Singapore Med J 2007; 48(10) : e262

Coronary artery fistula diagnosed by transthoracic Doppler echocardiography

Chee T S, Tan P J, Koh S K, Jayaram L

ABSTRACT

A 47-year-old Malay woman complained of an episode of shortness of breath after a shower. There was no previous complaint of shortness of breath or chest pain. Physical examination revealed a wide pulse pressure. Blood pressure was 160/66 mmHg, and heart rate was 77/minute and regular. What was initially thought to be a loud pansystolic murmur was heard over the precordium. Electrocardiography showed left ventricular hypertrophy with a volume overload pattern. Transthoracic Doppler echocardiography revealed a right coronary artery-right ventricular fistula, arising from the right coronary artery and draining into the right ventricular cavity.

Keywords: congenital cardiac anomalies, coronary artery fistula, coronary-ventricular fistula, Doppler echocardiography, transthoracic echocardiography

Singapore Med J 2007;48(10):e262–e264

INTRODUCTION

The evolution of the coronary artery network involves complex embryology. In early foetal development, persistence of myocardial sinusoids and the subsequent connection with the endothelial buds that originated from the base of truncus arteriosus forms the basis for abnormal coronary artery fistulae. These variations may be associated with some underlying congenital heart defects. The diagnosis of coronary artery fistula is challenging as its prevalence is low, yet it should be considered in many symptomatic or asymptomatic patients presenting with cardiac murmurs. The diagnosis is usually confirmed on invasive cardiac catheterisation and coronary angiography. Many patients can now be correctly diagnosed with this condition using transthoracic Doppler echocardiography currently available.

CASE REPORT

A 47-year-old Malay woman presented with a single episode of breathlessness after her shower. She gave a previous history of questionable "hole in the heart" when she was young. There was no previous echocardiography or definitive diagnosis made, neither was there any regular follow-up. There was a history of recent cough and upper respiratory tract infection. She did not complain of any chest pain or palpitations. Clinical examination revealed normal heart sounds. What was initially thought to be a loud pansystolic murmur was heard over the precordium. Heart rate was 77/min and regular, and blood pressure was 160/66 mmHg. Her lungs were clear and there was no elevated jugular venous pressure. Other systems were unremarkable. Electrocardiography (ECG) showed left ventricular hypertrophy (LVH) with a diastolic volume overload pattern (i.e. LVH by voltage criteria, with horizontal ST and upright T waves. There was no ST strain pattern. A complete standard two-dimensional (2D) echocardiography, Doppler interrogation and colour flow imaging were performed. 2D echocardiography showed a markedly dilated right coronary artery (RCA) (Figs. 1 & 2) with a turbulent and abnormal flow within the RCA. This turbulent jet started from RCA, traversing anterior to the right ventricular (RV) outflow tract and finally drained into the RV cavity. The course was clearly

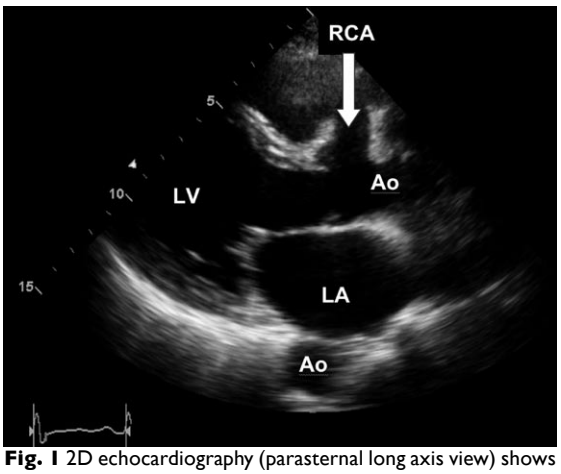

Fig. 1 2D echocardiography (parasternal long axis view) shows a markedly dilated RCA (white arrow). LA: left atrium; LV: left ventricle; Ao: aorta; RCA: right coronary artery.

