# Acute Hepatic Porphyria (AHP) Random Urine Testing Reference Guide

Random (spot) urine tests can help inform a diagnosis of AHP. This guide provides testing information and protocols from national laboratories in the US for porphobilinogen (PBG), aminolevulinic acid (ALA), and total porphyrins when considering an AHP diagnosis.

## This testing and protocol information is provided for educational purposes only and is not intended to replace the independent medical judgment of any healthcare professional.

<table>
<thead>
<tr>
<th>Lab/ Test Code</th>
<th>CPT Code</th>
<th>Test Name</th>
<th>Synonyms/Related Terms</th>
<th>Specimen Requirements and Storage/Transport Instructions</th>
<th>Specimen Container and Collection</th>
</tr>
</thead>
<tbody>
<tr>
<td>ARUP-PBG/ 2011476</td>
<td>84110</td>
<td>Porphobilogen (PBG), Urine</td>
<td>PBG; Quantitative Porphobilogen; Watson-Schwartz Test</td>
<td>Urine (random), frozen and protected from light</td>
<td></td>
</tr>
<tr>
<td>ARUP-ALA/ 2011474</td>
<td>82135</td>
<td>Aminolevulinic Acid (ALA), Urine</td>
<td>ALA; 5-Aminolevulinic Acid; Delta Aminolevulinic Acid; Delta ALA</td>
<td>Urine (random), refrigerated or frozen</td>
<td></td>
</tr>
<tr>
<td>ARUP-Porphyrins/ 2002181</td>
<td>84120</td>
<td>Total Porphyrins, Urine</td>
<td>Porphyrins, Quantitative Urine; Coproporphyrin; Heptacarboxyl Porphyrin; Hexacarboxyl Porphyrin; Pentacarboxyl Porphyrin; Uroporphyrin</td>
<td>Urine (random), frozen and protected from light; Urine (24 hour collection), frozen and protected from light</td>
<td></td>
</tr>
</tbody>
</table>

The porphyrins test includes both porphyrins and PBG, and is associated with CPT codes 84120 (porphyrins) and 84110 (PBG).

For additional educational resources, visit [https://arupconsult.com/content/porphyrias](https://arupconsult.com/content/porphyrias).

<table>
<thead>
<tr>
<th>LabCorp-PBG/ 003053</th>
<th>84110</th>
<th>Porphobilogen (PBG), Quantitative, Random Urine</th>
<th>PBG Random Urine, Quantitative</th>
<th>Causes for rejection: specimen exposed to light, specimen not received frozen</th>
<th>Transfer a 3-mL specimen to a plastic urine container with 0.5 mL of 30% glacial acetic acid (optional preservative, but required if ordered with Δ-ALA) and amber plastic frozen transport tube and cap (LabCorp No. 78656)</th>
</tr>
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<tbody>
<tr>
<td>LabCorp-ALA/ 007351</td>
<td>82135</td>
<td>Δ-Aminolevulinic Acid, Random Urine</td>
<td>ALA, Delta, Random Urine; Delta-Aminolevulinic Acid, Random Urine</td>
<td>Causes for rejection: specimen not protected from light, use of preservative other than 30% acetic acid, use of sodium carbonate preservative</td>
<td>Transfer a 3-mL specimen to a plastic urine container with 0.5 mL of 30% acetic acid and amber plastic frozen transport tube and cap (LabCorp No. 78656)</td>
</tr>
<tr>
<td>LabCorp-Porphyrins/ 120980</td>
<td>84120</td>
<td>Total Porphyrins, Quantitative, Random Urine</td>
<td>Coproporphyrin; Uroporphyrin</td>
<td>Causes for rejection: stored specimen not refrigerated, specimen exposed to light, acid preservative, pH &lt;3</td>
<td>Transfer a 2-mL specimen to a 12-mL amber plastic transport tube without preservative (LabCorp No. 23598)</td>
</tr>
</tbody>
</table>

For the most up-to-date testing and protocol information from LabCorp, please visit [www.labcorp.com](http://www.labcorp.com).

Links to the sites for these independent laboratories are provided as a reference. Alnylam Pharmaceuticals does not endorse and is not responsible for the content on sites that are not owned and operated by Alnylam Pharmaceuticals.


