Cambridge Isotope Laboratories, Inc.
isotope.com

MS/MS STANDARDS

Eurisotop
A CAMBRIDGE ISOTOPE LABORATORIES COMPANY

MS/MS Standards

For Screening and Tuning

Cambridge Isotope Laboratories, Inc.
North America: 1.800.322.1174 cilsales@isotope.com | **International:** +1.978.749.8000 intlsales@isotope.com | fax: 1.978.749.2768 | isotope.com

Table of Contents

Compounds

Please visit **isotope.com** for pricing and availability. *For research use only. Not for use in diagnostic procedures.*

Amino Acid Reference Standards (NSK-A and NSK-A1)

These sets contain 10 vials of 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. Individual vials are also available.

Reconstitution Recommendations for Concentrated Stock

- Solubilize the dried-down mix in 1 mL of 50:50 purified water: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 according to the following instructions, or stored as indicated below.

Dilution Recommendations for Working Stock

To prepare working stock solutions, one of the following procedures is suggested:

- Dilute 1 mL (reconstituted vial contents per instructions above) of the concentrated amino acid stock standard with pure solvent.
- If Set B (Acylcarnitine Reference Standards) was purchased, mix 1 mL (reconstituted vial contents) of concentrated standards from Set A with 1 mL of the concentrated standards from Set B.

Composition

NSK-A		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
Glycine (Gly)	2-^{13}C, 99%; ^{15}N, 98%	2500
DL-Glutamatic acid (Glu)	2,4,4-D_3, 98%	500
L-Alanine (Ala)	2,3,3,3-D_4, 98%	500
L-Arginine·HCl (Arg)	5-^{13}C, 99%; 4,4,5,5-D_4, 95%	500
L-Aspartatic acid (Asp)	2,3,3-D_3, 98%	500
L-Citruline (Cit)	5,5-D_2, 98%	500
L-Leucine (Leu)	5,5,5-D_3, 99%	500
L-Methionine (Met)	methyl-D_3, 98%	500
L-Ornithine·HCl (Orn)*	5,5-D_2, 98%	500
L-Phenylalanine (Phe)	ring-$^{13}C_6$, 99%	500
L-Tyrosine (Tyr)	ring-$^{13}C_6$, 99%	500
L-Valine (Val)	D_8, 98%	500

*NSK-A1 contains Orn 3,3,4,4,5,5,-D_6, 98%. The remaining components are equivalent in NSK-A.

Note: A complementary set of these unlabeled amino acid reference standards (NSK-A-US-1) and a combined set with NSK-B (i.e., NSK-AB) is also available.

Usage Specifications

NSK-A	
Criteria	Recommendation
Use	960 samples/vial
Before reconstitution:	
Storage	room temperature and light protected
Recommended retest	4 years
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

Carnitine and Acylcarnitine Reference Standards (NSK-B)

These sets contain 10 vials of a dry mixture of eight stable isotope-labeled free carnitine and 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. Individual vials are also available.

Reconstitution Recommendations for Concentrated Stock

- 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 according to the following instructions, or stored as indicated below.

Dilution Recommendations for Working Stock

To prepare working stock solutions, one of the following procedures is suggested:

- Dilute 1 mL (reconstituted vial contents per instructions above) of the concentrated acylcarnitine stock standard with pure solvent.
- If NSK-A (Amino Acid Reference Standards) was purchased, mix 1 mL (reconstituted vial contents) of concentrated standards from Set A with 1 mL of the concentrated standards from Set B.

