

# Implantable cardioverter-defibrillator in a child with isolated noncompacted myocardium

## *Cardioversor desfibrilador implantável em criança com miocárdio não-compactado isolado*

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### CLINICAL DATA

9-year-old male child, 25 kg, from Barretos, São Paulo. Full-term patient presenting cardiorespiratory arrest (CRA) without defined cause and remained for 30 days in Intensive Care Unit (ICU). At 11 months presented episode of fainting, followed by cyanosis and CRA. He had undergone cardioversion due to ventricular fibrillation (sic). At the time, corticoid and digitalis were used, and the patient evolved well until the last two months, when presented new fainting episode at rest, requiring cardiac massage and new cardioversion to revert this presentation. Amiodarone, spironolactone, aspirin and suspension of digitalis were administered. After about 10 episodes of syncope and fainting, the child was referred to the ICU of our Service. On physical examination the patient was in good general condition, afebrile, acyanotic and eupneic. Precordium unchanged, regular heart rhythm, with normal and rhythmic sounds. Pulmonary auscultation was normal. Flaccid abdomen, liver at 2 cm from the right costal margin, spleen not palpable and presence of bowel sounds. Good peripheral perfusion, without edema, palpable and symmetrical pulses in all limbs, without motor sequels with only mild mental retardation, resulting from the CRA at the time of parturition (sic).

### ELECTROCARDIOGRAM

Sinus rhythm, heart beat of 88 bpm, SÂP 0°, SÂQRS - 30°, PR interval 0.24s. First-degree AV block, atrial and left ventricular overload, and left branch block.

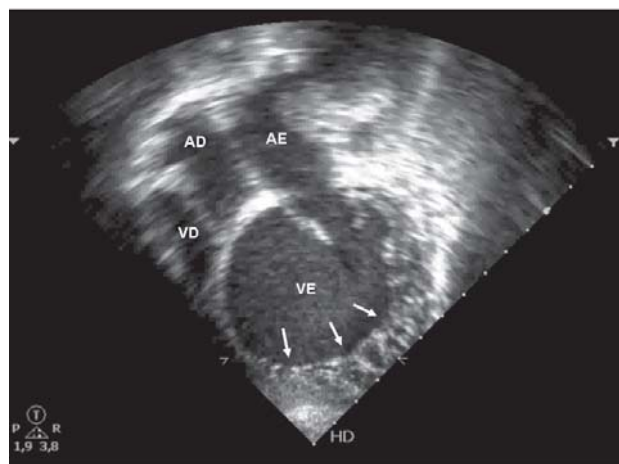


Fig. 1 - Apical four-chamber view showing dilated left ventricle with exuberant trabeculation in lateral and apical walls (arrows,) featuring noncompacted myocardium

### RADIOGRAM

Visceral *situs solitus* in levocardia. Increased cardiac area with left ventricle affected, presenting a cardiothoracic index of 0.68 and second arc slightly increased. Pulmonary vascular network without apparent changes.

### ECHOCARDIOGRAM

*Situs solitus* in levocardia. Normal venoatrial, atrioventricular and ventriculoarterial connections. Right ventricle with thin walls at trabecular portions and outflow

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tract, left ventricular myocardium with spongiform appearance (Figure 1), left ventricular contractile dysfunction of significant degree with an ejection fraction of 33.8% and mild mitral valve insufficiency. Absence of intracavitary thrombi.

### DIAGNOSIS

The patient was referred to our Service due to repeated episodes of ventricular fibrillation and recovery from sudden death [1], the treatment was quickly directed to the implantation of a cardioverter defibrillator. The diagnosis of noncompacted myocardium and not associated with other abnormalities is extremely rare, with few patients described in the literature and caused by a rare disorder of endomyocardial morphogenesis [2].