Division of Cardiology, Changi General Hospital, 2 Simei Street 3, Singapore 529889

Chee TS, MMed, FAMS, FASE
Visiting Consultant

Tan PJ, ARDMS
Senior Cardiac Technologist

Koh SK, ARDMS
Senior Cardiac Technologist

Jayaram L, MMed, FRCP, FACC
Senior Consultant and Head

Correspondence to:
Dr Chee Tek Siong
Chee Heart Specialists Clinic, East Shore Medical Centre, Joo Chiat Place, Singapore 427989
Tel: (65) 6447 0919
Fax: (65) 6447 3430
Email : tschee@singnet.com.sg

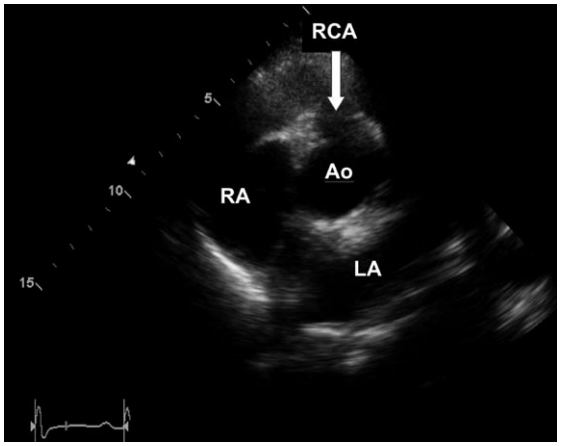

Fig. 2 2D echocardiography (parasternal short axis view) shows a marked dilated RCA (white arrow). LA: left atrium; LV: left ventricle; Ao: aorta; RCA: right coronary artery.

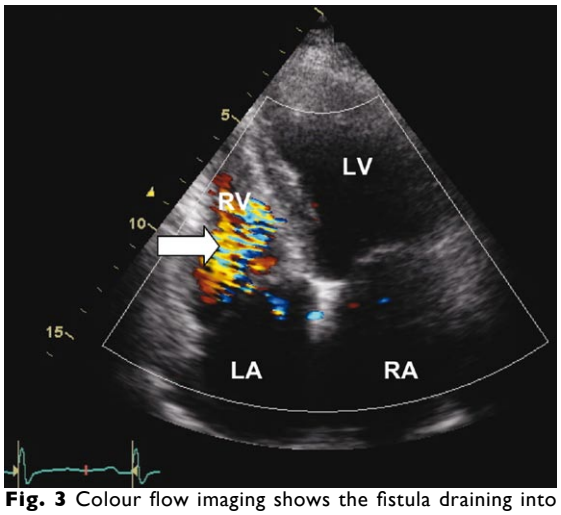

Fig. 3 Colour flow imaging shows the fistula draining into RV cavity (white arrow). LA: left atrium; RA: right atrium; LV: left ventricle; RV: right ventricle.

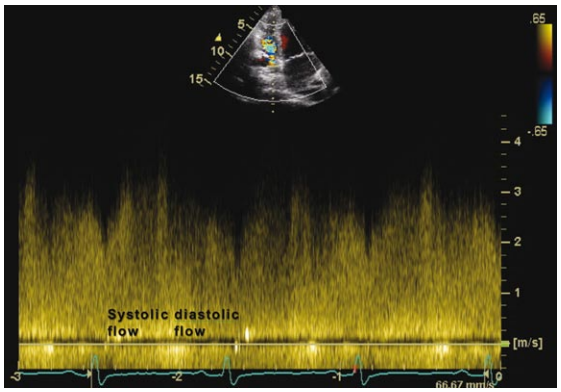

Fig. 4 Doppler echocardiography confirms the continuous flow from the fistula into the right ventricular cavity.

demonstrated by colour flow imaging (Fig. 3). Doppler interrogation demonstrated a continuous flow signal (Fig 4). These 2D, Doppler and colour flow echocardiographical features were consistent with the diagnosis of RCA-RV fistula. Subsequently, the patient refused further evaluation, treatment and follow-up, because she felt comfortable and well.

DISCUSSION

Coronary artery fistula is an uncommon clinical entity. Its incidence in selected series ranges from 0.26% to 0.40% of congenital cardiac anomalies.[1,2] The aetiology may be congenital or traumatic (such as stab or projectile injuries, post-coronary angioplasty), but more typically, it arises as a congenital anomaly. It may occur as an isolated abnormality in an otherwise structurally-normal heart or in association with congenital outflow obstruction. Multiple fistulous communications to cardiac chambers are a rare anomaly.[3] Many adults are asymptomatic if the fistulae are small. Some present with symptoms of fatigue, dyspnoea, angina (due to "steal" phenomenon), atrial arrhythmia, signs of congestive heart failure, pulmonary hypertension or infective endocarditis.[4] Our patient presented with an episode of breathlessness, but whether this was directly related to the fistula was not fully understood because she refused further evaluation. It remained uncertain whether her breathlessness was due to transient myocardial ischaemia from the 'coronary steal' effect. She had no clinical sign of heart failure, nor was there any abnormality in LV systolic function.

