



## What is Acute Intermittent Porphyria (AIP)?

AIP is one of a group of rare, inherited disorders called porphyrias that all involve the overproduction and buildup of chemicals called porphyrins or porphyrin precursors. AIP is caused by a partial deficiency of a specific enzyme (porphobilinogen deaminase) and is characterized by acute attacks, often triggered by environmental factors or hormone changes. In some cases, the cause of attacks can't be identified.

## Living with AIP

Because the symptoms of AIP mimic other common conditions, the diagnostic process can be long and difficult with some patients suffering for years before receiving a proper diagnosis.

This educational material is provided by the American Porphyria Foundation. For more information, visit [porphyriafoundation.com](http://porphyriafoundation.com)

## References

1. Anderson KE, Bloomer JR, Bonkovsky HL, et al. Recommendations for the diagnosis and treatment of the acute porphyrias. *Ann Intern Med.* 2005;142:439-450.
2. Bonkovsky HL, et al. Acute porphyrias in the USA: features of 108 subjects from porphyrias consortium. *The Amer. Jnl of Med.* 2014;127: 1233-1241.
3. Crimlisk HL. The little imitator-porphyria: a neuropsychiatric disorder. *J Neurol Neurosurg Psychiatry.* 1997;62:319-328.

## Effective Management of Acute Intermittent Porphyria:

# Patients Deserve Access to Appropriate Care

### Patients Diagnosed with AIP Deserve Access to Treatment

For people living with Acute Intermittent Porphyria (AIP), there are treatments available, but many still encounter barriers to receiving accessible care at their preferred hospitals.

The American Porphyria Foundation (APF) urges all healthcare institutions to provide timely access to medically necessary care. We encourage everyone involved in the care of patients with AIP to demand timely access to FDA-approved treatments. We believe:

- Regulators** should **advocate for patient access** to approved treatments
- Providers** need to understand AIP and **provide prompt, appropriate treatment**
- Patients** **deserve access and coverage** for available, life-saving treatment options

Signs and Symptoms			
Gastrointestinal	Neurologic	Cardiovascular	Urinary
<ul style="list-style-type: none"> <li>• Severe, intense abdominal pain</li> <li>• Nausea and vomiting</li> <li>• Constipation</li> <li>• Diarrhea</li> </ul>	<ul style="list-style-type: none"> <li>• Pain in the extremities, back, chest, neck, or head</li> <li>• Muscle weakness</li> <li>• Convulsions</li> <li>• Mental symptoms</li> <li>• Respiratory paralysis</li> </ul>	<ul style="list-style-type: none"> <li>• Fast heart beat</li> <li>• High blood pressure</li> </ul>	<ul style="list-style-type: none"> <li>• Dark reddish or purple urine</li> </ul>

## Managing AIP

AIP is a genetic disease and there is no cure. However, there are strategies that can be used to prevent the onset or severity of attacks:

- Eating a balanced diet, including sufficient carbohydrates
- Avoiding dieting or fasting, even for short periods
- Avoiding certain medications that can trigger an attack
- Limiting physical and emotional stress
- Avoiding alcohol

People with frequent attacks can also consider wearing a medical alert bracelet.

## Prompt Treatment is Critical

Even with proper diet and control of environmental factors, some patients still have recurring, and often severe attacks. Acute attacks typically require hospitalization and delays in treatment can be life-threatening and result in serious complications such as irreversible nerve damage.

## Treatments can include:

- Medication to manage pain and other symptoms
- Glucose and carbohydrates given orally or intravenously
- Intravenous heme if the patient's symptoms fail to improve within 36 hours of glucose treatment