

# Successful treatment of electrical storm in a child with early repolarization syndrome with orciprenaline and radiofrequency ablation

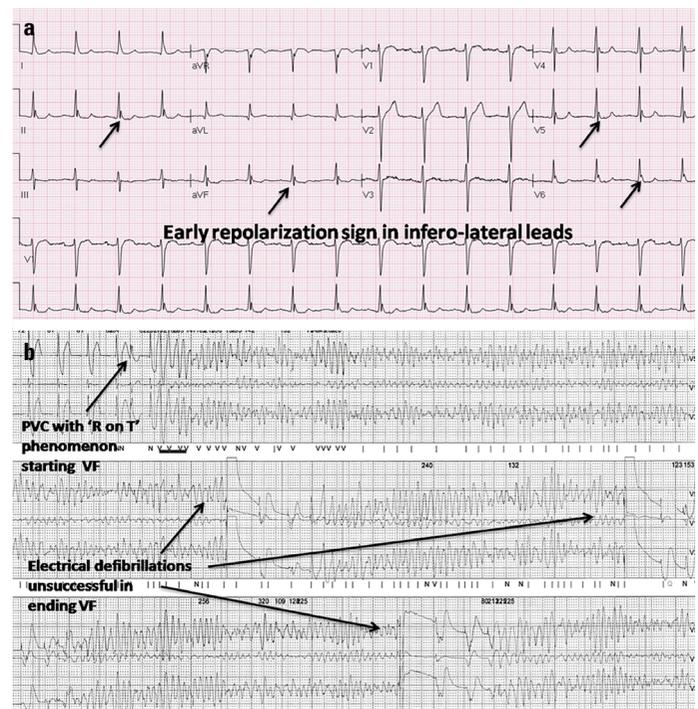
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## Introduction

Early repolarization (ER) is a benign electrographic pattern and described as J-point elevation  $\geq 1$  mm in two or more adjacent inferior and/or lateral derivations, resulting in QRS slurring or notching (1-3). Some researchs have shown that ER might be one of the arrhythmogenic reasons for idiopathic ventricular fibrillation (VF) (1, 3). Here we present the case of a boy with inferolateral ER complicated by an electrical storm and treated with orciprenaline and radiofrequency ablation of the Purkinje network.

## Case Report

A 14-year-old boy was referred to our center after having a survived sudden cardiac arrest at rest. The patient's electrocardiogram (ECG) yielded inferolateral ER (Fig. 1a). He had received an implantable cardioverter defibrillator (ICD; Medtronic Evera MRI SVR Sure scan) after the arrest, and mexiletine (3.5 mg/kg/day), sotalol (200 mg/m<sup>2</sup>), and metoprolol (1 mg/kg/day) had been prescribed because of three ICD shocks for VF. The device had been programmed VF only mode (with 35 joule) and no anti-tachycardia pacing. One month later, he experienced 14 shocks and 3 external defibrillations due to VF without success, despite amiodarone infusion (10 mg/kg/24 h) and taking mexiletine, sotalol, and metoprolol (Fig. 1b). Finally, orciprenaline infusion (0.03 mg/min) was started, which eliminated VF by increasing heart rate. In addition, disopyramide (6 mg/kg/day) was started. However, the patient experienced three shocks again when the orciprenaline dose was reduced. We decided to perform an electrophysiologic study for the ablation of premature ventricular contractions and the Purkinje fibers. The distal Purkinje fiber foci in the right and left ventricles were successfully ablated using an irrigated radiofrequency ablation catheter via the EnSite three-dimensional electroanatomic mapping system (St. Jude Medical, Inc.). The radiofrequency ablation (30 W, 43°C, 96–104 ohm) was implemented empirically in a linear style over 1–2 cm in the mid-right and mid-left ventricles at the site of presystolic Purkinje potentials without any complication (Fig. 2). Total procedure and fluoroscopy time were 210 and 24.7 min,



