JACC: CASE REPORTS VOL. 2, NO. 5, 2020

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

INTERMEDIATE

CASE REPORT: CLINICAL CASE

TGA With Interarterial Course and Athletes Heart

A Diagnostic Dilemma Answered by Stress CMR

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ABSTRACT

We report a case of coronary artery compression and athlete's heart in a patient with a history of transposition of the great arteries. We present the diagnostic dilemmas and demonstrate the use of cardiac magnetic resonance imaging and cycle-ergometer stress cardiac magnetic resonance in the management of our patient. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:740–4) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 20-year-old Chinese male was referred to our adult congenital heart disease clinic for routine follow-up. He led an active lifestyle reporting an exercise duration of >12 h/week, running half-marathons and playing competitive tennis at an amateur level. Physical examination revealed a pansystolic murmur radiating to the left axilla.

LEARNING OBJECTIVES

- To make a differential diagnoses in the evaluation of abnormal cardiac size and function in a D-transposition of great arteries post-ASO patient.
- To understand the role of different imaging modalities, and in particular, the role of CMR in the workup of patients with surgically corrected transposition of the great arteries.
- To recognize features of athlete's heart and its overlap with ischemic and dilated cardiomyopathy.

MEDICAL HISTORY

He had a history of D-transposition of the great arteries and underwent a balloon atrial septostomy on day 1 of life, followed by arterial switch operation (ASO) or the Jatene procedure using the Lecompte technique on day 3 of life.

INVESTIGATIONS

Electrocardiogram showed a normal sinus rhythm with left ventricular hypertrophy by voltage criteria. A transthoracic echocardiogram (TTE) showed biventricular dilatation (left ventricle end-diastolic diameter [LVEDD] = 66 mm; indexed LVEDD [iLVEDD] = 38 mm/m^2, right ventricular basal diameter = 43 mm) with eccentric hypertrophy (LV mass = 206 g; LV mass index = 118 g/m^2) and mildly reduced biventricular systolic function (left ventricular ejection fraction (LVEF) = 52%, tricuspid annular plane systolic excursion = 13 mm) with wall motion abnormalities of the septum and

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

Manuscript received June 20, 2019; revised manuscript received February 26, 2020, accepted March 27, 2020.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.03.013

anteroseptal segments and normal diastolic function (Table 1). There was also a prolapsed anterior mitral leaflet with moderate mitral regurgitation (vena contracta = 5 mm).

Because of the concerns of ischemic cardiomyopathy, a computed tomography coronary angiogram was performed and showed a single coronary button at the right coronary cusp, giving rise to both left and right coronary arteries. The left main coronary artery (LMCA) had a compressed inter-arterial course, lying anterior to the ascending aorta and posterior to the pulmonary trunk (Figure 1). A review of the patient's surgical records showed that because of their proximity with each other, the left and right coronary buttons were unable to be dissected and instead the surgeon implanted both at the right coronary cusp.

DIFFERENTIAL DIAGNOSES

Differential diagnosis for the clinical and imaging findings included: 1) ischemic cardiomyopathy secondary to the LMCA compression; 2) dilated cardiomyopathy; 3) underestimation of MR; or 4) athlete's heart.

MANAGEMENT

The case was discussed at the center's cardiovascular case conference involving congenital cardiothoracic surgeons. The following options were considered: 1) stress imaging with echocardiogram or myocardial perfusion imaging; 2) invasive coronary angiogram; 3) transesophageal echocardiogram for evaluation of MR; and 4) CMR with cycle ergometer stress testing.

A bicycle ergometer-stress CMR (1.5-T, Siemens MAGNETOM Aera) with intravenous gadolinium (Gadavist) contrast was subsequently performed. It confirmed a moderately dilated left ventricle and right ventricle (iLVEDD = 147 ml/m^2 and right ventricular end-diastolic volume index = 132 ml/m^2) with an LVEF of 53%. There were no wall motion abnormalities at rest and stress. The mitral regurgitation was confirmed to be mild with a regurgitation fraction of 7%. On late gadolinium enhancement (LGE) imaging, there was no evidence of infarction, infiltration, or fibrosis. Using T1-modified Look-Locker Inversion recovery sequences, the basal precontrast myocardial T1 (967 ms) and extracellular volume (ECV) (22%) were both normal (Figure 2).

