

Treating Postpartum Refractory Long QT Syndrome With Cervical Ganglion Sympathectomy

Nikhil A. Mehta MBBS

Lehigh Valley Health Network, Nikhil.Mehta@lvhn.org

Mario Caruso DO

Lehigh Valley Health Network, Mario.Caruso@lvhn.org

Syed Rafay Ali Sabzwari MD

Lehigh Valley Health Network, Rafay.Sabzwari@lvhn.org

Lohit Garg MD

Lehigh Valley Health Network, lohit.garg@lvhn.org

Jeffrey Gordon MD

Lehigh Valley Health Network, Jeffrey.Gordon@lvhn.org

See next page for additional authors

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Authors

Nikhil A. Mehta MBBS, Mario Caruso DO, Syed Rafay Ali Sabzwari MD, Lohit Garg MD, Jeffrey Gordon MD, and Talha Nazir MD

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Nikhil Mehta, MBBS, Mario Caruso, DO, Syed Rafay Ali Sabzwari, MD, Lohit Garg MD, Jeffrey Gordon, MD, Talha Nazir, MD
Lehigh Valley Health Network, Allentown, Pa.

BACKGROUND

- Congenital long QT syndrome (LQTS) type 2 is associated with an increased risk of ventricular arrhythmias and sudden cardiac death.
- Risk is increased in the postpartum period, and treatment consists of beta-blockers and ICDs in high risk patients.
- Left cervical ganglion sympathectomy can be considered in cases refractory to beta-blocker therapy.

