

The role of microbiome in urea cycle defect disorders

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In urea cycle defect patients the protein-restricted diet, in combination with essential amino acid supplementation, results in a different gut microbial composition compared to healthy individuals on a normal diet. Selection of ammonia producing...

Ethische beoordeling	Niet van toepassing
Status	Werving nog niet gestart
Type aandoening	-
Onderzoekstype	Observationeel onderzoek, zonder invasieve metingen

Samenvatting

ID

NL-OMON25258

Bron

NTR

Verkorte titel

UCD

Aandoening

UCD; Urea cycle defect; ureumcyclusdefect; PKU; Pheylketonuria; fenyylketonurie; microbiome; microbiom; gut bacteria; darmbacterien

Ondersteuning

Primaire sponsor: AMC

Overige ondersteuning: ESN (erfelijke stofwisselingsziekten Nederlands taalgebied).

Onderzoeksproduct en/of interventie

Uitkomstmaten

Primaire uitkomstmaten

differences in the abundance of bacterial strains between healthy volunteers and UCD

patients and between PKU and UCD patients

Toelichting onderzoek

Achtergrond van het onderzoek

Urea cycle disorders (UCDs) are a group of rare inherited metabolic diseases causing hyperammonemic encephalopathy. Despite intensive dietary and pharmacological therapy, outcome is poor in subset of UCD patients. Because of very low protein tolerance, UCD patients follow a strict low protein diet. Both the composition of the diet, as well as the amino acid supplementation these patients use could change gut microbial composition, with a potential negative effect on metabolic control.

Hypothesis:

In urea cycle defect patients the protein-restricted diet, in combination with essential amino acid supplementation, results in a different gut microbial composition compared to healthy individuals on a normal diet. Selection of ammonia producing bacteria can negatively influence metabolic regulation in UCD patients.

Objective:

To detect differences between microbiome composition of UCD patients, healthy controls and phenylketonuria (PKU) patients. PKU patients are included to differentiate between the effect of a low protein diet and the UCD itself on microbial composition)

Doel van het onderzoek

In urea cycle defect patients the protein-restricted diet, in combination with essential amino acid supplementation, results in a different gut microbial composition compared to healthy individuals on a normal diet. Selection of ammonia producing bacteria can negatively influence metabolic regulation in UCD patients

Onderzoeksopzet

single measurement

Onderzoeksproduct en/of interventie

one day food diary
and
a one time stool (feces) sample

Contactpersonen

Publiek

C. Timmer
Amsterdam
The Netherlands

Wetenschappelijk

C. Timmer
Amsterdam
The Netherlands

Deelname eisen

Belangrijkste voorwaarden om deel te mogen nemen (Inclusiecriteria)

Patients:

- age 18 years and over.
- confirmed diagnosis of a urea cycle defect or phenylketonuria (enzymatic or mutation analysis and amino acid spectrum)
- treatment with a low protein diet (natural protein intake 0.8 gram/kg/day or lower) and the use of amino acid supplements

Healthy subjects:

- age 18 and over.

Belangrijkste redenen om niet deel te kunnen nemen (Exclusiecriteria)

Patients:

- known non-adherence to the diet
- known incompliance with taking the amino acid supplements
- use of antibiotics three months prior to sample collection
- other severe conditions that may influence gut microbiome composition (e.g. liver cirrhosis, renal failure, inflammatory bowel disease)

Healthy subjects:

- illnesses known to influence microbiome composition (e.g. liver disease, bowel disease, diarrhea)
- medication use known to influence microbiome

Onderzoeksopzet

Opzet

Type:	Observationeel onderzoek, zonder invasieve metingen
Onderzoeksmodel:	Parallel
Toewijzing:	N.v.t. / één studie arm
Blinding:	Open / niet geblindeerd
Controle:	N.v.t. / onbekend

Deelname

Nederland	
Status:	Werving nog niet gestart
(Verwachte) startdatum:	01-03-2017
Aantal proefpersonen:	45
Type:	Verwachte startdatum

Ethische beoordeling

Niet van toepassing

Soort:

Niet van toepassing

Registraties

Opgevolgd door onderstaande (mogelijk meer actuele) registratie

Geen registraties gevonden.

Andere (mogelijk minder actuele) registraties in dit register

Geen registraties gevonden.

In overige registers

Register	ID
NTR-new	NL6106
NTR-old	NTR6447
Ander register	NL61031 : ABR nummer

Resultaten