# RED GUMMY BEAR OR RED HERRING? WHEN CARDIAC ARREST ETIOLOGY ISN'T WHAT IT SEEMS

Close, Victoria, MD; Svetlana Duvidovich; Irfan Warsy, MD; Aalok Singh, MD

WMC Health | Maria Fareri Children's Hospital, Westchester Medical Center Health Network

Boston Children's Health Physicians, New York & Connecticut

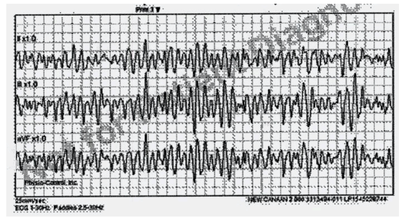
New York Medical College, Chartered 1860

## INTRODUCTION

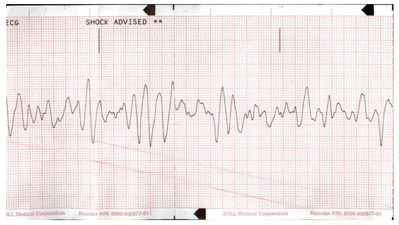
- We present a case of a 4-year-old-male with history of one prior cardiac arrest attributed to choking on a foreign body, later determined to have Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) after a second cardiac arrest prompted a genetic workup.

## CASE DESCRIPTION

- This is a single patient case study of a 4-year old male with two cardiac arrest events. Initial cardiac arrest was attributed to respiratory arrest due to story of possible choking on a red gummy bear.
- His rhythm at the scene showed coarse polymorphic ventricular tachycardia which reverted to normal sinus rhythm after multiple defibrillations.



- Approximately 3 months after the initial cardiac arrest, he was found unresponsive and pulseless. CPR was initiated and the patient was transported to our Emergency Department.
- The patient's cardiac rhythm showed polymorphic ventricular tachycardia, requiring multiple shocks and magnesium sulfate boluses for return of spontaneous circulation.



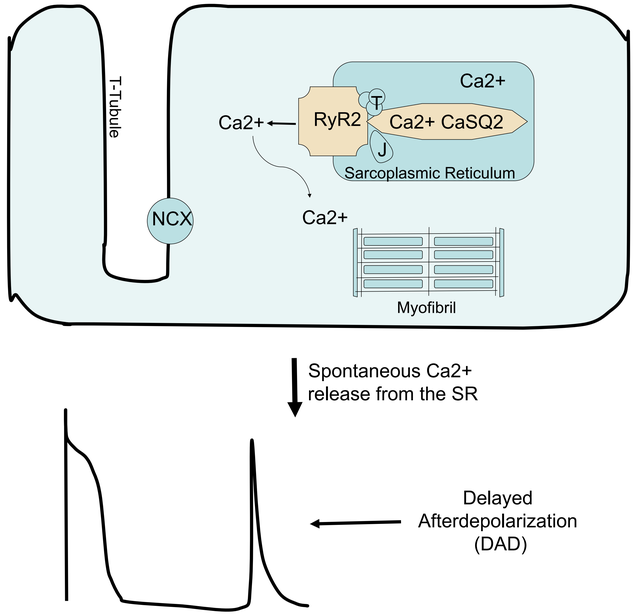

- Initial electrocardiogram demonstrated prolonged QTc that resolved the following day. Echocardiogram showed normal anatomy and function.

## CASE DESCRIPTION (CONT.)

- Telemetry showed intermittent ventricular ectopy, trigeminy, bigeminy, and couplets.
- A genetic workup revealed heterozygosity for a variant of the CASQ2 gene, c.538A>C (p.Lys180Gln), a variant of uncertain significance that is associated with CPVT.
- To prevent future cardiac arrests, a joint decision was made with the family for a sympathetic ganglion denervation.

## Pathology

Mutations in the RyR2 and CaSQ2 proteins lead to Ca leak from the sarcoplasmic reticulum in diastole, causing delayed afterdepolarizations




## DISCUSSION

- In patients without pre-existing cardiac conditions, the etiology of cardiac arrest remains unknown in half of all cases. Of these patients with initially unexplained cardiac arrest, a comprehensive workup identifies long QT syndrome and CPVT as the most common causes (Cunningham et al.)
- CPVT is a rare inheritable cardiac channelopathy which can result in arrhythmia and cardiac arrest in the setting of elevated emotional or physical stress, such as choking or near choking events. This may have occurred in our patient at the time of initial cardiac arrest.
- CPVT often presents in childhood and requires high clinical suspicion given its rarity as well as normal physical exam, echocardiogram, and resting electrocardiogram.
- A diagnosis of CPVT provides valuable benefit in guiding treatment and counseling families regarding genetic risk.
- Treatment can include a combination of beta blockers with or without sodium-channel blockers, implantable cardioverter defibrillator, and sympathetic ganglion denervation.

## CONCLUSION

- Standardized cardiac arrest workup including genetic testing in young children with initially unexplained cardiac events may play a valuable role in diagnosis and management.
- Even in seemingly straightforward cases, it is essential to avoid premature closure and to follow a standardized investigational approach including genetic testing.

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