



آغا خان یونیورسٹی ہسپتال، کراچی

The Aga Khan University Hospital, Karachi

Stadium Road,

P.O. Box 3500, Karachi 74800, Pakistan

Fax: (92) 21 34934294, Telephone: 34930051

THE AGA KHAN UNIVERSITY HOSPITAL CLINICAL LABORATORIES

UPDATE URINE AMINO ACID (QUANTITATIVE)

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INTRODUCTION:

Quantitative amino acid analysis in urine is helpful for the diagnosis and monitoring of individuals with inborn error of amino acid metabolism. This test is primarily useful to diagnose disorders of renal amino acid transport such as cystinuria or generalized aminoaciduria due to renal Fanconi syndrome. Creatinine is analyzed concurrently and used to normalize results in urine specimens. Depending on the natural history of the disorder, symptoms may be minimized or prevented by early diagnosis and treatment.

PRINCIPLE:

The test is performed by Cation-Exchange HPLC (Biochrom 30+).

SPECIMEN COLLECTION:

- 2-5 ml of urine, early morning samples preferred, otherwise random urine in a container with no preservatives.
- **Minimum acceptable volume of urine is 0.2 ml (200 µl).**
- Freeze sample (-20°C) if it cannot be sent to the laboratory within 4 hours and transport sample frozen in dry ice to the laboratory to prevent bacterial overgrowth.

UNACCEPTABLE CONDITIONS: Unfrozen Sample >24 hrs.

SPECIAL PRECAUTION: Biohazard specimen; to be handled with care.

SCHEDULE:

Reporting will be 10 days after receiving the sample.

NOTE:

- It is essential to fill in the request form related to inborn error of metabolism (inherited metabolic disease) provided at the reception of AKU Clinical Laboratory, Collection Points and Consulting Clinics.
- Instruct patient's attendant (parents or guardians) to provide previous reports related to inherited metabolic disease if available.
- Encourage the patients to get their test charged against the initial laboratory number each time so that a laboratory record related to patient can be readily available.

PLEASE FILE FOR QUICK REFERENCE