

## Aunt Cathy's Guide to Nutrition:

## Sanford Medical Center

### Comments Regarding the

### Prader-Willi Syndrome Association's Food Guide Pyramid for Weight Control



**Aunt Cathy 4-16**

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Children with a genetic condition called Prader Willi Syndrome usually struggle with their weight all of their lives. They appear to use up fewer calories each day, so just eating “normally” can still cause them to gain weight extremely fast. Losing weight is extremely difficult. The following is a 2003 Food Guide Pyramid that was designed for people with PWS, but potentially useful for any people with very low calorie requirements (such as people whose movement is impaired.) This pyramid has some helpful ideas (especially the suggestion to place vegetables as the “base” of the pyramid instead of the grains and cereals food group in the “regular” USDA Food Guide Pyramid.)

**In addition to calorie concerns, however, I am very concerned about assuring that micronutrients (vitamins and minerals) and protein are provided in appropriate amounts in spite of the decreased total food intake.** There are some other serious concerns as well. Unfortunately, there are some problems in that area in the 2003 Prader Willi Food Guide presented below. As it appears to continue to be used in practice in spite of these important issues, I have taken the liberty of interjecting my thoughts on this issue as you look over this otherwise helpful way of adjusting the base of the pyramid.

**My comments are clearly delineated from those of the PWS Pyramid designer (Beverly Ekaitis, DTR, registered dietetic technician) by brackets [ ] and by bold print and blue color.**  
**Start of Original Article: Prader-Willi Syndrome Association (USA)**

### **A PRADER-WILLI FOOD PYRAMID**

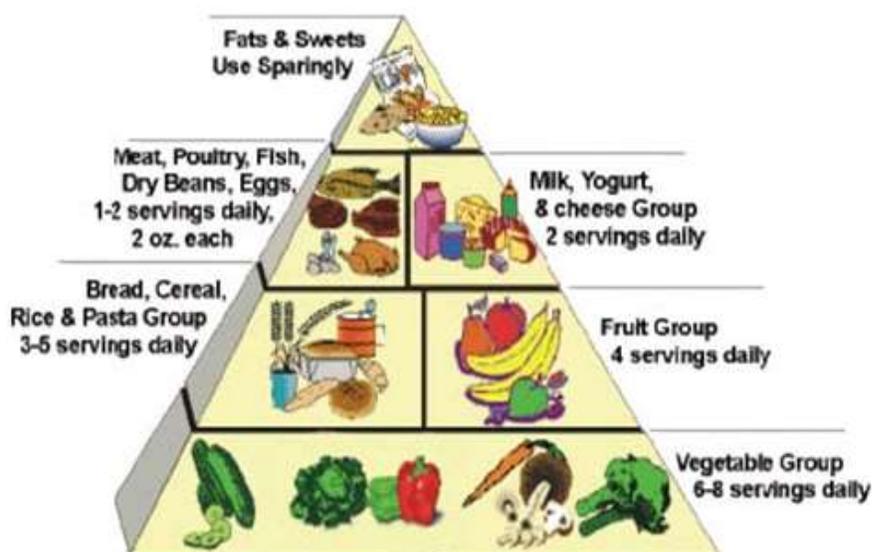
by Beverly Ekaitis, DTR, dietetic technician The Children's Institute of Pittsburgh (TRI)

**PWSA Editors' Note:** The USDA's Food Guide Pyramid provides an appealing graphic tool for thinking about a day's food portions, but it simply adds up to too much food for someone on a Prader-Willi diet. We asked the Children's Institute if they could adapt the new pyramid to the typical PW diet for families that might wish to use it as an alternative to the Exchange System, the Red-Yellow-Green (Stoplight) Diet, or other methods of counting calories.

The Institute was glad to oblige but urges those who have been through the Institute's program to continue using the Red-Yellow-Green Diet that they learned there. The Prader-Willi Food Pyramid that follows may not be appropriate for young children or for those on growth hormone therapy, and it should not be considered substitute for individualized dietary guidance. Dietary guidance preferably should come from a nutritionist who is familiar with PWS.

The Food Pyramid Guide to Daily Food Choices, designed by the U.S. Department of Agriculture for adults who need 1,600 to 2,800 calories a day, represents the relative portions of foods to eat each day to maintain a healthy weight and body. To make the Food Pyramid usable for people with Prader-Willi Syndrome, a few changes have to be made.

