Metabolic Product Formulary

SUMMARY OF PRESCRIBABLE METABOLIC PRODUCTS AVAILABLE IN NHS GREATER GLASGOW & CLYDE

The quantity of all nutritional products used in the management of metabolic disorders, are determined by the Metabolic Specialist Team managing the patient. All products contained in this list are ACBS approved.

Other nutritional products may be required for the management of this patient group, further information can be found within the

NHSGGC Low Protein food Formulary http://www.ggcprescribing.org.uk/media/uploads/other_formularies/low_protein_formulary__final_01062016.pdf

and the NHSGGC Paediatric Nutrition Formulary
http://www.ggcprescribing.org.uk/media/uploads/other_formularies/infant_and_paediatric_oral_and_enteral_nutrition_formulary_final_01062016_(2).pdf

Phenylketonuria (Pages 5-12)

- PKU Anamix® Infant
- PKU Anamix® First Spoon
- PKU Anamix® Junior
- PKU Anamix® Junior LQ
- Lophlex®
- PKU Lophlex® LQ 10
- PKU Lophlex® LQ 20
- PKU Lophlex® Sensation 20
- Phlexy-10® Exchange System
- PK Aid-4®
- PKU Start®
- Easiphen®
- XP Maxamaid®
- XP Maxamum®
- PKU Squeezie®
- PKU Gel®
- PKU Cooler10®
- PKU Cooler15®
- PKU Cooler20®
- PKU Express15®
- PKU Express20®
- PKU Air 15®
- PKU Air 20®
- XPhe Jump 10 ®
- XPhe Jump 20®
- PKU Sphere®
- L-Tyrosine
- Glytactin RTD 10®
- Glytactin RTD 15®
- GlytactinBettermilk 15®

Other Inborn errors of metabolism
(Pages 13-17)

- S.O.S. 10
- S.O.S. 15
- S.O.S. 20
- S.O.S. 25
- Basecal100
- Basecal200
- Energivit®
- EAA® Supplement
- DocOmega®
- KeyOmega®
- Cystine500®
- Valine50®
- UCD Amino 5®
- Isoleucine50®
- Leucine100®
- Phenylalanine50®
- Tyrosine1000®
- Infasoy®
- Wysoy®

Vitamin Supplements (Page 18)

- Fruitivits®
- Phlexy-Vits®
- Paediatric Seravit®

Miscellaneous Page (Page 19)

- FlavourPac
- Modjul® Flavour Powder
- Flavour Mix

Milk Replacements (Page 20)

- Loprofin® PKU Drink
- Dalia liquid®
- Loprofin®Sno-Pro
- Prozero®

GlutaricAciduria (Pages 21-22)

- GA1 Anamix® Infant
- GA1 Anamix® Junior
- GA Gel®
- GA amino5®
- XLYS, TRY Glutaridon®
- XLYS,LowTRY,Maxamaid®

Glycogen storage disease (Page 22)

- Glycosade®

Homocystinuria or hypermethioninaemia (Page 23-26)

- HCU Anamix® Infant
- HCU Lophlex® LQ 20
- HCU LV®
- HCU Anamix® Junior
- HCU gel®
- HCU Cooler® 10
- HCU Cooler® 15
- HCU Cooler® 20
- HCU Express® 15
- HCU Express® 20
- XMET Homidon®
- XMET Maxamaid®
- XMET Maxamum®

Hyperlysinaemia (Page 27)

- HYPER LYSAnamix® Infant
- XLYS Maxamaid®

Isovalericacidaemia (Page 28)

- IVA Anamix® Infant
- IVA Anamix® Junior
- XLEU Faladon®
- XLEU Maxamaid®

Maple Syrup Urine Disease (Page29-32)

- MSUD Anamix® Infant
- MSUD Anamix® Junior
- MSUD Anamix® Junior LQ
- MSUD Aid III®
- MSUD Lophlex® LQ 20
- MSUD Maxamaid®
- MSUD Maxamum®
- MSUD Gel®
- MSUD Cooler® 10
- MSUD Cooler® 15
- MSUD Cooler® 20
- MSUD Express® 15
- MSUD Express® 20
- MSUD amino5®

Methylmalonic or propionic acidaemia (Page 33-34)

- MMA/PA Anamix® Infant
- MMA/PA Anamix® Junior
- MMA/PA amino5®
- XMTVI Asadon®
- XMTVI Maxamaid®
- XMTVI Maxamum®

Tyrosinaemia (Page 35-38)

- TYR Anamix® Infant
- TYR Anamix® Junior
- TYR Anamix® Junior LQ
- TYR Lophlex® LQ 20
- TYR Gel®
- TYR Cooler® 10
- TYR Cooler® 15
- TYR Cooler® 20
- TYR Express15®
- TYR Express20®
- XPHEN TYR Maxamaid®
- XPHEN TYR Maxamum®
- XPHEN TYR Tyrosidon®
- XPTM Tyrosidon®

Fatty Acid Oxidisation Disorder (Page 39)

- Monogen®
- MCT Procal®
- Lipistart®

CHO Disorder (Page 40)

- Galactomin 17
- Galactomin 19

Ketogenic Diets(Page 41-42)

- Ketocal®
- Ketocal® 3:1
- Ketocal® 4:1 LQ
- Betaquik®
- Carbzero
- Keyo®
- KetoClassic Bar

A2.7 Nutritional supplements for metabolic diseases

Phenylketonuria

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven phenylketonuria in children from birth to 3 years

