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CASE REPORT
Congenital heart disease

Exercise cardiac magnetic resonance imaging to assess dynamic right ventricular outflow tract obstruction in congenital heart disease: a case report

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Background Right ventricular outflow tract obstruction in patients with congenital heart disease is usually assessed using echocardiographic peak instantaneous gradient at rest. Since right ventricular outflow tract obstruction may change during exercise (dynamic right ventricular outflow tract obstruction), we present a case emphasizing the potential use of exercise cardiac magnetic resonance imaging (CMR).

Case summary We discuss a 15-year-old patient with repaired mid-ventricular sub-pulmonary stenosis type double-chambered right ventricle causing right ventricular outflow tract obstruction and symptoms on exertion. In this case, exercise CMR imaging provided additional information, allowing adequate surgical planning.

Discussion The additional value of exercise CMR imaging in a case of right ventricular outflow tract obstruction was described. Although exercise cardiac magnetic resonance imaging did not show a significant increase in peak gradient across the right ventricular outflow tract obstruction, shifting and D-shaping of the interventricular septum with subsequent insufficient left ventricular filling (preload) was observed in the patient with recurrent double-chambered right ventricle. This case demonstrates how exercise CMR imaging can be helpful in the clinical decision beyond standard echocardiographic evaluation by providing additional evidence of adverse haemodynamics during exercise.

Keywords Pressure loaded ventricle • Right ventricular outflow tract • Obstruction • Congenital heart disease • Double-chambered right ventricle • Cardiac magnetic resonance imaging • Case report

Learning points

- Exercise cardiac magnetic resonance (CMR) has the potential to evaluate physiological circumstances during exercise and to correlate symptoms with dynamic right ventricular outflow tract obstruction (RVOTO).
- Haemodynamic assessment during exercise is as important as increase in peak gradient in dynamic RVOTO.
- Exercise CMR may be used in clinical decision to opt in or out surgery, especially in borderline cases.

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Introduction

Right ventricular outflow tract obstruction (RVOTO) in congenital heart disease (CHD) is assessed using non-invasive echocardiographic peak instantaneous gradient (PIG), stress echocardiography or invasive peak-to-peak gradient on cardiac catheterization. Since symptoms often occur during exercise and the degree of RVOTO may vary during exercise (dynamic RVOTO), we present a case emphasizing the potential use of exercise cardiac magnetic resonance (exCMR) imaging.[1,2]

Double-chambered right ventricle (DCRV) is characterized by mid-ventricular sub-pulmonary stenosis (subPS) due to aberrant hypertrophied septomarginal trabeculations or an abnormal moderator band.[3] Shortness of breath on exertion is a common symptom of RVOTO, due to the dynamic nature of RVOTO and secondary haemodynamic effects during exercise. According to the European Society of Cardiology (ESC) guidelines, indications for surgical repair for symptomatic patients with RVOTO are PIG >64 mmHg (class I/C recommendation) provided that right ventricle (RV) function is normal and no valve substitute is required.[4,5] Interventions in patients with gradient <64 mmHg should be considered when DCRV is present (class IIa recommendation).[5] Given its high temporal and spatial resolution, exCMR could be a useful diagnostic. If technically feasible, exercise testing is more physiological and, in most cases, preferred over pharmacological CMR. We used an exCMR protocol with an in-scanner cardiac magnetic resonance (CMR) compatible supine ergometer.

Timeline

0–1 year	Diagnosed at birth with a ventricular septal defect (VSD) and a mild mid-ventricular sub-pulmonary stenosis type double-chambered right ventricle causing a right ventricular outflow tract obstruction (RVOTO) (PIG 25 mmHg at rest on transthoracic echocardiography).
1–2 years	Progression to severe RVOTO (PIG 98 mmHg), spontaneous VSD closure—surgical resection of the muscular band.
7 years	A symptomatic residual RVOTO PIG 26 mmHg.
11 years	Asymptomatic RVOTO PIG 41 mmHg at rest.
15 years—August 2019	Shortness of breath on exertion with an RVOTO of 44 PIG mmHg at rest.
December 2019	ExCMR showed significant shifting of the interventricular septum and D-shaping of the left ventricle during exercise.
December 2019	Resection of muscular band. Short period of ventricular arrhythmia which stopped after 24 h. The patient was discharged from the hospital with an eventless clinical course.
February 2020	Latest follow-up the patient was asymptomatic and showed no signs of RVOTO post-operative on transthoracic echocardiography.

Case presentation

A 15-year-old boy presented with shortness of breath on exertion [New York Heart Association functional classification (NYHA) II], without syncope. His medical history included perimembranous VSD at birth that closed spontaneously at the age of 2 years and right orchidopexy at the age of 11 years old, no other medical history. From the age of 3 months, he was followed for progressive mild to severe mid-ventricular subPS resulting in DCRV (maximal PIG gradient of 98 mmHg) and first resection took place at the age of 2 years. Post-operative PIG gradient over the right ventricular outflow (RVOT) was 68 mmHg and regressed over the years to 26 mmHg at rest. At the age of 11 years, maximal systolic gradient was 41 mmHg whilst asymptomatic.