The first change needed is to adjust the number of daily servings for each food group in order to reduce the total calorie level to 800 to 1,200 a day. These lower levels will provide for weight loss or PWS.



[CB note:

**It is potentially useful for others with very low caloric requirements as well, with the same caveats described below. Actual calorie goals for PWS and non-PWS individuals will vary considerably. Also, when establishing such a low calorie goal, consider that the daily values and other guidelines are usually based on a 2000 calorie diet. It is important to remember that such low calorie levels will invariably be inadequate in a number of nutrients unless careful supplementation is done. Failure to replace these nutrients is not benign.]**

Second, although the five main food groups — bread, vegetable, fruit, meat, and milk — remain the same, the positions of two of the groups need to be changed on the pyramid to reflect a change in the recommended number of servings. Each group has a specific number of servings that determines its position on the pyramid.

### **The Food Groups**

The first USDA Food Pyramid (which had horizontal lines) has a base of the **Bread group**, which would provide the highest number of daily servings. The PW Pyramid, on the other hand, has as its base the **Vegetable group**, with 6-8 servings a day. For those familiar with the Red-Yellow-Green Diet, these would be "GO" foods, i.e., foods low in calories and fat. Making the vegetable group the base of the pyramid and the bulk of the diet will allow a large volume of food to be eaten without many additional calories.

The **Bread** group, which includes cereal, pasta, and rice, moves up the pyramid with a decrease in number of servings to three to five per day. We would also include starchy vegetables like corn, peas, and potatoes in this group because they have the same amount of calories per serving as breads.

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**[CB note: Try to use whole grains and foods that are naturally high in fiber whenever able to improve the micronutrient content of the diet (especially magnesium, chromium and natural forms of vitamin E) and to decrease the potential for insulin resistance problems. (See my “Magnesium” and “Top Five Recommendations” papers for more on this.)]**

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The **Fruit** group includes fresh fruit, canned fruit, juice, and dried fruits. Many people think of fruit as a "free" food. While it is a good snack and a good source of fiber and vitamins, it does have calories that should be counted if one is on a restricted diet. The daily servings should be four—one at each meal and one for snack.

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**[CB note:**

**Choose whole fruit as much as possible instead of juices. If you use canned fruit the juice-packed or water-packed are preferred over syrup-packed products. Liquid carbohydrate calories consumed may be less well recognized as calories consumed by the body of some individuals, and so additional calories may be accidentally taken in. Excessive juice and “regular” pop consumption is suspected of being contributory to increased weight gain in children in general.**

**Interestingly, some people with carnitine inadequacy problems (as discussed later) will be particularly unable – and therefore unwilling -- to**

limit carbohydrate of any kind (starch, fruit, sugar, etc.) because they cannot use fat for fuel well, and so carbohydrate foods are the only source of energy that they can reliably access. An example of this is a very heavy person who simply cannot switch from “regular” soda to diet soda (or to other sugar-free beverages like water) no matter how many times we point out those “150 empty calories per 12 oz can” labels and tell them they really need to quit drinking all that pop.

Unfortunately this is often interpreted as lack of will-power and a character flaw instead of recognizing that there is sometimes a unique pattern of what kind of foods are especially sought out. In a number of cases, I have found that correcting the fat metabolism problem with supplemental carnitine can hugely facilitate the ability to make this change to a lower intake of carbohydrates.

I think this situation also has application in the model of some heavy people being described as “addicted to sugar.” As some folks in the general population are now being identified as having compromised carnitine production (when we check) we may find that the same carnitine replacement intervention takes the pressure off people with this problem who are often actively seeking “carbs!”]

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The **Milk group** includes yogurt, milk, and cheese. To fit the needs of the person with PWS, the servings per day should be two, and the products chosen should be nonfat or low in fat. Fat-free, sugar-free frozen yogurt also can be used as a milk serving.

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[CB note: Be sure to provide additional calcium and vitamin D, as the amount provided by the serving number shown here is clearly inadequate for optimal health. MANY people (and over-weight individuals in particular) have been shown to require an intake of vitamin D well above the present RDA level.

Now that blood levels are beginning to be checked more often, the very large number of people with inadequate vitamin D levels in their blood is being identified, and inadequacy is now recognized as being very detrimental to heart health, muscle function, immune system function, prevention of cancer, autoimmune disorders, osteoporosis and bone pain. More on vitamin D will be discussed later.

