DIETETIC NATIONAL FORMULARY FOR INHERITED METABOLIC DISORDERS
ADULTS AND PAEDIATRICS

Acknowledgement:

On behalf of the BIMDG dietitians, I would like to thank all those who have contributed to this formulary: Harriet Churchill, Barbara Cochrane, Anne Daly, Charlotte Ellerton, Suzanne Ford, Camille Newby and Anita MacDonald.

Anne Daly
Chair of BIMDG-DG

BIMDG DIETETIC METABOLIC FORMULARY

INTRODUCTION

This formulary contains specialised dietetic formulae and low protein foods essential in the dietary management of inborn errors of metabolism. Each condition has a specific dietary management, which may be used in conjunction with a drug therapy or as a single treatment approach. Products which are not listed by the Advisory Committee on Borderline Substances (ACBS) but not blacklisted can be prescribed by general practitioners.

Further information on illness management or medications used in IMD can be found on ***www.bimdg.org.uk*.**

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CARBOHYDRATE DISORDERS

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- Energy supplements
- Vitamin, mineral and essential fatty acid supplements
- Flavour sachets
- Single use amino acids
- Low protein foods

GLOSSARY OF TERMS

ACBS	Advisory Committee for Borderline Substances
BIMDG	British Inherited Metabolic Disease Group
AA	Arachidonic acid
CHO	Carbohydrate
CGMP	Casein glycomacropeptide
DHA	Docosahexaenoic acid
EPA	Eicosapentaenoic acid
FOS	Fructose oligosaccaharide
GOS	Galactose oligosaccaharide
LCPUFA	Long chain polyunsaturated fatty acids
LCT	Long chain triglyceride
MCT	Medium chain triglyceride
PHE	Phenylalanine

AMINO ACIDOPATHIES

HOMOCYSTINURIA (HCU)

Background information
Homocystinuria is a rare amino acid disorder with an inability to metabolise the amino acid methionine. Without attentive and appropriate dietary treatment, it can lead to learning difficulties, lens dislocation, and thrombotic events.

Dietary treatment requires:

- a lifelong low protein diet
- supplementation with a protein substitute free of methionine, usually supplemented with vitamins, minerals and DHA
- access to low protein special foods

Protein substitutes
Protein substitutes are essential by helping maintain metabolic control and providing essential nutrients.

Dosage of protein substitutes:
The dose of protein equivalent from amino acid supplements prescribed/kg body weight/day (also considering natural protein tolerance) is:

- children 0-3y: 3g/kg/day
- children 4-6y: 2.5g/kg/day
- children 7-12y: 1.5-2g/kg/day
- over 12y and adults: 1-1.5g/kg/day up to a maximum 80g per day (unless extreme needs or athletes)
- pregnancy >70g/day total

Protein substitute dose is given three to four times per day.

Administration: orally or via nasogastric/gastrostomy tube.

Blood cystine may be low and require cystine supplementation. The dose is dependent on regular blood tests and is adjusted by the specialist dietitian.

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential. Dietary treatment must be sustained throughout pregnancy to protect the mother from thrombotic events, and also to ensure adequate protein intake and overall nutrition to support foetal growth. ***Pregnancy may occur without planning with unsafe metabolic control on patient presentation. Regaining metabolic control is considered a matter of urgency, with strict dietary management being implemented immediately. Therefore, dietary prescription products may be needed on an urgent basis having not been used before/in adulthood.***

Table 1

Name	Unit size	Age	Flavour	Source of protein	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
HCU Anamix infant	400g tin powder	0-3y	Neutral	L - amino acids	13.1g	2/100 mL	70 /100 mL	7.5/ 100 mL	3.5/ 100 mL	Yes	DHA	344-8776
HCU Anamix Junior	36g sachet powder	1-10y	Neutral	L-amino acids	28	10	135	11.5	4.5	Yes	DHA	399-0181
HCU Anamix Junior LQ	125mL liquid bottle	1-10y	Neutral	L-amino acids	8	10	119	8.8	4.8	Yes	DHA	399-0181
HCU Lophlex LQ 10	62.5mL liquid pouch	4y-adult	Juicy Berries	L-amino acids	8	10	60	4.4	0.22	Yes	DHA	410-1655
HCU Lophlex LQ 20	125mL liquid pouch	4y-adult	Juicy Berries	L-amino acids	16	20	120	8.8	0.44	Yes	DHA	370-7544

HCU LV	27.8g sachet powder	8y-adult	Neutral Tropical	L-amino acids	72	20	371	1.4	0.19	Yes	None	313-0639
XMET Homidon	500g tin powder	0-adult	Neutral	L-amino acids	77	77	326	4.5	0	No	None	406-2220
HCU Maxamum	500g tin powder	8y-adult	Neutral	L-amino acids	39	39	10/25g powder	74/25g powder	8.5/25g powder	Yes	None	None
COMPANY: VITAFLO												
HCU Gel	24g sachet powder	6m-10y	Neutral	L-amino acids	41.7	10	81	10.3	0	Yes	None	364-4093
HCU Explore 5	12.5g sachet powder	6m-5y	Neutral	L-amino acids	40	5	43	5.3	0.2	Yes	DHA	413-1793
HCU Express 15	25g sachet powder	3y-adult	Neutral	L-amino acids	60	15	74	3.4	0	Yes	None	305-1836
HCU Express 20	35g sachet powder	3y-adult	Neutral	L-amino acids	60	20	101	4.7	0	Yes	None	369-3215
HCU Cooler 10	87mL liquid pouch	3y-adult	Red	L-amino acids	11.5	10	65	4.4	0.8	Yes	DHA	378-5896
HCU Cooler 15	130mL liquid pouch	3y-adult	Red Orange	L-amino acids	11.5	15	97	6.6	1.2	Yes	DHA	Red 355-9531 Orange

												339-7528
HCU Cooler 20	174mL liquid pouch	3y-adult	Red	L-amino acids	11.5	20	130	8.9	1.6	Yes	DHA	378-5888
Cystine supplementation may be required see – table 17												

MAPLE SYRUP URINE DISEASE (MSUD)

Background information
MSUD is a rare amino acid disorder with an inability to metabolise the amino acids: leucine, valine and isoleucine. Without attentive and appropriate dietary treatment, it can lead to irreversible brain damage and death.

Dietary treatment requires:

- a lifelong low protein diet
- supplementation with a protein substitute free of leucine, valine and isoleucine, usually supplemented with vitamins, minerals and DHA
- access to low protein special foods

Protein substitutes
Protein substitutes are essential by helping maintain metabolic control and providing essential nutrients.

Plasma valine and isoleucine may be low and this is rate limiting for the synthesis of leucine, therefore supplementation with valine and isoleucine is recommended.

Dosage of protein substitute:
The daily dose of protein equivalent from amino acid supplements prescribed/kg body weight/day (also considering natural protein tolerance) is:

- children 0-3y: 3g/kg/day
- children 4-6y: 2.5g/kg/day
- children 7-12y: 1.5-2g/kg/day
- over 12y and adults: 1-1.5g/kg/day up to a maximum 80g per day (unless extreme needs or athletes).
- pregnancy >70g/day total

Protein substitute is given three to four times per day.

Dosage of valine and isoleucine supplementation
The dose of valine and isoleucine supplementation is dependent on regular blood tests and is adjusted by the specialist dietitian.

Administration: orally or via nasogastric/gastrostomy tube, taken 3 or 4 times a day.

Pregnancy
Throughout pregnancy, dietary treatment must be sustained to protect the mother from metabolic decompensation during pregnancy and the post-partum period, and also to ensure adequate protein intake and overall nutrition to support foetal growth. ***Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential*.** In MSUD, peripartum and post-partum management involve continued use of specific dietary products.

EMERGENCY MANAGEMENT.
In illness or trauma blood leucine levels may rise rapidly and an emergency regimen is required to help manage this.

Ingredients added to emergency regimens include:
- amino acid supplements without leucine, valine, and isoleucine
- valine and isoleucine supplementation
- glucose polymer
- fat emulsion
- combined glucose/fat supplement

Dose: of each emergency product is determined by the age and weight of the patient.

Administration: orally or continuous feed via nasogastric or gastrostomy tube**.**

See BIMDG emergency guidelines for further information

Table 2

Product name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
MSUD Anamix infant	400g tin powder	0-3y	Neutral	L amino acids	13.1	2/ 100mL	70 / 100mL	7.5/ 100mL	3.5/ 100mL	Yes	DHA	344-8743
MSUD Anamix Junior	36g sachet powder	1-10y	Neutral	L- amino acids	28	10	135	11.5	4.5	Yes	DHA	395-0359
MSUD Anamix Junior LQ	125mL liquid bottle	1-10y	Orange	L- amino acids	8	10	118	8.8	4.8	Yes	DHA	350-7480
MSUD Lophlex LQ 20	125mL liquid pouch	4y-adult	Juicy Berries	L- amino acids	16	20	120	8.8	0.44	Yes	DHA	370-7536
MSUD Lophlex LQ 10	62.5mL liquid pouch	4y-adult	Juicy Berries	L-amino acids	16	10	60	4.4	0.22	Yes	DHA	410-1648
MSUD Maxamum	500g tin powder	8y-adult	Neutral Orange	L-amino acids	39	10/25g powder	74/25g powder	8.5/25g powder	0	Yes	None	Neutral 211-9204

												Orange 228-6722
*MSUD aid III	500g tin powder **5g unit**	0- adult	Neutral	L-amino acids	77	3.9	16	0.23	0	No	None	259-2079
COMPANY: VITAFLO UK												
MSUD gel	24g sachet powder	6m-10y	Neutral	L-amino acids	41.7	10	81	10.3	0	Yes	None	364-4119
MSUD explore 5	12.5g sachet powder	6m-5y	Neutral	L-amino acids	40	5	43	42	1.5	Yes	DHA ARA	413-1819
MSUD express 15	25g sachet powder	3y-adult	Neutral	L-amino acids	60	15	74	3.4	0	Yes	None	302-2571
MSUD express 20	35g sachet powder	3y- adult	Neutral	L-amino acids	60	20	101	4.7	0	Yes	None	369-3272
MSUD cooler 10	87mL liquid pouch	3y- adult	Red	L-amino acids	11.5	10	65	4.4	0.8	Yes	DHA	378-5854
MSUD cooler 15	130mL liquid pouch	3y- adult	Red Orange	L-amino acids	11.5	15	97	6.6	1.2	Yes	DHA	Red 355-9549 Orange 328-1797
MSUD cooler 20	174mL liquid pouch	3y- adult	Red	L-amino acids	11.5	20	130	8.9	1.6	Yes	DHA	378-5904

*MSUD amino 5	6g sachet powder	3y- adult	Neutral	L-amino acids	83	5	20	0	0	No	None	384-9320

*

- MSUD aid III/ MSUD amino acids are titrated to individual requirements in illness management.
- Valine, isoleucine 50 (Vitaflo Ltd) are routinely used to supplement dietary intake. See single amino acid supplement-table 9
- Valine 1000 or isoleucine 1000 (Vitaflo) are used for emergency management in MSUD. See single amino acid supplements-table 9

PHENYLKETONURIA (PKU)

Background information
This is a rare amino acid disorder (1 in 12,000) with an inability to metabolise the amino acid phenylalanine, leading to irreversible brain damage if untreated or if treatment is sub-optimal.

