Congenital long QT syndrome treated by renal sympathetic denervation

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A 14-year-old girl was diagnosed as congenital long QT2 syndrome presented with recurrent syncope despite the use of beta-blocker. Her genetic analysis revealed a de novo KCNH2 gene mutation (c.G1714A). Holter monitor showed significantly prolonged corrected QT interval (QTc) of about 600 ms and frequent multiform premature ventricular beats (PVCs) (Figure 1A). Although left cardiac sympathetic denervation (LCSD) is recommended to more effectively inhibit sympathetic activity, the related complications could not be ignored. As a less invasive treatment to achieve intense anti-sympathetic effect, renal sympathetic denervation (RDN) was performed after getting the informed consent of her parents. Operation was performed under general anaesthesia. High-frequency stimulation (HFS) (10 Hz, 15 mA,
 Pulse width 2 ms) was performed from distal to proximal renal artery for a maximum of 60 s in each site, during which blood pressure elevation and ventricular arrhythmias (VAs) were induced was defined as positive response. In four positive sites (three on the left and one on the right renal artery), frequent multiform PVCs the same morphologies as preoperation were induced following the end of HFS (Figure 1B, denoted by asterisks). After ablation, no PVCs could be induced anymore at these positive sites. The QTc got significantly shortened from 600 ms before RDN to 500 ms 1 month after RDN (Figure 1A). The patient was syncope-free in the follow-up of 5 months, and no VA was observed in two consecutive Holter monitor.

It is commonly recognized that most cardiac events of VAs are related to sympathetic activation. While renal sympathetic efferent and afferent nerves are important mediators of cardiac and whole-body sympathetic activity. In a cesium-induced long QT canine model, renal sympathetic stimulation was revealed to promote left stellate ganglion (LSG) activity, which could decrease VAs threshold. In contrast, RDN significantly reduced whole-body norepinephrine spillover and LSG activity, which may exert protective effects on VAs. In a recent international multicentre research, RDN was demonstrated to significantly reduce VA burden in heart failure patients presented with refractory VA. In addition, a worldwide research containing 147 long QT syndrome (LQTS) patients observed QTc got significantly shortened by an average of 39 ms 6 months after LCSD, and a post-LCSD QTc < 500 ms predicted a very low risk of recurrent symptoms.

In this case, we introduced RDN as a potential technique for the treatment of congenital LQTS, and the VAs induced by HFS may be a crucial index to determine ablation target sites. The role of RDN in the treatment of LQTS needs to be confirmed by more studies in the future.

**Conflict of interest:** none declared.

**References**