PKU Anamix® Infant

Unflavoured, 400g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 13.1 g, carbohydrate 49.5 g, fat 23 g, fibre 5.3 g, energy 1915 kJ (457 kcal)/100g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2 g, carbohydrate 7.4 g, fat 3.5 g, fibre 800 mg, energy 287kJ (69kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven phenylketonuria in children from 6 months to 5 years

PKU Anamix® First Spoon

12.5g Sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 5 g, carbohydrate 4.8 g, fat 150 mg, fibre nil, energy 168 kJ (41 kcal)/12.5-g sachet, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children 1–10 years

PKU Anamix® Junior

Flavours:Chocolate, Berry, Orange, Vanilla, Neutral, 30×36g sachets

Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 10 g, carbohydrate 11.5 g, fat 4.5 g, fibre 4g, energy 566 kJ (135 kcal)/36-g sachet, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary supplement of phenyketonuria in children 1–10 years

PKU Anamix® Junior LQ

Flavours:berry, orange, or unflavoured, 125ml carton
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 10 g, carbohydrate 8.8 g, fat 4.8 g, fibre 310 mg, energy 497 kJ (118 kcal)/125 ml, with vitamins, minerals, and trace elements. Lactose-free.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven phenylketonuria in children over 8 years and adults including pregnant women

Lophlex®

Flavours: berry, orange or unflavoured, 30×27.8g sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 20 g, carbohydrate 2.5 g, fat 60 mg, fibre 220 mg, energy 385 kJ (91 kcal)/27.8-g sachet, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 4 years and adults including pregnant women

PKU Lophlex® LQ 10

Flavours: berry, orange, juicy citrus, juicy tropical, juicy berries, juicy orange (energy 246 kJ (58 kcal), 62.5 ml carton
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 10 g, carbohydrate 4.4 g, fibre 250 mg, energy 245 kJ (58 kcal)/62.5 ml, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 4 years and adults including pregnant women

PKU Lophlex® LQ 20

Flavours: berry, orange, juicy tropical, juicy citrus, juicy berries, juicy orange (fibre 500 mg, energy 493kJ (115kcal), 125ml carton
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 20 g, carbohydrate 8.8 g, fibre 500 mg, energy 490 kJ (115 kcal)/125ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 4 years and adults including pregnant women

PKU Lophlex® Sensation 20

Flavours: berry or orange, 3×109g pot
Semi-solid, protein equivalent (containing essential and non-essential amino acids except phenylalanine) 20g, carbohydrate 20.2g, fibre 1g, energy 706kJ (166kcal)/109 g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria. Suitable for 8 years and over.

Phlexy-10® Exchange System

Capsules, protein equivalent (essential and non-essential amino acids except phenylalanine) 416.5mg/capsule, 200-cap pack **(Average 120 capsules/day)**

Tablets, protein equivalent (essential and non-essential amino acids except phenylalanine) 833 mg tablet, 75-tab pack **(Average 60 tablets/day)**
Drink Mix, powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 8.33 g, carbohydrate 8.8 g/20-g sachet. Apple-black currant, citrus, or tropical flavour, 30×20-g sachet. **(Average 6 sachets/day)**

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children and adults

PK Aid-4®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 79g, carbohydrate 4.5g, fat nil, energy 1420kJ (334 kcal)/100 g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria from birth to 1 year

PKU Start®

Unflavoured, 500ml bottle
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 2g, carbohydrate 8.3g, fat 2.9g, energy 286kJ (68 kcal) / 100ml,

with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven phenylketonuria in children over 8 years

Easiphen®

Flavours: forest berries, orange, or tropical fruit, 250 ml carton
Liquid, protein equivalent (containing essential and non-essential amino acids except phenylalanine) 6.7g, carbohydrate 5.1g, fat 2g, energy 275kJ (65 kcal)/100ml with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children 1– 8 years

XP Maxamaid®

Flavours: orange or unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 25g, carbohydrate 51g, fat less than 500mg, energy 1311kJ (309 kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 8 years

XP Maxamum®

Flavours: orange, unflavoured. 30×50g sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 39g, carbohydrate 34g, fat less than 500mg, energy 1260kJ (297kcal)/100g, with vitamins, minerals, and trace elements.

Unflavoured, 30×25g sachets
Powder, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 15g, carbohydrate 3.8g, fat less than 100mg, energy 315kJ (75 kcal)/25g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of phenylketonuria in children from 6 months to 10 years

PKU Squeezie®

Flavour: apple-banana, 30×85g pouch

Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 10g, carbohydrate 22.5g, fat 500mg, energy 565kJ (135 kcal)/85g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

For use as part of the low-protein dietary management of phenylketonuria in children 1–10 years

PKU gel®

Flavours: orange, raspberry, or unflavoured, 30x24g sachets

Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 10g, carbohydrate 8.9g, fat less than 100mg, energy 318kJ (76 kcal)/24g, with vitamins, minerals, and trace elements. Orange, raspberry, or unflavoured (carbohydrate 10.3g, energy 339 kJ (81 kcal)/24g),

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Cooler10®

Unflavoured (white) or flavoured (orange, purple, or red), 30×87 ml

Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 10g, carbohydrate 5.1g, energy 258kJ (6kcal)/87-ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Cooler15®

Unflavoured (white) or flavoured (orange, purple, or red), 30x130 ml

Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 15g, carbohydrate 7.8g, energy 386 kJ (92kcal)/130-ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Cooler20®

