

Name: @NAME@
DOB: @DOB@

Date issued:

EMERGENCY LETTER
TANGO2-Related Metabolic Encephalopathy and Arrhythmias

[ADD NAME] is a [ADD AGE] year old [ADD GENDER] with mutations in the gene TANGO2, which causes a rare genetic disorder of Golgi apparatus metabolism. During times of fasting and metabolic stress such as gastroenteritis, respiratory infections, prolonged fasting, or a similar illness, [ADD NAME] can develop the following ACUTE complications:

- ***LIFE THREATENING severe cardiac arrhythmias. Cardiac arrest can occur due to lethal ventricular arrhythmias, most commonly in the setting of marked QT prolongation. ECGs during acute crisis typically demonstrate QTc prolongation and can also demonstrate intermittent Brugada Type I changes.***
- Metabolic crises (hypoglycemia, high lactic acid, and hyperammonemia). Individuals with this disease present acutely with symptoms of low blood sugar include lethargy and possibly seizures.
- Swallowing difficulties with drooling can occur with fluctuating symptoms, increasing risk of aspiration
- Profound muscle weakness and ataxia.
- Severe rhabdomyolysis and elevated creatine phosphokinase are also seen in acute decompensation. Liver enzymes including AST and ALT are elevated indicating muscle injury.
- Chronic symptoms of TRMEA include hypothyroidism and intellectual disability. Treatment with intravenous fluids/glucose may stabilize the acute process.
- In rare patients, pancreatitis and adrenal insufficiency have been seen.

****EMS: – Assess cardiac rhythm and assess for hypoglycemia and begin treatment immediately. If safe for the patient, please bring patient to a hospital, which is equipped to care for this rare genetic condition, or nearest tertiary care hospital.**

EMERGENCY ROOM PHYSICIAN:

1. [ADD NAME] should be triaged as soon as possible upon arrival to the Emergency Room even if the patient does not appear to be ill, because hypoglycemia and life-threatening arrhythmias can occur rapidly.
2. Place the child immediately on continuous rhythm monitoring and obtain an ECG. The ECG should be assessed for QTc prolongation and Brugada Type I pattern. Avoid QT prolonging drugs if the QTc is prolonged. Any premature ventricular contractions should warrant immediate intervention because ventricular arrhythmias can rapidly develop. Patients with PVCs should be transferred to an intensive care unit. Page Cardiology and review ECG and cardiac status.
3. **Start IVF immediately (do not wait on lab results):** Please provide IV fluids, D10 with added age-appropriate electrolytes, at 1.5x maintenance rates.
4. **Labs:** STAT fingerstick glucose, STAT Ammonia- should be placed on ice and sent to lab for immediate analysis, CK level, lactate, venous blood gas, Chemistry panel with glucose, amylase, lipase, and TSH.
5. **Please call or page genetics/metabolic service to inform of ER or hospital admission. Please page Cardiology service for concerns of high risk of arrhythmia during acute metabolic crises.**