

PF-PTD-228

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
Part of Profile / See Also	neurological signs.
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Request Form	Combined Pathology manual Blood form or ICE request
Availability / Frequency of	Referred test: Analysed by Chemical Pathology, Great Ormond Street Hospital
Analysis	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	Comprehensive interpretation and advice provided by the reference
Decision Value (if applicable)	laboratory.

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References

Test code

Lab Handling

http://www.labs.gosh.nhs.uk/laboratory-services/chemical-

pathology/tests/urine-organic-acids

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 15189:2022