Ruptured Sinus of Valsalva

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THE ENTITY of ruptured sinus of Valsalva has been known to pathologists for many years, the first report having been published in 1840 by Thurnam. Since then the condition has been described on many occasions, but it was not until 1956 that this defect was successfully repaired by operation. The increasingly widespread use of extracorporeal circulation and a growing awareness of the clinical manifestations and operability of this syndrome have resulted in a moderate number of cases being operated on. Because the mortality rate of operation using extracorporeal circulation is remarkably low, it seems worthwhile to report three additional cases as a means of reemphasizing the pitfalls in diagnosis and treatment, and at the same time to encourage operation as a safe and satisfactory means of treating this condition.

Report of Cases

CASE 1.—A 40-year-old man had been symptomatically well most of his life. However, early in his youth he was told that he had a cardiac murmur which was believed to be due to rheumatic heart disease and he was refused entrance to the Army on this account. He was first seen at Colorado General Hospital in May, 1957, with a complaint of some dyspnea on exertion for about two years, and he had been receiving digitalis. He had not had any chest pain nor had he had any sudden episodes of dyspnea.

On examination he was found to be a healthy looking man with a good color. BP was 132/88. His heart was not clinically enlarged and there were no thrills. A continuous machinery murmur was audible over the second left interspace and a grade 3 diastolic murmur was also audible along the lower left sternal border. There was no evidence of congestive heart failure. His electrocardiogram was that of left ventricular hypertrophy, and his x-ray showed a slight increase in pulmonary vascularity with a definite enlargement of both main pulmonary arteries.

A clinical diagnosis of patent ductus arteriosus was made and the patient was operated upon on July 9, 1957. However, the maximal thrill found at operation was over the base of the pulmonary artery and no thrills were present over the ligamentum. The ligamentum was ligated, but a diagnosis of aortopulmonary window was made and the chest closed with the idea of reoperating at a later date.

The second operation was carried out on Sept. 12, 1957, with the patient under moderate total body hypothermia (29.5 °C [85.1 °F]). A bulge was noticed just below the pulmonary valve and a thrill was present which was maximal over the bulge and extended up the pulmonary artery. No aortic thrills could be felt nor were there thrills low over either ventricle or over either atrium. Therefore, after reconsideration it was decided that the correct diagnosis was that of a fistula from aorta to right ventricle. Under inflow occlusion the pulmonary artery was opened, and by looking down through the pulmonary valve, a 5 mm defect could be seen high in the right ventricle immediately below the pulmonary valve. This was closed with two mattress sutures. Circulation was restarted after six minutes of occlusion. A few minutes later ventricular fibrillation occurred from which the patient was successfully resuscitated. It was then noticed that there was still a persistent but diminished thrill over the upper part of the right ventricle, but because of the development of ventricular fibrillation it was not thought advisable to have a second period of inflow occlusion to attempt complete closure of the defect. The patient made an uneventful recovery. The continuous murmur was still present but much less than preoperatively. The patient has since been lost to follow-up, and his subsequent progress is not known.

CASE 2.—A 6-year-old girl was first admitted to Colorado General Hospital on Oct. 6, 1955. She had been born prematurely and when six weeks old, was noticed to have a murmur. Her development was slow but generally satisfactory.

She was a thin, poorly developed girl but alert and without cyanosis. BP was 120/10. Her arterial pulsations were all readily palpable and overly active, and her heart was enlarged. A long prominent systolic thrill was palpable over the precordium but was maximal along the left sternal border. A harsh obstructive murmur was audible in the same area with a decrease diastolic murmur, and a blowing machinery murmur was present in the second left interspace.
There was no evidence of congestive failure. The electrocardiogram was that of biventricular hypertrophy, and x-rays showed the pulmonary vascularity to be greatly increased. A cardiac catheterization was carried out and she was found to have only minimal evidence of a left-to-right shunt, but the pulmonary artery pressure was 44/25 and her systemic and pulmonary blood flows were both increased. A retrograde aortogram did not show filling of either pulmonary or coronary arteries and was interpreted as ruling out a patent ductus.