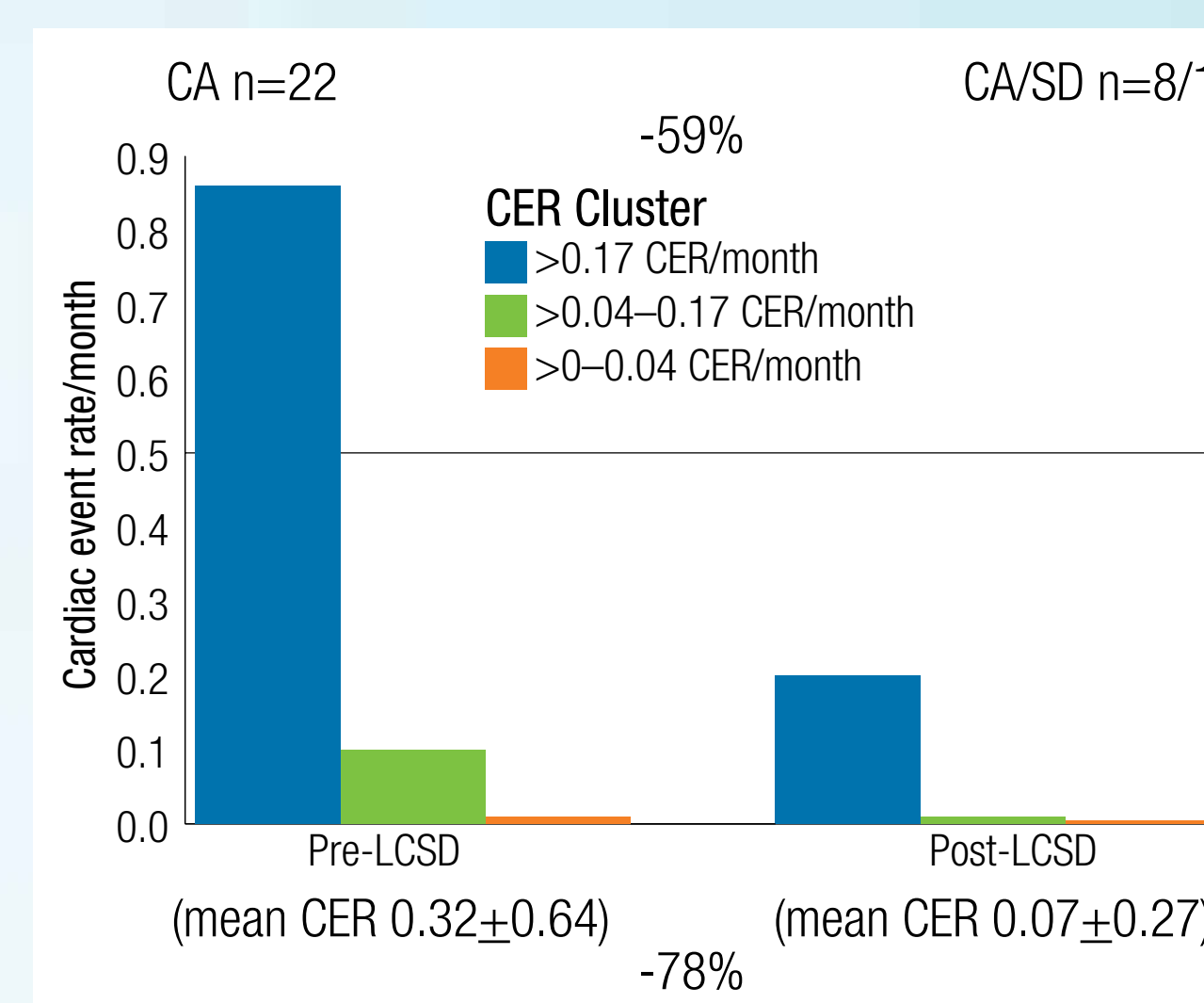
