Ventricular tachyarrhythmias are known sequelae among adults with repaired tetralogy of Fallot. The incidence of sustained ventricular tachycardia and sudden cardiac death in patients 35 years after corrective surgery is estimated at 11.9% and 8.3%, respectively. Studies have shown that right ventricular enlargement resulting from chronic pulmonary regurgitation is the most common hemodynamic substrate. QRs prolongation (>180 milliseconds) is one of the most sensitive ECG predictors and in turn correlates with right ventricular (RV) dilation. The exact mechanism is not as well studied as that of left ventricular arrhythmias, but it is purported that RV pressure and volume overload leads to RV myocardial stretching and RV fibrosis and hence serves as an arrhythmogenic focus.

The anatomic location of the RV pathology leading to ventricular tachyarrhythmias after tetralogy of Fallot surgical repair is not well defined. The basal portions of the RV and the subpulmonary regions of the RV are difficult to visualize on transthoracic echocardiography. Furthermore, 2-dimensional echocardiography is not capable of myocardial scar imaging. Cardiac magnetic resonance imaging (MRI) is capable of not only visualizing all anatomic segments of the RV but also delineating areas of myocardial fibrosis with late-gadolinium-enhancement imaging.

The value of cardiac MRI is demonstrated in this case of a 50-year-old man with a history of tetralogy of Fallot surgical repair as an infant. While playing recreational hockey, he suddenly developed presyncope with severe lightheadedness, dizziness, diaphoresis, and palpitations. On evaluation by emergency medical services personnel, he was found to have a wide-complex tachycardia at a rate of 300 bpm, suspicious for ventricular tachycardia (Figure 1). Normal sinus rhythm was restored after administration of amiodarone 150 mg IV. A postconversion ECG revealed normal sinus rhythm with a right bundle-branch block and a QRs duration of 175 milliseconds (Figure 2). The estimated total duration of the wide-complex tachycardia was 5 minutes. A transthoracic echocardiogram revealed evidence of both RV pressure and volume overload, with RV hypertrophy and abnormal septal motion.

A cardiac MRI study showed moderate RV dilation with severe RV systolic dysfunction, noting a global RV ejection fraction of 20%. Left ventricular function was normal with a global LV ejection fraction of 62%. The basal portions of the RV and RV outflow tract were aneurysmal (Figures 3 and 4 and Movie I in the online-only Data Supplement). This was not as well appreciated on the transthoracic echocardiogram (Movie II in the online-only Data Supplement). The RV aneurysmal area was found to have discrete subendocardial fibrosis on late-gadolinium delayed-hyperenhancement imaging (Figure 5). Late-gadolinium-enhanced imaging also illustrated fibrosis at the LV–RV hinge point (Figure 6) but not in the left ventricle itself. Hence, the arrhythmogenic focus was thought to be the RV outflow tract. The patient subsequently underwent automatic implantoable cardioverter-defibrillator placement.

Cardiac MRI is capable of acquiring true short-axis views of the ventricles from base to apex, allowing detection of segmental RV wall motion abnormalities that are not evident with 2-dimensional echocardiography because of its thoracic window limitations. In addition, 2-dimensional echocardiography is usually not capable of visualizing the RV outflow tract in the coronal view, which is easily accomplished with cardiac MRI. This case demonstrates the value of cardiac MRI in the assessment of patients with surgically repaired tetralogy of Fallot.

Disclosures
Gadolinium was used for delayed-enhancement imaging of the area of fibrosis. Dr Biederman has received significant funding from the National Institutes of Health/National Heart, Lung, and Blood Institute and the American Heart Association and a significant research grant from Merck. Dr Biederman also has modest contributions from the Merck Honorarium and Osiris Steering Committee. The other authors report no conflicts.

References

From the West Penn Allegheny Health System, Pittsburgh, PA (A.C., R.W.B., R.C., S.R., R.W.W., J.Y.); and Northeast Ohio Medical University, Youngstown, OH (J.R.M.).

The online-only Data Supplement is available with this article at http://circ.ahajournals.org/lookup/suppl/doi:10.1161/CIRCULATIONAHA.112.000644/-/DC1.

Correspondence to Anisa Chaudhry, MD, 29 Sterling Ct, Hershey, PA 17033. E-mail Anisa.Chaudhry@gmail.com

Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.112.000644

185
Figure 1. Rhythm strip illustrating wide-complex tachycardia suspicious for monomorphic ventricular tachycardia and its transition to normal sinus rhythm (red arrow) within 5 minutes of administration of 150 mg IV amiodarone.

Figure 2. Baseline ECG demonstrates normal sinus rhythm with a right bundle-branch block pattern and prolonged QRS of 175 milliseconds.

Figure 3. Magnetic resonance imaging short-axis view of the right ventricular outflow tract aneurysm.

Figure 4. Magnetic resonance imaging 4-chamber view of the right ventricular outflow tract aneurysm.

Figure 5. Late gadolinium enhancement illustrating endocardial fibrosis of the aneurysmal right ventricular outflow tract.
Figure 6. Late gadolinium enhancement illustrating fibrosis at the left ventricle–right ventricle hinge point.
Ventricular Tachycardia and Right Ventricular Fibrosis After Tetralogy of Fallot Surgical Repair
Anisa Chaudhry, Robert W. Biederman, Roberto Candia, Sahadev Reddy, Ronald W. Williams, June Yamrozik and J. Ronald Mikolich

doi: 10.1161/CIRCULATIONAHA.112.000644

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/128/2/185

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2014/10/23/128.2.185.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/