National PKU Alliance
P.O. Box 501 • Tomahawk, WI 54487-0501 • Phone: 715-437-0477 • Fax: 715-453-7670 • www.npkua.org

Treatment recommendations	2013	2000	Before 2000
Blood Phe levels	Maintaining blood phenylalanine (Phe) levels in the range of 120–360 µmol/L (2–6 mg/dL) in all patients of all ages is essential for people with PKU [ref PKU Guidelines 2013]	Lifelong treatment of PKU was first officially recommended with Phe levels maintained at: • **120–360 µmol/L** (2–6 mg/dL) for newborns and children up to the age of 12 years • **120–900 µmol/L** (2–15 mg/dL) after 12 years of age	Most clinics recommended lifelong management of PKU. No consensus existed for optimal blood Phe levels.
Phe levels warranting treatment	Infants with blood Phe levels >600 µmol/L (>10 mg/dL) require treatment. Those with sustained levels >360 µmol/L (>6 mg/dL) are recommended to have treatment. Those with levels of 120–360 µmol/L **(2–6 mg/dL)** should be monitored, but not treated. Patients with PKU who were treated in early life but stopped treatment should resume treatment. They may not be aware of the need for lifelong treatment and may be suffering from neurocognitive impairment due to poor metabolic control.	Individuals with blood Phe levels <360 µmol/L (<6 mg/dL)	Infants with PKU who have blood Phe levels >600 µmol/L (>10 mg/dL) should be started on treatment to establish metabolic control of Phe levels by seven to 10 days of age. Most US clinics targeted 120–360 µmol/L (2–6 mg/dL) for individuals younger than 12 years of age, and 120–600 µmol/L (2–10 mg/dL) for persons older than 12 years. Medical nutritional therapy should be initiated in newborns with levels between 420–600 µmol/L (7–10 mg/dL) that persist more than a few days.

Monitoring recommendations	Blood Phe and tyrosine (Tyr) monitoring should be performed: At least weekly until the age of one, and then with increased surveillance during periods of rapid growth and dietary changes. From age one until 12 years, on a twice-monthly to monthly basis In adolescents and adults who have stable and well-controlled Phe levels, on a monthly basis People with PKU should also be tested for plasma amino acids (full panel), transthyretin, albumin, complete blood count, ferritin, 25-OH vitamin D, vitamin B12, red blood cell, essential fatty acids, trace minerals (zinc, copper, and selenium) vitamin A, comprehensive metabolic panel, and folic acid	• Once weekly during the first year • Twice monthly from one to 12 years of age • Monthly after 12 years of age	During the first year, once a week to once a month, with once a week being more common. After the first year, move from monitoring once every month to once every three months.