Ventricular fibrillation in a girl with heterozygous SCN5A V1777M mutation presented bradycardia dependent prolonged QT interval and exercise-provoked two-to-one atrioventricular block: Case report.

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**Introduction**: Two-to-one atrioventricular block (2:1 AVB) are usually observed in severe phenotypes of congenital long QT syndromes in neonatal periods.

**Methods**: An 11-year-old girl suffered from ventricular fibrillation during swimming. Resting ECG was almost normal. Treadmill test provoked 2:1 AVB without prolongations of QT interval. Holter monitoring showed marked prolongation of QT interval while asleep (HR 49bpm, QT 584ms, QTcB 524ms) and no evidence of any grade of AVB. Heterozygote of SCN5A V1777M missense mutation was identified. After administration of propranolol and mexiletine, exercise-provoked AVB disappeared accompanied by suppression of maximum atrial rate.

**Result**: Lupoglazoff et al. reported a young boy with homozygous SCN5A V1777M mutation who presented 2:1 AVB associated with severe prolongation of QT interval. They also showed patient’s electrophysiological studies confirmed the location of the block was in the His-Purkinje system rather than at the ventricular level.

**Conclusion**: We speculated that 2:1 AVB in our patient related to a potential disturbance in the cardiac conduction system.