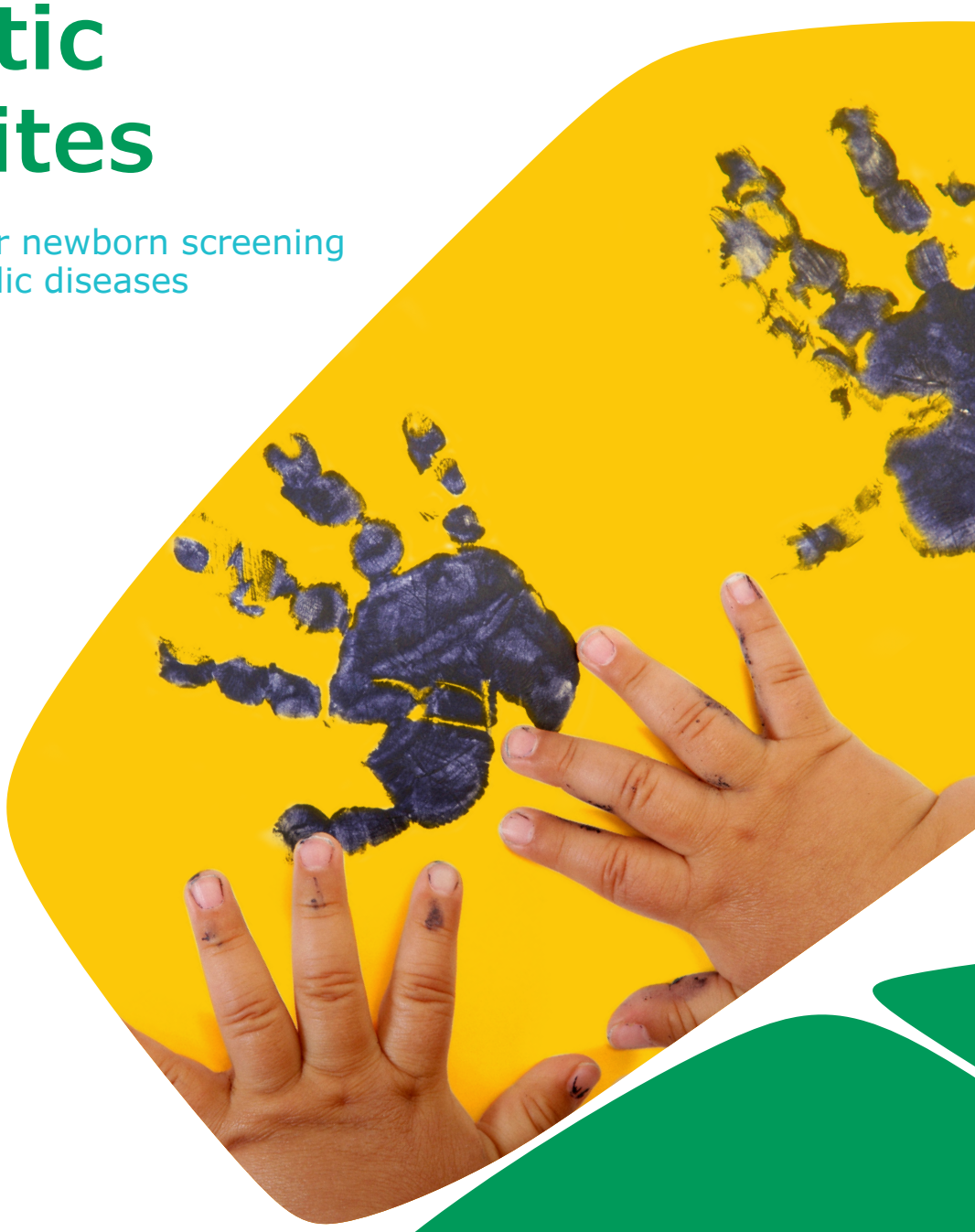
MilliporeSigma

Diagnostic Metabolites

Reference Materials for newborn screening & detection of metabolic diseases



Supelco®
Analytical Products

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Inborn Errors of Metabolism (IEMs) are a group of complex and heterogeneous monogenic disorders with a wide array of clinical symptoms. They are the result of an error in a genetic code causing lowered or deficient activity of an enzyme in a metabolic pathway. These metabolic disorders often cause specific metabolites to accumulate in the patient's serum and urine. The severe clinical consequences of IEMs make the detection of involved diagnostic metabolites of vital importance. Any delay in the diagnosis and treatment of these disorders, leads to a variety of adverse outcomes, that includes moderate-to-severe neuropsychological dysfunction, mental retardation, and death.

Newborn screening aims to detect these congenital, genetic, and metabolic disorders early among the newborns. The early detection of these inborn metabolic disorders can help preventing the morbidity or mortality associated with them by early disease management using dietary or drug interventions in the affected newborns. The different diseases include defects in amino acid metabolism such as phenylketonuria (PKU) and maple syrup urine disease (MSUD), defects in fatty acid metabolism and organic acidurias (isovaleric acidemia).

Our Supelco® portfolio offers the widest selection of high-quality reference materials (RMs) of acylcarnitines, acylglycines, amino acids, fatty acids, and organic acids, along with their stable-labeled internal standards for use in newborn screening and detection of IEMs by MS, GC/MS, or LC/MS methods.