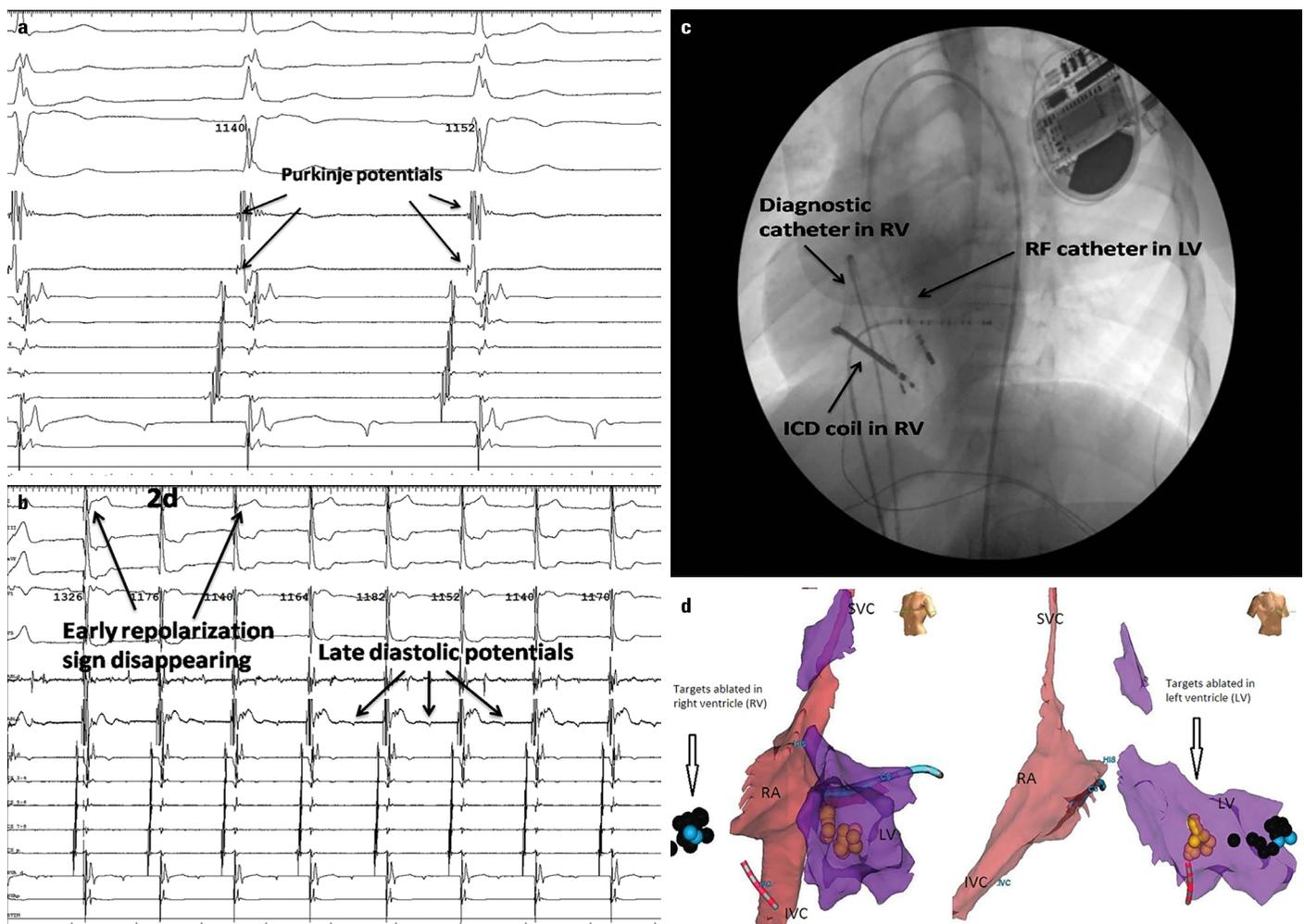
**Figure 1.** (a) The 12-lead surface electrocardiogram (ECG) on admission with the signs of malignant early repolarization in the inferolateral leads. (b) This Holter ECG record shows the episodes of ventricular fibrillation and implantable cardioverter defibrillator shocks in the pediatric intensive care unit

respectively. After the procedure, the J-point elevation on the ECG was almost disappeared (Fig. 3). The patient has remained symptom-free with quinidine (5 mg/kg/day) and amiodarone (5 mg/kg/day) for 5 months.

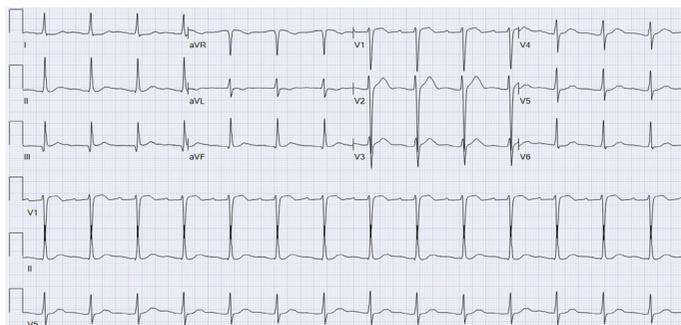
## Discussion

Three types of ERs have been determined by Antzelevitch et al. (2) according to risk stratification. In type 1, ER is seen only in the lateral leads. It is common in healthy athletes and is usually benign regarding arrhythmias. In type 2, ER is seen in the inferolateral leads and is associated with a higher risk for malignant arrhythmias. Finally, type 3 is characterized by ER in the inferolateral plus anterior or right ventricular leads, and it is significantly associated with malignant dysrhythmias and electrical storms. In our patient, the ER pattern was probably similar to type 2.

