Images in Cardiovascular Medicine

Atriopulmonary Fontan Spontaneous Echo Contrast Improved after Cardioversion

Wilson W. Lam, MD
Mehul B. Patel, MD
Fred H. Rodriguez III, MD
Dhaval R. Parekh, MD
Wayne J. Franklin, MD

A 21-year-old woman with tricuspid atresia who had undergone an atriopulmonary Fontan operation at 3 years of age presented with intra-atrial reentrant tachycardia and tachycardia-induced cardiomyopathy. She had done well on no medications, and systolic function was normal on echocardiograms 3 months before this presentation. Since then, she had experienced exertional palpitations, increasing abdominal girth, loose bowel movements, vague abdominal pain, fatigue, and decreased appetite. Examination revealed a blood pressure of 120/70 mmHg, a regular tachycardic pulse, clear lungs, a pulse oximetry saturation of 97%, and a distended abdomen with ascites and hepatomegaly. Laboratory tests showed elevated alanine transaminase of 115 IU/L, brain natriuretic peptide of 556 pg/mL, an international normalized ratio of 1.4, negative stool antitrypsin (no protein-losing enteropathy), and normal renal function, lactate, and cardiac enzyme levels. An electrocardiogram showed flutter waves and 2:1 atrioventricular conduction (ventricular rate, 122 beats/min) (Fig. 1). A transthoracic echocardiogram revealed a dilated left ventricle with severely depressed systolic function, an ejection fraction of 0.25, and spontaneous echo contrast in the dilated inferior vena cava. A transesophageal echocardiogram showed no left atrial appendage thrombus; however, dense contrast was present in the dilated right atrium of the Fontan circuit (Fig. 2).

We presumed that tachycardia-induced cardiomyopathy was exacerbating the patient's Fontan circuit stasis, so we recommended terminating the tachycardia. Cardio-

Section Editor:
Raymond F. Stainback, MD, Department of Adult Cardiology, Texas Heart Institute at St. Luke's Hospital, 6624 Fannin St., Suite 2480, Houston, TX 77030

From: *Department of Pediatrics, Section of Cardiology, Texas Children's Hospital; Department of Pediatric Cardiology, Texas Heart Institute; and Departments of Internal Medicine and Pediatrics, Section of Cardiology, Baylor College of Medicine; Houston, Texas 77030*

Dr. Rodriguez is now at Sibley Heart Center Cardiology and Department of Pediatrics, Division of Cardiology, Emory University School of Medicine, Atlanta, Georgia.

Address for reprints:
Wilson W. Lam, MD, 6621 Fannin St., MC 19345-C, Houston, TX 77030

E-mail: *wlam@bcm.edu*

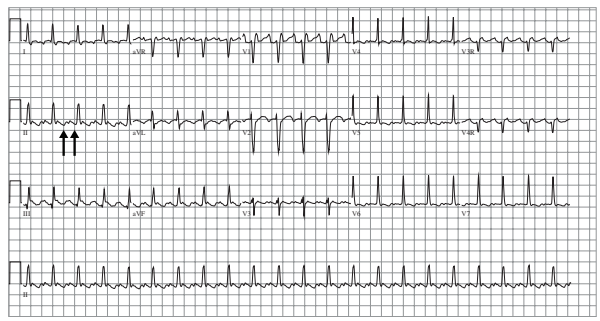

Fig. 1 *A 12-lead electrocardiogram shows intra-atrial reentrant tachycardia with 2:1 atrioventricular block; arrows point to flutter waves.*

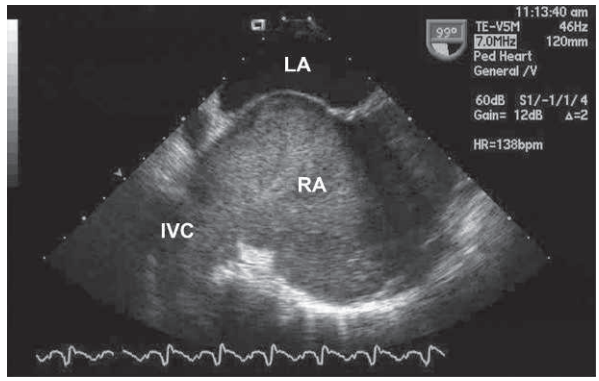

Fig. 2 *Transesophageal echocardiogram (bicaval view) shows 2:1 intra-atrial reentrant tachycardia with dense spontaneous echo contrast.*

IVC = inferior vena cava; LA = left atrium; RA = right atrium/atriopulmonary Fontan circuit to pulmonary arteries

Supplemental motion image is available for Figure 2.

version yielded improvement (Fig. 3); however, contrast was still seen on cardiac magnetic resonance images (Figs. 4 and 5). The patient was placed on amiodarone, anticoagulation, and anticongestive therapy. An echocardiogram 4 months later showed improvement; the patient had mildly depressed systolic function with maintained sinus rhythm.

