"Primum non nocere..." Explanting implantable cardioverter-defibrillators in patients with inherited arrhythmia syndromes - Case series

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Abstract

Introduction: Patients with Inherited arrhythmia syndromes (IAS) are at risk for life-threatening arrhythmias that may present as a sudden cardiac arrest (SCA) or a cardiac syncope. The benefit of Implantable cardioverter-defibrillators (ICD) in patients with IAS should be weighted against the risk for adverse events. This case-series describes patients diagnosed with an IAS, who were eventually reclassified in a low risk category and subsequently got their ICD explanted. **Methods:** We selected patients who, 1) were diagnosed with an IAS, 2) presented with either a documented arrhythmic event or syncope presumed to be arrhythmic syncope, 3) followed by an ICD implant, and 4) in whom we decided to explant the pulse generator. **Results:** Overall, eight patients fulfilled the inclusion criteria. Mean age at ICD implantation was 35±11.6 years. Mean length of ICD in situ was 4.9±3.6 years. None of the patients experienced a cardiac event. 2 patients presented with a presumed cardiac syncope, this diagnosis was rejected after guideline guided syncope evaluation including Tilt table testing. Alternative (pharmacological) therapy was started in six patients. **Conclusion:** These cases illustrate the importance of re-evaluating ICD therapy in patients with IAS. This should be integrated in standard clinical care, even in patients with IAS who survived a SCA and long term critical follow up is available. In patients with IAS presenting with an presumed cardiac syncope, extensive guideline guided syncope evaluation, can be of additional value when syncope event remains unknown

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