

Thoracoscopic Left Cardiac Sympathetic Denervation for the Treatment of Congenital Long QT Syndrome

To the Editor:

Congenital long QT syndrome (LQTS) is a hereditary disease characterized by QT prolongation and syncope due to malignant cardiac arrhythmia, especially *torsade de pointes*, and sudden death from ventricular fibrillation. The syncope is usually triggered by an increase in sympathetic activity. Management of LQTS includes treatment with beta blockers, a permanent atrial pacemaker, an implantable defibrillator, or left cardiac sympathetic denervation (LCSD). Using thoracoscopy for LCSD reduces any associated side effects to a minimum.¹⁻³

The patient was a 12-year-old girl with no personal or family history of note. She was referred to our hospital after five episodes of syncope in the previous month. The first episode occurred after physical exercise, the next two while she was resting, and the last two while she was being monitored at the other hospital. She had QT prolongation and her syncope was secondary to *torsade de pointes* polymorphic ventricular tachycardia. Treatment was started with esmolol, magnesium and AAI stimulation. Arrhythmia occurred on

changing to oral propranolol after 48 h. On admission physical examination was normal, the ECG showed sinus bradycardia at 48 bpm, QT of 600 ms and QTc of 540 ms (Figure 1a). LCSD was undertaken via a single 11 mm incision in the lower part of the left axilla through the fourth intercostal space. A 10 mm 0°-angled lens with a 5 mm working channel (Hopkins 26038AA, K. Storz-Endoskope, Tuttlingen, Germany) was used to visualize, in the upper left hemithorax, the stellate ganglion and sympathetic dilatations of the second to fifth thoracic nerve roots, situated subpleurally in the head of the ribs. With a monopolar coagulation hook the nervous dilatations of these ganglia were electrocoagulated until sectioned and the lower two-thirds of the stellate ganglion resected. There were no complications. Electrocardiogram after the procedure showed sinus rhythm of 72 bpm, QT of 480 ms and QTc of 500 ms (Figure 1b). The patient was discharged three days after the operation under treatment with 40 mg/d nadolol, which had been used instead of propranolol since her admission. Three months later, she was asymptomatic except for mild left palpebral ptosis.

Congenital LQTS is a hereditary genetic disorder characterized by episodes of syncope secondary to *torsade de pointes* ventricular arrhythmia and sudden death due to ventricular fibrillation.⁴ The mortality rate in untreated symptomatic patients is estimated to be 70%.⁵ Syncope is frequently triggered by physical or emotional stress, and clinical and experimental studies have demonstrated the role of sympathetic



Fig. 1a.

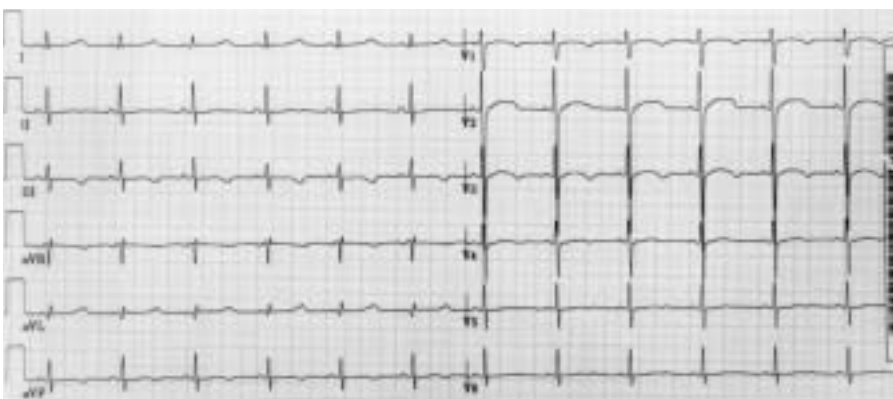


Fig. 1b.

stimulation in the onset of syncope. LCSD prevents the onset of fatal arrhythmia in patients with LQTS not only by suppressing the trigger but also by modifying the substrate, as QT dispersion, a marker of electrical instability, is reduced.⁶ The first surgical LCSD was performed in 1969 by Moss and in 1996 Chen undertook LCSD via a left thoracoscopy following his experience in patients with hyperhidrosis. This technique has since become popular due to its simplicity, low rate of morbidity and short hospital stay.^{1-3,7}

The treatment of choice in patients with symptomatic LQTS is drug therapy with beta blockers. If these are not beneficial, as in our case, LCSD is indicated because of the good results reported with this technique. Side effects of LCSD include Horner syndrome, though this is usually mild and transitory. Clear improvement was noted in our patient during follow-up.

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