The effect of slow releasing carbohydrates on the variability of plasma amino acid concentrations in PKU-patients in childhood.

Published: 19-12-2008
Last updated: 10-05-2024

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Ethical review	Approved WMO
Status	Pending
Health condition type	Protein and amino acid metabolism disorders NEC
Study type	Interventional

Summary

ID

NL-OMON31050

Source
ToetsingOnline

Brief title
Slow releasing carbohydrates and Phenylketonurie

Condition

- Protein and amino acid metabolism disorders NEC

Synonym
aangeboren stofwisselingsziekte, Phenylalanine hydroxylase deficiency

Research involving
Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Groningen

Source(s) of monetary or material Support: bedrijf,dieetpreparaten: Vitaflo International

Intervention

Keyword: Diurnal variation Phe concentration, Phenylalanine, Phenylketonuria, Slow releasing carbohydrates

Outcome measures

Primary outcome

Differences in blood Phe concentrations (and other amino acids?) between early morning (A) and end of the day (B) in the two study periods under the different conditions (without intervention, with SRC and with SRC) will be compared.

Secondary outcome

Relationship between the severity of the disease defined as the individual Phe tolerance at 5 years of age, and the resulting mean and SD of blood Phe concentrations of each period will be tested with univariate linear regression analysis (ANOVA).

Study description

Background summary

Metabolic control in PKU is based on regular blood sampling to measure plasma Phe concentration. The intake of Phe is adjusted when plasma Phe concentrations are not within the target range, taking into account other factors that influence metabolic control: e.g. intercurrent infections, growth, energy intake and amino acid supplementation. Studies on the diurnal variation of plasma Phe concentrations, showed an inverse pattern when compared with healthy controls: in PKU patients Phe concentrations measured after an overnight fast are the highest, and decrease during the day. A hypothesis for the rise in blood Phe concentration overnight is the occurrence of catabolism during the fasting period resulting in a higher amount of Phe available than

the amount used for protein synthesis.

Study objective

A reduction of the fasting period may prevent or reduce the rise in plasma Phe concentration. Therefore, the aim of the study is to measure the effect of the application of slow releasing carbohydrates (SRC) as evening meal before bedtime on the overnight fasting blood Phe concentration in comparison to fast releasing carbohydrates (FRC).

Study design

This pilot study has a randomized double-blind cross-over design

Intervention

Addition of SRC or FRC to the 3d portion of amino-acid supplementation at bedtime: the additional carbohydrates will be diluted in the fluid amino acid supplement with some extra water. When the amino acid supplement is not taken as a fluid, the additional carbohydrates will be solved in water and (optional) flavouring (flavor sachets Vitaflo).

Study burden and risks

Extra risico's ten opzichte van de gebruikelijke behandeling worden bij de studie niet verwacht. De extra inname van koolhydraten kan de uitgesproken smaak van de gebruikelijke aminozuren wel enigszins beinvloeden en zal ook het volume van deze suppletie vergroten, maar de verwachting is dat dit geen grote problemen zal geven.
Participating the study includes during 2 periods of 8 days:
- twice daily a fingerpuncture, daily report of stool and body weight
- during 2 of the 8-days period food record registration
- taking the individual diet as usual for the patient with exception of the third portion amino acid supplementation, to which ad bedtime in the intervention period slow and fast releasing carbohydrates will be added conform the study protocol.
The study will be performed at home, and the extra effort of the participants concerns mainly the higher frequency of bloodsampling with a fingerpuncture. Risks associated with the study are not to be expected. May be the extra intake of carbohydrates will influence the taste and volume of the amino acid mixture but is not expected as problematic as the mixture itself has a very pronounced taste.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Children (2-11 years)

Inclusion criteria

They are in good metabolic control, defined as a blood Phe concentration measurement within the target range of 120-360 µmol/l (Dutch PKU Advice Committee) one month prior to the study period. Apart from the PKU the patients are in a healthy state during the study periods. Their growth figures are within +/- 2 SD of the national growth charts. Patients take their amino acid supplementation divided into three portions each day. Participants / parents or caretakers are capable of routinely home skin puncturing without problems.

Exclusion criteria

Blood Phe concentrations frequently beyond the target range, bad compliance in intake of the amino acid mixture, growth retardation > -2SD, obesity > +2SD, insufficient knowledge of the Dutch language of the caretaker, not taking blood samples at home

Study design

Design

Study type:	Interventional
Intervention model:	Crossover
Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Treatment

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	01-11-2007
Enrollment:	6
Type:	Anticipated

Ethics review

Approved WMO

Application type:	First submission
Review commission:	METC Universitair Medisch Centrum Groningen (Groningen)

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL18797.042.07