Supelco® analytical standards and certified reference materials (CRMs) are manufactured and tested under the highest industry standards of ISO 17034, ISO/IEC 17025, and ISO 9001. The Cerilliant® certified solution standards and Certified Spiking Solutions® products are supported by a comprehensive Certificate of Analysis (CoA). The accompanying Certificate of Analysis (CoA) provides all analytical data on stability, homogeneity, accuracy, uncertainty, and traceability to support the regulatory requirements. The Cerilliant® certified reference materials are designed to promote a laboratory's efficiency, accuracy and reliability of results, besides the regulatory requirements. These accurate reference materials can be used as starting materials for the preparation of controls in calibration and quantitation, including isotope dilution methods by GC/MS or LC/MS.

Acylcarnitines

Acylcarnitines are intermediate compounds formed during the oxidation of fatty acids and amino acids in the tissues and body fluids. They serve as important diagnostic biomarkers in the detection of inherited peroxisomal and mitochondrial oxidation disorders. Abnormal concentrations of specific acylcarnitines are used in the diagnosis of carnitine deficiency, fatty acid oxidation defects, and organic acidurias, such as carnitine-acylcarnitine translocase deficiency (CACTD) or Isobutyryl-CoA dehydrogenase deficiency. Acylcarnitines used as primary markers on dried blood filter papers (DBS) in newborn screening can help in the detection of more than 40 different inborn errors of metabolism. Liquid chromatography-tandem mass spectrometry (LC-MS/MS) has significantly increased screening possibilities in newborn screening programs and has established itself as an efficient and robust methodology for the analysis of acyl-L-carnitines. MS detection provides the required sensitivity and direct infusion enough to detect several disorders with a single injection. For more detailed analysis and for measuring acylcarnitine isomers that are closely related, separation by liquid chromatography before detection is important. Supelco® analytical products offers the widest selection of high-quality analytical standards and certified reference materials (CRMs) of acylcarnitines including the native, stable isotope-labeled internal standards, and solution-based multi-analyte acylcarnitine CRM mixes suitable for use in newborn screening and the detection of metabolic diseases by MS, GC/MS, or LC/MS.

Cerilliant® portfolio is the first to offer certified reference materials (CRMs) of native and stable isotope-labeled, routinely tested carnitine and acetylcarnitine as single solutions and multi-component solution mixes. Our Certified Spiking Solutions® products are suitable for various critical quantitative applications in research and clinical environments such as:

- Preparation of internal standards, controls, calibrators, and linearity standards for LC-MS/MS assays
- As system tuning/suitability standards
- Linearity verification
- Internal proficiency testing.

The stable and quantitative solution format provides significant benefits to the user such as the convenience of use, material savings, and increased efficiency of labor. Certified concentration values for each component provides increased consistency and efficiency from less time remaking pools due to dilution errors or rerunning samples.

Acylcarnitines Certified Reference Materials

Product no.	Product name	Pack Size
A-141-1ML	Acylcarnitines Mix 1 (C0,C2)	10.0 mg/mL (each analyte as free base) in methanol
A-144-1ML	Acylcarnitines Mix 2 (C4,iC4,C5,iC5,C6,C8,C10,C12,C14,C18)	100-200 µg/mL in 90:10 methanol:1% aqueous formic acid
A-145-1ML	Acylcarnitines Mix 3 (C8:1,C10:1,C12:1,C14:1,C14:2,C16:1,C18:1,C18:2)	100-200 ug/mL in 90:10 methanol:1% aqueous formic acid
A-148-1ML	Acylcarnitines IS Mix 1 (C0,C2)	1.0 mg/mL each analyte in methanol
A-147-1ML	Acylcarnitines IS Mix 2 (C4,iC5,C6,C8,C10,C12,C14,C18)	100-200 µg/mL in 90:10 methanol:1% aqueous formic acid
A-176-1ML	Glutaryl-L-Carnitine	100 µg/mL in 90:10 methanol:1% aqueous formic acid
A-181-1ML	Glutaryl-L-Carnitine-(N-Methyl-D_3)	100 µg/mL in 90:10 methanol:1% aqueous formic acid
A-162-1ML	Palmitoyl-L-Carnitine	5.0 mg/mL in 90:10 methanol:dmso
A-164-1ML	Palmitoyl-L-Carnitine (N-Methyl-D_3)	500 µg/mL in 90:10 methanol:dmso
A-161-1ML	Propionyl-L-Carnitine	5.0 mg/mL in 90:10 methanol:dmso
A-163-1ML	Propionyl-L-Carnitine-(N-Methyl-D_3)	500 µg/mL in 90:10 methanol:dmso

We offer an extensive selection of high-quality analytical standards comprising of native and stable isotope-labeled acylcarnitines, suitable for qualitative and quantitative applications involving MS, GC/MS, or LC/MS in research environments and for method development. Our unique portfolio consists of all clinically relevant acylcarnitines of highest purity, including- saturated, unsaturated, dicarboxy-, 3-hydroxy-, unsaturated 3-hydroxy-, and aromatic acylcarnitines.