In PKU, the treatment is a lifelong low phenylalanine diet. It is the only proven treatment that prevents irreversible brain damage, significant delays in development, and hyperactive behaviour with autistic features in children. In adults it prevents neuropsychiatric co-morbidities and maternal PKU syndrome.

Dietary treatment requires:

- severe restriction of natural protein
- supplementation with low or phenylalanine free protein substitute usually supplemented with vitamins, minerals and DHA
- low protein special foods

Role of protein substitute
The protein substitutes are essential: not only do they supply essential amino acids; they help maintain metabolic control and usually provide a source of other nutrients. Poor adherence leads to loss of metabolic control. In patients with classical PKU, they are likely to provide 80% of protein requirements.

Dosage of protein substitute
The daily dose of protein equivalent from amino acid supplements prescribed /kg body weight/day (also considering natural protein tolerance) is:

- children 0-3 years: 3g/kg body weight/day
- children 4-6 years: 2.5g/kg body weight/day

- children 7-12 years: 1.5-2g/kg body weight/day
- over 12 years and adults: 1-1.5g/kg body weight/day usually to an upper amount of 80g day (unless extreme needs/athletes)
- pregnancy: >70g/day total

Protein substitutes are usually taken 3 or 4 times daily.

The dose and type of product should always be advised by the specialist dietitian according to the needs of the patient.

Administration of protein substitutes: oral.

Pregnancy

High blood phenylalanine levels during pregnancy have a teratogenic effect on the developing foetus that can result in growth retardation, microcephaly, intellectual disabilities, and birth defects, including congenital heart defects. It is particularly important that women maintain a very strict diet pre-conception and throughout pregnancy. ***Pregnancy may occur without planning with unsafe metabolic control on patient presentation. Regaining metabolic control is considered a matter of urgency, with strict dietary management being implemented immediately.*** Therefore, prescription products may be needed on an urgent basis having not been used recently/in adulthood.

Additional phenylalanine supplementation may be necessary during pregnancy.

Additional notes

There is a subsection of older adults with PKU that have stopped diet either because they were advised that it was safe in teenage years (in the 1970's -80's) or they could not tolerate the traditional amino acid products. More adults who have elected to stop dietary treatment are now presenting with high phenylalanine concentrations and symptoms such as headaches, tiredness, lack of concentration, low mood, anxiety, short term memory loss and executive function deficits. They have white matter changes on brain MRI scans concurrent with high brain phenylalanine concentrations. All adult patients are encouraged to return to a low phenylalanine diet.

Amino acid supplements have a poor taste and smell. It is necessary to have a wide choice of products available to find a single product that a patient may be able to tolerate. Please note inability to take protein substitutes or lack of access to supply could lead to irreversible brain damage.

Table 3

Name	Unit size	Age	Flavour	Protein source	Protein g per 100 g per 100mL	Protein g per unit size	Phe content mg per unit size	Energy kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs Novel ingredients	ACBS listed PIP code
COMPANY: CAMBROOKE THERAPEUTICS													
Glytactin Bettermilk 15 Original	49g sachet powder	3y-adult	Neutral	CGMP	31	15	23	160	23	4.5	Yes	DHA Probiotics	404-3378
Glytactin Bettermilk 15	52g sachet powder	3y-adult	Strawberry Crème Orange Crème	CGMP	29	15	24	200	26	3.9	Yes	DHA Probiotics	Strawberry Crème 406-9563 Orange Crème 406-9555
Glytactin Build 10	16g sachet powder	3y-adult	Neutral	CGMP	67	10	10	50	0.4	0.6	Yes	DHA Probiotics	407-0660
Glytactin Build 20/20	30g sachet powder	3y-adult	Neutral	CGMP	67	20	20	100	0.7	1.3	Yes	DHA Probiotics	414-0828
Glytactin Bettermilk Lite 20	46g sachet powder	3y-adult	Neutral	CGMP	43	20	35	150	12	2	Yes	DHA Probiotics	407-0389

Glytactin Complete 15 Fruit Frenzy	81g bar	3y-adult	Fruity taste chewy bar	CGMP	19	15	32	330	35	12	Yes	No	407-0348
Glytactin Complete 15 Peanut Butter	81g bar	3y-adult	Peanut butter taste chewy bar	CGMP	19	15	37	320	42	8	Yes	No	407-0355
Glytactin RTD 10	250mL liquid carton	3y-adult	Original Chocolate	CGMP	4g/100mL	10	18	153	21 22	3.5	Yes	No	Original 404-3287 Chocolate 404-3279
Glytactin RTD 15	250 mL liquid carton	3y-adult	Original, Chocolate	CGMP	6g/100mL	15	27	200	23	5	Yes	No	Original 404-3303 Chocolate 404-3303
Glytactin Restore Powder 5	20g sachet powder	3y-adult	Orange Berry	CGMP	25	5	9	73	14	0	No Incomplete	No	Orange 407-0363 Berry 407-0371
Glytactin Restore Lite Powder 10 Orange	19g sachet powder	3y-adult	Orange	CGMP	53	10	18	65	6.4	0	No Incomplete	No	407-0652
Glytactin Restore Lite Powder 20 Orange	38g sachet powder	3y-adult	Orange	CGMP	53	20	36	130	12.8	0	No Incomplete	No	407-0819

All nutritional analysis is for *neutral* products.

Name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 100mL	Protein g per unit size	Phe content mg per unit size	Energy kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs novel ingredients	ACBS listed PIP code
COMPANY: GALEN													
PKU Easy Liquid	130mL liquid pouch	3y-adult	Orange Citrus Mixed Berries	L-amino acids	11.5g/ 100mL	15	0	87	5.5	0.5	Yes	DHA EPA	Berry 407-6329 Orange 407-6311
PKU Easy Shake and Go	34g powder bottle	3y-adult	Orange	L-amino acids	45	15	0	125	14	0.1	Yes	No	407-6303
PKU Easy Microtabs	110g tablets bottle	8y-adult	Neutral	L-amino acids	70.8	10g/ 14g tablets	0	55Kcal/ 14g tablets	1.8/ 14g tablets	0.5/ 14g tablets	No	No	414-6288
PKU Go	20g powder	9m-10 y	Neutral	L-amino acids	50	10	0	65	5	>0.5	Yes	No	413-9978

All nutritional analysis is for *neutral* products.

Name	Unit size	Age	Flavour	Protein source	Protein g/100 g	Protein g per unit size	Phe content mg per unit size	Energy kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs novel ingredients	ACBS listed PIP code
COMPANY: METAX													
XPHE Jump 10	63mL liquid pouch	3y-adult	Neutral Cola (without caffeine) Orange Wild berry Vanilla Tropical	L-amino acids	16g/ 100mL	10 10	0 0	56 63	3.6 5	0.5 0.3	Yes	DHA EPA (except Neutral)	Neutral 397-8418 Cola 397-8384 Orange 397-8392 Wild berry 397-840
XPHE Jump 20	125mL liquid pouch	3y-adult	Neutral Cola (without caffeine) Orange Wild berry	L-amino acids	16g/ 100mL	20 20	0 0	111 126	7 10	0 0.6	Yes	DHA EPA (except Neutral)	Neutral 400-2614 Cola 400-2648 Orange 400-2630 Wild Berry 400-2622

All nutritional analysis is for neutral products.

Name	Unit size	Age	Flavour	Protein source	Protein g/100 g	Protein g per unit size	Phe content mg per unit size	Energy kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs novel ingredients	ACBS listed PIP code
COMPANY: MEVALIA													
PKU Motion 10	70mL liquid pouch	3y-adult	Red fruits	L-amino acids	14g/ 100 mL	10	0	56	3.9	0	Yes	0	413–7212
PKU Motion 20	140mL liquid pouch	3y-adult	Red fruits	L-amino acids	14g/ 100 mL	20	0	111	7.7	0	Yes	0	413–7220
PKU Motion 10	70mL liquid pouch	3y-adult	Tropical	L-amino acids	14g/ 100 mL	10	0	54	3.5	0	Yes	0	413–7204
PKU Motion 20	140mL liquid pouch	3y adult	Tropical	L-amino acids	14g/ 100 mL	20	0	108	7.0	0	Yes	0	413–7238
PKU GMPower	23.4g sachet powder	3y adult	Vanilla	CGMP	43	10	14	75	7.4	0.4	Yes	DHA GOS FOS	414-7245

All nutritional analysis is for *neutral* products.

Name	Unit size	Age	Flavour	Protein source	Protein g pre100 g per 100mL	Protein g per unit size	Phe content mg per unit size	Energy kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA													
PKU Anamix Infant	400g tin powder	From birth	Neutral	L-amino acids	13.1	2.0/ 100mL	0	70/ 100mL	7.5/ 100 mL	3.5/ 100mL	Yes	DHA, AA Prebiotics	344-8735
PKU Anamix First Spoon	12.5g sachet powder	6m-5y	Neutral	L-amino acids	40	5	0	41	4.8	0.2	Yes	DHA AA	357-8895
PKU Anamix Junior LQ	125mL liquid bottle	1-10y	Orange Berry	L-amino acids	8/ 100 mL	10	0	118	8.8	4.8	Yes	DHA	Orange 322-0621 Berry 315-7336
PKU Anamix Junior	36g sachet powder	1-10y	Orange Berry Vanilla Chocolate Neutral	L-amino acids	28	10	0	135	11.5	4.5	Yes	DHA	Orange 395-0326 Berry 395-0300 Vanilla 395-0342 Chocolate 395-0318 Neutral 395-0334
PKU Lophlex LQ 10	62.5mL liquid	4y-adult	Juicy Orange	L-amino acids	16/ 100 mL	10	0	60	4.4	0.2	Yes	DHA	Juicy orange 370-0374

	pouch		Juicy Berry Juicy Tropical Juicy Citrus Berry										Juicy berry 370-0382 Juicy tropical 344-8669 Juicy citrus 344-8644 Berry 315-7336
PKU Lophlex LQ 20	125mL liquid pouch	4y-adult	Juicy Orange Juicy Berry Juicy Tropical Juicy Citrus Berry Orange	L-amino acids	16/ 100 mL	20	0	120	8.8	0.4	Yes	DHA	Juicy orange 370-0390 Juicy berry 370-0408 Juicy tropical 344-8818 Juicy citrus 322-0613 Berry 322-0639 Orange 322-0621
PKU Lophlex Powder	27.8g sachet powder	8y-adult	Berry Orange Neutral	L-amino acids	72	20	0	91	2.5	0.06	Yes	No	Orange 313-6777 Berry 313-6769 Neutral 313-6751
PKU Lophlex Sensation	109g Semi-solid pot	4y-adult	Berries	L-amino acids	18.3	20	0	166	20.2	0.4	Yes	DHA	Berry 377-1128
PKU GMPro	33.3g sachet	3y - adult	Vanilla	CGMP	30	10	18	128	12.5	3.9	Yes	DHA	409-5261

