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SCN5A mutations: special ventricular lead capture features in children?

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Unstable and changing lead capture threshold are a recognized difficulty when implanting cardiac devices in adult with loss-of-function SCN5A mutation.
We aim to study our pediatric cohort.

Case#1: 7y boy. PM at 3y due to a febrile cardiogenic syncope and detection of complete heart block not detected previously.First Flecainide test did not show ECG changes.VVIR implantation with oscillating capture during the implantation.Final ventricular capture:1.5V at 0.31ms, but during follow-up capture was increased until 2.7 mV at 0.31ms.ICD implantation at 7y due to syncope and an atrial flutter.Mutation in SCN5A c.659C>T identified and type-1 Brugada ECG under flecainide. Both final atrial and ventricular capture:1.25V at 1.0 ms.
Case#2: 8y girl. PM at 3y due to complete heart block. According to loss-of-function SCN5A double mutation, electrophysiological test and clinical features, Lev-Lenegre syndrome was diagnosed. VVIR implanted with with oscillating ventricular capture during implantation and follow-up
Case#3: 11y girl. Prenatal echocardiograms showed foetal tachycardia accompanied of foetal hydrops. During first years, fascicular tachycardia was diagnosed and controlled with flecainide.Despite of the ECG improvement, complete heart block was alternating with fascicular tachycardia. Pacemaker implantation Endurity ™ SR mode VVI, at 9 years old, due to bradycardia symptoms, was performed. During pacemaker implantation newly capture issue was detected, with better initial compared to final procedure poor or absent stimulation despite of high voltages within the apex; stimulation improvement was detected within right ventricular outflow tract and then we obtained better capture.Final ventricular capture: 0.5V at 0.4ms. During 1 year follow-up, no issues were detected.
Genetic features: de novo SCN5A c.4783G>A, missense.

Conclusions:
As well as data published for adult patients, undulated capture thresholds are also an important issue in paediatric patients with loss-of-function SCN5A mutation carrying a pacemaker or implantable cardioverter defibrillator (ICD).
Genetic testing is a useful tool not just for the clinical management or familial segregation, but also it is an outstanding tool for the electrophysiologist.
Recognition of this clinical entity may help to understand the electrophysiological behaviour of SCN5A-related diseases and planning pre-implantation and follow-up strategies.