Cardioaortic Fistula
A Case Diagnosed in Life and Treated Surgically


A case of cardioaortic fistula, due to rupture of an aneurysm of a sinus of Valsalva into the right ventricle, is described. This case was diagnosed in life and treated surgically one year later when the patient was admitted to hospital with bacterial endocarditis. The clinical features are discussed and the role of special methods of investigation in the diagnosis of the condition is defined. The importance of cardiac catheterization is stressed. The operative technique is described briefly. Following the patient's death from the uncontrolled infection an autopsy examination was carried out and the findings in the heart and small pulmonary vessels are given.

Many examples of congenital aortic sinus aneurysms have been described since this cardiac anomaly was first recorded by Thurnam, Morgan Jones and Langley, who described four cases and reviewed 23 others, considered that it was difficult to diagnose in life both the unruptured aneurysm and that which has given rise to a cardioaortic fistula and thought that it was almost certainly impossible to distinguish clinically a cardioaortic fistula from an aortopulmonary septal defect or even from a patent ductus arteriosus. They felt that the development of bacterial endocarditis in a patient with no previous evidence of heart disease should suggest the possibility of an underlying unruptured congenital sinus aneurysm. Falholt and Thomsen have since shown that this lesion may be demonstrated in life by aortography. Venning reported a case of cardioaortic fistula which had been recognised clinically but thought that the diagnosis was a matter of academic interest rather than of practical importance. However, recent advances in cardiac surgery have made treatment of a cardioaortic fistula a possibility. The purpose of this paper is to describe such a case diagnosed in life by cardiac catheterization and subsequently treated by surgery.

Case Report

A girl, aged 17 years, was admitted in January, 1953, a pyrexia emergency. At the age of 9, a cardiac murmur was found during a routine school medical inspection. She was referred to one of us (J. W. B.) and on the basis of a continuous left parasternal murmur was diagnosed as a case of patent ductus arteriosus. At that time her general health was excellent, she was not breathless on exertion, had no cough and she continued symptom free until two weeks before admission when she began to feel weak and shivery and complained of aching in her muscles. On admission she was pale and febrile (103 F.) and blood culture grew a streptococcus viridans sensitive to penicillin. She was treated by injection of 2 million units of penicillin daily for six weeks. After seven days she was apyrexial and recovered without incident.

After treatment, the underlying cardiac lesion was investigated. On clinical examination the radial pulse was regular at 100 per minute and was collapsing. The blood pressure in the arms was 140/0 mm. Hg. Both femoral pulses were palpable. The jugular venous pressure was not raised. The apex beat was palpable in the fifth left intercostal space four inches to the left of the midline and the character of the cardiac impulse suggested left ventricular hypertrophy. There was no clinical evidence of pulmonary hypertension. On auscultation there was a loud precordial systolic murmur of maximum intensity in the fourth left intercostal space in the parasternal line, which was widely conducted throughout the chest and an early high-pitched diastolic murmur maximum in the fourth left intercostal space. At times the murmur appeared to be continuous in the fourth left intercostal space. The electrocardiogram indicated left ventricular hypertrophy (fig. 1). Teleradiographic examination was normal but on screening there was evidence of slight left ventricular hypertrophy (fig. 2). There was increased pulsation of the main pulmonary artery but no expansile pulsation was seen in the peripheral branches. Angiocardiography showed faint but definite reopacification of the pulmonary arteries at seven seconds and the contrast medium remained

From The Regional Cardiovascular Centre, City General Hospital and The University Department of Medicine, Royal Hospital, Sheffield, England.
in the pulmonary arteries for an abnormally long time. The hemoglobin was 10.5 Gm. per 100 ml. The results of cardiac catheterization are shown in table 1.

From these it was concluded that there was a left-to-right shunt of blood into the right ventricle and in view of the low systemic diastolic pressure, it was presumed that this was from the aorta rather than from the left ventricle. A diagnosis of a congenital aneurysm of a sinus of Valsalva which had ruptured into the right ventricle appeared to be in agreement with all the findings of clinical and special investigation. In view of the risk of reinfection, surgical repair was advised but the patient's mother decided against this.

