Ventricular Tachycardia Following Surgical Repair of Complex Congenital Heart Disease

Sherrie Joy Baysa, MD, Ronald J. Kanter, MD*

KEYWORDS

• Congenital heart disease • Tetralogy of Fallot • Double-outlet right ventricle • Pulmonary atresia • Ventricular tachycardia • Sudden cardiac death • Implantable cardioverter-defibrillator • Antiarrhythmic drugs

KEY POINTS

- Although uncommon, ventricular tachycardia can occur even in children following complex right ventricular outflow tract surgery.
- Programmed ventricular stimulation has reasonably high predictive value in patients having undergone right ventricular outflow tract surgery for complex congenital heart disease.
- Antiarrhythmic drug therapy still has a role in the management of the occasional youngster with congenital heart disease and ventricular tachycardia, although it does not provide the same level of protection of that provided by an implantable cardioverter-defibrillator.
- Cardiac MRI has an increasingly important role in surgical planning for patients having congenital heart disease. Even as MRI compatible implantable cardioverter-defibrillators are becoming available, the effect that these device components have on data acquisition may make their implantation problematic.

CLINICAL PRESENTATION

A 9-year old boy with surgically repaired complex congenital heart disease (CHD) presented with a 1-year history of self-limited palpitations, unassociated with syncope, light-headedness, shortness of breath, or chest pain. He was born with double-outlet RV, pulmonary atresia, subaortic ventricular septal defect (VSD), and normal-sized branch pulmonary arteries. Interventions included placement of 3.0-mm right modified Blalock-Taussig shunt as a neonate; shunt takedown, VSD closure, and right ventricle (RV) to pulmonary artery homograft conduit at 8 months; surgical revision of the stenotic conduit at 5 years; and complex interventional catheterization at 7 years because of distal conduit stenosis (dilation with stent), right pulmonary artery stenosis (dilation with stent), and severe pulmonary valve insufficiency (Melody valve placement [Medtronic; St Paul, MN]). Because attempts at documenting the rhythm during clinical episodes using an attached event recorder were unsuccessful, the patient underwent cardiac catheterization. The procedure was performed under propofol-based general anesthesia. He was found to have two-thirds systemic RV pressure, mild pulmonary valve insufficiency, moderate stenosis of both stents, and left ventricular end-diastolic pressure of 10 mm Hg. After balloon angioplasty of the stents, electrophysiologic study (EPS) was performed.

Division of Cardiology, The Heart Program, Nicklaus Children's Hospital, Miami Children's Hospital Health System, 3100 Southwest 62nd Avenue, Miami, FL 33155, USA
* Corresponding author.
E-mail address: Ronald.Kanter@mch.com

Card Electrophysiol Clin 8 (2016) 201–204
http://dx.doi.org/10.1016/j.ccep.2015.10.029

ELECTROPHYSIOLOGY STUDY

Baseline intervals showed incomplete right bundle branch block (RBBB) with a QRS duration of 132 milliseconds (Fig. 1), normal AH (atrium-to-His) and HV (His-to-ventricle) intervals, and absent VA (ventriculoatrial) conduction. Atrial flutter of 2-second duration was inducible at baseline, and no supraventricular tachyarrhythmias were inducible under the influence of isoproterenol. During ventricular programmed stimulation at baseline and using an output twice diastolic pacing threshold at 2 milliseconds pulse width, sustained rapid ventricular tachycardia (VT) (cycle length 200 milliseconds) was induced and reinduced by a drivetrain cycle length of 400 milliseconds and coupling intervals of 260, 210, and 190 milliseconds from the RV apex (Fig. 2). Ventricular diastole was difficult to discern, but the best estimation was that this had left bundle branch block morphology with superior QRS axis. It generated no blood pressure by arterial line, requiring cardioversion at 50 J.