	powder												
PKU GMPro LQ	250 mL liquid pouch	3y-adult	Vanilla	CGMP	4	10	18	112	8.5	4	Yes	DHA EPA	413-3773
PKU Synergy	33g sachet powder	10y-adult	Citrus	L-amino acids	60.6	20	<5	98	3.5	0.33	Yes	DHA	409-5253
Phlexy 10 Drink mix	20g sachet powder	8y-adult	Apple and Blackcurrant Citrus burst Tropical Surprise	L-amino acids	42	8.33	0	69	8.8	0	No	No	Apple B/currant 210-5427 Citrus 291-2822 Tropical 291-2830
Phlexy 10 tablets	75 tablets/ tub	8y-adult	Neutral	L-amino acids	58	10/12 tablets	0/12 tablets	54/12 tablets	0.78/ 12 tablets	0.24/12 tablets	No	No	290-3615
PK Aid 4	500g tin powder	0-adult	Neutral	L-amino acids	79	79 / 100 g	0	334/ 100g	4.5/ 100g	0	No	No	240-3988
Easiphen	250mL liquid carton	8y-adult	Forrest Berries	L-amino acids	9.7/ 100mL	16.8/ 250 mL	0	163	12.8	5	Yes	No	306-6008
PKU Maxamum	50g sachets powder 500g tin	8y-adult	Orange Neutral	L-amino acids	39	19.5/ sachet	0	149	17	<0.3	Yes	No	Orange (500g) 013-0443 Neutral (500g)

													340-4043 Orange (50g sachet) 303-5995 Neutral (50g sachet) 340-4043

All nutritional analysis is for *neutral* products.

Name	**Unit size**	**Age**	**Flavour**	**Protein source**	**Protein g per 100 g per 100mL**	**Protein g per unit size**	**Phe content mg per unit size**	**Energy kcal per unit size**	**CHO g per unit size**	**Fat g per unit size**	**Vitamins and minerals supplemented to provide complete profile**	**LCPUFAs novel ingredients**	**ACBS listed PIP code**
COMPANY: VITAFLO UK													
PKU Start	400g tin powder	From birth	Neutral	L-amino acids	14.3	2.0/ 100mL	0/ 100mL	68/ 100 mL	7.2/ 100 mL	3.5/ 100mL	Yes	DHA AA	406-9530
PKU Explore 5	12.5 g sachet powder	6m-5y	Neutral	L-amino acids	40	5	0	43	5.3	0.2	Yes	DHA, AA	409-5865
PKU Explore 10	25g sachet powder	1-5y	Orange Red	L-amino acids	40	10	0	83	9.8	0.4	Yes	DHA AA	Orange 409-5857 Red 409-5840
PKU Gel	24g sachet powder	Neutral 6m-10y Flavoured 1-10y	Neutral Orange Red	L-amino acids	41.7 41.7	10 10	0 0	81 76	10.3 8.9	0.02	Yes	No	Neutral 364-4069 Orange 364-4085

													Red 3644-077
PKU Squeezie	85g Semi-solid pouch	6m-10y	Apple and Banana	L-amino acids	12	10	0	135	22.5	0.5	Yes	DHA AA	363-6628
PKU Express 15	25g sachet powder	3y-adult	Neutral Lemon 0range Tropical	L-amino acids	60 60	15 15	0 0	70 74	2.4 3.4	0.05	Yes	No	Neutral 289-7007 Lemon 289-7015 Orange, 289-7023 Tropical 314-2809
PKU Express 20	34g sachet powder	3y-adult	Neutral Lemon Orange Tropical	L-amino acids	60 60	20 20	0 0	95 101	3.3 4.7	0.07	Yes	No	Neutral 369-3256 Lemon 369-3231 Orange 369-3223 Tropical 369-3249
PKU Cooler 10	87mL liquid pouch	3y-adult	Orange Purple Red White Yellow	L-amino acids	11.5/ 100 mL	10	0	65	4.4	0.8	Yes	DHA	Orange 325-6930 Purple 325-6948 Red 347-7718 White 333-8407 Yellow

													404-0085
PKU Cooler 15	130mL liquid pouch	3y-adult	Orange Purple Red White Yellow	L-amino acids	11.5/ 100mL	15	0	97	6.6	1.2	Yes	DHA	Orange 315-8482 Purple 315-8490 Red 347-9565 White 333-0768 Yellow 403-8386
PKU Cooler 20	174mL liquid pouch	3y-adult	Orange Purple Red White Yellow	L-amino acids	11.5/ 100mL	20	0	130	8.9	1.6	Yes	DHA	Orange 324-4753 Purple 324-4761 Red 347-7700 White 333-0776 Yellow 404-0002
PKU Air 15	130mL liquid pouch	3y-adult	**Red** (berry blast) **White** (Caribbean crush) **Green** (citrus twist) **Gold** (coffee	L-amino acids	11.5/ 100mL	15	0	75	2.0	0.8	Yes	DHA	Red 399-6394 White 399-6402 Green 388-5951 Gold 388-5969 Yellow 403-7776

			fusion contains <20 mg of caffeine per pouch) **Yellow** (mango breeze)										
PKU Air 20	174mL liquid pouch	3y-adult	**Red** (berry blast) **White** (Caribbean crush) **Green** (citrus twist) **Gold** (coffee fusion contains <20 mg of caffeine per pouch) **Yellow** (mango breeze)	L-amino acids	11.5/ 100mL	20	0	100	2.6	1.0	Yes	DHA	Red 399-6428 White 399-6436 Green 388-5977 Gold 388-5985 Yellow 403-9996
PKU sphere 15	27g sachet powder	4y-adult	Red Berry Vanilla Chocolate	CGMP	56	15	28	91	4.9	1.3	Yes	DHA	Red Berry 404-8005 Vanilla 404-8161

													Chocolate 409-7465
PKU sphere 20	35g sachet powder	4y-adult	Red berry Vanilla Chocolate	CGMP	56	20	36	120	6.3	1.6	Yes	DHA	Red Berry 404-0010 Vanilla 404-0028 Chocolate 409-7473

All nutritional analysis is for *neutral* products.

TYROSINAEMIA

Background information
There are three types of tyrosinaemia HTI, II and III depending on the enzyme deficiency in the tyrosine pathway. They are all rare amino acid disorders.

Tyrosinaemia type I is treated with a drug NTBC (Nitisinone), preventing hepatocellular carcinoma by blocking the pathway and production of toxic compounds. A low tyrosine diet is essential to prevent hypertyrosinaemia, which can result in ocular damage.

Tyrosinaemia type II is due to an enzyme deficiency further up the tyrosine pathway. It is not associated with hepatic carcinoma but high tyrosine concentrations lead to ocular lesions and photophobia, skin lesions or neurological complications may also be present.

Tyrosinaemia type III, only a few cases have been described. Clinical presentation is variable, neurological and intellectual disabilities have been described in some cases.

Dietary treatment for Tyrosinaemia type I and II requires:

- a lifelong low protein diet
- supplementation with a phenylalanine/tyrosine free protein substitute, usually supplemented with vitamins, minerals and DHA
- access to low protein special food

Dietary treatment for Tyrosinaemia type III

- as for Tyrosinaemia type I and II but diet may be relaxed in early childhood

Protein substitutes
Protein substitutes are essential by helping maintain metabolic control and providing essential nutrients.

Dosage of protein substitute

The daily dose of protein equivalent from amino acid supplements prescribed/kg body weight/day (also considering natural protein tolerance) is:

- children 0-3y: 3g/kg/day
- children 4-6y: 2.5g/kg/day
- children 7-12y: 1.5-2g/kg/day
- over 12y and adults: 1-1.5g/kg/day up to a maximum 80g per day (unless extreme needs or athletes)
- pregnancy >70g/day total

Protein substitutes are given three to four times daily.

Administration: orally or via nasogastric/gastrostomy tube.

Blood phenylalanine particularly in infancy may be low and require phenylalanine supplementation. The dose is dependent on regular blood tests and is adjusted by the specialist IMD dietitian.

Pregnancy

Dietary treatment must be sustained throughout pregnancy to protect both the mother and developing baby from hypertyrosinaemia, and to ensure adequate protein intake and overall nutrition to support foetal growth. ***Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential.***

Alkaptonuria (AKU)

Alkaptonuria is a rare disorder of the tyrosine pathway, rarely developing in childhood but more commonly in adults, leading to a multisystem disease affecting musculoskeletal and cardiac systems. NHS England approved the *off label* use of Nitisinone (NTBC) for the treatment of adult AKU to prevent disease progression. NTBC increases blood tyrosine concentrations. Patients on NTBC treatment require a low tyrosine diet using a phenylalanine/tyrosine free protein substitute and low protein diet.

Table 4

Name	Unit size	Age	Flavour	Source of protein	Protein g per 100g per 100ml	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
Tyr Anamix Infant	400g tin powder	0-12m	Neutral	L-amino acids	13.1	2/100mL	70/100mL	7.5 /100mL	3.5 /100mL	Yes	DHA Prebiotics	344-8768
Tyr Anamix Junior	36g sachet powder	1-10y	Neutral	L-amino acids	28	10	135	11.5	4.5	Yes	DHA	395-0367
Tyr Anamix Junior LQ	125mL liquid bottle	1-10y	Orange	L-amino acids	8	10	119	8.8	4.8	Yes	DHA	351-8677
Tyr Lophlex LQ 10	62.5mL liquid pouch	4y-adult	Juicy Berries	L-amino acids	16	10	60	4.4	0.2	Yes	DHA	410-1697
Tyr Lophlex LQ 20	125mL liquid pouch	4y-adult	Juicy Berries	L-amino acids	16	20	120	8.8	0.4	Yes	DHA	370-7528
Tyr Maxamum	500g tin powder	8y-adult	Neutral	L-amino acids	39	10/25g powder	74/25g powder	8.5/25g powder	0	Yes	None	272-5208
X Phen Tyr Tyrosidon	500g tin powder	0-adult	Neutral	L-amino acids	77	10g/13g powder	42/13g powder	0.6/13g powder	0	No	None	406-2246
COMPANY: VITAFLO UK												

Tyr gel	24g sachet powder	6m-10y	Neutral	L-amino acids	41.7	10	81	10.3	0.02	Yes	None	364-4101
Tyr explore 5	12.5g sachet powder	6m-5y	Neutral	L-amino acids	40	5	43	5.3	0.2	Yes	DHA ARA	413-1785
Tyr express 15	25g sachet powder	3y-adult	Neutral	L-amino acids	60	15	74	3.4	0.1	Yes	None	322-0647
Tyr express 20	34g sachet powder	3y-adult	Neutral	L-amino acids	60	20	101	4.7	0.1	Yes	None	369-3264
Tyr cooler 10	87mL liquid pouch	3y-adult	Red	L-amino acids	11.5	10	65	4.4	0.8	Yes	DHA	378-5870
Tyr cooler 15	130mL liquid pouch	3y-adult	Red Orange	L-amino acids	11.5	15	97	6.6	1.2	Yes	DHA	319-0089
Tyr cooler 20	174mL liquid pouch	3y-adult	Red	L-amino acids	11.5	20	130	8.9	1.6	Yes	DHA	378-5862
COMPANY: GALEN												
Tyr Easy shake and go	34g powder bottle	3y-adult	Orange	L-amino acids	45	15	123	13	0	Yes	None	414-6429
Supplementation with phenylalanine may be required see single amino acid supplements table 9.												

