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### TITLE: Adequacy of natural protein intake in patients with phenylketonuria: bibliographic review

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#### **ABSTRACT**

#### **Introduction:**

Phenylketonuria (PKU) is the most frequent inborn error of protein metabolism. The dietary treatment consists of a diet with a restricted natural protein intake, a free or low phenylalanine (Phe) protein substitute, and the intake of low protein food. The main objective of this work is to analyze if it is possible to increase the natural protein (NP) intake prescribed to PKU patients while maintaining blood Phe dosages within safe range. Materials and method: studies published were searched in 6 electronic data-basis. A total of 154 Pub Med articles were identified by range of years from 1999 to 2020. Fifteen articles which met the inclusion and exclusion criteria and responded to the objective of this bibliographic review were chosen.

Results: several factors may influence Phe tolerance, such as severity of the patient's phenotype, age, blood Phe safe range, Phe prescription and adherence to protein substitute. If Phe blood levels remain constantly within safe range and for a certain period, an increase of Phe intake should be considered. Increase of NP intake must be carried out in a controlled manner, individually and constantly evaluating blood Phe levels.

Conclusion: optimizing NP intake offers the PKU patient an improvement in quality of life, facilitates the patient's ability to socialize and contributes to a better adherence to the diet.

#### **BIOGRAPHY**

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