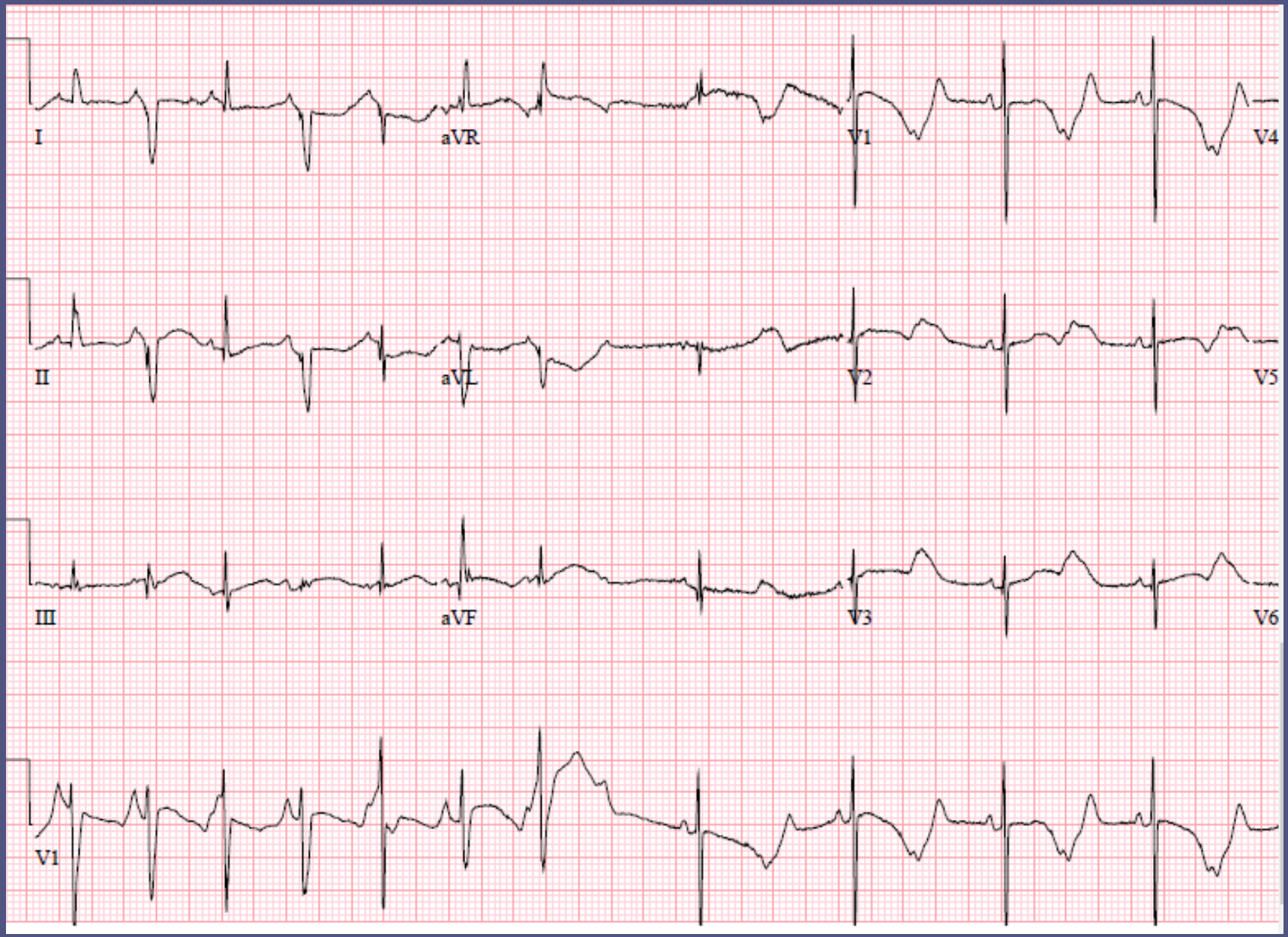
Case: Long QT 3

- Pregnancy complicated at 37 weeks gestational age by irregular heart beat
- C section due to concerns for non-reassuring fetal heart rate.
- Initial EKG showed normal sinus rhythm with markedly prolonged QTc at 650 msec.