Composition

NSK-B		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
L-Carnitine (C0)	trimethyl-D_9, 98%	152.0
O-Acetyl-L-carnitine·HCl (C2)	*N*-methyl-D_3, 98%	38.0
O-Propionyl-L-carnitine·HCl (C3)	*N*-methyl-D_3, 98%	7.6
O-Butyryl-L-carnitine·HCl (C4)	*N*-methyl-D_3, 98% (CP 97%)	7.6
O-Isovaleryl-L-carnitine·HCl (C5)	*N,N,N*-trimethyl-D_9, 98%	7.6
O-Octanoyl-L-carnitine·HCl (C8)	*N*-methyl-D_3, 98%	7.6
O-Myristoyl-L-carnitine·HCl (C14)	*N,N,N*-trimethyl-D_9, 98%	7.6
O-Palmitoyl-L-carnitine·HCl (C16)	*N*-methyl-D_3, 98%	15.2

Note: A complementary set of these unlabeled carnitine/acylcarnitine standards (NSK-B-US-1) and a combined set with NSK-A (i.e., NSK-AB) is also available.

Usage Specifications

NSK-B	
Criteria	Recommendation
Use	960 samples/vial
Before reconstitution:	
Storage	-5 to 5°C and light protected
Recommended retest	2 years
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

Example References

Haynes, C.A.; De Jesús, V.R. **2016**. Simultaneous quantitation of hexacosanoyl lysophosphatidylcholine, amino acids, acylcarnitines, and succinylacetone during FIA-ESI-MS/MS analysis of dried blood spot extracts for newborn screening. *Clin Biochem, 49(1-2),* 161-165.

Wang, Q.; Sun, T.; Cao, Y.; et al. **2016**. A dried blood spot mass spectrometry metabolomic approach for rapid breast cancer detection. *Onco Targets Ther, 9,* 1389-1398.

Huang, T.; Cao, Y.; Zeng, J.; et al. **2016**. Tandem mass spectrometry-based newborn screening strategy could be used to facilitate rapid and sensitive lung cancer diagnosis. *Onco Targets Ther, 9,* 2479-2487.

George, R.S.; Moat, S.J. **2016**. Effect of dried blood spot quality on newborn screening analyte concentrations and recommendations for minimum acceptance criteria for sample analysis. *Clin Chem, 62(3),* 466-475.

Held, P.K.; Haynes, C.A.; De Jesús, V.R.; et al. **2014**. Development of an assay to simultaneously measure orotic acid, amino acids, and acylcarnitines in dried blood spots. *Clin Chem Acta, 436,* 149-154.

Note: These references utilize NSK-A and NSK-B.

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

continued ➤

Supplemental Acylcarnitine Reference Standards (NSK-B-G1)

These sets contain 10 vials of a dry mixture of five stable isotope-labeled 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. Individual vials are also available.

Reconstitution Recommendations for Concentrated Stock

- 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 according to the following instructions, or stored as indicated below.

Dilution Recommendations for Working Stock

To prepare working stock solutions, mix 1 mL (vial contents) of concentrated standards from NSK-A with 1 mL of the concentrated standards from NSK-B and 1 mL of the concentrated standards from NSK-B-G1.

Composition

NSK-B-G1		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
O-Glutaryl-L-carnitine·CLO_4 (C5DC)	*N*-methyl-D_3, 98% (CP 97%)	152.0
3-Hydroxyisovaleryl-L-carnitine·CLO_4 (C5OH)	*N*-methyl-D_3, 98%	7.6
O-Dodecanoyl-L-carnitine·HCl (C12)	*N,N,N*-trimethyl-D_9, 98%	7.6
O-Octadecanoyl-L-carnitine·HCl (C18)	*N*-methyl-D_3, 98%	15.2
O-3-DL-Hydroxypalmitoyl-L-carnitine·CLO_4 (C16OH)	*N*-methyl-D_3, 98%	15.2

Note: A complementary set of these unlabeled acylcarnitine standards (NSK-B-G1-US) is also available.

Usage Specifications

NSK-B-G1	
Criteria	Recommendation
Use	960 samples/vial
Before reconstitution:	
Storage	-5 to 5°C and light protected
Recommended retest	2 years
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

Example References

Simcox, J.; Geoghegan, G.; Maschek, J.A.; et al. **2017**. Global analysis of plasma lipids identifies liver-derived acylcarnitines as a fuel source for brown fat thermogenesis. *Cell Metab, 26(3),* 509-522.