Currently, the isoproterenol infusion or rapid cardiac pacing is the most useful treatment approach in the early phase of an electrical storm (4). The drug of choice for the long-term treatment of recurrent idiopathic VF is quinidine, an inhibitor of fast sodium channel ( $I_{Na}$ ) (1, 4, 5). In our patient, we started with orciprenaline to control the electrical storm. Isoproterenol and orciprenaline are beta-adrenergic agonists, and both of them can decrease the electrical gradient and reduce the J-point elevation by activating  $I_{Ca}$ ; moreover, because heart rate can affect  $I_{Na}$  functions, these drugs may inhibit  $I_{Na}$  by accelerating heart rate, which results in normalization of repolarization abnormalities. Kyriazis et al. (6)



**Figure 2.** (a) This intracardiac record demonstrates the Purkinje potential before the ablation of the Purkinje network. (b) After the ablation, the J-point elevation almost disappeared in the 12-lead electrocardiogram and the late diastolic potentials from the ventricle were seen in the intracardiac record. (c) The fluoroscopy image shows the irrigated-tip radiofrequency ablation catheter in the left ventricle as well as the diagnostic catheter and the implantable cardioverter defibrillator coil in the right ventricle. (d) Three-dimensional mapping determining the ablated Purkinje potentials in the right (black points) and left (yellow points) ventricles



**Figure 3.** This surface electrocardiogram after the ablation shows no early repolarization characterized by the J-point elevation

have reported the effectiveness of orciprenaline in a 42-year-old patient with an electrical storm. Our case is the first report in terms of usefulness of orciprenaline for early treatment of electrical storm in a child.

Catheter ablation at experienced centers is useful for the treatment of idiopathic VF (7, 8). It has been shown that the

Purkinje network might be a driver for VF due to its feature of a very rapid burst activity, and the Purkinje–myocardial junction can be part of a re-entry circle (8). Unfortunately, we could not perform any voltage mapping regarding any scar or fibrosis due to lack of premature ventricular contractions, which was a limitation in our case. We successfully performed the Purkinje network ablation.

## Conclusion

This pediatric case report shows the efficiency of orciprenaline infusion in the termination of recurrent VFs in the ER syndrome. In addition, the ablation of the distal Purkinje system seems feasible in a child with idiopathic VF triggered by ER.

**Informed consent:** Informed consent was obtained from the single patient described in this case report.

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at [www.anatoljcardiol.com](http://www.anatoljcardiol.com)  
DOI:10.14744/AnatolJCardiol.2019.37085



## A rare cause of dyspnea: Left atrial angiosarcoma

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### Introduction

Primary malignant cardiac neoplasms are extremely rare tumors. In the present report, a case of a patient with a large car-

diac mass in an unusual location is described after obtaining his and his relative's approval.

## Case Report

A 64-year-old male patient was admitted to the cardiology clinic with dyspnea that increased over 1 month. The patient suffered from no other known condition. Electrocardiography showed sinus rhythm, heart rate of 85/bpm, and blood pressure of 134/84 mm Hg. His laboratory parameters and NT-proBNP level were within normal limits (NT-proBNP=121 pg/mL, upper limit for adjusted gender and age <210 pg/mL). A transthoracic echocardiogram showed a large (4.6×5.4 cm) mass originating from the left atrium (Videos 1 and 2). Thorax computed tomography confirmed a 4.8×5.4 cm mass with smooth borders located at the inferior site of the left atrium (Fig. 1, 2). Two radiologists reviewed CT images and the mass was considered extracardiac, with central low-density characteristics, probably a complex cystic lesion such as a bronchogenic cyst or an esophageal duplication cyst. A transesophageal echo confirmed a probable extracardiac mass as well. Preoperative cardiac MRI and PET-CT were not performed because the initial diagnosis did not reveal a malignant tumor. Coronary angiogram showed normal coronary arteries and a mass image with arterial blood supply from the distal circumflex artery. Thereafter, the patient underwent cardiac surgery for removal of the mass. Because the mass was considered extracardiac, the first surgical cut was left anteroaxillary. Hence, the surgical exploration was compatible with an intracardiac mass originating from the left atrial basis; sternotomy was performed and surgery was continued under cardiopulmonary bypass. The 6.5×5.5×4 cm left atrial mass



**Figure 1.** Pre-contrast thoracic CT cross-sectional image of the mass