## Back to the specifics of the Pyramid:

Note that the two cups of milk suggested will provide only 200 iu of vitamin D, but PWS individuals should be provided with at least 2000-5000 iu daily simply to maintain levels associated with optimal health. Even more may be shown to be needed for some individuals. Also, in this context, the 2000-5000 iu is a maintenance level, not a level to treat deficiency, which could be much more than that.

Most cheese, yogurt and other dairy products do not provide any vitamin D at all ... and if some has been added in certain brands, it is still at the same too-low amount as is added to milk (i.e. 100 iu/cup.)

Vitamin D deficiency is extremely common, generally unrecognized, and contributory to increased risk of cancer, muscle weakness, muscle pain, heart disease, MS, arthritis, diabetes, compromised immune function and more. It is now being described as an “unrecognized epidemic” in the general public, and the World Health Organization (WHO) estimates that about half of the world’s population is vitamin D deficient.

People who are overweight have an additional risk of vitamin D deficiency. A good plan would be to check the vitamin D level in the winter and if deficiency is found, the physician would then order a special high-dose catch-up amount (a “therapeutic dose”), which would be followed by an intake equivalent to at least a 2000 iu/daily maintenance level. Some people require 5,000 iu for maintenance ... it is quite variable among individuals. One example of a therapeutic dose is 50,000 iu/week for 8 weeks, followed by a re-check and sometimes another round of that level of supplementation.

The serum (blood) level that is associated with optimal health is a minimum of 40 iu; the older lab sheets say that 25 is the lower end of normal, but that level has been shown to be insufficient for many of the known functions of vitamin D, such as cancer prevention and muscle strength. The current lower end of the normal range is 30 ... but it looks like there are many health benefits associated with levels in the range of 40-60.

People with PWS have several additional risk factors for deficiency that lead me to recommend that it would be wise to get a vitamin D level measured annually in the winter (the lab to order is the “25-hydroxy vitamin D” measurement) unless the individually reliably takes a 5,000 iu

**vitamin D as a gel cap or tablet daily. (See my “Top Five Recommendations” handout or my more detailed paper just on vitamin D for more information.)]**

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The **Meat** group includes meat, fish, poultry, eggs, peanut butter, and cooked dried beans. The USDA also includes nuts in this group, but due to their high fat content they should be eliminated from the PW Pyramid.

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**[CB note: Although nuts and peanut butter are higher in calories than some foods, they are also quite nutrient-dense and they are among the richest source of dietary magnesium, a mineral that is often low in American diets, and which is extremely metabolically important.**

**In children and adults who are significantly overweight, magnesium may make a difference between whether or not one goes on to develop insulin resistant (type 2) diabetes. Additionally, magnesium inadequacy can contribute to obesity itself by making it hard to convert fuel from food into usable energy. (See “Magnesium” handout and “Top Five Recommendations” handout.)**

**The fat in nuts and legumes is of the more “heart friendly” type – rich in the type of fat called “monounsaturated” fat. Also, although fat has more calories per ounce than carbohydrate or protein, it is useful to note that in non-PWS individuals a more generous fat content per se does not seem to be associated with increased fatness or decreased weight loss as long as one stays within the target caloric level.**

**However, people with PWS appear to have difficulty doing anything with the fat they eat except store it. Restricting the proportion of calories from fat is reasonable, but as is discussed later, there are ways to help them use the fat more normally. In that situation, restricting fat specifically would become a much less important goal. Similarly, limiting consumption of fat does not prevent the production of fat from extra calories taken in from protein and carbohydrate.**

**Nuts and peanuts also provide protein and the protein and fat content both contribute to a sense of satiety for most people. It helps them not get hungry again so soon, and it has been shown to be true for many other**

**groups of people who are trying to “watch calories.” Whether this is true for people with PWS is not known ... it appears that the drive to eat is controlled by a different mechanism than the sense of satiety associated with stomach fullness, as described further below.]**

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And the USDA suggests two to three meat servings per day of 2½-to-3-oz. portions. To decrease the calories for the PW meal plan, we changed the portion size to 2 oz. and suggest one to two servings a day. This means that a person on 800 calories could divide the 2 oz. serving to provide 1 oz. at lunch and 1 oz. at dinner, and a person on 1,200 calories could have 2 oz. at lunch and 2 oz. at dinner.