Haynes, C.A.; De Jesús, V.R. **2016**. Simultaneous quantitation of hexacosanoyl lysophosphatidylcholine, amino acids, acylcarnitines, and succinylacetone during FIA-ESI-MS/MS analysis of dried blood spot extracts for newborn screening. *Clin Biochem, 49(1-2),* 161-165.

George, R.S.; Moat, S.J. **2016**. Effect of dried blood spot quality on newborn screening analyte concentrations and recommendations for minimum acceptance criteria for sample analysis. *Clin Chem, 62(3),* 466-475.

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

Amino Acid and Carnitine/Acylcarnitine Tuning Standards (NSK-A-TS and NSK-B-TS)

These standards are designed to tune the mass spectrometer and are intended to be conducted independent of experimental samples and their preparation. After simple reconstitution in 1 mL of solvent, the working solutions are ready for instrument tuning (as part of regular maintenance), MS/MS troubleshooting, and/or system suitability checks (i.e., before, during, and after batch analysis).

Composition

NSK-A-TS		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
L-Alanine (Ala)	2,3,3,3-D_4, 98%	25
L-Phenylalanine (Phe)	ring-$^{13}C_6$, 99%	25
L-Citrulline (Cit)	5,5-D_2, 98%	25
DL-Glutamic acid (Glu)	2,4,4-D_3, 98%	25
L-Methionine (Met)	methyl-D_3, 98%	25
NSK-B-TS		
L-Carnitine (C0)	trimethyl-D_9, 98%	7.6
O-Propionyl-L-carnitine (C3)	*N*-methyl-D_3, 98%	0.38
O-Octanoyl-L-carnitine (C8)	*N*-methyl-D_3, 98%	0.38
O-Palmitoyl-L-carnitine (C16)	*N*-methyl-D_3, 98%	0.76

Note: The reconstituted tuning standards are not intended to replace the NSK-A and NSK-B reference standards. These tuning standards are available individually and as a set (i.e., NSK-AB-TS-2X1).

Usage Specifications

NSK-A-TS and NSK-B-TS	
Criteria	Recommendation
Use	10 µL/min direct infusion at a frequency of 1-2× per week
Before reconstitution:	
Storage	room temperature and light protected
Recommended retest	4 years for NSK-A-TS and 2 years for NSK-B-TS
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

CIL Application Note

NSK-A-TS and NSK-B-TS Instructions for Use (#41)

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

continued ➤

Succinylacetone Reference Standards (NSK-T)

This set contains 10 vials of dried-down stable isotope-labeled succinylacetone. 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. Individual vials are also available.

Reconstitution Recommendations for Concentrated Stock

- 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 according to the following instructions, or stored as indicated below.

Dilution Recommendations for Working Stock

To prepare working stock solutions, dilute 1 mL (reconstituted vial contents per instructions above) of the concentrated succinylacetone standard with pure solvent.

Composition

NSK-T		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
Succinylacetone (SUAC)	3,4,5,6,7-$^{13}C_5$, 99%	1000

Note: Unlabeled SUAC (ULM-7995) is also available.

Usage Specifications

NSK-T	
Criteria	Recommendation
Use	9,600 samples/vial
Before reconstitution:	
Storage	room temperature, protected from light and moisture
Recommended retest	5 years, if stored as recommended
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

Example References

de Sain-van der Velden, M.G.M.; van der Ham, M.; Gerrits, J.; et al. **2017**. Quantification of metabolites in dried blood spots by direct infusion high resolution mass spectrometry. *Anal Chim Acta, 979,* 45-50.

Haynes, C.A.; De Jesús, V.R. **2016**. Simultaneous quantitation of hexacosanoyl lysophosphatidylcholine, amino acids, acylcarnitines, and succinylacetone during FIA-ESI-MS/MS analysis of dried blood spot extracts for newborn screening. *Clin Biochem, 49(1-2),* 161-165.

