



CellBio NBS kit

Amino Acids and Acylcarnitines
from Dried Blood Spot





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Metabolism is the process by which the body get or make energy from the food and nutrient material. A metabolic disorder occurs when unnatural chemical reactions disrupt the metabolic process. Metabolic disorders are different groups of disorders some effects the decomposition of amino acids, carbohydrates or lipids, or effects the energy producing in the cell.

This product is an In Vitro Diagnostic device that allow the reliable determination of acylcarnitines and amino acids from DBS using tandom mass spectrometry technique.

This product can be used in newborn screening program to identify metabolic disorders as follows:

- **Aminoacidopathies**

Are caused by lack or partial deficiency of biological activity of enzymes involved in amino acid metabolism, that resulting in serious effect on infant's health.

- **Organic acidemia (OA) disorders**

Are arising due to defect in intermediary metabolic pathways of carbohydrates, amino acids and fatty acid oxidation, that resulting in numerous clinical symptoms, such as metabolic acidosis, ketosis, hyperammone-mia, failure to thrive, sepsis or coma.

- **Fatty acid oxidation (FAO) disorders**

Are cause by deficiency of enzymes which needed to decomposition or oxidation of fatty acids, that resulting in delayed physical and mental thrive.



Table 1: List of measurable analytes using CellBio NBS kits

	ANALYTE	ABBREVIATION
Amino acids	Alanine	Ala
	Arginine	Arg
	Aspartic Acid	Asp
	Citrulline	Cit
	Glutamic Acid	Glu
	Glycine	Gly
	Leucine/ Isoleucine/ Hydroxyproline	Leu/Ile/Pro-OH
	Methionine	Met
	Ornithine	Orn
	Phenylalanine	Phe
	Proline	Pro
	Tyrosine	Tyr
	Valine	Val
Acylcarnitines and free Carnitine	Carnitine	C0
	Acetylcarnitine	C2
	Propionylcarnitine	C3
	Malonylcarnitine/3-Hydroxybutyrylcarnitine	C3DC/C4OH
	Butyrylcarnitine	C4
	Methylmalonylcarnitine/3-Hydroxyisovalerylcarnitine	C4DC/C5OH
	Isovalerylcarnitine	C5
	Tiglylcarnitine	C5:1
	Glutaryl carnitine/3-Hydroxyhexanoylcarnitine	C5DC/C6OH
	Hexanoylcarnitine	C6
	Adipylcarnitine	C6DC
	Octanoylcarnitine	C8
	Octenoylcarnitine	C8:1
	Decanoylcarnitine	C10
	Decenoylcarnitine	C10:1
	Decadienoylcarnitine	C10:2
	Dodecanoylcarnitine	C12
	Dodecenoylcarnitine	C12:1
	Tetradecanoylcarnitine	C14
	Tetradecenoylcarnitine	C14:1
	Tetradecadienoylcarnitine	C14:2
	3-Hydroxy-Tetradecanoylcarnitine	C14OH
	Hexadecanoylcarnitine	C16
	Hexadecenoylcarnitine	C16:1
	3-Hydroxy-Hexadecanoylcarnitine	C16OH
	3-Hydroxy-Hexadecenoylcarnitine	C16:1OH
	Octadecanoylcarnitine	C18
	Octadecenoylcarnitine	C18:1
	Octadecadienoylcarnitine	C18:2
	3-Hydroxy-Octadecanoylcarnitine	C18OH
	3-Hydroxy-Octadecenoylcarnitine	C18:1OH

kits' components

Table 2. Ordering information for CellBio NBS kit (non-derivatized)

DESCRIPTION	QUANTITY
CellBio NBS kit (non-derivatized), for Amino Acids and Acylcarnitines, From Dried Blood spot for 960 assays	1 pcs
Internal Standard	4 vials
Mobile Phase	1 bottle
Rinsing Solution	1 bottle
Extraction Buffer	2 bottles
Reconstitution Buffer	2 bottles
Protective Sheets for 96 Well Plates, aluminum foil	10 pcs
96 Well Plates, flat bottom	10 pcs
96 Well Plates, conical bottom	10 pcs
Dried Blood Spot Control Level I , II	1 pcs
Manual	

Table 3. Ordering information for CellBio NBS kit (derivatized)

DESCRIPTION	QUANTITY
CellBio NBS kit (derivatized), for Amino Acids and Acylcarnitines, From Dried Blood spot for 960 assays	1 pcs
Internal Standard	4 vials
Mobile Phase	1 bottle
Rinsing Solution	1 bottle
Extraction Buffer	2 bottles
Derivatization Reagent	3 bottles
Reconstitution Buffer	2 bottles
Protective Sheets for 96 Well Plates, aluminum foil	10 pcs
96 Well Plates, flat bottom	10 pcs
96 Well Plates, conical bottom	10 pcs
Dried Blood Spot Control Level I , II	1 pcs
Manual	

