

Frequently Asked Questions

This letter is parent created and it is not intended as medical advice. TANGO2 disease is not well known and more research is needed to understand all of the implications of the disease.

What is TANGO2 disease?

TANGO2 is the name of the gene that contains a mutation also called genetic change. Our genes are like sentences made of the letters ATCG. A genetic change can include replacing a single letter with the wrong letter, insertion or deletion of a single letter, and even large deletions of multiple letters. These genetic changes can reduce the function of the gene or stop the function completely.

TANGO2 is an autosomal recessive genetic disease. Both mother and father have to carry a change in one of their *TANGO2* genes for a baby to inherit this condition. Parents who are both carriers have a one in four or 25% chance of having a child with TANGO2 disease in each pregnancy. TANGO2 disease has a variety of symptoms and some children with the disease seem severely affected while others have milder symptoms. The biggest risk for children and young adults with TANGO2 is rhabdomyolysis (muscle break down) and life-threatening cardiac arrhythmia (irregular heart rhythm) brought on by metabolic crisis (low blood sugar and toxins in the blood stream). Many children experience cognitive and speech delays, trouble with coordination and balance, and episodes of lethargy.

Some children with TANGO2 disease have also been diagnosed with DiGeorge syndrome (22q11 deletion syndrome). No two cases of Tango2 are exactly alike.

What are the symptoms of TANGO2 disease?

TANGO2 disease is a very newly discovered disease. The symptoms experienced vary in severity and also in the number of symptoms experienced. Some children experience several complex symptoms, while others seem to experience milder symptoms.

Many parents have reported episodes of lethargy and loss of motor control. Episodes of muscle weakness have also been described, including intermittent episodes of loss of head control resulting in the head to be positioned down or to the side. Parents have reported that sometimes these episodes tend to resolve themselves, especially after the child naps. Most children have developmental delay of varying severity. Some children have weak oral musculature and have difficulty eating enough food to grow and may

struggle with episodes of drooling and slurred speech. Most children with TANGO2 disease have neurologic involvement, including brain changes on MRI and some children have seizures. Health deterioration in TANGO2 kids has been associated with long periods of exposure to sun, viruses such as the common cold or influenza, constipation, over exertion, and not eating or drinking enough fluids. Metabolic crisis (low blood sugar and release of toxins in the blood) is a risk for TANGO2 kids and often results in dangerous heart conditions and rhabdomyolysis.

What are recommendations that may help to keep my child with TANGO2 disease healthy?

There are no proven treatments or diet recommendations for TANGO2 disease. TANGO2 symptoms seem to appear when the body is stressed in any way. With that said, sometimes there does not seem to be a direct reason why worsening symptoms are experienced. Some parents have reported that they see less symptoms when their children:

1. Stay hydrated.
2. Eat regular, frequent meals that include carbohydrates and healthy low-fat proteins.
3. Have regular bowel movements. Some families report that their doctors recommend laxatives on an as needed basis to ensure that constipation doesn't weaken the health of Tango2 kids.
4. Exercise frequently without overheating or over exertion
5. Take the "mito cocktail" * see below*
6. Wash hands regularly to prevent illness.
7. Avoid public places when possible during the winter season to prevent illness.
8. Some cardiologists have recommended that patients have a LINQ monitor to notate any irregular heart rates

Surveillance and Management of TANGO2 disease.

As there are no known treatments for TANGO2 disease, symptoms are managed by a team of doctors and medical professionals that treat symptoms as they arise. Most patients see a **cardiologist**, a **geneticist or a metabolic physician**, an **endocrinologist** and a **neurologist** every six months.

Children may also receive:

1. Yearly blood draws to check for hypothyroidism.
2. Initial audiology exam is recommended as hearing loss has been seen in some cases. Follow-up exams may be yearly.

3. Initial eye exam completed by a pediatric ophthalmologist is recommended, as some children experience vision loss and eye muscle problems. Follow-up exams may be yearly.
4. Having your child evaluated and treated by a Physical Therapist, Occupational Therapist, and Speech-Language Pathologist can help manage developmental delay and maximize the child's ability to participate in activities of daily living.
5. Some children benefit from seeing a dietician, to ensure proper growth and adequate nutrition/hydration.
6. A gastroenterologist is a doctor that can help if your child is experiencing chronic constipation or other digestive problems.

Parents have also seen varying results in health from the "mito cocktail," consisting of B vitamins, CoQ10, and levocarnitine. This vitamin mixture has had significant positive health impacts for some children and not much impact or improvement for others. Some doctors have recommended the liquid form of CoQ10 stating that it is more effective than the powder. Many parents mix the vitamins with chocolate pudding or put it in a smoothie to disguise the taste. This should be discussed with the metabolic specialist or geneticist caring for your child prior to starting any new medications or supplements.

What do I do if my child is having symptoms?

When children are ill or symptomatic, notify the doctor and go in for care. It is very important that children who are ill do not go long periods without eating/drinking. If your child is not eating/drinking or is unable to keep fluids down, they need to be evaluated by a doctor as soon as possible. Anytime you need to bring your child to the ER, it is recommended that you bring the emergency letter (example included) with you to the hospital explaining TANGO2 disease. When you arrive at the hospital, request that your child's heart is monitored. Blood sugar and CPK levels should also be monitored. Hydration, nutrition, and regular bowel movements are very important for TANGO2 kids.

How did tango2research.org get started?

When Tyson and Veronica's daughter, Thea, was diagnosed at age 2 in 2016 with TANGO2 disease, they weren't sure where to turn for help. The disease was not well known and they were told that they would have to be their own advocates for care and information. Their genetic counselor recommended that they use social media to try to find other families affected by the disease. Veronica created a Facebook group and slowly found more and more families to connect with and share information. The Morris family joined the group in 2017. Their 15-year old son Ryan had been recently diagnosed. The Morris family created and began the tango2research foundation with the goal of funding research for TANGO2 disease. They organized a board of tango2 parents and formed a scientific advisory board of doctors and researchers connected with Tango2 patients. The foundation has partnered with Dr. Seema Lalani and Dr. Christina Miyake from Baylor to create more research opportunities for TANGO2 disease.

What's being done for TANGO2 children now?

Since the *TANGO2* gene was discovered in 2016 at Baylor, several papers have been written about the disease. Currently, there is a new TANGO2 disease study underway and the first TANGO2 conference is scheduled for June 2019. Mouse models are available at Baylor for study. Please visit <https://tango2research.org/> for more information.