Pankowicz, F.P.; Barzi, M.; Legras, X.; et al. **2016**. Reprogramming metabolic pathways *in vivo* with CRISPR/Cas9 genome editing to treat hereditary tyrosinaemia. *Nat Commun, 7,* 12642.

Congenital Adrenal Hyperplasia (CAH) Reference Standards (NSK-S-CAH)

This set contains 10 vials of dried-down stable isotope-labeled steroids. 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. Individual vials are also available.

Reconstitution Recommendations for Concentrated Stock

- 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 according to the following instructions, or stored as indicated below.

Dilution Recommendations for Working Stock

To prepare working stock solutions, dilute 1 mL (reconstituted vial contents per instructions above) of the concentrated succinylacetone standard with pure solvent.

Composition

NSK-S-CAH		
Standard (Abbreviation)	Label and Enrichment	Conc. (µM)
17α-Hydroxyprogesterone (17-OHP)	2,2,4,6,6,21,21,21-D_8, 98%	20
4-Androstene-3,17-dione (A4)*	2,2,4,6,6,16,16-D_7, 97%	20
11-Deoxycortisol (11-S)	2,2,4,6,6-D_5, 98%	20
21-Deoxycortisol (21-S)	2,2,4,6,6,21,21,21-D_8, 97%	20
Cortisol (F)	9,11,12,12-D_4, 98%	100

Note: More concentrated stocks are also available.

*Controlled substance. CIL has a DEA exemption for this product.

Usage Specifications

NSK-S-CAH	
Criteria	Recommendation
Use	48 samples/vial
Before reconstitution:	
Storage	-5 to 5°C and light protected
Recommended retest	5 years, if stored as recommended
After reconstitution:	
Storage	store in a tightly sealed vial at 4°C. To maintain the integrity of the solution, store the sealed vials in a second sealed container.
Recommended retest	1 month

Example References

Gervasoni, J.; Schiattarella, A.; Primiano, A.; et al. **2016**. Simultaneous quantification of 17-hydroxyprogesterone, androstenedione, testosterone and cortisol in human serum by LC-MS/MS using TurboFlow online sample extraction. *Clin Biochem, 49(13-14),* 998-1003.

Hicks, R.A.; Yee, J.K.; Mao, C.S.; et al. **2014**. Precursor-to-product ratios reflect biochemical phenotype in congenital adrenal hyperplasia. *Metabolomics, 10(1),* 123-131.

Dhillon, K.; Ho, T.; Rich, P.; et al. **2011**. An automated method on analysis of blood steroids using liquid chromatography tandem mass spectrometry: application to population screening for congenital adrenal hyperplasia in newborns. *Clin Chem Acta, 412(23-24),* 2076-2084.

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

continued ➤

Additional Standards for Screening

The additional products outlined below are available as individual dried-down mixtures of their substrate and internal standard.

Lysosomal α-Glucosidase Substrate and Internal Standard (NSK-PO-1)

Each vial contains the following compounds at a molar ratio of 100:1.

	Substrate	Internal Standard
Compound Name	(7-Benzoylamino-heptyl)-{2-[4-(3,4,5-trihydroxy-6-hydroxymethyl-tetrahydro-pyran-2-yloxy)-phenylcarbamoyl]-ethyl}-carbamic acid *tert*-butyl ester	(7-D_5-Benzoylamino-heptyl)-[2-(4-hydroxy-phenyl-carbamoyl)-ethyl]-carbamic acid *tert*-butyl ester
Chemical Formula	$C_{34}H_{49}N_3O_{10}$	$C_{28}H_{34}N_3O_5D_5$
MW (Da)	659.8	502.7

Reconstitution Recommendations

- Solubilize with 1.8 mL of 100 g/L CHAPS in water. Vortex.
- Add 15.9 mL of buffer – 0.34 M sodium phosphate (monobasic), 0.17 M sodium citrate (tribasic) dihydrate, pH 4.0. Vortex.
- Add 0.3 mL of 0.8 mM acarbose in water.