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**[CB note :**

**Dried beans and peas – kidney beans, navy beans, lentils, split peas, etc. – are terrific foods for people with weight problems. They are generous in magnesium, chromium and many other nutrients that help us use our calories appropriately, and they are also low in fat and high in fiber. They are a good source of protein, they are ‘filling,’ and their carbohydrate is in a form that has a low ‘glycemic index.’**

**The amount of total protein needs to be at least adequate to maintain muscle mass and many protein-dependent functions. That mean that after removing servings of meat, etc., it is important for people designing the diet to check that the total protein is not inadequate. It easily could be if the calorie allowance is quite restrictive.**

**The World Health Organization suggests that a minimum of about 1 gram protein per kg body weight is needed by everyone. In comparison, most Americans eat two to three times this amount.**

**Some people calculate this protein requirement in PWS using the individual’s actual weight, some use the average weight for people of the person’s height, and some folks split the difference and find the mid-range between those two numbers. If the lowest protein-allowance version above is chosen (i.e. 1 g/kg using average weight for height,) follow-up evaluation of protein status is essential. Nobody benefits from protein deficiency.**

Diets based on 'low glycemic index' foods appear to result in more weight loss than a "low fat" diet even when both diets provided the same number of calories. However ... as before ... these observations may not be that useful in coping with the metabolic problems of people with PWS. However, it may be helpful to people with PWS as well if it results in fewer episodes of elevated blood sugar after eating, perhaps by avoiding excessive stimulation of insulin. Hyperinsulinemia itself can be contributory to excessive weight gain.

It is also reasonable to provide at least 1000 mg fish oil daily or to eat "fatty fish" twice weekly (as is recommended for everyone by the American Heart Association.) There are many reasons for this ... please see my "Top Five Recommendations" paper for more general explanation. The reason it would be particularly reasonable in PWS is because missing part of a chromosome (or other metabolic conditions like diabetes) can affect production of many things we usually make for ourselves, such as the long-chain omega-3 fats EPA and DHA. That is, they become "essential."

These fats have many special roles in health, including eye health, brain development and decreasing inflammatory conditions. The "ready-to-go" form of EPA and DHA needed for many of these activities is in fish oil. Vegetable oils have none. Meat, eggs, and dairy fats generally have little or none. (Some specially produced products are becoming available by feeding animals differently, but the amount is not enough to rely on in this situation, and they are pricey.) The caloric contribution is very small compared to the potential health benefit of providing some.

This small amount is designed to assure adequacy of key substances to perform important metabolic roles; it is not primarily being provided as a calorie source nor should it be counted as one. I standardly recommended it for all the children I work with who have metabolic problems ... and also for all those who do not have any particular health problem. I take it myself and make my husband take it too.

One additional caveat is that some people with PWS develop Type II diabetes and may be put on the medication Metformin/Glucophage. This medication has been shown to cause significant impairment of vitamin B12 status, which can cause neurologic injury. I recommend that anyone on this medication at least be monitored with an annual vitamin B12 level.

Note that if the level is dropping even though still in the normal range, it suggests that they are using up stored vitamin B12 and are likely to develop deficiency even if it is not seen now. A blood test showing high MCV (Mean Cell Volume) is a very late-appearing symptom of vitamin B12 deficiency. The changes in cell size occur when the person is no longer able to make adequate DNA ... this should not be used as a screening test.)

If generous oral vitamin B12 does not correct the problem (which it definitely may not do with this medication in the picture,) consider a vitamin B12 shot to bypass the intestinal impairment of vitamin B12 absorption that is associated with use of this medication. Vitamin B12 deficiency is very dangerous. It causes neurologic damage and impairs production of DNA. The latter problem can't be good since DNA is needed to make nearly all one's cells.

I have found overt vitamin B12 deficiency in several adolescents with PWS who were new to our clinic and who had never had it evaluated. Neurologic symptoms are often not recognized in people who have many other health issues. It is assumed to be just a symptom of the syndrome.

As noted earlier, another potential diabetes-related factor is that inadequacy of the minerals magnesium and chromium will make insulin receptors on the cells be less able to work. In other words, micronutrient inadequacy can be an additional contributor to the increased risk of people with PWS developing and having trouble controlling type 2 diabetes. Assuring adequacy instead of assuming it is always the way to go.

Please see my "Vitamin B12" paper or my "Other Nutrition Issues in Diabetes" paper for more detail on this phenomenon, and some other monitoring issues.]

