Cardiology in the Young

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Left ventricle implantable cardioverter defibrillator: a dextro-transposition of the great arteries case

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Brief Report

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Abstract

We present a case of a patient with dextro-transposition of the great arteries palliated with a Senning procedure and a long-term arrhythmic complication that required an intervention, with an Implantable Cardioverter Defibrillator (ICD) implantation in the sub-pulmonary ventricle (morphologically left). This case highlights the need to perform off-label procedures to deal with the long-term complications of these complex patients.

The current surgical treatment of dextro-transposition of the great arteries is arterial switch;[1] before this was the standard of care, the atrial switch surgery was performed, leaving the morphologically right ventricle as the systemic ventricle. Nowadays, we must be prepared to deal with the long-term complications of this procedure.

Clinical case

A 33-year-old male, with good functional status, was submitted to a Senning operation[2] by the age of 16 months. He was referred to our Adult CHD Clinic in 2003. He was on permanent junctional rhythm with frequent pauses (the longest 5200 ms), with a mean heart rate of 50 bpm, asymptomatic and with a good functional capacity on serial treadmill exercise tests. It was decided to maintain surveillance. In 2013, he performed an echocardiogram that showed a moderately dilated systemic ventricle, with severe hypertrophy and moderated systolic dysfunction. He was medicated with lisinopril 5 mg once daily (dosage was limited by symptomatic hypotension). On February 2019, the patient was admitted to the Emergency Department complaining of episodic shortness of breath, chest pain and palpitations that lasted for 2 minutes, without syncope. The patient was admitted for rhythm monitoring, as he had a complex CHD with systemic ventricular dysfunction and high risk for arrhythmic events and sudden cardiac death.[3] The patient presented 2 episodes of symptomatic monomorphic non-sustained ventricular tachycardia, the longest one for 14 seconds (Fig 1), and a basal slow junctional rhythm (minimum heart rate 23 bpm), with frequent premature beats and bigeminy. In the context of a post-Senning patient, with systemic ventricular dysfunction, symptomatic non-sustained ventricular tachycardia and a slow junctional rhythm, a single chamber Implantable Cardioverter Defibrillator (ICD) was implanted in the sub-pulmonary ventricle (morphologically left). An after-procedure echocardiogram was performed with no atrioventricular (AV) valve dysfunction or worsening of systemic ventricle function. He was discharged with medical therapy for heart failure, including low-dose beta-blocker. Therapy was limited by symptomatic hypotension. During a 12-month follow-up, the patient was on New York Heart Association (NYHA) functional class I, with mild systemic atrioventricular valve insufficiency, with a percentage of ventricular pacing of 28% and no arrhythmic events (Figs 2 and 3).

Conclusions

Ventricular arrhythmias are the main cause of late-mortality in d-TGA.[3] Although the complex CHD imposes difficult technical issues, many patients may require cardiac device implantation during adulthood, with important clinical and prognostic impact.

