UNIKLINIK RWTH AACHEN
Clinic for Gastroenterology,
Metabolic Diseases and
Medical Intensive Care (Med. Klinik III)

UNIKLINIK RWTH AACHEN
Alpha1 Liver Center

Liver involvement in AAT deficiency – an underestimated problem

Alpha1-antitrypsin deficiency (AAT deficiency) is a frequently overlooked metabolic disorder. Apart from lung disease, AAT deficiency leads to liver disease in up to 50% of patients. This can affect people of all ages. Although involvement of the liver is the second most common cause of decreased quality of life and life expectancy in Alpha1 patients, no preventative care plan, like that one implemented to avoid lung involvement, has yet been drawn up.
Chronic liver involvement therefore often remains undetected until a very late stage even in patients with diagnosed AAT deficiency. This alarming shortcoming is facilitated by the fact that the affected patients generally have only unspecific symptoms if any at all. Moreover, routine diagnostic measurements (e.g. liver function tests) often reveal no abnormalities. In the case of a late diagnosis, the diverse complications of liver disease can no longer be effectively prevented.
Liver involvement in AAT deficiency can lead to liver cirrhosis or liver cancer. Liver cirrhosis is the life-threatening consequence of many liver disorders and carries a poor prognosis. Besides AAT deficiency, many other – potentially treatable – conditions, such as viral hepatitis, excessive alcohol consumption and diabetes, can cause liver damage. Liver cirrhosis itself leads to many, often life-threatening secondary disorders such as heavy bleeding or liver cancer. It is therefore crucial that liver disorders such as AAT deficiency are diagnosed at an early stage, so as to prevent complications and to treat concomitant risk factors.

As **the European Reference Network for AAT deficiency-associated liver disease**, we are the only multicentre study group in Europe carrying out research into liver involvement in AAT deficiency, and are collaborating in these studies with various patient support groups and other hospitals specialising in AAT deficiency-associated lung disease. Together with our collaborators, we hope to improve the care of affected patients and help to bring about diagnostic and therapeutic advances.

The aim of our study is to clarify how liver function is modified in patients with AAT deficiency. For this, among other things, we use a modern ultrasound-based scanner (e.g., FibroScan®) for non-invasive measurement of the degree of liver scarring. In addition, we analyse parameters in blood samples, which also provide information on any existing liver disorders, and round up this information with a questionnaire. Study participants receive a full analysis of their individual liver involvement and are given appropriate recommendations for disease prevention. Many of these examinations can only be offered in specialised centres and are not covered by most health insurance providers. Every study participant receives these examinations **without any cost**.
The first data from more than 600 patients show that over 80% of participants did not receive sufficient liver check-ups. This is particularly worrisome given that patients with a PiZZ-genotype not rarely display advanced liver damage. On the other hand, our examinations provide a relief to many and to everybody, we are able to offer an individual liver damage prevention plan.

We are looking forward to include you in our study. Please contact us if you are interested and we will do our best to answer all your questions. We study and advise all adult patients (of every genotype) as well as any family members interested. Besides our specialist clinic in Aachen/Germany (European Reference Centre for AAT deficiency) we offer free examinations throughout Europe.

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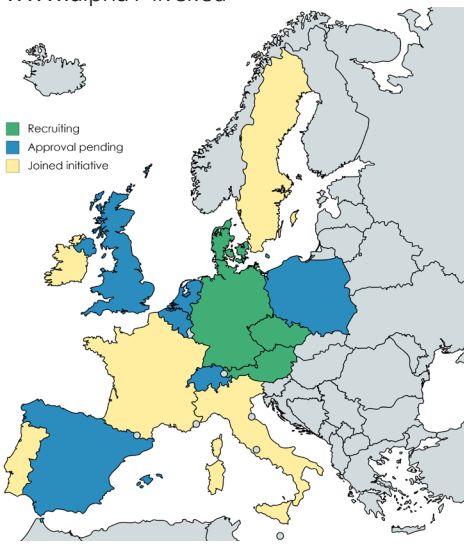

Upcoming regional exams:
Waidhofen (Austria) – 23 to 24 June 2017
Essen (Germany) – 23 September 2017
Cologne (Germany) – tba
Nijmegen (The Netherlands) – tba
Warsaw (Poland) – tba
Bern (Switzerland) – tba

Further Links:
www.alpha1.org
http://bit.ly/a1l-film

European Reference Networks
EASL European Association for the Study of the Liver
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