

Reporting Title: PBG Deaminase, WB  
Performing Location: Rochester

**Ordering Guidance:**  
This test is for diagnosis of acute intermittent porphyria. Porphobilinogen deaminase, also known as uroporphyrinogen I synthase, is commonly confused with uroporphyrinogen III synthase, the enzyme deficient in congenital erythropoietic porphyria (CEP). For CEP cases, order UPGC / Uroporphyrinogen III Synthase (Co-Synthase), Erythrocytes.

- Necessary Information:**
- 1. Patient's age is required
  - 2. Include a list of medications the patient is currently taking.

**Specimen Requirements:**  
**Patient Preparation:** Abstinence from alcohol for at least 24 hours prior to specimen collection is essential as ethanol induces porphobilinogen deaminase activity, which may lead to a false-normal result.  
**Container/Tube:**  
**Preferred:** Green top (sodium heparin)  
**Acceptable:** Lavender top (EDTA) or green top (lithium heparin)  
**Specimen Volume:** 4 mL  
Collection Instructions: **Refrigerate specimen as soon as possible.**

- Forms:**
- 1. **New York Clients-Informed consent is required.** Document on the request form or electronic order that a copy is on file. The following documents are available:  
[-Informed Consent for Genetic Testing](#) (T576)  
[-Informed Consent for Genetic Testing-Spanish](#) (T826)
  - 2. If not ordering electronically, complete, print, and send a [Biochemical Genetics Test Request](#) (T798) with the specimen.

Specimen Type	Temperature	Time	Special Container
Whole blood	Refrigerated (preferred)	8 days	
	Ambient	7 days	

**Result Codes:**

Result ID	Reporting Name	Type	Unit	LOINC®
4022	PBG Deaminase, WB	Alphanumeric	nmol/L/sec	12810-8
28400	Interpretation	Alphanumeric		59462-2
606470	Reviewed By	Alphanumeric		18771-6

LOINC® and CPT codes are provided by the performing laboratory.

**Supplemental Report:**  
No

**CPT Code Information:**

82657

**Reference Values:**

Reference ranges have not been established for patients who are younger than 16 years of age.

- > or =7.0 nmol/L/sec
- 6.0-6.9 nmol/L/sec (indeterminate)
- <6.0 nmol/L/sec (diminished)