Acylcarnitines Analytical Standards

Product no.	Product name	ID	Pack Size
94954	L-Carnitine hydrochloride	C0	10 mg/50 mg
92107	O-Acetyl-L-carnitine hydrochloride	C2	10 mg/50 mg
91275	Propionyl-L-carnitine	C3	10 mg/50 mg
08984	Butyryl-L-carnitine	C4	10 mg/50 mg
91299	Isobutyryl-L-carnitine	iC4	10 mg/50 mg
04265	Valeryl-L-carnitine	C5	10 mg/50 mg
91403	Isovaleryl-L-carnitine	iC5	10 mg/50 mg
91388	Hexanoyl-L-carnitine	C6	10 mg/50 mg
06206	Octanoyl-L-carnitine	C8	10 mg/50 mg
91521	Decanoyl-L-carnitine	C10	10 mg/50 mg
91432	Lauroyl-L-carnitine	C12	10 mg/50 mg
91582	Myristoyl-L-carnitine	C14	10 mg/50 mg
91503	Palmitoyl-L-carnitine	C16	10 mg/50 mg
08084	Stearoyl-L-carnitine	C18	10 mg/50 mg
91519	2-Methylbutyryl-L-carnitine		10 mg/50 mg
07441	Malonyl-L-carnitine lithium salt	C3DC	10 mg/50 mg
04609	O-Succinyl-L-carnitine lithium salt	C4DC	10 mg/50 mg
91499	Glutaryl-L-carnitine lithium salt	C5DC	soon available
07481	Adipoyl-L-carnitine lithium salt	C6DC	soon available
91926	Adipoyl-L-carnitine	C6DC	10 mg/50 mg
15868	Suberoyl-L-carnitine lithium salt	C8DC	10 mg/50 mg
92070	Suberoyl-L-carnitine	C8DC	10 mg/50 mg
16329	Sebacoyl-L-carnitine lithium salt	C10DC	10 mg/50 mg
95187	Methylmalonyl-L-carnitine lithium salt		10 mg/50 mg
39588	Tigloyl-L-carnitine	C5:1	10 mg/50 mg
55184	trans-2-Octenoyl-L-carnitine	C8:1	10 mg/50 mg
56613	trans-2-Decenoyl-L-carnitine	C10:1	10 mg/50 mg
74023	trans-2-Dodecenoyl-L-carnitine	C12:1	10 mg/50 mg
40733	trans-2-Tetradecenoyl-L-carnitine	C14:1	10 mg/50 mg
41003	trans-2-Hexadecenoyl-L-carnitine	C16:1	10 mg/50 mg
19945	Oleoyl-L-carnitine	C18:1	10 mg/50 mg
79669	2-trans,4-cis-Decadienoyl-L-carnitine	C10:2	soon available
19188	cis,cis-5,8-Tetradecanedienoyl-L-carnitine	C14:2	10 mg/50 mg
76771	cis,cis-9,12-Octadecadienoyl-L-carnitine	C18:2	10 mg/50 mg
30172	Arachidonyl-L-carnitine	C20:4	10 mg/50 mg
28986	3-Methylcrotonyl-L-carnitine		10 mg/50 mg
90941	[(R)-3-Hydroxybutyryl]-L-carnitine	C4OH	soon available

Acylcarnitines Analytical Standards (continued)

Product no.	Product name	ID	Pack Size
91298	3-Hydroxyisovaleryl-L-carnitine	iC5OH	10 mg/50 mg
54253	[(3R)-3-Hydroxyhexanoyl]-L-carnitine	C6OH	10 mg/50 mg
07305	[(3R)-3-Hydroxyoctanoyl]-L-carnitine	C8OH	10 mg/50 mg
92372	[(3R)-3-Hydroxydecanoyl]-L-carnitine	C10OH	10 mg/50 mg
49853	[(3R)-3-Hydroxydodecanoyl]-L-carnitine	C12OH	10 mg/50 mg
52002	[(3R)-3-Hydroxytetradecanoyl]-L-carnitine	C14OH	10 mg/50 mg
52096	[(3R)-3-Hydroxyhexadecanoyl]-L-carnitine	C16OH	10 mg/50 mg
52413	[(3R)-3-Hydroxyoctadecanoyl]-L-carnitine	C18OH	10 mg/50 mg
52018	[(3R)-3-Hydroxy-cis-tetradec-9-enoyl]-L-carnitine	C14:1OH	1 mg/10 mg
52383	[(3R)-3-Hydroxy-cis-hexadec-9-enoyl]-L-carnitine	C16:1OH	1 mg/10 mg
51624	[(3R)-3-Hydroxy-cis-octadec-9-enoyl]-L-carnitine	C18:1OH	1 mg/10 mg
55282	[(3R)-3-Hydroxy-cis,cis-octadeca-9,12-dienoyl]-L-carnitine	C18:2OH	soon available
94558	trans-Cinnamoyl-L-carnitine	C9Ar	10 mg/50 mg

