Upon receipt of a letter from the clinic explaining the Simplified Diet approach, we ask the families to contact us to discuss implementation. A diet change will need to be made to reduce daily phenylalanine or protein <u>allowance from food</u> by 30%. This new diet prescription will accommodate for the change of no longer counting fruits, specific vegetables, and low protein foods.

A baseline level will need to be drawn prior to starting the Simplified Diet and then a weekly level for one month after the diet has been started. We do NOT change the diet prescription at all even if there is an increase in phe levels the first few weeks because we have found that they often return to treatment range when the newness of not counting subsides. Ongoing frequency of Phe level monitoring will be assessed on an individual basis. Based on severity of the individual phenylalanine restriction, the diet may need to be adjusted to maintain levels within treatment range. For example we may need to limit a higher phe fruit (bananas) to 2/day versus unlimited.

We start the following approach on all ages. With infants it makes the introduction to solids easier for families.

Patients already following traditional PKU diet

- Phe from food allowance, not total phe, is reduced by 30%
- Patient/family is given the Simplified Diet Foods list (list we created within our clinic) to begin diet
 - Fruits and vegetables <0.75 mg phe/g are considered Free (uncounted)
 - Low Protein Foods made by low protein manufactures that are <20 mg* phe/serving are considered Free (uncounted)
- Repeat phe levels are requested weekly x 4 weeks to monitor response, adjustments are made if needed to maintain patient on this diet.

* Less than not equal to.

Infants starting solid foods

- Simplified Diet is started immediately when solid foods are introduced (~6 months of age).
- We typically give 15 mg phe from food to start, however, since patient is following the Simplified Diet more should be taken out of the formula to account for the free foods. In this case, 20 mg phe would be reduced from the formula, but only 15 mg would be allowed from the food. So, 15 mg is the approximate yield of: 20 mg 30% = 14 mg phe (15 mg is easier for counting and prescribing).
 - $\,\circ\,$ Another way to do this is:
 - Determine how much phe from food you want to give. Add 30% to that amount. This will determine how much phe you should remove from the formula.

Example: Want to give 15 mg phe from food 15 mg Phe + 30% = 19.5 mg Phe (remove 20 mg Phe from formula)