



PH TECH

Paya Hamsan Technologies

PH NBS complete kit

Amino Acids and Acylcarnitines
From Dried Blood Spot

- Is designed to meet the specific requirement of newborn screening laboratories
- Analysis method is according to NBS 04
- Validation method



- **Mission Statement**

We are committed to promoting public health and reducing medical costs by producing goods and providing services in the field of medical diagnosis.

- **Vision & Values**

Achieving a reputable national and international position in the field of production and development of medical diagnostic kits.

- **Social responsibility**

We are aware of our social responsibilities in the fields of environmental protection and natural resources, customer privacy, adherence to ethical principles in business and we do our best to play our role in these areas.



PH NBS/NBS-D

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- **Clinical Background**

Newborn screening is a preventive measure to detect the genetic metabolic deficiency. Free carnitine and acylcarnitines are markers for fatty acid oxidation (FAO) disorders or organic aciduria (OA) and amino acids are marker for amino acidopathies.

- Fatty acid oxidation (FAO) disorders

Fatty acid oxidation disorders, are a group of about 20 defects in fatty acid transport and mitochondrial β -oxidation that are inherited as autosomal recessive disorders.

FAO disorders are caused by a lack or deficiency of the enzymes needed to break down or oxidation of fatty acids, resulting in delayed mental and physical development.

- Organic aciduria (OA) disorders

Organic acidurias (OA) are an important class of inherited metabolic disorders arising due to defect in intermediary metabolic pathways of carbohydrate, amino acids and fatty acid oxidation. It leads to accumulation of organic acids in tissues and their subsequent excretion in urine, that resulting in numerous clinical symptoms, including metabolic acidosis, ketosis, hyperammonemia, failure to thrive, sepsis or coma.

- Amino acidopathies

In this inherited defect is reflected downstream as a lack or a partial biological activity of enzymes involved in amino acids metabolism. As a result, the concentration of the affected amino acids and their metabolites, increases in the infant's body.

These excesses can have severe deleterious effects on the infant's health including death. Amino acid disorders are managed by medical support and nutritional restrictions, supplements and medical foods that limit consumption of an offending amino acid or in some cases protein consumption.

• Intended Use

The PH NBS/PH NBS-D Complete kits are intended for the semi-quantitative measurement and evaluation of amino acids, free carnitine and acylcarnitines concentrations in dried blood samples on the filter paper for newborn screening.

The evaluated values of these analytes and their relationship with each other is intended to provide analyte concentration profiles that interpret metabolic disorder in newborn screening.



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• **General description of the assay**

The PH NBS/PH NBS-D kits are supplied with 26 internal standards and controls for the measurement of carnitines and amino acids. Therefore, the PH NBS/PH NBS-D kits have capability of measuring 13 amino acids and 31 carnitine species, because of acylcarnitines with same chain length have similar performance characteristics. In this way, C16 internal standard can be used to determine the concentrations of the acylcarnitine series C16, C16:1, C16OH and C16:1OH.

Additionally, unlabeled C16 can be used as an external control for all C16 acylcarnitine series. The analysis is done semi-quantitatively from dried blood spots and sample is spiked using the internal standard, the analytes are extracted from the dried blood matrix and analyzed with and without derivatization step using a tandem mass spectrometry system.

The response of each analyte relative to their corresponding stable-isotope labeled internal standard is proportional to analyte concentration.

Components of the complete kits

Table 1. Ordering information for PH NBS complete kit, order no. PH 2001

Order No.	Description	Quantity
PH 2001	PH NBS complete kit, 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	



PH NBS/NBS-D

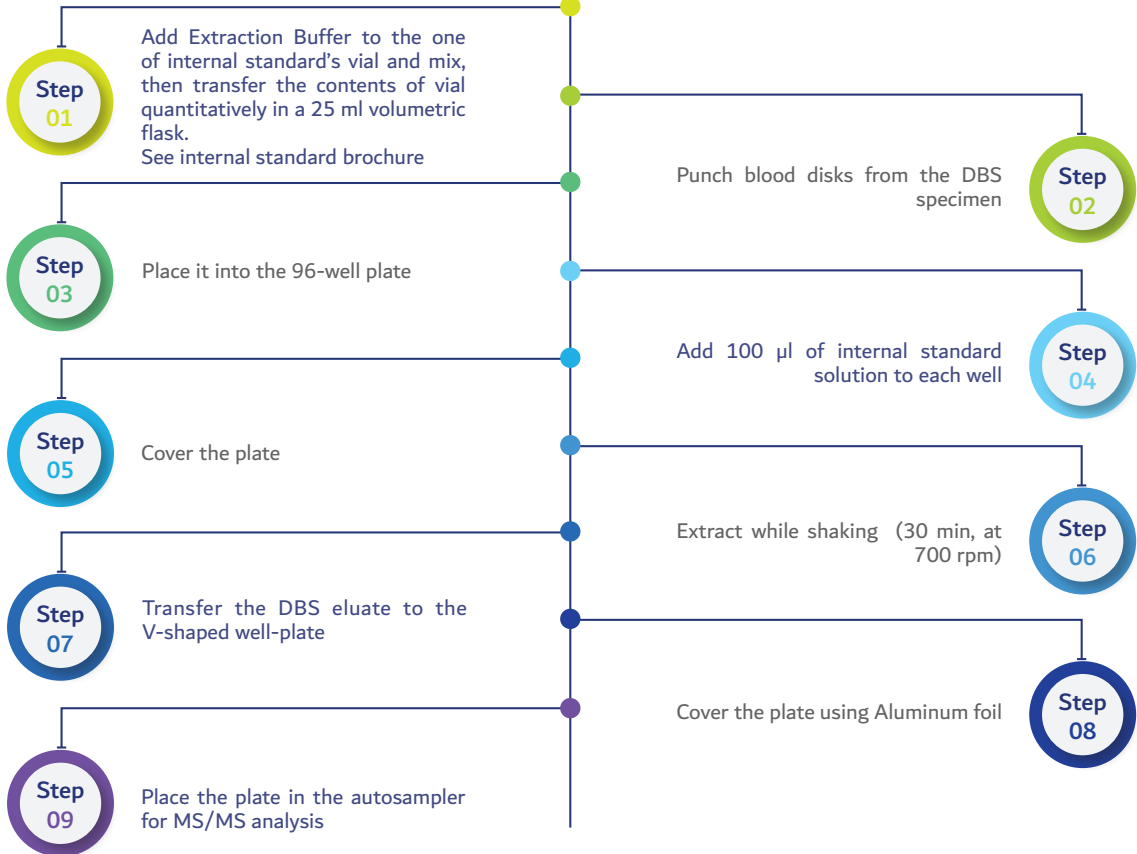
Amino Acids and Acylcarnitines
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Table 2. Ordering information for PH NBS-D complete kit, order no. PH 2002