ORGANIC ACIDURIAS

GLUTARIC ACIDURIA TYPE 1

Background information

Glutaric aciduria Type 1 (GA1) is a rare organic acidaemia. The enzyme deficiency causes an inability to metabolise the amino acids lysine and tryptophan. Without appropriate dietary treatment, severe movement problems, irreversible brain damage and death may result. The aim of dietary treatment is to avoid a metabolic crisis by limiting lysine and tryptophan intake, and to promote normal growth and development.

Dietary treatment requires:

- strict low protein diet (commonly less than 1g/kg body weight/day)
- lysine free/low tryptophan protein substitute
- vitamins, minerals and DHA supplements if not added to the protein substitute
- energy supplements to support weight gain, growth and anabolism
- access to low protein foods

Protein substitutes

Protein substitutes maintain metabolic control, preventing neurological crisis by limiting entry of lysine at the blood brain barrier. Poor adherence or lack of access to protein substitute could lead to an encephalopathic crisis and severe neurological damage.

Dosage of protein substitute

The daily dose of protein equivalent from amino acid supplements prescribed/kg body weight/day (also considering natural protein tolerance) is:

- children 0-6m: 0.8-1.3g/kg/day
- children 7-12m: 0.8-1g/kg/day
- children 1-6 y: 0.8g/kg/day
- pregnancy>70g/day total

Protein substitute is given three to four times per day.

Administration: orally or via nasogastric/gastrostomy tube.

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential (particularly protein substitutes). Pregnancy may occur with no dietary treatment in place. However, in the second and third trimesters and during the peri- and post-partum period dietary prescription products may be required even if they have not been used before in adulthood.

EMERGENCY MANAGEMENT

An **emergency regimen** during illness is essential. Catabolism of lysine could cause irreversible brain damage (encephalopathic crisis). The emergency regimen is based on:

- glucose polymer
- **lysine-free, low tryptophan amino acid supplements**
- ± fat emulsion

The emergency feed is administered orally or via a nasogastric/gastrostomy tube at home or hospital. It is important that a glucose and ± fat source together with the lysine free low tryptophan amino acid supplement is administered. The specialist dietitian will determine the dose and frequency of administration.

See BIMDG emergency guidelines for further information.

Table 5

Name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
GA 1 Anamix Infant	400g tin powder	0-12m	Neutral	L-amino acids	13.1	2/ 100mL	70/ 100mL	7.5/ 100mL	3.5/ 100mL	Yes	DHA Prebiotic fibres	344-8800
GA Anamix Junior	18g sachet powder	1-10y	Neutral	L-amino acids	28	5	66	5.4	2.3	Yes	DHA	399-0173
GA1 Maxamum	500g tin powder	8y-adult	Neutral	L-amino acids	39	7.8/20g	59/20g	6.8/20g	0	Yes	None	Not ACBS prescribed
*XLysTry Glutaridon	500g tin powder	0-adult	Neutral	L-amino acids	79	4/5g powder	17/5g powder	0.2/5g powder	0	No	None	343-9874
Flavour sachets to flavour the neutral taste of protein substitute - see table 18												
COMPANY: VITAFLO UK												
GA gel	24g sachet powder	6m-10y	Neutral	L-amino acids	41.7	10	81	10.3	0.02	Yes	None	364-9296
GA explore 5	12.5g sachet powder	6m-5y	Neutral	L-amino acids	40	5	43	5.3	0.2	Yes	DHA AA	413-1801
GA Express	25g	3y-	Neutral	L-amino	60	15	74	3.4	0	Yes	None	Not ACBS

	sachet powder	adult		acids								prescribed
*GA amino 5	6g sachet powder	0-adult	Neutral	L-amino acids	83	5	20	0	0	No	None	384-9304
* Used with energy supplements - see table 16 Flavour sachets to flavour the neutral taste of protein substitute - see table 18												

METHYLMALONIC ACIDAEMIA AND PROPIONIC ACIDAEMIA (MMA/PA)

Background information

Methylmalonic and propionic acidaemias are rare inborn errors of metabolism commonly grouped as organic acidaemias. Both conditions have an inability to metabolise methionine, valine, threonine and isoleucine, and left untreated result in the accumulation of toxic metabolites.

Dietary treatment

The conditions are treated by a low protein diet. A methionine, valine, threonine and isoleucine free amino acid infant feed may be used in infancy to supplement protein and to aid growth. It may also be used in older children/adults if metabolic control is poor or safe levels of natural protein intake meeting WHO/FAO/UNU 2007 recommendations are not tolerated. Many children and adults need tube-feeding to meet target energy and protein requirements.

Protein substitutes

Infancy An infant formula without the precursor amino acids valine, leucine, methionine and isoleucine may be given. This is used in combination with a reduced and controlled intake of breast milk or normal infant formula. These special formulas are usually complete in vitamins and minerals and all the other amino acids to support growth.
Alternatively, a protein free infant feed formula containing fat, carbohydrate, vitamins and minerals is given in combination with a measured intake of breast milk or normal infant formula to control and lower protein intake.

Children/Adults/Pregnancy: An amino acid supplement free from the precursor amino acids valine, leucine, methionine and isoleucine may be used if safe levels of protein intake defined by WHO/FAO/UNU 2207 are not tolerated. To achieve adequate energy requirements, energy supplements may be recommended (see section *energy supplements*)

Dosage of protein substitute/precursor free amino acids

The dose is dependent on the age and natural protein tolerance. The protein substitute would usually provide ≤ 20% of total protein requirements. It is equally distributed throughout the day.

Administration: orally or via nasogastric or gastrostomy tube.

EMERGENCY REGIMENS

See BIMDG emergency guidelines for further information.

Table 6

Name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 10mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
MMA/PA Anamix Infant	400g tin powder	0-12m	Neutral	L-amino acids	13.1	2/ 100mL	70/ 100mL	7.5/ 100mL	3.5/ 100mL	Yes	DHA	344-8750
MMA/PA Anamix Junior	18g sachet powder	1-10y	Neutral	L-amino acids	28	5	66	2.3	12.5	Yes	DHA	399-0199
XMTVI Asadon	200g tin powder	0-adult	Neutral	L-amino acids	77	3.9/5g powder	16/5g powder	0.2	0	No	None	406-2170
MMA/PA Maxamum	500g tin powder	8y-adult	Neutral	L-amino acids	39	7.8/20g powder	59/20g powder	6.8/20g powder	0	Yes	None	212-0863
COMPANY: VITAFLO UK												
MMA/PA	12.5g	6m-5y	Neutral	L-amino	40	5	43	5.3	0.2	Yes	DHA	Non ACBS

explore 5	sachet powder			acids							AA	
MMA/PA gel	24g sachet powder	6m10y	Neutral	L-amino acids	41.7	10	81	10.3	0	Yes	None	Non ACBS
MMA/PA express	25g sachet powder	3y adult	Neutral	L-amino acids	60	15	74	3.4	0	Yes	None	Non ACBS
MMA/PA Cooler	130mL liquid pouch	3y-adult	Red	L-amino acids	11.5	15	97	6.6	1.2	Yes	DHA	Non ACBS
MMA/PA amino 5	6g sachet powder	3y-adult	Neutral	L-amino acids	83	5	20	0	0	No	None	384-3912
Energy supplements A range of energy supplements with or without fat, vitamins and minerals are used to support nutritional requirements in these conditions - see table 16												

ISOVALERIC ACIDAEMIA (IVA)

Background information
IVA is due to an enzyme deficiency in the leucine pathway and may present either acutely in the neonatal period or as a chronic presentation later in childhood. In the neonatal form symptoms appear in the first few weeks associated with metabolic acidosis, while in the chronic form non-specific symptoms of faltering growth and developmental delay occur.

IVA is diagnosed as part of the newborn screening programme. Some infants may have a milder phenotype, which does not require dietary restriction, but an emergency regimen in illness is required.

Dietary treatment
IVA is treated by a low protein diet. A leucine free amino acid infant feed may be used in infancy. In older children/adults, a leucine-free amino acid supplement may be used if metabolic control is poor or if safe levels of natural protein intake, based on WHO/FAO/UNU 2007 recommendations are not tolerated. Some patients need tube-feeding to meet target energy and protein requirements.

Protein substitutes
Infancy: If recommended, an infant formula without the precursor amino acid leucine may be given. This is used in combination with a lower and controlled intake of breast milk or normal infant formula.

Alternatively, a protein free infant formula containing fat, carbohydrate, vitamins and minerals is used in combination with a lower and controlled intake of breast milk or normal infant formula.

Children/Adults/Pregnancy: An amino acid supplement without the precursor amino acid leucine may be used if WHO/FAO/UNU 2207 recommendations for safe levels of protein intake are not tolerated. In order to achieve adequate energy requirements, energy supplements

may be recommended (*energy supplements table 16*).

Dosage of protein substitute

The dose is dependent on the age and natural protein tolerance. Protein substitute usually provides ≤ 20% of total protein requirements. It is equally distributed throughout the day.

Administration: orally or via nasogastric or gastrostomy tube.

EMERGENCY REGIMEN

See BIMDG emergency guidelines for further information.

