Rare disease

A neonatal case of congenital coronary artery fistula

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Coronary artery fistulae (CAF) are rare forms of congenital heart disease with an incidence of one in 50 000 live births. The authors present the case of an asymptomatic neonate with a precordial murmur. Pre and postductal saturations, blood pressure and ECG were normal. Echocardiography revealed a large right coronary artery fistula to the right ventricle (4.5 mm). At 11 months, transcatheter occlusion of the fistula with a vascular plug was performed. A year on, the child was thriving, ECG and echocardiogram remained normal. CAF complications and symptoms (including aneurysm, myocardial ischaemia, angina, heart failure and dyspnoea) are commoner in older patients, so traditionally we intervene early. With increasing case reports of spontaneous closure of even large and symptomatic fistulae, management of especially asymptomatic children is unclear. Long-term complications of intervention also remain largely unknown. As such more information is required on the conditions natural history to better manage patients and counsel parents.

BACKGROUND

Coronary artery fistulas (CAF) are defined as abnormal communications between coronary arteries and cardiac chambers or vessels, bypassing the capillary network.^{1 2} The majority of cases are congenital; hypothesised to represent persistence of embryonic intratrabecular spaces and coronary sinusoids. The incidence of isolated fistula is 1 in 50 000 live births,³ and they account for 0.2–0.4% of congenital heart defects.⁴

We report a rare case of a large, asymptomatic CAF presenting in the neonatal period, treated successfully with percutaneous transcatheter intervention. This leads to a discussion of the problems surrounding CAF management due to insufficient knowledge of the condition's natural history, and the difficulties this creates when counselling parents.

CASE PRESENTATION

A baby boy born at term by elective caesarean section, had a birth weight of 3.48 kg, and Apgar scores of 9 and 10 at 1 and 5 min, respectively. No postnatal resuscitation was required, and there was no history of prenatal complications or abnormalities.

On routine neonatal examination, a precordial systolic murmur was audible. Initial clinical cardiac assessment including pre and postductal saturations, blood pressure and pulse check was normal. The baby was therefore discharged with an outpatient local cardiac clinic appointment and instructions to the mother to seek medical attention if the baby exhibited signs of developing heart failure.

INVESTIGATIONS

At 5 weeks of age the baby was still asymptomatic, feeding and growing well, pink and well perfused, with palpable femoral pulses. Both heart sounds were audible with a grade 2/6, long systolic murmur at the left sternal edge. Echocardiogram revealed a small perimembranous ventricular septal defect (VSD) and an unexpected, significantly dilated right coronary artery (RCA) (figures 1 and 2).

ECG showed sinus rhythm with no evidence of ST segment or T wave abnormalities.

DIFFERENTIAL DIAGNOSIS

A coronary artery fistula was suspected and these findings were explained to the parents with some difficulty. A tertiary cardiology review was arranged where it was confirmed that the infant had a coronary artery fistula draining from the RCA in to the right ventricle (figure 3). Due to the size of the fistula (4.54 mm) cardiological opinion was that it was unlikely to close spontaneously but would require surgical closure. However, as the baby was asymptomatic it was decided to allow the baby to grow and plan the procedure electively once the baby was older and bigger to minimise the procedure related risk.

TREATMENT

At 11 months of age, although still asymptomatic, a transcatheter occlusion was performed with a 4 mm AMPLATZER vascular plug II (figure 4).

OUTCOME AND FOLLOW-UP

Following this there was decent filling of the side branches of the RCA providing some myocardial perfusion to the right ventricle. Postprocedure ECG did not show any ischaemia. He made an uneventful recovery and discharged the following day on aspirin (50 mg daily) for 6 months.

A year on, the child was thriving, ECG and echocardiogram remained normal.

DISCUSSION

Heart murmurs are not uncommon in the neonatal period and often picked up during routine neonatal examination. However, CAF as the cause of neonatal heart murmur is extremely rare. Clinically the position of a CAF murmur is suggestive of VSD, whereas its 'spill-over' into diastole resembles murmurs due to patent ductus arteriosus. The diagnosis may be suspected on an echocardiogram which

Figure 1 Dilated right coronary artery.

Figure 2 Colour Doppler of dilated right coronary artery.

reveals a dilated coronary artery (this has even been detected in utero).⁵ Definitive diagnosis is with coronary angiography and ECG will reveal any evidence of ischaemia.