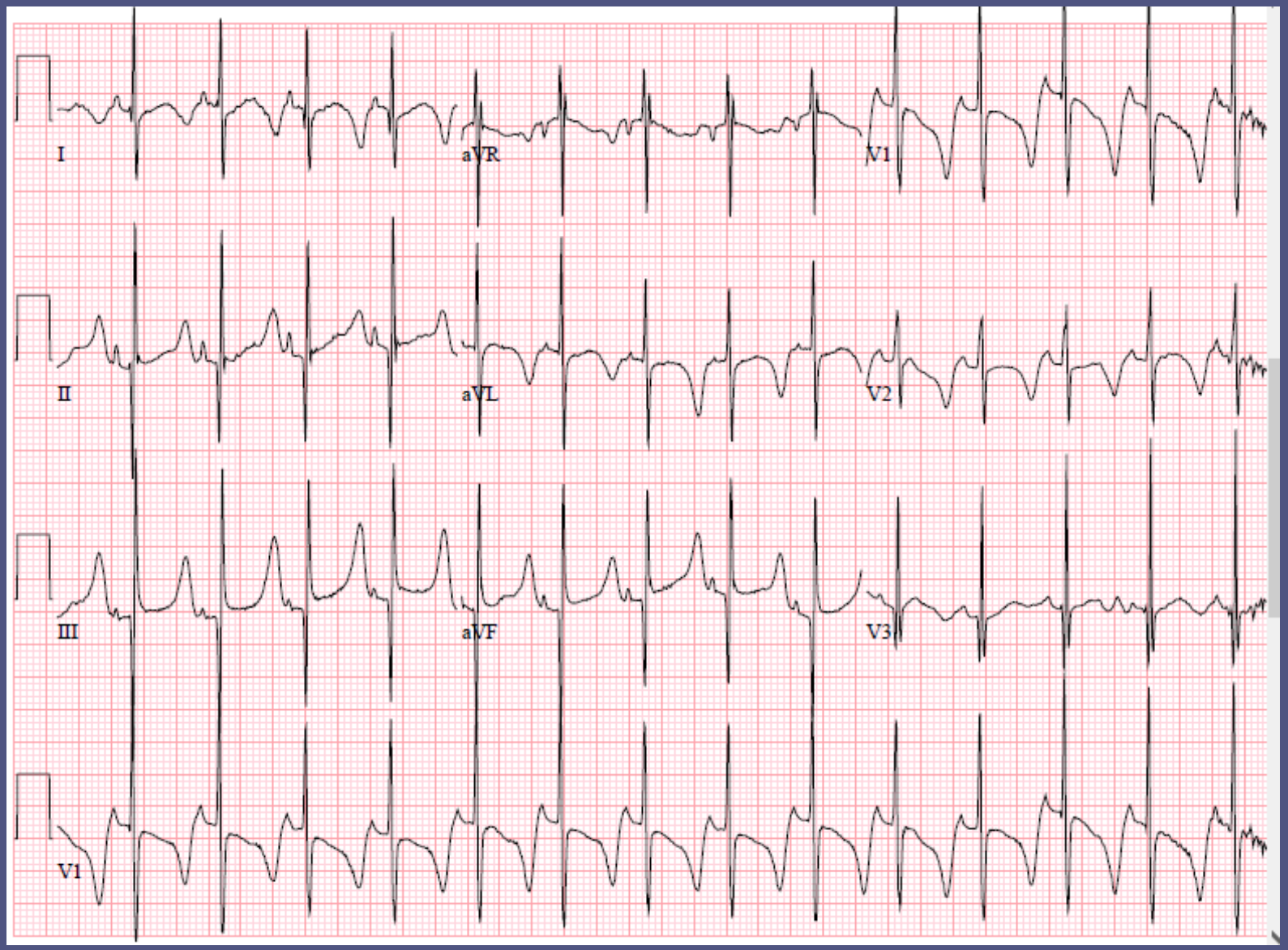
Newborn ECG



Clinical course

- In the first 2 days of life, he had repeated episodes of 2:1 AV block and multiple brief episodes of torsade de pointes
- After starting propranolol and repleting Mg^{++} and K^{+}, torsade abated and conduction went to 1:1
- Discharged on CR monitor and with an AED
- Genetic testing: long QT3 mutation in SCN5A at V1763M, a mutation associated with a severe phenotype of LQT3
- Mexiletine added
- ICD implanted at 1 year (bipolar RV sense/pacing lead, transverse ICD coil, abdominal generator)

Current ECG



Clinical course

- Single appropriate ICD discharge, repeated non-sustained torsade
- Multiple admissions for hypoglycemia, sometimes with seizures, often associated with not being fed regularly
- Propranolol decreased to ~2 mg/kg/day
- Mexiletine levels low and dose increased most recently to 80 mg 3 times daily (current weight 17 kg)
- CYP2D6 metabolizer testing for rapid mexiletine metabolism negative
- Last ICD interrogation showed no further recorded torsade

Question

- Consider stellate gangliectomy, not a proven therapy in long QT 3?
- Hold the course?
- Any other suggestions?