

# Myotonic Dystrophy Disaster: An Outside Hospital Cardiac Arrest



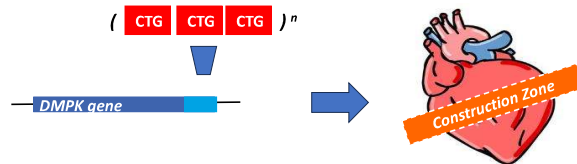
<sup>1</sup>Alexis Janoczkin DO, <sup>2</sup>Dominic Parfianowicz DO, <sup>3</sup>Daniel Makowski DO  
<sup>1</sup>Internal Medicine Residency Program, Lehigh Valley Health Network  
<sup>2</sup>Cardiovascular Disease Fellowship Program, Lehigh Valley Health Network  
<sup>3</sup>Department of Cardiology, Lehigh Valley Health Network



## Introduction

- Myotonic dystrophy (DM) is an inherited multisystem disorder characterized by progressive weakness, myotonia, and muscle wasting.
- 50% of patients can develop cardiopulmonary complications.
  - Cardiomyopathies, conduction and rhythm defects, early-onset heart failure
  - Arrhythmias = 2nd most common cause of death
- Current guidelines for DM patients only recommend implantable cardioverter defibrillator (ICD) placement post – VT/VF or EF < 35% patients despite medical therapy.

## Pathophysiology



## Case Presentation

46 YOF with outside hospital cardiac arrest

- 10-15 minutes without CPR
- 45-minute total downtime
- Initial rhythms per EMS: Ventricular tachycardia and ventricular fibrillation

**PMHx:** DM, recently new LBBB, QTc prolongation, thyroid cancer s/p thyroidectomy, obesity, multiple DVTs during pregnancy

**FHx:** mother with DM, own two children who died from DM complications

## Hospitalization

- Day 1:** Multiple vasopressor shock, ventilated, and targeted temperature management
- Day 3:** MRI without anoxic brain injury (Figure 2), started showing neurologic recovery
- Day 5:** Weaned off vasopressors
- Day 6 – 21:** Complications including arrhythmias, duodenal perforation, *C. diff* infection, heart failure exacerbation
- Day 22:** Tracheostomy and PEG tube placed
- Day 29:** Biventricular ICD placed
- Day 46:** Discharged to rehabilitation facility

## Imaging

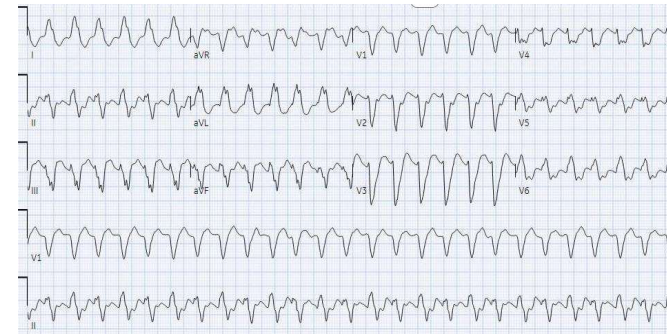


Figure 1: Initial EKG with wide complex tachycardia and previously seen LBBB.

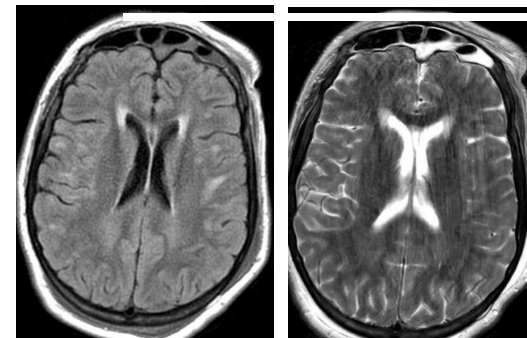


Figure 2: MRI T2 FLAIR 72 hours post-arrest with bilateral anterior encephalomalacia without evidence of diffuse anoxic injury.

## Discussion

- Current recommendations for cardiac death primary prevention in DM in the absence of cardiac abnormalities on invasive or non-invasive testing modalities remains a **gray area**.
- Patients should undergo **regular screening** with EKG and echocardiography for risk stratification and cardiac involvement monitoring
- DM care should include monitoring for cardiopulmonary, ocular, gynecologic / obstetric, endocrine, and musculoskeletal sequelae.

## References

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