Summary - Medical Foods for Mito

DeFries & Moore

The history of Medical Foods can be traced to 1963 when the Guthrie Test, a blood test, was first formulated as a means of diagnosing phenylketonuria (PKU) at birth. Without treatment this disease causes severe brain damage. By the 1970's, all newborns in the US were being tested for PKU and if found positive (about 400 infants per year), they were treated with a very specific diet which included specially formulated medical foods. These products were originally classified as drugs but were later reclassified as "Medical Foods" by the FDA. There are now about 40 different types of products which are used to prevent or treat severe or crippling diseases. They are often produced in formulas meant for infants, adolescents or adults. This has revolutionized how patients are treated for many rare disorders. In order to be classified as Medical Foods these products must meet specific criteria:

- formulated for oral or enteral administration
- made for dietary management of those with limited ability to absorb nutrients
- provide nutritional support for management of unique medical conditions
- be used with strict medical supervision
- only to be used by a specific classification of patients - not the general public
- be GRAS - Generally Recognized As Safe (this means they must be nontoxic and safe to be used)

Solace Nutrition is pursuing many areas and is quite involved both with patients and professionals who deal with Mitochondrial Disease Mark was involved with the family of Lorenzo Odone whose condition was made known through the movie Lorenzo's Oil.

Coenzyme Q10 (Ubiquinol)

Solace Nutrition produces a product called Cyto-Q. It is a bioavailable form of CoQ10, a Medical Food for Mitochondrial patients which differs from the over-the-counter (OTC) version of CoQ10 (Ubiquinone). Ubiquinol is unoxidized; it has not yet been exposed to oxygen. Cyto-Q comes in a liquid orange/pineapple flavor dosed for the pediatric population of 80 mg/10 ml (2 teaspoons). Coenzyme Q10 supports mitochondrial energy production in the electron chain by carrying electrons from cytochrome to cytochrome in order for ATP to be produced in the mitochondria. Without CoQ10, there is no electron transfer.

In order to obtain Cyto-Q (and have it reimbursed) a physician must prescribe this medical food using one of these codes: 57771-0070-10 or 57771-0007-01. These numbers are also relevant to the pharmacist who will match it to the insurance coverage to determine if this medical food is covered. Soon (August 2009), a more concentrated version of Cyto-Q will be available. The more potent product will contain 100mg/ml and will have to be mixed with juice. It is highly concentrated and has not been pre-flavored.

Creatine Monohydrate

Cytotine is the creatine monohydrate product offered by Solace, and is a very unique form of stabilized creatine monohydrate. Over-the-counter (powdered) creatine
immediately changes to creatinine as soon as it is added to liquid (water, juice, gastric acids). Subsequently, creatinine is a toxic by-product naturally produced in the muscles and filtered thru the kidneys. Increased creatinine levels can cause headaches and decreased energy, and is generally associated with kidney failure. As a result, many physicians have concern about use of high doses of creatine used for mitochondrial disease patients, and recognize that use of OTC requires vigilant monitoring of kidney function. Solace Nutrition suggests that their product, Cytotine, remains stable (even with contact with water or gastric acid) until it is absorbed into the bloodstream, and has better absorption levels overall. In addition, there are usually no side effects like bloating or headaches which often occur with the OTC versions. Creatine is occasionally used in the "mito cocktail" because it is a necessary part of ATP production: creatine phosphate holds on to the phosphorous, then is able to give it up in order for ADP to make ATP. In other words, the creatine phosphate acts as a storage mechanism for phosphorous on the outer mitochondrial membrane.

D-Ribose
Cytose is the Solace product name for D-Ribose. Since cytos refers to "cellular" all Solace products which have the prefix cyto are mitochondrial products. Cytose was developed because in 2007 the National Institutes of Heart, Lung and Blood identified the connection between mitochondrial dysfunction and cardiac disease. NIH then decided that there needed to be more research into the interconnection between the two conditions especially since mitochondrial disease is essentially a disease of energy depletion. Ribose had been used primarily in cardiovascular diseases but cardiovascular issues appear to be a secondary condition in 60% of adult mitochondrial patients.

Primary mitochondrial disorders are often complicated by depressed levels of key components in the mitochondrial tissues or even in the cardiac tissues which subsequently reduce heart function. Ribose is a sugar which enhances the cellular function of ATP. Since the heart, brain and digestive systems have the highest use of energy in the body, it is not surprising that these systems may be affected when someone has mitochondrial disease.

How to Obtain Products
Because the products discussed here are labeled by the FDA as medical products, use must be under medical supervision and requires a prescription from a physician. (Solace Nutrition products are not available over the counter.) They may be purchased directly from the website (www.solacenutrition.com ). You will be asked to fill in your physician's name, phone number and your diagnosis. Whether you obtain the products from the website or from your pharmacist, a special national drug code (NDC) will be required (as noted with the discussion of Cyto-Q). All Solace Medical Food products have an NDC. This information can be obtained from the website or you can request help from your pharmacist. If you are denied coverage by your insurance company, you can ask Solace to send your physician a Medical Necessity Letter which then may be used for reimbursement from your insurance company. If the products are still not covered, purchasing them out of pocket may be less expensive if done directly
from Solace online, but you will need to provide your physician's a name and phone number - the products can then be sent to you directly. Within the US they should come within 5-7 business days (Solace is based in Maryland); if in Canada it may take a week and will be billed in US dollars.

Dosage ranges for all these products are very individual and must be prescribed by the patient's physician. Since patients with Mitochondrial Disease are often their own best advocates, reporting of symptoms is the best guide to dosage. There is no universal "Mitochondrial cocktail"; for example, some patients need higher doses of niacin while others need riboflavin or selenium.

The Solace Nutrition website may be visited for further reading, links to publications and detailed information all Solace products (including controversial issues). Patients, caregivers, and healthcare providers may also email info@solacenutrition.com or call 1-888-8solace (1-888-876-5223) and speak to Nancy or Mark.