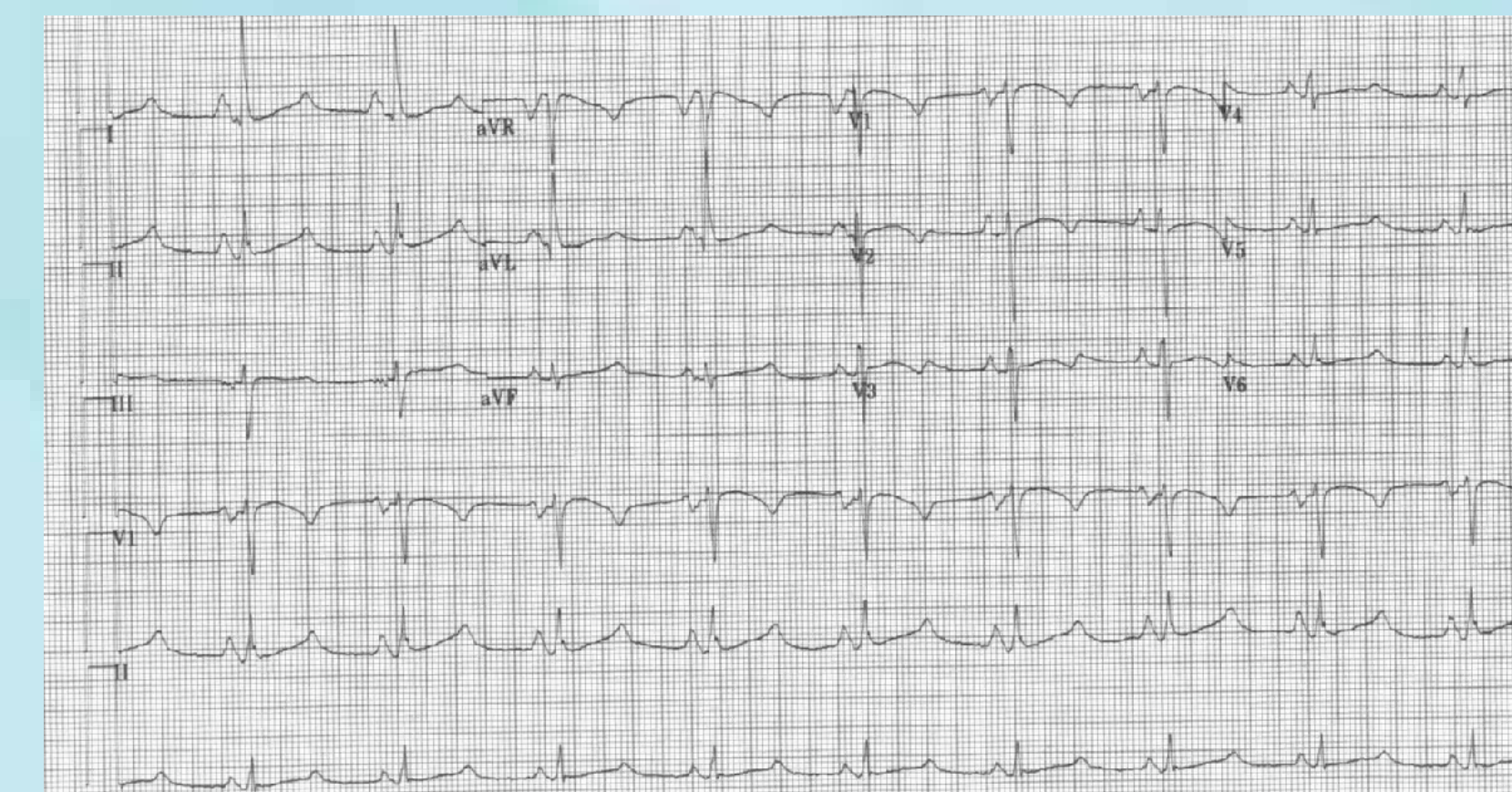
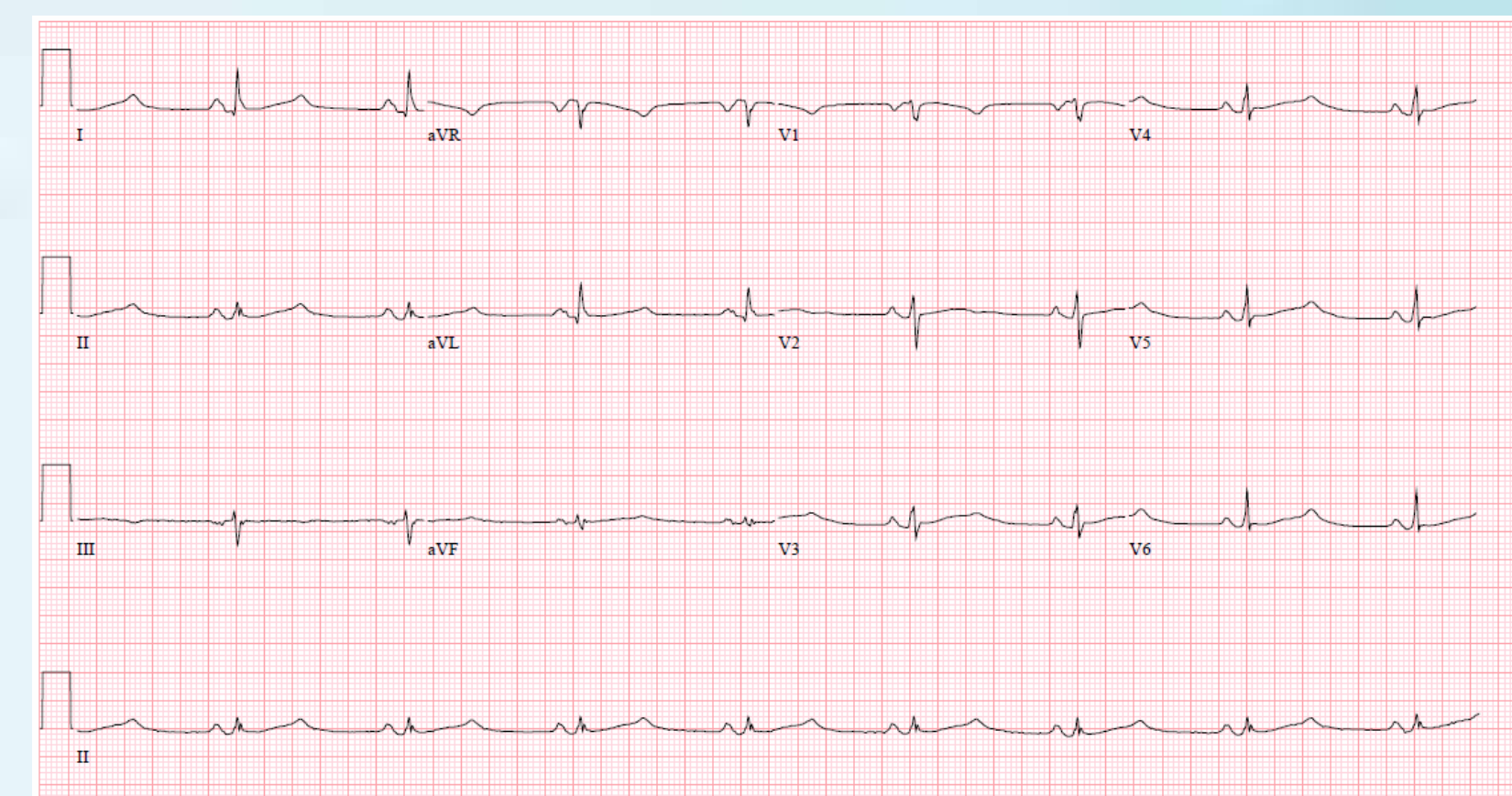