With ergometer stress, the cardiac index increased from 6 to 12.8 l/min/m^2 at peak stress. This was compared with a cohort of individuals taken from our local center and his peak stress cardiac index corresponded to the 95th percentile when matched for age and gender. The patient was diagnosed with athlete's heart. Because of surgical considerations, our center's surgeons deemed the surgical risks to outweigh its benefits. These considerations were shared with the patient and a joint decision was made to avoid strenuous exercise. After detraining, a repeat echocardiogram in 6 months showed an LVEF of 53%, with a reduction in LV size (LVEDD = 60 mm; iLVEDD = 35 mm/m^2) and LV mass (mass = 171 g; LV mass index = 98 g/m^2).

DISCUSSION

This case demonstrates the complexities of surgically corrected complex congenital heart disease and the role of CMR in their management. The ASO or Jatene procedure, has become the preferred approach for repair of transposition of great arteries (TGA) with intact ventricular septum since its introduction in the 1980s and is recommended to be performed in the first 3 weeks of life (Class I, Level of Evidence: B) (1). The Lecompte technique involves placing the pulmonary artery bifurcation in front of the ascending aorta and is the current routine surgical technique for ASO. It has excellent clinical results; however, complications include stenosis at arterial anastomotic sites, aortic root dilatation and regurgitation, and coronary obstruction. The 2018 American College of Cardiology/American Heart Association guidelines recommend (Class I, Level of Evidence: C) baseline and serial imaging with CMR as an alternative to echocardiography in a patient with TGA with arterial switch. It has the added benefit of other recommended evaluations like coronary artery patency and physiological tests of myocardial perfusion (2).

TABLE 1 TTE Summary

Parameter	Result
LVEF	52% with RWMA
LVEDD	66 mm
Indexed LVEDD to BSA	38 mm/m^2
LV mass	206 g
LVMI	118 g/m^2
Regional wall thickness	0.24
MR vena contracta	5 mm
Effective regurgitant orifice area	0.2 cm^2
Conclusions	Dilated left ventricle with low-normal LV systolic function and RWMA. Moderate MR

Summary of the patient's TTE, which showed a moderately dilated left ventricle, with mildly reduced LVEF and moderate MR.

LVEDD = left ventricular end-diastolic diameter; LVEF = left ventricular ejection fraction; LVMI = left ventricular mass index; MR = mitral regurgitation; RWMA = regional wall motion abnormality; TTE = transthoracic echocardiogram.

ABBREVIATIONS AND ACRONYMS

ASO = arterial switch operation

DCM = dilated cardiomyopathy

ECV = extracellular volume

iLVEDD = indexed left ventricular end-diastolic diameter

LGE = late gadolinium enhancement

LMCA = left main coronary artery

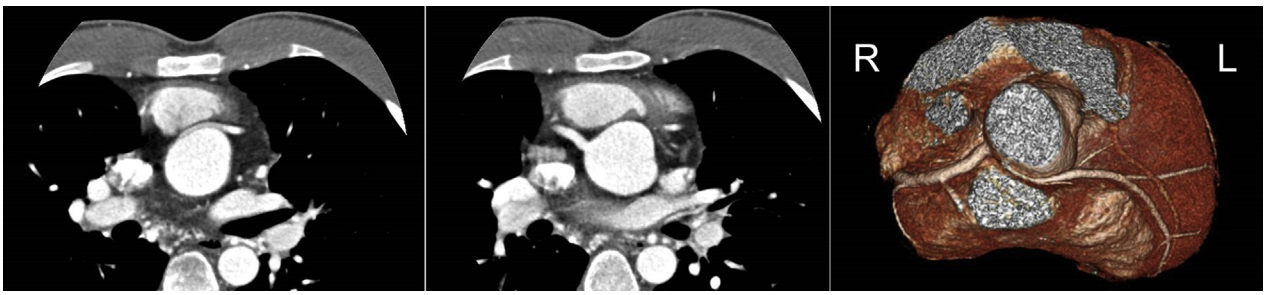
LVEDD = left ventricular end-diastolic diameter

LVEF = left ventricle ejection fraction

TGA = transposition of great arteries

TTE = transthoracic echocardiogram

FIGURE 1 Patient's CTCA



Computed tomography angiography showed the LMCA button arising from the right-sided cusp, with an inter-arterial course between the pulmonary trunk and ascending aorta in cross-sectional **(left)** and 3-dimensional composite imaging **(right)**. **(Center)** Normal right coronary artery button. CTCA = computed tomography coronary angiogram; LMCA = left main coronary artery.

