



## NeoMass AAAC Plus

Detection of Amino Acids, Acylcarnitines, Succinylacetone  
and Argininosuccinic Acid by Tandem Mass Spectrometry

## Extended Newborn Screening by LC-MS/MS

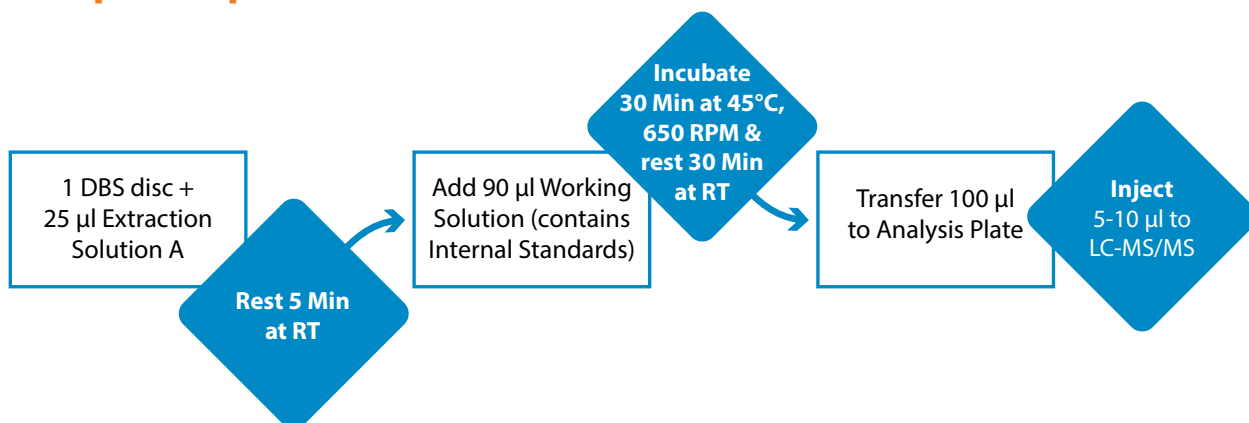
Inborn Errors of Metabolism (IEM) are a genetically heterogeneous group of disorders caused by enzyme defects in the various metabolic pathways. The result is an accumulation of toxic metabolic intermediates and associated metabolic disorders. These concentration changes of metabolites of the amino acid and fatty acid metabolism as well as those of the urea cycle can be determined with the NeoMass AAAC Plus Kit.

The LC-MS/MS NeoMass AAAC Plus kit can be used for the quantitative determination of 15 amino acids, free carnitine, 12 acylcarnitines,

argininosuccinic acid (ASA) and succinylacetone (SUAC) in whole blood dried on filter paper. The determination requires the extraction of Dried Blood Spot samples with a solution containing stable isotope labelled Internal Standards and analysis on a tandem mass spectrometer (MS/MS). The response of each analyte with respect to its Internal Standard is proportional to the concentration of the analyte in the sample. Data is acquired using MRM (Multiple Reaction Monitoring) acquisition. The acquisition and processing of data are carried out by software supplied with the LC-MS/MS system.

- ◆ Ready-to-use CE-IVD kit for 960 analyses
- ◆ Multiple diagnosis (> 50 disorders) in a single run
- ◆ Sample preparation without derivatisation
- ◆ No use of cancerogenic hydrazine or derivatives of it
- ◆ 30 Isotopically labelled Internal Standards
- ◆ With argininosuccinic acid for detection of complete urea cycle disorders
- ◆ Kit also available with 96 Well Filter Plates for customized automation solution
- ◆ All components are also separately available including Internal Standards and Controls
- ◆ Three levels of Dried Blood Spot Controls covering the clinically relevant range

## Sample Preparation Procedure





## NeoMass AAAC Plus Analytes & Internal Standards

Amino acids	
Analytes	Internal Standards
Alanine (Ala)	$^2\text{H}_4$ - Alanine
Arginine (Arg)	$^2\text{H}_4$ - $^{13}\text{C}$ -Arginine
Aspartic acid (Asp)	$^2\text{H}_3$ - Aspartic acid
Citrulline (Cit)	$^2\text{H}_2$ - Citrulline
Glutamic acid (Glu)	$^2\text{H}_3$ - Glutamic acid
Glycine (Gly)	$^{15}\text{N}$ - $^{13}\text{C}$ -Glycine
Leucine (Leu)	$^2\text{H}_3$ - Leucine
Lysine (Lys)	$^{13}\text{C}_6$ - $^{15}\text{N}_2$ -Lysine
Methionine (Met)	$^2\text{H}_3$ - Methionine
Ornithine (Orn)	$^2\text{H}_6$ - Ornithine
Phenylalanine (Phe)	$^{13}\text{C}_6$ - Phenylalanine
Proline (Pro)	$^{13}\text{C}_5$ - Proline
Serine (Ser)	$^{13}\text{C}_3$ - Serine
Tyrosine (Tyr)	$^{13}\text{C}_6$ - Tyrosine
Valine (Val)	$^2\text{H}_8$ - Valine
Argininosuccinic acid (ASA)	$^{15}\text{N}_4$ - $^{13}\text{C}_6$ - Argininosuccinic acid
Succinylacetone (SUAC)	$^{13}\text{C}_5$ - Succinylacetone

