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Letter to the Editor

Prophylactic Implanted Cardioverter Defibrillator Implantation in Kearns-Sayre Syndrome Patients

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With interest we read the article by Imamura, T. et al (2018) about a 19 years old female with Kearns-Sayre syndrome (KSS) who received an implantable cardioverter defibrillator (ICD) after symptomatic ventricular arrhythmias and QT-prolongation (Imamura, T. et al 2018). We have the following comments and concerns.

A shortcoming of the study is that the mutation responsible for the phenotype was not provided. Though single mtDNA deletions are the most frequent cause of KSS, the syndrome may be due mitochondrial DNA (mtDNA) point mutations in some patients (Seneca, S. *et al.*, 2001). It is also crucial to know if the mother was genetically tested, since in 4% of the cases mtDNA deletions are transmitted via the maternal line (Poulton, J. *et al.*, 2017).

Since sudden death (SCD) in KSS may not only be due to ventricular arrhythmias, but also due to acute heart failure (Rossello, X. *et al.*, 2018), we should be informed about the echocardiographic findings at the time of the two syncopes. We also should know the pro-brain natriuretic protein (BNP) and troponin levels. It is also crucial to revise echocardiography for the presence of left ventricular hypertrabeculation, also known as noncompaction (LVNC), which can be a prominent feature of mitochondrial disorders and may be complicated by cardioembolism, heart failure, or ventricular arrhythmias including SCD (Captur, G., & Nihoyannopoulos, P. 2010).

Patients with KSS may also manifest with seizures, and generalised seizures may be associated with sudden unexplained death in epilepsy (SUDEP) and with Takotsubo syndrome. Since SUDEP is suspected due to ventricular arrhythmias induced by the seizure, we should know if the history of the index case was positive for epilepsy and if she was taking antiepileptic medication. Lastly, we should be informed about the electrolyte serum levels at the time of the ventricular arrhythmias.

In summary, this case could be more meaningful if the genetic background of the presented case and her first degree relatives was presented, if echocardiographic findings were presented, and if the history was completed for epilepsy and electrolyte disturbances.

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