CFTR protein expression in cells from CF patients: correlation with clinical, functional, and inflammatory parameters.

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Main objective is to assess CFTR protein expression in nasal epithelial cells and in different peripheral blood immune cell subsets from CF patients with different CFTR gene mutations. Secondary objective is to correlate these expression levels with...

Ethical review	Approved WMO
Status	Recruitment stopped
Health condition type	Chromosomal abnormalities, gene alterations and gene variants
Study type	Observational non invasive

Summary

ID

NL-OMON38139

Source
ToetsingOnline

Brief title
CFTR protein expression in CF patient cells

Condition

- Chromosomal abnormalities, gene alterations and gene variants

Synonym
cystic fibrosis

Research involving
Human

Sponsors and support

Primary sponsor: Universitair Medisch Centrum Utrecht

Source(s) of monetary or material Support: WKZ onderzoeksfonds

Intervention

Keyword: CFTR protein expression, cystic fibrosis, immune cells, nasal epithelial cells

Outcome measures

Primary outcome

CFTR protein expression in nasal epithelial cells and in immune cells

Secondary outcome

Cytokines and chemokines in sputum, blood and bronchoalveolar lavage fluid.

Functional testing of lymphocytes, monocytes and granulocytes.

Clinical parameters including lung function, chest radiograph score and sputum

culture

Study description

Background summary

Cystic fibrosis (CF) is the most common lethal autosomal recessive disorder in the Caucasian population, with an overall birth prevalence in the Netherlands of 1 in 4750 live births from 1974 to 1994. CF patients with a similar CFTR gene mutation display a broad clinical spectrum, and gene modifiers only partly explain these differences. Active transcription of the CFTR gene and CFTR mRNA transcripts are detectable in a variety of epithelial cells (lung, nose, intestine) and also in cells of non-epithelial origin. CFTR expression in human immune cell subsets has been reported for macrophages, neutrophils, and T cells. There are however no studies that compare levels of CFTR protein expression in these different human immune cells, nasal epithelial cells or in human alveolar macrophages or compare these CFTR protein expression levels with clinical and inflammatory parameters.

Study objective

Main objective is to assess CFTR protein expression in nasal epithelial cells and in different peripheral blood immune cell subsets from CF patients with different CFTR gene mutations.

Secondary objective is to correlate these expression levels with clinical parameters (lung function, sputum culture, chest radiograph scoring systems), functional tests measuring CFTR channel function (sweat chloride test, nasal potential differences), inflammation measured in serum and sputum (including different cytokines and chemokines), and functional testing of lymphocytes, monocytes and granulocytes.
Third objective is to assess CFTR protein expression in alveolar macrophages isolated from bronchoalveolar lavage fluid (BALF). CFTR expression levels will be correlated with clinical parameters (lung function, sputum culture, chest radiograph scoring systems), functional tests measuring CFTR channel function (sweat chloride test, nasal potential differences), and inflammation measured in BALF (cytokines and chemokines).

Study design

Cross-sectional observational study

Study burden and risks

Annual blood sampling, sputum sampling, and BALF obtained by bronchoscopy is part of good clinical practice in children with CF. Nasal swabs are conducted by the ear nose and throat specialist on a regular basis and are part of standard care. There are no additional risks in obtaining nasal epithelial cells by nasal swabs.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)
Adults (18-64 years)
Children (2-11 years)
Elderly (65 years and older)

Inclusion criteria

All children with cystic fibrosis currently treated in the UMCU

Exclusion criteria

Acute infectious exacerbation for which treatment with intravenous antibiotics is needed.

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	Active
Primary purpose:	Diagnostic

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	04-04-2011

Enrollment:	234
Type:	Actual

Ethics review

Approved WMO Date:	31-08-2010
Application type:	First submission
Review commission:	METC Universitair Medisch Centrum Utrecht (Utrecht)
Approved WMO Date:	19-06-2012
Application type:	Amendment
Review commission:	METC Universitair Medisch Centrum Utrecht (Utrecht)

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	NL31966.041.10