α-Galactosidase Substrate and Internal Standard (NSK-FA-1)

Each vial contains the following compounds at a molar ratio of 500:1.

	Substrate	Internal Standard
Compound Name	(6-Benzoylamino-hexyl)-{2-[4-(3,4,5-trihydroxy-6-hydroxymethyl-tetrahydro-pyran-2-yloxy)-phenylcarbamoyl]-ethyl}-carbamic acid *tert*-butyl ester	(6-D_5-Benzoylamino-hexyl)-[2-(4-hydroxy-phenyl-carbamoyl)-ethyl]-carbamic acid *tert*-butyl ester
Chemical Formula	$C_{33}H_{47}N_3O_{10}$	$C_{27}H_{32}N_3O_5D_5$
MW (Da)	645.7	488.6

Reconstitution Recommendations

- Solubilize with 0.45 mL of aqueous sodium taurocholate (120 g/L sodium taurocholate, ≥95% purity). Vortex.
- Add 14.67 mL of 0.174 M sodium acetate buffer (pH 4.6). Vortex.
- Add 2.88 mL of 1 M N-acetylgalactosamine in water.

Glucocerebrosidase Substrate and Internal Standard (NSK-GA-1)

Each vial contains the following compounds at a molar ratio of 50:1.

	Substrate	Internal Standard
Compound Name	D-Glucosyl-β1-1′-N-dodecanoyl-D-*erythro*-sphingosine [C12-glucocerebroside]	*N*-Myristoyl-D-*erythro*-sphingosine [C14-ceramide]
Chemical Formula	$C_{36}H_{69}NO_8$	$C_{32}H_{63}NO_3$
MW (Da)	643.9	509.8

Reconstitution Recommendations

- Solubilize with 2.4 mL of aqueous sodium taurocholate (120 g/L sodium taurocholate, ≥95% purity). Vortex.
- Add 15.6 mL of a 0.72 M phosphate, 0.36 M citrate buffer, pH 5.1 (prepared with sodium phosphate monobasic and sodium citrate tribasic dihydrate). Vortex.

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

Additional Standards for Screening (continued)

Galactocerebrosidase Substrate and Internal Standard (NSK-KR-1)

Each vial contains the following compounds at a molar ratio of 150:1.

	Substrate	Internal Standard
Compound Name	D-Galactosyl-β1-1'-octanoyl-D-*erythro*-sphingosine [C8-galactosylceramide]	*N*-Decanoyl-D-*erythro*-sphingosine [C10-ceramide]
Chemical Formula	$C_{32}H_{61}NO_8$	$C_{28}H_{55}NO_3$
MW (Da)	587.8	453.7

Reconstitution Recommendations

- Solubilize with 1.8 mL of aqueous sodium taurocholate (96 g/L sodium taurocholate, ≥95% purity and 12 g/L oleic acid). Vortex.
- Add 16.2 mL of a 0.2 M phosphate, 0.1 M citrate buffer, pH 4.4 (prepared with sodium phosphate monobasic and sodium citrate tribasic dihydrate). Vortex.

α-L-Iduronidase Substrate and Internal Standard (NSK-MP-1)

Each vial contains the following compounds at a molar ratio of 150:1.

	Substrate	Internal Standard
Compound Name	(7-(1-Iduronic acid)-oxycoumarin-4-methylamine-(5'-*N*-boc-aminopentanoyl)-amide)	(7-Hydroxycoumarin-4-methylamine- (4'-*N*-boc-aminobutanoyl)-amide)
Chemical Formula	$C_{26}H_{34}N_2O_{12}$	$C_{19}H_{24}N_2O_6$
MW (Da)	566.55	376.4

Reconstitution Recommendations

- Solubilize with 0.5 mL of inhibitor solution (3.0 mM D-saccharic acid 1,4 lactone monohydrate in water). Vortex.
- Add 17.5 mL of sodium formate buffer (0.11 M sodium formate and 0.16 M formic acid, pH 3.6). Vortex.

