Idiopathic non-cirrhotic intrahepatic portal hypertension (INCIPH): assessment of thrombophilia and histopathology in a multicentre cohort study and HIV patients.

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Ethical review	Approved WMO
Status	Recruiting
Health condition type	Coagulopathies and bleeding diatheses (excl thrombocytopenic)
Study type	Observational invasive

Summary

ID

NL-OMON35427

Source
ToetsingOnline

Brief title
Idiopathic non-cirrhotic intrahepatic portal hypertension (INCIPH)

Condition

- Coagulopathies and bleeding diatheses (excl thrombocytopenic)
- Hepatic and hepatobiliary disorders

Synonym
hepatoportal sclerosis

Research involving
Human

Sponsors and support

Primary sponsor: Erasmus MC, Universitair Medisch Centrum Rotterdam
Source(s) of monetary or material Support: financiering via fondsen van de stichting lever onderzoek.

Intervention

Keyword: HIV infection, idiopathic, portal hypertension, thrombophilia

Outcome measures

Primary outcome

Prevalence of thrombophilic factors.

Clinical evolution.

Pathologic features

Fibroscan and ultrasound elements.

Presence of INCIPH in HIV patients

Secondary outcome

Not applicable

Study description

Background summary

Non-cirrhotic intrahepatic portal hypertension (NCIPH) is characterised by portal hypertension with patent portal vein, in the absence of cirrhosis. In the western countries the disorder is rare and its cause remains unknown. NCIPH is therefore referred to as idiopathic non-cirrhotic intrahepatic portal hypertension (INCIPH). Due to the rarity of the disease in Europe, solid scientific knowledge on various aspects of NCIPH in this region is limited. In 1994, a Belgian cohort of 42 patients with incomplete septal cirrhosis (form of INCIPH) was reported (including patients seen between 1968-1987). Since this publication only one European cohort study (28 patients) has been performed assessing manifestations, clinical course and causes, demonstrating a prevalence of 50% of prothrombotic disorders in the studied cohort. This study supports the theory of obstructive portal vasculopathy, posing injury

(thrombosis) to portal venules initiating alterations in liver architecture and deposition of fibrin [6]. Nevertheless this thrombosis theory remains a controversial issue.

Study objective

Objevtive of the study is to analyze a European cohort of patients with INCIPH based on scientific analyses of clinical, laboratory and morphological data prospectively collected. HIV patients with known splenomagaly will be invited for abdominal ultrasound in order to diagnose portal hypertension. All patients will be screened for thrombophilia and thrombin production (by use of the thrombin generation test). Additionally fibroscan and abdominal ultrasound will be performed. Histology available from previous performed liver biopsy will be reassessed by 2 pathologists. Additional immunohistochemical stainings will be performed to assess stellate cell activation.

Study design

Retrospective observational European multicentre study combined with assessment of thrombophilia and performance of fibroscan and abdominal ultrasound in all patients. Presence of portal hypertension in HIV patients with splenomegaly.

Study burden and risks

Patients participating the study (INCIPH patients and HIV patients with splenomegaly) are invited for an interview at the hospital. Additionally an abdominal ultrasound, fibroscan and blood examination will be performed (time: 1 hour). There are no risks associated with participation.

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

1) Evidence of portal hypertension (oesophageal varices, hypersplenism or ascites)
2) Doppler ultrasound showing patent portal and hepatic veins
3) Liver biopsy showing no cirrhosis
4) Exclusion of conditions causing cirrhosis according to conventional diagnostic criteria (chronic viral hepatitis, alcoholic liver disease, NASH, obesity, hemochromatosis, autoimmune hepatitis or Wilson*s disease)
5) Exclusion of chronic vitam A intake, professional exposure to copper sulphate, vinyl chloride monomer, Spanish toxic oil or arsenic salts
6) Older than 18 years at diagnosis.
7) HIV patients

Exclusion criteria

1) Presence of cirrhosis
2) Presence of portal thrombosis at diagnosis
3) Presence of conditions causing cirrhosis according to conventional diagnostic criteria (chronic viral hepatitis, alcoholic liver disease, NASH, obesity, hemochromatosis, autoimmune hepatitis or Wilson*s disease).
4) Presence of chronic vitam A intake, professional exposure to copper sulphate, vinyl chloride monomer, Spanish toxic oil or arsenic salts
5) Younger than 18 years old at diagnosis.
6) intake oral anticoagulation

Study design

Design

Study type: Observational invasive

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

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	24-11-2009
Enrollment:	50
Type:	Actual

Ethics review

Approved WMO Date:	22-04-2009
Application type:	First submission
Review commission:	METC Erasmus MC, Universitair Medisch Centrum Rotterdam (Rotterdam)

Approved WMO Date:	19-04-2010
Application type:	Amendment
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
CCMO	NL24296.078.08