Single coronary artery—right ventricle fistula

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A patient is presented with a single coronary artery arising from the left aortic sinus with a fistulous communication to the right ventricular outflow tract. The lesion was shown by selective coronary arteriography and was successfully corrected surgically.

There are only five well-documented cases of single coronary artery—right ventricle fistula (Murray, 1963; Michaud et al., 1963; Noonan and Spencer, 1965; Hallman et al., 1965). None of these patients had selective coronary arteriography.

In this communication, we describe a patient with a fistula between a single coronary artery and the right ventricle; the fistula was demonstrated by selective coronary arteriography and the lesion was successfully corrected by operation.

Case report

A 5-year-old Negro girl was referred for evaluation of a heart murmur which was detected on a routine examination. She was asymptomatic and on physical examination the abnormal findings were limited to the heart. Both heart sounds were normal. A grade 3/6 continuous murmur, high pitched in character, was heard in the third intercostal space at the left sternal border and along the right sternal border. The murmur was conducted to other areas of the precordium, but was not well heard at the left infraclavicular region. Peripheral pulses were normal. Electrocardiogram and chest x-ray were within normal limits.

On cardiac catheterization, all the intracardiac and intravascular pressures were normal. Oxygen saturations showed a 2 per cent step-up at the right ventricular level, giving a pulmonary/systemic flow ratio of 1:2:1. Selective coronary arteriograms showed a coronary artery arising from the left coronary sinus (Fig. 1). It gave off a normal appearing anterior descending branch. The circumflex branch was much dilated and very tortuous. It continued as the posterior descending branch and proceeded anteriorly to the area normally supplied by the right coronary artery. Some of its dilated branches entered the outflow portion of the right ventricle (Fig. 1). Diagnosis of a coronary artery—right ventricle fistula was made and surgical correction was recommended.

At operation, a single coronary artery arising from the left aortic sinus was present. It was much dilated and tortuous. A thrill was felt at the site of the fistula and pressure at this point obliterated the thrill. With further dissection, a 3 mm fistula could be seen arising from the side of the artery and entering the right ventricular outflow tract. It was obliterated with multiple suture ligations. The thrill disappeared after the procedure.

After operation the patient did well and there were no complications. She was readmitted approximately one year later for a follow-up study. At this time, only an aortic root injection was done in the posteroanterior and left anterior oblique views (Fig. 2). A single coronary artery was seen arising from the left aortic sinus and it gave off the anterior descending and the circumflex branches. The circumflex branch was almost normal in size but was still slightly tortuous. There was no fistulous communication.

Discussion

Though McNamara and Gross (1969), in a review of the published reports, collected 172 cases of fistula between a coronary artery and a cardiac chamber or a great vessel, only 6 cases have been reported where a single coronary artery communicated with the right ventricle or pulmonary artery. However, the case reported by Anselmi et al. (1961) had severe pulmonary stenosis and the right ventricle communicated with the aorta through several intramyocardial sinusoids. At necropsy, a single coronary artery was found. Haemodynamically, this is altogether a different anomaly from the one under discussion. Murray (1963) reported 2 cases, in one of which
the fistulous communication was to the right ventricle, while in the other it was to the pulmonary artery. Since then, 3 more cases have been reported (Michaud et al., 1963; Noonan and Spencer, 1965; Hallman et al., 1965). In all of these cases the fistula communicated with the right ventricle. None of these cases had selective coronary angiography.

The present patient was investigated because of a continuous murmur. Cardiac catheterization data were equivocal. Selective coronary arteriogram showed a fistula between a single coronary artery and the outflow tract of the right ventricle. Since the anatomy was so clearly delineated, no problems were encountered at operation.

The left-to-right shunt in such cases may be small and there are no haemodynamic ill effects. The dilated and tortuous coronary artery could be more prone to premature degenerative changes. The fistula provides a low resistance outflow and, therefore, the left ventricular myocardium may become more easily ischaemic, a type of 'steal syndrome'. Further, there could be a risk of developing bacterial endocarditis. Since operation can be accomplished without much risk, all these patients should have the lesion corrected. Selective coronary angiography demonstrates the lesion very precisely.

Most of the cases reported, including the present case, had ligation of the fistula. The case reported by Hallman et al. (1965) had ligation of the fistula and an anastomosis between the aorta and the right portion of the coronary artery using a Dacron graft, thus providing a dual coronary artery system. It is not clear whether the latter technique provides any added advantages.

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References
FIG. 2 Postoperative study. Aortic root injection (anteroposterior and left anterior oblique views). The coronary artery is seen arising from the left aortic sinus. The circumflex branch is much reduced in size compared to Fig. 1.

two-coronary system with dacron graft. Circulation, 32, 293.

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