

## Supplements

Supplements are popular therapies in PWS. Some families report responses such as increased strength in infancy, while others do not see changes with these products. Be aware of herb/drug interactions. Herbs and drugs that have similar purposes (such as sedatives, blood thinners, or stimulants) should not be taken together, nor should herbs and drugs with opposite actions (a sedative herb taken with a stimulant drug, for instance). Ask your local pharmacist about any known adverse reactions, and inform your child's physician of all supplements being used.

### Coenzyme Q-10

PWS Specialist Moris Angulo, MD wrote the following information for PWSA | USA which was reviewed by PWS Specialist Jennifer Miller, MD:

“Coenzyme Q10 (CoQ10) is an important vitamin-like substance that is vital to the “powerhouse” of cells – the mitochondria - for energy. It is created in the body but is also present in foods. CoQ10 is now a very popular supplement with a wide range of claimed benefits. Its role in muscle energy is clearly proven, but taking additional supplements has shown varied results in a variety of conditions. For example, CoQ10 may be recommended to a heart patient who is on medications that can decrease natural CoQ10 levels. With muscle energy so diminished in PWS, it was hoped that CoQ10 supplementation would be a boost, a low-risk supplement worth trying. With many parents reporting positive results, many PWS specialists now routinely suggest CoQ10, not as essential or due to a deficiency, but as a supplement that may be worth a try.

**Are people with PWS deficient in CoQ10?** There is no clear evidence of a CoQ10 deficiency in PWS. There has not been a research trial which shows the benefits of CoQ10 in PWS by comparing treated children to those who did not take CoQ10. Lacking that evidence, PWSA | USA cannot provide scientific information in terms of dosing and the right formula of CoQ10 for children and adults with PWS, or state that it is an essential medication for people with PWS.

**Is CoQ10 supplementation effective in persons with PWS?** We know from communications with parents and physicians who have used this product that the response to CoQ10 can vary - from substantial changes in energy to no effect at all. We are aware of no known adverse side effects if taken in an appropriate dose. There are not clear dosing guidelines for CoQ10 especially in childhood, and suggested doses are not standardized across brands. The information we have on the effectiveness of CoQ10 is all "anecdotal" data (i.e., from parents commenting on their personal experiences with their child). CoQ10 supplementation is not a substitute for growth hormone treatment which has clearly been shown to have multiple benefits for individuals with PWS, and is the only FDA approved treatment for PWS.

**How can I choose the most effective brand of CoQ10?** Over-the-counter supplements and vitamins are easy to obtain, but may not be effective because the ingredients must be in the right formulation for the body to absorb. Vitamins can be hydrophilic (dissolve in water) or lipophilic (dissolve in fat), so other ingredients may need to be added to improve absorption. For example, vitamin D may be an ingredient in a typical dry vitamin tablet, but without additional fat, it is not well absorbed by the body. CoQ10 is a lipophilic cofactor and requires fat for absorption. Powder-based CoQ10 supplements do not contain fat and are poorly absorbed. To overcome this, most CoQ10 supplements are available in softgels with vitamin E and other lipids (fats) to help the body absorb the CoQ10. While the addition of fats

improves absorption, a CoQ10 supplement that is both water and fat-soluble can provide even better absorption.

**What are safe doses of this supplement?** Unlike multivitamins, there is not an official recommended starting supplemental dose of CoQ10. Each manufacturer may suggest different doses and package the supplement in different strengths. Most manufacturers suggest starting doses between 1-30 mg/per kg/ per day for infants, and no more than 180-200 mg per day for older children. In non PWS older children and adults who have had low blood levels of CoQ10, 60-100 mg per day is sufficient to raise the blood level to the normal range. CoQ10 softgels typically come in 60- or 100-mg doses. These gel caps can be sliced open, and the contents squeezed out. Tiny softgels may be a choking hazard for some individuals, but there are also liquid preparations. CoQ10 can be purchased over-the-counter at most pharmacies or can be purchased on line. Blood tests of CoQ10 need special handling and results are difficult to interpret. Some physicians still recommend a CoQ10 level before treatment and may want to monitor blood levels. Be sure to give your health care providers a complete list of supplements and the doses that your child is receiving at each visit.”

### **Vitamins in High Doses**

PWS Specialist Merlin Butler, MD, PhD, shares, “It is not a good idea to have excess quantity of any specific agent particularly a vitamin. Although the B vitamins are not fat soluble (such as A, D, E, K) and should be excreted if in excess if the metabolic and physiology of an individual is not altered and we cannot say this with certainty in PWS. Therefore, I would not recommend it particularly in developing infants (PWS or not PWS). B12 treatment can mask specific types of anemias so one would need to follow blood counts as well. As recent data on nutrition (particularly vitamins such as folic acid, another B vitamin) and its effect on gene expression and activity (which we know is already altered in PWS) could further complicate the gene alterations in PWS. For example, adding folic acid may decrease gene expression in living cells further complicating the altered genetic network and gene-gene interaction in PWS based on the one carbon metabolism pool in living cells. Therefore, I would not recommend megadoses of B12 in [patients with] PWS, particularly infants; it is an area of concern for me regarding long term effects for the PWS individual. We should be cautious.”