After discharge in March, 1953 the patient remained in good health until August, 1954 when she was readmitted with a second attack of endocarditis. Shortly before admission she developed a boil on the left side of her face and this was probably the source of her infection. On this occasion the infecting organism proved to be a coagulase positive Staphylococcus aureus.

The cardiac signs were the same as on her previous admission, except that cardiac enlargement had occurred, the apex beat being in the sixth left intercostal space in the anterior axillary line. The cardiac enlargement was confirmed by radiologic examination (fig. 2). As the staphylococcus was resistant to penicillin in vitro, the infection was treated initially by streptomycin and later, when there was no response, by aureomycin and erythromycin. After one month, when it was evident that the infection was uncontrolled by the antibiotic therapy, the only hope of cure lay in repairing the fistula and the patient was transferred to Professor P. R. Allison at Leeds General Infirmary for surgical treatment on September 27. Four days after admission to the surgical wards temperatures of 103 F. and 104 F. were recorded although she was still receiving erythromycin. The white cell count had risen from 8,700 on admission to 20,000 per cu.mm. Three days later the pulse became irregular and the electrocardiogram showed dissociation between the auricles and ventricles, the nodal rate slightly exceeding the sinus rate. Blood cultures still grew a Staphylococcus aureus, insensitive to peni-
cillin and streptomycin but sensitive to aureomycin, terramycin and erythromycin in vitro. After trans-
fusion with three pints of blood the patient was explored under hypothermia on Oct. 5, 1954. Blanket cooling was used, assisted by Largactil, Phenergan and Pethidine under general anesthesia. The cooling process was slow, presumably due to her pyrexia, and after three hours the operation was started, when her temperature was 84 F.

The sternum was split from end to end. A strip of pericardium was resected, doubled on itself and sutured into a button in case it was needed for closing the defect. The right ventricle was greatly enlarged and there was a marked expansile pulsation of the conus in diastole. There was a continuous thrill low down in the ventricle but nothing ab-
normal could be felt in the main vessels. There was no dilatation of the aorta or pulmonary artery. The two venae cavae were isolated and string passed round them for occlusion when necessary. The pulmonary artery was then separated from the aorta and a string passed around it. A clamp was applied to the aorta ready for closure. The vessels were occluded in the following order: venae cavae, pul-
monary veins, pulmonary artery and aorta. A long incision was then made in the right ventricle be-
tween two rows of holding sutures previously in-
serted. The pulmonary valves and the conus appeared normal. There was a hole about 1 cm. in diameter partly overshadowed by the uppermost chordae tendinae of the tricuspid valve and in fact not very far away from the upper part of the valve itself. A large vegetation was flapping from it. The heart was thoroughly washed out with a few pints of saline and the vegetation removed. The hole was then closed by means of a mattress suture of braided wire on an atraumatic intestinal needle and reinforced by a single wire suture alongside it. The occlusion seemed to be complete. Sutures of catgut on atraumatic intestinal needles were then passed through the ventricular wall, very frequent flushings with saline being used to try to get air out of the heart. As soon as the sutures had been inserted, the strings were released in the same order in which they had been applied. The heart was dilated and inert with very slight fibrillation. Cardiac massage was applied immediately and adrenaline injected into the left ventricle. Very little tone returned and defibrillation was tried without suc-

cess. A clamp was applied to the aorta, further adrenaline injected into the left ventricle and mas-
sage applied to get adrenaline into the coronary arteries. Such measures as these, combined with frequent defibrillation, were carried out until finally some normal but weak beats occurred. At this stage, calcium chloride was injected and mas-
sage continued until a strong beat developed. Of all the measures used, defibrillation seemed more effective than anything else, particularly when the electrodes were fairly wide part. Some further

sutures were inserted in the wall of the venae cavae and the chest closed with drainage of the mediasti-
num and pleural cavities. During the operation the circulation was occluded for seven and one half minutes when the patient’s temperature was 29 C. Spontaneous and effective beats were produced 35 minutes after releasing the clamps.