Stable isotope labeled Acylcarnitines

Product no.	Product name	ID	Pack Size
94991	L-Carnitine-(N-methyl-d_3)	C0-d_3	1 mg/5 mg/10 mg
89927	L-Carnitine-(N,N,N,-trimethyl-$^{13}C_3$)	C0-$^{13}C_3$	1 mg/10 mg
93689	L-Carnitine-(N,N,N-trimethyl-d_9)	C0-d_9	1 mg/5 mg/10 mg
52437	Acetyl-L-carnitine-(N-methyl-d_3)	C2-d_3	1 mg/5 mg/10 mg
52941	Propionyl-L-carnitine-(N-methyl-d_3)	C3-d_3	1 mg/5 mg/10 mg
53099	Butyryl-L-carnitine-(N-methyl-d_3)	C4-d_3	1 mg/5 mg/10 mg
08196	Isobutyryl-L-carnitine-(N,N,N-trimethyl-d_9)	iC4-d_9	1 mg/10 mg
08459	Valeryl-L-carnitine-(N-methyl-d_3)	C5-d_3	1 mg/10 mg
52993	Isovaleryl-L-carnitine-(N,N,N-trimethyl-d_9)	iC5-d_9	1 mg/5 mg/10 mg
94344	Hexanoyl-L-carnitine-(N-methyl-d_3)	C6-d_3	1 mg/5 mg/10 mg
53230	Octanoyl-L-carnitine-(N-methyl-d_3)	C8-d_3	1 mg/ mg/10 mg
92439	Decanoyl-L-carnitine-(N-methyl-d_3)	C10-d_3	1 mg/5 mg/10 mg
94347	Lauroyl-L-carnitine-(N,N,N-trimethyl-d_9)	C12-d_9	1 mg/5 mg/10 mg
94346	Myristoyl-L-carnitine-(N,N,N-trimethyl-d_9)	C14-d_9	1 mg/5 mg/10 mg
55107	Palmitoyl-L-carnitine-(N-methyl-d_3)	C16-d_3	1 mg/5 mg/10 mg
53156	Stearoyl-L-carnitine-(N-methyl-d_3)	C18-d_3	1 mg/5 mg/10 mg
93035	Malonyl-L-carnitine-(N-methyl-d_3) lithium salt	C3DC-d_3	1 mg/5 mg/10 mg
92523	Succinyl-L-carnitine-(N-methyl-d_3) lithium salt	C4DC-d_3	1 mg/5 mg/10 mg
95334	Glutaryl-L-carnitin-(N-methyl-d_3) lithium salt	C5DC-d_3	1 mg/5 mg/10 mg
06691	Adipoyl-L-carnitine-(N-methyl-d_3) lithium salt	C6DC-d_3	1 mg/5 mg/10 mg
92586	Suberoyl-L-carnitine-(N-methyl-d_3) lithium salt	C8DC-d_3	1 mg/5 mg/10 mg
92661	Sebacoyl-L-carnitine-(N-methyl-d_3) lithium salt	C10DC-d_3	1 mg/5 mg/10 mg
41898	Methylmalonyl-L-carnitine-(N-methyl-d_3) lithium salt		1 mg/5 mg
94355	3-Hydroxyisovaleryl-L-carnitine-(N-methyl-d_3)	iC5OH-d_3	1 mg/5 mg
78884	[(3R)-3-Hydroxydecanoyl]-L-carnitine-(N-methyl-d_3)	C10OH-d_3	1 mg/10 mg
53297	[(R)-3-Hydroxyhexadecanoyl]-L-carnitine-(methyl-d_3)	C16OH-d_3	1 mg/5 mg

Acylglycines

Acylglycines are glycine conjugates of acyl-CoA species and are formed as intermediates in the metabolism of amino acids and fatty acids. Abnormal concentrations of acylglycines are used as biomarker in the detection of selected inborn errors of metabolism such as fatty acid oxidation disorders and organic acidemias. Testing of acylglycines is often done alongside the testing of plasma acylcarnitines and urine organic acids. The analysis of urine acylglycines is particularly effective for the biochemical diagnosis of selected IEMs. It measures the target urinary metabolites present in amounts below the detection limit in routine analysis of organic acids such as medium chain acyl-CoA dehydrogenase deficiency (MCADD), isovaleric acidemia, and β-ketothiolase deficiency (BKTD).

Our portfolio offers a comprehensive range of analytical standards consisting of native acylglycines, suitable for qualitative use and for method development in newborn screening and detection of metabolic diseases involving MS, GC/MS, or LC/MS.

Our portfolio consists of clinically relevant acylglycines, including saturated, unsaturated, dicarboxy-, and aromatic acylglycines of highest quality.

Acylglycines Analytical Standards

Product no.	Product name	ID	Pack Size
69563	N-Acetylglycine	C2	10 mg/50 mg
94274	N-Butyrylglycine	C4	10 mg/50 mg/250 mg
04513	N-Isobutyrylglycine	iC4	10 mg/50 mg/250 mg
04268	N-Valerylglycine	C5	10 mg/50 mg/250 mg
72672	N-Isovalerylglycine	iC5	10 mg/50 mg
93539	N-Heptanoylglycine	C7	10 mg/50 mg/250 mg
93529	N-Octanoylglycine	C8	10 mg/50 mg/250 mg
93204	N-Decanoylglycine	C10	10 mg/50 mg/250 mg
93586	N-Lauroylglycine	C12	10 mg/50 mg/250 mg
00970	N-Myristoylglycine	C14	10 mg/50 mg/250 mg
00886	N-Palmitoylglycine	C16	available soon
54364	N-Stearoylglycine	C18	10 mg/50 mg/250 mg
53461	N-Valproylglycine		10 mg/50 mg/250 mg
04478	N-(2-Methylbutyryl)glycine		10 mg/50 mg/250 mg
03535	N-Pivaloylglycine		10 mg/50 mg/250 mg
53438	N-Malonylglycine	C3DC	10 mg/50 mg/250 mg
55139	(±)-N-Methylmalonylglycin	3-MeC3DC	10 mg/50 mg/250 mg
01883	N-Succinylglycine	C4DC	10 mg/50 mg/250 mg
53795	N-Glutarylglycine	C5DC	10 mg/50 mg/250 mg
53603	N-Adipoylglycin	C6DC	10 mg/50 mg/250 mg
52888	N-Suberylglycine	C8DC	10 mg/50 mg/250 mg
52851	N-Sebacoylglycine	C10DC	10 mg/50 mg/250 mg
72656	N-Tigloylglycine	C5:1	10 mg/50 mg
69685	N-(3-Methylcrotonyl)glycine		10 mg/50 mg
96408	N-(2-Phenylacetyl)glycine	C2Ar	10 mg/50 mg/250 mg
94348	N-(3-Phenylpropionyl)glycine	C3Ar	10 mg/50 mg/250 mg
98956	N-Cinnamoylglycine	C9Ar	10 mg/50 mg/250 mg
03417	N-[2-(4-Hydroxyphenyl)acetyl]glycine		10 mg/50 mg/250 mg

