Discussing Acute Intermittent Porphyria with Your Family

Discussing your acute intermittent porphyria (AIP) with family members can be very helpful—for you and for them. When your family understands your disorder and how it affects you, they can help provide support. The questions and answers in this discussion guide can be used to encourage everyone to talk about AIP.

What is AIP?

AIP is a rare inherited disorder caused by a partial lack of an enzyme 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. "Acute intermittent" means that symptoms, or "attacks," may occur for a set period of time, then go away, only to return later. Left untreated, AIP attacks can cause damage to your body such as to your brain and nervous system.

What are symptoms of AIP?

The most common symptom of AIP is severe abdominal pain. Other common symptoms include vomiting, constipation, a fast heart rate, pain in different areas of your body, muscle weakness, and mental symptoms like depression or changes in behavior. During AIP attacks, you may not have all of these symptoms. Talk to your family about your specific symptoms and how they make you feel.

When are AIP attacks more likely to occur?

AIP attacks are more likely to occur when you're exposed to things that upset your body's chemical balance. These "triggers" may include use of hormones or hormone fluctuations, use of certain prescription or illegal drugs, use of alcohol or cigarettes, fasting or crash dieting, infections, surgery, or stress. Talk to your family about your triggers and ways to help manage them.

How is AIP diagnosed?

Because the symptoms of AIP are common in other conditions and doctors are not always familiar with this rare disease, a diagnosis of AIP may be difficult to obtain. But an accurate diagnosis is critical for getting the right treatment. Diagnosis may involve laboratory tests that analyze chemicals in your urine and a genetic DNA test.

Are family members at risk for developing AIP?

AIP is inherited, so family members of someone who has AIP have a greater risk of developing the disorder. Most people with the enzyme deficiency never have symptoms, and some people may have only mild symptoms throughout life. But it's important to remember that symptoms can develop and become serious very quickly. Therefore, family members may want to talk to their doctors about genetic testing for AIP.

Is there treatment for AIP?

Although there is no cure for AIP, treatment is available. PANHEMATIN® (hemin for injection) is a prescription medication used to relieve repeated attacks of AIP related to the menstrual cycle in affected women, after initial carbohydrate therapy is known or suspected to be inadequate. The goal of PANHEMATIN is to reduce the chemical build-up that causes symptoms.

What are some good sources of information about AIP?

For more information about AIP, go to aiporphyria.com and click on the "Patient Support" page. There you'll find links to online resources that educate and support people with AIP and their families.

Indications and Usage

PANHEMATIN is a hemin for injection prescription medication used to relieve repeated attacks of acute intermittent porphyria related to the menstrual cycle in affected women, after initial carbohydrate therapy is known or suspected to be inadequate.

Limitations of Use

- Before giving PANHEMATIN, consider an appropriate trial of carbohydrates [i.e., 400 grams of glucose (a carbohydrate or sugar) per day for 1 to 2 days].
- Attacks of porphyria may progress to a point where irreversible nerve damage has occurred. PANHEMATIN therapy is intended to prevent an attack from reaching the critical stage of nerve breakdown. PANHEMATIN is not effective in repairing nerve damage that has already occurred.

Important Safety Information

- PANHEMATIN is not for patients known to be allergic to this drug.
- Vein inflammation is possible. Use a large arm vein or a central line to administer PANHEMATIN to minimize the possibility of vein inflammation.
- Elevated iron levels may occur. Your doctor must monitor your iron levels if you receive multiple courses of PANHEMATIN.
- PANHEMATIN has a passing and mild blood thinning effect. Avoid blood thinners during PANHEMATIN therapy.
- Reversible kidney shutdown has occurred when too high a dose of PANHEMATIN was given in a single dose.
- PANHEMATIN may carry a risk of transmitting agents that can cause infections, such as viruses, and theoretically, the Creutzfeldt-Jakob disease (CJD) agent.
- Most common side effects are headache, fever, infusion site reactions, and vein inflammation.
- **To report SUSPECTED SIDE EFFECTS, contact Recordati Rare Diseases Inc. at 1-888-575-8344, or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.**
- When taking PANHEMATIN, do not take drugs such as estrogens (e.g., oral contraceptives), barbiturates (drugs that help with sleep and used to treat epilepsy) or steroids (body hormone-like drugs), because such drugs can trigger an attack or make an attack worse.

For more information, please see Full Prescribing Information at www.Panhematin.com.

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