Treating Postpartum Refractory Long QT Syndrome With Cervical Ganglion Sympathectomy

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Published In/Presented At


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Treating Postpartum Refractory Long QT Syndrome With Cervical Ganglion Sympathectomy

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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.

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.

REFERENCES