Unflavoured (white) or flavoured (orange, purple, or red), 30x174 ml
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine) 20g, carbohydrate 10.2g, energy 517kJ (124kcal)/174-ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Air 15®

flavoured (green, gold, red, or white), 30x130 ml
Liquid: protein equivalent (essential and non-essential amino acids except phenylalanine) 15g, fat 0.8g, carbohydrate 2.0g, energy 316 kJ (75kcal)/130-ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Air 20®

flavoured (green, gold, red, or white), 30x174 ml
Liquid: protein equivalent (essential and non-essential amino acids except phenylalanine) 20g, fat 1.0g, carbohydrate 2.6g, energy 423 kJ (100kcal)/174-ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Express15®

Flavours: lemon, orange, tropical, or unflavoured, 30x25 g sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 15g, carbohydrate 2.4 g, energy 293 kJ (70kcal)/25g, with vitamins, minerals, and trace elements. For unflavoured, carbohydrate 3.4 g, energy 310 kJ (74kcal)/25g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Express20®

Flavours: lemon, orange, tropical, or unflavoured, 30x34 g sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 20g, carbohydrate 3.3g, energy 389kJ (93 kcal)/34g, with vitamins, minerals, and trace elements. For unflavoured, carbohydrate 4.7g, energy 416kJ (99kcal)/34g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

PKU Sphere®

Flavours: vanilla, berry
Powder: caesinegycomacropeptide isolate: protein equivalent (essential and non-essential amino acids except phenylalanine) 20g, Fat 1.6g; carbohydrate 6.3g, energy 508 KJ (120 cals) / 35g sachet with vitamins, minerals and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in pregnant women with low plasma tyrosine concentrations

L-Tyrosine

100g
Powder, L-tyrosine 20g, carbohydrate 76.8g, fat nil, energy 1612kJ (379kcal)/100g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

XPhe Jump10®

Flavours: cola, wild berries, orange, or neutral, 30x63 ml
Liquid,protein equivalent (essential and non-essential amino acids except phenylalanine) 10g, fat 0.3g, carbohydrate 3.6g neutral; 5g flavoured, energy 470 kJ neutral (533 Kj flavoured (56 kcal neutral / 63 kcal flavoured)/63-ml pouch, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

XPhe Jump 20®

Flavours: cola, wild berries, orange, or neutral, 30x125 ml
Liquid,protein equivalent (essential and non-essential amino acids except phenylalanine) 20g, fat 0.6g carbohydrate 7g neutral; 8g flavoured, energy 237 kJ neutral; 269 flavoured (111kcal flavoured; 126 kcal flavoured)/125-ml pouch, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

Glytactin RTD 10

Flavours: original and chocolate, 30 x 250ml
Liquid,protein equivalent (essential and non-essential amino acids except phenylalanine)10g, fat 3.5g, carbohydrate 22g, energy 642kJ (153kcal)/250 ml carton, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

Glytactin RTD 15

Flavours: original and chocolate, 30 x 250ml
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine)15g, fat 5.0g, carbohydrate 24g, energy 840 kJ (200cal)/ 250 ml carton, with vitamins, minerals, and trace elements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 3 years

GlytactinBettermilk 15-original

30 x 49g sachet
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine) 15g, carbohydrate 23g, fat 4.5g, energy 672 kJ (160kcal)/49g, with vitamins, minerals, and trace elements.

Other inborn errors of metabolism

A2.4.1 High-energy supplements: Carbohydrate
For use as an emergency regimen in the dietary management of inborn errors of metabolism in adults and children from birth

S.O.S. 10
30x21g sachet
Powder, carbohydrate 95g, fat nil, fibre nil, protein nil, energy 1590kJ (380kcal)/100g

A2.4.1 High-energy supplements: Carbohydrate
For use as an emergency regimen in the dietary management of inborn errors of metabolism in adults and children from birth

S.O.S. 15
30x31g sachet
Powder, carbohydrate 95g, fat nil, fibre nil, protein nil, energy 1590kJ (380kcal)/100g

A2.4.1 High-energy supplements: Carbohydrate
For use as an emergency regimen in the dietary management of inborn errors of metabolism in adults and children from birth

S.O.S. 20
30x 42g sachet
Powder, carbohydrate 95g, fat nil, fibre nil, protein nil, energy 1590kJ (380kcal)/100g

A2.4.1 High-energy supplements: Carbohydrate
For use as an emergency regimen in the dietary management of inborn errors of metabolism in adults and children from birth

S.O.S. 25
30x52g sachet
Powder, carbohydrate 95g, fat nil, fibre nil, protein nil, energy 1590kJ (380kcal)/100g

ACBS approved
Nutritional supplement used for the dietary management of inborn errors of metabolism requiring a low protein diet. Suitable for 3 years of age.

Basecal100®
21.5g sachet
Powder, Protein nil, carbohydrate 69.8g, fat 20.7g, energy 1952kJ (465kcal)/100g, with vitamins, minerals, and trace elements.

ACBS approved
Nutritional supplement used for the dietary management of inborn errors of metabolism requiring a low protein diet. Suitable for 3 years of age.

Basecal 200®
43g sachet
Powder, Protein nil, carbohydrate 69.8 g, fat 20.7g, energy 1952kJ(465 kcal)/100g, with vitamins, minerals, and trace elements.