Acid Sphingomyelinase Substrate and Internal Standard (NSK-NI-1)

Each vial contains the following compounds at a molar ratio of 50:1.

	Substrate	Internal Standard
Compound Name	*N*-Hexanoyl-D-*erythro*-sphingosylphosphorylcholine [C6-sphingomyelin]	*N*-butyroyl-D-*erythro*-sphingosine [C4-ceramide]
Chemical Formula	$C_{29}H_{59}N_2O_6P$	$C_{22}H_{43}NO_3$
MW (Da)	562.8	369.6

Reconstitution Recommendations

- Solubilize with 0.15 mL of aqueous sodium taurocholate (120 g/L sodium taurocholate, ≥95% purity). Vortex.
- Add 17.85 mL of 0.93 M sodium acetate, 0.604 mM zinc chloride buffer, pH 5.7. Vortex.

Usage Specifications

Criteria	Recommendation
Use	~600 samples/vial
Before reconstitution:	
Storage	-20°C and light protected
Recommended retest	2 years
After reconstitution:	
Storage	2-8°C or -20°C
Recommended retest	4 weeks

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

continued ➤

Additional Standards for Screening (continued)

Catalog No.	Standard (Abbreviation)	Label and Enrichment	Unit Size
CNLM-9007-CA	L-Argininosuccinic acid (ASA), barium salt·2H_2O	arginine-$^{13}C_6$, 99%; $^{15}N_4$, 99% (CP 90%)	0.1 mg, 0.5 mg
ULM-9008-CA	L-Argininosuccinic acid (ASA), barium salt·3H_2O	unlabeled (CP 90%)	100 mg
CLM-9426	Ethylmalonic acid (EMA)	$^{13}C_4$, 99%	Please inquire
DLM-6013	Ethylmalonic acid (EMA)	methyl-D_3, 98%	0.1 g
DLM-3619	DL-Homocystine (HCY)	3,3,3′,3′,4,4,4′,4′-D_8, 98%	0.5 g, 1 g
CLM-8742	allo-Isoleucine (alle)	$^{13}C_6$, 97-99%	Please inquire
DLM-1505	allo-Isoleucine (alle)	D_{10}, 98%	0.1 g
DLM-2312	Methylcitric acid (MCA)	methyl-D_3, 98% (CP 90%)	Please inquire
CLM-9426	Methylmalonic acid (MMA)	$^{13}C_4$, 99%	0.1 g
DLM-387	Methylmalonic acid (MMA)	methyl-D_3, 98%	250 mg
NLM-1048	Orotic acid (Oro)·H_2O	1,3-$^{15}N_2$, 98%	250 mg
CLM-2260	L-Proline (Pro)	$^{13}C_5$, 99%	0.1 g, 0.25 g, 0.5 g
DLM-487	L-Proline (Pro)	D_7, 97-98%	100 mg, 250 mg
ULM-8333	L-Proline (Pro)	unlabeled	50 mg, 100 mg
DLM-10520	Lysophosphatidylcholine 20:0 (LYSO-PC 20:0)	eicosanoyl-12,12,13,13-D_4, 98%	1 mg, 5 mg
ULM-10521	Lysophosphatidylcholine 20:0 (LYSO-PC 20:0)	unlabeled	5 mg, 10 mg
CLM-10499	Lysophosphatidylcholine 22:0 (LYSO-PC 22:0)	docosanoyl-1,2,3,4,5,6-$^{13}C_6$, 99%	1 mg, 5 mg
DLM-10500	Lysophosphatidylcholine 22:0 (LYSO-PC 22:0)	docosanoyl-12,12,13,13-D_4, 98%	1 mg, 5 mg
ULM-10498	Lysophosphatidylcholine 22:0 (LYSO-PC 22:0)	unlabeled	5 mg, 10 mg
CLM-10496	Lysophosphatidylcholine 24:0 (LYSO-PC 24:0)	tetracosanoyl-1,2,3,4,5,6-$^{13}C_6$, 99%	1 mg, 5 mg
DLM-10497	Lysophosphatidylcholine 24:0 (LYSO-PC 24:0)	tetracosanoyl-12,12,13,13-D_4, 98%	1 mg, 5 mg
ULM-10495	Lysophosphatidylcholine 24:0 (LYSO-PC 24:0)	unlabeled	5 mg, 10 mg
CLM-9792	Lysophosphatidylcholine 26:0 (LYSO-PC 26:0)	hexacosanoyl-1,2,3,4,5,6-$^{13}C_6$, 99%	1 mg, 5 mg
DLM-10501	Lysophosphatidylcholine 26:0 (LYSO-PC 26:0)	hexacosanoyl-12,12,13,13-D_4, 98%	1 mg, 5 mg
ULM-9791	Lysophosphatidylcholine 26:0 (LYSO-PC 26:0)	unlabeled	5 mg, 10 mg

