The Single Dose Amino Acid Handbook

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Introduction

This handbook is intended for healthcare professionals only. It is intended to provide useful product and clinical usage information on Vitaflo's range of Single Dose Amino Acids (SDAAs) in the form of individual fact files. Where available, this handbook directs the reader to further information in the relevant clinical guidelines.

The handbook is not intended for use by parents, caregivers or patients themselves. It is for general information only and must not be used as a substitute for professional medical advice.

Background

Dietary supplementation with individual amino acids is used in a wide range of disorders, particularly disorders of protein metabolism where restrictive therapeutic diets limit natural protein intake and potentially increase the risk of an essential amino acid deficiency.

In these disorder types, the quantities of amino acids supplemented are often small and impractical to weigh out accurately on domestic weighing scales. When this is the case, hospital or local pharmacies may be required to prepare 'doses' for patients, a practice which is inconvenient, time-consuming and, even in experienced hands, potentially prone to error.

Vitaflo's Single Dose Amino Acid (SDAA) range

In 2007 Vitaflo launched its SDAA range. Covering a wide range of amino acids, we spoke to global experts and asked them what quantities they typically used with their patients. We then pre-measured those quantities and placed them in sachets, on a carbohydrate base.

For many patients, families and caregivers, this eliminates the need to weigh out amino acids and thus brings convenience, reassurance and a reduced need to collect products from their pharmacy, all of which may support adherence. Additionally, the use of pre-measured sachets has been shown to decrease preparation error, compared to using scoops or weighing scales[1], which may, in the context of SDAAs, help reduce the risk of potentially serious dosage errors.

1. Gokmen-Ozel H, Daly A, Davies P, Chahal S, MacDonald A. Errors in Emergency Feeds in Inherited Metabolic Disorders: a Randomised Controlled Trial of Three Preparation Methods. Archives of Disease in Childhood. 2010; 95(10):776-80. DOI: 10.1136/adc.2010.161711.

Colour coding and naming system

The branding and the naming of each SDAA is clear and simple

The name indicates the **amino acid** and **quantity**, in milligrams (mg), per pre-measured sachet.

Phenylalanine50 provides **50mg of L-phenylalanine** per sachet

Tyrosine1000 provides **1000mg of L-tyrosine** per sachet

Distinct colour coding for safety

Each SDAA has a distinct colour, to help ensure patients consume the right product.

Red is the main colour used for our **Arginine** products.

Yellow is the main colour used for **Glycine**.

SDAAs with similar sounding names are branded with distinctively different colours.

For example: Citrulline, Cystine and Creatine are orange, grey and blue respectively.

SDAAs which could be harmful if consumed for specific disorders are also given distinctively different packaging colours to those likely to be used in those specific disorders.

For example: Leucine, which must be avoided in Maple Syrup Urine Disease (MSUD), is branded white, whereas Isoleucine, which is regularly supplemented in MSUD, is branded black.

Safety Message

Extra measures have been put in place to ensure patients use the correct SDAA and dose every time.

A safety message, reminding patients to check the product name and dose before consuming, can be found on the inside of the lid.

Mixing Instructions

Each amino acid fact file provides bespoke instructions for preparing the relevant amino acid with water. Where we provide more than one dose of an amino acid (e.g. Arginine500, Arginine2000 and Arginine5000), different mixing instructions are required for the different doses in some cases.

Generic instructions for mixing the SDAA products with both powder and liquid protein substitutes, plus flavouring options, are provided at the back of this handbook.

Important Notice

SDAAs are Food for Special Medical Purposes and must only be given to patients with proven inborn errors of metabolism under strict medical supervision.

Arginine5000, **Creatine5000**, **Cystine500** and **Tyrosine1000** are suitable from 3 years of age onwards.

All other SDAAs are suitable from birth. Children under 12 months, when made into solution or added to infant formula, use immediately and discard any remaining product within 1 hour.

SDAAs are not for use as a sole source of nutrition.

SDAAs are for enteral use only.

Arginine

Product range	Arginine500	Arginine2000	Arginine5000
Pack size	30 x 4g e	30 x 4g e	30 x 7.6g e
Ingredients	Dried Glucose Syrup, L-arginine.	Maltodextrin, L-arginine.	L-arginine, Acidity regulator (E296), Flavouring, Artificial sweetener (Sucralose), Colour (E160a).

Description

Powdered, pre-measured L-arginine amino acid supplements on a carbohydrate base.

