

LNAAs IN THE TREATMENT OF PKU

The four letters "LNAA" stand for "Large Neutral Amino Acids".

The Large Neutral Amino Acids (LNAAs) are Phenylalanine, Tryptophan, Tyrosine, Histidine, Methionine, Threonine, Leucine, Isoleucine, and Valine.

These LNAAs use the same transport pathway as phenylalanine to cross from the bloodstream into the brain. As a result, they compete with phenylalanine for the same transporter at the blood–brain barrier.

When consumed in sufficient amounts, LNAAs can therefore inhibit the transport of phenylalanine into the brain. Consequently, less phenylalanine reaches the brain, helping to reduce the harmful accumulation of phenylalanine in this critical organ.

Studies also show that LNAAs may help lower phenylalanine levels in the brain while improving the availability of other essential amino acids.

LNAA therapy may be particularly beneficial for individuals who have difficulty maintaining their phenylalanine levels within the recommended target range with the conventional PKU diet alone. Although blood phenylalanine levels often do not decrease during LNAA therapy, less phenylalanine reaches the brain. This can help reduce the negative effects of elevated phenylalanine levels in the brain.

According to current European guidelines, there is still insufficient scientific evidence to recommend LNAA therapy for routine use for all individuals with PKU. In particular, this therapy is **not currently recommended** for **children**, as there is not yet enough scientific evidence regarding its effectiveness in this age group. **LNAA therapy must also not** be used during **pregnancy**.

MORE FLEXIBILITY IN FOOD CHOICE

LNAA therapy can broaden the range of foods that individuals with PKU are able to enjoy. For example, regular bread, rice, and pasta can often be eaten in adequate amounts.

However, foods rich in protein, such as meat, fish, dairy products, eggs, and legumes still need to be limited. In most cases, these restrictions are generally less strict than with the classical PKU diet.



PRODUCT INFORMATION



XPHE **LNAA**
minis

TABLETS
WITH LNAAs

SUPPLEMENTED WITH
Vitamins
Minerals
Trace elements

FROM 18 YEARS OF AGE



XPHE **LNAA**
minis

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36/06/24 09/11/21

FOOD FOR SPECIAL MEDICAL PURPOSES
FOR THE DIETARY MANAGEMENT OF

**PHENYLKETONURIA (PKU)
OR HYPERPHENYLALANINEMIA (HPA)**

XPHE LNAA minis



TABLETS — EASY & PRACTICAL

- protein supplement free of Phenylalanine in tablet form
- enriched with a high content of Large Neutral Amino Acids (LNAA)
- with vitamins, minerals and trace elements
- without carbohydrates and only a small amount of fat
- suitable from 18 years of age – must not be used in the time period before and during pregnancy

highly concentrated and balanced

CONVENIENT

- ready for application, absolutely no preparation necessary
- can be consumed discreetly
- do not draw attention to your dietary product – this is what makes good compliance rather easy
- packed in sachets – convenient and time saving – quick to calculate

1 Sachet $\triangleq$ 24 tablets $\triangleq$ 10 g protein eqv.

DISCREET

- no unpleasant aftertaste
- no awkward bad breath
- no annoying belching

very pleasant – even after taking

PERFECT „TO GO“

- simply take along – fits in every pocket
- ideal for on the go – for school and job, for leisure and sports
- ideal for traveling (e.g. by plane) – little pack size, low weight, instantly ready to take

comfortable and always handy

DOSAGE

- The daily total amount of amino acid mixture depends on age, body weight and individual medical condition and/or protein tolerance. The dosage should be re-examined and adjusted according to the results of regular medical monitoring.
- The daily dosage of protein substitute should at best be divided into 3 – 5 single portions.
- The diet with XPhe LNAA minis can be supplemented with other amino acid mixtures from the XPhe-products if protein intake from food is insufficient.
- The PKU-diet must be supplemented with energy, natural protein and other nutrients in prescribed quantities.

INTAKE

- XPhe LNAA minis should be swallowed unchewed as whole tablets. They should always be taken with a sufficient quantity of liquid. The tablets are not meant to dissolve in the mouth. Do not dissolve the tablets in water or other liquids. We recommend to take them in an upright position. Once sachet is opened, tablets should be taken within the next 6 hours.
Note: XPhe LNAA minis are coated with a flavour-neutral layer. This outer coating makes the tablets easy to swallow. The protective coating is extremely thin and water-soluble. If this coating were to dissolve, the tablets would taste like a typical amino acid mixture.
- XPhe LNAA minis should preferably be taken along with calculated amounts of Phe-containing food according to individual protein tolerance.

TOOTH-FRIENDLY

- the coated tablets are tooth-friendly
- taking them with (mineral) water makes an important contribution to oral hygiene
- helps to prevent possible damage to the teeth caused by caries

IMPORTANT NOTICE Must only be used under medical supervision. Not for use as a sole source of nutrition. For enteral use only. Only for people with proven Phenylketonuria (PKU) or Hyperphenylalaninemia (HPA). Suitable from 18 years of age. Should not be used in the time period before and during pregnancy.

NUTRITION INFORMATION

		100 g	24 tablets (1 sachet)
• Energy	kJ	1130	187
	kcal	268	44
• Fat	g	1	0,2
of which saturates	g	1	0,2
• Carbohydrate	g	0	0
of which sugars	g	0	0
• Fibre	g	14	2,3
• Protein eqv.	g	60	10
Amino acids	g	72	12
• Salt	g	0	0

- supplemented with
- ✓ vitamins
 - ✓ minerals
 - ✓ trace elements



30 sachets at 24 tablets