

Could It Be Acute Intermittent Porphyria?

A checklist for you and your doctor

Acute intermittent porphyria (AIP) is a rare inherited disorder caused by a partial lack of enzyme activity needed to make heme, a substance that carries oxygen to all parts of your body. This enzyme deficiency results in the build-up of certain chemicals in your body, causing symptoms to develop. If not treated, AIP symptoms, also called AIP “attacks,” can cause damage to your body such as to your brain and nervous system.

Recognizing AIP can be hard for two reasons. One, symptoms of AIP are similar to those of many conditions. And two, AIP is rare. So a more common diagnosis may be given.

Signs and Symptoms Commonly Associated with an AIP Attack

During AIP attacks, people often have some of the signs and symptoms listed below. **Check and write in any symptoms you are having.**

- Severe abdominal pain
- Nausea or vomiting
- Constipation
- Dark or reddish urine
- Muscle weakness
- Pain in extremities, back, chest, neck, or head
- Fast heartbeat
- High blood pressure
- Mental symptoms, such as minor behavioral changes, anxiety, confusion, or depression
- Other: _____

Symptoms and family history

Symptoms of AIP can occur throughout life. AIP is inherited, so you are more likely to develop it if you have a relative with AIP. **Answer the questions below about your symptoms and family history.**

1. How often do you have symptoms?

2. When was the first time you had symptoms?

3. About how long do your symptoms last?

4. Do you have any relatives with AIP or relatives who have symptoms listed above?

Things that can trigger AIP attacks

AIP attacks are “acute” and “intermittent,” meaning they tend to come and go. Certain things may trigger attacks. **Check and write in the things that seem to trigger your attacks.**

- The week or two before your periods start
- Dieting
- Smoking cigarettes
- Drinking alcohol
- Taking certain medicines: _____
- _____
- Feeling stressed
- Having an illness or infection
- Other: _____

Diagnosing AIP

Diagnosis of AIP may involve:

- Laboratory tests of chemicals in your urine
- A genetic DNA test

Working with your doctor

If you are diagnosed with AIP, you should work closely with your doctor to manage your disorder. There is no cure for AIP, but treatment is available.

IMPORTANT SAFETY INFORMATION

PANHEMATIN is contraindicated in patients with a known hypersensitivity to this drug.

Risk of Phlebitis: Phlebitis is possible. Utilize a large arm vein or a central venous catheter for administration to minimize the risk of phlebitis.

Iron and Serum Ferritin: Elevated iron and serum ferritin may occur. Monitor iron and serum ferritin in patients receiving multiple administrations of PANHEMATIN.

Anticoagulant Effects: PANHEMATIN has transient and mild anticoagulant effect. Avoid concurrent anticoagulant therapy.

Renal Effects: Reversible renal shutdown has been observed with an excessive hematin dose (12.2 mg/kg in a single infusion). Strictly follow recommended dosage guidelines.

Transmissible Infectious Agents: PANHEMATIN may carry a risk of transmitting infectious agents, e.g., viruses, and theoretically, the Creutzfeldt-Jakob disease (CJD) agent. There is also the possibility that unknown infectious agents may be present in the product.

The most common adverse reactions (>1% of patients) are headache, pyrexia, infusion site reactions, and phlebitis.

To report SUSPECTED ADVERSE REACTIONS, contact Recordati Rare Diseases Inc. at 1-888-575-8344, or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Drug Interactions: Avoid CYP inducing drugs such as estrogens, barbituric acid derivatives and steroid metabolites which induce δ -aminolevulinic acid synthetase 1 (ALAS1) through a feedback mechanism.

PANHEMATIN® (hemin for injection), for intravenous infusion only, is available as powder for reconstitution in 350 mg vials.

INDICATIONS AND USAGE

PANHEMATIN is a hemin for injection indicated for the amelioration of recurrent attacks of acute intermittent porphyria temporally related to the menstrual cycle in susceptible women, after initial carbohydrate therapy is known or suspected to be inadequate.

Limitations of Use

- Before administering PANHEMATIN, consider an appropriate period of carbohydrate loading (i.e., 400 g glucose/day for 1 to 2 days).
- Attacks of porphyria may progress to a point where irreversible neuronal damage has occurred. PANHEMATIN therapy is intended to prevent an attack from reaching the critical stage of neuronal degeneration. PANHEMATIN is not effective in repairing neuronal damage.

Please see the accompanying full Prescribing Information at www.PANHEMATIN.com.