Example References

Monostori, P.; Klinke, G.; Richter, S.; et al. **2017**. Simultaneous determination of 3-hydroxypropionic acid, methylmalonic acid and methylcitric acid in dried blood spots: Second-tier LC-MS/MS assay for newborn screening of propionic acidemia, methylmalonic acidemias and combined remethylation disorders. *PLoS One, 12(9),* e0184897.

Prinsen, H.C.M.T.; Schiebergen-Bronkhorst, B.G.M.; Roeleveld, M.W.; et al. **2016**. Rapid quantification of underivatized amino acids in plasma by hydrophilic interaction liquid chromatography (HILIC) coupled with tandem mass-spectrometry. *J Inherit Metab Dis, 39(5),* 651-660.

Held, P.K.; Haynes, C.A.; De Jesús, V.R.; et al. **2014**. Development of an assay to simultaneously measure orotic acid, amino acids, and acylcarnitines in dried blood spots. *Clin Chem Acta, 436,* 149-154.

Please visit isotope.com for a list of additional compounds.

Chemical purity (CP) is 98% or greater, unless otherwise indicated.

MS/MS Parameter Examples (all *m/z* as $[M+H]^+$)

Neutral loss (NL) scan (for NSK-A and NSK-A1 standards)

Compound	Abbreviation	Underivatized		Butyl Ester Derivatized	
		Precursor Ion *m/z*	NL *m/z*	Precursor Ion *m/z*	NL *m/z*
Alanine (D_4)	Ala	94	46	150	102
Arginine (^{13}C; D_4)	Arg	180	105	236	161
Aspartate (D_3)	Asp	137	46	249	102
Citrulline (D_2)	Cit	178	63	234	119
Glutamate (D_3)	Glu	151	46	263	102
Glycine (^{13}C; ^{15}N)	Gly	78	46	134	102
Leucine (D_3)	Leu	135	46	191	102
Methionine (D_3)	Met	153	46	209	102
Ornithine (D_2)	Orn	135	63	191	119
Ornithine (D_6)	Orn	139	63	195	119
Phenylalanine ($^{13}C_6$)	Phe	172	46	228	102
Tyrosine ($^{13}C_6$)	Tyr	188	46	244	102
Valine (D_8)	Val	126	46	182	102

Note: The MS/MS fragmentation mechanism of amino acids during NL scan is well established (e.g., PMID: 14578311). For example, the losses for the underivatized amino acids reflect HCOOH (*m/z* 46), HCOOH and NH_3 (*m/z* 63), and HCOOH and $H_2NCNHNH_2$ (*m/z* 105).