Physical examination showed a harsh systolic murmur parasternal left. His electrocardiogram displayed sinus rhythm, right bundle branch block, and right axis. Doppler-echocardiography showed a turbulent flow over the RVOT with a PIG of 44 mmHg at rest. There were no signs of right-sided heart failure. Bicycle stress-test electrocardiogram revealed no ischaemia nor arrhythmias. Since the patient was symptomatic on exertion despite no significant increase in PIG at rest (+3 mmHg over 4 years), exCMR was performed using Philips Achieva 1.5-T CMR real-time sequencing. The patient did not take any medication at time of exCMR. Anatomical obstruction was confirmed showing a muscular band (19 × 6 mm, 1.2 cm^2) at the RVOT with a PIG of 40 mmHg at rest (see *Figure 1*). The RV was dilated [RV end-diastolic volume (RVEDV) 224 ml, RV end-systolic volume (RVESV) 147 mL] and hypertrophied with decreased RV systolic function, ejection fraction (EF) of 34%. The volumes of the left ventricle (LV) were within normal limits with an EF of 50%. The cardiac output in rest was 4.2 L/min.

During exercise, there was an increase in RVEDV (+16%) and RVESV (+18%), with no contractile reserve of the RV (ΔRVEF -1%). For the LV, a decrease in LV end-diastolic volume (LVEDV) (-9%) and LV end-systolic volume (LVESV) (-36%) was noticed but with a normal contractile reserve (ΔLVEF +15%). There was an equal rise of stroke volume (SV) both right and left in the first stage of the exercise followed by a decrease of SV with increased exercise load. The shifting of the interventricular septum (IVS) and D-shaping of the LV during exercise was remarkable (see *Video 1*). These haemodynamic changes during exercise could explain the symptoms and the patient was referred for surgical re-intervention. The patient had a redo resection of the muscular band at the crista supraventricularis. Post-operative during his stay at ICU, there was a short period of ventricular extrasystoles in bigeminy and betablockade was started. The patient became asymptomatic (NYHA I) and transthoracic echocardiography showed a residual peak gradient of 20 mmHg on discharge. The patient was discharged from the hospital with an eventless clinical course. At follow-up consultation, the patient remained asymptomatic without signs of residual obstruction of the RVOT (no D-shaping of the LV, no turbulent flow over the RVOT) with normal function of the RV on transthoracic echocardiography.

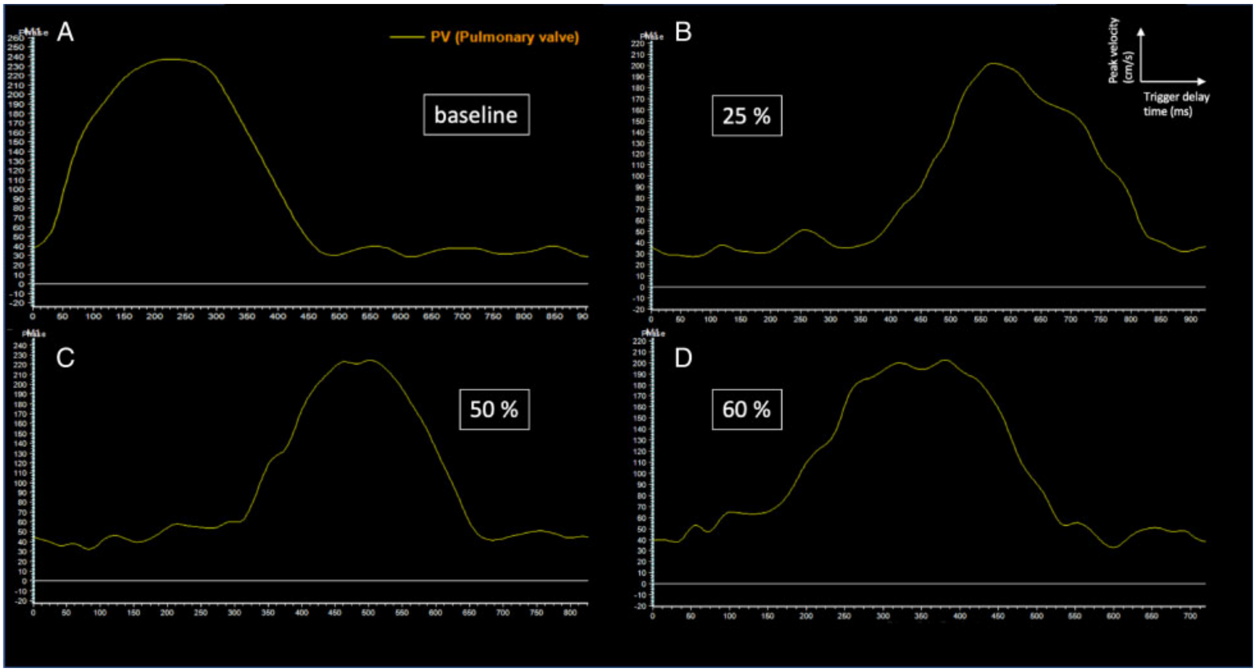
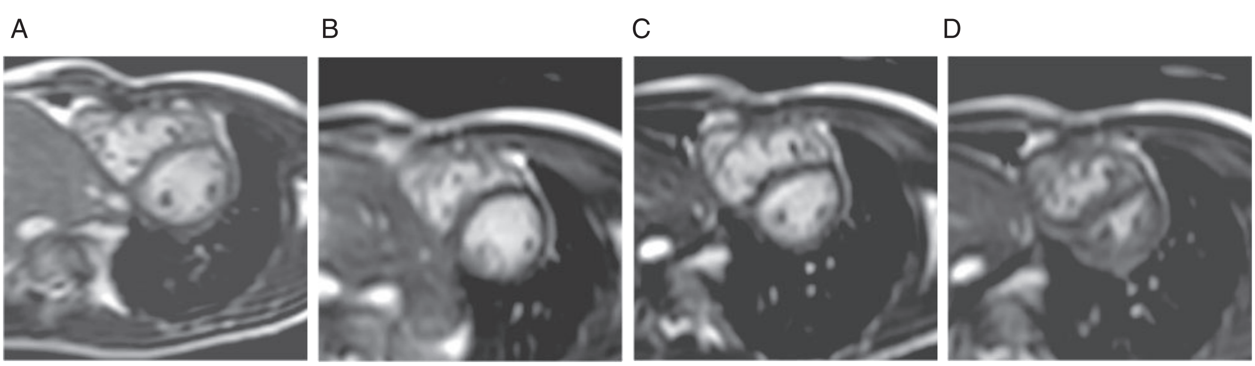


