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VALVULAR HEART DISEASE MINI-FOCUS ISSUE

INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

4D Flow Cardiac Magnetic Resonance Uncovers the Cause of Bioprosthetic Pulmonary Valve Dysfunction



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ABSTRACT

We present a case of a 20-year-old man with tetralogy of Fallot status post-repair with a transannular patch and left pulmonary artery reconstruction at age 1 and subsequent pulmonary valve replacement with bioprosthetic valve 3 years ago. A transthoracic echocardiogram revealed severe bioprosthetic valve insufficiency. Cardiac magnetic resonance with 4-dimensional flow revealed a paravalvular leak. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:485-6) © 2019 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 man was referred to our multidisciplinary adult congenital heart disease (ACHD) program. He had a history of tetralogy of Fallot status post-repair with a transannular patch and left pulmonary artery (LPA) reconstruction at 1 year of age. He underwent placement of an LPA stent with subsequent multiple stent dilatations. He then underwent pulmonary valve replacement with a bioprosthetic valve 3 years before presentation. The patient was referred to the multidisciplinary ACHD program to establish care and for further evaluation. He was symptomatic with effort intolerance. Routine blood work was noncontributory. A transthoracic echocardiogram revealed severe bioprosthetic pulmonary valve insufficiency as well as moderate right ventricular dilatation. He was referred for cardiac magnetic resonance (CMR) with 4-dimensional (4D) flow for further evaluation; 4D flow CMR has been shown previously to be valuable for quantification of mitral and tricuspid valve regurgitation (1), congenital heart disease in general (2), and intracardiac and extracardiac defects not identifiable on other modalities (3). CMR was performed on a 3T Discovery 750 magnet (GE Healthcare, Chicago, Illinois) with an injection of 18 ml Multihance (Bracco Diagnostics, Monroe Township, New Jersey) contrast. Sequences gathered included cine steady-state free precession, first-pass magnetic resonance angiography, and volumetric phase-contrast 4D flow CMR after contrast administration. CMR with 4D flow demonstrated a paravalvular leak along both the anterior and posterior aspects of the bioprosthetic valve (Video 1). 4D flow was also used to quantify the leak at a volume of 63-ml/beat and a regurgitation fraction of 35% (graded as moderate to severe). 4D flow also revealed residual LPA stenosis with a peak velocity of 3 m/s, corresponding to a pressure gradient of 36 mm Hg. Subsequent cardiac computed tomography angiography with 3-dimensional reconstruction (Supplemental Figure 1) was then used to confirm the suspected leaks and further localize the areas of dehiscence, measured as 9.9 × 7.6 mm anteriorly and 11.3 × 7.0 mm posteriorly. Cardiac computed tomography angiography was performed on a Revolution 256 scanner (GE Healthcare) at 80 kv (peak) covering the whole chest and cardiac cycle with prospective cardiac gating and retrospectively

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Informed consent was obtained for this case.

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**ABBREVIATIONS
AND ACRONYMS****4D** = 4-dimensional**ACHD** = adult congenital heart disease**CMR** = cardiac magnetic resonance**LPA** = left pulmonary artery**TOF** = tetralogy of Fallot

gated image reconstruction using 93 ml Omnipaque (GE Healthcare) 350 ml intravenous contrast injected at a rate of 4 ml/s. The total radiation dose of computed tomography was 326.75 mGy·cm, corresponding to an estimated ionizing radiation dose of 4.6 mSv.


4D flow CMR is an emerging technique to visualize and quantify blood flow throughout an imaging volume, performed as a 10-min free-breathing acquisition. This allows quantification of blood flow throughout the cardiac cycle and is increasingly used clinically to evaluate structural heart disease. In this case, it was helpful to pinpoint the etiology of what was previously thought to be simple valvular regurgitation and accurately quantify an effective regurgitant volume for decision making. Although CMR may also be used to quantify regurgitant volume, 4D flow was instrumental to characterize this as a paravalvular leak. This case highlights the emerging and irreplaceable role of advanced cardiac imaging, especially 4D flow, in the workup and management of adults with congenital heart disease. Our group uses this technology routinely for all patients with ACHD referred for CMR. An interventional approach was discussed at the ACHD conference; however, the committee felt that the patient was at low surgical risk for repeat operation, and a percutaneous approach was unlikely to eliminate the leaks in their entirety. Knowing the valve dehiscence allowed the surgeon to plan the surgical approach more effectively by using a homograft rather than a simple bioprosthesis to replace the pulmonary valve.

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KEY WORDS congenital heart defect, pulmonary valve, valve replacement

 **APPENDIX** For a supplemental figure and a video, please see the online version of this paper.