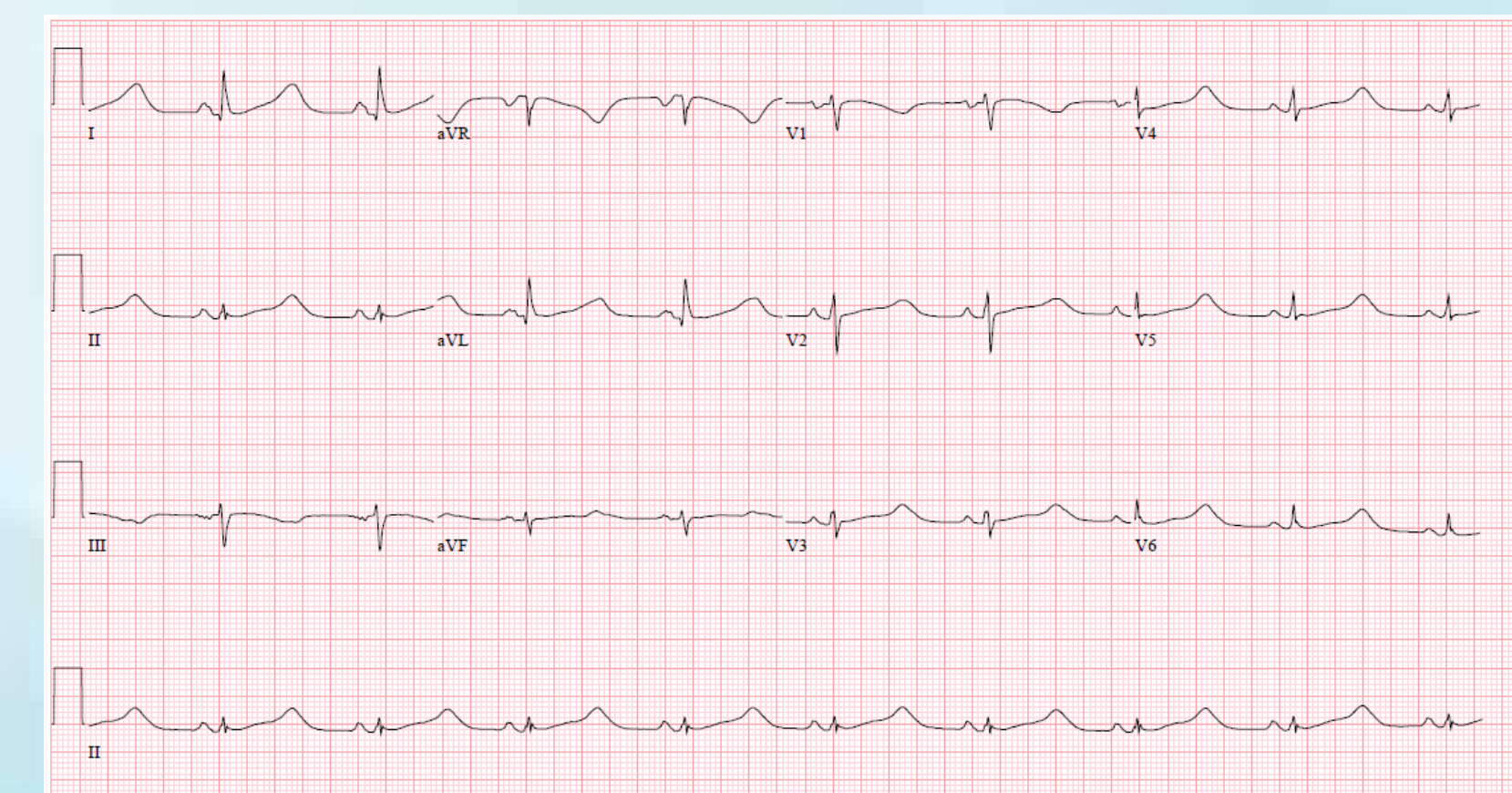