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**Serving Sizes:**

Except for the meat group, the serving sizes on our PW Pyramid are unchanged from the USDA Food Pyramid. They are as follows:

**Vegetable:** ½ cup cooked or 1 cup raw

**Bread:** 1 slice bread; ½ C. rice, pasta, or starchy vegetable; 1 oz. Cereal

**Fruit:** ½ C. canned, ½ C. or 1 piece fresh, 1/4 C. dried; ½ C. juice

**Milk:** 1 C. skim milk or lite yogurt, 1 oz. cheese, ½ C. frozen fat-free sugar-free yogurt

**Meat:** 2 oz. cooked lean meat, fish, poultry; 1 egg; ½ C. cooked dried beans; 2 T peanut butter

### **Fats, Oils, and Sweets**

The top of the USDA Pyramid shows fats, oils, and sweets. These are denoted by symbols that are concentrated in this area and dispersed throughout the other groups. The USDA suggests that these foods be used sparingly to add extra calories. These foods include butter, margarine, regular dressings, candy, sugars, sweets, fatty desserts, gravy, and fried foods, to name a few. The foods from this group add unwanted calories and few nutrients to the Prader-Willi diet. They should be limited to once a month for an 800-calorie plan and once a week for a 1,200-calorie plan. We have deleted the fat symbols throughout the PW Pyramid, because all foods chosen should be low in fat and sugar.

Using this modified pyramid as a guide to weight loss and maintenance, in conjunction with a favorite exercise program, can be an easy way to ensure a healthy, nutritious diet for the person with Prader-Willi syndrome.

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**[CB note: Following this plan will result in very poor micronutrient balance unless a number of vitamins and minerals are supplemented. This includes inadequacies of many nutrients that are not included in a standard multivitamin with minerals.**

**Other nutrition-related substances are also looking to be important in PWS, including certain special forms of fat and some “conditionally essential” substances that we normally make in adequate amounts ourselves. Conditionally essential simply means that in certain conditions ... like having PWS, for example, ... ready-made versions of the substances are actually essential.**

**An extremely important consideration is that there is clearly a metabolic component to weight issues in Prader Willi Syndrome. They are not being “piggy” or “lazy.” Something in their bodies is not working right and they are trying desperately to compensate for it ... with predictable results:**

**The well-recognized features of PWS include an intense drive to eat even when allowed to consume a very generous amount of food.**

Additionally, they often accumulate very generous body fat, and they tend to have hypotonia (low muscle tone) and very poor exercise endurance. These features of the syndrome make it necessary to look at more than severely cutting their calories and pushing for increase physical activity in an effort to avoid fatness.

Restricting calories does not help the drive to eat at all, nor does it truly solve the excess fat accretion or muscle issues. This collection of symptoms can often reflect a metabolic problem that requires the person to be provided with a substance called carnitine, and our experience with children who have received carnitine supplementation has borne this out.

Carnitine is normally made in ample amounts in one's body, but there are a number of situations in which it is not made in normal amounts, or in which a person has much higher than usual needs. In a nutshell, carnitine has an important role in everyone's ability to burn fat for fuel.

Failure to efficiently burn fat for fuel means that fat will be stored but then it is unable to be made available for use as fuel. That means that for practical purposes, the fat consumed with a meal and the fat already stored in the body do not contribute to the job of running a person's body.

One's brain then makes "getting some fuel" an extremely high priority ... hence the intense food-seeking behavior even in the presence of clearly generous energy stores and a generous amount of food eaten.

Maintaining normal muscle tone also requires carnitine because muscles preferentially burn fat for fuel, especially in endurance activities. Muscle weakness and poor exercise tolerance result from inability to burn fat fuel normally, which then impairs the pursuit of the physical activity encouraged as part of a weight management program. Further, the heart is a muscle ... cardiomyopathy and hypotonia (low muscle tone) is often associated with carnitine insufficiency.

When health care professionals only focus on the caloric restriction issue, they can contribute to big problems for families. They often put the responsibility for preventing obesity on the shoulders of the caretakers with messages like "It's your job to just tell her 'no' when she wants more food." Consider that when health professionals say this kind of thing, they are asking a parent to consistently refuse food to a child who is begging and pleading for it.

Imagine the hungriest you have ever felt, and then imagine that people with PWS feel that way all the time. They experience intense discomfort from hunger ... cutting food intake down to half the amount other people get to eat does not solve this problem. It may prevent obesity to some degree, but it makes their discomfort worse and it can have a very negative on behavior and relationships.