On Oct 11, 1956, she was catheterized again, and the information obtained was the same as that acquired previously. It was felt, on reviewing all the data, that the diagnosis of a ventricular septal defect (VSD) and aortic insufficiency (AI) was most likely with a patent ductus next for consideration. However, by April, 1959, the diagnosis of a sinus of Valsalva fistula into the right atrium was considered to be the correct interpretation of her findings, because there was a slight but definite increase in oxygen saturation at the right atrial level.

On April 9, 1959, she was operated upon under moderate hypothermia (30.5°C [86.9°F]). The right auricle was found to be big and there was a prominent thrill over the lateral atrial wall: digital exploration confirmed a defect just above the tricuspid valve. A short period of inflow occlusion was used to reconnoiter the situation, and a 6 mm hole above the tricuspid valve was seen through which a view of the upper aspect of the aortic valve could be obtained. During a second period of inflow occlusion this defect was closed with three mattress sutures. After the circulation was restarted a diastolic thrill became noticeable over the posterior aspect of the left ventricle, but the systolic thrill had disappeared. A decision was made not to attempt any repair of the aortic insufficiency at this operation. Postoperatively the child progressed well but six months later still had both aortic insufficiency and a continuous murmur. During the next few months her heart enlarged somewhat, and because it was felt that she still had a communication between her right ventricle and aorta, reoperation was advised.

A second operation was carried out on Sept 15, 1960, using extracorporeal circulation and mild hypothermia (30.5°C [86.9°F]). The right auricle was found to be big and there was a prominent thrill over the lateral atrial wall: digital exploration confirmed a defect just above the tricuspid valve. A short period of inflow occlusion was used to reconnoiter the situation, and a 6 mm hole above the tricuspid valve was seen through which a view of the upper aspect of the aortic valve could be obtained. During a second period of inflow occlusion this defect was closed with three mattress sutures. After the circulation was restarted a diastolic thrill became noticeable over the posterior aspect of the left ventricle, but the systolic thrill had disappeared. A decision was made not to attempt any repair of the aortic insufficiency at this operation. Postoperatively the child progressed well but six months later still had both aortic insufficiency and a continuous murmur. During the next few months her heart enlarged somewhat, and because it was felt that she still had a communication between her right ventricle and aorta, reoperation was advised.

A second operation was carried out on Sept 15, 1960, using extracorporeal circulation and mild hypothermia. There was a diastolic thrill around the root of the aorta and a systolic thrill could also be felt over the outflow tract of the right ventricle. During perfusion, the heart was stopped with cold and the aorta opened. The right coronary cusp of the aortic valve seemed to be unusually large and prolapsed somewhat into the ventricle; but a careful search of all the sinuses did not reveal any communication between aorta and right atrium or ventricle. The commissures of the right coronary cusp were closed slightly to increase the competency of the aortic valve and the aorta was then closed. The right ventricular outflow tract was opened and a series of small openings were observed high under the tricuspid valve through which bright red blood flowed on unclamping the aorta. These were closed with buttressed mattress sutures, after which no more red blood entered the right ventricle. After the heart was closed the systolic thrill was noticed to have disappeared. The patient's immediate postoperative condition was satisfactory, but she developed a bleeding diathesis with profuse oozing. Her general condition deteriorated, and she died eight hours postoperatively.

At autopsy a healed scar was found in the right atrium over the area adjacent to the right coronary aortic sinus. No patent communication could be demonstrated between aorta and the right side of the heart. It was felt that at the first operation an aorto-right atrial defect was closed and that at the second operation a small VSD was closed with partial repair of the aortic insufficiency. Death was due to diffuse hemorrhage.

Case 3.—A 22-year-old Spanish American male was first seen on Dec 13, 1961. He had always been rather more breathless on exertion than his peers but had never required medical attention. One month before he was first seen he was examined for induction into the Army and was told that there was something wrong with his heart. A week later, while driving a truck, he suddenly experienced severe crushing central chest pain, extending through from his sternum to the spine and aggravated by breathing and movement. He was admitted to a hospital where he remained for five days. The pain did not recur, but he was referred to Colorado General Hospital for a further opinion.