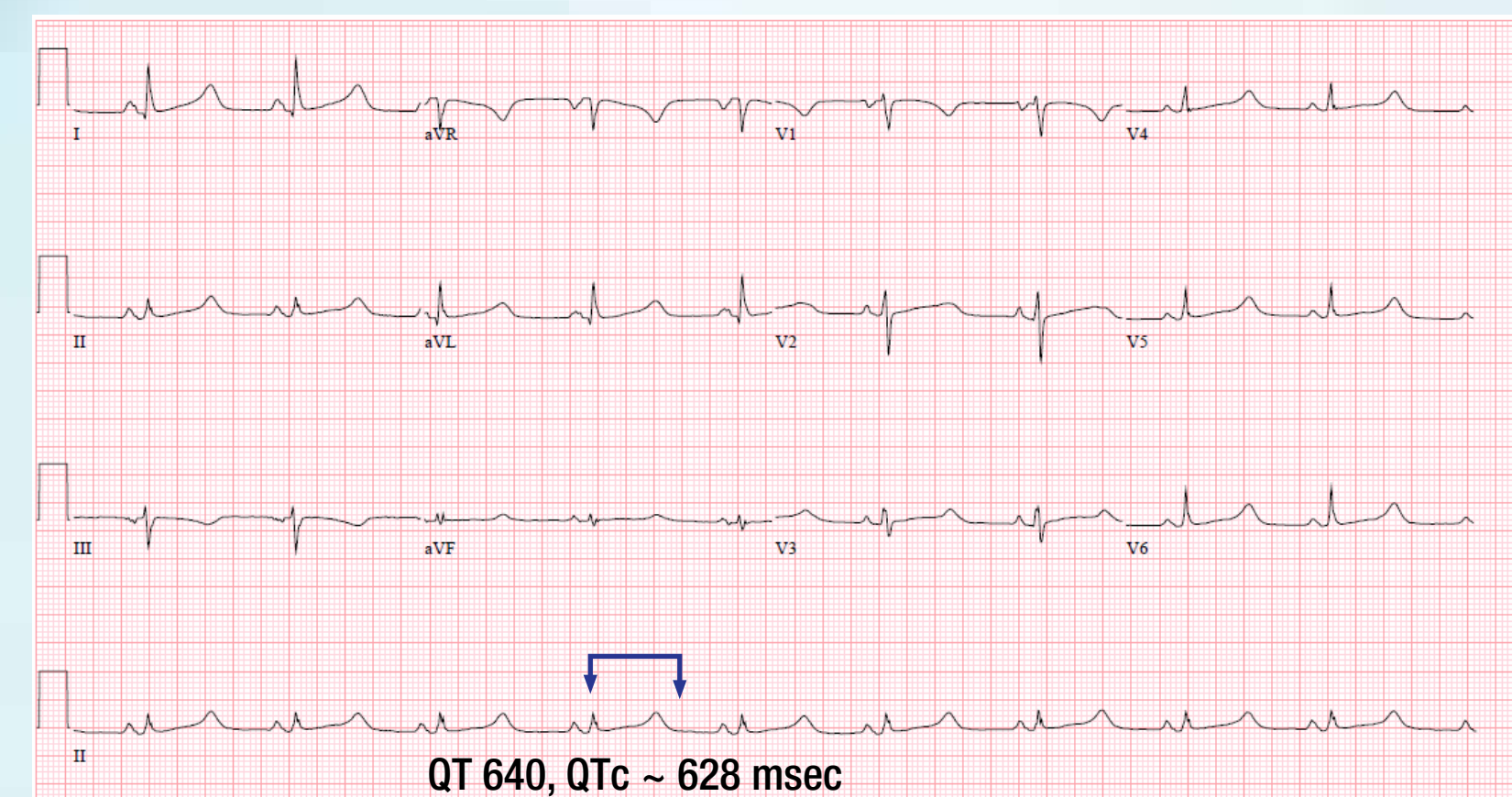
CASE SUMMARY

