Left Cardiac Sympathectomy Denervation in Patient with LQTS Unresponsive to Beta-blockers and Cardiac Pacing

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BACKGROUND

Left cardiac sympathetic denervation (LCSD) has been proposed for treating channelopathies such as congenital long QT syndrome in which betablockers as first line therapy has not been successful. An implantable cardioverter defibrillator (ICD) is appropriate for patients at high-risk or refractory to medical treatment. However, it cannot prevent recurrence of ventricular arrhythmias,

CASE

A 5-year-old female presented with generalized muscle weakness and history of nonspecific normokalemic channelopathy. She had a brother who died at age 4 from rhabdomyolysis.

Bradycardia was seen, and an EKG showed QTc of 487ms, on the sixth in-hospital day. Rhythm Holter reported long QTc, T wave electric alternation and 14 seconds of ventricular fibrillation.

Beta-blocker propranolol is started (0.6mg/kg/day). We placed a temporary pacemaker with HR of 70 bpm, then, definitive pacemaker at HR 90 bpm programed in AAI mode. Despite this, electrical instability manifested by VT and T wave electric alternation continued, finally VF started, it was resolved with CPR. Then she had left lateral sympathectomy with video-assisted thoracoscopic surgery (VATS) as rescue treatment, without VT or VF recurrence. The patient however, died of complications associated with mechanical ventilation..

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Quick Decision making by heart team is crucial for our patients at risk of sudden cardiac death.

They can be treated with Beta-blockers, ***every so often*** they can not work.

Cardiac pacing can be an option, so it avoids bradycardia and QT prolongation.

Left cardiac sympathectomy denervation terminates electrical instability as a second and ***definitive line treatment.***

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DECISION-MAKING:

The incessant VT that degenerated to VF despite the pacemaker and beta-blocker treatment led the heart team decided the LCSD as a salvage measure with VATS, a minimally invasive procedure, due to imminent risk of sudden cardiac death; a decision also supported by comorbidities and unresponsiveness to medical treatment and pacemaker. LCSD with VATS has been successful in observational studies with significant decrease in mortality.

CONCLUSION

There are few described cases of LCSD in congenital long QT syndrome. LCSD demonstrates the total resolution of the lethal arrhythmic episodes so this practice could be extended because of the safety and effectiveness of this procedure. In this case, we certainly show that this surgical procedure is effective in a patient with significant comorbidities with poor prognosis in which all the first-line therapeutic alternatives had failed.

FIGURE 1

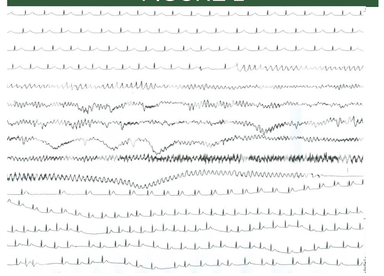

Figure 1. Torsades de pointes and Ventricular fibrillation, start and termination

FIGURE 2

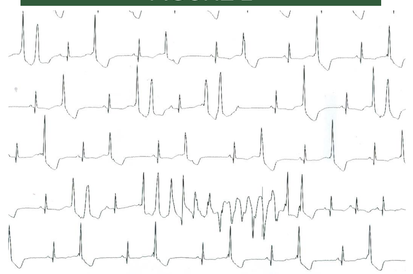

Figure 2. T wave alternation (Electrical instability)

FIGURE 3

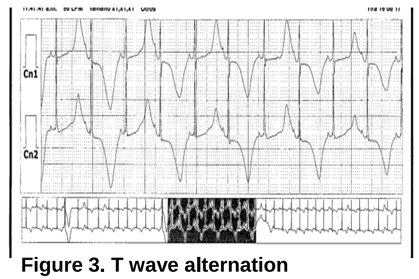

Figure 3. T wave alternation

FIGURE 4

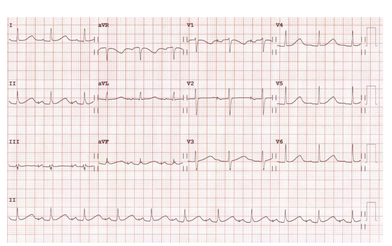

Figure 4. Atrial pacing