Arginine500 and **Arginine2000** are unflavoured.

Arginine5000 is orange flavoured and contains sweetener.

Indication

Arginine500, **2000** and **5000** are used in the dietary management of conditions requiring arginine supplementation, such as urea cycle disorders (UCD).

Arginine500 and **Arginine2000** are suitable from birth.

Arginine5000 is suitable from 3 years of age onwards.

Clinical use

Urea Cycle Disorders[1] (UCD)

UCD is an umbrella term encompassing six genetic disorders of protein metabolism, all of which result in impairment of the body's ability to remove waste nitrogen. UCD can have acute, chronic and intermittent clinical manifestations occurring at any age. Their clinical hallmark is the hyperammonaemia crisis.

Due to impaired synthesis, L-arginine becomes an essential amino acid in all of the UCDs, with the exception of Arginase 1 deficiency. It must therefore be supplemented as either L-arginine or its precursor, L-citrulline.

Pyridoxine Dependent Epilepsy[2] (PDE)

Pyridoxine-dependent epilepsy (PDE-ALDH7A1) is an autosomal recessive condition due to a deficiency of α-aminoadipic semialdehyde dehydrogenase, which is a key enzyme in L-lysine oxidation.

Although pyridoxine monotherapy is often effective in reducing seizure activity, most patients experience developmental delay and intellectual disability. Recent guidelines recommend 'triple therapy'; pyridoxine hydrochloride, a lysine-restricted diet, and supplementing the diet with L-arginine.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4

Arginine500 & Arginine2000

For both **Arginine500** and **Arginine2000**, wait 5 minutes*, shake again and serve.

Arginine5000

Arginine5000 is now ready to serve. Water or permitted drinks should be consumed after **Arginine5000**.

* The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Arginine500, **Arginine2000** & **Arginine5000** can be used in conjunction with other Vitaflo products for the dietary management of UCD:

Further reading

1. Häberle J, Burlina A, Chakrapani A, Dixon M, Karall D, Lindner M et al. Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision. Journal of Inherited Metabolic Disease. 2019:42 (6); 1192-1230. DOI:10.1002/jimd.12100

2. Coughlin CR II, Tseng LA, Abdenur JE, Ashmore C, Boemers F, Bok LA, et al. Consensus guidelines for the diagnosis and management of pyridoxine-dependent epilepsy due to α-aminoadipic semialdehyde dehydrogenase deficiency. Journal of Inherited Metabolic Disease. 2021;44 (1):178–192. DOI: 10.1002/jimd.12332

Arginine500, Arginine2000 and Arginine5000 are Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
For use under medical supervision. For healthcare professional use only.

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Citrulline

Product range	Citrulline200	Citrulline1000
Pack size	30 x 4g e	30 x 4g e
Ingredients	Maltodextrin, L-citrulline.	Maltodextrin, L-citrulline.

Description

Powdered, unflavoured, pre-measured L-citrulline amino acid supplements on a carbohydrate base.

Indication

Citrulline200 and **Citrulline1000** are for the dietary management of inborn errors of amino acid metabolism such as Urea Cycle Disorders (UCD), suitable from birth.

Clinical use

Urea Cycle Disorders[1] (UCD)

UCDs is an umbrella term encompassing six genetic disorders of protein metabolism, all of which result in impairment of the body's ability to remove waste nitrogen. UCD can have acute, chronic and intermittent clinical manifestations occurring at any age. Their clinical hallmark is hyperammonaemia crises.

Due to impaired synthesis, L-arginine becomes an essential amino acid in all of the UCDs, with the exception of Arginase 1 deficiency. It must therefore be supplemented as either L-arginine or its precursor, L-citrulline. In some of the UCDs, low plasma levels of L-citrulline are found. Supplementing the diet with L-citrulline will help manage both low L-citrulline and low L-arginine levels.

Preparation guidelines

Step 1 Measure out 100ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4

Citrulline200

Wait 5 minutes*, shake again and serve.

Citrulline1000

Wait 10 minutes*, shake again and serve.

* The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Citrulline200 & **Citrulline1000** can be used in conjunction with other Vitaflo products for the dietary management of UCD:

Further reading

1. Häberle J, Burlina A, Chakrapani A, Dixon M, Karall D, Lindner M et al. Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision. Journal of Inherited Metabolic Disease. 2019:42 (6); 1192-1230. DOI:10.1002/jimd.12100.

