daily whereas tripterygium and hydroxychloroquine were remained the primary dosage. The patient came to the cardiology department for return visit on May 20th, 2016. Her BP was 100/70mmHg, heart rate was 65/min. It was surprising that the QRS interval became normal and CLBBB disappeared (Figure 1B, upper panel). The echocardiography revealed the left ventricular contractility recovered to normal. The LVDd was decreased to 5.7cm and LVEF was increased to 50% (Figure 1B, lower panel). The cardiac remodeling was reversed to a certain degree. We suspected the patient suffered from IgG4 related heart disease and suggested her myocardial biopsy. She rejected our recommendation for fear of the procedural risk.
During the following half a year, the patient maintained the medication. On December 15th, 2016, the blood test showed IgG4 level was 5.18g/L and another echocardiography demonstrated that LVDd 5.6cm and LVEF 53%. In December 2017, methylprednisolone was reduced to 6mg once daily while tripterygium and hydroxychloroquine were still kept the primary dosage. Perindopril was kept 8mg once daily and Metoprolol succinate was kept 47.5mg once daily. The patient had no symptoms of heart failure. In May 2017, the IgG4 was retested 4.78g/L. The echocardiography showed LVDd 5.65cm, LVEF 50% and mild regurgitation of mitral and tricuspid valves. The patient was required to maintain current medication.

UNIQUE SITUATIONS IN THE MANAGEMENT OF VENTRICULAR ARRHYTHMIAS – FINDING SHELTER FROM THE STORM

4985
Heart team in electrical storm treatment

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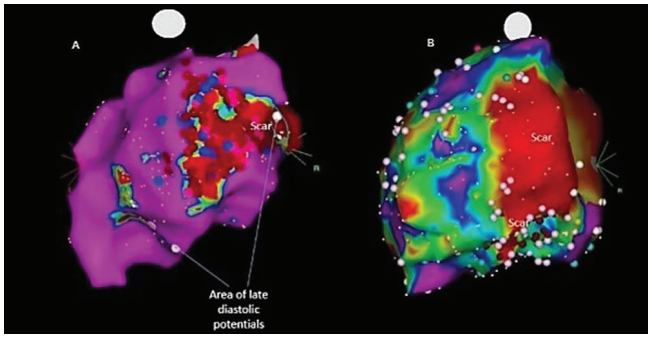
Introduction: In the settings of recurrent arrhythmic storms substrate-based catheter ablation is considered to be life-saving approach. Case report. A 51-year-old male was admitted for management of electrical storm (ES) with more than 40 appropriate ICD shocks within 2 hours due to recurrent hemodynamically unstable VT and VF episodes. He had history of 2 endocardial and 3 endo/epicardial ablations of VT substrate. Endomyocardial biopsy didn't demonstrate any specific abnormalities or acute inflammatory changes but revealed viral expression with possible Enterovirus etiology. During last 2 years the patient presented multiple electrical storms and underwent two redo ICD implantations due to battery depletion and then bilateral thoracopic sympathectomy which was partially effective. Stellate ganglion biopsy demonstrated neuropathy without specific viral expression. Several months ago he underwent PTCA of left anterior descending artery due to hemodinamically significant stenosis but despite complete revascularization and antiarrhythmic treatment he continued to experience sustained VT and VF. We considered new procedure of endo/epicardial bipolar catheter ablation under intracardiac echo control. Intraoperative electroanatomical mapping and EP study demonstrated epicardial origin of VT from LV lateral wall and mitral annulus region (Fig. 1) with rapid progression of arrhythmogenic substrate since last ablation episode. Substrat-based cooled RF ablation (40 W, 45C) was performed with the elimination of clinical VTs. Endomyocardial biopsy revealed possible prior myocarditis without acute inflammatory process. During 1 year follow up period the patient remained stable.