The most common vessel of origin for congenital CAF is the RCA (50–55%), followed by the left coronary artery (LCA) (35–42%), with 5% from both.^{6–7} The fistulas predominantly drain into low pressure structures; 92% to the right side of the heart, into the right ventricle in 41% of cases, right atrium in 26%, and pulmonary artery in 17%.^{8–9}

Complications include aneurysm formation, myocardial ischaemia and infarction (due to coronary steal phenomenon or thrombus formation), congestive cardiac failure

(due to left to right shunting), arrhythmias and rarely bacterial endocarditis and even rupture, resulting in haemopericardium and sudden death.^{7–14} Symptoms (angina, fatigue, dyspnoea), incidence of death prior to surgical intervention and complications and mortality related to intervention, have been shown to increase with age.¹¹

The traditional viewpoint is to intervene early. Transcatheter occlusion is the preferred therapeutic method as it avoids the need for surgical intervention and cardiopulmonary bypass. Starting postop patients on antiplatelet therapy and bacterial endocarditis prophylaxis is recommended by some.^{15–16}

Figure 3 Catheterisation image showing blood flowing from dilated right coronary artery into right ventricle.

In recent times there is increasing debate as to whether asymptomatic children need intervention. Arguments for include the increased incidence of complications, symptoms and mortality with age, including worse surgical complication rate and mortality; and generally low mortality and morbidity of intervention which provides immediate resolution of pathology and negligible recurrence rate.

Arguments against are based on two points. The first is that there are increasing number of case reports and series revealing spontaneous closure of congenital CAF. Schleigh *et al* presented six cases of complete closure of congenital CAF.¹⁷ Ages of patients ranged from 2–18 months and all were asymptomatic. In three cases, each the fistula was from the RCA and LCA respectively; and all drained to the right ventricle. With a follow-up range of 1.5–7 years, repeat echocardiogram revealed normal coronary artery calibre, and lack of abnormal right ventricle flow. Follow-up of a further 42 patients revealed an overall spontaneous closure rate of 17% (7/42 patients). A similar study by Sherwood *et al* revealed a closure rate of 23% in 31 asymptomatic paediatric patients (age range 7.2±8.4 years), and all patients remained asymptomatic, without complications or evidence of ischaemia for the durations of follow-up (mean age 9.3±9.1 years).

This phenomenon is not just isolated to asymptomatic cases, or those without complications. Farooki *et al* and Aggoun *et al* reported cases of spontaneous resolution on fistulas in infants with symptoms of congestive cardiac failure.^{18 19} Malcic *et al* presented a 7-day-old infant with continuous precordial murmur, diagnosed as a RCA to RV fistula; definitively diagnosed at 1 month of age with aortography.²⁰ Radioscintigraphy and oximetric analysis

Figure 4 Postintervention catheterisation image showing blood flow in the right coronary artery.

revealed 47% left to right shunt, and so surgical intervention was planned, but in the mean time the murmur disappeared, and at 6 months repeat aortography showed spontaneous closure of the fistula. The RCA showed morphological changes (stenosis and aneurysm), but repeat aortography at 1 year of age showed regression of these pathological findings.

The other point is that the long-term complications of intervention remain largely unknown. Cheung *et al* looked at medium- to long-term outcome (range from 5 to 28 year) of 41 patients with CAF who underwent surgical repair in the last 30 years at the Department of Surgery, Grantham Hospital, Hong Kong (age range 2.3 to 58 years).²¹ Among the 21 patients who agreed to a second catheterisation there was a relatively high incidence of recurrent/residual fistula (19%), albeit asymptomatic. Initially dilated coronary arteries either remained dilated for the follow-up period, or were completely thrombosed, with retrograde circulation filling the distal coronaries. Therefore these patients were still at risk of rupture, or embolisation and ischaemia.

Not only does this lack of evidence based information on the natural history of congenital CAF and potential long-term complications of intervention cause problems regarding management; it also makes counselling of parents particularly troublesome; as we experienced. Which parent would like to expose their child to 'heart surgery' when there is the potential for spontaneous resolution of the problem; especially when we are unsure about long-term complications of intervention? Alternatively, how can we quantify the continued level of anxiety experienced by parents watching and waiting for their child to get 'better' from a cardiac condition which has the increasing

potential for significant morbidity and mortality with age, eventually requiring surgery anyway but then with increased associated risks?

So was our patient treated optimally? It would be difficult to argue that he was not, since a year on from intervention he is well, with no evidence of complications. But given that he was asymptomatic we cannot say the fistula would not have spontaneously resolved, regardless of its size. So the pertinent question may instead be whether we could have gotten away without intervening; which we cannot answer with hindsight.