Precursor (Pre) ion scan (for NSK-B and NSK-B-G1 standards)

Compound	Abbreviation	Underivatized		Butyl Ester Derivatized	
		Precursor Ion *m/z*	Product Ion *m/z*	Precursor Ion *m/z*	Product Ion *m/z*
Carnitine (D_9)	C0	171	85	227	85
Acetylcarnitine (D_3)	C2	207	85	263	85
Propionylcarnitine (D_3)	C3	221	85	277	85
Butyrylcarnitine (D_3)	C4	235	85	291	85
Isovalerylcarnitine (D_9)	C5	255	85	311	85
Glutarylcarnitine (D_3)	C5DC	279	85	391	85
Hydroxyisovalerylcarnitine (D_3)	C5OH	265	85	321	85
Octanoylcarnitine (D_3)	C8	291	85	347	85
Dodecanoylcarnitine (D_9)	C12	353	85	409	85
Myristoylcarnitine (D_9)	C14	381	85	437	85
Palmitoylcarnitine (D_3)	C16	403	85	459	85
Octadecanoylcarnitine (D_3)	C18	431	85	487	85

Note: The common fragment ion of *m/z* 85 corresponds to $^+CH_2$-CH=CH-COOH and is consistent between non-derivatized and derivatized acylcarnitines (e.g., PMID: 9365395 for background and fragmentation mechanism).

Please visit isotope.com for a complete list of isotope-labeled compounds.

Research products are distributed and sold worldwide via our extensive network.

To request a quotation or place an order, please contact CIL Sales at
email: cilsales@isotope.com | telephone: 1.978.749.8000 | 1.800.322.1174 (North America)

For our international customers

Please contact CIL International Sales at
email: intlsales@isotope.com | telephone: +1.978.749.8000

CIL's distributor listing is available at: isotope.com

Setting the Standard...

Donald H. Chace, PhD MSFS FACB
Medolac Laboratories

"In clinical chemistry-based applications of mass spectrometry, the first lesson the laboratory learns is the requisite nature of stable isotope-enriched standards for quantification of metabolites in biological fluids. In newborn screening of amino acids and acylcarnitines, Cambridge Isotope Laboratories, Inc., set the standard for quantification of these metabolites in dried blood spots. As research and development of the newborn screening analysis by mass spectrometry progressed, it was clear that a half dozen isotope-labeled internal standards would not be adequate for the analysis of an amino acid and acylcarnitine profile, together comprising a range of 500 separate mass units and more than 30 important metabolites, most of which require accurate quantification. When screening began to expand beyond research, it was clear that weighing out small quantities of individual standards would reduce accuracy and introduce unnecessary error. Therefore, together, we set out to develop sets of standards for amino acids and acylcarnitine analysis that would enable quantification. We started this development more than 20 years ago adding, changing and improving these standards. CIL, together with the early developers of tandem mass spectrometry-based newborn screening, set the standard by which all other laboratories follow. CIL's commitment to supporting the metabolic and newborn screening community is exceptional. It is our good fortune in the clinical chemistry and mass spectrometry community to have CIL as part of our laboratory solutions."

For research use only. Not for use in diagnostic procedures.

The references in this document were chosen to represent the wide range of applications that CIL products support. Papers not referenced in this document do not imply a lesser impact on the scientific community.

These articles do not imply that materials produced by CIL are suitable for any intended purpose.

To request a quotation or place an order, please contact our sales team

Euriso-Top International

Parc des Algorithmes
Bâtiment Homère, route de l'orme
F-91194 Saint-Aubin Cedex
Tel : +33 (0)1 69 41 95 96
Fax : +33 (0)1 69 41 93 52
sales@eurisotop.com

Euriso-Top GmbH

Lebacher Str. 4
66005 Saarbrücken
Tel : +49 (0) 681 99 63 338
Fax : +49 (0) 681 99 63 438
sales@eurisotop.de