A2.4.1 High-energy supplements:Fat
Liquid supplements should be diluted before use in child under 5 years

Energivit®
400g can
Powder provides: carbohydrate 66.7g, fat 25g, energy 2059kJ (492kcal)/100g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of disorders of protein metabolism including urea cycle disorders in children over 3 years

EAA® Supplement
50x 12.5g sachets
Powder, protein equivalent (essential amino acids) 5g, carbohydrate 4g, fat nil, energy 151kJ (36kcal)/12.5g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of disorders of protein metabolism including urea cycle disorders in children over 3 years

UCD amino5®
6g sachets
Powder, protein equivalent (essential amino acids supplement) 5 g, carbohydrate nil, fat nil, energy 85 kJ (20kcal)/6.6 g

Nutritional supplement for the dietary management of inborn errors of metabolism for adults and children from birth

DocOmega®
30x4g sachets
Powder, protein (cows' milk, soya) 100mg, carbohydrate 3.2g, fat 500mg (of which docosahexaenoic acid 200mg), fibre nil, energy 74kJ (18kcal)/4g, with minerals

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of inborn errors of metabolism. Suitable from birth.

KeyOmega®
30x4g sachets
Powder, protein (cows' milk, soya) 170mg, carbohydrate 2.8g, fat 800mg (of which arachidonic acid 200mg, docosahexaenoic acid 100mg), energy 80kJ (19kcal)/4g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of inborn errors of amino acid metabolism in adults and children from birth

Cystine500®
30x4 g sachets
Powder, cystine 500mg, carbohydrate 3.3g, fat nil, energy 63 k (15 kcal)/4g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of inborn errors of amino acid metabolism in adults and children from birth

Valine50®
30x4g sachets
Powder, valine 50mg, carbohydrate 3.8g, fat nil, energy 63kJ (15kcal)/4g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for use in the dietary management of inborn errors of amino acid metabolism in adults and children from birth

Isoleucine50®
30x4 g sachets
Powder, isoleucine 50mg, carbohydrate 3.8g, fat nil, energy 63kJ (15kcal)/4g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of inborn errors of amino acid metabolism in adults and children from birth

Leucine100®
30x 4g sachets
Powder, leucine 100mg, carbohydrate 3.7g, fat nil, energy 63kJ (15kcal)/4 g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for use in the dietary management of inborn errors of metabolism in adults and children from birth

Phenylalanine50®
30x4g sachets
Powder, phenylalanine 50mg, carbohydrate 3.8g, fat nil, energy 63kJ (15kcal)/4g

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of inborn errors of amino acid metabolism in adults and children from birth

Tyrosine1000®
30x4g sachets
Powder, tyrosine 1g, carbohydrate 2.9g, fat nil, energy 63kJ (15 kcal)/4g sachet

A.2.3.1 Specialised formulas: Infant and child: Soya based formula
Suitable for galactokinase deficiency and galactosaemia

Infasoy®
900g can
Powder provides: protein 12.8g, carbohydrate 54.5g, fat 27.3g, energy 2150 kJ(514 kcal)/100g

A.2.3.1 Specialised formulas: Infant and child: Soya based formula
Suitable for galactokinase deficiency and galactosaemia

Wysoy®
430g can
Powder provides: protein 14g, carbohydrate 54g, fat 27g, energy 2155kJ (515 kcal)/100g

Vitamin Supplements

A2.4.2 Vitamin and mineral supplements
Vitamin, mineral and trace element supplement for children 3-10 years with restrictive therapeutic diets

Fruitivits®
Sachet 30 x 6g
Powder

A2.7 Nutritional supplements for metabolic diseases
For use as a vitamin and mineral component of restricted therapeutic diets in children over 11 years and adults with phenylketonuria and similar amino acid abnormalities

Phlexy-Vits®
Powder, vitamins, minerals, and trace elements, 30 × 7-g sachets
Tablets, vitamins, minerals, and trace elements, 180-tab pack

A2.4.2 Vitamin and mineral supplements
Vitamin, mineral and trace element supplement in fants and children with restrictive therapeutic diets

Paediatric Seravit®
Unflavoured and pineapple(not suitable for under 6 months), 200g tub
Powder

Miscellaneous

A2.5.3 Flavour preparations
For use with Vitaflo's range of unflavoured protein substitutes for metabolic diseases; not suitable for child under 1 year

FlavourPac®
30x4g sachets
Powder

A2.5.3 Flavour preparations
For use with unflavoured SHS products based on peptides or amino acids; not suitable for child under 6 months

Modjul® Flavour Powder
Flavours: blackcurrant, orange, pineapple, 100g; cherry-vanilla, grapefruit, lemon-lime, 20×5g sachets
Powder

A2.5.3 Flavour preparations

Flavour Mix®
Flavours: banana, chocolate, coffee, lemon-lime, strawberry, 60g tub
Powder

Milk Replacements

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria in children over 1 year

Loprofin® PKU Drink

200 ml carton
Liquid, protein (cows' milk) 400 mg (phenylalanine 10 mg), lactose 9.4 g, fat 2 g, energy 165 kJ (40 kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria, chronic renal failure and other inborn errors of amino acid metabolism

Loprofin®Sno-Pro

200 mlcarton
Liquid, protein (cows' milk) 220 mg (phenylalanine 12.5 mg), carbohydrate 8 g, fat 3.8 g, energy 273 kJ (65 kcal)/100 mL. Contains lactose.

A2.7 Nutritional supplements for metabolic diseases
A protein-free nutritional supplement for the dietary management of inborn errors of metabolism in children over 6 months and adults

ProZero®

18 x 250 ml; 6 x 1 litre
Liquid, carbohydrate 8.1 g (of which sugars 3.5 g), fat 3.8 g, energy 278 kJ (66 kcal)/100 ml. Contains lactose.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of phenylketonuria, chronic renal failure and other inborn errors of amino acid metabolism

Dalia liquid®

200ml carton
Liquid, protein (cow's milk) 0.2g (phenylalanine 6.4mg), carbohydrate 6.4 g (of which sugars 4.8 g), fat 2.6 g, energy 208 kJ (50 kcal)/100 ml. Contains lactose.