Amino Acids

Amino acids are the building blocks of proteins occurring throughout the body. Amino acid disorders, also called as aminoacidopathies, are a group of inborn errors of metabolic diseases caused by the inherited defects in amino acid metabolic pathways. Many inborn errors in amino acid metabolism, such as phenylketonuria, tyrosinemia, citrullinemia, non-ketotic hyperglycinemia, maple syrup urine disease, and homocystinuria have been identified to either cause accumulation or deficiency of one or more amino acids in the biological fluids, causing the different clinical presentations of the specific amino acid disorders. In addition, analysis of amino acids in plasma may be clinically important to the evaluation of several acquired conditions such as endocrine disorders, liver diseases, muscle diseases, neoplastic diseases, neurological disorders, nutritional disturbances, renal failure, and burns. The amino acid analysis required for the diagnosis of a variety of inborn errors of metabolism and metabolic diseases, is routinely performed in diagnostic testing and newborn screening programs. We offer a broad selection of analytical standards and certified reference materials (CRMs) of amino acids including their native, stable isotope-labeled internal standards, and solution-based multi-analyte mixes suitable for use in newborn screening and detection of metabolic diseases by GC/MS or LC/MS.

We offer TraceCERT® certified reference materials of routinely tested amino acids in both native and stable isotope-labeled formats, as single solution and multi-component solution mixes. Our products are suitable for several critical quantitative applications in research and clinical environments such as:

- Preparation of internal standards, controls, calibrators, and linearity standards for LC-MS/MS assays
- As system tuning/suitability standards
- Linearity verification
- Internal proficiency testing

The stable and quantitative solution format provides significant benefits to the user such as the convenience of use, material savings, and increased efficiency of labor. Certified concentration values for each component provides increased consistency and efficiency from less time remaking pools due to dilution errors or rerunning samples.

Amino Acids Certified Reference Materials

Product no.	Description	Pack Size
79248	Amino Acids Mix Solution certified reference material, TraceCERT®	17 AAs at 2500 nmol/mL in 0.1M HCl
96378	Stable Isotope Labeled Amino Acid Mix Solution 1 certified reference material, TraceCERT®	17 SIL AAs at 1250-2500 nmol/mL in 0.1M HCl
01428	Stable Isotope Labeled Amino Acid Mix Solution 2 certified reference material, TraceCERT®	11 SIL AAs at 500-2500 nmol/mL in 0.1M HCl
96072	Stable Isotope Labeled Amino Acid Mix 3 2500 nmol certified reference material, TraceCERT®	available soon
44526	L-Alanine certified reference material, TraceCERT®	100 mg
43811	γ-Aminobutyric acid certified reference material, TraceCERT®	50 mg
90538	L-Arginine monohydrochloride certified reference material, TraceCERT®	100 mg
51363	L-Asparagine, certified reference material, TraceCERT®	100 mg

Amino Acids Certified Reference Materials (continued)

Product no.	Description	Pack Size
51572	L-Aspartic acid certified reference material, TraceCERT®	100 mg
95437	L-Cysteine certified reference material, TraceCERT®	100 mg
49603	L-Cystine certified reference material, TraceCERT®	100 mg
72816	3,4-Dihydroxy-L-phenylalanine certified reference material, TraceCERT®	50 mg
95436	L-Glutamic acid certified reference material, TraceCERT®	100 mg
76523	L-Glutamine certified reference material, TraceCERT®	100 mg
73767	L-Histidine certified reference material, TraceCERT®	100 mg
56241	L-Isoleucine certified reference material, TraceCERT®	100 mg
76526	L-Leucine certified reference material, TraceCERT®	100 mg
67448	L-Lysine monohydrochloride certified reference material, TraceCERT®	100 mg
39496	L-Methionine certified reference material, TraceCERT®	100 mg
91016	L-Methionine sulfoximine certified reference material, TraceCERT®	100 mg
40541	L-Phenylalanine certified reference material, TraceCERT®	100 mg
93693	L-Proline certified reference material, TraceCERT®	100 mg
54763	L-Serine certified reference material, TraceCERT®	100 mg
61506	L-Threonine certified reference material, TraceCERT®	100 mg
51145	L-Tryptophan certified reference material, TraceCERT®	100 mg
91515	L-Tyrosine certified reference material, TraceCERT®	100 mg
50848	L-Valine certified reference material, TraceCERT®	100 mg

We also offer a wide selection of high-quality analytical standards of amino acids, suitable for qualitative and quantitative applications involving GC/MS or LC/MS in research environments and for method development. Our portfolio consists of clinically relevant amino acids of highest purity.