One promising non-nutritional intervention is the use of growth hormone. There are nutrition-related substances under investigation as potentially helpful in muscle operation in PWS as well, such as CoQ-10. In our clinic, we have seen tremendous benefit to individual babies and children from the combination of growth hormone, carnitine and CoQ-10. It has worked better in combination than with the (already standardly used) growth hormone therapy alone. The metabolic adjustment seems to make that expensive growth hormone more effective.

(Whether or not growth hormone therapy would be useful for a child with PWS is something his/her physician would need to determine. My only point here is that IF hormone therapy is tried, it might work better if some of these identified metabolic nutrition issues are addressed before the trial period.)

The benefits include better lean body mass, BMI or weight-for-length, and muscle tone. It has significantly improved the classic intense food-seeking behavior. Severely hypotonic babies who started the carnitine and CoQ-10 upon diagnosis in the nursery have been able to suck well enough to go home without the gastrostomy tube placement often needed in early life. They move more vigorously.

There is a lot more to learn, but the carnitine and CoQ-10 are very safe and very promising in some reports and small studies for children with PWS and other serious medical conditions with metabolic derangements. But large-scale long-term randomly-assigned prospective studies will take a long time to get. For that reason the use of carnitine and CoQ-10 in this application is not the official recommendation of large medical groups yet.

Please note that I do not sell anything nor do I have any relationship with manufacturers of these substances. Additionally, I never go ANYWHERE near “scary” in my recommendations. But if it were MY child who had PWS, I would surely do a trial on these substances while we await the long-term large research studies that are always needed. There

are supportive preliminary reports in the scientific literature, and also anecdotal information from parents and from people like me who follow patients with PWS. For any particular child it will either help or it won't, and I think it is certainly worth a trial. Stay tuned!

One other very new addition to the collection of possibly conditionally essential substances of interest in PWS is "NAC" -- N-acetylcysteine (called Pharma-NAC®) It is showing some promise in addressing the problem of skin-picking that is common in PWS. No recommendation is being made here ... this is not my territory ... but because this is so new, here is the abstract of a study investigating its use so that interested pharmacy and physician folks can look into it further.

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Am J Med Genet A. 2014 Feb;164A(2):421-4. **An open-label pilot study of N-acetylcysteine for skin-picking in Prader-Willi syndrome.**  
Miller JL, Angulo M. Dept of Pediatrics-Endocrinology, University of Florida, Gainesville, Florida.

Prader-Willi syndrome (PWS) is a complex neurodevelopmental disorder caused by an abnormality on the long arm of chromosome 15 (q11-q13) that results in a host of behavioral characteristics including excessive interest in food, skin picking, difficulty with a change in routine, and obsessive and compulsive behaviors. Skin-picking can result in serious and potentially life-threatening infections. Recent evidence suggests that the excitatory neurotransmitter glutamate is dysregulated in obsessive-compulsive behaviors, and modulation of the glutaminergic pathway may decrease compulsive behaviors, such as recurrent hair pulling or skin-picking behaviors. N-acetylcysteine (NAC), a derivative of the amino acid cysteine, is thought to act either via modulation of NMDA glutamate receptors or by increasing glutathione in pilot studies. Thirty-five individuals with confirmed PWS (ages 5-39 years, 23 females/12 males) and skin-picking behavior for more than 1 year were treated with N-acetylcysteine (Pharma-NAC®) at a dose of 450-1,200 mg/day. Skin-picking symptoms and open lesions were assessed after 12 weeks of treatment by counting and measuring lesions before and after the medication. All 35 individuals had improvement in skin-picking behaviors. Ten (29%) individuals (six males and four females) did not have complete resolution of skin-picking behavior, but had significant reduction in the number of active lesions. Longer-term, placebo-controlled trials are needed to further assess the potential benefit of this treatment.

## **Summary about the Use of the PWS Pyramid:**

The PWS pyramid above is a reasonable starting place for approaching a weight problem in general ... it illustrates a simple way to identify the food groups with various calorie levels.

However, the individual who designed it was essentially only addressing the issue of how to cut calories, as she had been requested to do. This discussion is not a criticism of her ... she would not be expected to be aware of the other complex issues of nutrient inadequacy and metabolic disturbance such as those described above. She was asked to help with arranging a pyramid by the calorie content of food groups and she did that nicely.

However, as pointed out in the comments above, there are many other issues besides calories and weight in optimizing the health of individuals with PWS.

There is the potential for doing great harm by just cutting calories back when these other issues are not corrected.

It is very important that the nutrition regimens of people with PWS be carefully evaluated by a health professional familiar with these particular problems.