SCN5A mutations: special ventricular lead capture features in children?

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Introduction

Unstable and changing lead capture threshold are a recognized difficulty when implanting cardiac devices in adult population with loss-of-function SCN5A mutation.

Material and methods

We aim to study our paediatric cohort with SNC5A mutation whom required a cardiac device.

Results

Case 1

7 years-old boy required a pacemaker (PM) due to 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 procedure. Final ventricular capture 1.5V at 0.31ms, but during follow-up capture was increased until 2.7 mV at 0.31 ms. ICD implantation at 7 years old due to syncope and atrial flutter. Mutation in SCN5A c.659C>T was identified and type-1 Brugada ECG under flecainide was detected. Both final atrial and ventricular capture 1.25V at 1.0 ms.

Case 2

8 years-old girl required a PM at 3 years old due to complete heart block. According to loss-of-function SCN5A double mutation, electrophysiological test and clinical features, Lev-Lenegre Syndrome was diagnosed. VVIR mode PM was implanted with oscillating ventricular capture during implantation and follow-up.

Case 3

11 years-old 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. PM implantation Endurity™ SR mode VVI, at 9 years old, due to bradycardia symptoms, was performed. During PM 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 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.

Discussion / Conclusions

1. 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 PM or ICD.

2. Genetic testing is an useful tool not just for the clinical management or familial segregation, but also it is an outstanding tool for the electrophysiologist.

3. Recognition of this clinical entity may help to understand the electrophysiological behavior of SCN5A-related diseases and planning pre-implantation and follow-up strategies.