Table 7

Name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
IVA Anamix Infant	400g tin powder	0-12m	Neutral	L-amino acids	13.1/ 100mL	2/ 100mL	70/ 100mL	7.5/ 100mL	3.5/ 100mL	Yes	LCPFA DHA Prebiotic fibres	344-8792
IVA Anamix Junior	400g tin powder	1-10y	Neutral	L-amino acids	28	10/36g powder	132/36g powder	10.8/36g powder	4.4/36g powder	Yes	DHA	398-9480
COMPANY: VITAFLO UK												
IVA Cooler 15	130mL liquid pouch	3y-adult	Red	L-amino acids	11.5	15	97	6.6	1.2	Yes	DHA	Not ACBS prescribed
Energy supplements A range of energy supplements with or without fat, vitamins and minerals are used to support nutritional requirements in these conditions – see table 16												

PYRIDOXINE DEPENDENT EPILEPSY (PDE)/ ANTIQUITIN DEFICIENCY

Background information

Pyridoxine or vitamin B6 is activated to pyridoxal 5 phosphate (PLP) and is one of the most abundant cofactors for cellular neurotransmitter metabolism. PDE leads to reduced availability of PLP resulting in seizures, which are not responsive to anticonvulsant medication. Antiquitin is involved in the cerebral lysine degradation, and restricting dietary lysine is a treatment option together with B6 supplementation. A low lysine diet similar to the treatment of GA1 is recommended (***see GA1 dietary treatment).***

Dietary treatment requires:

- strict low protein diet
- lysine free protein substitute (using protein substitutes designed for GA1 as no specific PDE protein substitute available)
- vitamins, minerals and DHA supplements if not added to protein substitute
- energy supplements to support weight gain, growth and anabolism
- access to low protein special foods

Protein substitutes

Protein substitutes are lysine free and low in tryptophan.

Dosage of protein substitute

The daily dose of protein equivalent from amino acid supplement prescribed/kg body weight/day (also considering natural protein tolerance) is:

- children 0-6m: 0.8-1.3g/kg/day
- children 7-12m: 0.8-1g/kg/day
- children 1-6 y: 0.8g/kg/day
- children 7-12y: total protein equivalent intake: 1.5-2g/kg body weight/day
- over 12y and adults: total protein equivalent: 1-1.5g/kg body weight/day usually to an upper amount of 80g day

UREA CYCLE DISORDERS

Background information
Urea Cycle Disorders (UCD) are rare disorders of protein metabolism, in which there is reduced function of the urea cycle to metabolise waste nitrogen to urea. Without attentive and appropriate medical and dietary treatment it can lead to irreversible brain damage and death.

Dietary treatment requires:

- a lifelong low protein diet
- essential amino acid supplementation
- vitamins, minerals and trace element supplementation
- use of low protein special foods
- use of energy supplements

Essential amino acids
Essential amino acid supplements are a key aspect of dietary management to maintain metabolic control, provide essential nutrients and prevent protein malnutrition. The dose of protein equivalent from amino acid supplements are titrated based on weight and clinical need.

Children/Adults/Pregnancy: Essential amino acid supplements are used if WHO/FAO/UNU 2007 recommendations for safe levels of protein intakes are not tolerated.

Essential amino acids: dose is dependent on weight, biochemical, clinical and dietary evaluation and are adjusted by the specialist IMD dietitian/clinician.

Essential amino acids are given at least three to four times per day.

Administration: orally or via nasogastric/gastrostomy tube.

Energy supplements

A range of energy supplements may be required depending on nutritional status and clinical need. These may be protein free formulations providing energy, fat, vitamin and minerals or individual carbohydrate and fat-based supplements.

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential. Pregnancy may occur with no dietary treatment in place. However, regaining metabolic control is considered a matter of urgency, thus prescription products may be needed on an urgent basis having not been used before/in adulthood. In some conditions peripartum and post-partum management involve continued use of specific dietary products such as glucose polymers.

EMERGENCY REGIMEN

See BIMDG emergency guidelines for further information.

Table 8

Name	Unit size	Age	Flavour	Source of protein	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFAs Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
Dialamine	400g tin powder	6m-adult	Orange	L – amino acids	25	4.3	62	11.2	0	No	None	326-3845
Essential Amino Acid Mix	200g tin powder **Dose 5g**	0-adult	Neutral	L- amino acids	79	4g /5g powder	16Kcal/5g powder	0	0	No	None	Not ACBS prescribed
COMPANY: VITAFLO UK												
UCD amino 5	6.6g sachet powder	3y-adult	Neutral	L-amino acids	75	5.0	20	0	0	No	None	384-9361
EAA Supplement	12.5g sachet powder	3y-adult	Tropical	L-amino acids	40	5g/sachet	36 Kcal /sachet	4g/ sachet	0	No	None	338-4724
Energy supplements A range of energy supplements with or without fat, vitamins and minerals are used to support nutritional requirements in these conditions – see table 16												

OTHER CONDITIONS NEEDING PROTEIN RESTRICTION

3 HYDROXY 3 METHYLGLUTARYL HMG-COA LYASE DEFICIENCY

Background information

A rare inherited metabolic disorder presenting with hypoglycaemia, acidosis and reduced consciousness. The enzyme HMG CoA lyase plays an essential role in breaking down dietary proteins and fats for energy. The enzyme is responsible for processing leucine and its deficiency causes reduced ketogenesis and accumulation of toxic leucine metabolites. Some patients may need overnight tube feeding to prevent hypoglycaemia due to limited fasting tolerance.

Dietary treatment requires:

- a low protein diet
- moderate fat restriction
- energy supplementation
- vitamin and mineral supplementation
- low protein special foods
- emergency regimen in illness

Administration: orally or via a nasogastric or gastrostomy tube.

EMERGENCY MANAGEMENT

See BIMDG emergency guidelines for further information.

GYRATE ATROPHY / ORNITHINE AMINOTRANSFERASE DEFICIENCY

Background information

Gyrate atrophy is a rare inborn error of ornithine metabolism, resulting in hyperornithinaemia due to ornithine aminotransferase deficiency. It causes myopia and night blindness, resulting in worsening visual acuity or blindness between 45 to 65 years of age.

Dietary treatment aims to reduce ornithine concentrations by a low protein diet restricting arginine, the precursor of ornithine.

Dietary treatment requires:

- a low protein diet
- essential amino acids supplements
- vitamin and mineral supplementation
- low protein special foods
- energy supplements to meet any energy deficit for growth, avoid weight loss and catabolism

Essential amino acids

These help achieve protein requirements and prevent amino acid deficiency associated with severe restriction of natural protein.

Dose of essential amino acids: this will be dependent on the severity of protein restriction and patient age.

Administration orally or nasogastric/ gastrostomy tube.

Products used in the treatment of gyrate atrophy are found in tables 16,17 and 8

Energy supplements - see table 16
Vitamin and mineral/ essential fatty acid supplements - see table 17
*Essential amino acid supplement*s - see table 8

SINGLE DOSE L- AMINO ACIDS

Background information
Some inherited metabolic disorders require additional supplementation with one or more single L-amino acids. This is due to the risk of deficiency due to an impaired metabolism.

Examples are provided below of single dose amino acids that may need to be supplemented in specific conditions:

- phenylalanine supplementation in HT1
- isoleucine and valine supplementation in MSUD
- cystine supplementation in HCU

The dose is dependent on regular blood tests and is adjusted by the specialist IMD dietitian.

Administration: orally or via nasogastric/gastrostomy tube.

Emergency management: in MSUD, during illness the dose of valine and isoleucine may increase; the dose is adjusted by a specialist IMD dietitian.

Table 9

Name	Unit size	Age	Flavour	Source of protein	Protein g / 100g	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
L-alanine	100g tub powder	0-adult	Neutral	L-amino acids	79.7	4/5g powder	319	0	0	Incomplete profile	None	Not ACBS prescribed
L-arginine	100g tub powder	0-adult	Neutral	L-amino acids	89.7	4.5/5g powder	359	0	0	Incomplete profile	None	Not ACBS prescribed
L-carnitine*	100g tub powder	0-adult	Neutral	L-amino acids	89.7	4.5/5g powder	359	0	0	Incomplete profile	None	Not ACBS prescribed
L-carnitine*	1g sachet	0-adult	Neutral	L-amino acids	89.7	4.5/5g powder	359	0	0	Incomplete profile	None	Not ACBS prescribed
L-citrulline	100g tub powder	0-adult	Neutral	L-amino acids	89.7	4.5/5g powder	359	0	0	Incomplete profile	None	Not ACBS prescribed
L-cystine	100g tub powder	0-adult	Neutral	L-amino acids	92.5	5.1/6g powder	370	0	0	Incomplete profile	None	Not ACBS prescribed
L-glutamine (Adamin G)	5g sachet powder	0-adult	Neutral	L-amino acids	87.7	4.4gsachet	17.6	0	0	Incomplete profile	None	Not ACBS prescribed
Glycine	100g tub powder	0-adult	Neutral	L-amino acids	76	3.8/5g powder	304	0	0	Incomplete profile	None	Not ACBS prescribed
L- isoleucine	100g tub powder	0-adult	Neutral	L-amino acids	86.3	4.3/5g powder	345	0	0	Incomplete profile	None	Not ACBS prescribed
L-leucine	100g tub powder	0-adult	Neutral	L-amino acids	86.3	4.3/5g powder	345	0	0	Incomplete profile	None	Not ACBS prescribed

L-lysine hydrochloride	100g tub	0-adult	Neutral	L-amino acids	70	3.5/5g powder	280	0	0	Incomplete profile	None	Not ACBS prescribed
L-ornithine hydrochloride	100g tub powder	0-adult	Neutral	L-amino acids	67.7	3.4/5g powder	271	0	0	Incomplete profile	None	Not ACBS prescribed
L-serine	100g tub powder	0-adult	Neutral	L-amino acids	82.9	4.1/5g powder	332	0	0	Incomplete profile	None	Not ACBS prescribed
L-tyrosine	100g tub powder	0-adult	Neutral	L-amino acids	90.1	4.5/5g powder	360	0	0	Incomplete profile	None	Not ACBS prescribed
L-valine	100g tub powder	0-adult	Neutral	L-amino acids	84.6	4.2/5g powder	338	0	0	Incomplete profile	None	Not ACBS prescribed
COMPANY: VITAFLO UK												
L-Arginine 500	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	11.2	0.4 L-arginine/ 500mg	15	3.3	0	Incomplete profile	None	Not ACBS prescribed
L-Arginine 2000	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	44.8	1.8 L-arginine/ 2000mg	15	1.9	0	Incomplete profile	None	Not ACBS prescribed
L-Citrulline 200	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	4.5	0.2 L citrulline/ 200mg	15	3.6	0	Incomplete profile	None	Not ACBS prescribed
L-Citrulline 1000	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	22.4	0.9 Lcitrulline/ 1000mg	15	2.9	0	Incomplete profile	None	Not ACBS prescribed
L-Creatine 5000	30 x 6g sachet powder	3y-adult	Neutral	L-amino acids	70	4.2 L-Creatine/ 5000mg	18	0.2	0	Incomplete profile	None	Not ACBS prescribed
L-Cystine 500	30 x 4g sachet powder	3y-adult	Neutral	L-amino acids	11.6	0.5 L-Cystine/ 500mg	0.8	3.3	0.2	Incomplete profile	None	333-8423
L-Glycine 500	30 x 4g	0-	Neutral	L-amino	9.5	0.4 L-Glycine/	15	3.3	0	Incomplete	None	Not ACBS

