Reviewer’s report

Title: Refractory ventricular fibrillations after surgical repair of atrial septal defects in a patient with CACNA1C gene mutations - Case report

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Reviewer: Johan Bos

Reviewer's report:

This case report by Kojima and colleagues presents the serious complications caused by a missed pre-operative diagnosis of long QT syndrome type 8 in a patient with multiple atrial septal defects and complete right bundle branch block. While QT prolongation was evident (albeit masked by RBBB) in the absence of QT prolonging medications or electrolyte imbalances, follow up on this ECG finding in preparation for an elective procedure was limited leading to a missed family history of (LQTS-associated) arrhythmias. Following the surgery, the patient experienced recurrent episodes of VF that needed CPR and multiple cardioversion. The authors highlight the importance of a careful family history, the difficulty of QTc assessment in setting of RBBB as well as evaluation of QT in general (review of causes other than congenital LQTS, such as electrolyte disturbances and QT prolonging medications.

Comments:

- Additional information on the CACNA1C mutation must be provided. Was the mutation novel or previously published? What is the variants ACMG classification? Is the variant seen in public exome databases such as ExAC or GnomAD?

- Was this variant identified in the family members that had reported arrhythmias?

- Ref#5 is not first to associate CACNA1C in pathogenesis of LQTS (https://www.ncbi.nlm.nih.gov/pubmed/23677916). Additionally, mutations in CACNA1C have been associated with QT prolongation, cardiomyopathy and congenital heart defects. This should be referenced as well (https://www.ncbi.nlm.nih.gov/pubmed/26253506).

- What are the post-operative risk in patients with LQTS and how do they relate to general risks after the procedures in this patient?

- Please provide normal values for potassium/magnesium. Reader now has to assume they were normal post-operatively. Also, what medications were used for anesthesia and post-operatively. Aby of these risk for QT prolongation?

- Line 43: Fredericia should be Fridericia
Level of interest
Please indicate how interesting you found the manuscript:

An article of importance in its field

Quality of written English
Please indicate the quality of language in the manuscript:

Not suitable for publication unless extensively edited

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