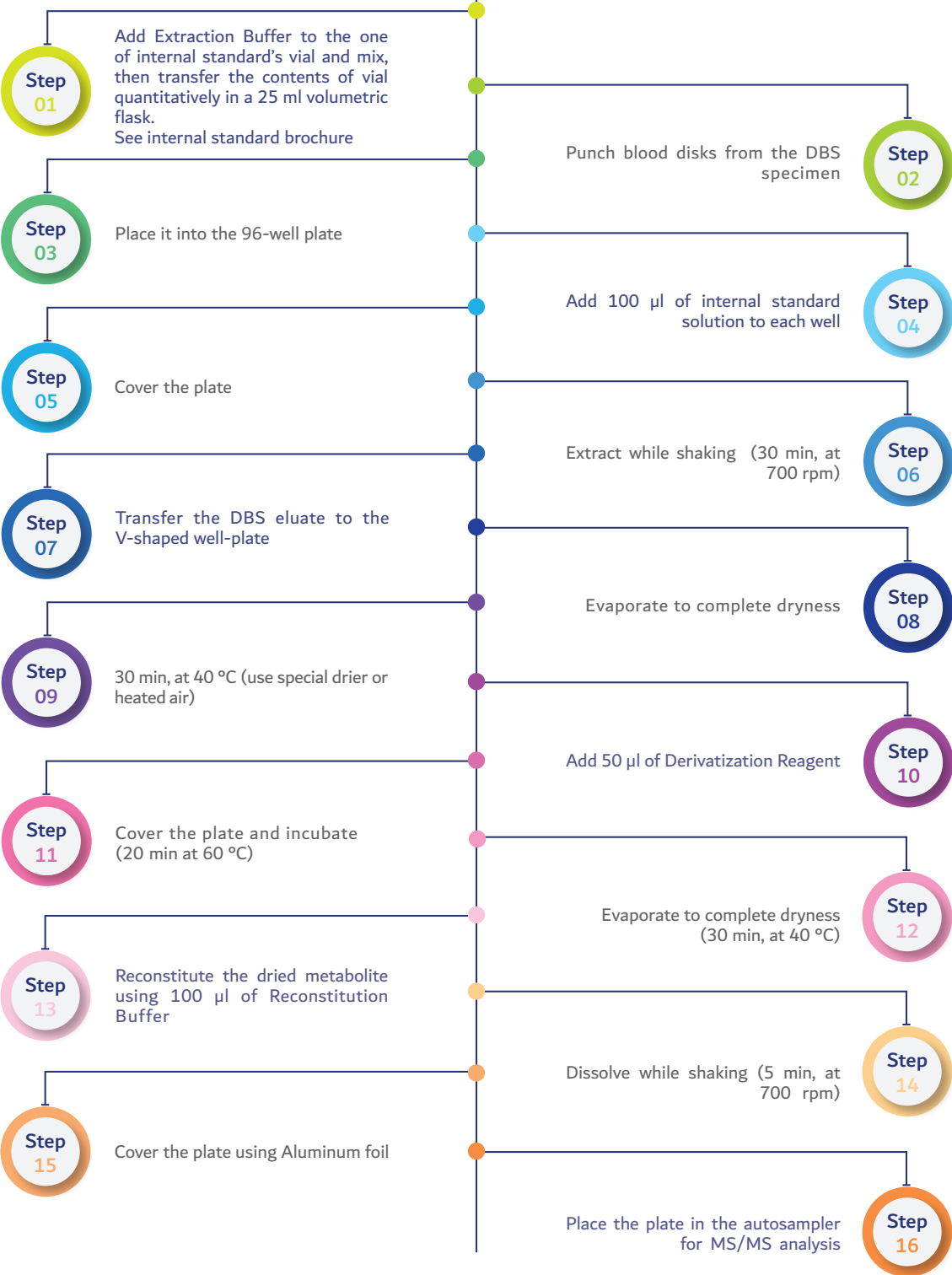
Order No.	Description	Quantity
PH 2002	PH NBS-D complete kit, for Amino Acids and Acylcarnitines, From Dried Blood spot for 960 assays	1 pcs
	Internal Standard	4 vials
	Mobile Phase	1 bottle
	Rinsing Solution	1 bottle
	Extraction Buffer	2 bottles
	Derivatization Reagent	3 bottles
	Reconstitution Buffer	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	

The expiry date of all components of kit is stated on the labels. Store each component as indicated on the corresponding labels.

Sample preparation steps
1. Sample preparation steps for PH NBS kit



Sample preparation steps
2. Sample preparation steps for PH NBS-D kit





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