Optimising screening for early disease detection in familial pulmonary fibrosis

Published: 03-02-2021
Last updated: 06-05-2024

Our primary objective is to determine the prognostic value of parameters for early disease detection of familial pulmonary fibrosis in the study population. All putative parameters for detection of preclinical disease have been shown to be...

Ethical review	Approved WMO
Status	Recruiting
Health condition type	Respiratory disorders congenital
Study type	Observational invasive

Summary

ID

NL-OMON51138

Source
ToetsingOnline

Brief title
Screening for early (Famillial) long (LO) fibRosIS (FLORIS)

Condition

- Respiratory disorders congenital
- Lower respiratory tract disorders (excl obstruction and infection)

Synonym
familial lung fibrosis; familial pulmonary fibrosis

Research involving
Human

Sponsors and support

Primary sponsor: Sint Antonius Ziekenhuis
Source(s) of monetary or material Support: TopZorg van ZonMW (TZO projectnummer 10070012010004),Boehringer Ingelheim

Intervention

Keyword: Biomarkers, Familial pulmonary fibrosis, Pulmonary function test, Screening

Outcome measures

Primary outcome

Presence of Interstitial Lung Disease (ILD) changes on HRCT are indicative of preclinical interstitial lung disease and will be determined at baseline.

Putative parameters of early lung disease that will be investigated include lung function, exercise tests, blood based biomarkers, MUC5B rs35705950 genotype, physical examination for digital clubbing and crackles, and self-reported cough and dyspnoea.

The primary endpoint is the difference in these parameters between the group with ILD changes on HRCT as compared to the group without ILD changes on HRCT.

Secondary outcome

The secondary endpoints are differences in values for parameters between the baseline and follow-up screening after one and two years for the ILD changes and no ILD changes cohort and differences between these groups.

Study description

Background summary

Familial pulmonary fibrosis (FPF) is a fatal lung disease that is often not diagnosed until a significant portion of the lung function is lost. Median survival after diagnosis is 3 to 5 years. As treatment can only slow down lung function decline, early disease detection is essential to provide timely therapeutic support. As first-degree relatives of patients with FPF are at high risk of developing pulmonary fibrosis as well, a screening protocol has been put in place. However, the value of current screening parameters to detect early asymptomatic disease as well as the optimal interval between screening

appointments are unknown. A prospective study into the prognostic value of these screening markers in the target population and the appropriate clinical setting is needed to develop an evidence-based screening protocol. There will be an emphasis on easily operable parameters that may allow for redirection of (part of the) screening activities to the general practice in the future.

Study objective

Our primary objective is to determine the prognostic value of parameters for early disease detection of familial pulmonary fibrosis in the study population. All putative parameters for detection of preclinical disease have been shown to be potentially informative due to their association with disease severity in pulmonary fibrosis and observed presence in patients with preclinical disease. As a secondary objective we aim to gain insight in the natural history of early pulmonary fibrosis, as this is currently incompletely understood. In addition we aim to determine the necessary interval between screening visits.

Study design

The study will be a prospective cohort study whereby subjects will visit the hospital for screening at three time points: at baseline, after 1 year and after 2 years.

Study burden and risks

Participants will visit the St Antonius Hospital 3 times in two years, visits will take approximately 2:20 hour for the baseline visit and 1:50 hour for the two subsequent visits. The health-related burden for participants is minimal and consists of physical examination (including auscultation with electronic stethoscope), three times blood withdrawal, 6-minute walking test and pulmonary function testing and one HRCT scan. Except for the 6-minute walking test and the auscultation with the electronic stethoscope, these procedures would otherwise also be performed as part of the routine lung screening for pulmonary fibrosis in the context of familial disease. Measures are taken to facilitate the practical burden of visiting and costs.

Contacts

Public
Sint Antonius Ziekenhuis

Koekoekslaan 1
Nieuwegein 3435 CM
NL

Scientific
Sint Antonius Ziekenhuis

Koekoekslaan 1
Nieuwegein 3435 CM
NL

Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

Subjects must be a first-degree relative of a patient with familial pulmonary fibrosis
Subjects are reffered by a medical doctor for screening of familial pulmonary fibrosis

Exclusion criteria

- A previous diagnosis of an interstitial lung disease (ILD)
- Pregnancy

Study design

Design

Study type: Observational invasive
Masking: Open (masking not used)

Control:	Uncontrolled
Primary purpose:	Diagnostic

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	16-06-2021
Enrollment:	200
Type:	Actual

Medical products/devices used

Registration:	No

Ethics review

Approved WMO Date:	03-02-2021
Application type:	First submission
Review commission:	MEC-U: Medical Research Ethics Committees United (Nieuwegein)
Approved WMO Date:	29-04-2021
Application type:	Amendment
Review commission:	MEC-U: Medical Research Ethics Committees United (Nieuwegein)
Approved WMO Date:	09-02-2022
Application type:	Amendment
Review commission:	MEC-U: Medical Research Ethics Committees United (Nieuwegein)
Approved WMO Date:	24-04-2024
Application type:	Amendment
Review commission:	MEC-U: Medical Research Ethics Committees United (Nieuwegein)

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
ClinicalTrials.gov	NCT05367349
CCMO	NL75303.100.20