Registry of Sarcoidosis Associated Pulmonary Hypertension (RESAPH)

Published: 02-05-2013
Last updated: 23-04-2024

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Ethical review	Not approved
Status	Will not start
Health condition type	Pulmonary vascular disorders
Study type	Observational non invasive

Summary

ID

NL-OMON38714

Source
ToetsingOnline

Brief title
RESAPH

Condition

- Pulmonary vascular disorders

Synonym
hypertension of the lungvessels, in sarcoidosis patiente, pulmonary hypertension

Research involving
Human

Sponsors and support

Primary sponsor: Erasmus MC, Universitair Medisch Centrum Rotterdam
Source(s) of monetary or material Support: Ministerie van OC&W

Intervention

Keyword: pulmonary hypertension, sarcoidosis

Outcome measures

Primary outcome

Primary Objective

Characterize the demographics, clinical course, hemodynamics, pulmonary physiology, and disease management of sarcoidosis associated pulmonary hypertension on the United States compared to non-US sites.

Endpoints

1. Progression of disease to death or transplant
2. Hospitalization
3. Institution of new therapy for pulmonary hypertension
4. Six minute walk distance
5. Change in Saint George Respiratory Questionnaire of >4 points

Secondary outcome

1. Determine what factors are associated with a bad prognosis in patients diagnosed with SAPH
2. Determine if there is a difference in survival in patients treated with different management strategies
3. Determine the role of immunosuppressive therapy in management of SAPH

Study description

Background summary

Sarcoidosis is a multi systemic disease of unknown etiology with a highly variable clinical course that pathologically is characterized by non-caseating granulomas. Any organ may be involved, but the lungs and draining lymphnodes are most commonly affected. In the Netherlands nearly 5000 people suffer from sarcoidosis. Treatment options are limited and mainly aimed at preventing organ damage. The majority of patients with sarcoidosis do well, but up to 9,7% of the patients will eventually die of sarcoidosis.

The development of pulmonary hypertension is a severe complication of sarcoidosis. The cause of pulmonary hypertension in sarcoidosis is multi-factorial, including diastolic dysfunction, vessel compression by adenopathy, lung destruction by fibrosis, and granulomatous vasculitis. Sarcoidosis Associated Pulmonary Hypertension (SAPH) is an important risk factor for early mortality in sarcoidosis patients. Prospective studies have suggested that five to ten percent of all sarcoidosis patients have pulmonary hypertension.

There is limited information about the management of pulmonary hypertension in sarcoidosis. Most sarcoidosis centers have diagnosed few cases and no standard therapy has evolved. Clinical trial for treatment of SAPH have been limited to small series. Some patients improve when treated with pulmonary vasodilators. Thus the optimal management of these patients remains unclear.

Study objective

We propose to develop a multicenter registry of sarcoidosis associated pulmonary hypertension (SAPH). With this registry, we will characterize the demographics, clinical course, hemodynamics, pulmonary physiology, and disease management of sarcoidosis associated pulmonary hypertension in the United States. We will also compare these features to non-US sites.

Study design

An open label registry consisting of two populations of patients with SAPH. One group will be the patients with known SAPH who are under care at the study center. The second group will be a prospective study of newly diagnosed cases of SAPH. the goal of the registry will be to include approximately 150 patients in both groups for a total of 300 patients. A total of 6 centers from the United States will be used and one of Europe (The Netherlands)

Study burden and risks

Next to the regular outpatient visits and investigations that are part of the regular care for patients with SAPH, the additional burden associated with participation in this study consists of:
- Filling in questionnaires: Patients will be asked to complete 3 questionnaires (St george*s respiratory questionnaire, FAS fatigue score and SF-36 short-form health survey) that should take no more than 20-30 minutes of their total time per visit.
- Next to the regular yearly six minute walk test, one additional six minute walk test is performed per year in the first two years by the patient with walking as far as they can in six minutes.
- During regular phlebotomy, additional blood is drawn

Information about the patient's underlying health will be collected. That information will be stored in a secure database and identifiers will be used (REDCap). There will be no specific information about a participant published or otherwise made available.

The risk of the study is minimal.

There is no direct benefit for the patient in participating in this study. The benefit would be to provide more information regarding this complication of sarcoidosis.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

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

Inclusion criteria

Patients with sarcoidosis as defined by the ATS/WASOG statement
Patients with pulmonary hypertension as confirmed by right heart catheterization
Patients willing to provide written informed consent

Exclusion criteria

Unwillingness to provide assurance that they will complete the follow up visits for the study.

Study design

Design

Study type: Observational non invasive

Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Other

Recruitment

NL

Recruitment status:	Will not start
Enrollment:	10
Type:	Anticipated

Ethics review

Not approved

Date:	02-05-2013
Application type:	First submission
Review commission:	METC Erasmus MC, Universitair Medisch Centrum Rotterdam (Rotterdam)

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	NCT01467791
CCMO	NL43198.078.13