



Urinary Organic Acids

Synonyms

UOA, MMA, Orotate, Orotic acid, IVA, PA, succinylacetone

Clinical Indication

Most organic acids are cleared efficiently by the kidney and urine is used for screening.

Urine organic acids by GC-MS can be used to diagnose specific organic acidurias, including methylmalonic aciduria, propionic aciduria and isovaleric aciduria. It is also a comprehensive screening method for a great number of metabolic disorders that can be detected either directly or indirectly. These include disorders of amino acid metabolism (e.g. MSUD, urea cycle defects), organic acidaemias and fatty acid oxidation defects.

Although not an exhaustive list, organic acid disorders often present with nonspecific symptoms such as unexplained metabolic acidosis, reduced levels of consciousness, hypoglycaemia, hyperammonaemia, hypotonia or neurological signs.

Part of Profile / See Also

Request Form

Combined Pathology manual Blood form or ICE request

Availability / Frequency of

Analysis

Referred test: Analysed by Chemical Pathology, Great Ormond Street Hospital 8692, if specific criteria met.

Turnaround Time

2-4 weeks

Patient Preparation

Samples should be collected when the patient is unwell and before treatment or a change in diet alters the pattern of metabolites.

A fasting or pre-breakfast urine is required for investigation of fatty acid oxidation defects.

Sample Requirements

Specimen Type

Urine

Volume

5.0 ml

Container



White Capped Universal

Sample should be transported to laboratory immediately. Samples that are received in the laboratory >1 hour post collection may not be suitable for analysis.

Reference Range & Units

Comprehensive interpretation provided by the reference laboratory.

Interferences

The circumstances under which a sample is collected is of critical importance since in cases of an incomplete metabolic block, characteristic metabolites may only be detected at times of metabolic decompensation (e.g. infection). Acidified samples are **not** acceptable.

Interpretation & Clinical

Decision Value (if applicable)

Comprehensive interpretation and advice provided by the reference laboratory.



PF-PTD-228

References

http://www.labs.gosh.nhs.uk/laboratory-services/chemicalpathology/tests/urine-organic-acids

Test code

Lab Handling

UOA

Freeze primary sample at -20°C immediately. If a sample is received at over one hour since collection, still freeze the sample and enter the test code but also record on the form the time at which the sample was received. This information can then be passed on to the referral laboratory. Samples are sent at -20C by courier to Great Ormond Street Camelia Botnar Labs.



Accredited to ISO 15189:2012