Learning points

- ▶ Although there are an increasing number of case reports of spontaneous closure of congenital coronary artery fistulae, we do not yet have the kind of evidence base required for guidelines regarding management of this rare condition.
- ▶ A significant proportion of young, asymptomatic children with fistula may go on to have spontaneous resolution, but regular follow-up and imaging would be required.
- ▶ Children who present with symptoms and/or complications due to their fistula, or develop them during follow-up, warrant intervention.
- ▶ There is more of a grey area with regard to large asymptomatic fistula where a case by case approach is best; as our case has shown intervention is relatively safe.
- ▶ For better future management and counselling of parents evidence is needed on the natural progression of asymptomatic congenital CAF if no intervention takes place, and long-term follow-up is needed of CAF patients who have intervention to look at potential complications.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Sapin P**, Frantz E, Jain A, *et al*. Coronary artery fistula: an abnormality affecting all age groups. *Medicine (Baltimore)* 1990;**69**:101–13.
2. **Angelini P**. Coronary-to-pulmonary fistulae: what are they? What are their causes? What are their functional consequences? *Tex Heart Inst J* 2000;**27**:327–9.
3. **Xu L**, Xu ZY, Jiang SL, *et al*. Transcatheter closure of coronary artery fistula in children. *Chin Med J* 2010;**123**:822–6.
4. **Chen CC**, Hwang B, Hsiung MC, *et al*. Recognition of coronary arterial fistula by Doppler 2-dimensional echocardiography. *Am J Cardiol* 1984;**53**:392–4.
5. **Sharland GK**, Tynan M, Qureshi SA. Prenatal detection and progression of right coronary artery to right ventricle fistula. *Heart* 1996;**76**:79–81.
6. **Vavuranakis M**, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. *Cathet Cardiovasc Diagn* 1995;**35**:116–20.
7. **Gowda RM**, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. *Int J Cardiol* 2006;**107**:7–10.
8. **Levin DC**, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. *Circulation* 1978;**58**:25–34.
9. **Schumacher G**, Roithmaier A, Lorenz HP, *et al*. Congenital coronary artery fistula in infancy and childhood: diagnostic and therapeutic aspects. *Thorac Cardiovasc Surg* 1997;**45**:287–94.
10. **Daniel TM**, Graham TP, Sabiston DC Jr. Coronary artery-right ventricular fistula with congestive heart failure: surgical coection in the neonatal period. *Surgery* 1970;**67**:985–94.
11. **Liberthson RR**, Sagar K, Berkoben JP, *et al*. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation* 1979;**59**:849–54.
12. **Bauer HH**, Allmendinger PD, Flaherty J, *et al*. Congenital coronary arteriovenous fistula: spontaneous rupture and cardiac tamponade. *Ann Thorac Surg* 1996;**62**:1521–3.
13. **Habermann JH**, Howard ML, Johnson ES. Rupture of the coronary SINUS with hemopericardium. A rare complication of coronary arteriovenous fistula. *Circulation* 1963;**28**:1143–4.
14. **Moro-Serrano C**, Martinez J, Madrid AH, *et al*. Ventricular tachycardia in a patient with congenital coronary arteriovenous fistula. *Am Heart J* 1992;**124**:503–5.
15. **Zenooz NA**, Habibi R, Mammen L, *et al*. Coronary artery fistulas: CT findings. *Radiographics* 2009;**29**:781–9.
16. **McMahon CJ**, Nihil MR, Kovalchin JP, *et al*. Coronary artery fistula. Management and intermediate-term outcome after transcatheter coil occlusion. *Tex Heart Inst J* 2001;**28**:21–5.
17. **Schleich JM**, Rey C, Gewillig M, *et al*. Spontaneous closure of congenital coronary artery fistulas. *Heart* 2001;**85**:E6.
18. **Farooki ZQ**, Nowlen T, Hakimi M, *et al*. Congenital coronary artery fistulae: a review of 18 cases with special emphasis on spontaneous closure. *Pediatr Cardiol* 1993;**14**:208–13.
19. **Aggoun Y**, Bonhoeffer P, Sidi D, *et al*. [Congenital coronary-cardiac fistula in children. Effects of surgical occlusion and percutaneous embolization]. *Arch Mal Coeur Vaiss* 1997;**90**:605–9.
20. **Malci I**, Belina D, Gitter R, *et al*. [Spontaneous closure of fistula between right coronary artery and right ventricle in an infant]. *Lijec Vjesn* 2009;**131**:65–8.
21. **Cheung DL**, Au WK, Cheung HH, *et al*. Coronary artery fistulas: long-term results of surgical correction. *Ann Thorac Surg* 2001;**71**:190–5.

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