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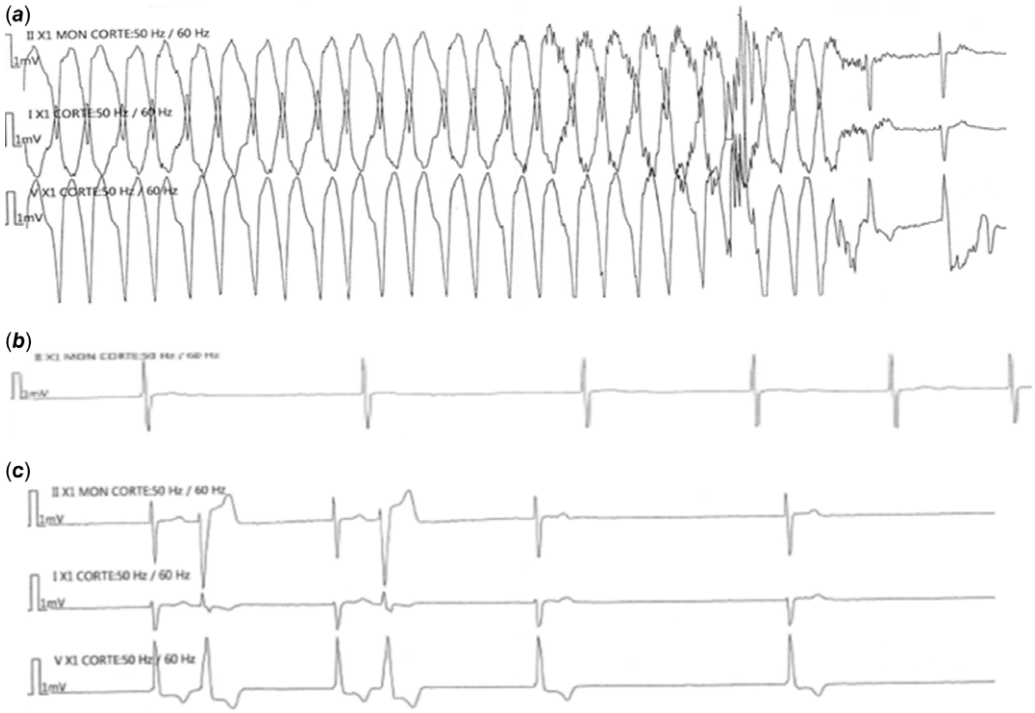

Figure 1. Telemetry monitoring: (***a***) Non-sustained ventricular tachycardia; (***b*** and ***c***) basal heart rhythm, minimum heart rate 23 bpm, frequent premature beats, bigeminy and pauses.

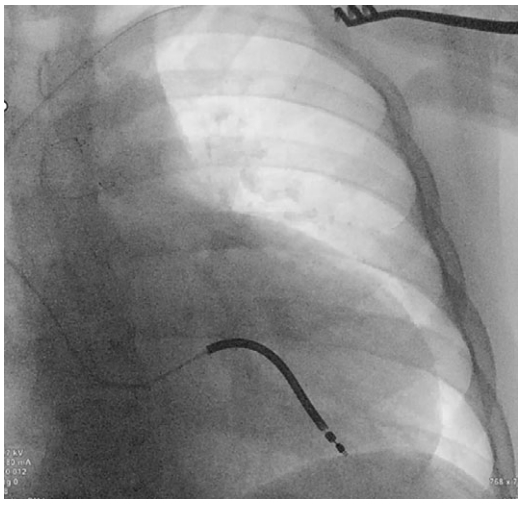

Figure 2. ICD implantation.

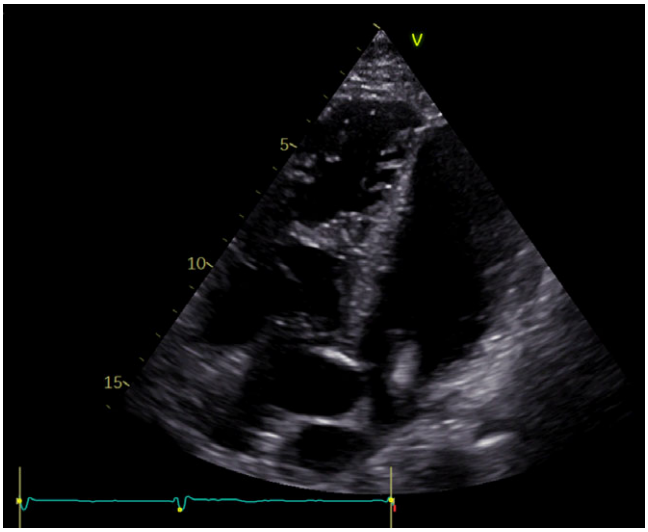

Figure 3. Echocardiographic follow-up.

Conflicts of interest. None.

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