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THE AGA KHAN UNIVERSITY HOSPITAL CLINICAL LABORATORIES

UPDATE URINE ORGANIC ACID (QUALITATIVE)

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INTRODUCTION:

Organic acids are compounds that are intermediates or end product of amino acids, carbohydrates, lipids or biogenic amine metabolism. An enzyme defect in a metabolic pathway can lead to accumulation and increased excretion of one or more of these metabolites in urine. Organic acidurias are a group of inborn errors of metabolism that may lead to acute life-threatening illness, developmental delays, and metabolic decompensation. Some examples of organic acidurias are propionic aciduria, methylmalonicaciduria, isovalerylglycinuria, glutaricaciduria type I and type II, etc.

Organic acid analysis by GC-MS is critical for the diagnosis of organic acidurias. Individuals with these disorders may present with sudden catastrophic life threatening illness, associated with metabolic acidosis, hypoglycemia, hyperammonemia, liver dysfunction, cardiomyopathy, or coma. 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 Gas chromatography/ Mass Spectrometry (GC/MS).

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 1.5 ml (1500 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 7 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