Maternal PKU

Background

Pregnant women with PKU offer extraordinary risks to their unborn child. Damage is done to the developing fetus if the mother's plasma phenylalanine (phe) level is not well controlled. The higher the mother's phe, the greater the degree of damage is to the fetus. The effects of untreated or uncontrolled maternal PKU include spontaneous abortion, microcephaly, congenital cardiac anomalies, poor intrauterine growth, and mental retardation. Phe levels at the time of conception and in early pregnancy are the most critical. As the pregnancy progresses, the maternal phe tolerance typically increases, often making metabolic control easier to attain.

*** A pregnant woman with PKU is **not** at risk of an emergent metabolic decompensation so hospital admission or transfer to VGH would not normally be required for dietary adjustment. Rather, the AMDC staff speak to such patients almost daily to adjust their diet, especially in the first trimester. If you get called about a pregnant woman with PKU who is vomiting or a patient who believes she is newly pregnant, simply follow the instructions below and let the metabolic dieticians know at 6048755965 and they will take it from there.*

Goals:

1. **Phe in target range of 2-6 mg/dl (120-360 umol/L) – patients think of their levels in mg/dL so if you are talking to them about it, convert umol/L to mg/dL by dividing by 60**
2. 100% of formula intake (and additional supplements when prescribed)
3. Normal weight gain
4. Frequent blood monitoring – patients do this at home using a finger lancet and a blood dot card which then they send to the lab by mail

Treatment:

1. **Phe-free formula**. 100% of protein needs and most micronutrients are met with formula. Remaining calories and nutrients are met through diet and often with additional vit/min supplements. A list of the formulae and their kcal/protein content is attached.

   *** Patients order their own formula (as prescribed by the Metabolic Clinic RD) from Special Products Distribution Centre at BC Children's Hospital 604-875-3020 Hours of operation M-F 10am to 6pm.*

2. **Phe-restricted diet** (very low protein: no meat, dairy, legumes/beans/nuts, eggs, limited starches). Diet is primarily fruit, vegetables, small amount of starches and special low protein food products.

3. **Adequate calorie and protein** intake through formula and diet is necessary to keep phe levels well-controlled as a deficiency of either total calories or protein can lead to elevated phe levels from breakdown of body tissues to meet energy and pro demands.

<u>If Your Phe is Over 6 mg% (360umol/L) and you are ill or if you think you might be newly pregnant and don't yet know what your Phe level is:</u>

1. Take a blood dot measurement and send to lab immediately
2. Reduce dietary phe to 200mg (consume only low protein foods/starches, fruits and vegetables)
3. Take 100% of your formula prescription (this meets 100% of pro needs)
4. Consume phe-free foods to meet your calorie needs (list below)
5. Keep detailed food records and send to clinic dietitian ***daily***
6. Daily blood work

<u>If you are Vomiting</u>

1. See your family physician to discuss treatment of the cause of vomiting. They may be able to provide a safe medication that you can take in pregnancy (e.g. Diclectin for morning sickness)
2. Concentrate on trying to get all of your PKU formula in each day – this is the main priority. Take small sips every 5-10 minutes. Chilling the formula (or blending it with ice cubes) can make it easier to get down.
3. Ensure you stay well hydrated – drink low or no phe beverages (preferably with lots of calories), if you can tolerate them. Regular Ginger ale is a good choice – sip on this throughout the day (see list below for other ideas)
4. Send in daily blood dots so that the clinic can monitor your phe levels
5. Keep daily food records and send them to your dietitian *daily*

<u>If Your Phe is Over 6 and you aren't sick with flu/fever:</u>

Phe above 6 mg%: follow these steps

1. Identify any foods which may have contributed to excess dietary phe and stop using (large portions of starches or high protein foods)—if you aren't sure if a food caused a high level, contact your dietitian and review your daily food records with her
2. Make sure you are taking your full formula prescription, and that you are taking it 3-4 x/day and that you eat a calorie containing food with the formula i.e. formula and applesauce or formula and low protein crackers/margarine
3. Reduce dietary phe by 10% for 3 days and then redo a blood dot
   i.e. if you are aiming for 250 mg phe and have been hitting

that 250 on the mark but then end up with a high phe, you should take in 250 – 10% = 250 – 25 = 225 mg phe for 3 days
NOTE: if after 3 days the phe comes back still high, further reduce phe by another 10% i.e. 225 x 10% = 225 – 23 = 202 mg phe/day

4. Make sure your calories are meeting or slightly above prescribed calories for pregnancy (your dietitian will tell you how many calories you need) - **too few calories can cause high phe levels**
5. Keep detailed food records and send to clinic dietitian ***daily***
6. Daily blood work
7. If none of this works, your dietitian may increase how much protein you are getting from your formula - **too little protein can cause high phe levels**

Free Foods

Food	Phe (all items listed as 0 are 0-1 mg per serving indicated)	Calories	Protein
Gatorade ½ c	0	25	0
Hawaiian punch 1/2c	0	60	0
Kool-aid ½ c	0	50	0
Ocean spray cran-rasperry ½ c (other flavours to)	0	85	0
Powerade ½ c	0	35	0
Regular 7-up/sprite 355 ml can	0	145-150	0
Iced tea (sweetened) ½ c	0	45-55	0
Tang crystals (1 tbsp)	0	90	0
Lemon drops (1ea)	0	10	0
Ener-g low pro cookies (1 ea)	0	120-160	0
Starburst fruit chews 1 piece	0	20	0
Sweet tart candy (8 pieces)	0	60	0
Juicy jells(3.5 oz cup)	0	78	0
Hunts lemon	0	125	0

pudding snack pak(3.5 oz cup)			
Ready to spread icing (1 tb)	0	70	0
Popsicles (scribblers, juice jets, ice pops)	0	45-60	0
Cotton candy (1 cone)	0	300	0
Energy Option Bars	Ask your dietitian		

The following are estimated recommendations from the Protocol for Nutrition Support of MPKU and can be used as a guide until you discuss with your dietitian:

Calories: 1st trimester: 2000-3000 avg: 2200 protein >74 g/d
2nd trimester: 2000-3000 avg: 2500 protein >74 g/d
3rd trimester: 2000-3000 avg: 2500 protein >74 g/d

Never eat less than 35 calories/kg of your body