Radboudumc
university medical center

International registry
Polycystic Liver Disease

An international cohort study on the progression of polycystic liver disease

RESEARCH GROUP

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INTRODUCTION & AIMS

Polycystic liver disease (PLD) is a condition characterized by the manifestation of multiple cysts in the liver. PLD is part of the phenotype of two genetic disorders: 1) autosomal dominant polycystic kidney disease (ADPKD) where liver cysts are present as an extrarenal manifestation in addition to polycystic kidneys and 2) isolated polycystic liver disease (ADPLD) in which liver cysts are the primary phenotype and renal cysts are absent.
Due to growth and proliferation of liver cysts patients can develop severe hepatomegaly leading to symptoms such as abdominal pain, early satiety and dyspnea or complications like cyst infection or rupture.

We have created an international cohort study in order to study the natural course of a large population of PLD patients. Secondary we aim to identify potential antecedents and risk factors for progressive PLD. The main question we want to answer is: how can we predict which patients will become symptomatic?

TIMEFRAME, WHERE ARE WE NOW?

We created an electronic data management registry (Castor EDC). The benefit of a web-based registry is that it allows decentralized data entry and thus facilitates national and international collaboration. To date, approximately 725 Dutch and Belgium patients have been enrolled. Currently, the first data analysis are being performed. Next step is to expand our database by including more patients from national and international centers. Our goal is to achieve the milestone of 1.500 patients at the end of 2016.

If you are interested in a collaborating with us, please don't hesitate to contact me; rene.vanaerts@radboudumc.nl.