Citulline200 and Citrulline1000 are Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Creatine

Description

A powdered, unflavoured, pre-measured L-creatine amino acid supplement on a carbohydrate base.

Product range	**Creatine5000**
Pack size	**30 x 6g** e
Ingredients	Creatine monohydrate, Maltodextrin, Thickener (E415).

Indication

Creatine5000 is for the dietary management of inborn errors of amino acid metabolism, suitable from 3 years of age onwards.

Clinical use

Cerebral creatine deficiency syndrome (CCDS)[1]

CCDS is a group of inborn errors of L-creatine metabolism. Individuals with these disorders experience global developmental delay, intellectual disability and associated behavioural and neurological manifestations such as seizures, movement disorders and myopathy.

Within CCDS, there are two disorders of L-creatine biosynthesis; guanidinoacetate methyltransferase (GAMT) deficiency and L-arginine:glycine amidinotransferase (AGAT) deficiency. The diet is supplemented with oral creatine monohydrate to replenish L-creatine levels in the brain and other tissues in individuals with GAMT and AGAT.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds and serve. Water or permitted drinks should be consumed after **Creatine5000**.

For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Further reading

1. Cerebral Creatine Deficiency Syndromes. https://rarediseases.org/rare-diseases/cerebral-creatine-deficiency-syndromes/#:~:text=Cerebral%20creatine%20deficiency%20syndromes%20(CCDS,all%20cells%20in%20the%20body. Accessed March 2022.

Creatine5000 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Cystine

Product range	**Cystine500**
Pack size	**30 x 4g** e
Ingredients	Dried glucose syrup, L-cystine, Thickener (E415).

Description

A powdered, unflavoured, pre-measured L-cystine amino acid supplement on a carbohydrate base.

Indication

Cystine500 is for the dietary management of inborn errors of amino acid metabolism, such as Homocystinuria (HCU), suitable from 3 years of age onwards.

Clinical use

Homocystinuria (HCU)[1]

HCU, also known as cystathionine beta-synthase (CBS) deficiency, is a rare inherited disorder in the methionine catabolic pathway. Patients with severe HCU usually present in childhood with ectopia lentis, learning difficulties and skeletal abnormalities, whilst those with mild disease tend to present as adults with thromboembolism.

HCU results in raised levels of plasma methionine, homocysteine and other sulphur-containing metabolites, and low levels of plasma cysteine and cystathionine. Cysteine is an essential amino acid in HCU, and low concentrations may contribute to the pathogenesis.

Case reports suggest that cysteine deficiency can cause poor weight gain and growth, even in the presence of adequate energy intake. Protein substitutes used in the dietary management of HCU are enriched with L-cystine (a cysteine dimer), so additional supplementation is not usually necessary. In patients with severe deficiency, L-cystine may be added to the diet.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds and serve.

For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Cystine500 can be used in conjunction with other Vitaflo products for the dietary management of HCU:

Further reading

1. Morris AA, Kožich V, Santra S, Andria G, Ben-Omran TI, Chakrapani AB, et al. Guidelines for the diagnosis and management of cystathionine beta-synthase deficiency. Journal of Inherited Metabolic Disease. 2017;40(1):49-74.

Cystine500 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Glycine

Description

A powdered, unflavoured, pre-measured glycine amino acid supplement on a carbohydrate base.

Product range	**Glycine500**
Pack size	**30 x 4g** e
Ingredients	Maltodextrin, Glycine.

Indication

Glycine500 is for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Serine deficiency disorders[1]

Serine deficiency disorders are neurometabolic disorders caused by a defect in one of the three synthesizing enzymes of the L-serine biosynthesis pathway. Congenital microcephaly, seizures and severe psychomotor retardation are all symptoms of serine deficiency. As glycine can be synthesized directly from L-serine and vice versa, L-serine deficiency disorders tend to also present with low or low to normal glycine concentrations.

Supplementing the diet with L-serine and glycine can help address potential deficiencies and may improve clinical outcomes.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds and serve.

For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Further reading

1. De Koning TJ. Treatment with amino acids in serine deficiency disorders. Journal of Inherited Metabolic Disease 2006:29(2-3); 347-351. https://doi.org/10.1007/s10545-006-0269-0

Glycine500 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Isoleucine

Product range	Isoleucine50	Isoleucine1000
Pack size	30 x 4g e	30 x 4g e
Ingredients	Dried glucose syrup, L-isoleucine.	Dried glucose syrup, L-isoleucine.

