



Identifying and Diagnosing Acute Hepatic Porphryia

Full abbreviations, accreditation, and disclosure information available at [PeerView.com/DQY40](https://www.peerview.com/DQY40)

Clinical Criteria to Establish an AHP Diagnosis¹⁻⁴

Initial Assessment

- Personal and family history
- Physical examination
- Neurologic assessment (baseline)
- Baseline assessment (blood count, ferritin, hyponatremia, renal function, hepatic function)

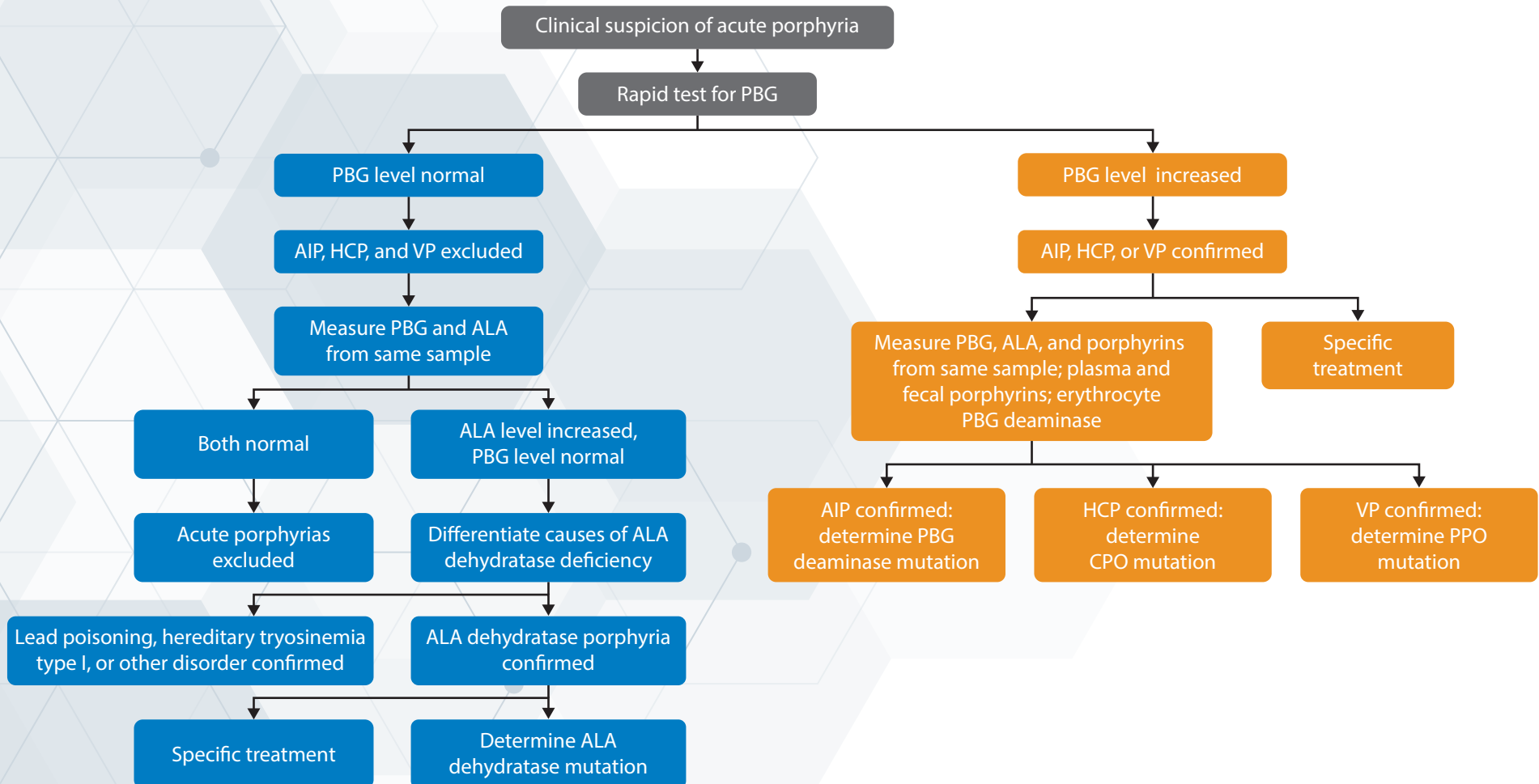
Biochemical Testing

- First line: Urinary PBG, ALA, porphyrins, creatinine
 - Quantitative spot urine sample normalized to creatinine
 - Normal ALA: 1.47 mmol/mol Cr (9.3- to 12-fold higher in AHP)
 - Normal PBG: 0.137 mmol/mol Cr (238- to 336-fold higher in AHP)
 - Should be performed when patients are sick, since levels may be normal between acute episodes
- Second line: More thorough porphyrin assessment

Genetic Testing

- After biochemical testing (not recommended for first-line testing)
- *ALAD*, *HMBS*, *CPOX*, and *PPOX* genes
- Helps confirm or exclude porphyria diagnosis (~95% sensitivity and 99% specificity)
- To determine specific AHP
- To help identify at-risk family members

Algorithm for Diagnosing an Acute Porphryia Attack⁵



Results are most accurate when patients are symptomatic or have had recent symptoms (within last 10 days)



Preventive Measures for Acute Hepatic Porphyrria Attacks

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Precipitating and Aggravating Factors That Can Trigger an Increase in ALA and PBG¹⁻⁴

- Luteal phase of menstrual cycle
- Pregnancy/postpartum
- Stress/exhaustion
- Fasting/dietary restriction
- Infection
- Smoking
- Alcohol —binge drinking
- Surgery/anesthesia

Implicated Medications

- Anticonvulsants (ie, phenytoin, barbiturates, valproic acid, carbamazepine, primidone)
- Hormones (eg, estrogen, progestogens)
- Some antibiotics/antifungals (eg, trimethoprim, erythromycin, fluconazole, nitrofurantoin, rifampicin)
- Inducers of cytochromes P-450



Harmful and Safe Drugs for Patients With AHP⁵

Harmful Drugs

Barbiturates
Trimethoprim
Many tranquilizers and sedatives
(eg, meprobamate, methyprylon, glutethimide)
Griseofulvin
Some antiepilepsy drugs (phenytoin, etc)
Birth control pills
Alcohol
Ergots
Metoclopramide
Rifampin
Diclofenac
Danazol

Safe Drugs

Narcotic analgesics
(morphine, meperidine, codeine, etc)
Aspirin and acetaminophen
Phenothiazines
(eg, chlorpromazine)
Penicillin and derivatives
Chloral hydrate
Streptomycin
Glucocorticoids
Bromides
Insulin
Atropine
Cimetidine
Serotonin reuptake inhibitors (antidepressants)

All drug lists are mainly based on expert opinion, not evidence-based,
so there is an ongoing need for the testing of drugs and chemicals