GlutaricAciduria

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven glutaricaciduria (type 1) in children from birth to 3 years

GA1 Anamix® Infant

Unflavoured, 400 g
Powder, protein equivalent (essential and non-essential amino acids except lysine, and low tryptophan) 13.1 g, carbohydrate 49.5 g, fat 23 g, fibre 5.3 g, energy 1915 kJ (457 kcal)/100 g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2 g, carbohydrate 7.4 g, fat 3.5 g, fibre 800 mg, energy 287 kJ (69 kcal)/100 ml

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven glutaricaciduria (type 1) in children aged 1-10 years

GA1 Anamix® Junior

Neutral, 30 x 18g sachets
Powder, protein equivalent (essential and non-essential amino acids) 28g, carbohydrate 30g, fat 12.5g, fibre 11.2g, energy 367kcal /100 g with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for type 1 glutaricaciduria in children and adults; requires additional source of vitamins, minerals and trace elements

XLYS, TRY Glutaridon®

Unflavoured, 2 × 500 g
Powder, protein equivalent (essential and non-essential amino acids except lysine and tryptophan) 79g, carbohydrate 4 g, energy 1411 kJ (332 kcal)/100 g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for dietary management of type 1 glutaricaciduria. Suitable for children aged 1 to 8 years

XLYS, Low TRY, Maxamaid®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids except lysine, and low tryptophan) 25 g, carbohydrate 51 g, fat less than 500 mg, energy 1311 kJ (309 kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for dietary management of type 1 glutaricaciduria in children 6 months–10 years

GA Gel®

Unflavoured 30 × 24-g sachets
Gel, protein equivalent (essential and non-essential amino acids except lysine, and low tryptophan) 10 g, carbohydrate 10.3 g, fat trace, energy 339 kJ (81 kcal)/24 g, with vitamins, minerals, and trace elements.

ACBS approved
Nutritional supplement for dietary management of type 1 glutaricaciduria. Suitable for 3 years

GA amino5®

6g sachets
Powder, protein equivalent (essential and non-essential amino acids except lysine, and has Low Tryptophan) 5 g, carbohydrate nil, fat less than nil, energy 85 kJ (20kcal)/6g

Glycogen storage disease

A2.7 Nutritional supplements for metabolic diseases
A nutritional supplement for use in the dietary management of glycogen storage disease and other metabolic conditions where a constant supply of glucose is essential. Not suitable for use in children under 2 years

Glycosade®

30 × 60 g sachets
Powder, protein 200 mg, carbohydrate (maize starch) 47.6 g, fat 100 mg, fibre less than 600 mg, energy 803 kJ (192 kcal)/60g

Homocystinuria or hypermethioninaemia

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of proven vitamin B_6 non-responsive homocystinuria or hypermethioninaemia in children from birth to 3 years

HCU Anamix® Infant

Unflavoured, 400 g

Powder, protein equivalent (essential and non-essential amino acids except methionine) 13.1 g, carbohydrate 49.5 g, fat 23 g, fibre 5.3 g, energy 1915 kJ (457 kcal)/100 g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2 g, carbohydrate 7.4 g, fat 3,5 g, fibre 800 mg, energy 287 kJ (69 kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of homocystinuria in children over 3 years

HCU Lophlex® LQ 20

Juicy berries flavour, 125 ml

Liquid, protein equivalent (essential and non-essential amino acids except methionine) 20 g, carbohydrate 8.8 g, fat 440 mg, energy 509 kJ (120 kcal)/125 ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of hypermethioninaemia or vitamin B_6 non-responsive homocystinuria in children over 8 years

HCU LV®

Unflavoured or tropical flavour, 30 × 27.8 g sachets

Powder, protein (essential and non-essential amino acids except methionine) 20 g, carbohydrate 2.5 g, fat 190 mg, energy 390 kJ (92 kcal)/27.8-g sachet, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of hypermethioninaemia or vitamin B_6 non-responsive homocystinuria in children 1 – 10 years

HCU Anamix® Junior

Neutral flavour, 30x 36g sachets

Powder, protein (essential and non-essential amino acids except methionine) 28g, carbohydrate 32g, fat 12.5g, fibre 11.2g, energy 375kcal /100g ,with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of hypermethioninaemia or homocystinuria in children

XMET Homidon®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids, except methionine) 77 g, carbohydrate 4.5 g, fat nil, energy 1386 kJ (326 kcal)/100 g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of hypermethioninaemia or homocystinuria. Suitable for children aged 1-8 years

XMET Maxamaid®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids except methionine) 25 g, carbohydrate 51 g, fat less than 500 mg, energy 1311 kJ (309 kcal)/100 g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of hypermethioninaemia or homocystinuria. Suitable for children over 8 years, adults and pregnant women.