Carnitine & Acylcarnitines	
Analytes	Internal Standards
Carnitine (C0)	$^2\text{H}_9$ - Carnitine
Acetylcarnitine (C2)	$^2\text{H}_3$ - Acetylcarnitine
Propionylcarnitine (C3)	$^2\text{H}_3$ - Propionylcarnitine
Butyrylcarnitine (C4)	$^2\text{H}_3$ - Butyrylcarnitine
Isovalerylcarnitine (C5)	$^2\text{H}_9$ - Isovalerylcarnitine
Glutaryl carnitine (C5-DC)	$^2\text{H}_3$ - Glutaryl carnitine
Hexanoylcarnitine (C6)	$^2\text{H}_3$ - Hexanoylcarnitine
Octanoylcarnitine (C8)	$^2\text{H}_3$ - Octanoylcarnitine
Decanoylcarnitine (C10)	$^2\text{H}_3$ - Decanoylcarnitine
Lauroylcarnitine (C12)	$^2\text{H}_3$ - Lauroylcarnitine
Myristoylcarnitine (C14)	$^2\text{H}_9$ - Myristoylcarnitine
Palmitoylcarnitine (C16)	$^2\text{H}_3$ - Palmitoylcarnitine
Stearoylcarnitine (C18)	$^2\text{H}_3$ - Stearoylcarnitine

## Detection of more than 50 disorders in a single run

Amino Acid Disorders	
Argininemia (ARG1 Deficiency)	N-Acetyl Glutamate Synthetase Deficiency (NAGS Deficiency)
Argininosuccinic Aciduria (ASL Deficiency)	
Carbamoylphosphate Synthetase Deficiency 1 (CPS1 Deficiency)	Ornithine Transcarbamoylase Deficiency (OTC Deficiency)
Citrullinemia I (ASS Deficiency)	5-Oxoprolinuria
Citrullinemia II	Phenylketonuria Classical/Hyperphenylalaninemia Defects of Biopterin Cofactor Biosynthesis Defects of Biopterin Cofactor Regeneration
Homocystinuria	
Hypermethioninemia	
Hyperammonemia, Hyperornithinemia, Homocitrullinemia Syndrome 1	Tyrosinemia [detected by SUAC] Transient Neonatal Tyrosinemia
Hyperornithinemia with Gyral Atrophy 1	Tyrosinemia Type I Tyrosinemia Type II
Maple Syrup Urine Disease	Tyrosinemia Type III

Organic Acid Disorders	Fatty Acid Oxidation Disorders
Adenosylcobalamin Synthesis Defects	Carnitine/Acylcarnitine Translocase Deficiency
Glutaric Acidemia Type I, II	Carnitine uptake Deficiency
Isobutyryl-CoA Dehydrogenase Deficiency	Carnitine Palmitoyl Transferase Ia Deficiency
Isovaleric Acidemia	Carnitine Palmitoyl Transferase Ib Deficiency
Malonic Aciduria	Carnitine Palmitoyl Transferase Deficiency Type II
Maternal Vitamin B12 Deficiency	2,4-Dienoyl-CoA Reductase Deficiency I
2-Methylbutyryl-CoA Dehydrogenase Deficiency	3-Hydroxy Long Chain Acyl-CoA Dehydrogenase Deficiency (LCHAD)
3-Methylcrotonyl-CoA Carboxylase Deficiency	Medium Chain Acyl-CoA Dehydrogenase Deficiency (MCAD)
3-Methylglutaconyl-CoA Hydratase Deficiency	Medium Chain Ketoacyl-CoA Thiolase Deficiency
Methylmalonic Acidemias	Medium/Short Chain Hydroxy Acyl-CoA Dehydrogenase Deficiency
Methylmalonyl-CoA Mutase Deficiency	Short Chain Acyl-CoA Dehydrogenase Deficiency
Mitochondrial Acetoacetyl-CoA Thiolase Deficiency	Trifunctional Protein Deficiency
Multiple-CoA Carboxylase Deficiency	Very Long Chain Acyl-CoA Dehydrogenase Deficiency (VLCAD)
Propionic Acidemia	

## Ordering Information

Product Code	Product
7100110	<b>NeoMass AAAC Plus</b> For 960 tests
<b>Kit Contents</b>	Extraction Plate, 10 plates Analysis Plate, 10 plates Dried Blood Spot Controls Level, C1-C2-C3, 5 x 3 spots Internal Standard, labelled amino acids Internal Standard, labelled acylcarnitines Internal Standard, labelled succinylacetone Internal Standard, labelled argininosuccinic acid Extraction Solution A, 50 ml Extraction Solution B, 150 ml Eluent Solution, 1000 ml Extraction Plate Covers, 10 pcs Analysis Plate Covers, 10 pcs Instruction for use

### Kit components are available separately

710011003	DBS Controls, C1-C2-C3, 3 x 3 spots
710011004	Internal Standard, labelled amino acids, 1 Vial
710011005	Internal Standard, labelled acylcarnitines, 1 Vial
710011006	Internal Standard, labelled succinylacetone, 1 Vial
710011007	Internal Standard, labelled argininosuccinic acid, 1 Vial
710011008	Extraction Solution A, 50 ml
710011009	Extraction Solution B, 150 ml
710011010	Eluent Solution, 1000 ml
2205	Sample Preparation Plates, U-bottomed, 10 plates
2605	Sample Analysis Plates, V-bottomed, 10 plates
1541891	Extraction Plate Cover, 10 pcs
1541850	Analysis Plate Cover, 10 pcs



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