



Adult PKU Program
University of Washington, Box 357920
Seattle, WA 98195

Dear.....

Given your diagnosis of phenylketonuria (PKU), we recommend that you continue to follow the University of Washington Adult PKU Program Treatment Guidelines:

1. Maintain the phenylalanine level in your blood between 1 to 10 mg/dl.
2. Submit a monthly sample of your blood to the laboratories at Children’s Hospital (CHRMC) or the Washington State Newborn Screening Program for testing.
3. Be seen in the University of Washington Adult PKU Program at least once a year.
4. Attend one or two meetings per year for adults with PKU.

It is our understanding that you have made a decision not to follow these treatment recommendations. We are concerned that you have made this decision because, regardless of how well your metabolic disorder was managed during childhood and adolescence, any adult with PKU whose blood phenylalanine levels run consistently above 10 mg/dl increase their risk of experiencing any or all of the following:

- Headaches
- Increased body odor
- Increased hair loss
- Mild to severe skin rash
- Impairment of short term memory
- Increased irritability (crankiness)
- Inability to cope with life
- Greater prevalence of thought disorders and mood disturbances
 - Increased problems with depression
 - Increased problems with impulse control
 - Increased fear of leaving the house
 - Increased problems with violent behavior
- Possible blackouts or periods of loss of consciousness
- Possible uncontrolled seizures
- Potential spasticity. This is involuntary muscular contraction and/or rigidity. This could result in loss of mobility and potentially may require you to use a wheelchair.
- Potential significant damage to the central nervous system and the brain. This could result in a decrease in your ability to use your brain effectively.

Females of childbearing years

For women with PKU who become pregnant with blood phenylalanine levels of greater than 4 mg/dl, there are additional consequences to your unborn child.

- There is increased risk of spontaneous abortion.
- There would be a high risk of organ abnormalities (problems with the heart and lungs).
- There is a high risk of microcephaly (small brain) which could cause mental retardation in your child.

It is not known how long it will take an individual with PKU who has blood phenylalanine levels greater than 10 mg/dl to experience some, or all, of these difficulties. It has been documented that each individual off treatment will experience some, or all, of these medical, intellectual, and emotional problems. As an adult, it is your decision whether or not to manage your PKU by conforming to the recommended treatment.

By signing your name at the bottom of this page you signify that you understand the possible consequences of terminating treatment and that you do so against the advice if the University of Washington’s Adult PKU Program.

Name

Date

Signature of Health Care Professional

Date

If you as an adult with PKU choose to return to recommended treatment, many of the emotional, medical, and intellectual difficulties can be greatly decreased if blood phenylalanine levels drop to less than 10 mg/dl. The resources of the Adult PKU Program are available to you should you decide to resume recommended treatment. Call the Social Worker for the Adult PKU Program at (206) 543-2499.



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Patient Agreement

Please read the following information carefully. If you have any questions regarding the program or its services, please contact the PKU Program Social Worker at (206) 543-2499.

Program benefits are available to all adults with phenylketonuria (PKU) in the State of Washington. The Adult PKU Program's annual recommended treatment includes:

1. **Nutrition:** nutrition assessments, nutrition counseling as needed and provision of education materials
2. **Medical:** yearly neurological exam, mandatory visit for **annual prescription** for formula and coordination of general health care with primary care physician
3. **Social work:** help with financial support for formula and clinic visits, coordinating clinic appointments and interfacing with appropriate community agencies
4. **Psychometric testing:** evaluation of cognitive functioning
5. Reporting blood phenylalanine levels to you
6. 1-2 meetings per year for adults with PKU (social, support group, research updates)

I, _____ request Adult PKU Program services from the Lipid and Nutrition Clinic at the University of Washington. I understand that if any of the following occur, I will lose my eligibility for services through the University of Washington Adult PKU Program including some benefits funded by the State of Washington.

1. If I move away from the State of Washington.
2. If I fail to make an annual clinic visit or follow-up appointments as requested by the Adult PKU program.
3. If I fail to do blood phenylalanine levels as requested by the Adult PKU Program.

Note: Blood levels should be done once per month. Recent blood levels are needed before formula can be shipped to you.

4. If I fail to take personal responsibility for the cost of my PKU formula by failing to secure insurance coverage for formula and clinic visits through: a) my work or b) my parents' policy or c) have failed to apply for medical coupons or d) the Washington Basic Health Plan.

We ask that you notify us when you change addresses and/or telephone numbers. For billing purposes, we must know about changes in insurance coverage.

I understand that if I choose not to participate in the Adult PKU Program, I will be responsible for finding another qualified health care provider to assist me with the management of my PKU.

By signing this partnership agreement, I agree to comply with the conditions of the Patient Agreement.

Signature of Client

Date

Signature of Health Care Professional

Date

Original to medical record

Copy to client

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