Cognitie en het ontwikkelende brein in galactosemie.

No registrations found.

Ethical review	Positive opinion
Status	Recruitment stopped
Health condition type	-
Study type	Observational non invasive

Summary

ID

NL-OMON22209

Source
NTR

Health condition

Classic galactosemia (Klassieke galactosemie)

Sponsors and support

Primary sponsor: Dr. M.E. Rubio-Gozalbo, academisch Ziekenhuis Maastricht
Source(s) of monetary or material Support: Galactosemie Onderzoek Fonds (GOF)

Intervention

Outcome measures

Primary outcome

1. The performance on the language-related behavioural paradigms (four paradigms, each focusing on one aspect of speech processing or production);

2. The event-related potentials (ERPs) extracted from the EEG recorded during the behavioural tasks and.

Secondary outcome

The performance on the neuropsychological test battery (i.e. a composition of tests focusing on the major cognitive domains: memory, attention, speech and language, visuo-motor skills).

Study description

Background summary

Rationale:

Classic galactosemia is a hereditary disorder caused by the body's inability to breakdown galactose, a sugar mainly found in milk. A galactose-restricted diet (soy diet) is the basis of therapy. This, however, does not prevent the emergence of long-term complications. One of these long-term complications is related to cognition. Reduced intelligence quotient (IQ) scores as well as impairments in speech and language have been reported. In fact, ninety percent of affected children have speech problems, usually diagnosed as Childhood Apraxia of Speech (CAS) (i.e. an impairment of motor programming of speech musculature). However, at present, CAS is over-diagnosed. This means that there are patients that receive a treatment for CAS, while other speech disabilities are being overlooked and not addressed properly resulting in suboptimal treatment. Our preliminary research concerning speech shows that syntax is a major problem in these children. The innovative approach in this study, examining the different levels of speech (e.g. semantic and syntactical speech) in combination with behavioral data and simultaneous brain activity recordings, aims to elucidate which levels of speech are specifically impaired in these children. Pinpointing at which level speech and other cognitive functions are affected is imperative to design successful treatment approaches with a better outcome.

Objectives:

To elucidate which areas of speech are specifically affected in children and adolescents with classic galactosemia; and to sketch a profile of the galactosemic patients' general cognitive functions and of the development of these cognitive functions.

Study design:

Observational case control design consisting of a neuropsychological assessment and a behavioural paradigm with simultaneous electroencephalographic brain activity recording.

Study objective

Specific areas of language production are affected in children and adolescents with classic galactosemia, in particular syntactic production. Further, it is hypothesized that the galactosmic patients will show a profile of general cognitive functions deviating from typically developing children and adolescents.

Study design

Two sessions will be scheduled: One for the neuropsychological test and one for the EEG study.

Intervention

N/A

Contacts

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Eligibility criteria

Inclusion criteria

For patients, a diagnosis of classic galactosemia as diagnosed by GALT enzyme activity assay or GALT-gene mutation analysis (information obtained from treating physician); Age between 10 and 18 years old.

Exclusion criteria

Any other disorder or disease that could affect cognitive functioning independently of classic

galactosemia (an exception is made for the diagnosis of ADHD, because this disorder is common in this group).

Study design

Design

Study type:	Observational non invasive
Intervention model:	Factorial
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	N/A , unknown

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	01-08-2009
Enrollment:	52
Type:	Actual

Ethics review

Positive opinion

Date:	26-04-2011
Application type:	First submission

Study registrations

Followed up by the following (possibly more current) registration

ID: 32979
Bron: ToetsingOnline
Titel:

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

No registrations found.

In other registers

Register	ID
NTR-new	NL2731
NTR-old	NTR2869
CCMO	NL27398.068.09
ISRCTN	ISRCTN wordt niet meer aangevraagd.
OMON	NL-OMON32979

Study results

Summary results
N/A