He was found on examination to be a robust young man. BP was 125/50; pulse regular and full. A continuous thrill was palpable over the third left interspace and a grade 4 to 5 continuous murmur was audible in the same area. This murmur was louder in the fourth space than might have been expected with a patent ductus; but a patent ductus was at first considered clinically to be the most likely diagnosis. Fluoroscopy confirmed that there was increased pulmonary vascularity, and the EKG showed left ventricular hypertrophy. The patient was admitted for cardiac catheterization, and both right and left heart catheterizations were carried out. A left-to-right shunt was demonstrated at the right ventricular level and a catheter passed retrograde down the aorta took an unusual course and passed directly into the right ventricle. A gradient of 15 mm Hg was noted across the pulmonary valve. Dye studies excluded a patent ductus, but the differential diagnosis between a ruptured sinus of Valsalva and aortic insufficiency and a VSD could not be made without a retrograde aortogram. This clearly demonstrated a large right coronary sinus and a fistula leading from this into the right ventricle (Figure).

Operation was carried out through a midline sternotomy on March 21, 1962, using extracorporeal circulation. When the right ventricle was opened a small high membranous interventricular septal defect was found (diameter 2-3 mm), and above this was a second defect, ending in a fibrous cone-shaped protusion, was observed. When the apex of the cone was excised a 5 mm hole was delineated which entered the aorta above the aortic valve. Both defects were closed with interrupted sutures of silk, buttressed with Teflon felt.
Retrograde aortogram showing flow from the anterior sinus of Valsalva into the right ventricle.

In the immediate postoperative phase he developed hypertension of 160/120 mm Hg and simultaneously began to bleed from his chest tube. Four hours later, after continuing hemorrhage, his chest was reopened and a retrosternal periosteal arterial bleeder was found and secured. His progress thereafter was uneventful.

During the next month his functional capabilities improved markedly, and his only difficulty was with mild recurrent nonspecific pericarditis. He was recatheterized in June, 1963, and closure of the defects was confirmed. His BP was then 120/70. He still had a 10-15 mm Hg RV/PA gradient believed on this occasion to be infundibular in site.

Comment

The introduction of cardiopulmonary bypass for the treatment of ruptured aneurysms of the sinus of Valsalva has greatly improved the treatment of this condition, and thereby stimulated its more frequent diagnosis. In 1957, Sawyers et al reviewed the literature on the subject of congenital aneurysms with or without rupture back to Thurnam's original description and found only 47 cases. However, since the first operations for this lesion, there have been reports of 91 ruptured aneurysms operated on, and at least 67 other cases of this type have been described, including congenital cases both ruptured and not ruptured, 13 of aneurysm associated with Marfan's syndrome, and 12 cases of aneurysms associated with coarctation of the aorta. These reports also include cases acquired from penetrating wounds.

The anatomical defects responsible for aneurysms and fistulae of the coronary sinuses are now well known. Sawyers et al found that rupture had occurred in 37 of 45 autopsied cases, the fistulae terminating in the right ventricle in 19, in the right atrium in 14, and once in each of the following: pulmonary artery, pericardial sac, and both right atrium and ventricle. In contrast, of the 73 operated cases published in which the details are available, 43 (58.9%) ruptured into the right ventricle; 23 (31.5%) into the right atrium; 3 (4.2%) into the left atrium and 2 (2.7%) into the left ventricle. Two others (2.7%) had ruptured into both right atrium and right ventricle. Obviously cases rupturing into the pericardium would not get into this group.

The typical story of onset is that of a male in the late 20's or early 30's who after exertion suffers severe retrosternal chest pain which gradually passes off, but who then develops breathlessness or cardiac failure and is found to have a murmur. Alternatively the onset may be without pain but associated with sudden dyspnea and palpitation, and occasionally sudden death. Aortic insufficiency is found on examination and sometimes tricuspid insufficiency. A high-pitched continuous murmur is audible and is usually maximal one or more spaces lower than the classical murmur of a patent ductus. Cardiac catheterization demonstrates a left-to-right shunt at the level of atrium or ventricle, but the diagnosis is best confirmed by retrograde aortography. Without treatment, death occurs usually from congestive cardiac failure, but sometimes from other causes such as bacterial endocarditis.