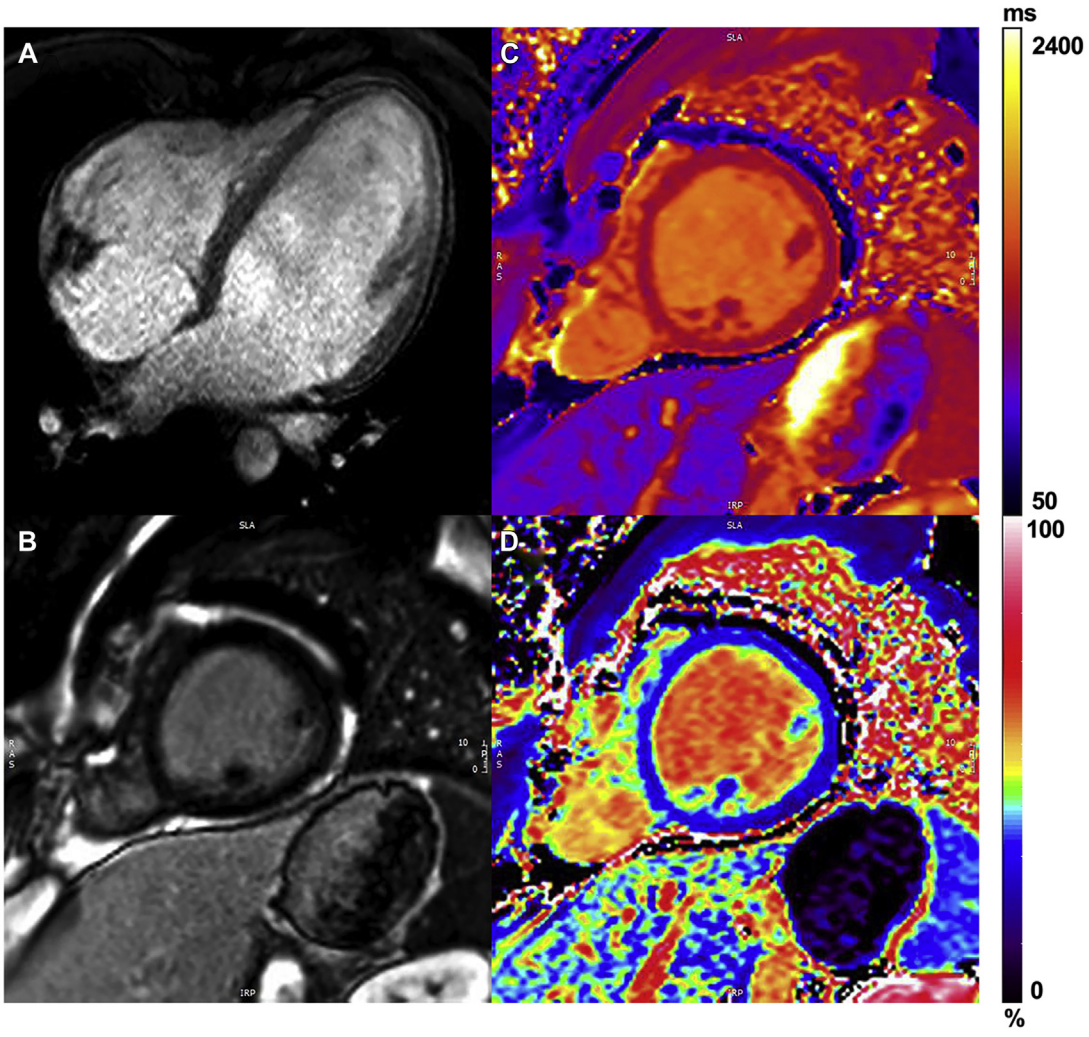
In this case, the left coronary and right coronary arteries were closely adjacent and the surgeon was unable to translocate the coronary artery buttons separately. The single coronary button was reimplanted onto the right coronary cusp. This led to an inter-arterial course of the left coronary artery (**Figure 1**). In this case, where the inter-arterial course is iatrogenic and after ASO, it is less certain whether the prognosis is similar to uncorrected congenital anomalous coronary arteries. It is still, however, prone to compromised blood flow resulting from scarring and angulation. There is no guideline that addresses coronary anomalies post-ASO, but in anomalous coronary arteries, the 2018 American College of Cardiology/American Heart Association guidelines recommend a symptom-guided approach, with surgical correction in patients with left coronary arteries from the right sinus as a Class I, Level of Evidence: A recommendation when ischemia is proven on diagnostic testing, and Class II, Level of Evidence: A if there is none (2). In post-ASO cases, we recommend a prudent approach involving multidisciplinary teams when making clinical decisions on coronary reimplantation surgery.

