Management of electrical storm in a 14-year-old girl with Andersen-Tawil syndrome

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Introduction:
Andersen-Tawil syndrome (ATS) / Long QT syndrome Type 7:
• Predisposition for ventricular arrhythmias: isolated premature ventricular contractions (PVC), bidirectional or polymorphic ventricular tachycardia (VT)
• Generally low risk of Torsade de Pointes tachycardia or sudden cardiac death
• First report of management of electrical storm in patient with ATS

Case Presentation:
• 14-year-old girl, clinical features and positive genetic testing for ATS
• At age 5 ICD implantation due to a syncope, recurrent bidirectional VT and short non-sustained VT (Fig. 1)
• Treatment with Flecainide (2mg/kg) and Metoprolol (2mg/kg)
• Parents refused change of antiarrhythmic medication over the years despite increase of arrhythmia burden (56,8%) and reduced shortening fraction (SF) during clinical course (Fig. 2)
• Begin 2016: reduced physical fitness – approval to change antiarrhythmic medication

Conclusion: Electrical storm is a rare but life-threatening complication of Andersen-Tawil syndrome which was acutely treated by Lidocain infusion in our patient. As described once before Imipramin may help to reduce arrhythmia burden in these patients.