



Patiëntenvereniging
ACUTE PORFYRIE

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Information meeting Acute Porphyria July 5th 2022

At last, after a two year delay due to the COVID pandemic, the first Information meeting on acute porphyria has taken place on July 5th 2022. This was organized by the Dutch Patient Association Acute Porphyria (PVAP) in cooperation with Isala Hospital Zwolle and the Porphyria Expertisecenter Rotterdam, Erasmus Medical Center.

Acute Porphyria is a very rare inborn metabolic disease. The symptoms are often not recognized or misdiagnosed, which can lead to life threatening situations or irreversible neurological damage. Therefore it is essential that awareness is raised and information about acute porphyria is spread to all patients and doctors. The number of patients in the Netherlands is estimated at about 750. Most of these carriers of the enzyme deficiency have no medical complaints, but can get an acute attack by a number of triggers. Ten percent of the carriers get sick, varying from 1 attack in a lifetime to recurrent attacks and chronic complaints.

The meeting was intended for patients and their relatives. Everyone was provided with a PVAP flyer with information about the disease, warnings about triggers and advices what to do when an attack occurs.

The program consisted of three lectures and a panel discussion by five specialists in the field of acute porphyria. The chairwoman of the PVAP opened the meeting with a speech in which she illuminated the origin of this association. She emphasized the importance of founding the first Dutch association for this rare disease, and she spoke about the mission of the PVAP to bring acute porphyria more to the attention of general practitioners, internists and First Aid Departments.

The first speaker informed attendees about the previous generation of Dutch specialists, the pioneers in the field of acute porphyria, who cared about the fate of these patients and invested in proper treatment. The transmission of their knowledge to the next generation of specialists has evolved to the establishment of the Porphyria Expertisecenter Rotterdam, Erasmus MC. Additionally, the pharmaceutical industry invested in developing therapeutical strategies that made treatments possible.



The lecture 'General information about acute porphyria' followed the question "What do patients with acute porphyria perceive to be important characteristics of their attending doctor". To answer this question a survey had been sent to the members of the association to fill in. Among conclusions were the necessity of the doctor to have extended knowledge of the disease, to be empathetic and to be easily accessible, preferably by mobile phone. Also the question was raised and answered why acute porphyria is so hard to recognize for physicians and why it can take five years before a patient gets the correct diagnosis.

The director of the Porphyria Expertisecenter shed some light on both the current standard treatment as well as the newest treatment with the medicine Givosiran (Givlaari). This is a groundbreaking drug for patients with recurring attacks, which are reduced greatly by using a mRNA technique that hinders the accumulation of toxic heme precursors in the body.

During the break patients got to know fellow sufferers and exchanged personal experiences and we were delighted to see speakers and attendees in conversation with each other while enjoying a drink and a snack.

After the break there was a panel discussion with all the speakers. Various aspects and consequences of acute porphyria were explored in depth. A lively exchange of knowledge erupted that also allowed for some humor. It was good to see so many questions being asked by the attendees.

The evening was ended with a call to carriers, patients and their family members to share their personal stories with the PVAP. By sharing their story other patients and medical experts can learn more about acute porphyria. Our first information meeting was rated by the attendees at an average of 8,7!

We thank everyone who contributed to the evening and made it a success! Especially we thank SSIEM for granting us with the Patient Support Award 2022.

List of speakers and scientific panel (alphabetically)

M.A. Alleman, Md Phd, non-practicing internist

J.G. Langendonk, Md Phd, Porphyria Expertisecenter Rotterdam, Netherlands

G.S. Mijnhout, Md Phd, Department of Internal Medicine. Zwolle, Netherlands

C.G. Vermey, Md Phd, Department of Internal Medicine, Deventer, Netherlands

J.H.P. Wilson, Emeritus Professor of Internal Medicine