This clinical picture of the young adult male as the main sufferer obscures the fact that this diagnosis is now frequently made from infancy to old age. Ainger and Paton reported a case of a 6-week-old female infant successfully treated while other cases have been described in children.
The main differential diagnosis lies between those conditions that cause a continuous murmur, and rupture of other structures in the heart. The commonest error is to diagnose a patent ductus arteriosus, and several of the reported cases including our first case were subjected to thoracotomy with this diagnosis and subsequently reoperated upon. The murmur of a patent ductus is usually in the second interspace and the rapid onset of symptoms with chest pain or congestive failure affords a clue to the correct diagnosis. If a misdiagnosis occurs then two alternatives are present: either the thoracotomy is closed and a second operation is undertaken at a later date; or, if a perfusion system is available using a disposable bag bubble oxygenator which could be assembled and primed while the incision was being extended and cannulations performed, the definitive closure of the defect could be carried out at the same operation. Aortopulmonary window, coronary artery aneurysm, pulmonary arteriovenous fistula, and ventricular septal defect associated with aortic insufficiency must also be considered. Occasionally the rupture of an aortic cusp or chordae tendinea due to bacterial endocarditis may give rise to similar symptoms and signs. The association of a ventricular septal defect with aortic insufficiency can likewise cause difficulty in differentiation.

In the recorded pathological data the association of a ruptured sinus with a ventricular septal defect has been a very common one, and this has also been so in the cases from the literature which have been operated upon. Of the 73 cases with sufficient details reviewed, 14 also had VSD's. Several authors have reported repairing the combined defect but the majority of cases operated on have had a ruptured sinus alone repaired. In the third case reported here the VSD was closed simultaneously without difficulty. The exact anatomical relationship of the two defects is clarified by seeing the inferior aspect of the aortic valve through the VSD and the superior aspect of the valve through the aortoventricular fistula. Certain other abnormalities were also found, eg, two atrial septal defects (ASD) and a bicuspid pulmonary valve with stenosis.

Aortic regurgitation with a ventricular septal defect is probably more difficult to repair than a ruptured sinus, but as extracorporeal circulation is required for both these operations, the surgeon has the opportunity of making the diagnosis and repairing it at the time of operation.

The most important technical aspect of the surgical repair is adequate excision of the fistula to normal aortic wall and then repair by direct suture or patch depending upon the size of the defect, its situation in relation to the aortic valve cusps, and the possibilities of distorting the valve cusps by crimping of the aortic wall.

The recorded mortality rate in operations using hypothermia alone has been high (ie, four out of ten), probably because of the difficulty of repairing the defect adequately in the time available. In our first case, a complete closure was not obtained, and in the second only part of the defect was recognized and closed. The mortality rate with extracorporeal circulation has been low (seven of 59) not including the cases about which details have not been published. Therefore, there is no doubt that the use of a pump-oxygenator is the method of choice.

Recently a series of iatrogenically acquired aortocardiac defects has been described. The diagnosis of these lesions presents the same problems as in the defects with a congenital origin. The development of aortic regurgitation and the persistence of a left-to-right shunt after the closure of a VSD should raise the suspicion that an aortocardiac defect has been created by damage to the root of the aorta or the aortic valve.

An additional cause of aorto-right ventricular or right atrial fistula is trauma, and a number of cases have been reported in which a successful cure has been effected using total cardiopulmonary bypass. Most of the wounds have been caused by stabbing, and the defect has usually been closed only after a period of congestive failure or hemodynamic difficulty associated with the development of a continuous murmur.

Bacterial infection of a sinus with subsequent erosion and rupture has often been reported. This has not, however, been the usual presenting history in those patients who have undergone operation. Now that aortic valve replacement by prosthetic valves is being
widely carried out, occasional cases of bacterial infection of the prosthetic valves will occur. For the same anatomical reasons that erosion through the normal aortic sinuses leads into the right ventricle or atrium it may be anticipated that bacterial erosion from a prosthetic valve could lead to aortoventricular or aortoatrial fistulas.

**Summary**

The clinical and pathological features of ruptured sinus of Valsalva have been reviewed and three cases reported representing some of the problems in diagnosis and treatment.

The treatment of choice is surgical, using extracorporeal circulation and a meticulous closure of the defect. The mortality rate from operation should be acceptably low.

**REFERENCES**

2. Cooley, D. A., in discussion on Sawyers et al., p 41.

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