	sachet powder	adult		acids		500mg				profile		prescribed
L-Iso-leucine 50	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	1	0.04 L-Isoleucine/ 50mg	15	3.8	0	Incomplete profile	None	304-2280
L-Iso-leucine 1000	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	20.8	0.8 L-Isoleucine/ 1000mg	15	2.9	0	Incomplete profile	None	Not ACBS prescribed
L-Leucine 100	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	2.2	0.1 L-Leucine/ 100mg	15	3.7	0	Incomplete profile	None	333-8431
L-Lysine 4000	30 x 5.6g sachet powder	0-adult	Neutral	L-amino acids	62.2	3.5 L-Lysine/ 4000mg	14	0	0	Incomplete profile	None	Not ACBS prescribed
L-Methionine 100	30 x 4g sachet powder	0-adult	Neutral	L-amino acids	2.2	0.1 L-Methionine/ 100mg	15	3.7	0	Incomplete profile	None	Not ACBS prescribed
L-Phenyl-alanine 50	4g sachet powder	0-adult	Neutral	L-amino acids	1.1	0.04 L-phenylalanine /50mg	15	3.8	0	Incomplete profile	None	345-3263
L-Tyrosine 1000	4g sachet powder	0-adult	Neutral	L-amino acids	22.5	0.9 L-Tyrosine/ 1000mg	15	2.9	0	Incomplete profile	None	328-2209
L-Valine 50	4g sachet powder	0-adult	Neutral	L-amino acids	1	0.04 L-Valine/ 50mg	15	3.8	0	Incomplete profile	None	304-2298
L-Valine 1000	4g sachet powder	0-adult	Neutral	L-amino acids	21.1	0.8 L-Valine/ 1000mg	15	2.9	0	Incomplete profile	None	Not ACBS prescribed

DISORDERS OF FAT METABOLISM

FATTY ACID OXIDATION DISORDERS

DISORDERS OF LONG CHAIN FATTY ACID OXIDATION

Defects of the carnitine cycle

- Carnitine palmitoyltransferase I (CPTI) and II (CPTII) deficiency
- Carnitine acylcarnitine translocase (CACT) deficiency

Defects of β-oxidation

- Very long chain acyl-CoA dehydrogenase deficiency (VLCADD)
- Long chain 3-hydroxylacyl-CoA dehydrogenase deficiency (LCHADD)
- Mitochondrial trifunctional protein deficiency (MTPD)

Background information

Disorders of long chain fatty acid oxidation have an inability to metabolise long chain fatty acids. Without attentive and appropriate dietary treatment, it can lead to irreversible brain damage and death.

Treatment requires:

- limiting lipolysis/avoidance of long fasting times (uncooked cornstarch may be necessary)
- provision of adequate energy (60% of energy is from CHO)
- limiting long chain triglyceride intake to provide 5-15% fat calories (condition dependent)
- supplementation with an MCT oil/emulsions/powder (provide 20-25% of calories)

- supplementation with vitamins, minerals, essential fatty acids and long chain fatty acids including DHA and AA.

MCT products for long chain fatty acid oxidation disorders

MCT based products are essential to meet energy requirements on a low LCT diet, providing MCT is tolerated.

Dose: The dose and type of product should always be advised by the specialist IMD dietitian according to individual patient needs.

Administration: orally or via nasogastric/gastrostomy tube.

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential. Pregnancy may occur with no dietary regimen in place, however, regaining metabolic control is considered a matter of urgency, thus prescription products may be needed on an urgent basis having not been used before/in adulthood. In some conditions peripartum and post-partum management involve continued use of specific products. Increased use of dietary products (including MCT and cornstarch) may be required to support foetal growth in mothers with FAODs.

EMERGENCY MANAGEMENT

During illness patients are at risk of metabolic decompensation due to increased lipolysis and fatty acid oxidation and an emergency regimen is required.

See BIMDG emergency guidelines for further information.

In carnitine transporter deficiency, an emergency regimen only is required during illness, trauma and prolonged fasting.

Table 10

Name	Unit size	Age	Flavour	Fat source	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD											
Monogen	400g tin powder	0-adult	Neutral	MCT 84% LCT 16%	2.2/100 mL	75/ 100 mL	11.6/ 100 mL	2.2/ 100mL	Yes	DHA AA	264-3971
Low Fat Module	500g tin powder	0-adult	Neutral	MCT 4.6% LCT 89.1%	1.6/100mL	67.3/ 100mL	14.9/ 100mL	0.14/ 100mL	Yes	None	Not ACBS prescribed
MCT Oil	500mL bottle liquid	0-adult	Neutral	MCT 100%	0	855/ 100mL	0	95/ 100mL	No	None	272-5125
Liquigen	250mL bottle liquid	0-adult	Neutral	MCT 96.4%	0	450/ 100mL	0	50/ 100mL	No	None	003-9040
COMPANY: VITAFLO UK											
Lipistart	400g tin powder	0-10y	Neutral	MCT 80% LCT 20%	2.1/100mL	69/ 100mL	8.3/ 100mL	3.1/ 100mL	Yes	DHA AA	369-9832
MCT procal	16g sachet powder	1y-adult	Neutral	MCT 99%	2.0	112	3.3	10.1	No	None	369-8669
In exercise / illness a glucose polymers are used. See Emergency regimens/ Energy supplements table 16											

Medium chain acyl CoA dehydrogenase (MCADD) deficiency

Background information

MCADD is the most common fatty acid disorder and is part of the newborn screening programme. This is a disorder of medium chain fatty acid oxidation resulting in an inability to metabolise medium chain fats. Without attentive, prompt and appropriate treatment it can lead to irreversible brain damage and death. When well, patients can eat a normal diet (with controlled fasting times) but require an emergency regimen based on glucose polymers during illness/trauma.

Guidelines are provided on dietary and clinical management in illness and safe fasting times, which increase with age reference BIMDG :: British Inherited Metabolic Disease Group

Emergency dietary management in illness – see Emergency regimens table 16 **MCT products are contraindicated.**

KETOGENIC DIETS (KD) AND GLUCOSE TRANSPORTER 1 (GLUT1) DEFICIENCY

KETOGENIC DIETS

Background information

Ketogenic diets are successfully used and recommended by NICE for the treatment of childhood and adult epilepsy. They are high fat, very low carbohydrate with adequate protein for normal growth. A ketogenic diet produces ketosis, and ketone bodies are used as an alternative energy source for brain tissue, preventing seizures.

Type of ketogenic diets

- classical ketogenic diet: 90% of calories from fat
- MCT diet: MCT produces more ketones per calorie of energy than long chain fat: 45-55% calories from MCT, 21-25% from LCT
- modified ketogenic diet/modified Atkins diet: protein is unrestricted, very low in carbohydrate, 75% of calories from fat

GLUT 1 DEFICIENCY

Background information

GLUT 1 is a protein allowing transportation of glucose into the brain; deficiency of this important carrier results in low brain glucose / energy concentrations leading to epilepsy, movement disorders and developmental delay. Management of the condition is by a ketogenic diet

Table 11

Name	Unit size	Age	Flavour	Fat source	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD											
KetoCal 3:1	300g tin powder	0-adult	Neutral	LCT	1.5/100mL	66/ 100mL	0.68/ 100mL	6.4/ 100mL	Yes	No	409-1104
KetoCal LQ 4:1	200mL liquid carton	1-adult	Neutral Vanilla	LCT	3.1/100mL	150/ 100mL	0.6/ 100mL	14.8/ 100mL	Yes	DHA AA	Neutral 409-1096 Vanilla 409-1286
KetoCal 4:1	300g tin powder	0-adult	Neutral Vanilla	LCT	2/100mL	100/ 100mL	0.4/ 100mL	9.8/ 100mL	Yes	DHA AA	Neutral 409-1088 Vanilla 409-1294
KetoCal 2.5:1	237mL liquid	8y-adult	Neutral	MCT 25%	10.7g/ 237mL	363/237mL	5.2g/ 237mL	34g/ 237mL	Yes	DHA	409-1062

				LCT							
MCT Oil	500mL bottle liquid	0-adult	Neutral	MCT 100%	0	855/100mL	0	95/ 100mL	No	None	272-5125
Liquigen	250mL bottle liquid	0-adult	Neutral	MCT 96.4%	0	450/100mL	0	50/ 100mL	No	None	003-9040
COMPANY: VITAFLO LTD											
MCT procal	16g sachets powder	1y-adult	Neutral	MCT 99%	2.0	112	3.3	10.1	No	None	369-8669
Keyo	100g semi solid pot	3y-adults	Chocolate	LCT 3.1	8	310	2	30	Yes	DHA ARA	405-6719
Betaquik	225mL liquid bottle	3y-adult	Neutral	MCT 95% LCT 5%	0	189/ 100mL	0	21/ 100mL	No	No	406-6742

X LINKED ADRENOLEUKODYSTROPHY (X-ALD)/ ADRENOMYELONEUROPATHY (AMN)

Background information

These are both X linked chromosomal disorders causing severe neurodegeneration. They are due to reduced very long chain fatty acid oxidation in the peroxisomes. This leads to accumulation of saturated and mono-unsaturated very long chain fatty acids (C 26:0 and C24:0) in the nervous system.

Dietary treatment requires:

- Lorenzo oil (made up of Glycerol trioleate GTO and Glycerol trierucate GTE)
- GTO oil
- moderate fat restriction
- energy supplementation
- fat soluble vitamins
- essential fatty acid supplements

Dose of Lorenzo/GTO oil

The dose of each product is dependent on patient weight and age and will be determined by the specialist IMD dietitian.

Administration: orally or via nasogastric or gastrostomy tube.

Table 12

Name	Unit size	Age	Flavour	Source of fat	Fat per 100g	Fat g per unit size	Energy Kcal per unit size	CHO g per unit size	Protein g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
GTO oil Glycerol trioleate oil	500mL	18m-adult	Neutral	90% Oleic acid	91		819 Dose dependent	0	0	0	0	Not ACBS prescribed
*Lorenzo's oil	500mL	18m-adult	Neutral	Glycerol Trioleate Glycerol Trierucate Ratio 4:1 part	90		807 Dose dependent Up to 20% of energy intake	0	0	0	0	Not ACBS prescribed
Energy supplements - see table 16												
Vitamin and mineral/essential fatty acid supplements - see table 17												

* not recommended before 18m of age. It lowers blood platelet levels and DHA levels, which may affect retinal and brain development.

DISORDERS REQUIRING A MODIFIED CARBOHYDRATE DIET

GLYCOGEN STORAGE DISEASES (GSD)

Background information

There are 11 different GSDs, with varying levels of severity and dietary treatments. The different GSD types relate to specific enzyme defects in the glycolysis pathway, leading to impaired production of glucose and hypoglycaemia. The glycogen disorders commonly have a limited fasting tolerance needing overnight feeding and in illness, an emergency regimen based on glucose polymer. Uncooked cornstarch is a slow release carbohydrate necessary to enhance fasting tolerance in some GSD types. Dietary treatment depends on the specific type of GSD. It commonly requires supplementation with glucose polymer and cornstarch.