The presence of ischemia was a key diagnostic question in our assessment. CMR can assess for ischemia in a combination of perfusion and wall motion imaging. It is also important that clinicians be cognizant of the limitations of stress CMR, especially in young patients with excellent cardiac reserve. In such cases where the clinical scenario is complex and does not fit in the clinical guidelines, we recommend a prudent approach involving multidisciplinary teams.

The last challenge was in the differentiation of dilated cardiomyopathy from athlete's heart. Dilated cardiomyopathy (DCM) is characterized by dilated LVEF in the absence of ischemia, valvular disease, or hypertension (3). Athlete's heart may also have features that overlap with DCM, with chamber dilatation resulting from repetitive increased stroke volumes and pressure overload from exercise and a resulting lower resting EF (4). A study on elite cyclists showed an average left ventricle internal diameter of 60.1 ± 3.9 mm with LVEF <60% in approximately 40% of participants (5). Because of the overlapping features of DCM and athlete's heart, the diagnosis may be difficult to ascertain. Key features in differentiating the 2 conditions are a normal diastolic function and an appropriate hyperdynamic cardiac response to exercise in athlete's heart, which patients with DCM are rarely able to achieve (3,4). Stress-imaging modalities like echocardiography and myocardial perfusion imaging would be able to assess for these features. Although echocardiography is the standard modality to assess diastolic function, this can also be assessed via CMR. CMR, however, may have a growing role as the imaging modality of choice for patients. CMR has the added benefit of LGE assessing for fibrosis, a feature that may be present in DCM but not seen in athlete's heart. T1 and ECV mapping may also have an additional role, especially in cases where the diagnosis is uncertain. Higher native T1, ECV, and T2 relaxation times have also been reported in patients with DCM compared with controls and athletes, with native T1 showing to be the best discriminator for both conditions (3).

In our case, the patient had no increased uptake on LGE. Additionally, he showed a hyperdynamic response to exercise with no wall motion abnormalities, and T1 and ECV mapping values were normal.

FIGURE 2 **Composite Image of Patient CMR**



(A) The CMR confirmed biventricular dilatation as shown on the apical 4-chamber view. **(B)** LGE imaging with no myocardial enhancement along with **(C)** normal T1 and **(D)** ECV value precludes DCM and ischemia as the underlying pathology. CMR = cardiac magnetic resonance; DCM = dilated cardiomyopathy; ECV = extracellular volume; LGE = late gadolinium enhancement.

This rules out ischemic cardiomyopathy and DCM, giving the diagnosis of athlete's heart. The effects of detraining in our patient (i.e., reduction in LV mass with no change in LVEF) were also consistent with findings in another study (6).

FOLLOW-UP

After detraining, a repeat echocardiogram showed a normal LV systolic function with a reduction in LV size. Because of considerations mentioned previously, a strategy of watchful waiting with advice of strictly avoiding vigorous physical activities has been used.

CONCLUSIONS

The multiple diagnostic considerations in this case highlight the advantages of ergometer stress CMR as a safe and comprehensive imaging modality in the assessment of a patient with complex congenital heart disease. With a noninvasive test without

radiation exposure, we have demonstrated that the patient has normal cardiac physiology and an adequate response to physical stress, clinching the diagnosis of athlete's heart. CT angiography, with its superior spatial resolution to magnetic resonance angiography, also played a critical role in the diagnostic assessment. To our knowledge, this is the first reported case of cycle ergometer stress CMR used in this setting.

ACKNOWLEDGMENTS The authors acknowledge the support of the contributing authors and the National Heart Centre, Singapore, without which, this case report would not be possible.

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KEY WORDS athlete's heart, cardiac magnetic resonance, stress CMR, transposition of great arteries