
Lab Dept: Urine/Stool

Test Name: PORPHYRINS, TIMED URINE

General Information

Lab Order Codes: PORU

Synonyms: Coproporphyrins; Porphobilinogen; Uroporphyrins

CPT Codes: 84110 – Porphobilinogen, quantitative
84120 – Porphyrins, quantitation and fractionation

Test Includes: Includes quantitation of coporphyrins, uroporphyrins, porphobilinogen, hexacarboxylporphyrins and heptacarboxylporphyrins reported in nmol/24 hours.

Logistics

Test Indications: Evaluate patients for porphyrias. Porphyrins are by products of porphyrinogens. Accumulations of either cause porphyrias; hereditary enzyme disorders. They affect the heme biosynthetic pathway and are characterized by increased excretion of porphyrins, porphyrinogens, or their precursors. Early precursors are delta aminolevulinic acid and porphobilinogen. These are water soluble and appear in urine, in common with coproporphyrin and uroporphyrin.

Preferred test during symptomatic periods for acute intermittent porphyria (AIP), hereditary coproporphyria (HCP), and variegate porphyria (VP).

Preferred test to begin assessment for congenital erythropoietic porphyria (CEP) and porphyria cutanea tarda (PCT).

Lab Testing Sections: Urine/Stool - Sendouts

Referred to: Mayo Medical Laboratories (MML Test: PQNU/8562)

Phone Numbers: MIN Lab: 612-813-6280

STP Lab: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: 2 – 3 days, test set up Monday – Friday

Special Instructions:

Submit an entire 24-hour urine collection with sodium carbonate obtained from the laboratory as a preservative. Refrigerate specimen during and after collection. Protect specimen from the light.

Please **include a list of medications** the patient is currently taking and forward with the specimen.

Note: Starting and ending times of collection are required for a timed urine collection and must be documented electronically or on the proper request form.

See [Patient Preparation](#).

Specimen**Specimen Type:**

Urine, timed collection

Container:

Plastic leakproof container with 5 g of sodium carbonate (Mayo Supply T272) added before the start of the collection. Urine GUARD® collection container is preferred for a timed urine sample.

Note: Do not substitute sodium bicarbonate for sodium carbonate.

Draw Volume:

Submit an entire 24-hour urine collection

Processed Volume:

20 - 50 mL (Minimum: 15 mL) urine from a well mixed 24 hour collection

Collection:

For timed urine collections, empty the bladder, discard the voided sample, and note the start time. Collect all urine voided for the specified time period. The specimen must remain covered and protected from light at all times. At the end of the period, note the finishing time; add the last voided sample to the container by emptying the bladder. Bring the refrigerated container to the lab. Make sure all specimens submitted to the laboratory are properly labeled with the patient's name, medical record number and date of birth.

Special Processing:

Lab Staff: Add 5 g sodium carbonate as preservative at start of collection. This preservative is intended to achieve a pH of >7.0.

Measure total urine volume. Remove 50 – 60 mL (Minimum: 15 mL) from a well mixed 24 hr urine specimen. Send aliquot frozen in a plastic, 60 mL amber urine bottle (Mayo Supply T596) to protect specimen from light. **Do Not** substitute Sodium Bicarbonate for Sodium Carbonate.

The pH will generally fall between 8 - 10 if the proper preservative (5 g sodium carbonate) was added to the container before the collection was started.

Send a list of medications with the specimen.

24 hour volume is required on the request form.

Patient Preparation: Avoid alcohol 24 hours prior and during collection.

Sample Rejection: Specimen not protected from light; warm specimens; mislabeled or unlabeled specimens; pH <5.0

Interpretive

Reference Range:

Uroporphyrins (octacarboxyl):
< or =30 nmol/24 hours
Heptacarboxylporphyrins:
< or =9 nmol/24 hours
Hexacarboxylporphyrins:
< or = 8 nmol/24 hours
Pentacarboxylporphyrins:
< or = 10 nmol/24 hours
Coproporphyrin (tetracarboxyl):
Males: < or = 230 nmol/24 hours
Females: < or = 168 nmol/24 hours
Porphobilinogen:
< or = 2.2 mcmol/24 hours

Critical Values: N/A

Limitations:

Porphobilinogen and porphyrins are susceptible to degradation at high temperature, at pH below 7.0, and on exposure to light. Twenty four hour urine collections should be preserved by adding 5.0 g of sodium carbonate to a light resistant collection container prior to beginning the collection.

Ethanol and a variety of medications are known to interfere with heme synthesis leading to elevations in urine porphyrins, particularly coproporphyrin. Coproporphyrin elevation without concomitant porphobilinogen elevation should not be used as the basis for the diagnosis of porphyria, but may warrant follow-up testing with fecal porphyrin analysis.

Methodology:

High performance liquid chromatography (HPLC) with fluorometric detection

References:

[Mayo Medical Laboratories](#) May 2013

Updates:

4/20/2010: Updated reference ranges and units.