Description

Powdered, unflavoured, pre-measured L-isoleucine amino acid supplements on a carbohydrate base.

Indication

Isoleucine50 and **Isoleucine1000** are for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Maple syrup urine disease (MSUD)[1,2]

MSUD is an inherited metabolic disorder caused by branched-chain α-ketoacid dehydrogenase (BCKD) deficiency, resulting in the accumulation of branched chain amino acids (BCAAs) L-leucine, L-isoleucine and L-valine. MSUD varies considerably in disease severity, with clinical features including seizures, ketoacidosis, hypoglycaemia, apnoea, ataxia and coma.

Elevated L-leucine levels are toxic and associated with abnormal brain morphology and cognitive impairment. This may impact on social and psychomotor function. The mainstay of dietary management is the restriction of dietary BCAAs, especially L-leucine, and the use of BCAA-free protein substitutes (PS).

To promote anabolism of L-leucine, when L-leucine blood concentrations are high, additional supplementation with L-isoleucine may be required. In addition, patients on dietary management can develop an L-isoleucine deficiency, which is absent from their PS. This can lead to a rash and can also involve the eye and gut epithelium. Supplementation of the diet with L-isoleucine supplements is used to manage this.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4

Isoleucine50

Isoleucine50 is now ready to serve.

Isoleucine1000

For **Isoleucine1000,** wait 10 minutes*, shake again and serve.

* The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Isoleucine50 & **Isoleucine1000** can be used in conjunction with other Vitaflo products for the dietary management of MSUD:

Further reading

1. Frazier DM, Allgeier C, Homer C, Marriage BJ, Ogata B, Rohr F, et al. Nutrition management guideline for maple syrup urine disease: an evidence-and consensus-based approach. Molecular Genetics and Metabolism. 2014;112(3):210-7.
2. Hollak CE, Lachmann R. Inherited Metabolic Disease in Adults: A Clinical Guide: Oxford University Press; 2016.

Isoleucine50 and Isoleucine1000 are Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Leucine

Description
A powdered, unflavoured, pre-measured L-leucine amino acid supplement on a carbohydrate base.

Product range	**Leucine100**
Pack size	**30 x 4g** e
Ingredients	Dried glucose syrup, L-leucine.

Indication
Leucine100 is for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Leucine deficiency[1]

Many disorders of protein metabolism are managed through the restriction of natural protein. Such restrictions can increase the risk of essential amino acid deficiency, particularly if one or more of the following factors apply:

- Tolerance for natural protein is low.
- Due to feeding difficulties, lack of appetite or aversion, the patient consumes even less natural protein than recommended.
- The biological value of the natural protein consumed is low.
- Limited or no use of protein substitutes (for example, in some patients with organic acidaemias) or essential amino acid supplements (for example, in some patients with UCD).

Ongoing monitoring of plasma amino acids is required in patients following low protein diets as part of their dietary management. L-leucine is an essential amino acid; it should be supplemented in the case of an L-leucine deficiency, where other methods of increasing L-leucine intake (e.g. increasing natural protein) are not feasible.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4 Wait 5 minutes*, shake again and serve.

*The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Further reading

1. Dixon M, MacDonald A, White FJ. 28. Disorders of Amino Acid Metabolism, Organic Acidaemias and Urea Cycle Disorders. Clinical Paediatric Dietetics, 5th edition: Wiley Blackwell; 2020.

Leucine100 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
For use under medical supervision. For healthcare professional use only.

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Methionine

Description
A powdered, unflavoured, pre-measured L-methionine amino acid supplement on a carbohydrate base.

Product range	Methionine100
Pack size	30 x 4g e
Ingredients	Dried glucose syrup, L-methionine.

Indication
Methionine100 is for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Remethylation disorders[1]

Remethylation defects are rare inherited disorders in which a deficient activity of methionine synthase results in impaired remethylation of homocysteine to L-methionine, leading to the accumulation of homocysteine and decreased L-methionine levels.

Although remethylation disorders affect multiple systems, and thus often presents in a multifaceted manner, a few patterns of clinical presentations can be identified. These include developmental and neurocognitive impairment, feeding problems, neurological symptoms including seizures, movement disorders, abnormal muscle tone, visual impairment, neuropathy and haematological abnormalities.