During the postoperative period the patient remained comatose but reacted to pain. On the fourth day she spoke to her parents and then re-

gaped into a semicoma again. Throughout this period her systolic blood pressure was 130 but the diastolic pressure was zero. Erythromycin and Terramycin were given throughout and on the fourth day gastroscopy was performed for feeding purposes. However, the infection was uncontrolled and she died on the fourteenth postoperative day with multiple visceral staphylococcal abscesses.

Autopsy Report

The heart was enlarged, mainly due to hyper-
trophy of the right ventricle, and weighed 400 Gm. The right and posterior (noncoronary) sinuses of Valsalva had undergone aneurysmal enlargement, especially the former (figs. 3 and 4). The left sinus of Valsalva was normal. The aneurysm of the right sinus continued as a fistula 33 mm. long to open into the right ventricle just anterior to the anterior cusp of the tricuspid valve on the crista supra-
ventricularis. The aortic valve was incompetent due to partial detachment of the right coronary cusp. The tricuspid and mitral valves were normal, admitting three and two fingertips, respectively. The pulmonary valve was normal. The pulmonary artery arose normally from the right ventricle, was not dilated and was free of atheroma. There were no septal defects. The aorta was free from atheroma. The thickness of the ventricular walls in the region of the apex was 1 cm. in the case of the right ventri-
cle and 1.5 cm. in the case of the left. There were multiple septic infarcts of both lungs and the spleen but not in the other organs, including the brain, kidneys and liver. Swabs from the vegetations on the aortic valve grew Staphylococcus aureus.

<table>
<thead>
<tr>
<th>Site</th>
<th>Blood Oxygen Saturation, Per Cent</th>
<th>Blood Pressure in mm. Hg</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femoral artery</td>
<td>90.5</td>
<td>140/0</td>
</tr>
<tr>
<td>Left pulmonary artery</td>
<td>91.5</td>
<td>—</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>93.6</td>
<td>12 (mean)</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>92.4</td>
<td>—</td>
</tr>
<tr>
<td>High right ventricle</td>
<td>91.0</td>
<td>9 (mean)</td>
</tr>
<tr>
<td>Low right ventricle</td>
<td>93.1</td>
<td>—</td>
</tr>
<tr>
<td>Low right atrium</td>
<td>78.0</td>
<td>—2 (mean)</td>
</tr>
<tr>
<td>High right atrium</td>
<td>83.8</td>
<td>—</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>75.0</td>
<td>—</td>
</tr>
</tbody>
</table>
FIG. 3. View of the sinuses of Valsalva from the aorta. The right, left and noncoronary cusps and the origins of the right and left coronary arteries are shown. The right sinus of Valsalva has undergone aneurysmal enlargement and is covered with vegetations. LCA = Left coronary artery. RCA = Right coronary artery. LAC = Left aortic cusp. RAC = Right aortic cusp. NCC = Noncoronary cusp. V = Vegetations.

FIG. 4. View of the course of the cardioaortic fistula. The fistula opens from the right sinus of valsalva (1) passes through the wall of the right ventricle (2) and opens into the sinus of the right ventricle (3). A matchstick has been passed through the fistula from the aorta to the right ventricle. AV = Aortic valve.
FIG. 5. Two pulmonary vessels stained by Verhoeff/Van Gieson stain to demonstrate muscle, elastic and fibrous tissue. The vessel on the right is an abnormal pulmonary arteriole with a distinct media lying between thick elastic membranes. There is slight intimal proliferation of fibrous tissue. These changes indicate pulmonary hypertension. The vessel on the left is a muscular pulmonary artery. There is no intimal proliferation of fibrous tissue and the thickness of the media is within normal limits.

