DESCRIPTION

A murmur was noted in a 2-year-old patient who was otherwise asymptomatic. The patient was born by caesarean section at 37 weeks gestation. No significant family history was present. Clinical examination revealed a continuous murmur at the sternal edge. Echocardiography did not reveal any abnormality. Cardiac catheterisation revealed a communication between the right coronary artery (RCA) and the right ventricle (RV).

On median sternotomy, a dilated tortuous acute marginal branch communicating with the RV was evident (figure 1). After placing the patient on cardiopulmonary bypass, the vessel was tied off with no associated haemodynamic compromise. Postoperative period was uneventful. Follow-up echocardiography showed good left ventricle function.

Coronary artery fistula (CAF) was first described by Krause in 1865. The congenital fistulous connections, between the coronary system and the cardiac chambers, are rare conditions that apparently represent the persistence of the embryonic and sinusoid intertrabecular spaces. Congenital CAFs account for only 0.27–0.4% of all congenital cardiac defects.

The aetiology of CAFs may be congenital, traumatic or iatrogenic, that is, after coronary intervention or valve replacement. Around 55–65% of congenital CAFs arise from the RCA and usually drain into a right chamber; a RCA into a left chamber is less frequent. The most prevalent receiving chamber of CAF (45%) is the RV, followed by the right atrium (25%) and the pulmonary artery (20%).

Children with CAF are generally asymptomatic; in adults a variety of complications may occur ranging from cardiac insufficiency, myocardial ischaemia, infective endocarditis, arrhythmias and rupture. Diagnosis of CAF is based on angiography. Echocardiographic examination may be helpful for diagnosis, usually demonstrating the drainage site and sometimes the dilated fistula itself.

In conclusion, RCA with RV drainage and giant coronary artery aneurysm are rarely seen in congenital CAF.
The optimal treatment for CAF should be coronary surgery, even in the asymptomatic patient.

Competing interests None.

Patient consent Not obtained.

REFERENCES

