NeoMass AAAC 3.0

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NeoMass AAAC 3.0

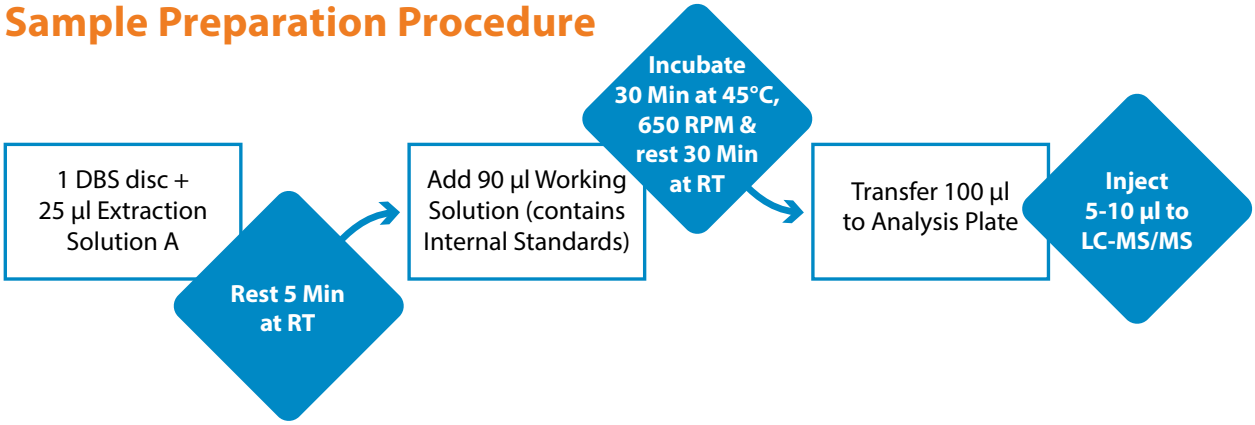
Detection of Amino Acids, Acylcarnitines, Argininosuccinic Acid, Succinylacetone, Adenosine, Deoxyadenosine and Lysophospholipids by Tandem Mass Spectrometry

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 acids, fatty acids and purine metabolisms as well as those of the urea cycle can be determined with the new NeoMass AAAC 3.0 Kit an upgraded version of the former NeoMass AAAC Plus kit. The kit has been extended with regard to the determination of the parameters adenosine, C26 and C26:0 Lysophosphatidylcholine, which now additionally allows the screening for Severe Combined Immunodeficiency due to Adenosine deaminase (ADA-SCID) deficiency as well as X-linked Adrenoleukodystrophy (X-ALD) which is an inherited disorder that mainly affects the nervous system and the adrenal glands. Compared to the NeoMass AAAC Plus Kit, there are no changes in sample preparation. Only the number of Internal Standards has been extended by an additional reaction tube containing the new Internal Standards $^{13}C_5$-Adenosine, $^{15}N_5$-Deoxyadenosine, $^{2}H_3$-C26 and $^{2}H_4$-C26:0-Lysophosphatidylcholine. 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**
- **34 Isotopically labelled Internal Standards including the new Internal Standards $^{13}C_5$-Adenosine, $^{15}N_5$-Deoxyadenosine, $^{2}H_3$-C26 and $^{2}H_4$-C26:0-Lysophosphatidylcholine**
- **With argininosuccinic acid for detection of complete urea cycle disorders**
- **Kit also available with 96 Well Filter Plates for customized automation solution**
- **Three levels of Dried Blood Spot Controls covering the clinically relevant range**

Sample Preparation Procedure



NeoMass AAAC 3.0 Analytes & Internal Standards

Amino acids	
Analytes	**Internal Standards**
Alanine (Ala)	$^{2}H_4$ - Alanine
Arginine (Arg)	$^{2}H_4$ - ^{13}C-Arginine
Aspartic acid (Asp)	$^{2}H_3$ - Aspartic acid
Citrulline (Cit)	$^{2}H_2$ - Citrulline
Glutamic acid (Glu)	$^{2}H_3$ - Glutamic acid
Glycine (Gly)	^{15}N - ^{13}C-Glycine
Leucine (Leu)	$^{2}H_3$ - Leucine
Lysine (Lys)	$^{13}C_6$ - $^{15}N_2$-Lysine
Methionine (Met)	$^{2}H_3$ - Methionine
Ornithine (Orn)	$^{2}H_6$ - Ornithine
Phenylalanine (Phe)	$^{13}C_6$ - Phenylalanine
Proline (Pro)	$^{13}C_5$ - Proline
Serine (Ser)	$^{13}C_3$ - Serine
Tyrosine (Tyr)	$^{13}C_6$ - Tyrosine
Valine (Val)	$^{2}H_8$ - Valine
Argininosuccinic acid (ASA)	$^{15}N_4$ - $^{13}C_6$ - Arginino-succinic acid
Succinylacetone (SUAC)	$^{13}C_5$ - Succinylacetone