Amino Acids Analytical Standards

Product no.	Description	Pack Size
AAS18	Amino Acid Standard	17 AAs at 1.25-2.5 µmoles per mL in 0.1N HCl
A9781	Amino acid standards for protein hydrolysates	18 AAs at 0.25-0.5 µmole/mL in 0.2N sodium citrate, pH 2.2
A2908	Amino acid standards for protein hydrolysates containing norleucine	Amino acids at 0.25-0.5 µmole/ml in 0.2N sodium citrate, pH 2.2
A9906	Amino acid standards, physiological analytical standard, acidics, neutrals, and basics	38 component mix
A6407	Amino acid standards, physiological analytical standard, acidics and neutrals	Amino acids at 1.25-2.5 µmole/ml
A6282	Amino acid standards, physiological analytical standard, basics	Amino acids and related compounds at 2.5 µmole/mL
41144	β-Alanine	100 mg
55329	L-Alanine hydrochloride solution	100 mM amino acid in 0.1M HCl
08163	L-Arginine hydrochloride solution	100 mM amino acid in 0.1M HCl
57579	L-Cystine hydrochloride solution	10 mM amino acid in 0.1M HCl
55097	Glycine hydrochloride solution	100 mM amino acid in 0.1M HCl
43011	L-Histidine hydrochloride solution	100 mM amino acid in 0.1M HCl
44925	DL-Homocysteine	25 mg
50271	L-Isoleucine hydrochloride solution	100 mM amino acid in 0.1M HCl
80687	L-Leucine hydrochloride solution	100 mM amino acid in 0.1M HCl
23128	L-Lysine	100 mg
44208	L-Lysine hydrochloride solution	100 mM amino acid in 0.1M HCl
50272	L-Methionine hydrochloride solution	100 mM amino acid in 0.1M HCl
53721	DL-Norvaline	100 mg
91331	L-Phenylalanine	25 g
44026	L-Phenylalanine hydrochloride solution	100 mM amino acid in 0.1M HCl
94321	L-Proline hydrochloride solution	100 mM amino acid in 0.1M HCl

Fatty Acids

Free fatty acids (FFAs), also called nonesterified fatty acids (NEFA), occur as albumin-bound and account for 2% to 5% of all the serum fatty acids circulating in the blood plasma. Most of the free fatty acids are derived from either dietary sources or are mobilized from adipose tissues. The free fatty acids found in plasma are elevated in many obesity-related disorders and may contribute to insulin resistance in peripheral tissues. A knowledge of the individual levels of FFAs in blood plasma can be helpful in the diagnosis and management of certain diseases and disorders of metabolism such as uncontrolled diabetes mellitus, hyperlipoproteinemia, secondary hyperlipoproteinemia, and conditions such as sepsis and lipoactive hormone producing tumors.

We offer analytical standards and CRMs of fatty acids suitable for use in qualitative and quantitative testing applications involving GC/MS or LC/MS. Our portfolio comprises of clinically relevant free fatty acids of highest quality.

Fatty Acids Certified Reference Material

Product #	Description	ID	Pack Size
07754	Octanoic acid	C8:0	50mg
43051	Palmitic acid	C16:0	100mg
76137	Stearic acid	C18:0	100mg
L-039	α-Linolenic Acid	C18:3w3	1.0 mg/mL in ethanol
A-122	Arachidonic acid	C20:4w6	1.0 mg/mL in ethanol
E-113	Eicosapentaenoic acid (EPA)	C20:5w3	500 µg/mL in ethanol
D-120	Docosapentaenoic acid (DPA), C22:5w3	C22:5w3	500 µg/mL in ethanol
D-117	Docosahexaenoic acid (DHA)	C22:6w3	500 µg/mL in ethanol

Fatty Acids Analytical Standard

Product #	Description	ID	Pack Size
21639	Octanoic acid	C8:0	5 mL
21409	Decanoic acid	C10:0	5 g
69638	cis-2-Decenoic acid (Decenoic acid)	C10:1	10 mg/100 mg
61609	Lauric acid	C12:0	5 g
73079	11-Dodecenoic acid (Lauroleic acid)	C12:1	10 mg/100 mg
70079	Myristic acid	C14:0	5 g
41788	Myristoleic acid	C14:1	100 mg
69573	cis,cis-5,8-Tetradecadienoic acid	C14:2	10 mg/100 mg
72691	Pristanic acid (2,6,10,14-Tetramethylpentadecanoic acid)	C15:0(CH3)4	soon available
76119	Palmitic acid	C16:0	5 g
43306	Phytanic acid (3,7,11,15-Tetramethylhexadecanoic acid)	C16:0(CH3)4	soon available
76169	Palmitoleic acid	C16:1w7	1 g
43284	cis-7-Hexadecenoic acid (Hypogeic acid)	C16:1w9	10 mg/100 mg
43067	cis,cis-9,12-Hexadecadienoic acid (Palmitolinoleic acid)	C16:2	soon available

Fatty Acids Analytical Standard (continued)

