Fistulae between the coronary arteries and the right cavities of the heart

Report of three cases treated surgically

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Three cases of fistula between coronary arteries and the right cavities of the heart were studied. Outstanding findings were a continuous murmur with late systolic reinforcement, cardiomegaly ranging from slight to considerable, and lack of correlation between the symptoms of angina and electrocardiographic changes.

Communications between coronary arteries and the venous side of the circulation are a rare type of congenital malformation (2.7 per thousand in the experience of Gasul et al. (1960) and 4 per thousand in that of Nora, McNamara, and Fraser (1967)). Krause (1865) first reported a case of coronary fistula, Abbott (1908) described the illness, and Biörck and Crafoord (1947) reported the continuous murmur.

The right coronary artery is most usually affected, and the fistulae occur, in order of frequency, at the level of the right ventricle, right atrium, pulmonary artery, coronary sinus, and vena cava (Edwards, 1958; Neufeld et al., 1961). The communicating artery is found to be considerably dilated, its course is tortuous, and there may be one communication or many.

Materials and methods

A series of 1200 congenital malformations were studied in the Vizcayan Foundation for Persons Suffering from Heart Disease. Of these, three showed a fistula between the coronary arteries and the cavities of the right side of the heart. Two of the patients were girls and one was a boy. Their ages were 9 months, 13 months, and 9 years.

The diagnosis was established by selective angiocardiography and confirmed at operation. A clinical examination in all cases included case history, physical examination, electrocardiography, phonocardiography, and simple radiology. Cardiac catheterization was performed in all three, pressures being determined with a Schwarcher polygraph and oxygen saturation in the blood samples by the Van Slyke method.

In each case angiocardiography was performed

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A continuous murmur was detected in the right ventricle by means of the intracardiac phonocardiograph.

The aortogram showed considerable increase in calibre of the left coronary artery (both circumflex and anterior descending branches) as the right ventricle and the pulmonary artery opacified through the fistula.

Case 2  A 13-month-old girl had had recurring bronchitis since the age of 5 months. On auscultation she was found to have a continuous murmur of 3/6 intensity with late systolic accentuation, the epicentre being in the 2nd left intercostal space. A third heart sound was present. The arterial tension was 95/60 mm. Hg.

Radiographs showed a moderate cardiomegaly due to enlargement of the left ventricle. The electrocardiogram showed in the frontal plane an axis of an A QRS at 0° and of A T at 20°. There was biventricular hypertrophy (Fig. 2).

FIG. 2 Case 2. The aortogram shows dilatation of both branches of the coronary artery, and simultaneous opacification of the right ventricle and pulmonary artery. A, aorta; P, pulmonary artery; C, coronary artery.

FIG. 3 Case 3. A continuous murmur was detected in the right ventricle by means of the intracardiac phonocardiograph.

The electrocardiogram showed on the frontal plane an A QRS at +30° and the A T at +100°. Signs of right ventricular hypertrophy were recognized, and there were primary alterations of repolarization of the type seen in extensive anterior and high lateral ischaemia.

In the haemodynamic examination peak systolic pressures of 9 mm. Hg in the right atrium and 50 mm. Hg in the right ventricle were observed. Contrast injection at the level of the aortic sigmoid plane showed (Fig. 1) that the left circumflex artery was considerably dilated and that there was a right juxta-auricular aneurysmal dilatation. The pulmonary fields darkened slowly with recirculation of the contrast.

Case 3  A 12-year-old boy was asymptomatic. On auscultation a continuous murmur of 4/6 intensity was heard, the epicentre being in the 2nd left intercostal space. A third heart sound was present.

The arterial tension was 95/60 mm. Hg.

Radiographs showed a moderate cardiomegaly due to enlargement of the left ventricle. The electrocardiogram showed in the frontal plane an axis of an A QRS at 0° and of A T at 20°. There was biventricular hypertrophy (Fig. 2).

FIG. 4 Case 3. The aortogram shows a huge right coronary artery terminating in the right ventricle. AO, aorta; COR, right coronary artery; VD, right ventricle.
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Intensity was heard in the 4th and 5th right intercostal spaces with late systolic accentuation radiating to the whole praecordium. The second heart sound was widely split. The arterial tension was 110/60 mm Hg.

In the radiographs (Fig. 3) cardiomegaly was seen in which the growth of the left ventricle pre-

**FIG. 5** Case 3. Posteroanterior chest film. Cardiac enlargement.

...continued clearly. The hila appeared to be dilated and pulmonary vascularity increased.

