Arrhythmia management in pregnant patient with Andersen-Tawil syndrome

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Introduction: Andersen-Tawil syndrome (ATS) is a rare inherited disease characterized by ventricular arrhythmias (VAs), periodic paralysis, and dysmorphic features. In the condition with pregnancy and childbirth, there is no guideline for providing effective treatment to suppress VAs.

Methods: We report a case detailing the management of a pregnant patient with ATS safely undergoing childbirth.

Result: A 32-year-old Thai woman came to seek an advice regarding to planning for pregnancy. She had a history of abnormal cardiac rhythm detected by doctor without symptoms, ECG was evaluated and revealed multiform PVCs and a non-sustained polymorphic VT. Her sister and mother had a similar history of abnormal cardiac rhythm, however, there was no history of sudden cardiac arrest in her family. Her sister was fine without any medical attention and her mother also delivered totally 5 children without adverse event. She denied her psychological problem. Physical examination revealed only micrognathia. At that time, Andersen-Tawil syndrome was suspected according to her unique feature of VAs, autosomal dominant inheritance, and minor dysmorphic feature. Genetic testing confirmed the missense mutations of KCNJ2 with novel mutations including c.557C>G (p.Pro186Arg) and c.436G>C (p.Gly146Arg) in our patient, her sister, and mother. Choice of treatment had been discussed with the patient. Catheter ablation was offered but she denied due to financial issue and no symptom, so medical treatments were given. Firstly, propranolol was given at dose 30 mg daily but the patient was poorly tolerated due to fatigue then it was switched to metoprolol which are well tolerate up to dose of 100 mg daily, however, there was no difference in her VAs assessed by physical examination and random ECG. Eventually, the patient decided to begin pregnancy without taking any medication despite extensive discussion about risk of adverse cardiac event due to her ventricular dysfunction, VAs and a chance of inheritance to her child. During pregnancy, VAs became less frequent. Due to contracted pelvis, we planned elective cesarean section as a route of delivery. However, the labor pain occurred spontaneously together with irregular fetal heart beat, an emergency low transverse cesarean section was performed under spinal anesthesia. She had no episode of malignant VAs which need to be treated with additional treatment. The patient and baby were doing well postdelivery. After delivery, she developed multidirectional VT all the time so we decided to start flecainide 100 mg per day as an additional therapy. 5 months after pregnancy, ECG showed polymorphic PVCs without VT.

Conclusion: Combination between flecainide and metoprolol effectively suppressed VAs in patient with ATS especially when were used during pregnancy and childbirth.