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Prolonged QTc and Hypokalemia: A Bad Combination Before Surgery

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A 34-year-old female with a past medical history of asthma presented to our perioperative medical center for medical clearance for carpal tunnel release surgery. The patient is a schoolteacher and reported frequent episodes of palpitations and syncope for a few years alternating with painful, tetanic muscle spasms followed by flaccid paralysis. Her syncopal episodes were short and she usually regained consciousness in a few minutes. The patient's mother never witnessed seizure activity during any of her episodes. She had been seeing a neurologist and a cardiologist in her hometown, but they had not been able to figure out the cause of these symptoms. Her physical examination was completely normal and her vitals stable. Her blood work showed hypokalemia of 3.1 mEq/L. Electrocardiogram showed prolonged QT/QTc.

Because of her syncope and prolonged QTc, the patient was referred to cardiology for further work-up. Exercise stress testing was done and she developed ventricular arrhythmias that terminated the test. She was at risk for sudden cardiac death, so an AICD was inserted. Repeated blood work in subsequent visits continued to show low potassium and magnesium. The patient was referred to neurology, which raised the suspicion for hypokalemic periodic paralysis. She was started on an oral potassium supplement, and her paralysis attacks became less frequent and shorter in duration. The combination of periodic paralysis and long QT made the diagnosis of Andersen-Tawil syndrome more likely, and genetic testing for mutation in the *KCNJ2* gene was positive.

Andersen-Tawil syndrome is a hereditary syndrome that consists of a triad of periodic paralysis, prolonged QT, and characteristic physical features (low-set ears and small mandible, among others). About two-thirds of patients with Andersen-Tawil syndrome have mutations in the *KCNJ2* gene, which codes for potassium channels.

A few weeks later the patient returned to our preoperative clinic for her carpal tunnel surgery. The recommendation from the neurologist and cardiologist was to maintain her potassium level in the high-normal rage (> 5 mEq/L), as this would shorten the QTc, lessening the chances of a malignant arrhythmia, as well as help control her muscle symptoms. Furthermore, it is necessary to regularly check the serum potassium level when patients like this are hospitalized and acutely ill; if the patient starts to have painful muscle spasms, potassium has to be checked and replaced if necessary, and benzodiazepines work better than other options in controlling pain.

Our patient underwent her surgery with no complications.