A 26-year-old woman with congenital deafness and a lifelong “seizure disorder” presented to the emergency department with 15 episodes of syncope. She was 1 week postpartum. The ECG showed normal sinus rhythm, markedly prolonged QT interval (900 ms), and remarkably large and broad T waves (Figure). In the emergency room, the patient had another syncopal episode associated with tonic-clonic activity, and monitoring showed polymorphic ventricular tachycardia (torsade de pointes), which terminated spontaneously. The patient was started on a β-blocker and underwent implantation of a cardioverter-defibrillator.

The diagnosis of Jervell-Lange-Nielsen syndrome, the autosomal recessive form of the long-QT syndrome associated with congenital deafness, was confirmed by identification of 2 different mutations in the potassium channel gene KCNQ1 (KvLQT1), resulting in A341V and K362R; one was a de novo mutation, and the other was inherited from her father. The peripartum period is one of increased arrhythmia risk in this syndrome. Marked QT prolongation is characteristic, but the remarkably large-amplitude T waves shown here are unusual.
Himalayan T Waves in the Congenital Long-QT Syndrome
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