L-methionine is an essential amino acid. Maintaining plasma levels in the normal range is recommended and can be achieved by oral L-methionine supplementation where necessary.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4 Wait 5 minutes*, shake again and serve.

*The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Further reading

1. Huemer M DD, Schwahn B, Schiff M, Bandeira A, Benoist J-F, Burlina A, Cerone R, Couce ML, Garcia-Cazorla A, la Marca G, Pasquini E, Vilarinho L, Weisfeld-Adams JD, Kožich V, Blom H, Baumgartner MR, Dionisi-Vici C. Guidelines for diagnosis and management of the cobalamin-related remethylation disorders cblC, cblD, cblE, cblF, cblG, cblJ and MTHFR deficiency. Journal of Inherited Metabolic Disease. 2017;40:21-48.

Methionine100 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
For use under medical supervision. For healthcare professional use only.

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Phenylalanine

Product range	**Phenylalanine50**
Pack size	**30 x 4g** e
Ingredients	Dried glucose syrup, L-phenylalanine.

Description

A powdered, unflavoured, pre-measured L-phenylalanine amino acid supplement on a carbohydrate base.

Indication

Phenylalanine50 is for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Hereditary tyrosinaemia type I (HT-1)[1,2]

HT-I is an autosomal recessive inherited disorder of the amino acid L-tyrosine. The main organs affected are the liver, kidneys and peripheral nervous system, with death in childhood common in untreated individuals.

The management of HT-I includes the dietary restriction of both L-tyrosine and its precursor amino acid, L-phenylalanine. The restriction of L-phenylalanine can lead to low plasma levels of this essential amino acid. Low L-phenylalanine concentrations may be detrimental to development. Supplementing the diet with enough L-phenylalanine to manage low levels, without leading to an excessive rise in L-tyrosine levels, is sometimes indicated.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4 Wait 5 minutes*, shake again and serve.

*The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Phenylalanine50 can be used in conjunction with other Vitaflo products for the dietary management of HT-1:

Further reading

1. de Laet C, Dionisi-Vici C, Leonard JV, McKiernan P, Mitchell G, Monti L et al. Recommendations for the management of tyrosinaemia type I. Orphanet Journal of Rare Diseases 2013: 8:8. DOI: 10.1038/gim.2017.101.

2. Chinsky JM, Singh R, Ficicioglu C, van Karnebeek CDM, Grompe M, Mitchell G et al. Diagnosis and treatment of tyrosinemia type I: a US and Canadian consensus group review and recommendations. Genetics in Medicine. 2017; 19(12): doi: 10.1038/gim.2017.101

Phenylalanine50 is a Food for Special Medical Purposes for the dietary management of inborn errors of metabolism.
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Tyrosine

Product range	**Tyrosine1000**
Pack size	**30 x 4g** e
Ingredients	Dried Glucose Syrup, L-Tyrosine, Thickener(E415).

Description

A powdered, unflavoured, pre-measured L-tyrosine amino acid supplement on a carbohydrate base.

Indication

Tyrosine1000 is for the dietary management of inborn errors of amino acid metabolism, suitable from 3 years of age onwards.

Clinical use

Phenylketonuria (PKU)[1]

PKU is an autosomal recessive inborn error of L-phenylalanine (Phe) metabolism, caused by deficiency in the enzyme phenylalanine hydroxylase (PAH), which converts Phe to tyrosine. PAH deficiency leads to accumulation of Phe in the blood and brain. Symptoms of untreated PKU include irreversible intellectual disability, microcephaly, motor deficits, seizures and developmental delay.

Dietary management of PKU involves a low Phe diet in combination with Phe-free protein substitutes (PS). As Phe is a precursor to L-tyrosine, L-tyrosine becomes an essential amino acid when Phe intake is restricted. L-tyrosine is important in the production of brain neurotransmitters, thyroxin and melanin skin pigments. As L-tyrosine is already added to Phe-free PS, additional L-tyrosine supplementation is not usually required in routine care.

Individuals who do not take the prescribed amount of PS may have an inadequate intake of L-tyrosine. It is recommended that supplemental L-tyrosine be provided if blood L-tyrosine concentrations are consistently below the normal range.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds and serve.