Carnitine & Acylcarnitines	
Analytes	**Internal Standards**
Carnitine (C0)	$^{2}H_9$ - Carnitine
Acetylcarnitine (C2)	$^{2}H_3$ - Acetylcarnitine
Propionylcarnitine (C3)	$^{2}H_3$ - Propionylcarnitine
Butyrylcarnitine (C4)	$^{2}H_3$ - Butyrylcarnitine
Isobutyrylcarnitine (C5)	$^{2}H_9$ - Isobutyrylcarnitine
Glutarylcarnitine (C5-DC)	$^{2}H_3$ - Glutarylcarnitine
Hexanoylcarnitine (C6)	$^{2}H_3$ - Hexanoylcarnitine
Octanoylcarnitine (C8)	$^{2}H_3$ - Octanoylcarnitine
Decanoylcarnitine (C10)	$^{2}H_3$ - Decanoylcarnitine
Lauroylcarnitine (C12)	$^{2}H_3$ - Dodecanoylcarnitine
Myristoylcarnitine (C14)	$^{2}H_9$ - Myristoylcarnitine
Palmitoylcarnitine (C16)	$^{2}H_3$ - Palmitoylcarnitine
Stearoylcarnitine (C18)	$^{2}H_3$ - Stearoylcarnitine
Hexacosanoyl-carnitine (C26)	$^{2}H_3$ - Hexacosanoyl carnitine

Lysophospholipid	
Analytes	**Internal Standards**
Lysophosphatidyl-choline (C26:0-LPC)	$^{2}H_{4}$ - Lysophosphatidyl-idylcholine (C26:0-LPC)

Nucleoside	
Analytes	**Internal Standards**
Adenosine (ADO)	$^{13}C_{5}$ - Adenosine (ADO)
	$^{15}N_{5}$ - Deoxyadenosine (dAdo)

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 - Tyrosinemia Type I - Tyrosinemia Type II - Tyrosimenia Type III
Hyperornithinemia with Gyral Atrophy 1	
Maple Syrup Urine Disease	

Organic Acid Disorders	Fatty Acid Oxidation Disorders
Adenosylcobalamin Synthesis Defects	Carnitine/Acylcarnitine Translocase Deficiency
Glutaric Acidemia Type I, II	Carnitine uptake Deficiency
3-Hydroxy-3-Methylglutaryl-CoA Lyase Deficiency	Carnitine Palmitoyl Transferase Ia Deficiency
Isobutyryl-CoA Dehydrogenase Deficiency	Carnitine Palmitoyl Transferase Ib Deficiency
Isovaleric Acidemia	Carnitine Palmitoyl Transferase Deficiency Type II
Malonic Aciduria	2,4-Dienoyl-CoA Reductase Deficiency I
Maternal Vitamin B12 Deficiency	3-Hydroxy Long Chain Acyl-CoA Dehydrogenase Deficiency (LCHAD)
2-Methylbutryl-CoA Dehydrogenase Deficiency	
3-Methylcrotonyl-CoA Carboxylase Deficiency	Medium Chain Acyl-CoA Dehydrogenase Deficiency (MCAD)
3-Methylglutaconyl-CoA Hydratase Deficiency	Medium Chain Ketoacyl-CoA Thiolase Deficiency

Organic Acid Disorders	Fatty Acid Oxidation Disorders
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	

Nucleoside Disorder	Lysophospholipid Disorders
Adenosine Deaminase (ADA) Deficiency	X-Linked Adrenoleukodystrophy (X-ALD)

Ordering Information

Product Code	Product
7100120	**NeoMass AAAC 3.0** For 960 tests
Kit Contents	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, 1 vial Internal Standard, labelled acylcarnitines, 1 vial Internal Standard, labelled succinylacetone, 1 vial Internal Standard, labelled argininosuccinic acid, 1 vial Internal Standard, labelled adenosine, deoxyadenosine, hexacosanoylcarnitine and lysophosphatidylcholine, 1 vial 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

LABSYSTEMS
DIAGNOSTICS
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Labsystems Diagnostics Oy
Tiilitie 3, 01720 Vantaa,
Finland

Phone: +358 20 155 7530
Fax: +358 20 155 7521
www.labsystemsdx.com
sales@labsystemsdx.com

NeoMass AAAC 3.0 7100120