XMET Maxamum®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids except methionine) 39 g, carbohydrate 34 g, fat less than 500 mg, energy 1260 kJ (297 kcal)/100 g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement for the dietary management of children 1–10 years with homocystinuria

HCU gel®

Unflavoured, 30 × 24 g sachets
Powder, protein (essential and non-essential amino acids except methionine) 10 g, carbohydrate 10.3 g, fat 20 mg, energy 339 kJ (81 kcal)/24 g with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement in children over 3 years with homocystinuria

HCU Cooler® 10

Red Flavour, 30 x87ml
Liquid, protein (essential and non-essential amino acids except methionine) 10 g, carbohydrate 4.7 g, fat 0.3 mg, energy 261kJ (62kcal)/87ml with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement in children over 3 years with homocystinuria

HCU Cooler® 15

Orange or red flavour, 30 × 130 ml pouch
Liquid, protein (essential and non-essential amino acids except methionine) 15 g, carbohydrate 7 g, fat 500 mg, energy 393 kJ (92 kcal)/130 ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement in children over 3 years with homocystinuria

HCU Cooler® 20

Red Flavour, 30 x174ml
Liquid, protein (essential and non-essential amino acids except methionine) 10 g, carbohydrate 4.7 g, fat 0.3 mg, energy 526kJ (124kcal)/174ml with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement in children over 8 years with homocystinuria

HCU Express® 15

Unflavoured, 30 × 25 g sachets
Powder, protein (essential and non-essential amino acids except methionine) 15 g, carbohydrate 3.8 g, fat 30 mg, energy 315 kJ (75.3 kcal)/25 g with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
A methionine-free protein substitute for use as a nutritional supplement in children over 8 years with homocystinuria

HCU Express® 20

Unflavoured, 30 × 34 g sachets
Powder, protein (essential and non-essential amino acids except methionine) 20 g, carbohydrate 4.7 g, fat 70 mg, energy 416 kJ (99 kcal)/34 g with vitamins, minerals, and trace elements.

Hyperlysinaemia

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven hyperlysinaemia in children from birth to 3 years

HYPER LYS Anamix® Infant

Unflavoured, 400 g
Powder, protein equivalent (essential and non-essential amino acids except lysine) 13.1 g, carbohydrate 49.5 g, fat 23 g, fibre 5.3 g, energy 1915 kJ (457 kcal)/100 g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2 g, carbohydrate 7.4 g, fat 3.5 g, fibre 800 mg, energy 287 kJ (69 kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of hyperlysinaemia. Suitable for children aged 1-8 years

XLYS Maxamaid®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids except lysine) 25 g, carbohydrate 51 g, fat less than 500 mg, energy 1311 kJ (309 kcal)/100 g with vitamins, minerals, and trace elements.

Isovalericacidaemia

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven isovalericacidaemia or other proven disorders of leucine metabolism in children from birth to 3 years

IVA Anamix® Infant

Unflavoured, 400g
Powder, protein equivalent (essential and non-essential amino acids except leucine) 13.1g, carbohydrate 49.5g, fat 23g, fibre 5.3g, energy 1915kJ (457kcal)/100g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2g, carbohydrate 7.4g, fat 3.5g, fibre 800mg, energy 287kJ (69kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven isovalericacidaemia or other proven disorders of leucine metabolism in children from 1 – 10 years

IVA Anamix® Junior

Unflavoured, 400g
Powder,protein equivalent (essential and non-essential amino acids except leucine) 28g, carbohydrate 30g, fat 12.5g, fibre 11.2g, energy 367kcal /100g, with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of isovalericacidaemia in children.

XLEU Faladon®

Unflavoured, 200g
Powder, protein equivalent (essential and non-essential amino acids except leucine) 77g, carbohydrate 4.5g, fat nil, energy 1386kJ (326kcal)/100g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of isovalericacidaemia. Suitable for children aged 1-8 years

XLEU Maxamaid®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except leucine) 25g, carbohydrate 51g, fat less than 500mg, energy 1311kJ (309 kcal)/100g with vitamins, minerals, and trace elements.

Maple syrup urine disease

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven maple syrup urine disease in children from birth to 3 years

MSUD Anamix® Infant

Unflavoured, 400g
Powder, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 13.1g, carbohydrate 49.5g, fat 23g, fibre 5.3g, energy 1915 kJ (457 kcal)/100g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2 g, carbohydrate 7.4g, fat 3.5g, fibre 800mg, energy 287kJ(69kcal)/100ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children 1–10 years

MSUD Anamix® Junior

Unflavoured, 30×29g sachets
Powder, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 8.4g, carbohydrate 11g, fat 3.9g, energy 474kJ (113 kcal)/29g sachet, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children 1–10 years

MSUD Anamix® Junior LQ

Orange flavour, 125ml carton
Liquid, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, andvaline) 10g, carbohydrate 8.8g, fat 4.8g, fibre 310mg, energy 497kJ(118kcal)/125ml, with vitamins, minerals, and trace elements. Lactose-free.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of maple syrup urine disease and related conditions in children and adults where it is necessary to limit the intake of branched chain amino acids

MSUD Aid III®

Unflavoured, 500g

Powder, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 77g, carbohydrate 4.5g, fat nil, energy 1386kJ (326kcal)/100g.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of maple syrup urine disease in children over 3 years

MSUD Lophlex® LQ 20

Juicy berries flavour, 125ml

Liquid, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 20g, carbohydrate 8.8g, fat less than 500mg, energy 509kJ (120kcal)/125ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of maple syrup urine disease. Suitable for children aged 1-8 years

MSUD Maxamaid®

Unflavoured, 500g

Powder, protein equivalent (essential and non-essential amino acids except isoleucine,leucine, and valine) 25g, carbohydrate 51g, fat less than 500mg, energy 1311kJ (309kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases

Nutritional supplement for the dietary management of maple syrup urine disease. Suitable for children over 8 years of age, adults and pregnant women.

