










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.

	Lab/ Test Code	CPT Code	Test Name	Synonyms/Related Terms	Specimen Requirements and Storage/Transport Instructions	Specimen Container and Collection
	ARUP-PBG/ 2011476	84110	Porphobilinogen (PBG), Urine	PBG; Quantitative Porphobilinogen; Watson-Schwartz Test	 Urine (random), frozen and protected from light	Please visit ARUP Laboratories' online test directory www.aruplab.com for current specimen collection, transport, and stability guidelines
	ARUP-ALA/ 2011474	82135	Aminolevulinic Acid (ALA), Urine	ALA; 5-Aminolevulinic Acid; Delta Aminolevulinic Acid; Delta ALA	 Urine (random), refrigerated or frozen	
	ARUP- Porphyrins/ 2002181	84120	Total Porphyrins, Urine	Porphyrins, Quantitative Urine; Coproporphyrin; Heptacarboxyl Porphyrin; Hexacarboxyl Porphyrin; Pentacarboxyl Porphyrin; Uroporphyrin	 Urine (random), frozen and protected from light  Urine (24 hour collection), frozen and protected from light	

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>.

	LabCorp-PBG/ 003053	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/ 007351	82135	Δ-Aminolevulinic Acid, Random Urine	ALA, Delta, Random Urine; Delta-Aminolevulinic Acid, Random Urine	 Urine (random), frozen and protected from light. Causes for rejection: specimen not protected from light, use of preservative other than 30% acetic acid, use of sodium carbonate preservative	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	Total 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 a 12-mL 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 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 coproporphyrin; 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, frozen (preferred) or refrigerated, protected from light; no preservative necessary, but pH must be >5.0  	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 and during testing 	Transfer a 2-mL specimen to a 10-mL urine tube (T068)
	Mayo- Porphyrins/ PQNRU	84120	Total Porphyrins, Quantitative, Random, Urine	Acute Intermittent Porphyria (AIP); Congenital Erythropoietic Porphyria (CEP); Coproporphyrin; Hereditary Coproporphyrin (HCP); PBG (Porphobilinogen); Porphyria Cutanea Tarda (PCT); Uroporphyrin; Variegate Porphyria (VP)	 Urine (random), frozen and protected from light; patient should abstain from alcohol for 24 hours prior to collection 	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.mayoclinic.org.

	Quest-PBG/ 6329	84110	Porphobilinogen, Quantitative, Random Urine	PBG	 Urine (random) or random clean-catch urine, protected from light (wrap sample in aluminum foil);  ship frozen or refrigerated (cold packs); do not use  preservatives; optimal pH is 6-7	Transfer a 10-mL specimen to a transport tube
	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 to a sterile plastic screw-cap container
	Quest- Porphyrins/ 36592	84120	Total 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 www.QuestDiagnostics.com.

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