Product #	Description	ID	Pack Size
21639	Octanoic acid	C8:0	5 mL
85679	Stearic acid	C18:0	500 mg/5 g
69301	cis-Vaccenic acid	C18:1w7	10 mg/100 mg
75090	Oleic acid	C18:1w9	5 mL/25 mL
62230	Linoleic acid	C18:2w6	5 mL/25 mL
62160	α-Linolenic Acid	C18:3w3	1 mL/5 mL
62174	γ-Linolenic acid	C18:3w6	100 mg/500 mg
39383	Arachidic acid	C20:0	100 mg
69154	Homo-γ-linolenic acid	C20:3w6	50 mg
43059	Mead acid (cis,cis,cis-5,8,11-Eicosatrienoic acid)	C20:3w9	soon available
23401	Arachidonic acid	C20:4w6	50 mg
44864	Eicosapentaenoic acid (EPA)	C20:5w3	100 mg/500 mg
11909	Docosanoic acid	C22:0	100 mg/5 g
45629	Docosenoic acid	C22:1	1 g/5 g
43044	all-cis-7,10,13,16-Docosatetraenoic acid (DTA)	C22:4w6	soon available
43002	all-cis-7,10,13,16,19-Docosapentaenoic acid (DPA)	C22:5w3	soon available
18566	Docosapentaenoic acid (DPA)	C22:5w6	10 mg
53171	Docosahexaenoic acid (DHA)	C22:6w3	10 mg
42893	Tetracosanoic acid	C24:0	10 mg/100 mg
87117	Nervonic acid	C24:1w9	100 mg
06922	Hexacosanoic acid	C26:0	10 mg
72556	cis-17-Hexacosenoic acid	C26:1	soon available

Organic Acids

Organic acids occur as physiologic intermediates in a variety of metabolic pathways. Organic acid disorders are a group of inherited metabolic conditions caused by the deficiency of a specific enzyme and resulting in the accumulation of organic acids in blood and urine. More than 65 specific organic acids affecting these pathways have been identified. The accumulated compounds or their metabolites are of toxic nature and are responsible for the clinical presentations of these disorders. Diagnostic specificity of inborn errors of metabolism via organic acids analysis is routinely performed in diagnostic tests and newborn screening programs.

Our analytical products offer high-quality analytical standards and CRMs of organic acids including native and stable isotope-labeled internal standards suitable for the preparation of calibrators, controls, and internal standards for use in the detection of inborn errors of metabolism by GC/MS or LC/MS.

Organic Acids Certified Reference Material

Product #	Potential New Products	Pack Size
M-080	Methylmalonic acid	1.0 mg/mL in acetonitrile
M-105-1ML	Methyl-D_3-malonic acid	1.0 mg/mL in acetonitrile
M-173-1ML	Methylmalonic acid-$^{13}C_4$	100 µg/mL in acetonitrile
V-006	Valproic acid	1.0 mg/mL in methanol
V-029	Valproic acid-D_6	1.0 mg/mL in methanol

Organic Acids Analytical Standard

Product #	Potential New Products	Pack Size
89143	Adipic acid	100 mg
52458	α-Aminoadipic acid (DL-2-Aminoadipic acid)	10 mg/50 mg
55398	α-Aminoadipic acid (L-2-Aminoadipic acid)	10 mg/50 mg
03835	γ-Aminobutyric acid	250 mg
43811	γ-Aminobutyric acid	50 mg
67338	α-Aminobutyric acid (DL-2-Aminobutyric acid)	10 mg/50 mg
43954	β-Aminoisobutyric acid (L-3-Aminoisobutyric acid)	10 mg/50 mg
95148	L-Argininosuccinic acid lithium salt	10 mg/50 mg
51572	Aspartic acid	100 mg
07596	Butyric acid	100 mg
08089	Butyric acid	100 mL
19215	Butyric acid	5 mL
90833	3-Carboxy-4-methyl-5-propyl-2-furanpropionic acid	10 mg/50 mg
46933	Citric acid	500 mg
94676	Citric acid	100 mg
01949	3-Dehydroquinic acid lithium salt	10 mg/50 mg
05283	3-Deoxy-2-keto-6-phosphogluconic acid lithium salt	10 mg/50 mg

