Cambridge Isotope Laboratories, Inc.
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MS/MS STANDARDS

NSK-A-CE and NSK-B-CE

IVD

ISO13485:2016

Amino Acid Reference Standards and Free Carnitine/Acylcarnitine Reference Standards For MS/MS Newborn Screening

To aid the screening, diagnosis, and monitoring of neonates for inherited metabolic disorders (e.g., phenylketonuria, maple syrup urine disease, medium-chain and very long-chain acyl-CoA dehydrogenase deficiency), **Cambridge Isotope Laboratories, Inc. (CIL) is pleased to offer two sets of CE-marked standards – amino acid reference standards (NSK-A-CE) and carnitine/acylcarnitine reference standards (NSK-B-CE)**. When used as directed (see instructions of use below), these products provide solutions of stable isotope-labeled standards at defined concentrations. Note: the metrics surrounding the concentrations have been rigorously validated for quality system compliance as a CE-marked, *in vitro* diagnostic (IVD) medical device for newborn screening. The ready-to-use assays can be used to measure and evaluate the concentrations of targets (amino acids in NSK-A-CE; free carnitine/acylcarnitines in NSK-B-CE) in a range of biosamples (e.g., dried blood spot, plasma, urine) by a variety of analytical techniques (e.g., FIA-MS/MS, LC-MS/MS, LC-MRM/MS).

NSK-A-CE Device Description and Intended Use

Each vial of the NSK-A-CE reference standard (packaged 10 vials per box) contains a dry mixture of 12 stable isotope-labeled amino acids. Accurate and complete reconstitution of one vial's contents in 1 mL of high-purity solvent will produce the concentrations listed in the table below.

Composition

Reference Standard	Mark	Conc. (µM)
L-Alanine (2,3,3,3-D_4, 98%)	CE	500
L-Arginine·HCl (5-^{13}C, 99%; 4,4,5,5-D_4, 95%)	CE	500
L-Aspartic acid (2,3,3-D_3, 98%)	CE	500
L-Citrulline (5,5-D_2, 98%)	CE	500
DL-Glutamatic acid (2,4,4-D_3, 98%)	CE	500
Glycine (2-^{13}C, 99%; ^{15}N, 98%)	CE	2500
L-Leucine (5,5,5-D_3, 99%)	CE	500
L-Methionine (methyl-D_3, 98%)	CE	500
L-Ornithine·HCl (5,5-D_2, 98%)	CE	500
L-Phenylalanine (ring-$^{13}C_6$, 99%)	CE 0050	500
L-Tyrosine (ring-$^{13}C_6$, 99%)	CE 0050	500
L-Valine (D_8, 98%)	CE	500

For sale in European Economic Area (EEA) – EU and EFTA – only.

Method of Reconstitution

- Solubilize the dried-down mix in 1 mL of purified water:methanol (1:1).
- Vortex manually for 1 minute then auto-vortex for 30 minutes or until complete reconstitution is achieved.

Aliquots of this concentrated stock can then be processed, diluted, or stored (see usage specifications for guidelines).

Usage Specifications

Criteria	Recommendation
Use	960 samples/vial
Before reconstitution:	
Storage	≤25°C; protect from light
Recommended shelf life	4 years
After reconstitution:	
Storage	Store refrigerated (2-8°C) in a tightly sealed vial. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended shelf life	1 month

Warning: Irritant 25°C CE 0050 CE

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NSK-B-CE Device Description and Intended Use

Each vial of the NSK-B-CE reference standard (packaged 10 vials per box) contains a dry mixture of stable isotope-labeled carnitine/acylcarnitines. Accurate and complete reconstitution of one vial's contents in 1 mL of high-purity solvent will produce the concentrations listed in the table below.

Composition

Reference Standard	Abbrev.	Conc. (µM)
L-Carnitine (trimethyl-D_9, 98%)	C0	152.0
O-Acetyl-L-carnitine·HCl (*N*-methyl-D_3, 98%)	C2	38.0
O-Propionyl-L-carnitine·HCl (*N*-methyl-D_3, 98%)	C3	7.6
O-Butyryl-L-carnitine·HCl (*N*-methyl-D_3, 98%)	C4	7.6
O-Isovaleryl-L-carnitine·HCl (*N,N,N*-trimethyl-D_9, 98%)	C5	7.6
O-Octanoyl-L-carnitine·HCl (*N*-methyl-D_3, 98%)	C8	7.6
O-Myristoyl-L-carnitine·HCl (*N,N,N*-trimethyl-D_9, 98%)	C14	7.6
O-Palmitoyl-L-carnitine·HCl (*N*-methyl-D_3, 98%)	C16	15.2

Method of Reconstitution

- Solubilize the dried-down mix in 1 mL of highly pure methanol.
- Vortex manually for 1 minute then auto-vortex for 30 minutes or until complete reconstitution is achieved

Aliquots of the concentrated stock can then be processed, diluted, or stored (see usage specifications for guidelines).

Usage Specifications

Criteria	Recommendation
Use	960 samples/vial
Before reconstitution:	
Storage	≤8°C; protect from light
Recommended shelf life	1 year
After reconstitution:	
Storage	Store refrigerated (2-8°C) in a tightly sealed vial. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended shelf life	1 month

Warning: Irritant

Causes skin irritation. Causes serious eye irritation. May cause respiratory irritation. Avoid breathing dust/fumes/gas/mist/vapors/spray. Wash skin thoroughly after handling. Use only outdoors or in a well-ventilated area. Wear protective gloves, protective clothing/eye protection/face protection. IF ON SKIN: Wash with soap and water. IF INHALED: Remove the victim to fresh air. Keep at rest in a comfortable position. Call a POISON CENTER or doctor/physician if you feel unwell. IF IN EYES: Rinse with water for several minutes. Remove contact lenses.

For sale in European Economic Area (EEA) – EU and EFTA – only.

For professional use only.
In the US, CIL products are labeled "For Research Use Only. Not for use in diagnostic procedures."
Please visit isotope.com or see our "MS/MS Standards" catalog for additional information.

Keep away from sunlight

Upper temperature limit

Do not use if package is damaged

IVD *in vitro* diagnostic medical device

Manufactured by:

EC REP **Authorized representative:**
ABX Advanced Biochemical Compounds, Heinrich-Glaeser-Strasse 10-14, D-01454 Radeberg, Germany

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Supersedes all previously published literature