For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Tyrosine1000 can be used in conjunction with other Vitaflo products for the dietary management of PKU:

Further reading

1. Genetic Metabolic Dietitians International (GMDI). PKU Nutrition Management Guidelines 2016. Available from: https://managementguidelines.net/guidelines.php/90/overview/0/0/PKU%20Nutrition%20Guidelines/Version%201.12/Overview

2. van Wegberg AMJ, MacDonald A, Ahring K, B langer-Quintana A, Blau N, Bosch AM, et al. The complete European guidelines on phenylketonuria: diagnosis and treatment. Orphanet Journal of Rare Diseases. 2017;12(1):1-56.

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Valine

Product range	Valine50	Valine1000
Pack size	30 x 4g e	30 x 4g e
Ingredients	Dried glucose syrup, L-valine.	Dried glucose syrup, L-valine.

Description

Powdered, unflavoured, pre-measured L-valine amino acid supplements on a carbohydrate base.

Indication

Valine50 and **Valine1000** are for the dietary management of inborn errors of amino acid metabolism, suitable from birth.

Clinical use

Maple syrup urine disease (MSUD)[1,2]

MSUD is an inherited metabolic disorder caused by branched-chain α-ketoacid dehydrogenase (BCKD) deficiency, resulting in the accumulation of branched chain amino acids (BCAAs) L-leucine, L-isoleucine and L-valine. MSUD varies considerably in disease severity, with clinical features including seizures, ketoacidosis, hypoglycaemia, apnoea, ataxia and coma. Elevated L-leucine levels are toxic and associated with abnormal brain morphology and cognitive impairment. This may impact on social and psychomotor function.

The mainstay of dietary management is the restriction of dietary BCAAs, especially L-leucine, and the use of BCAA-free protein substitutes (PS). To promote anabolism of L-leucine, when L-leucine blood concentrations are high, additional supplementation with L-valine may be required. In addition, patients on dietary management can develop a L-valine deficiency, which is absent from their PS. This can lead to a rash and can also involve the eye and gut epithelium. Supplementation of the diet with L-valine is used to manage this.

Preparation guidelines

Step 1 Measure out 50ml of water into a shaker.

Step 2 Add one sachet.

Step 3 Shake vigorously for 30 seconds.

Step 4 Wait 1 minute*, shake again and serve.

*The waiting time will enhance dispersion of the powder in the water.
For alternative serving suggestions, please refer to 'Mixing with protein substitutes and flavouring options' at the end of this handbook.

Combinations

Valine50 & **Valine1000** can be used in conjunction with other Vitaflo products for the dietary management of MSUD:

Further reading

1. Frazier DM, Allgeier C, Homer C, Marriage BJ, Ogata B, Rohr F, et al. Nutrition management guideline for maple syrup urine disease: an evidence-and consensus-based approach. Molecular Genetics and Metabolism. 2014;112(3):210-7.
2. Hollak CE, Lachmann R. Inherited Metabolic Disease in Adults: A Clinical Guide: Oxford University Press; 2016.

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Mixing with Protein Substitutes and Flavouring Options

Can the SDAAs be STIRRED instead of shaken?

We don't advise it. They should be shaken not stirred. Some of our SDAA range form lumps when stirred.

Can SDAAs be prepared with a protein substitute?

Yes. The process is different for powdered and liquid protein substitutes. Please follow the relevant instructions below.

Is it possible to use less water?

For some SDAAs, yes. Please contact your Vitaflo representative if you require information on the suitability of preparing particular SDAA products in a lower volume of fluid.

Is it possible to change the flavour?

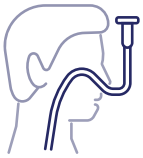
Yes. We recommend using 100ml of apple juice or reconstituted cordial (orange or lime).

Adding SDAAs to a powdered protein substitute:

1. Mix the dry SDAA and protein substitute powders together.
2. Follow the protein substitute preparation instructions. You may wish to use extra water.
3. Serve.

Adding SDAAs to a liquid protein substitute:

1. Pour the liquid protein substitute into a shaker.
2. Add in the SDAA.
3. Shake for 30 seconds.
4. Serve.



SDAAs and tube feeding

- SDAAs can be used as ingredients in modular feeds. Blend with other powder ingredients before adding liquid ingredients.
- SDAAs can also be administered individually via a feeding tube. With the SDAA range varying significantly in solubility, it is advised to administer using the bolus method. Prepare the SDAA with water and flush with water pre- and post-bolus.

Single Dose Amino Acids are Food for Special Medical Purposes for the dietary management of inborn errors of metabolism. Please refer to individual product information for age indications. For use under medical supervision. For healthcare professional use only.

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