The Pulmonary Vessels. The elastic pulmonary arteries (>1000 μ in diameter) had no atheromatous deposits on the intima and there was no medial necrosis.

The muscular pulmonary arteries (100 to 1000 μ in diameter) were normal in appearance (fig. 5). The thickness of the media was 10 per cent of the external diameter of the vessel and there was no subintimal proliferation of fibrous tissue. The adventitia was normal. The elastic membranes were thick.

The pulmonary arterioles (<100 μ in diameter) were abnormal (fig. 5). This type of vessel normally has no media but consists solely of a single elastic membrane lying between intima and adventitia. In this case, however, all the arterioles were characterised by a thick media lying between thick elastic membranes and this abnormal media was present in arterioles with as small a diameter as 30 μ.

The veins and venules were normal. There was no thrombosis in any type of pulmonary vessel.

Discussion

Symptomatology

Morgan Jones and Langley2 thought that patients with unruptured congenital aortic sinus aneurysms were characteristically free from symptoms until they developed subacute bacterial endocarditis. Such was the history of this patient. When investigated after treatment of her first attack of subacute bacterial endocarditis, she was found to have a cardioaortic fistula. It is impossible to say whether the fistula in this case had developed as a complication of the infection or was congenital in origin. The absence of any dramatic incident suggests that the fistula was congenital in origin rather than an acquired lesion. Rupture of hitherto silent aneurysms described by Eppinger3 and Venning4 has been characterized by sudden intense breathlessness associated with severe chest pain and vomiting. However, the aneurysm may have ruptured during the first attack of endocarditis without the dramatic symptoms usually associated with this event and this is supported by the fact that the heart was of normal size at this time and then enlarged rapidly in the following year (fig. 2). The continuous murmur which was noted at the age of 9 years is of no diagnostic value since such murums have been noted in the unruptured congenital aortic sinus aneurysms5 as well as in patients with a cardioaortic fistula.

Signs

A high pulse pressure, a murmur, which is at times continuous and at other times “to and fro” in character and of maximum intensity in the fourth left intercostal space, cardiac enlargement and a cardiac impulse suggestive of left ventricular hypertrophy, as were observed in this case, are classical signs of a cardioaortic fistula. Edwards and co-workers6 described a continuous murmur maximum in the second left intercostal space in a case with a systemic blood pressure of 180/0–60 mm. Hg where a congenital aortic sinus aneurysm had ruptured into the right ventricle. Herson and Symons7 and Venning4 also noted continuous murmurs and high pulse pressures in patients with cardioaortic fistula opening into the right auricle but Hirschboek8 and Macleod9 observed only systolic and diastolic murmurs in the region of the sternum in such patients. It is interesting to note that in the present patient the murmurs were at times to-and-fro and at other times continuous in character. The continuous murmur in cardioaortic fistula may be due to the passage of blood through the fistula from the aorta throughout the cardiac cycle but Falholt and Thomsen8 noted that such murmurs have been described with unruptured aneurysms and thought that these were due to the rush of blood in and out
of the aorta in systole and diastole. Although the signs observed in the present case are characteristic of a cardioaortic fistula they are not pathognomonic of one since identical signs occur in the much commoner anomalies of patent ductus arteriosus and aortopulmonary septal defect where a left-to-right shunt occurs distal to the aortic valve and in patients with aortic stenosis and incompetence. It may be impossible to distinguish clinically a cardioaortic fistula from these conditions.

Electrocardiography

The electrocardiogram in the present patient indicated left ventricular hypertrophy (fig. 1). In general, electrocardiograms provide little positive evidence in the diagnosis of the anomaly under discussion. In seven of the patients described or reviewed by Morgan Jones and Langley\textsuperscript{2} where electrocardiograms were available there was left axis deviation in three, right axis deviation in two, auricular fibrillation in one and heart block in one. Venning\textsuperscript{1} first case showed right bundle branch block, first degree heart block and widened bifid P waves, the second a digitalis effect only, the third left bundle branch block and the fifth the Q\textsubscript{3}T\textsubscript{3} pattern of infarction. The electrocardiogram in the case of Edwards and associates\textsuperscript{6} showed sinus rhythm and left ventricular hypertrophy.