INITIAL PRESENTATION:

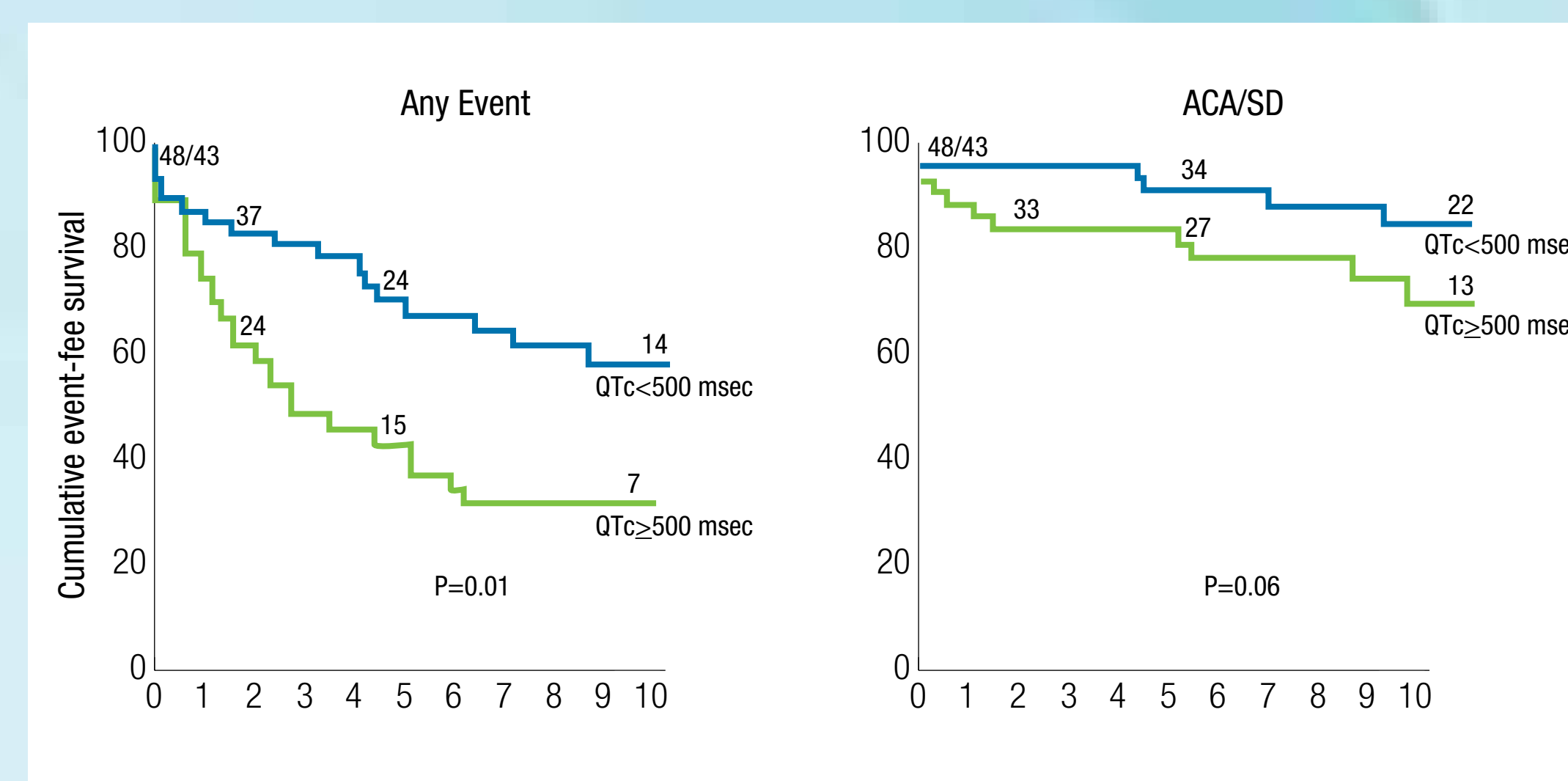
- A 36 year old woman (G5P0313) one month postpartum with history of LQTS type 2 presented to the ER with syncope and ICD shock from polymorphic ventricular tachycardia (PMVT). Her 4 children all have LQTS type 2 with class 1 KCNH2 mutation.
- During her last pregnancy, she was found to have significant QTc prolongation to 628 msec for which she underwent subcutaneous ICD implantation. [ECG 1]
- She was admitted and treated with increasing doses of beta-blocker (nadolol) until episodes of PMVT subsided, and was finally discharged to outpatient follow up. Episodes attributed to increased anxiety and her postpartum state.

