**Title:** **Severe genetic arrhythmia (CPVT) studied with iPSC-cardiac model derived from pediatric patients**

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**Abstract**
Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a genetic cardiac condition causing severe risk for sudden cardiac death. Structure of a patient's heart is normal, and no arrhythmias are detected at rest. When the heart rate increases due to emotional stress or exercise severe arrhythmias occur causing symptoms like loss of consciousness, and even sudden death. Approximately 30% of the patients receive symptoms before age of 10 and prognosis of death before age of 30 is 30-35%.
We have derived iPSC lines from the blood samples of six patients from 10 to 18 years old carrying severe form of the CPVT. These iPSC lines are differentiated into cardiomyocytes and their electrical properties are measured with calcium imaging method. We have studied calcium release from the calcium storages which is essential for beating of the cardiomyocytes. At the moment we are optimizing increasement of the beat rate so we could observe electrical events on a moment where beat rate starts to elevate. As the severe clinical phenotype of the patients predicts there are also challenges in a cell model when the beat rate starts to increase. Previously we have studied cardiomyocytes derived from adult CPVT patients in 3D bioprinted cardiac structures which showed normal functionality, $Ca^{2+}$ handling properties and disease phenotypic response to adrenaline treatment. This cardiac iPSC model could be used to determine why the clinical phenotype, especially with these patients, is so life threatening and in addition to test drugs to treat arrhythmias caused by increased beat rate.