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Editorial

On the Issue of Acute Hepatic Porphyria

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Porphyria (porphyriae; Greek. porphyra (purple dye) is a group of hereditary diseases based on the insufficiency of any one enzyme for the synthesis of the organic part of heme-protoporphyrin [1]. Porphyria are not endemic diseases and occur with the same frequency among the population of all continents [2]. The most frequent of acute porphyria is acute intermittent porphyria (AIP), which accounts for more than 85% of all cases of acute porphyria. The frequency of AIP was the same in all countries (0.13 per million per year; 95% CI: 0.10 - 0.14), with the exception of Sweden (0.51; 95% CI: 0.28 - 0.86) [3]. The first attack of acute porphyria can develop at the age of 14 - 16 years [4].

All acute porphyria, except for the sporadic form of late cutaneous porphyria, are hereditary. Therefore, their complete cure is impossible. However, competent prevention of AIP attacks makes it possible to achieve a long-term latent course of the disease [5].

The purpose of treatment of AIP is to prevent attacks of the disease and the development of irreversible changes in the nervous system. Prevention of AIP attacks involves limiting the effects of provoking factors on the body, namely: alcohol intake, medications with increased porphyrinogenic activity, insolation, the development of bacterial and viral HCV, HBV, CMV infections, hypoglycemia, in women with a proven link between the onset of menses and frequent seizures - prevention of menstruation [6].

The decision on the selection of drug therapy for AIP is relevant not only during the treatment of an acute attack of the disease, the main purpose of which is to prevent repeated attacks against the background of treatment, but also at the outpatient stage [7]. Patients who suffer from recurrent seizures report a low quality of life and a negative impact on some aspects of everyday life, including unemployment, personal relationships and long-term disability [8].

Thus, AIP are severe diseases that cause suffering to patients and have high mortality and lethality. Many aspects of this disease require resolution. There is insufficient data on the epidemiology of AIP in some regions of the world. Mutations and other causes leading to this disease, complicating its course, have not been studied in many regions. Due to the rarity of AIP, the features of its clinic are insufficiently represented, there are no clear criteria for the precursor of an attack.

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