SUBSEQUENT PRESENTATION:

- Six months postpartum, she came back with another episode of syncope and ICD shock due to PMVT, QTc markedly prolonged to 660 msec. [ECG 2]
- By now she was taking maximum doses of beta-blocker (Nadolol), and an anxiolytic, Valium.



Effect of sympathectomy on cardiac event rates (CER) in 78 patients under β -blocker therapy before and after procedure. [1]



Kaplan-Meier curves of event-free survival and survival according to post-procedure QTc. [1]

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DECISION-MAKING

- Since our patient was 6 months postpartum, markedly anxious, and having recurrent PMVT with ICD shocks despite maximum doses of nadolol, we elected to pursue left T2-T4 cervical and lower stellate ganglion sympathectomy.
- This interrupts the release of norepinephrine in the heart, thereby reducing symptoms and shortening the QT.
- Sympathectomy has been shown to reduce the incidence of aborted cardiac arrest and syncope, with mean yearly events per patient dropping by as much as 91%. [1, 2] A post procedure QTc <500 ms predicts low risk of future events.
- Current guidelines recommend sympathectomy for patients with LQTS in whom: (I) ICD therapy is contraindicated or refused (Ic), (II) β -blockers are either not effective, not tolerated, or contraindicated (Ic), or (III) patients with breakthrough events despite β -blockers/ICD (IIa). [3]
- She successfully underwent the procedure without any complications. [ECG 3]
- At 12-month follow up, QTc measured 510 msec. [ECG 4]

CONCLUSION

- This case highlights the management strategies for ventricular arrhythmias in patients with LQTS type 2 in the postpartum period.
- When patients experience recurrent malignant arrhythmias despite β -blocker use, left cervical sympathectomy could be considered to prevent future events.

DISCLOSURES

Nikhil Mehta and Talha Nazir – No disclosures
Contact Info: nikhil.mehta@lvhn.org