Radiological Examination

In the seven cases of cardioaortic fistula reviewed by Morgan Jones and Langley\textsuperscript{2} there was enlargement of both ventricles on radiologic examination but no localized swellings were noted to indicate an aneurysm although Roesler\textsuperscript{10} and Ostrum and his colleagues\textsuperscript{11} have described small projections from the vascular pedicle in patients with unruptured aneurysms. Morgan Jones and Langley\textsuperscript{2} made no comment on the pulmonary arterial pulsations but Edwards and colleagues\textsuperscript{6} noted increased pulsation of the pulmonary arteries in their case and Venning\textsuperscript{1} also found expansile pulsation of the small branches of the pulmonary artery which he considered as marked as in patients with auricular septal defects. This latter author regarded radioscopy of diagnostic importance in cardioaortic fistula but this was not so in the present case where there was no expansile pulsation in the peripheral branches of the pulmonary artery and where the increased pulsation of the main pulmonary arteries and the left ventricular hypertrophy were not pathognomonic of the anomaly.

Angiocardiography

On angiocardiographic examination there was slight reopacification of the pulmonary arteries seven seconds after the injection. The contrast medium must have re-entered the right ventricle from the aorta via the cardioaortic fistula to produce this reopacification but there was no conclusive evidence of this flow and such reopacification as was seen in the pulmonary arteries could theoretically have occurred from an aortopulmonary septal defect or from a patent ductus arteriosus. Unruptured congenital aortic sinus aneurysms have been demonstrated by aortography\textsuperscript{3} and with venous angiocardiography by the present authors.

Cardiac Catheterization

As would be expected, Falholt and Thomsen\textsuperscript{5} found nothing abnormal at cardiac catheterization in a patient with an unruptured sinus aneurysm and without any evidence, they believed, that this investigation would be of no assistance in differentiating a ruptured aneurysm from a patent ductus arteriosus or an aortopulmonary septal defect unless the ductus or septal defect were actually catheterized. However, Edwards and co-workers\textsuperscript{6} catheterized a patient in whom an aneurysm had ruptured into the right ventricle and found a blood oxygen saturation of 78 per cent in the right ventricular inflow tract and 94 per cent in the right ventricular outflow tract which they considered of diagnostic importance, indicating that blood of high oxygen saturation was shunting into the right ventricle. Since the blood pressure in the arms in this patient was 180/0 mm. Hg, cardioaortic fistula was diagnosed. In the case now reported, the diagnosis was based on similar findings. The results of cardiac catheterization (table 1) indicated a shunt of blood of high oxygen saturation into the right ventricle and there was also a high systemic pulse pressure. In contrast to the case of Edwards and associates\textsuperscript{6} in
whom the left pulmonary-artery blood pressure was 60:23 the pulmonary-artery blood pressure was normal when recorded following her first attack of endocarditis a year before death.

**Histology of Pulmonary Vessels**

The hemodynamic changes with this anomaly are similar to those occurring with patent ductus arteriosus. The pulmonary-artery blood pressure in the present case was normal one year before death and, since Heath and Whitaker\(^a\) found that the pulmonary arterioles were normal in eight patients with patent ductus arteriosus with pulmonary artery mean blood pressures of 33 mm. Hg or less, it is believed that the pulmonary arterioles in this case would have been normal with no media if they had been examined at this time. However, after death they were abnormal, with a thick media and thick elastic membranes suggesting that the patient had developed pulmonary hypertension in the year following cardiac catheterization (fig. 5). If the cardioaortic fistula formed during the first attack of bacterial endocarditis, the resultant hemodynamic changes could account for the subsequent development of pulmonary hypertension and the rapid enlargement of the left ventricle.

**SUMMARY**

A description is given of a patient with cardioaortic fistula which was diagnosed in life and treated surgically.

