Implantable cardioverter defibrillator in a young child with left ventricular noncompaction

Défibrillateur automatique implantable chez un jeune enfant atteint de non-compaction ventriculaire

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KEYWORDS
Left ventricular noncompaction; Ventricular fibrillation; Implantable cardioverter defibrillator; Sudden death

Abbreviations

ICD implantable cardioverter defibrillator
LVNC left ventricular noncompaction

We report the case of a 33-month-old boy, who was referred to our emergency care unit because he presented with syncope during effort. There was no family history of sudden cardiac death. Initial clinical examination and electrocardiogram were strictly normal. After a sudden cardiac arrest due to ventricular fibrillation in the emergency room, he was defibrillated. Subsequently, the patient was shocked several times due to repeated episodes of ventricular fibrillation. Two-dimensional echocardiography established the diagnosis of isolated LVNC (Fig. 1). A few days after the cardiac arrests occurred, left ventricular function was normal. An automatic ICD (Virtuoso II VR; Medtronic, Inc., Minneapolis, MN, USA) was implanted in the chest of this 15 kg child (Fig. 2). A sternotomy was performed to facilitate epicardial placement of the shocking leads. The ICD generator was placed into the abdomen. Antiarrhythmic drug therapy (amiodarone) was started immediately after the cardiac arrests occurred and was continued in association with aspirin at an antiplatelet dose. Six months later, three episodes of ventricular fibrillation were successfully shocked. The ICD did not produce inappropriate shocks and appeared to be well tolerated.

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Figure 1. Apical four-chamber two-dimensional (A) and three-dimensional (B) echocardiographic views showing typical aspect of left ventricular noncompaction (LVNC): presence of a two-layer structure with a thin compacted epicardial layer and a thicker noncompacted endocardial layer consisting of numerous prominent ventricular trabeculations (white arrows) and deep intertrabecular recesses. (C) Parasternal short-axis end-systolic two-dimensional echocardiographic view showing the two-layer structure. In this view, the ratio of noncompacted to compacted (NC/C) thickness was 2.2; in children, an NC/C ratio > 1.4 is recognised as a diagnostic criterion for LVNC. (D) Apical four-chamber Doppler colour view showing blood flow from the left ventricular cavity into the intertrabecular recesses. LV: left ventricle.

Ventricular fibrillation is so rare in children that it should systematically bring LVNC to the physician’s mind. Left ventricular failure, systemic thromboembolic events, arrhythmias and sudden cardiac death are well known complications of LVNC. Although the prognosis of this rare cardiomyopathy is closely correlated with the occurrence of cardiac events, the secondary prevention of sudden cardiac death using an ICD remains controversial.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.