Product #	Potential New Products	Pack Size
89143	Adipic acid	100 mg
19367	3-Deoxy-D-arabino-hexonic acid lithium salt	10 mg/50 mg
90061	4-Deoxy-D-erythronic acid	10 mg/50 mg
06932	4-Deoxy-L-erythronic acid	10 mg/50 mg
91423	N^{1},N^{12}-Diacetylspermine hydrochloride	50 mg/100 mg
89783	(2R,3S)-2,3-dihydroxybutanoic acid	10 mg/50 mg
89921	2,4-Dihydroxybutanoic acid	10 mg/50 mg
90948	(2S,3R)-2,3-dihydroxybutanoic acid	10 mg/50 mg
61932	DL-3,3-Dimethylmalic acid	10 mg/50 mg
07303	(2RS)-2-Ethyl-2-methylpentanoic acid	50 mg
69474	3-Ethylmalic acid	10 mg/50 mg
43975	Ethylmalonic acid	10 mg/50 mg
52229	Ferulic acid	50 mg
90034	Ferulic acid	100 mg
76635	Fumaric acid	100 mg
00251	Fumarylacetone	10 mg/50 mg
53439	Glutaconic acid	10 mg/50 mg
89147	Glutaric acid	100 mg
00380	Glutaric semialdehyde	10 mg/50 mg
59969	L-Glyceric acid sodium salt	10 mg/50 mg
68069	Hippuric acid	10 mg/50 mg
93739	Hippuric acid-(phenyl-$^{13}C_6$)	10 mg/50 mg
67862	Homogentisic acid	10 mg/50 mg
69673	Homovanillic acid	25 mg
69433	4-hydroxy-3,3-dimethyl-2-oxo-butanoic acid (2-Dehydropantoic acid)	10 mg/50 mg
61076	DL-4-Hydroxy-2-ketoglutaric acid lithium salt	10 mg/50 mg
80529	2-Hydroxy-3-methylvaleric acid, mixture of diastereomers	10 mg/50 mg
92826	2-Hydroxyadipic acid	10 mg/50 mg
55397	(R)-3-Hydroxybutyric acid	10 mg/50 mg
04725	3-Hydroxyglutaric acid	10 mg/50 mg
61382	D-α-Hydroxyglutaric acid disodium salt (D-2-Hydroxyglutaric acid /D-α-Hydroxyglutaric acid)	10 mg/50 mg
61313	L-α-Hydroxyglutaric acid disodium salt (L-2-Hydroxyglutaric acid / L-α-Hydroxyglutaric acid)	10 mg/50 mg
36106	D-α-Hydroxyglutaric acid-$^{13}C_5$ disodium salt ([$^{13}C_5$]-D-2-Hydroxyglutaric acid)	1 mg/10 mg
80559	L-α-Hydroxyglutaric acid-$^{13}C_5$ disodium salt ([$^{13}C_5$]-L-2-Hydroxyglutaric acid)	1 mg/10 mg
55697	5-Hydroxyindole-3-acetic acid (5HIAA)	10 mg/50 mg
55188	Sodium (R)-β-hydroxyisobutyrate	10 mg/50 mg
55578	4-Hydroxyisovaleric acid	10 mg/50 mg
96045	2-(Hydroxymethyl)butyric acid sodium salt	10 mg/50 mg
00537	3-(4-Hydroxyphenyl)-DL-lactic acid-$^{13}C_9$	1 mg/10 mg
91779	3-(3-Hydroxyphenyl)propionic acid	10 mg/50 mg/250 mg
91378	(2RS)-2-(4-Hydroxyphenyl)propionic acid	10 mg/50 mg/250 mg
06704	3-(3-Hydroxyphenyl)-3-hydroxypropanoic acid	10 mg/50 mg
55206	4-Hydroxyphenylacetic acid	10 mg/50 mg
07491	4-Hydroxyphenylpyruvic acid IS (4-Hydroxyphenylpyruvic acid-$^{13}C_9$)	1 mg/10 mg
78651	Isovaleric acid	1 mL/5 mL
74194	2-Keto-3-deoxy-D-galactonic acid lithium salt	10 mg
61234	α-Ketoglutaric acid / 2-Ketoglutaric acid	10 mg/50 mg
67667	Kynurenic acid	10 mg/50 mg

Product #	Potential New Products	Pack Size
89143	Adipic acid	100 mg
67653	L-Kynurenine	10 mg/50 mg
46937	L-(+)-Lactic acid	100 mg
92816	Maleic acid	1 g
46940-U	Malic acid	100 mg
68714	Malonic acid	100 mg
06997	(R) 2-Methyl-3-ureidopropionic acid	10 mg/50 mg
42641	(Z)-3-Methylglutaconic acid	10 mg/50 mg/250 mg
44108	(E)-3-Methylglutaconic acid	10 mg/50 mg/250 mg
53873	3-Methylglutaconic acid	10 mg/50 mg
74626	(E)-2-Methylglutaconic acid	10 mg/50 mg
75606	(Z)-2-Methylglutaconic acid	10 mg/50 mg
49954	3-Methylglutaric acid-$^{13}C_4$ (3-Methyl-^{13}C-glutaric acid-1,2,3-$^{13}C_3$)	1 mg/5 mg
92988	(2RS,3SR)-2-Methylisocitric acid	10 mg/50 mg
69369	3-Methylmalic acid	10 mg/50 mg
18802	2-Methyl-trans-aconic acid	10 mg/50 mg
42147	(RS)-Mevalonic acid	5 mg
43987	(S)-Mevalonic acid lithium salt	10 mg/50 mg
89163	(±)-Mevalonic acid-d_3	1 mg/10 mg
43536	trans,trans-Muconic acid-$^{13}C_6$	1 mg/10 mg
07754	Octanoic acid	50 mg
21639	Octanoic acid	5 mL
92675	trans-2-Octenedioic acid (2-Octene-1,8-dioic acid)	10 mg/50 mg
57197	L-Ornithine	100 mg
50899	2-Oxoadipic acid	10 mg/50 mg
74346	Phenylpyruvic acid	100 mg
94425	Propionic acid	1 mL/5 mL
69499	3-Propylmalic acid	10 mg/50 mg
93693	Pyroglutamic acid; L-Proline	100 mg
49893	Succinic acid	100 mg
39584	3-Sulfopyruvic acid	10 mg/50 mg
73829	(±)-Ureidoglycolic acid sodium salt	10 mg/50 mg/250 mg
74005	DL-3-Ureidoisobutyric acid	10 mg/50 mg
69301	cis-Vaccenic acid	10 mg/100 mg
89127	DL-Vanillactic acid	10 mg/50 mg/250 mg
68654	Vanillic acid	50 mg
73538	Vanillylmandelic acid	25 mg

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