GSD IIIa is due to deficient enzyme activity in the muscle and liver, leading to cardiac and skeletal myopathy. A high protein diet using a protein supplement may improve symptoms and is commonly recommended.

Dietary treatment requires:

- slow release carbohydrate (cornstarch/ *Glycosade*) to prevent hypoglycaemia and maintain normoglycemia.
- protein supplementation ± cornstarch*/ Glycosade* in GSD Type III

Dose: Glucose polymer (dose per kg will vary depending on age and metabolic control**)**
Infants and children: 0.5g/kg/h
Adults: 0.3g/kg /h

Slow release carbohydrate (cornstarch/ *Glycosade* depending on age and metabolic control**)**
Infants and children: 1 to 2g/kg.
Adults 1g/kg

Administration: orally or via nasogastric/ gastrostomy tube.

Protein supplementation

Infants and children: 2 to 3g/kg depending on GSD type, age and metabolic control. Aim to provide 20-25% of energy from protein.
Adults: 1 to 2g /kg depending on metabolic control and weight.

Pregnancy

Dietary treatment must be sustained throughout pregnancy to protect both the mother and developing baby from hypoglycaemia, and to ensure adequate nutrition to support foetal growth. ***Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential.*** Pregnancy may occur with no dietary regimen in place, however, regaining metabolic control is considered a matter of urgency, thus prescription products may be needed on an urgent basis having not been used before/in adulthood.

EMERGENCY MANAGEMENT

In illness if it is not possible to sustain normoglycaemia by the usual feeding regimen, an emergency regimen using glucose polymer is required.

See BIMDG emergency guidelines for further information.

Table 13

COMPANY: NUTRICIA LTD											
Energy supplements - see table 16											
Protein supplements - see table 16											
COMPANY: VITAFLO UK											
Energy supplements - see table 16											
Protein supplements - see table 16											
Slow release carbohydrate supplements											
Name	**Unit size**	**Age**	**Flavour**	**Carbohydrate source**	**Energy kcal per unit size**	**CHO g per unit size**	**Protein g per unit size**	**Fat g per unit size**	**Vitamins and minerals supplemented to provide complete profile**	**LCPUFA Novel Ingredients**	**ACBS listed PIP code**
COMPANY: VITAFLO LTD											
Glycosade	60g sachet powder	2y-adult	Neutral Lemon	Amylopectin starch	212	53	0	0	No	None	Neutral 341-2780 Lemon 414-6775
COMPANY: SUPERMARKETS											
Cornstarch/ Cornflour	500g packet powder	6m-adult	Neutral	Starch	214/60g	53	0	0	No	None	BNF approved

GALACTOSAEMIA

Background information

Galactosaemia is a rare metabolic disorder with an inability to metabolise galactose. This is caused by a deficiency of the enzyme galactose-1-phosphate-uridyltransferase (GALT). This leads to the accumulation of the metabolites galactose-1-phosphate (Gal-1-P), galactitol, and galactonate. Without attentive and appropriate dietary treatment in infancy it causes life-limiting illness with hepatic, renal and cerebral involvement. Infants present with feeding difficulties, liver failure, E. coli sepsis and weight loss. Bilateral cataracts may be seen in the first weeks of life.

In the long-term, complications such as cognitive deficits, speech and language deficits, neurological abnormalities, and hypergonadotropic hypogonadism in females occur.

Dietary treatment requires:

- a lifelong galactose (lactose) restricted diet
- lactose free formula
- calcium and vitamin D supplementation

On presentation, standard milk based infant formula or breastfeeding should stop and lactose-free infant formula commence when galactosaemia is suspected. In the initial stages of treatment, it may be necessary to use an MCT based extensively hydrolysed casein formula until the liver function returns to normal. Soya infant formula is then introduced.

Protein hydrolysate milks based on whey hydrolysate contain residual lactose and are best avoided.

Dose of lactose free infant formula:

The amount of lactose-free infant formula is determined by fluid and nutrient requirements for age and weight.

Administration: orally or via nasogastric tube.

Table 14

Name	Unit size	Age	Source of protein	Protein g 100mL / unit size	Energy Kcal unit size	CHO g / unit size	Fat g / unit size	Vitamins and minerals supplemented to provide	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: SMA nutrition										
Wysoy	860g tin powder	0-2y	Soy bean	1.8/100 mL	67/100 mL	6.9/100 mL lactose free	3.6/ 100mL	Yes	DHA	408-6567
COMPANY: MEAD JOHNSON										
Pregestimil Lipil	400g tin powder	0-2y	Casein hydrolysate	1.9/100mL	70/100mL	6.9/100mL <5mg residual lactose	3.8/ 100mL (55% MCT)	Yes	DHA	043-4050
Nutramigen 1 with LGG	400g tin powder	0-2y	Casein hydrolysate	2.1/100mL	75/100mL	8.2/ 100mL <5mg residual lactose	3.7/ 100mL	Yes	DHA	019-8861
Puramino AA	400g tin powder	0-2y	L-amino acids	1.9/100mL	70/100mL	7.2/ 100mL lactose free	3.6/ 100mL	Yes	DHA AA	338-3304
COMPANY: NUTRICIA LTD										
Neocate LCP	400g tin powder	0-2y	L -amino acids	1.8/100mL	67/100mL	7.2/100mL lactose free	4.3/ 100mL	Yes	DHA AA	329-0301
Nutrison Soya	500mL / 1 L bottle liquid	1-6y	Soy bean	4.0/100mL	100/100 mL	12.3/ 100mL lactose free	3.9/ 100mL	Yes	No	500mL 379-4500 1 L 236-4123

CITRIN DEFICIENCY OR CITRULLINEMIA TYPE II (CTLN2)

Background information

This is a rare complex inborn error of metabolism and may present either neonatally, in childhood or adulthood. Severity of dietary treatment is determined by the symptoms. Citrin is a liver enzyme allowing transportation of aspartate into the urea cycle. Disturbance of this pathway can result in liver dysfunction and may require liver transplantation in adults.

Dietary treatment requires:

- low carbohydrate, high fat high protein diet
- soya products, which are rich in arginine, and L-arginine supplement
- high protein supplements
- fat (long and medium chain fat) based energy supplements

Protein and fat supplements

Protein and fat supplements provide substrates for the urea cycle.

Dose:

The dose of individual supplements is patient specific depending on symptoms. This will be determined by specialist IMD dietitians.

EMERGENCY REGIMEN

See BIMDG emergency guidelines for further information.

Table 15

COMPANY: NUTRICIA LTD											
Protein energy supplements - see table 16											
Fat energy supplements - see table 16											
COMPANY: VITAFLO LTD											
Protein energy supplements - see table 16											
Fat energy supplements - see table 16											
COMPANY: HOLLAND AND BARRETT	**Unit size**	**Age**	**Flavour**	**Source of protein**	**Protein g per 100g**	**Protein g per unit size**	**Energy Kcal per unit size**	**CHO g per unit size**	**Vitamins and mineral supplemented providing complete profile**	**LCPUFA Novel Ingredients**	**ACBS listed PIP code**
COMPANY: HOLLAND AND BARRET											
Natures Garden Pure soya protein isolate powder (low carbohydrate)	908g Tub powder	12m-adult	Neutral	Soya isolate	392	1.3	86	5	No	None	Not ACBS prescribable

EMERGENCY REGIMENS reference to energy supplements table 16

Background information

These are a key part of management in illness in conditions at risk of metabolic decompensation. They are given orally as a drink or via a nasogastric/gastrostomy tube. They are used to prevent acute decompensation by providing glucose as an energy source preventing the breakdown of protein/fat, which may lead to the accumulation of toxic metabolites, or providing a source of glucose in conditions in which fasting tolerance is limited and hypoglycaemia may occur.

They are usually based on a glucose polymer solution; the concentration is age dependent.

Administration

Orally or via a nasogastric/ gastrostomy tube, given over 24 hours.

Dosage

0-12m	10% carbohydrate
1-2y	15% carbohydrate
2-9y	20% carbohydrate
10y-adult	25% carbohydrate

In some conditions (MSUD and GA1) it is essential that the disorder specific precursor-free amino acids are added to the emergency regimen to prevent encephalopathy and metabolic decompensation.

See BIMDG emergency guidelines for further information.

KETOTIC HYPOGLYCAEMIA (KH)

Background

Ketotic hypoglycaemia is a common cause of hypoglycaemia with raised ketones in plasma and urine, often precipitated by a prolonged period of fasting due to illness. Many children become non symptomatic with age. A fasting tolerance test helps to establish a safe fasting time under normal conditions. An emergency regimen based on glucose polymer only is required during illness.

EMERGENCY REGIMEN table 16

See BIMDG emergency guidelines for further information.

ENERGY SUPPLEMENTS

Background information
Energy supplements in the form of carbohydrate polymers ± ***fat emulsions are essential in the treatment of inborn errors of metabolism.

Energy supplements:

- are an essential source of calories and are core ingredients in emergency regimens during illness/trauma to prevent/minimise catabolism and promote anabolism
- provide an energy source in severe dietary restrictions
- supplement energy intake in exercise when energy supply is limited e.g. fatty acid oxidation deficiency or glycogen storage diseases
- supply energy substrates for metabolic conditions with a limited fasting tolerance e.g. fatty acid oxidation and glycogen storage disorders
- supplement oral or tube feeding requirements promoting anabolism

Fat emulsions may be contraindicated in some metabolic conditions

Dose of energy supplements is dependent on the age and condition of the patient. This will be advised by a specialist IMD dietitian.

Administration: orally or via nasogastric/gastrostomy tube.

PROTEIN SUPPLEMENTS

As part of the dietary therapy some metabolic disorders require additional protein supplementation e.g. GSD type III.

