



## Porphobilinogen (PBG) ve Aminolevulinic acid (5-ALA) LC-MS/MS Analysis Kit

Porphyrias are inherited metabolic diseases caused by a defect in a specific enzyme in the heme synthetic pathway. Three autosomal dominant acute hepatic porphyrias (AHP) are defined by the occurrence of acute neurovisceral attacks: acute intermittent porphyria (AIP), the most frequent, variegata porphyria (VP), and hereditary coproporphyria (HCP). The precursors, 5-aminolevulinic acid (ALA) and porphobilinogen (PBG), are involved in the heme biosynthetic pathway. ALA and PBG concentrations are elevated in urine and plasma during an attack of acute porphyrias, such as acute intermittent porphyria (AIP), or porphyria variegata (VP). They are used as markers to screen for these conditions. Urinary ALA can also be elevated in cases of lead poisoning.

### Highlights of the Analysis Kit



Simultaneous analysis of PBG and 5-ALA



Total run time 11 min.



Just a few pipetting steps for sample preparation; “dilute and shoot”



Safeguarded by its own stable labelled isotope as internal standard



Long life span of HPLC column

## Parameters

Porphobilinogen (PBG), Aminolevulinic acid (5-ALA)

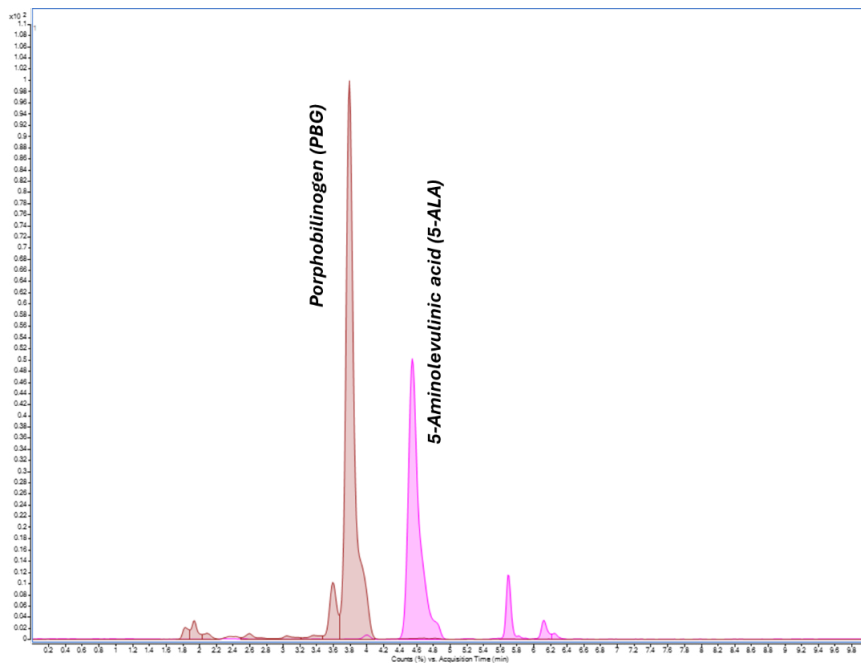
## Sample Type

Urine

## Sample Preparation

1	Pipette 100 $\mu$ L of urine sample into a HPLC vial then, add 25 $\mu$ L of internal standard solution and 375 $\mu$ L of Reagent-1 respectively, vortex 15 sec.
2	Inject to LC-MS/MS system.

## Example Chromatogram



Extracted ion chromatogram of PGB and 5-ALA

## Method Performance

All results were obtained using Agilent 6465 TQ (Ultivo) instrument

Analytes	LOQ (mg/L)	Linearity (mg/L)	Recovery		Repeatability			
			LLQC* (%)	HLQC** (%)	intra-day		inter-day	
					LLQC (%CV)	HLQC (%CV)	LLQC (%CV)	HLQC (%CV)
Porphobilinogen	0.033	1.41 – 15.4	111	100	3.27	2.19	5.06	2.61
5-ALA	0.21	4.12 – 17.9	106	98	4.45	0.80	6.46	0.95

\* LLQC: Low-level quality control

\*\* HLQC: High-level quality control



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