The electrocardiogram showed in the frontal plane an A QRS at +60° and A T +80°. Left ventricular, possibly right ventricular, and biastral hypertrophy were present. Primary alterations were detected of repolarization of the type seen in subendocardial lesions and anteroseptal subepicardial ischaemia (Fig. 4).

Cardiac catheterization allowed us to identify a large arteriovenous shunt at the level of the right ventricle (pulmonary:systemic flow ratio 2.36) and pulmonary arterial normotension (22 mm Hg systolic). A continuous murmur was detected in the right ventricle by intracardiac phonocardiography (Fig. 5). The aortogram showed a right coronary artery of large calibre which ended in the right ventricle (Fig. 6).

**FIG. 6** Case 3. Electrocardiogram showing subendocardial and anteroseptal subepicardial ischaemia.

**FIG. 7** Case 3. Phonocardiogram after operation. The continuous murmur has disappeared.
Surgery
Taking into account that ligation of the anomalous artery at its outlet allows correction of the defect by a simple technique, we believe that, in general terms, operation is indicated (Lee et al., 1968) provided that angiocardiography shows that the defect is easily accessible. Progressive cardiomegaly, or incapacitating symptoms, such as dyspnoea or angina pectoris, may make surgical correction an urgent matter.

The benefit of operation is not limited to suppressing an arteriovenous shunt, but operation also improves the coronary perfusion (Effler et al., 1967). The first and third cases were operated on by a closed technique and the second with the use of extracorporeal circulation. In the first case (communication in right atrium) a right thoracotomy was performed, and in the other two, median sternotomy. After dissecting the abnormal coronary artery up to the level of its outlet it was clamped for 10 minutes, the electrocardiogram being observed so that it was possible to proceed with simple ligation of the distal portion of the anomalous artery. Immediate postoperative observation in the three cases showed disappearance of the continuous blowing murmur (Fig. 7). So far we have not been able to identify significant changes in radiographs and electrocardiograms. In the last two cases this may be attributed to the short time that has elapsed between the surgical operation and the last observation.

Discussion
We have described three cases of fistulae between coronary arteries and right cavities of the heart. In two cases the communicating artery was the right coronary and in the other the left (Harris, Jefferson, and Chatterjee, 1969). The fistulae were at right ventricular level in the first two cases and at the right atrium in the third.

When examined for the first time none of the patients had significant symptoms. As in the majority of reports (Gasul et al., 1960; Neufeld et al., 1961; Edwards, Gladding, and Weir, 1958) we did not find evidence of angina pectoris or of cardiac insufficiency in this series. These complications are exceptional, and are rarely referred to (Steinberg, Baldwin, and Dotter, 1958; Cooley and Ellis, 1962; Nora et al., 1967).

As in other reports (Biörck and Crafoord, 1947; Paul, Sweet, and White, 1949; Gasul et al., 1960), we found a continuous murmur of variable intensity in all cases. In some reports the diastolic accentuation is pointed out as a distinctive characteristic of this malformation (Gasul et al., 1960; Sanger, Taylor, and Robicsek, 1959; Davison, McCracken, and McIlvain, 1955). We did not recognize this feature in our three cases.

All the cases had cardiomegaly, which varied from slight to considerable. Radiological analysis of the cavities showed left ventricular dilatation in the three cases, and slight dilatation of the right ventricle in two. Only in the case with a large communication between the right coronary artery and the inflow tract of the right ventricle did we observe a significant dilatation of the branches of the pulmonary artery and an increase of the circulation in the pulmonary fields.

Electrocardiographically we found in two cases left ventricular hypertrophy associated with a possible right ventricular hypertrophy. In one case isolated right ventricular hypertrophy was observed.

In two patients primary alterations of repolarization were found and in one of them they were intense and extensive.

Some authors (Abbott, Rivarola, and Logue, 1961; Knoblich and Rawson, 1956; Sabiston et al., 1963) have described angina in patients with electrocardiographic alterations of an ischaemic type. We did not find such a correlation in our three cases.

Of the two patients in whom oximetric determinations allowed us to quantify the arteriovenous shunt, only one showed a significant flow.

The diagnosis was suspected clinically in the three patients and was confirmed by angiocardiographic examination. The angiocardiograms showed where the communication was established and the dilated and tortuous character of the communicating coronary arteries.

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References
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