Anesthetic Planning for Delivery for a Patient with Titin-Truncating Variant Mutation Complicated by Complex Arrhythmia

Kiana C. Nguyen BS, Elliott Callahan MD, Peter Yeh MD

INTRODUCTION

- **Titin (TTN)**
  - Sarcomere organization
  - Striated muscle function
- **TTN-truncating variants (TTNtvs)**
  - Genetic dilated cardiomyopathy[1]
  - Peripartum cardiomyopathy[1]
  - Early-onset arrhythmia[2]

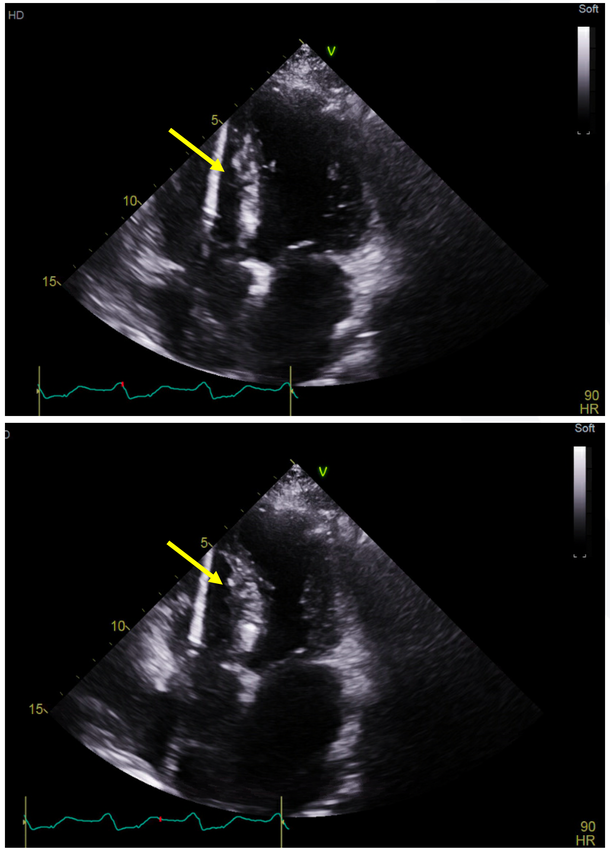


Figure 1. 30w TTE, apical 4 chamber view. End-diastole (above) and end-systole (below). Pacer/ICD lead indicated with arrow.

CASE

- **27 yo G3P0 at 33w3d**
  - TTNtv c.87852G>A (p.Trp29284*)
- **PMH**:
  - Sudden cardiac arrest, age 16
  - Atrial fibrillation
  - AICD placed
- **MEDS**: quinidine, cilostazol, metoprolol
- **mWHO class II-III, CARPREG II score 5**

Anesthetic Planning for Delivery for a Patient with Titin-Truncating Variant Mutation Complicated by Complex Arrhythmia

Kiana C. Nguyen BS, Elliott Callahan MD, Peter Yeh MD

DELIVERY PLAN

Discontinue cilostazol 48 hrs prior to neuraxial anesthesia

Monitor in ICU while holding cilostazol

Manage potential breakthrough arrhythmia with isoproterenol

IOL at 39 weeks with neuraxial anesthesia

LABOR COURSE

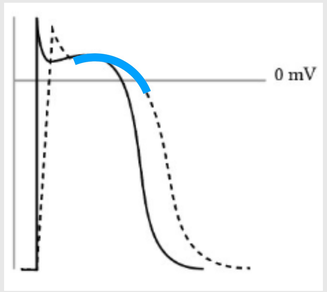
- Patient p/w SROM at 38w4d
- Insufficient window off cilostazol
  - Thus, no neuraxial anesthesia
- Pt received remifentanil with pudendal block for 2nd stage analgesia
- Patient had an uncomplicated delivery
  - No episodes of arrhythmia

Anesthetic Planning for Delivery for a Patient with Titin-Truncating Variant Mutation Complicated by Complex Arrhythmia

Kiana C. Nguyen BS, Elliott Callahan MD, Peter Yeh MD

DISCUSSION

Cilostazol



- PDE-3 inhibitor
- Anti-arrhythmic
- Inhibits platelet aggregation
- Contraindication to neuraxial procedures

Early epidural in cardiac disease

- Avoid sympathetic surges associated with uterine contractions
- May help prevent precipitating malignant arrhythmia during labor

CONCLUSION

Pregnancy is a cardiac stressor!

- "Second hit" in TTNtv patients at risk for PPCM & malignant arrhythmia[1]

Risk stratification

- Cardiac MRI assessing for midwall fibrosis is a known predictor of arrhythmic events and need for device therapy in DCM.[3]
- Explore future roles for cardiac MRI in PPCM

REFERENCES

1. Goli R, Li J, Brandimarto J, et al. Genetic and Phenotypic Landscape of Peripartum Cardiomyopathy. *Circulation*. 2021;143(19):1852-1862.
2. Choi SH, Weng LC, Roselli C, et al. Association Between Titin Loss-of-Function Variants and Early-Onset Atrial Fibrillation. *JAMA*. 2018;320(22):2354-2364.
3. Corden B, Jarman J, Whiffin N, et al. Association of Titin-Truncating Genetic Variants With Life-threatening Cardiac Arrhythmias in Patients With Dilated Cardiomyopathy and Implanted Defibrillators. *JAMA Netw Open*. 2019;2(6):e196520.