

Succesfull RF ablation for the treatment of electrical ICD storm in a child with early repolarization and idiopathic ventricular fibrillation

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Early repolarization(ER) may sometimes be associated with ventricular fibrillation(VF) and sudden cardiac death. A pattern of convex upward J wave, with ST-segment changes, especially in inferolateral leads on Electrocardiography(ECG) suggests “malignant” ER. We report a 14-year-old boy with ER associated idiopathic VF and electrical ICD storm controlled by intravenous isoproterenol and successfully treated by radiofrequecy(RF) ablation finally.

The patient was referred to our clinic with aborted sudden cardiac arrest, while resting at home. ECG yielded malignant type ER pattern and no significant structural defect was noticed on his echocardiograpy(Figure1-A). Coronary angiography, cardiac magnetic resonance, epinephrine-ajmalin challenge tests and electrophysiologic study(EPS) revealed no specific pathology. After that he was implanted a transvenous ICD(Medtronic-Evera, MRI-SVR Sure-scan) for secondary prevention and Metoprolol was initiated.

After 2 monts, he admitted to our ICU unit because of ICD storm with more then 20 shocks in a day(figure1-B). On İCU first day he further experienced a lot of appropriate ICD shocks, despite multiple antiarrhythmic medications(combination of beta bloker plus amiodaron, flecainide and mexiletine). Finally we strated intravenous isoproterenol infusion and VF storm was controled. Amiodarone plus oral disopyramide was initiated as maintenance therapy (because quinidin sulfate is not found routinely in our country). After an attempt to discontinue isoproterenol, the patient had further VF attacks and mutiple ICD shocks, so we decided for an urgent EPS and catheter ablation with ECMO backup ready. During the study we couldn’t stimulate any premature ventricular contraction or VF, so we ablated local purkinje signals in both ventricles with an irrigated RF catheter(Figure1-C and -D). Especially after the left ventricular purkinje signal substrate ablation, ER almost disappeared on inferolateral leads(Figure1-E) . After one week of successful ablation, the patient was discharged with oral amiodaron and disopramide therapy. He is still on follow-up without any VF attack and ICD shocks.

In conclusion; this case report represents the efficiency of the isoproterenol infusion for acute control of idiopathic VF associated with ER and electrical storm. However, if recurrent VF attacks and İCD shocks continue or recure after cessation of isoproterenol infusion, purkinje network ablation should be kepted in mind, even in children.

Figure-1:
 A- Twelve lead surface ECG on admission with 'malignant' early repolarization signs in inferior leads.
 B- ECG record of ventricular fibrillation and ICD shock.
 C- Flouroscopy image showing iirigated RF ablation catheter in LV and diagnostic catheter in RV, beside the ICD coil.
 D- 3-D Map showing ablated Purkinje potentials in both ventricles; in RV shown as black and in LV as yellow.
 E- 12 lead Surface ECG with almost lost of ER sign.

