

NeoPhe[®]

LNAA TABLETS[®]

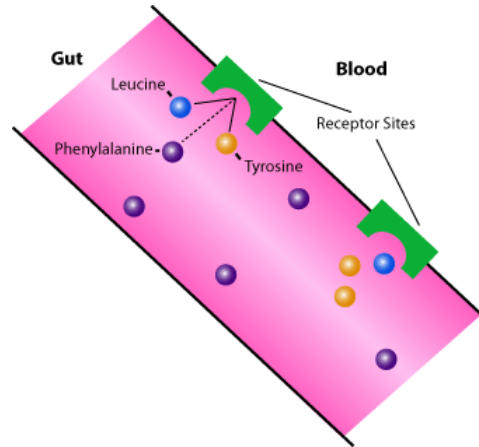


The clinically proven LNAA tablet that blocks Phenylalanine from entering the blood.

Mechanism of Action

NeoPhe's formulation (of large neutral amino acids and cationic amino acids) has been optimized to inhibit the transport of Phenylalanine from the gut into the blood.

Large neutral amino acids and cationic amino acids cross the intestinal mucosa by means of a carrier protein. High concentrations of LNAA compete with Phe at the GI transporter thus decreasing the amount of Phe that crosses through the GI tract¹.



Tyrosine, Leucine and other LNAA are known to be deficient in the PKU patient.

NeoPhe allows for lower blood Phe levels and corrects deficiencies of essential amino acids – the “gold standard” of treating PKU patients.

Pilot and DBPC Trials Showed

Blood Phe concentration in the 8 patients taking 0.5 g/kg per day of LNAAs decreased from an average of 957.4 $\mu\text{mol/dl}$ to an average of 458.4 $\mu\text{mol/dl}$, a decline of 52%. Three patients took 1 g/kg per day of LNAAs. Their baseline average was 1230 $\mu\text{mol/dl}$ compared to an average of 549.0 $\mu\text{mol/dl}$, an average decline of 55%. All patients experienced a decrease in blood Phe concentrations from baseline after NeoPhe. The average decrease was 601 $\mu\text{mol/dl}$ (SD = 370), and when analysed together (N = 11) this drop in blood Phe was highly significant ($p = 0.0003$). When treatment was discontinued, blood Phe concentrations increased to pre-trial levels¹.

Double blind studies performed in 7 different centers indicate that LNAA can compete with phe on the transporter in the GI tract, resulting in a decrease in the blood phe-level in patients on NeoPhe, averaging 39% from baseline levels.²

Starting a Patient on NeoPhe

The dosage that has proven to lower blood Phenylalanine is 1 tablet / kg / d¹.

- Have patient maintain the same diet plan.
- Start NeoPhe at 1 tablet/kg/d.
- Monitor blood Phe levels once every 1-2 weeks.

- As blood Phe levels decrease, adjust diet (*decrease NeoPhe or decrease medical food*)

Why is there a Dose Range?

Patients new to LNAA therapy should start on 1 tab/kg/d. Depending on response of Phe levels, NeoPhe can be decreased as low as 0.5 tablets/kg/d.

Patients transitioning from PreKUnil to NeoPhe should continue to take the same dose as PreKUnil (usually 0.5 tablets/kg/d).

How Long will it Take to Reach Optimal Control?

Every patient is different. Clinical trials showed an average of 7 days before goal blood Phe levels were achieved and optimal daily NeoPhe intake was determined¹.

When Can Protein from Regular Food be Introduced?

Once NeoPhe has lowered blood Phe to desired levels, traditional PKU diets can gradually be discontinued and regular food of the same protein category can be introduced.

Suggested Progression to Regular Protein Containing Foods

- Discontinue the equivalent of non animal protein medical foods first (low protein baked goods...). Replace protein from the medical food with regular food (grains, cereals).
- Discontinue the equivalent of animal sources of protein (Formulas) and replace the protein from the formula with animal sources of protein (dairy products, meat, poultry, fish, eggs).

Is there a Maximum Number of Tablets a Patient Can Take Per Day?

Although the recommended starting dose is 1.0 tablet/kg/d, some patients may require a larger dose to obtain goal blood Phe levels. Intake of NeoPhe should not exceed 2.0 tablets/kg/d.

Ordering NeoPhe

(888) 8-SOLACE (876-5223)

Fax: 401 633-6066

Reimbursement

Reimbursement code: 57771-0050-01



A Medical Food for the dietary management of PKU

REFERENCES

- 1) R. Matalon, K. Michals-Matalon, G. Bhatia, E. Grechanina, P. Novikov, D. J. McDonald, J. Grady, S.K. Tyring, F. Guttler, Large Neutral Amino Acids in the Treatment of Phenylketonuria (PKU), *J Inherit Metab Dis.* 2006 Dec;29(6):732-8.
- 2) R. Matalon, K. Michals-Matalon, G. Bhatia, A.B. Burlina, A.P. Burlina, C. Braga, L. Fiori, M. Giovannini, E. Grechanina, P. Novikov, J. Grady, S.K. Tyring, F. Guttler, Double blind placebo control trial of large neutral amino acids in treatment of PKU: Effect on blood phenylalanine. *J Inherit Metab Dis.* 2007, Feb 27.