Sample preparation procedure

1. Sample preparation procedure for CellBio NBS kit (non-derivatized)



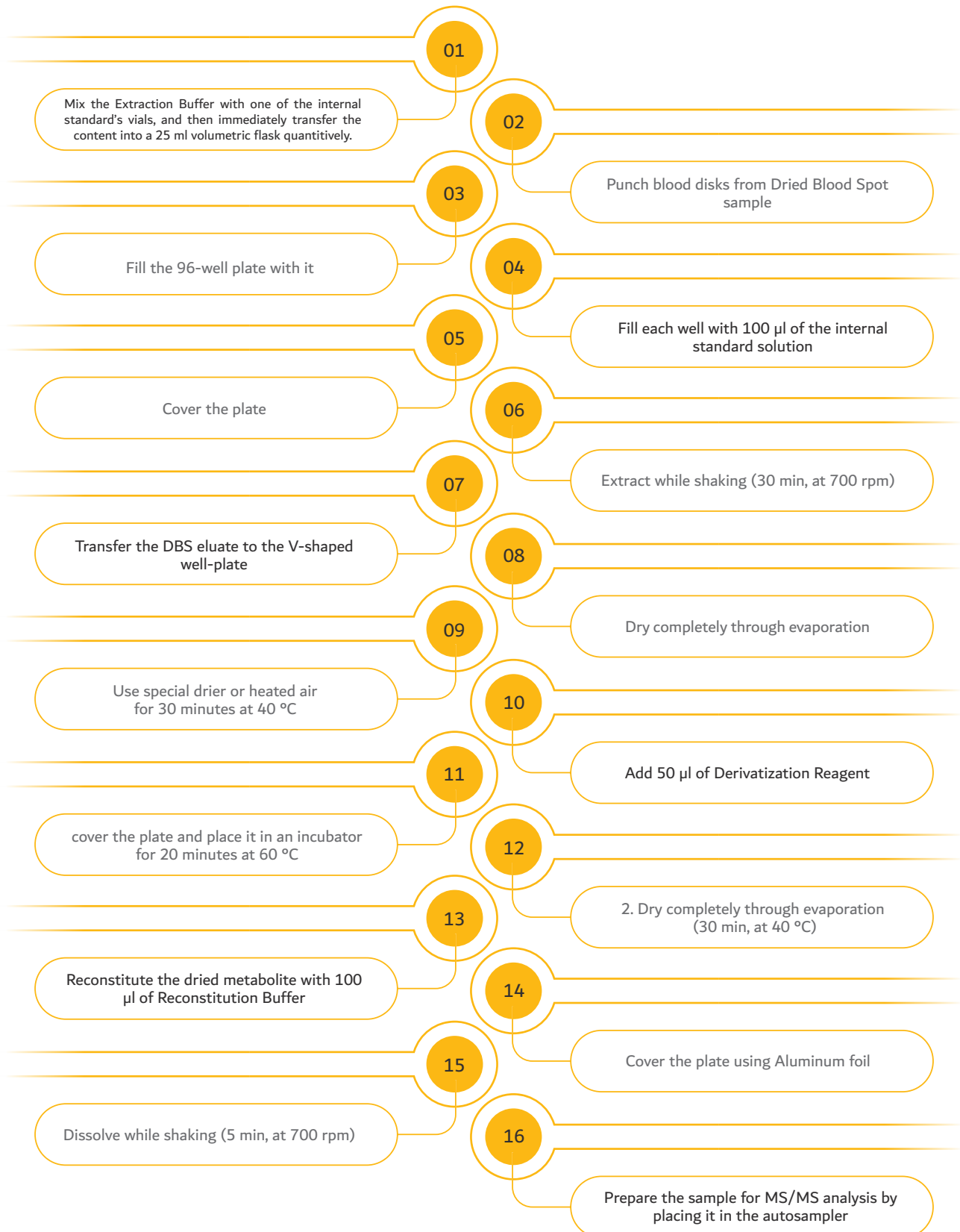


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Sample preparation procedure

2. Sample preparation procedure for CellBio NBS kit (derivatized)



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- **Description of the assay**

CellBio NBS kits include 26 internal standards and controls for the measurement of carnitines and amino acids. Since acylcarnitines with the same chain length would have similar performance characteristics, CellBio NBS kits can measure 13 amino acids and 31 carnitine species. By using the C16 internal standard, the concentration of C16, C16:1, C16OH, and C16:1OH can be determined.

Unlabeled C16 can also be used as an external control for all C16 acylcarnitine series. A semi-quantitative analysis is performed from dried blood spots, and the internal standard is used to spike the sample. Analytes are extracted from the dried blood matrix and each is analyzed with and without derivatization using tandem mass spectrometry.

Accordingly, each analyte responds proportionally to its stable-isotope labeled internal standard.




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