MANAGEMENT AND CLINICAL COURSE

The patient was treated with enteral phenytoin and, with a therapeutic plasma level, programmed ventricular stimulation was repeated. The study was negative for inducible VT using an aggressive pacing protocol: 2 drivetrains (600 and 400 milliseconds), 3 premature beats down to cycle length 180 milliseconds, and from 2 RV sites (apex and outflow tract). An insertable loop recorder (Reveal LINQ; Medtronic; St Paul, MN) was implanted for documentation of the patient's rhythm during subsequent symptoms, and the patient was discharged with an automated external defibrillator (AED).

CLINICAL CONSIDERATIONS

This patient's cardiac anatomy and subsequent interventions can logically be viewed from the perspective of a patient with tetralogy of Fallot (TOF), a condition for which far more experience has been accrued. Several key questions influenced this patient's management. What is the relationship between this patient's symptoms and the results of electrophysiologic testing? What is the risk of sudden cardiac death (SCD) or clinical VT in this patient? What is the positive predictive value of inducible VT in this disease? In addition, what are the risks and benefits of implantable cardioverter-defibrillators (ICDs) in this population?

DISCUSSION

It was not thought that the induced rapid VT could account for the patient's more mild symptoms. Although atrioventricular (AV) reciprocating tachycardia could not exist in the absence of VA

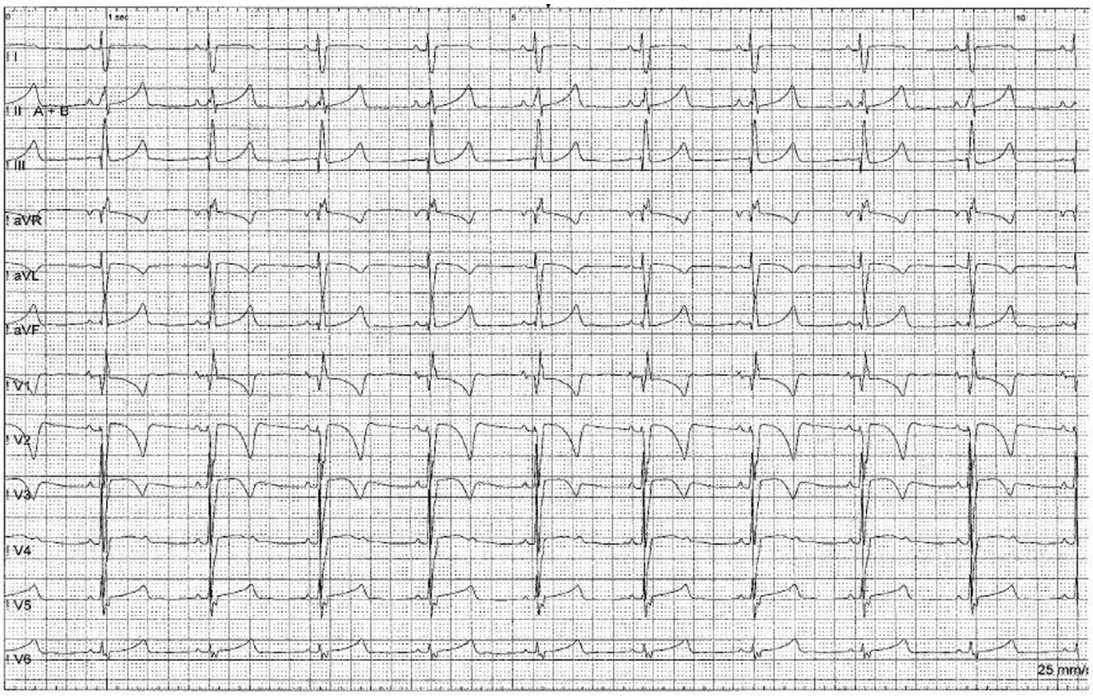

Fig. 1. Baseline surface electrocardiogram (ECG). Baseline intervals were normal, except for incomplete RBBB with QRS duration of 132 milliseconds.