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

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Introduction: One prominent feature of Anderson-Tawil syndrome (ATS) is a propensity to considerable ventricular arrhythmia such as isolated premature ventricular contractions (PVC), bidirectional or polymorphic ventricular tachycardia (VT). However, the risk of Torsade de Pointes tachycardia or sudden cardiac arrest seems to be low in ATS. To the best of our knowledge, this is the first report of management of electrical storm in a patient with ATS.
Case Presentation: In a 14-year-old girl with clinical features and a positive genetic testing for ATS an ICD was implanted in 2007 due to a syncope and recurrent bidirectional VT. Despite the treatment with Flecainide and Metoprolol arrhythmia burden increased over the years up to 70% in the Holter monitor. Although VT was clinically well tolerated, ventricular function became compromised in the last 6 months with a shortening fraction (SF) of 20-25%. Five days prior to hospital admission for a planned change of antiarrhythmic medication she suffered from a syncope due to self-limited VT. Some hours later the patient developed an electrical storm with recurrend hemodynamic instable VT. Despite sedation, intubation, and infusion of Esmolol and magnesium 11 internal and external (after deactivating the ICD to save battery life-time) defibrillations had to be performed. Left ventricular function was reduced to a SF of 15-20%. After the injection and subsequent continuous infusion of Lidocaine rhythm stabilized and sinusrhythm in alteration with ventricular bigeminus was noted. In the next five days Lidocain infusion could be stopped and oral antiarrhythmic medication was changed from Metoprolol to Propranolol (5 mg/kg/d) and from Felcainide to Imipramin (4 mg/kg/d) resulting in a reduction of arrhythmia burden to 20% with mainly PVC/ventricular bigeminus in the Holter monitor. In the next 3 months arrhythmia burden remained stable and ventricular function returned to normal with a SF of 30-35%.
Conclusion: Electrical storm is a rare but life-threatening complication of Anderson-Tawil syndrome which can acutely be treated by Lidocain infusion in our patient. As described once before Imipramin may play an important role in the chronic treatment of this channelopathy.