Figure 1 During exercise, there is no significant increase of peak gradient over right ventricular outflow tract. Baseline (*A*) and three stages of exercise are displayed. Initial workload was 25% of maximal capacity (*B*) followed by 50% (*C*) and 66% (*D*) of maximal capacity.



Video 1 Short-axis view during exercise CMR imaging showing shifting of the interventricular septum and D-shaping of the left ventricle during exercise. Baseline (*A*) and three stages of exercise are displayed. Initial workload was 25% of maximal capacity (*B*) followed by 50 (*C*) and 66% (*D*) of maximal capacity.

Discussion

We evaluated a pressure overloaded RV and the importance of exCMR for clinical decision-making. During exercise, we noticed an insufficient filling of the LV with shifting of the IVS and D-shaping of the LV during exercise due to (i) pressure overload caused by the muscular band, (ii) volume load secondary to the increase of systemic venous return during exercise, and (iii) inadequate preload of LV.

The diagnosis of the anatomical cause of RVOT in CHD is challenging because of difficulties in evaluating the RVOT and high-quality multimodality imaging is essential in this population.[6] CMR is gold standard in CHD for evaluating cardiac anatomy and function in rest.[7,8] According to the ESC guidelines, CMR is recommended in addition to echocardiography for evaluating the RVOT.[5,9] Although cardiovascular dysfunction is not always present at rest, stress testing may be required to unmask disease. In contrast to pharmacological testing, exercise stress testing is more physiological as exercise stress testing also induces the preload of the ventricle.

When real-time-ungated CMR is combined with *post-hoc* analysis incorporating compensation for respiratory motion, highly reproducible and accurate biventricular volumes can be measured during maximal exercise.[1,10] Therefore, real-time-ungated CMR represents a new gold standard in imaging during exercise.

In this case, at the age of 11 years, the patient had an asymptomatic RVOT gradient of 41 mmHg but 4 years later, he was symptomatic without a significant increase in gradient. According to the ESC guidelines, intervention in patients with gradient <64 mmHg should be considered in DCRV (class IIa recommendation).[5]

During exCMR, there was no significant increase of PIG but with an adequate increase of RVEDV and RVESV yet without sufficient RV performance. Notably, LVEDV decreased at 66% of maximal capacity which is compatible with inadequate preload to the LV, causing compromised LV filling with shifting of IVS and D-shaping of the LV during exercise. So, poor haemodynamics during exercise were observed. As a consequence, surgery was indicated. Note, PIG can be underestimated on exCMR since phase-contrast flow velocity to measure the PIG was used and flow velocity was measured during breath-holding phase of exCMR.

Thus, exCMR has an important place in RV imaging especially since RV evaluation is difficult with other imaging techniques during exercise. (Stress-) echocardiography will remain first-line examination but is limited by two-dimensional assessment of the RV and PIG on Doppler is overestimated.[11] This case demonstrates how multimodality imaging and exCMR and can be helpful in the clinical decision taking. Furthermore, exCMR is also able to assess wall motion abnormalities, perfusion defects, exercise capacity, and viability in a single examination. This holds important clinical potential in a variety of cardiovascular conditions for patients with CHD.[12]

Lead author biography

Béatrice Santens was born in South Carolina in 1992 and became a resident in internal medicine in 2016. She recently started as a resident in cardiology at the University Hospitals Leuven, Belgium. Her main interest lies in the field of congenital heart disease and started as a PhD researcher at the department of congenital and structural heart disease and cardiovascular science at the University Hospitals Leuven, Belgium in 2018. She is an author and co-author of numerous articles within the field of congenital cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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