

Triggers for Acute attacks in Acute Hepatic Porphyria

Author: Christopher V. Almario, MD, MSHPM; Cedars-Sinai Medical Center, Karsh Division of Gastroenterology and Hepatology

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What is Acute Hepatic Porphyria?

Acute hepatic porphyria (AHP) is a group of rare genetic conditions that can cause severe gastrointestinal (GI), neurologic, psychiatric, cardiovascular, and skin symptoms.

There are four types of AHP which include:

- acute intermittent porphyria,
- variegate porphyria,
- · hereditary coproporphyria, and
- ALAD-deficient porphyria.

It is estimated that 5 in every 100,000 persons have AHP. Most people with AHP start to develop symptoms when

they are adults. While AHP can affect anyone, it is more common in women and Caucasians.

People with AHP have a defective gene that leads to problems making heme, which is an important part of hemoglobin. Hemoglobin is a protein in red blood cells that carries oxygen throughout the body. This defect leads to the buildup of toxic chemicals in the body that can damage nerve cells and cause severe symptoms. Sometimes the AHP attacks can be life-threatening. Below are the most common symptoms of AHP, many other symptoms may occur.

Triggers for Acute Attacks in Acute Hepatic Porphyria

Patients with acute hepatic porphyria (AHP) can have sudden (acute) symptoms that are severe and require them to go to the hospital. At times, these acute attacks can be triggered by stress, medications (drugs), diet, and environmental factors, among others. Avoiding or reducing exposure to known triggers is an important way to prevent acute AHP attacks. However, some people experiencing an attack may not know what caused the attack to happen.

Drugs can be triggers for some patients with AHP. Some examples of drugs that should be avoided when possible include carbamazepine, danazol, diclofenac, estrogens, metoclopramide, phenytoin, progesterone, rifampin, sulfonamide antibiotics, and valproic acid, among many others. A full list of safe and unsafe medicines for AHP can be found at the American Porphyria Foundation Drug Database

(https://porphyriafoundation.org/drugdatabase/).

Crash dieting can also trigger acute attacks and should be avoided in those with AHP. Patients with AHP can also consider working with a dietician for help

IFFGD's **Dietitian Listing** is a resource that allows you to search for a dietitian that is in your area or treats a specific condition.

identifying foods that may be causing attacks. Alcohol, tobacco, marijuana, and recreational drugs also may lead to sudden symptoms in AHP and should be avoided.

Any infection in the body, including common ones like urinary tract infections and pneumonias, can trigger acute attacks. When a person with AHP is experiencing an acute attack and is found to have an infection, both the AHP and infection should be treated at the same time. Also, those with AHP should receive all appropriate vaccinations that prevent infections, such as the flu, pneumonia, and recombinant shingles vaccines.



Understanding Acute Hepatic Porphyria

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Any stress to the body such as surgery or physical/emotional exhaustion can also trigger acute AHP attacks. For women with AHP, some may experience frequent attacks during their menstrual cycle. As the menstrual cycle can be a trigger, some types of contraceptives (birth control) can help in preventing attacks. Common types include low-dose estradiols or low-dose estrogen-progestins which are available under many brand and generic names.

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IFFGD 537 Long Point Road, Suite 101 Mt Pleasant, SC 29464

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