MSUD Maxamum®

Orange flavour or unflavoured, 500g

Powder, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 39g, carbohydrate 34g, fat less than 500mg, energy 1260 kJ (297kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children 1–10 years

MSUD Gel®

Unflavoured, 30×24g sachets
Powder, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 10g, carbohydrate 10.3g, fat less than 100mg, energy 339kJ (81kcal)/24g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children over 3 years and adults

MSUD Cooler® 10

Red Flavour, 30 x 87ml
Liquid, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 10g, carbohydrate 4.7 g, fat 0.3g, energy 261 kJ (62 kcal)/87ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children over 3 years and adults

MSUD Cooler® 15

Orange or red flavour, 30×130 ml
Liquid, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 15g, carbohydrate 7g, fat 500mg, energy 393kJ (92kcal)/130ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children over 3 years and adults

MSUD Cooler® 20

Red Flavour, 30 x 174ml
Liquid, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 20g, carbohydrate 9.4g, fat 0.7g, energy 526kJ (124kcal)/174ml pouch, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children over 8 years and adults

MSUD Express® 15

Unflavoured, 30×25 g sachets
Powder, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 15g, carbohydrate 3.8g, fat less than 100mg, energy 315kJ (75kcal)/25g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of maple syrup urine disease in children over 8 years and adults

MSUD Express® 20

Unflavoured, 30×34 g sachets
Powder, protein equivalent (essential and non-essential amino acids except leucine, isoleucine, and valine) 20g, carbohydrate 4.7g, fat less than 100mg, energy 416kJ (99kcal)/34g, with vitamins, minerals, and trace elements.

ACBS approved
Nutritional supplement for the dietary management of maple syrup urine disease. Suitable from 3 years

MSUD amino5®

6g sachet
Powder, protein equivalent (essential and non-essential amino acids except isoleucine, leucine, and valine) 5g, carbohydrate nil, fat less than nil, energy 85kJ (20kcal)/6g

Methylmalonic or propionic acidaemia

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of methylmalonicacidaemia or propionic acidaemia

MMA/PA Anamix® Infant

Unflavoured, 400g
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, andvaline, and low isoleucine) 13.1g, carbohydrate 49.5g, fat 23g, fibre 5.3g energy1915kJ(457kcal)/100g, with vitamins, minerals, and trace elements. Standard dilution (15%) provides protein equivalent 2g, carbohydrate 7.4g, fat 3.5g, fibre 800mg, energy 287kJ(69kcal)/100ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of methylmalonicacidaemia or propionic acidaemia in children 1-10 years

MMA/PA Anamix® Junior

Unflavoured, 30x 18g sachets
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, and valine, and low isoleucine) 28g, carbohydrate 30g, fat 12.5g, fibre 11.2g, energy 367kcal /100g, with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of methylmalonicacidaemia or propionic acidaemia in children and adults

XMTVI Asadon®

Unflavoured, 200g
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, andvaline, and low isoleucine) 77g, carbohydrate 4.5g, fat nil, energy 1386kJ (326kcal)/100 g.
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, andvaline, and low isoleucine) 25g, carbohydrate 51g, fat less than 500mg, energy 1311kJ (309kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of methylmalonicacidaemia or propionic acidaemia

XMTVI Maxamaid®

Unflavoured, 500 g
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, and valine, and low isoleucine) 25 g, carbohydrate 51 g, fat less than 500 mg, energy 1311 kJ (309 kcal)/100 g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of methylmalonicacidaemia or propionic acidaemia.
Suitable for children aged over 8 years

XMTVI Maxamum®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except methionine, threonine, and valine, and low isoleucine) 39 g, carbohydrate 34 g, fat less than 500 mg, energy 1260 kJ (297 kcal)/100 g, with vitamins, minerals, and trace elements.

ACBS approved
Nutritional supplement for the dietary management of proven methylmalonicacidaemia or propionic acidaemia in children from 3 years

MMA/PA amino5®

6g sachet
Powder, protein equivalent (essential and non-essential amino acids except except methionine, threonine, and valine, and low isoleucine) 5 g, carbohydrate nil, fat less than nil, energy 85 kJ (20kcal)/6 g

Tyrosinaemia

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven tyrosinaemia where plasma-methionine concentrations are normal in children from birth to 3 years

TYR Anamix® Infant

Unflavoured, 400g (5g measuring scoop provided)
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 13.1g, carbohydrate 49.5g, fat 23g, fibre 5.3g, energy 1915kJ (457kcal)/100g, with vitamins, minerals, and trace elements; *standard dilution* (15%) provides protein equivalent 2g, carbohydrate 7.4g, fat 3.5g, fibre 800mg, energy 287kJ (69kcal)/100 ml.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of proven tyrosinaemia in children 1–10 years

TYR Anamix Junior

Unflavoured, 30x29 g sachets
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 8.4g, carbohydrate 11g, fat 3.9g, energy 475kJ (113 kcal)/29g sachet, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia type 1 (when nitisinone (NTBC) is used, see section 9.8.1), type II, and type III, in children over 1 year

TYR Anamix® Junior LQ

Orange flavour, 36×125 ml bottle
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 10g, carbohydrate 8.8g, fat 4.8g, fibre 310mg, energy 500kJ (119 kcal)/125 ml, with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 3 years

TYR Lophlex® LQ 20

Juicy berries flavour, 125ml
Liquid, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 20g, carbohydrate 8.8g, fat less than 500mg, fibre 500mg, energy 509kJ (120 kcal)/125ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children 1–8 years

XPHEN TYR Maxamaid®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 25g, carbohydrate 51g, fat less than 500mg, energy 1311 kJ (309kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 8 years of age and adults including pregnant women

XPHEN TYR Maxamum®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 20g, carbohydrate 34g, fat less than 500mg, energy 1260 kJ (297kcal)/100g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the management of tyrosinaemia in infants, children and adults where plasma-methionine concentrations are normal.