The patient was asymptomatic until an attack of subacute bacterial endocarditis at the age of 17 and evidence is presented which suggests that the cardioaortic fistula developed from the rupture of a congenital aortic sinus aneurysm at this time. On clinical examination after the first attack of endocarditis there was a high pulse pressure, a murmur which was at times continuous and at times “to and fro” in character, of maximum intensity in the fourth left intercostal space, and a cardiac impulse suggestive of left ventricular hypertrophy. In the absence of a classical history of rupture of a congenital aneurysm into the right ventricle or auricle the history and examination did not provide the diagnosis which could equally well have been one of patent ductus arteriosus, aortopulmonary septal defect or aortic stenosis with incompetence.

Electrocardiography and teleradiography were normal. Angiocardiography revealed faint but definite reopacification of the pulmonary arteries. Cardiac catheterization demonstrated a shunt of oxygenated blood into the right ventricle and established the diagnosis.

A second attack of endocarditis occurred a year after the first. In the intervening period cardiac enlargement had occurred. Since the staphylococcus causing the second infection was resistant to antibiotic therapy, an attempt was made to treat the infection surgically. The operative technic for repair of the fistula is described briefly. The patient died 14 days postoperatively from an uncontrolled staphylococcal pyemia. Autopsy findings in the heart and small pulmonary vessels are given.

**ACKNOWLEDGMENTS**

We wish to thank Professor P. R. Allison and his staff at the Department of Thoracic Surgery, Leeds General Infirmary, who performed the operation. We also wish to thank Miss E. K. Abbott, Consultant Radiologist, City General Hospital, for radiologic facilities and Mr. C. Lambourne for technical assistance.

**SUMMARIO IN INTERLINGUA**

Es describite le caso de un patiente con fistula cardioaortica que eseva diagnosticate in vivo e tractate chirurgicamente.

Le patiente eseva sin symptomas usque al etate de 17 annos quando ille habeva un attacco de subacute endocarditis bacterial. Nostre observationes suggere que le fistula cardioaortica resultava a ille tempore ab le ruptura de un congenite aneurysma del sinus aortica. Post le prime attacco de endocarditis le examine clinic revelava (1) un alte pression del pulso, (2) un murmure que eseva a vices contine e a altrce vices de character fluctuante, con un intensitate maximal in le quarte spatio intercostal sinistre, e (3) un impulso cardiac que pareva indicar hypertrophia sinistroventricular. Proque le caso non sequeva le historia classic de rupturas de aneurysmas congenite in le ventriculo o auriculu dextere, le diagnose non poteva esser establite super le base del examine o del historia del patiente. Instos eseva equal-
CARDIOAORTIC FISTULA

...mente indicative de un patente ducito arterioso o defecto del septo aortopulmonar o stenosis aortic con incompetencia.

Le constationes electrocardiographic e tele-radiographic eseva normal. Le examine angiocardiographic revelava un leve sed distincte re-opacification del arterias pulmonar. Cathe-terisation cardiac demonstrava un derivation de sanguine oxygenate a in le ventriculo. Isto establiva le diagnose.

Un secunde attacco de endocarditis occurreva un anno post le prime. Durante le intervallo allargamento cardiac habeva occurrite. Proque le staphylococco que causava le secunde infeccion se monstrava resistentente al therapia antibiotic, un tractamento chirurgic eseva tentate. Nos describe brevemente le technica operative que eseva usate in reparar le fistula.

Le patiente moriva 14 dies post le operation in consequentia de un non-controlabile pyemia staphylococcal. Es presentate constatationes autoptic in le corde e le parve vasos pulmonar.

REFERENCES

Cardioaortic Fistula: A Case Diagnosed in Life and Treated Surgically
J. W. BROWN, D. HEATH and W. WHITAKER

Circulation. 1955;12:819-826
doi: 10.1161/01.CIR.12.5.819
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1955 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/12/5/819

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/