

Symptoms of Dehydration



Infants Toddlers Young Child

Dry mouth and tongue
No tears when crying
Decrease in urination
Sunken eyes
Sunken cheeks
Listlessness
Increased irritability
Drowsiness
Rapid breathing
Concentrated urine
Dry skin

Adults

Headaches
Extreme thirst
Decrease in urination
Concentrated urine
Fatigue
Dizziness
Confusion
Racing heartbeat
Dry mouth
Muscle cramps
Dry skin

Barth syndrome (BTHS) is an X-linked genetic disorder presenting with a variable pattern of cardiomyopathy, skeletal myopathy, neutropenia, and growth delay. Pathological changes in the *tafazzin* gene are responsible for Barth syndrome. Tafazzin protein products are critical to mitochondrial structure and function.

Mitochondrial Risk

Heat intolerance or oversensitivity to extreme ambient temperature is common in those with mitochondrial disorders, such as Barth syndrome. In response to extreme temperatures, our bodies thermoregulate by sweating or shivering, which requires mitochondrial energy. Since mitochondrial function is impaired in Barth syndrome, someone with Barth syndrome has a lower tolerance to extreme heat than an otherwise healthy individual.

Cardiac Risk

Cardiomyopathy is a cardinal symptom of Barth syndrome and predisposes those with the condition to be particularly

vulnerable to heat injury. Ventricular dysfunction, which is often associated with Barth syndrome, similarly predisposes those with the condition to heat intolerance. Protective mechanisms such as sweating, the body's way to protect against heat-related injury, can quickly deplete the body's electrolyte balance. This affects the body's autonomic nervous system, which regulates heart rate, blood pressure, breathing and voiding.

Heat Intolerance


The Barth Syndrome Foundation www.barthsyndrome.org


DISCLAIMER: This fact sheet is designed for educational purposes only and is not intended to serve as medical advice. The information provided here should not be used for diagnosing or treating a health problem or disease. It is not a substitute for professional care.


Precautionary Measures


If you or your loved one has Barth syndrome, there are precautionary measures to manage heat intolerance. The person with Barth syndrome should stay well hydrated to replenish lost electrolytes and fluids. They need to have the ability to quickly cool from the extreme heat with nearby access to air conditioning. Make sure the person with Barth syndrome remains in the shade as much as possible to limit heat exposure. It will take less time for someone with Barth syndrome to experience heat exhaustion than an otherwise healthy person and longer for them to recover. Heat exhaustion recovery is not immediate and can sometimes take days for recovery.


The best approach to protection is prevention. Avoid environments or activities that may cause overheating, as someone with Barth syndrome has a higher risk of suffering from heat exhaustion or an even worse heat-related injury.

 **Spritzer Bottles and Fans:** When on an outing, fill a spritzer bottle with icy water and keep it in the cooler. Some spritzer bottles come in combination with a fan to create a constant misting. USB or battery-operated clip fans are great for strollers.

 **Neck Gaiters:** These are a compact quick cooling option that can be placed in a purse or a child's backpack. After soaking for just a minute, gaiters provide hours of cooling relief. They are very affordable and come in a variety of colors. It also may be an approach that the teen will be more agreeable to wearing.

 **Cooling Vests:** These come in a variety of sizes, colors, and cooling options. Vests can be specially made for people with ostomies and for people in a wheelchair. [MitoAction](#) is a great resource about personal cooling systems options.

 **Cooling Patches:** Gel cooling patches offer another approach to cool quickly. Patches are applied directly to the head. They can be used to reduce fever discomfort and headache pain relief. These are available as disposable patches or as reusable gel packs.

 **Swimming Pools:** Swimming pools and other water-related summer fun provide a fun and rapid way to cool down.

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