Challenges of Long QT Syndrome in Pregnancy

Case Presentation

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History of Present Illness

- 27 year old G1P0 at 18 weeks gestation with history of Long QT Syndrome (LQTS) presented for routine follow up in Obstetrics Cardiology clinic.
- The patient denied any symptoms of syncope or palpitations but had a significant family history.
- Was on atenolol 50mg daily and refused placement of a defibrillator in the past.

Physical Exam

HR: 72 **BP:** 106/62 mmHg **Weight:** 89.2Kg **BMI:** 34 kg/m^2

General: no acute distress, able to lay flat without difficulty

Neck: no jugular venous distention, normal carotid upstroke

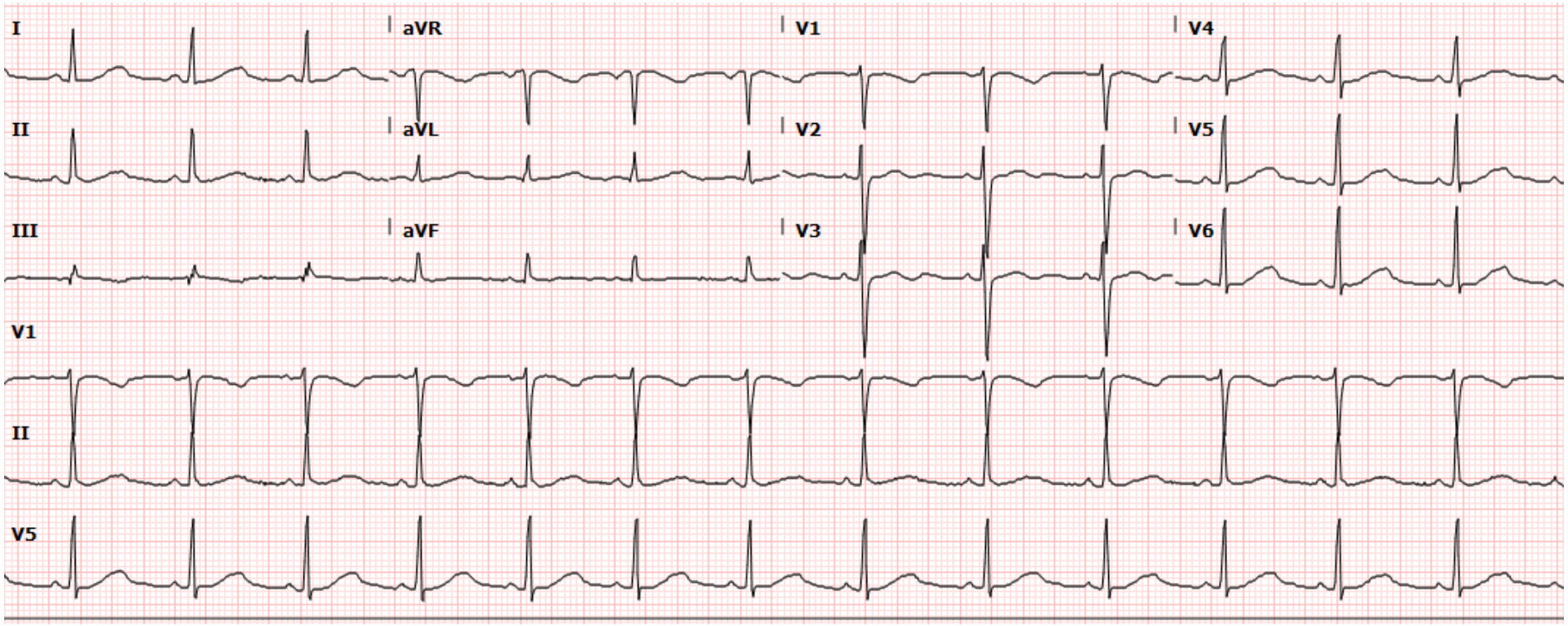
Cardiac: non displaced PMI, no right ventricular heave, normal s1, s2, no s3 or s4, no murmurs

