



Support, Educate, Advocate
Mitochondrial Disease Action Committee

888-MITOACTION (888-648-6228)

www.mitoaction.org

The Dosing Debate: CoQ10 and Creatine in mitochondrial disorders

by Cristy Balcells

Posted on <http://www.mitoaction.org/blog/the-dosing-debate-coq10-and-creatine-mitochondrial-disorders>

This month MitoAction welcomed Dr. Fran Kendall of Emory University's Department of Human Genetics mitochondrial program to discuss the dosing debate around CoQ10 and creatine.

A long-time advocate and clinician for mitochondrial disease patients, Dr. Kendall began the Mitochondrial Disorders Program at Boston Children's Hospital 18 years ago. Since that time, she has been involved in direct patient care as well as diagnostics and biochemical genetics research and currently follows patients through her Atlanta practice.

CoEnzyme Q10

CoQ10 is a cofactor in the electron transport chain as well as an antioxidant and has been a cornerstone of supportive mitochondrial disease treatment for many years. Typical dosing of CoQ10 is 5-15mg/kg of body weight, although "high dose" CoQ10 is common for patients with mitochondrial disease, as well as other mitochondrial-related disorders which are a result of secondary mitochondrial dysfunction (such as ALS).

Recent studies examining the effect of high dose (2700 mg/day) CoQ10 in patients with ALS concluded no improvement in patient symptoms with use of the high dose. Additional studies with mitochondrial patients using 500-800 mg/day of the Tischon brand CoQ10 (such as QGel from epic4health.com, a form of CoQ with better absorption and bioavailability) demonstrated mild to moderate improvements in symptoms such as fatigue and low muscle tone.

Dr. Kendall's experience as well as her clinical research suggests several general findings about use of CoQ10 in Mito patients.

1. Use of CoQ10 does increase blood levels in patients, and the Tischon brand does appear to be better absorbed

2. Most patients reported an improvement in muscle and fatigue related symptoms while using CoQ10; however, higher dosing did not necessarily translate into clinical improvements.
3. Symptom improvement may take 4-6 weeks or longer of CoQ10 use.
4. High dose CoQ10 actually caused muscle breakdown (as evidenced by increased CPK blood levels) in 10-20% of patients. CPK levels returned to normal when the CoQ10 dose was lowered.

As a result, Dr. Kendall suggests that when using CoQ10 to treat mitochondrial disease symptoms, "more is not necessarily better". Patients showed mild-moderate improvements even while on a moderate dose (specifically, 20 mg/kg/day in children, and 1000-2000 mg/day in adults). In addition, increased blood levels of CoQ10 do not necessarily correlate with increased symptom relief.

Side effects of CoQ10 may include:

- Headache (although CoQ10 can help with chronic migraines)
- Heartburn
- Insomnia (take in the morning to prevent this)
- Involuntary muscle movements

Creatine

Use of the supplement Creatine has recently gained more attention - as well as confusion - amongst mitochondrial disease patients, as well as the general population due to use by athletes.

Creatine is converted in the body to phosphocreatine (an anaerobic pre-cursor to ATP). This naturally occurring substance is thought to help generate extra energy when taken as a supplement in people with mitochondrial disorders. However, while creatine has shown positive short-term improvements in patients with Mito and neuromuscular diseases, there are documented side effects with creatine use.

Specifically, side effects of creatine can include:

- Diarrhea
- Dehydration (creatine is a form of a salt; water intake should be increased)
- Weight gain
- Kidney dysfunction (obviously, the most concerning potential side effect with long-term use)

Dr. Kendall suggests that Mito patients who primarily experience muscle-related symptoms, such as pain and hypotonia, may find creatine to be beneficial. However, Dr. Kendall cautions against using any form other than a medical grade product, such as Neotine, as other products are potentially toxic (dosing is typically 5g/day for children, 10g/day in adults). In addition, the potential for kidney dysfunction should always be discussed with the prescribing physician.

To learn more about other ingredients in the "Mito Cocktail", see the summary of the March 2008 MitoAction discussion with compounding pharmacist Saad Dinno, and an informative interview about CoQ10 supplementation with compounding pharmacist Arthur Margolis.