*Mayo Clinic Laboratories suggest that for total porphyrins testing, random tests should be ordered when specimens will reach the lab within 72 hours. If transportation will take longer than 72 hours, a 24-hour urine collection test should be ordered.

To ensure greater accuracy, experts recommend that urine tests require normalization to urinary creatinine concentration per gram, and that specimens be collected during acute episodes. In most cases, 24-hour urine collection is not required. Additional testing (genetic or biochemical) may be required to differentiate AHP type (AIP, acute intermittent porphyria; HCP, hereditary coproporphyria; VP, variegate porphyria; ADP, aminolevulinic acid dehydratase [ALAD]-deficiency porphyria).
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<tr>
<td>Mayo-PBG/ PBGU</td>
<td>84110</td>
<td>Porphobilinogen, Quantitative, Random, Urine</td>
<td>Acute Intermittent Porphyria (AIP); Hereditary Coproporphyria (HCP); Porphobilinogen (PBG); Variegate Porphyria (VP)</td>
<td>Urine, frozen (preferred) or refrigerated, protected from light; no preservative necessary, but pH must be &gt;5.0</td>
<td>Transfer a 20-mL specimen to an amber 60-mL urine container (T596)</td>
</tr>
<tr>
<td>Mayo-ALA/ ALAUR</td>
<td>82135</td>
<td>Aminolevulinic Acid, Urine</td>
<td>Acute Intermittent Porphyria (AIP); ALA Dehydratase Deficiency Porphyria (ADP); Delta-Aminolevulinic Acid; Hereditary Coproporphyria (HCP); Variegate Porphyria (VP); 5-Aminolevulinic Acid</td>
<td>Urine (random), refrigerated (preferred) or frozen; patient should abstain from alcohol for 24 hours prior to and during testing</td>
<td>Transfer a specimen to a 10-mL urine tube (T068)</td>
</tr>
<tr>
<td>Mayo-Porphyrins/ PQNRU</td>
<td>84120</td>
<td>Total Porphyrins, Quantitative, Random, Urine</td>
<td>Acute Intermittent Porphyria (AIP); Congenital Erythropoietic Porphyria (CEP); Coproporphyrin; Hereditary Coproporphyria (HCP); PBG (Porphobilinogen); Porphyria Cutanea Tarda (PCT); Uroporphyrin; Variegate Porphyria (VP)</td>
<td>Urine (random), frozen and protected from light; patient should abstain from alcohol for 24 hours prior to collection</td>
<td>Transfer a 20- to 50-mL specimen to an amber 60-mL urine container (T596)</td>
</tr>
<tr>
<td>Quest-PBG/ 6329</td>
<td>84110</td>
<td>Porphobilinogen, Quantitative, Random Urine</td>
<td>PBG</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Quest-ALA/ 6301</td>
<td>82135</td>
<td>Delta- Aminolevulinic Acid, Random Urine</td>
<td>Delta-ALA; Porphyria; Aminolevulinic Acid; D-Aminolevulinic Acid; D-ALA; ALA</td>
<td>Urine (random) with no preservative, or preserved with 1 mL 6N HCl or 1 mL concentrated glacial acetic acid, protected from light (wrap sample in aluminum foil); keep refrigerated and transport refrigerated (cold packs), Rejection criteria: received room temperature or not protected from light</td>
<td>Transfer 2 mL of random urine to a sterile plastic screw-cap container</td>
</tr>
<tr>
<td>Quest-Porphyrins/ 36592</td>
<td>84120</td>
<td>Total Porphyrins, Fractionated, Quantitative, Random Urine</td>
<td>Not listed</td>
<td>Urine (random) (preferred) or random clean-catch urine, refrigerated and protected from light, Rejection criteria: received room temperature, not protected from light, or with a pH &lt;4.0</td>
<td>Transfer a 2-mL specimen to a 5-g sodium carbonate container or container with no preservative</td>
</tr>
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The porphyrins test includes both porphyrins and PBG, and is associated with CPT codes 84120 (porphyrins) and 84110 (PBG).

For the most up-to-date testing and protocol information from Mayo Clinic, please visit www.mayoclinic.org.

For the most up-to-date testing and protocol information from Quest Diagnostics, please visit www.QuestDiagnostics.com.

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