Lungs: Clear to auscultation

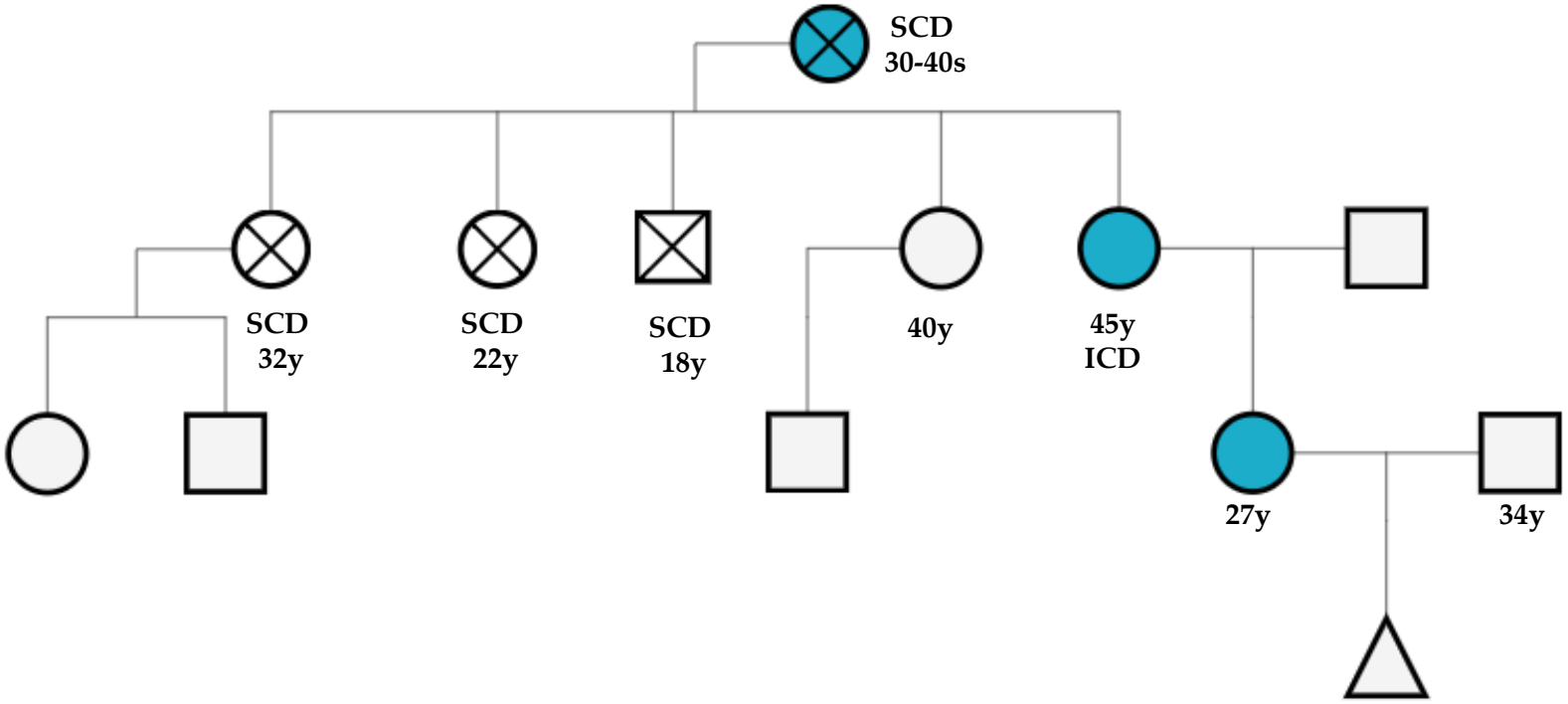
Abdomen: obese, gravid, non tender

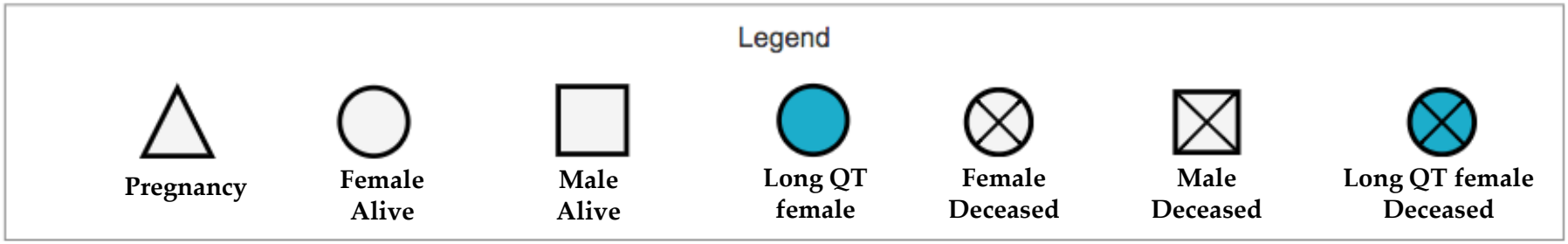
Extremities: no leg edema, 2+ distal pulses

ECG
QT/Qtc 440/514 ms



Family Genogram





Case Presentation

- She was referred for genetic testing and found to have heterzygous mutation of KCNH2 consistent with LQT2.
- She was switched to nadolol 80mg daily.
- A fetal echocardiogram was performed at 22 weeks gestation.

Fetal Echocardiography

- Situs Solitus
- HR: 134 beats/minute
- Normal four chamber with AV-VA concordance
- Normal pulmonic and systemic veins, normal ductal and aortic arches
- Normal right and left ventricular outflow tract

Clinical course

- Patient was diagnosed with gestational diabetes based on oral glucose challenge test 259mg/dl (normal <140mg/dl)
- Hemoglobin A1c 8.0%
- Started on an insulin regimen
- Started on aspirin 81mg daily
- Evaluated by electrophysiologist who recommended placement of a subcutaneous defibrillator after delivery

Clinical Course

- Patient presented to the emergency department at 36weeks 5days in active labor
- Decision was made to deliver in the emergency department
- A female infant was born by normal spontaneous vaginal delivery
- APGARS 9/9 weighing 2615g
- Baby sent for genetic testing found to be positive for the KCNH2 mutation

Clinical Questions

- Should she have an ICD?

2015 ESC Guidelines for the management of patients with ventricular arrhythmias

n of sudden cardiac death

ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.	I	B	436–438
Beta-blockers should be considered in carriers of a causative LQTS mutation and normal QT interval.	IIa	B	67
ICD implantation in addition to beta-blockers should be considered in LQTS patients who experienced syncope and/or VT while receiving an adequate dose of beta-blockers.	IIa	B	439

Implant of an ICD may be considered in addition to beta-blocker therapy in asymptomatic carriers of a pathogenic mutation in *KCNH2* or *SCN5A* when QTc is >500 ms.	IIb	C	67

Clinical Questions

- Should nadolol, which is the drug of choice for LQT2, be used in pregnancy?[1,2]

1. Ackerman, Michael J., et al. "Beta-blocker therapy for long QT syndrome and catecholaminergic polymorphic ventricular tachycardia: Are all beta-blockers equivalent?." *Heart rhythm*14.1 (2017): e41-e44.

2. Abu-Zeitone, Abeer, et al. "Efficacy of different beta-blockers in the treatment of long QT syndrome." *Journal of the American College of Cardiology* 64.13 (2014): 1352-1358.

Thank you