Progression of arrhythmogenic substrate

Questions, problems or possible differential diagnosis: This clinical case intended to evaluate the possible etiology, treatment tactics and outcome in patient with implanted ICD and recurrent ES due to episodes of sustained polymorphic VT/VF which were controlled only after several endo/epicardial ablation procedures. Possible differential diagnosis includes myocarditis, ion channel diseases, catecholaminergic polymorphic ventricular tachycardia (CPVT), ischemic heart disease and cardiomyopathies.
Answers and discussion: Etiology of recurrent VT and VF episodes and multiple ES in this patient remains controversial. Electroanatomical mapping revealed complex arrhythmogenic substrate with rapid progression over time which couldn't be explained only by ischemic heart disease considering complete revascularization.
Conclusions and implications for clinical practice: In this case report ES is considered to be polyetiological condition where potential ischemic origin of VT is combined with post-myocarditis fibrotic changes and autonomic disbalance with increased sympathetic tone. Heart team work and multidisciplinary approach with pharmacotherapy, bilateral thorascopic sympathectomy and repeated procedures of combined endocardial and epicardial catheter ablation appeared to be safe and effective strategy.

4986
LQTS type 7 evaluation in patient presenting with neurological signs: weakness and periodic paralyses of lower extremities

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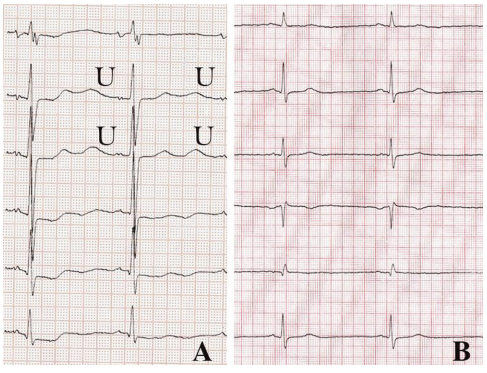
Introduction: Long QT syndrome (LQTS) is a large group of channelopathies. We provide a case of 12-year-old male with rare LQTS type 7 or Andersen–Tawil syndrome (ATS).
Case report description: Firstly our patient showed up to neurologist with muscle weakness and paresthesia in lower limbs with an inability to walk properly. After the routine ECG signs of QT prolongation was found and he was send to cardiologist. On the resting ECG U-wave and QUc-568ms appeared with single episodes of polymorphic ventricular premature beats detected by Holter. Echo and treadmill stress test showed no significant pathological abnormalities. There were no registered syncopes. Physical examination showed several face and skeletal dysmorphic features as, for example, broad forehead, micrognathia and clinodactyly of fourth fingers. The nervous system examination revealed weakness of proximal limb muscles (MRC grade 4 on the right and MRC grade 3 on the left) in both lower limbs with pyramidal signs of foot clonus.
Description of the techniques used: We provided a complete diagnostic examination: ECG, 24-hour Holter monitor, Echo, treadmill stress test, EEG, thyroid gland ultrasound, thyroid hormones measurements, blood electrolyte levels.
Questions and possible differential diagnosis: The main objective was to distinguish exact type of LQTS and to choose correct management. The controversial question was the selection of proper pharmacological rhythm control therapy and assessment of the necessity of ICD in asymptomatic patient. The second significant aim was to find a way how to cope with periodic paralysis. Finding the connection between the type of LQTS and response to treatment either to risk of SCD, seems to be of paramount importance. The possible differential diagnosis included other LQTS types, ventricular arrhythmias and periodic paralyses of hypokaliemic, hyperkaliemic or thyrotoxic nature.
Answers and discussion: Performed genetic testing detected heterozygous mutation in KCNJ2 that proves the ATS diagnosis suspected clinically. Paralysis attacks were not connected with potassium or calcium imbalance according to daily electrolyte monitoring. Paralysis developed after physical training as well as after long rest. No other specific trigger (as electrolyte imbalance, thyroid function abnormality etc) was detected. Commonly paralyses lasted from several hours to days and disappeared spontaneously. After detailed literature search and delicate evaluation of patient's family history (two cases of SCD in close relatives) the heart team decided to start rhythm control therapy with selective beta-blocker (atenolol in 1 mg/kg/per day). After two days of atenolol treatment U wave was no longer detected and QTc was in reference range (figure B). Patient's ECG pattern was normal in consecutive half year follow-up period. Unfortunately the nature of periodic paralyses remains unknown and patient still shows episodes of periodic weakness.



Conclusions and implications for clinical practice: Even though SCD beeing not common in patients with ATS, the electric myocardium instability and burdened family history make patient to fall in a high-risk group. To conclude we suggest the multidisciplinary team of specialists (pediatrician, neurologist, cardiologist, geneticist, cardiovascular surgeon) is becoming increasingly important in early diagnosis and maintenance of patients with ATS.