

Summary – Q&A With Compounding Pharmacists Saad Dinno, RPh and Dr. Virginia Tawa, PharmD

One of the front-line treatment approaches to mitochondrial disease is use of a combination, unique to each patient based on symptoms and diagnosis, of vitamins and supplements such as Coenzyme Q10, B-vitamins, L-Carnitine, Creatine and Alpha Lipoic Acid.

Compounding pharmacists Saad Dinno, RPh, and Dr. Virginia Tawa, PharmD, from [Acton Pharmacy](#) answer everything you wanted to know about the ingredients which make up the mysterious "Mito Cocktail."

What is a compound?

A compounding pharmacy assists mitochondrial disease patients by providing vitamins and supplements in a compounded capsule or liquid form depending on the needs of the adult or child patient. There are many benefits to working with a compounding pharmacist. Primarily, a compounding pharmacy can combine the vitamins/supplements in order to minimize the *number* of vitamins and supplements required, as well as to make the medication more patient more *palatable* in liquid or capsule form. They work closely with the patient's physician and take into consideration the patient's diet and diet restrictions as well as the overall medication plan. A compounded medication is then developed which is unique for each patient and his/her specific treatment plan or prescription. The formulas are based on multiple variables, including the prescription, the patient's symptoms, the patient's diagnosis, weight, allergies, physician recommendations, etc. The goal is to work with the physician, patient and pharmacy in order to develop an ideal mix (or "compound") of these vitamins and supplements that offers the patient the most ease and the least side effects.

Vitamins & Supplements

There are many supplements and vitamins that may be prescribed for Mito patients, but the following is an overview of the most commonly prescribed components used in the "Mito cocktail".

CoEnzyme Q 10

Commonly called **CoQ10** this substance is needed to make energy; every mitochondria in the body has CoQ10 (except red blood cells) and it is needed for energy. There is often a deficiency of CoQ10 in patients with Mito, which is one reason that a supplement is provided. Reactions to therapy vary but usually result in an increased energy, improved respiratory function, decreased muscle atrophy, decreased seizure activity, and improved mental alertness. In general, it takes at least 4 -6 weeks for patients to feel the benefits of CoQ10 (for some it may take longer for the effects to become apparent). Side effects are possible and commonly include stomach upset, diarrhea, and sleep disturbances; it is recommended that patients take CoQ10 with food

and to take all doses earlier in the day. Most patients take one dose with breakfast. The only adverse interaction patients need to be aware of is with coumadin/warfarin (blood thinning) medications.

B Complex Vitamins:

Vitamin B1 (Thiamin) This is a water-soluble vitamin which helps break down carbohydrates so the body can better use them, helps with growth and maintenance of muscle tone, and aids memory. The only possible side effect sometimes noted is drowsiness.

Vitamin B2 (Riboflavin) Also a water-soluble vitamin, B2 is necessary for energy production in the mitochondria and increases muscle performance as well as helping maintain healthy mucous membranes, skin, hair and nails. The only side effect noted is the tendency to turn urine an orange color. Given in the form of Riboflavin Biphosphate can improve the taste of this vitamin.

Vitamin B3 (Niacin) Occasionally used, B3 can often cause flushing of the face so it is generally given separately first to see if any side effects will occur before it is added to the cocktail.

Vitamin B6 (Pyridoxine) and Vitamin B12 (Cobalamine) These are other B vitamins which are frequently part of the compounded mix which patients with Mito may use.

Other AntiOxidants

Antioxidants decrease free radical accumulation in the cells and therefore are used for Mito patients as well. ***Alpha Lipoic Acid*** is probably the most commonly prescribed anti-oxidant used in the Mito cocktail.

Vitamin C This is used for its help in the healing process and to ward off infections but can cause some stomach upset and occasionally headaches when the dose is increased.

Vitamin E This protects cell membranes and improves neurological function. Usually the dose is no higher than 400 - 600 mg per day for an adult. Again, this can interfere with coumadin/warfarin medications, so be aware of this.

Vitamin K1 Another vitamin that may be added (but with caution as there is a very small safe range for the dosage of this vitamin) must be prescribed by a physician, and is not to be purchased over the counter.

L-Carnitine Also prescribed by a physician, L-Carnitine helps transport fatty acids and improve the strength and tone of muscles. It may upset the stomach and give off a fishy odor. It is taken in either tablet or liquid form and is usually taken separate from the compounded cocktail.

Creatine Creatine helps maintain muscle mass and increases energy for cells. Its side effects include diarrhea and drowsiness; the dose ranges from 5 grams/day for children to 10 grams/day for adults and is generally compounded into liquid or capsule form. All of the vitamins and supplements noted above are added or not added to a cocktail as specified by a patient's need; each cocktail is patient specific. The physician will prescribe the needed treatment then the pharmacist will mix the cocktail accordingly. _

Questions from listeners

What is the difference between Ubiquinol and Ubiquinone?

The form of CoQ10 which is dispensed by Acton Pharmacy in 90% of the cocktails is Ubiquinone. Though Ubiquinol is twice as bioavailable, it is very difficult to compound. There are no studies which indicate how to use it in this form especially because it is very unstable in air. It is the same chemical as Ubiquinone, but it is in a reduced form. Some patients also may be prescribed Idebenone, another form of CoQ10 that is under study for use in mitochondrial diseases.

How do I judge the quality of substances I purchase over the counter?

This can be difficult because there are many manufacturers, and price can also vary quite a bit. The best advice is to talk with your pharmacist about your available options. The potency of substances from different manufacturers may vary. Typically, the forms used in a compounded medication are more pure than those that could be purchased over the counter.

What about a terrible taste?

As an example, the B complex vitamins can be very bitter so they may be more palatable if compounded in a suspension vehicle. Ask if your pharmacist uses sugar-free, carbohydrate-free flavors, but may add a natural sweetener to mask the taste.

What is an MCT oil?

MCT stands for medium chain triglyceride. This is an oil occasionally used with CoQ10 to lessen the side effects and improve absorption, but some patients have difficulty with the taste. A common side effect is diarrhea. For best absorption and to decrease the risk of stomach upset, it is recommended that patients take CoQ10 with food. For those patients with a very restricted diet, the pharmacy can compound the substances so that the cocktail is casein free, gluten free, etc.

Why not just buy the vitamins over the counter?

For some patients, the dosages necessary in the Mito cocktail require one to take up to 30 pills a day; working with a compounding pharmacist could get this number of pills down to a more manageable number. Also, note that whenever medications/supplements etc are being compounded, be sure to get a print-out from the pharmacy of exactly what substances and what exact amounts are in your cocktail. It is very important to have this one page print out so that you can bring it to all physician appointments AND to have available and updated in case of an emergency.

Can a patient bring a request for vitamins and/or supplements directly to pharmacy him/herself or should a prescription come from physician?

You need a prescription. Not only is the amount of substance/chemical is weight specific, but different components of the compound may vary depending on the exact diagnosis/mitochondrial defect. In addition, the compounded cocktail is more typically covered by insurance when prescribed by an MD.

Any questions can be directed to Saad Dinno at Acton Pharmacy, **actonpharmacy@yahoo.com**