Comment

Usually 1 to 2 decades after initial palliation, the Fontan circuit is at risk for diminished cardiac output from multiple causes, including ventricular dysfunction, arrhythmia, anatomic obstruction, or valvular regurgitation. Elevated cardiac pressures can lead to elevated Fontan pressures that present clinically as ascites, protein-losing enteropathy, desaturation, and worsening arrhythmias. This hypercoagulable state can lead to further thrombosis.[1] Escalating levels of antiarrhythmic therapies include medications, electrophysiologic study with catheter ablation, or a maze procedure as part of Fontan conversion to the more modern lateral tunnel or extracardiac conduit system.[2] This operation should include placement of an epicardial pacing system, which can help to prevent or abort atrial arrhythmias.[3] In this complex congenital case, expedient recognition and management occurred in the setting of a collaborative team in a complex congenital heart center, as is recommended.[4]

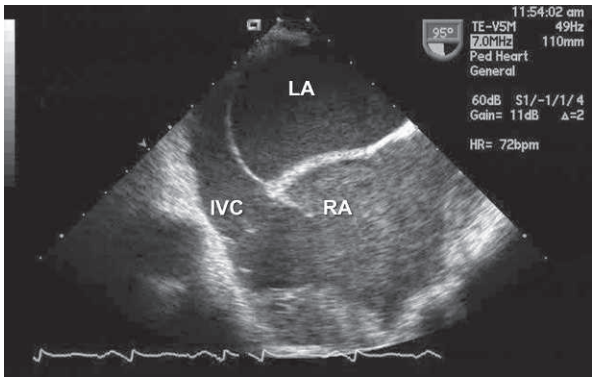

Fig. 3 *Transesophageal echocardiogram (bicaval view) after cardioversion to sinus rhythm shows improved but persistent spontaneous echo contrast.*

IVC = inferior vena cava; LA = left atrium; RA = right atrium/atriopulmonary Fontan circuit to pulmonary arteries

Supplemental motion image is available for Figure 3.

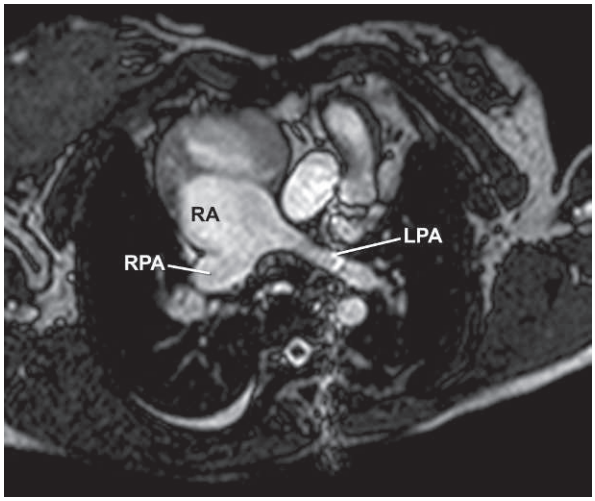

Fig. 4 *Cardiac magnetic resonance image (axial view) shows atriopulmonary Fontan anatomy (right atrium-to-bilateral branch pulmonary artery anastomoses with visible contrast).*

LPA = left pulmonary artery; RA = right atrium/atriopulmonary Fontan circuit to pulmonary arteries; RPA = right pulmonary artery

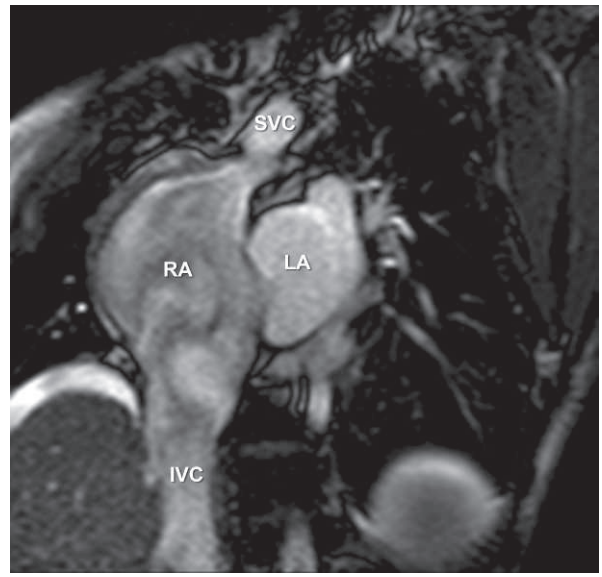

Fig. 5 *Cardiac magnetic resonance image (bicaval coronal view) shows the atriopulmonary Fontan circuit with swirling spontaneous echo contrast.*

IVC = inferior vena cava; LA = left atrium; RA = right atrium/atriopulmonary Fontan circuit to pulmonary arteries; SVC = superior vena cava

References

1. Cromme-Dijkhuis AH, Hess J, Hahlen K, Henkens CM, Bink-Boelkens MT, Eygelaar AA, Bos E. Specific sequelae after Fontan operation at mid- and long-term follow-up. J Thorac Cardiovasc Surg 1993;106(6):1126-32.
2. Ermis PR, Morales DL. The adult Fontan patient: update for 2011. Methodist DeBakey Cardiovasc J 2011;7(2):3-8.
3. Lam W, Friedman RA. Electrophysiology issues in adult congenital heart disease. Methodist DeBakey Cardiovasc J 2011; 7(2):13-7.
4. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report

of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults with Congenital Heart Disease). Developed in collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol 2008;52(23):e143-263.