XPHEN TYR Tyrosidon®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except phenylalanine and tyrosine) 77g, carbohydrate 4.5g, fat nil, energy 1386kJ (326 kcal)/100g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia type I in children and adults where plasma-methionine concentrations are above normal

XPTM Tyrosidon®

Unflavoured, 500g
Powder, protein equivalent (essential and non-essential amino acids except methionine, phenylalanine, and tyrosine) 77g, carbohydrate 4.5g, fat nil, energy 1386kJ (326 kcal)/100g.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children 1–10 years

TYR Gel®

Unflavoured, 30×24g sachets
Gel, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 10g, carbohydrate 10.3g, fat less than 100mg, energy 339 kJ (81kcal)/24g, with vitamins, minerals and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 3 years

TYR Cooler® 10

Red flavour 87ml
Liquid, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 10g, carbohydrate 4.7g, fat 0.3g, energy 261kJ (62 kcal)/87ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 3 years

TYR Cooler® 15

Orange or red flavour, 30x130 ml pouch
Liquid, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 15g, carbohydrate 7g, fat 500mg, energy 393kJ (92 kcal)/130ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 3 years

TYR Cooler® 20

Red flavour 174ml
Liquid, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 20g, carbohydrate 9.4g, fat 0.7g, energy 526kJ (124 kcal)/174ml, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 8 years

TYR Express15®

Unflavoured, 30×25 g sachets
Powder, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 15g, carbohydrate 3.4g, fat less than 100mg, energy 310kJ (74kcal)/25g, with vitamins, minerals, and trace elements.

A2.7 Nutritional supplements for metabolic diseases
Nutritional supplement for the dietary management of tyrosinaemia in children over 8 years

TYR Express20®

Unflavoured, 30×34 g sachets
Powder, protein equivalent (essential and non-essential amino acids except tyrosine and phenylalanine) 20g, carbohydrate 4.7g, fat less than 100mg, energy 416kJ (99kcal)/34g, with vitamins, minerals, and trace elements.

Fatty Acid Oxidation Disorder

A.2.3.1 Specialised formulas: Infant and child: MCT-enhanced formula
Specialised formulas are suitable for infants from birth unless otherwise indicated

Monogen®
Can:400g
Powder provides: protein equivalent 12.5g, carbohydrate 68g, fat 11g, energy 1769kJ (420kcal)/100g

A2.4.1.3 High-energy supplements: protein
Dietary management of disorders of long-chain fatty acid oxidation, fat malabsorption, and other disorders requiring a low LCT, high MCT supplement.
Not suitable for child under 1 year

MCT Procal®
30x16 g sachets
Powder 16g provides: protein 2g, carbohydrate 3.3g, fat 10.1g, energy 439 kJ (105kcal)

A.2.3.1 Specialised formulas: Infant and child: MCT-enhanced formula
Specialised formulas are suitable for infants from birth unless otherwise indicated

Lipistart®
Can:400g

CHO Disorder

A2.3.1 Specialised formulas: Infant and child: Residual lactose formula
Specialised formulas are suitable for infants from birth unless otherwise indicated

Galactomin 17®
Unflavoured, 400g can
Powder provides: protein equivalent 12.3g, carbohydrate 55.3g, fat 27.2g, energy 2155kJ (515kcal)/100g

A2.3.1 Specialised formulas: Infant and child: Fructose-based formula
Suitable for infants from birth

Galactomin 19®
400g can
Powder provides: protein equivalent 14.6g, carbohydrate 49.7g, fat 30.8g, energy 2233kJ (534kcal)/100g

Ketogenic Diets

A2.3.2 Specialised formulas for specific clinical conditions
Nutritional supplement as part of a ketogenic diet, suitable for children over 1 year

KetoCal®
Flavours: vanilla, Unflavoured, 300g can.
Powder provides: protein 15.25g, carbohydrate 3g, fat 73g, energy 3011kJ (730kcal)/100g

A2.3.2 Specialised formulas for specific clinical conditions
Nutritional supplement as part of a ketogenic diet, suitable for children over 6 years

KetoCal® 3:1
Unflavoured, 300g can.
Powder provides: protein 15.3g, carbohydrate 7.2g, fat 67.7g, energy 2927kJ (699 kcal)/100g

A2.3.2 Specialised formulas for specific clinical conditions
Nutritional supplement as part of a ketogenic diet, suitable for children over 10 years

KetoCal® 4:1 LQ
Vanilla, 237ml carton.
Liquid provides: protein 3.09g, carbohydrate 610mg, fat 14.8g, fibre 1.12g, energy 620kJ (150kcal)/100ml

ACBS approved
Nutritional supplement for use in the ketogenic diet or in dietary management of conditions requiring a source of MCT. Suitable from the age of 3 years

Betaquik®
250ml carton
Liquid, carbohydrate nil, fat 21 g, energy 777kJ (189 kcal)/100ml.

ACBS approved

Nutritional supplement for use in the ketogenic diet or in dietary management of conditions requiring a source of LCT, Suitable from the age of 3 years

Carbzero

250ml carton

Liquid, carbohydrate nil, fat 20g, energy 740kJ(180kcal)/100ml. Contains Soya.

ACBS approved

Nutritional supplement for use in the ketogenic diet, suitable from the age of 3 years

Keyo®

Flavour: chocolate, 100g pot

Semi-solid food, protein 8g, carbohydrate 2g, fat 30g, energy 1280kJ (310kcal) /100g, with vitamins, minerals, and trace elements .

ACBS approved

Nutritional supplement for use in the ketogenic diet

KetoClassic Bar