### OPERATION

Patient in dorsal decubitus position, under general anesthesia, incision in the left infraclavicular region, divulsion of the pectoralis major muscle and making of the pocket with appropriate dimensions for subsequent generator implantation. Puncture of the left subclavian vein,

passage of the guidewire, dilator and electrodes up to the right ventricle, with positioning at the most apical portion so that the coil may contact a large portion of the ventricular wall. Similarly as for the atrial electrode, it was possible to obtain appropriate measurements in the right atrium lateral wall. Both electrodes were of active-fixation type. The measurements of electrodes measures reveled as follows: the atrial electrode tip threshold was 1.1 mV, impedance of 426 ohms and return of 23.0. The right ventricular electrode presented threshold of 0.7 mV, impedance of 420 ohms and return loss of 10,0. After induction of ventricular fibrillation (Figure 2), two shocks were required to revert to sinus rhythm, the first was of 15J and the second 20J (Figure 3). With the system tested and the measures within the desired standards, the implantation of the generator was performed through layer suturing technique. The patient was referred to ICU and chest radiography (Figure 4) and echocardiogram were performed in order to prevent complications such as pneumothorax and tamponade. As a result of ventricular dysfunction and prior arrhythmia, the patient was discharged from hospital on the third postoperative day, under high doses of metoprolol and amiodarone.

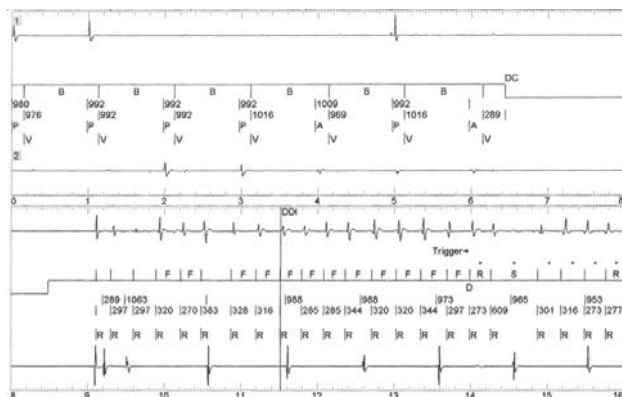


Fig. 2 - Induction of ventricular fibrillation

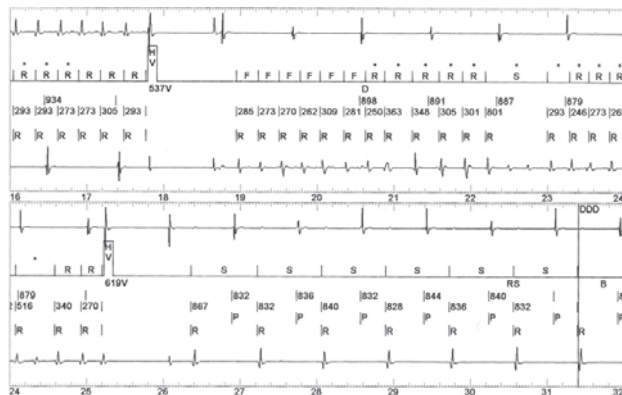


Fig. 3 - First unsuccessful ventricular defibrillation with 15J. Second ventricular defibrillation with 20J and reversion to sinus rhythm

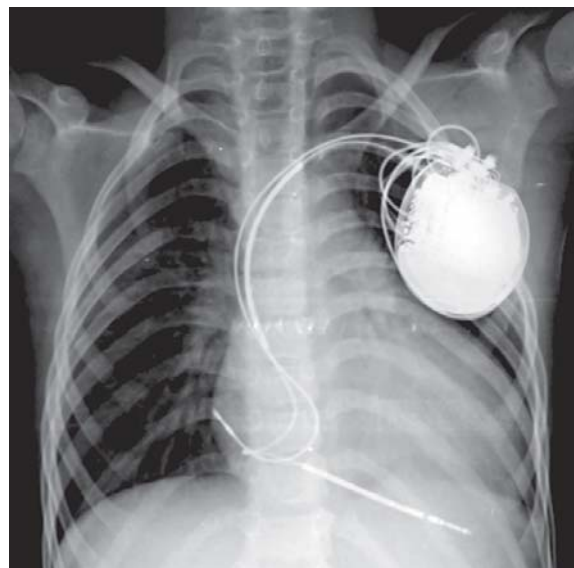


Fig. 4 – Chest radiography in the early postoperative period with the atrial and ventricular electrodes implanted

### REFERENCES

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2. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. Circulation. 1990;82(2):507-13.