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Rhabdomyolysis and cardiac depolarization anomalies: TANGO2 or not ?

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Introduction: acute rhabdomyolysis is a rare but potentially severe disease in childhood. Inherited metabolic disorders have to be searched for. Significant repolarisation abnormalities have been described in patients with acute rhabdomyolysis and TANGO2 mutation involved in the Golgi apparatus.
Methods: Between 12/2013 and 01/2017, 4 children (mean age 3,5 years at first crisis) presented in our department with 1 to 3 acute severe rhabdomyolysis crises (CPK > 10 000 UI/l) associated with significant ECG anomalies. All 4 had neuro-developmental delay (from mild speech delay to spastic tetraparesia, with microcephaly). Rhabdomyolysis occurred after acute viral illnesses or unusual fasting and were associated with neurological deterioration. Cardiac anomalies included significant lengthening of QT and QTc interval in all 4, concomitant bradycardia and life-threatening polymorphic ventricular tachycardia in patient 1, left bundle branch bloc in patient 3 and ventricular arrhythmia and cardiac arrest in patient 4 (figure 1). ECG anomalies were maximal around 10 days after the beginning of crisis and slightly delayed from the peak CPK level. Ventricular dysfunction was associated in patient 2 (mild) and in patient 3 (severe requiring inotrope support). Cardiac anomalies regressed completely in all between the rhabdomyolysis crises. Patient 1 died while admitted for her 3rd rhabdomyolysis crisis, with severe neurological worsening, prolonged QT but no ventricular arrhythmia. TANGO2 mutation was confirmed in patient 1, suspected but not yet confirmed for the 3 others.
Conclusion: Ventricular conduction and function have to be explored in children with acute rhabdomyolysis in the setting of neurodevelopmental delay. TANGO2 mutations may be responsible. The cardiac disorders are rapidly progressive, potentially life-threatening but reversible. The mechanism and treatment are poorly defined.

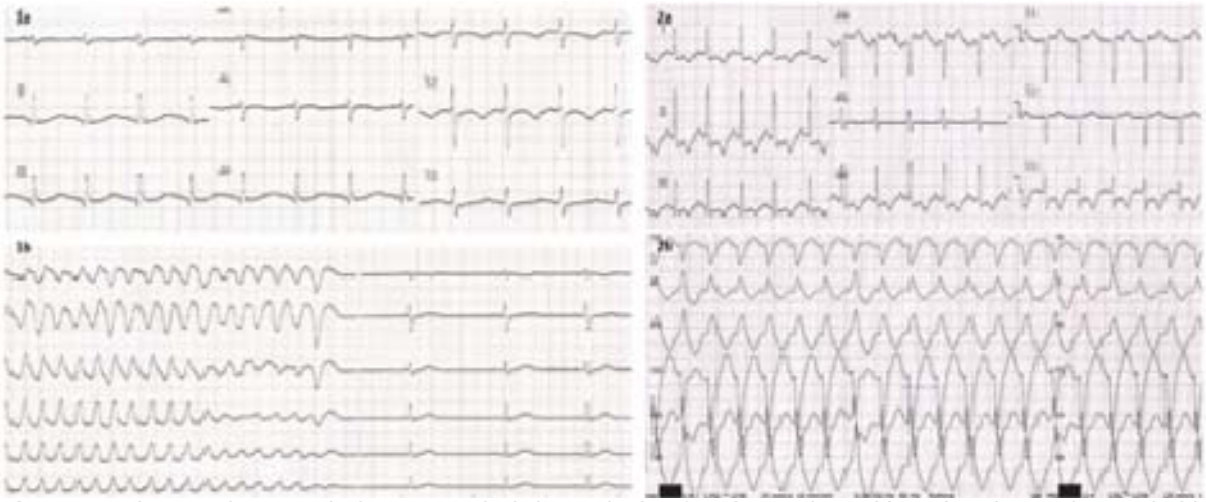

Figure 1: Abnormal ECGs during acute rhabdomyolysis of patient 1 (1a, 1b) and patient 3 (2a and 2b)