The Impact of Acute Hepatic Porphyria (AHP)

The debilitating burden of this disease can impact your patients' quality of life today, and long term.

What is AHP?

AHP is a rare genetic condition characterized by acute, potentially life-threatening attacks, as well as chronic ongoing symptoms. AHP patients often face a higher risk of long-term complications such as hypertension, chronic kidney disease, hepatocellular carcinoma, depression, and anxiety.[1,2]

These studies explored the impact of AHP

● = POWER study ■ = EXPLORE study

● **POWER*** was a study where **92 adults** with AHP who experienced 1 or more attacks in the last 2 years or were receiving hemin and/or glucose (prophylactically or on demand) participated in a survey to determine the impact of AHP on their well-being and emotional, mental, and physical needs.[3]

   ***POWER**=**Po**rphyria **W**orldwide Patient **E**xperience **R**esearch.

■ **EXPLORE** was a 2-part prospective natural history study of **112 patients** (Part A) and **136 patients** (Part B) with AHP to gather data on impact of AHP on symptoms, treatment, quality of life, and health status. Patients in Part A included those who experienced recurrent attacks (≥3 over the last year) or received prophylactic treatment. Including eligible patients from Part A, Part B also enrolled patients with relatively few attacks (<3 attacks without prophylactic treatment).[2]

Patients' quality of life can worsen due to chronic symptoms, regardless of attack recurrence[†4]

● Patients reported acute symptoms such as **pain, muscle weakness, and fatigue**[‡3]

● Pain limited the daily activities of nearly **95% of patients**[3]

■ A majority of patients with recurrent attacks reported **chronic symptoms** between attacks in the past 12 months[2]

■ **Elevated levels of ALA and PBG** (which are associated with long-term complications) were observed in patients **outside of attacks** and increased further during attacks[§1,2,5,6]

ALA, aminolevulinic acid; PBG, porphobilinogen; QOL, quality of life; ULN, upper limit of normal.

[†]Quality of life was measured by the EuroQol visual analog scale (EQ-VAS), which ranges from 0 (worst imaginable health state) to 100 (best imaginable health state). At baseline, scores were 62.9 and 71.8 in patients with ≥3 attacks or prophylaxis and in patients with <3 attacks and no prophylaxis, respectively.[4]

[‡]The most burdensome acute symptoms reported were acute pain (72%), acute muscle weakness (37%), and acute fatigue (28%).[3]

[§]Elevated ALA and PBG levels were above the ULN of 1.5 mmol/mol for urinary ALA and 0.14 mmol/mol for urinary PBG.[2]

Alnylam®
PHARMACEUTICALS

The emotional toll of AHP is immense[4]

- In patients who experienced 0-5 attacks in the past 2 years, **53%** and **44%** reported having moderate-to-severe depression and anxiety, respectively*[3]

  *Moderate-to-severe depression and anxiety scores were determined by PHQ-8 and GAD-7 scores ≥10, respectively. GAD-7 scores range from 0-21 and PHQ-8 scores range from 0-24, with higher scores indicating increases in severity.[3]

- **51%** of patients with recurrent attacks reported problems affecting their quality of life due to anxiety and depression†[2]

  †Anxiety/depression was measured by the EQ-5D-5L Index, a 5-dimension visual analog scale that goes up to 100, which signifies the maximal score and best overall health.[2]

From work to home, AHP doesn't let up

60% of patients reported often feeling lonely or isolated[3]
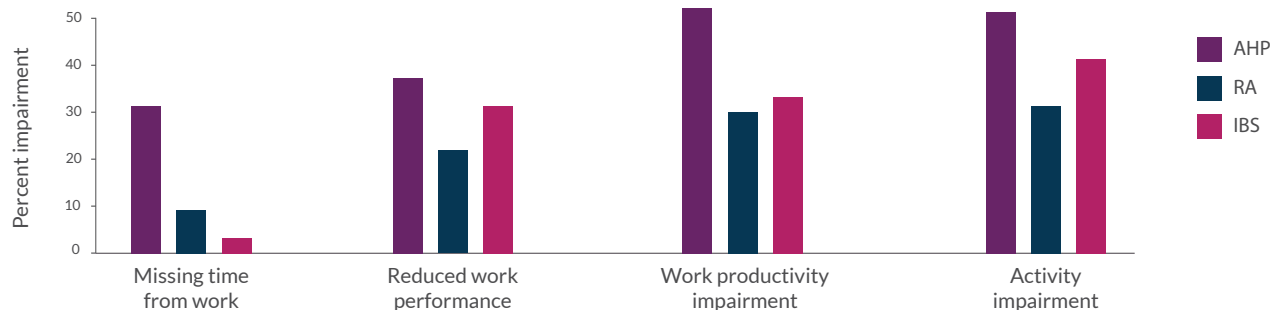
73% of patients agree that AHP has kept them from working at their full potential‡[3]

61% reported the disease affects their ability to keep a job at all‡[3]

Patients with AHP worry about how their illness affects the people closest to them

Approximately 3 out of 4 patients with AHP reported increased guilt and frustration about how their disease impacts the people around them.[3]

Patients with AHP report work productivity and activity impairment comparable to those with rheumatoid arthritis (RA) and irritable bowel syndrome (IBS)‡§[7-10]

Bar chart — Percent impairment (y-axis 0-50) across four categories for AHP, RA, and IBS:
- Missing time from work: AHP ~31, RA ~9, IBS ~3
- Reduced work performance: AHP ~37, RA ~22, IBS ~31
- Work productivity impairment: AHP ~52, RA ~30, IBS ~33
- Activity impairment: AHP ~51, RA ~31, IBS ~41

‡Data were based on the Work Productivity and Activity Impairment (WPAI) instrument, which was used to evaluate absenteeism, presenteeism, overall work productivity, and activity impairment.[7]

§The data are sourced from different studies (150 patients with RA from a 2010 study, 135 patients with IBS from a 2004 study).[9,10]

Learn more about AHP, and find resources for your patients

Learn about disease pathophysiology, symptoms, testing, and treatment information for AHP at **porphyriadiagnosis.com.**

References: 1. Wang B, Bonkovsky HL, Lim JK, Balwani M. *Gastroenterology.* 2023;164(3):484-491. **2.** Gouya L, Ventura P, Balwani M, et al. *Hepatology.* 2020;71(5):1546-1558. **3.** Dickey A, Wheeden K, Lyon D, et al. [Published correction appears in *JIMD Rep.* 2023;64(4):300]. *JIMD Rep.* 2023;64(1):104-113. **4.** Cassiman D, Kauppinen R, Monroy S, et al. *J Inherit Metab Dis.* 2022;45(6):1163-1174. **5.** Pallet N, Karras A, Thervet E, Gouya L, Karim Z, Puy H. *Clin Kidney J.* 2018;11(2):191-197. **6.** Pallet N, Mami I, Schmitt C, et al. *Kidney Int.* 2015;88(2):386-395. **7.** Dickey A, Wheeden K, Lyon D, et al. [Published correction appears in *JIMD Rep.* 2023;64(4):300]. Appendix S1. Supporting information. *JIMD Rep.* 2023;64(1):104-113. **8.** Dickey A, Meninger S, Mora J, et al. Poster presented at American College of Gastroenterology Annual Scientific Meeting; October 22-27, 2021; Las Vegas, NV. **9.** Zhang W, Bansback N, Boonen A, Young A, Singh A, Anis AH. *Arthritis Res Ther.* 2010;12(5):R177. **10.** Reilly MC, Bracco A, Ricci J-F, Santoro J, Stevens T. *Aliment Pharmacol Ther.* 2004;20(4):459-467.