosis. The anterior descending branch of the left coronary artery was occluded by an old sclerotic, fibrotic process 3 cm. from its origin. The myocardium showed no gross evidence of recent or old infarction.

Regions of atelectasis with small foci of bronchopneumonia and bronchitis were present in both lower lobes. The pulmonary blood vessels, microscopically, were congested but showed no significant sclerosis (fig. 4).

Severe arterial and arteriolar nephrosclerosis was evident on microscopic section. Most of the small arteries and arterioles of the adrenal glands showed thickened walls with intimal fibrosis.

**DISCUSSION**

The present case has interest from several points of view, including the mechanism of development of fistulas between the aorta and the chambers of the heart, the special dynamics induced by the abnormal blood flow in such instances, and the potential mechanism for protection against excessive pulmonary arterial pressures and flows.

Anomalous dilatations at the root of the aorta are known to extend in any direction and may produce a fistulous connection with any of the chambers of the heart, the pulmonary artery, and even with the pericardium. These abnormal communications have been attributed to specific developmental defects of the heart and great vessels, especially when the fistulae are found in the young. In older individuals, these lesions are variously considered to result from congenital defects in the ventricular septum or from arteriosclerotic or other degenerative processes. Since these aneurysms and fistulas may point in almost any direction, the development of a particular type of fistula may be ascribed to the combination of the mechanical forces and any local site of weakness, rather than to a congenital anomaly. The present ease history provides no evidence for a congenital defect. The presence of such a large fistula in the heart of a 79-year-old man whose history was that of a hard laborer strongly suggests that the lesion was acquired.

Erosion of the base of the aorta may lead to a burrowing through the ventricular septum, with the establishment of a new channel lined by endocardium. In the present instance, the extensive and specific pattern of development of endothelium, collagen, and elastic tissue suggests the action of mechanical forces in transforming the interventricular tissues into structures resembling a blood vessel under considerable mechanical tension.

Most cases with an acquired fistula report a signal event characterized by sudden pain in the chest or the upper abdomen and the immediate appearance of a loud continuous murmur near the sternum. The development of the fistula may be more gradual, since no report of an acute episode could be elicited in our patient and in other reported cases. Usually the development of a ventricular septal defect or an aortic-ventricular fistula leads to rapidly progressive congestive failure, sometimes associated with endocarditis. In the present instance, the fistula apparently was present for at least 18 months, and perhaps for the entire 6 years that the patient was under treatment for heart failure.

The murmurs and arterial pressures suggest that flow took place from the aorta into the right ventricle during most of the cycle. The machinery-like character of the murmur suggests that particularly during early ventricular systole, some of the flow may have
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been pumped by the right ventricle into the aorta. Cardiac nodules at the aortic and ventricular orifices of the fistulous tract provide evidence for the reaction of these tissues to a strong stream.

The compatibility of such a large fistula with long survival is of considerable interest. In the presence of the fistula, the high aortic pressure would tend to drive a rapid flow of blood into the right ventricle, with a resulting excessive pulmonary blood flow. This increased flow may account for some of the pulmonary congestion, dyspnea, and signs of right heart failure.

Of particular interest in the present case was the site of the opening of the fistula immediately below the remarkably well developed supraventricular crest of the right ventricle. This muscle band, interposed between the ventricular opening of the fistula and the outflow tract of the chamber, has been implicated on the basis of phylogenetic and cardiodynamic evidence as a mechanism protecting the pulmonary vasculature against excessive blood pressures and flows. In congenital ventricular septal defects, the high left ventricular pressures are transmitted directly to the right ventricle and then to the pulmonary vessels. Clinical evidence has suggested that in such cases, the supraventricular crest may act as a sphincter during midsystole to prevent these high pressures from being transmitted to the pulmonary vessels. The fact that the fistula entered the right ventricle below the markedly hypertrophied supraventricular crest would place the crest in a position to provide a degree of regulation of the potential volume of the shunt. A partial protective action of the crest might thus act to reduce the tendency to excessive pressures in the pulmonary arteries. Catheterization data in a case of Semler and Brandenburg demonstrated a definite increase in systolic pressure when the catheter was withdrawn from the outflow to the inflow part of the right ventricle, indicating the presence of some degree of obstruction to outflow from the right ventricle. Further, the evidence in our case for a long-standing large shunt (loud murmur, high pulse pressure, and low diastolic pressure), and the absence of significant pulmonary vascular sclerosis suggests protection of these vessels against such high pressures.

Definitive diagnosis of these defects through clinical study and cardiac catheterization has recently become possible. Attempts at surgical repair would appear to be indicated in some cases. A fistula between the sinus of Valsalva and the right atrium, diagnosed by aortography and cardiac catheterization, has been successfully closed by open-heart surgery.

SUMMARY

A man of 79 years with progressive congestive failure was found to have a low diastolic pressure, a wide pulse pressure, and a machinery-type murmur over the precordium. He responded repeatedly to medical therapy for congestive failure. A diagnosis of an intracardiac shunt was made about 18 months before death. Autopsy revealed a large heart with a fistula joining the sinus of Valsalva to the right ventricle immediately beneath a hypertrophied crista supraventricularis. Clinical, physiologic, and pathologic aspects of the case are discussed.

SUMMARIO IN INTERLINGUA

In un masculo de 79 annos de etate con progressive disfallimento congestive, basse tension diastolique esseva constatate insimul con un large pression de pulse e un murmure del typo-de locomotiva supra le precordio. Le patiente respondeva repetemente a thera-pia medical pro disfallimento congestive. Le diagnose de un shunt intracardiac esseva facete circa 18 menses ante morte. Le necro-pisia revelava un grande corde con un fistula inter le sino de Valsalva e le ventriculo dex-tere, immediatemente infra un hypertrophiade crista supraventricularis. Aspectos clinic, physi-ologice, e pathologic del caso es discutite.

REFERENCES

Harvey and Sydenham, types of the scientific and the practical physician, though contemporaries, were uninfluenced, so far as we know, by each other’s work or method. Harvey had little reputation as a practical physician, and Sydenham cared little for theories or experiment. Modern scientific medicine, in which these two great types meet, had its rise in France in the early days of this century. True, there had lived and worked in England the greatest anatomist and medical thinker of modern times; but John Hunter, to whose broad vision disease was but one of the processes of nature to be studied, was as a voice crying in the wilderness to the speculative, theoretical physicians of his day.—WILLIAM OSLER, M.D. *Influence of Louis on American Medicine.* Johns Hopkins Hospital Bulletin, 1897.
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