

Acute Hepatic Porphyrria (AHP) Random Urine Testing Reference Guide

Random (spot) urine tests for elevated levels of porphobilinogen (PBG), aminolevulinic acid (ALA), and porphyrins can help inform an AHP diagnosis.*† This guide provides testing information and protocols from national laboratories in the US for ordering tests for PBG, ALA, and porphyrins.

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.

	Lab/ Test Code	CPT Code	Test Name	Synonyms/Related Terms	Specimen Requirements and Storage/Transport Instructions	Specimen Container and Collection
	ARUP-PBG/ 0080260	84110	Porphobilinogen (PBG), Urine	Acute Porphyrria Screening Test; PBG, Quantitative Porphobilinogen	 Urine (random or 24-hour collection), frozen and protected from light	Please visit ARUP Laboratories' online test directory at www.aruplab.com for current specimen collection, transport, and stability guidelines
	ARUP-ALA/ 2011474	82135	Aminolevulinic Acid (ALA), Random Urine	ALA; 5-Aminolevulinic Acid; Delta Aminolevulinic Acid; Delta ALA	 Urine (random), refrigerated; patient should refrain from alcohol consumption 24 hours prior to collection	
	ARUP- Porphyrins/ 2002181	84120	Porphyrins and Porphobilinogen (PBG), Urine*	Uroporphyrin; Heptacarboxyl Porphyrin; Hexacarboxyl Porphyrin; Coproporphyrin; PBG; Pentacarboxyl Porphyrin; Porphobilinogen; Porphyrins, Quantitative, 24-Hour Urine; Urine Quantitative	 Urine (random or 24-hour collection), frozen and protected from light	

*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>.

	Labcorp-PBG/ 003065	84110	Porphobilinogen (PBG), Quantitative, Random Urine	PBG Random Urine, Quantitative	 Urine (random), frozen and protected from light. Causes for rejection: specimen exposed to light; specimen not received frozen	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)
	Labcorp-ALA/ 007364	82135	Δ-Aminolevulinic Acid, Random Urine	ALA, Delta, Random Urine	 Urine (random), frozen and protected from light. Causes for rejection: specimen exposed to light; specimen not received frozen	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)
	Labcorp- Porphyrins/ 120980	84120	Porphyrins, Quantitative, Random Urine	Coproporphyrin; Uroporphyrin	 Urine (random), refrigerated and protected from light. Causes for rejection: stored specimen not refrigerated, specimen exposed to light, acid preservative, pH <3	Transfer a 2-mL specimen to an amber plastic transport tube without preservative (Labcorp No. 23598)

For the most up-to-date testing and protocol information from Labcorp, please visit 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.

CPT=Current Procedural Terminology.

*Mayo Clinic Laboratories suggests 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 coproporphyrria; VP, variegate porphyria; ADP, aminolevulinic acid dehydratase [ALAD]-deficiency porphyria).

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	Lab/ Test Code	CPT Code	Test Name	Synonyms/Related Terms	Specimen Requirements and Storage/Transport Instructions	Specimen Container and Collection
	Mayo-PBG/ PBGU	84110	Porphobilinogen, Quantitative, Random, Urine	Acute Intermittent Porphyria (AIP); Hereditary Coproporphyria (HCP); Porphobilinogen (PBG); Variegate Porphyria (VP)	 Urine (random), frozen immediately following collection, protected from light; no preservative necessary, but pH must be >5.0. Include a list of medications patient is currently taking; patient should abstain from alcohol for 24 hours prior to collection	Transfer a 20-mL specimen to an amber 60-mL urine container (T596)
	Mayo-ALA/ ALAU	82135	Aminolevulinic Acid, Urine	Acute Intermittent Porphyria (AIP); ALA Dehydratase Deficiency Porphyria (ADP); Delta-Aminolevulinic Acid; Hereditary Coproporphyria (HCP); Variegate Porphyria (VP); 5-Aminolevulinic Acid	 Urine (random), refrigerated (preferred) or frozen; patient should abstain from alcohol for 24 hours prior to collection	Transfer a 2-mL specimen to a 10-mL urine tube (T068)
	Mayo- Porphyrins/ PQNRU*	84120	Porphyrins, Quantitative, Random, Urine	Acute Intermittent Porphyria (AIP); Congenital Erythropoietic Porphyria (CEP); Coproporphyrin; Hereditary Coproporphyria (HCP); PBG (Porphobilinogen); Porphyria Cutanea Tarda (PCT); Uroporphyrin; Variegate Porphyria (VP)	 Urine (random), protected from light; patient should abstain from alcohol for 24 hours prior to collection. Include a list of medications patient is currently taking	Transfer a 20- to 50-mL specimen to an amber 60-mL urine container (T596)

*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.mayocliniclabs.com.

	Quest-PBG/ 6329	84110	Porphobilinogen, Quantitative, Random Urine	PBG	 Urine (random) or random clean-catch urine, protected from light (wrap sample in aluminum foil); random urine sent with sodium bicarbonate or without preservative; transport refrigerated (cold packs); do not use preservatives; optimal pH is 6-7. Rejection criteria: received room temperature, not protected from light	Preferred specimen is 2 mL of urine collected in a plastic leak-proof container
	Quest-ALA/ 6301	82135	Delta Aminolevulinic Acid, Random Urine	Delta-ALA; Porphyria; Aminolevulinic Acid; D-Aminolevulinic Acid; D-ALA; ALA	 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	Transfer 2 mL of random urine
	Quest- Porphyrins/ 36592	84120	Porphyrins, Fractionated, Quantitative, Random Urine	Not listed	 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 <4.0	Transfer a 2-mL specimen to a 5-g sodium carbonate container or container with no preservative

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

**For labs not on this guide, confirm testing information and protocols in the lab's test directory by searching each of the three test names:
aminolevulinic acid (ALA), porphobilinogen (PBG), porphyrins.**

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