# Summary – The Mito Cocktail Explained Dr. Virginia Tawa

"Mito Cocktail" is the name given to a variety of vitamins and supplements which are commonly used by adults and children who have been diagnosed with mitochondrial disease. While there is no cure for mitochondrial disease, many types of the disease including mitochondrial myopathy, mitochondrial cytopathy, MELAS, PDCD, or Complex I, II, III, and IV are responsive to specific vitamin and supplement therapies. Please, consult with your physician before beginning or altering any medication, vitamin or supplement regimen.

The supplements and vitamins used by Mito patients are often high doses and could require a patient to take up to 50 different pills per day. A compounding pharmacist (find one in your area through the IACP, International Academy of Compounding Pharmacists) can create a "cocktail" in a liquid, capsule or other form that combines the pure powdered form of the prescribed supplements and vitamins. The final medication is usually a much smaller amount than if otherwise taken, and can even be flavored to improve palatability. By avoiding fillers common in over the counter tablets, an individual's allergy or dietary restrictions can be accommodated. The exact compound, including dosage and ingredients, is determined by the patient's physician and differs depending on an individual Mito patient's diagnosis, clinical symptoms, weight. Saad and Virginia discussed the most common ingredients used by compounding pharmacists who specialize in treating mitochondrial disease when putting together the "Mito Cocktail", and offered some advice about use and side effects as well.

## Coenzyme Q-10

Coenzyme Q-10 (Coenzyme Q10, CoQ10, CoQ-10, CoQ, ubiquinone, Q-Gel®), is a fat-soluble vitamin-like substance present in every cell of the body and serves as a coenzyme for several of the key enzymatic steps in the production of energy within the cell. It also functions as an antioxidant protecting against accumulation of harmful free radicals, which is important in its clinical effects. Many patients report increased energy while using Coenzyme Q-10, and thus it is a common "front-line" approach to supporting children and adults with mitochondrial disease. Frequently reported side effects include stomach upset and sleep disturbance, so the pharmacists recommend taking Co Q-10 doses earlier in the day and with food. Therapuetic levels may need time to be established, so patients may not see an immediate beneficial effect. In addition, the excess of the substance that is not used is stored in the fat cells, so proper dosing is important.

### L-Carnitine

Another common therapeutic supplement, L-Carnitine (levocarnitine, CARNITOR®) helps to break fat from the food that we eat into energy. Side effects may include diarrhea, and a fishy odor which may be excreted via the sweat glands. Some patients report decreased fatigue and energy improvements by taking L-Carnitine.

#### **B-Vitamins**

Some B-vitamins are cofactors which participate in important mitochondrial reactions. Most of the B-vitamins have a bitter taste and more palatable if flavored. B-vitamins are water soluble; that is, they are excreted if not used, and the benefit from taking these vitamins should be felt immediately.

## Thiamine (B1)

Thiamin is a cofactor for the pyruvate dehydrogenase complex, and in some patients has improved lactate and pyruvate levels. Thiamin has also helped some patients by improving their clinical symptoms.

## Riboflavin (B2)

Helpful for some patients with headache and migraine, Riboflavin is another important cofactor that participates in important mitochondrial reactions.

Other B vitamins that may be included in the "Mito Cocktail" include: **B3**, **B6**, **B12** and **folic acid** (B9). Many patients find folic acid a beneficial ingredient, as this supplement is key in prevention of anemia and assists with the production and health of new cells.

#### **Antioxidants**

A normal byproduct of the energy cycle are free radicals. Accumulation of free radicals may be especially harmful to mitochondrial disease patients. The use of antioxidants like **Vitamin C** (ascorbic acid) and **Vitamin E** (? tocopherol) can help to reduce free radical accumulation, which in some patients may mean improvements in energy and function.

Other common ingredients in the "Mito Cocktail" may include alpha-lipoic acid (another potent antioxidant), creatine, selenium or succinate.

Again, please work with your doctor to determine the exact ingredients to be used in the compound that will be most beneficial for you. One can see that, like some of the best recipes, the "Mito Cocktail" is created for each individual's unique taste!