Self-Management Skills and Treatment Knowledge in Patients with Phenylketonuria

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**Objective:** To describe patient-reported self-management behaviors and treatment knowledge in patients with PKU and explore how these correlate with blood phenylalanine (Phe).

**Results:**
- **n=43,** 53% female
- Treatment knowledge associated with participation in corresponding behaviors
- Learning impairment (n=11) not significantly associated with self-management (p=0.054)
- Behavior/knowledge scale not significantly associated with mean Phe after controlling for number of blood samples (R<sup>2</sup>=0.249; p=0.024)

**Methods:**
- Cross-sectional survey
- English-speaking patients with PKU aged 7-30 years attending clinic within the 1 year enrollment period
- All patients prescribed a low protein diet with medical food
- Administered age-specific self-management questionnaire (7-12, 13-17, and 18-30 years)
- Content adapted from the University of Washington’s PKU Self-Management Timeline

**Outcome Variables**
- Scale variables created as overall measures of self-management behaviors and treatment knowledge
- Regarding dietary Phe/protein, medical food, and blood Phe samples
- Blood Phe levels within 12 months of enrollment
- Individualized education program (IEP) eligibility used a proxy for learning impairment
- Data analysis: descriptive statistics, Spearman correlations, and multivariate linear regression
- Significant at alpha p<0.01

**Discussion and Conclusion:**
- Participation in recommended behaviors and treatment knowledge was lower than clinically recommended.
- Increased self-management behaviors and knowledge were not significantly associated with lower Phe levels.
- Results may reflect small sample size.
- Additional research is needed to investigate strategies to improve self-management behaviors.

**Practice Implications:**
We recommend using the surveys or a similar tool as a checklist in a clinical setting to facilitate regular discussion of self-management skills.