A continuous murmur is often present in such patients. Our patient was initially diagnosed to have a pansystolic murmur by a senior consultant. This auscultatory mistake was made because the systolic component of the continuous murmur was loud and the softer and higher pitch diastolic component was missed. Coronary artery fistula may also be mistaken as patent ductus arteriosus. Our patient demonstrated a wide pulse pressure (BP 160/66 mmHg). The differential diagnoses include significant aortic regurgitation, patent ductus arteriosus, or various hyperdynamic states. Transthoracic echocardiography by an experienced echocardiographer is an ideal first-line definitive diagnostic tool in the evaluation and diagnosis of such patients.

About 50% of the coronary artery fistulae arise from the RCA. Majority of these terminate in the right side of the heart. The most frequent sites of termination, in descending order, are the right ventricle, right atrium, pulmonary artery, coronary sinus, left atrium, left ventricle or superior vena cava.[5] Our patient demonstrated the common variety of

coronary artery fistula. A dilated RCA, continuous turbulent flow from RCA into the RV cavity and the absence of other causes of systolic-diastolic flow clinches the diagnosis of RCA to RV coronary fistula. Traditionally, coronary artery fistula has been diagnosed by invasive investigations such as coronary angiography. Recently, non-invasive methods such as comprehensive transthoracic echocardiography, computed tomography coronary angiography and magnetic resonance imaging, have been employed. Transthoracic echocardiography, supplemented by Doppler and colour flow imaging, are non-invasive, portable, easily available and fairly accurate in such circumstances. Moreover, the result of the test will be known immediately. Cardiac function and any associated cardiac lesions can also be directly visualised. Repeated studies during follow-up can also be easily and non-invasively performed.

Advances in echocardiographical technology over the past ten years have markedly improved the spatial and temporal resolution. We are now able to visualise and interrogate blood flow in proximal and mid portions of the coronary arteries in the majority of the patients. Septal branches and bypass grafts have been visualised with the modern echocardiographic machines. Our patient had a dilated RCA and Doppler signals of continuous flow. This can be considered pathonomonic for RCA fistula. A similarly-dilated RCA present in Kawasaki disease may have a turbulent but not continuous flow on Doppler interrogation. These simple echocardiographical views can be easily and rapidly obtained with the transthoracic echocardiogram. Our patient's echocardiographic study was so characteristic and informative that other tests, including coronary angiography, became redundant.

The natural history of the disease is that the larger fistulae will dilate over time, with increasing risk of thrombosis, endocarditis or rupture. The LV or RV may also progressively dilate with subsequent development of heart failure. Fistula flow may jeopardise myocardial perfusion due to coronary "steal" effect, resulting in significant myocardial ischaemia.[6] Therefore, larger fistulae should be treated with transcatheter embolisation or surgical repair. The coronary artery fistula in our patient was of considerable size. There was already LV dilatation, although the systolic function remained normal. Our patient chose not to undergo further evaluation or follow-up. Her LV dilatation could progress over time. She might require percutaneous or surgical treatment when she becomes symptomatic. Small coronary artery fistula, on the other hand, is generally well-tolerated and should anticipate no significant restriction on daily routine and activities. However, antibiotic prophylaxis to prevent infective endocarditis prior to dental or surgical procedures is still advisable.

REFERENCES

1. Gillebert C, Van Hoof R, Van de Werf F, Piessens J, De Geest H. Coronary artery fistulas in an adult population. Eur Heart J 1986; 7:437-42.
2. Reeder GS, Tajik AJ, Smith HC. Visualization of coronary artery fistula by two-dimensional echocardiography. Mayo Clin Proc 1980; 55:185-9.
3. Black IW, Loo CKC, Allan RM. Multiple coronary artery-left ventricular fistulae: clinical, angiographic and pathologic findings. Cathet Cardiovasc Diagn 1991; 23:133-5.
4. Kasravi B, Reid CL, Allen BJ. Coronary artery fistula presenting as bacterial endocarditis. J Am Soc Echocardiogr 2004; 17:1315-6.
5. Lin FC, Chang HJ, Chern MS, et al. Multiplane transesophageal echocardiography in the diagnosis of congenital coronary artery fistula. Am Heart J 1995; 130:1236-44.
6. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. Circulation 1979; 59:849-54.