Spontaneous closure of coronary artery fistula

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SUMMARY A coronary artery fistula diagnosed in a 1 year old girl closed spontaneously during childhood. This outcome has been documented in only three previous cases.

The natural history and management of coronary arterial fistulae remain incompletely defined, particularly with respect to the role and timing of surgery in asymptomatic patients.¹⁻³ We report a case of a right coronary artery to right ventricle fistula which underwent spontaneous closure. This outcome has been only rarely reported.⁴⁻⁷

Case report

A West Indian girl was referred in 1976 at the age of 1 year for investigation of an asymptomatic murmur, which had been discovered on a routine infant examination. Her medical history, birth and development, was unremarkable, and no murmurs had previously been noted. Examination at that time detected bounding arterial pulses and a systolic thrill with a grade 4/6 continuous murmur at the lower right sternal edge. The clinical diagnosis, based on the character and site of the murmur, was a coronary fistula. An electrocardiogram showed changes of biventricular hypertrophy, and a chest radiograph indicated minimal cardiomegaly and pulmonary plethora. Cardiac catheterisation in January 1977 (aged 1½ years) confirmed a left to right shunt at the right ventricular level with a calculated pulmonary to systemic flow ratio of 1.4. Haemodynamic pressures in the right atrium, right ventricle, pulmonary artery, left ventricle, and aorta were all normal. Angiography of the aortic root showed a dilated proximal right coronary artery with a fistulous communication to the right ventricle just below the outflow tract (Figure). It was decided to manage this conservatively, and advice regarding prophylaxis against infective endocarditis was given.

During the period February 1977 to February 1979 the patient's mother reported that the systolic thrill had disappeared. The patient remained asymptoma-

tic. At the end of this period, when aged 4 years, she had no evidence of bounding pulses, systolic thrill, or any continuous murmur. Both the electrocardiogram and chest radiograph were normal. A treadmill exercise test (Bruce protocol) was negative for symptoms or electrocardiographic changes. Repeat cardiac catheterisation in August 1983 (at 8 years) again showed normal haemodynamic pressures, but no evidence of any left to right shunting could be detected. Angiography again showed a dilated right coronary artery ostium but no fistula was evident. A small right coronary artery distal to the previous fistula site was found (Figure).

Discussion

Congenital coronary arterial fistulae, first reported in 1865, were considered rare curiosities before the advent of angiography and selective coronary arteriography.⁸ In recent years, however, increasing numbers of cases have been reported, and almost 300 have now been published.³ The anatomical and clinical variations and haemodynamic profiles have been well reviewed elsewhere.²

Principles of long term management of coronary arterial fistula have not been clearly defined. The natural history is poorly understood, and the proportion of symptomatic patients varies in different series.¹⁻³ Nevertheless, symptoms do appear to relate to increasing age. Furthermore, the incidence of fistula related complications, such as congestive heart failure, myocardial ischaemia/infarction, and death, appears to increase with older age groups.¹ In contrast the risk of infective endocarditis seems constant at 3-4% of patients with coronary arterial fistula. For these reasons routine early elective closure of the fistula has been recommended.¹⁻³ Previous reports of surgical treatment have resulted in an appreciable mortality (4%) and morbidity (10-15%), the latter mainly being due to recurrence of the fistula and to

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Figure Angiograms of the aortic root in a child at (a) aged 1½ years showing a fistula from the right coronary artery to the right ventricle and (b) at age 8 years showing closure of the fistula.

myocardial ischaemia and infarction.¹² With the availability of the cardiopulmonary bypass technique and since the introduction of coronary artery bypass surgery recent series have emphasised the absence of

any operative related mortality and a low long term morbidity (3–5%).^{3,9}

Spontaneous closure of coronary arterial fistula has been angiographically documented in only three previously reported cases. All were children (4, 5, and 14 years old), and all had fistulae terminating in the right ventricle.^{4,5,7} In another case, a continuous murmur from a confirmed coronary artery fistula terminating in the right ventricle disappeared at the age of 4 years, but the presumed fistula closure was not confirmed angiographically.⁶ Jaffe *et al* have reported a case of coronary arterial fistula in a 29 year old woman whose continuous murmur had disappeared by the age of 44 years, but this was found to be a result of atherosclerotic occlusion of the right coronary artery.¹⁰ Our case is the fourth reported to have undergone documented spontaneous closure, the mechanisms of which remain obscure. It has been postulated that a high flow leads to shear induced endothelial damage leading to premature atherosclerosis and thrombosis.¹⁰ The right coronary artery distal to the occluded fistula is small and irregular (Figure) and indeed may give rise to future ischaemia. Thrombosis could also occur independently in the intramyocardial and endocardial venous sinusoids. All the reports of spontaneous closure of coronary arterial fistula have been in children, and all such fistulae have terminated in the right ventricle. Thus a further possible mechanism of spontaneous closure, similar to that occurring in muscular ventricular septal defects in children, is by presumed fibrosis and local myocardial hypertrophy.

Routine elective surgical closure of coronary arterial fistula can be recommended in adults, either on the basis of symptoms or to prevent future complications. Surgical closure in symptomatic children is also indicated.¹ The role of surgery in asymptomatic children with coronary arterial fistula remains to be defined, both because of the uncertain natural history and the (small) possibility of spontaneous closure.

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