Table 16

Name	Unit size	Age	Flavour	Carbohydrate, fat or protein source	Energy kcal per unit size	CHO g per unit size	Protein g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA											
Carbohydrate energy supplements											
Polycal powder	400g	0-adult	Neutral	Maltodextrin	19/5g powder	4.8/5g powder	0	0	No	None	027-6816
Polycal liquid	200mL liquid bottle	3y-adult	Neutral Orange	Maltodextrin Glucose syrup	74/30mL	18.6/ 30mL	0	0	No	None	Neutral 377-1334 Orange 377-1326
Super soluble maxijul powder	200g tin powder 4x132g powder sachet	0-adult	Neutral	Dried glucose syrup	76/20g powder	19/20g powder	0	0	No	None	200g tin 024-6827 Sachet (132g) 033-0548
Fat energy supplements											
*Calogen Liquid	200mL 500mL liquid	Neutral 0-adult Flavoured	Neutral Strawberry	Vegetable oil	135/30mL 140/30mL	0 1	0 0	15/30mL 15/30mL	No	None	Neutral 327-3414 Strawberry

	bottle		Banana		140/30mL	1	0	15/30mL			327-3422 Banana 327-3406
**Liquigen Liquid	250mL liquid bottle	0-adult	Neutral	Palm / coconut oil MCT	135/30mL	0	0	15/30mL	No	None	003-9040
MCT oil	500mL liquid bottle	0-adult	Neutral	Coconut/ palm oil MCT	171/20mL	0	0	19/20mL	No	None	272-5125
Carbohydrate and fat energy supplements											
***Duocal powder	400g tin powder	0-adult	Neutral	Dried glucose fat contains MCT	50/10g powder	7.2/10g powder	0	2.2 /10g powder contains some MCT	No	None	031-4989
Energivit	400g tin powder	0-12m	Neutral	Carbohydrate Fat	74/ 100mL	10/ 100mL	0	3.8/ 100mL	Yes	None	280-3245
Protein energy supplements											
Protifar	225g tin powder	3y-adult	Neutral	Cows milk protein	37/10g powder	0.1/10g powder	8.7/10g powder	0.1/10g powder	No	None	040-1653
Calogen extra	200mL liquid bottle	3y-adult	Neutral Strawberry	Fat Cows milk Maltodextrin	160/ 40mL	1.8/ 40mL	2.0/ 40mL	16.1/ 40mL	Yes	None	346-3882
Calogen extra shots	6x40mL liquid	3y adult	Neutral Strawberry	Fat Cows milk	160/shot	1.8	2.0	16.1	Yes	None	Neutral 373-1684

	bottle			Maltodextrin							Strawberry 373-1866
COMPANY: VITAFLO UK											
Name	**Unit size**	**Age**	**Flavour**	**Carbohydrate, fat or protein source**	**Energy Kcal per unit size**	**CHO g per unit size**	**Protein g per unit size**	**Fat g per unit size**	**Vitamins Minerals supplemented to provide complete profile**	**LCPUFA Novel Ingredients**	**ACBS listed PIP code**
Carbohydrate energy supplements											
SOS 10,15,20,25 sachet powders	10=21g 15=31g 20=42g 25=52g	0-12m 1-2y 3-10y 10y-adult	Neutral	Maltodextrin	80/sachet 120/sachet 160/sachet 200/sachet	20 30 40 50	0 0 0 0	0 0 0 0	No	None	**353** **10** -5903 **15**-5911 **20**- 5929 **25**-5895
Vitajoule	500g tin powder	0-adult	Neutral	Maltodextrin	380/100g powder	9.5/10g powder	0	0	No	None	092-9299
Fat energy supplements											
Beta quick	225mL liquid bottle	3y-adult	Neutral	MCT 95% LCT 5%	189/ 100mL	0	21/ 100mL	MCT 20g LCT 1g/ 100mL	No	None	406-6742
Carbohydrate and fat energy supplements											
Basecal 200	30x 43g sachet powder	12m-adult	Neutral	Carbohydrate Fat	200	30	0	8.9	Yes	DHA AA	384-8637

Protein energy supplements											
MCT Procal	30 x 16g sachets powder	3y-adult	Neutral	MCT fat Carbohydrate Protein	112/ 16g	3.3	2.0	10	No	None	369-8669
Procal	30 x 15g sachets powder	3y-adult	Neutral	LCT Fat Carbohydrate Protein	100	4.3	2.0	8.3	No	None	410-6993
COMPANY: Nutrinovo											
Protein supplement											
ProSource TF	45mL sachet powder	3y-adult	Neutral	Protein hydrolysed collagen	44	1	11	0	No	None	348-9622
* Contraindicated in the use of LCHADD and LCFAD (long and vey long chain acyl CoA dehydrogenase deficiencies) **Contraindicated for the use of MCADD (medium chain acyl dehydrogenase deficiency) *** Contraindicated for both MCADD/LCHADD and VLCAD											

VITAMIN AND MINERAL / ESSENTIAL FATTY ACID SUPPLEMENTS

Background information
Many inherited metabolic disorders of metabolism require dietary treatment, which may restrict intake of macro- and micronutrients. These specialist diets may be nutritionally incomplete and vitamin and mineral supplementation is required to prevent deficiency. Supplementation is required in conditions such as urea cycle disorders, amino acid disorders, organic acidaemias, disorders of fat metabolism and glycogen storage disorders.

Dose The dose is dependent on the biochemical, clinical and dietary evaluation and is adjusted by the specialist IMD dietitian.

Vitamin and mineral supplements are usually given once or twice daily.

Administration orally or via nasogastric/gastrostomy tube.

Essential fatty acids (EFA) can only be provided by dietary sources. In diets restricted in fat, protein or carbohydrate these may be sub-optimal and supplementation necessary. There are two EFA: alpha linolenic (omega 3, ALA), which produces docosahexaenoic acid (DHA) and linoleic acid (omega 6, LA) which produces arachidonic acid (AA). DHA and AA supplements are available for diets at risk of EFA deficiency.

Table 17

Name	Unit size	Age	Flavour	Source of protein	Protein g 100g	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins Minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
Paediatric Seravit	200g tin powder	0- 6m (neutral only) 6-12m 1-7y 7-14y	Neutral Pineapple	0	0	0	15/5g 42/14g 51/17g 75/25g 105/35g	3.8/5g 11/14g 13/17g 19/25g 26/35g	0 0 0 0	Complete profile	None	Neutral 233-4662 Pineapple 233-4654
Phlexy Vits tablets	60 tablets/tub Dose 5 tablets	11y-adult	Neutral	0	0	0	5	-	0	Complete profile	None	322-5414
Phlexy Vits sachets	30x7g sachet powder	11y-adult	Neutral	0	0	0	1	-	0	Complete profile	None	272-6834
COMPANY: VITAFLO UK												
Fruiti Vits	30 x6g sachet powder	3y-adult	Orange	0	0	0	2	0.5	0	Complete profile	None	371-5067
Doc Omega	30 x 4 g sachet	0-adult	Neutral	Milk protein	3.1	0.1	18	3.2	0.5	No	DHA 200mg	349-0182

	powder											
Key Omega	30 x 4 g sachet powder	0-adult	Neutral	Milk protein	4.4	0.2	19	2.8	0.8	No	DHA 100mg AA 200mg	343-7795
COMPANY: ALLIANCE PHARMACEUTICALS												
Forceval Capsule	Capsule (liquid)	12y-adult	Neutral	0	0	0	-	-	0	Complete profile	None	008-1489
Forceval Soluble (adult)	30/pack Effervescent tablet	12y-adult	Forest Fruits	0	0	0	8	0.8	0	Complete profile	None	369-1524
Forceval Soluble (junior)	30/pack Effervescent tablet	6y-12y	Forest Fruit	0	0	0	8.5	0.9	0	Complete profile	None	369-1524
COMPANY: ESSENTIAL PHARMECEUTICALS LTD												
Ketovite tablets	100/pack Dose 3 tablets	1m-adult	Neutral	0	0	0		0	-	Incomplete profile	None	014-7306
Ketovite liquid	150ml liquid bottle Dose 5mL	1m-adult	Orange	0	0	0		0		Incomplete profile	None	013-1953

FLAVOUR SACHETS

These are sachets of flavoured powders, which are added to protein substitutes to mask their bitter taste.

Table 18

Name	Unit size	Age	Flavour	Protein source	Protein g per 100g per 100mL	Protein g per unit size	Energy Kcal per unit size	CHO g per unit size	Fat g per unit size	Vitamins and minerals supplemented to provide complete profile	LCPUFA Novel Ingredients	ACBS listed PIP code
COMPANY: NUTRICIA LTD												
Flavour sachets	20x5g sachet powder	12m-adult	Lemon/ lime Cherry/ vanilla Grape fruit	Sugar Sweeteners	0 0 0	0 0 0	15 13 15	1.3 1.8 1.4	0 0 0	No	None	Not ACBS prescribed
COMPANY: VITAFLO UK												
Flavour pac	30x4g sachet powder	12m-adult	B/current R/berry Orange Tropical	Sugar Sweeteners	0 0 0 0	0 0 0 0	13 13 14 12	3.3 3.2 3.4 3.1	0 0 0 0	No	None	296-1068 296-1084 296-1050 296-2817

SPECIAL LOW PROTEIN FOODS FOR USE WITH LOW PROTEIN DIETS

Background information

Patients on a low protein diet are reliant upon ACBS prescribed low protein foods. These products are not available to purchase in supermarkets. They are important to:

- improve variety in the diet
- help attain acceptable metabolic control and growth by providing essential calories
- help dietary adherence

Inability to access low protein special foods will lead to poor metabolic control which may lead to irreversible brain damage.

The amount of low protein products prescribed will vary from patient to patient and should be determined on an individual basis. However, guidelines are given which are calculated according to energy requirements. Low protein special foods are expected to provide up to 50% of estimated daily energy requirements in patients on a very low protein diet (natural protein: up to 10g/day).

The maximum number of monthly food units by age for special low protein foods (except low protein milk replacements) are identified in Table 19

Table 19
Recommended maximum **monthly** number of units of low protein foods for each age group

Age of patient with PKU	Recommended maximum number of low protein items to prescribe each month
4 months -3 years	20 units
4-6 years	25 units
7-10 years	30 units
11-18 years	50 units
Adults	50 units
Pre-pregnancy/pregnancy	50 units

PLEASE NOTE: low protein milk replacements are excluded from this monthly unit allocation

Table 20
The definition of one unit of low protein foods

ACBS low protein prescribed foods	Definition of one unit	Suggested size of unit
Pasta/Rice	1 box	Up to 500g
Pasta pots / potato pots/pasta in sauce/XPots	1 pack/box	Up to 300g
Flour mix/cake mix/waffle mix	1 pack/box	Up to 500g
Bread/bread rolls	1 pack	Up to 600g
Pizza bases/pizza base mixes	1 pack	Up to 300g
Crackers/crispbread /mini crackers /croutons/ bread sticks/savoury snacks	1 pack/box	Up to 200g
Sausage/burger mixes/fish substitutes	1 pack/box	Up to 360g dry powder
Cheese sauce mix	1 pack	Up to 225g dry powder
Soups	1 box	Up to 112g dry powder
Breakfast cereals	1 box/pack	Up to 400g
Egg replacer/egg white replacer	1 tub/box	Up to 500g
Yoghurt substitute	1 tub	Up to 400g
Biscuits/biscuit bars	1 pack/box	Up to 200g
Cakes/breakfast bars/dessert powder mixes /jelly powder/ chocolate spread substitute	1 pack/box/tub	Up to 300g
Readymade desserts	1 pack	Up to 500g
Energy bars (Vitabite or Chocotino)	1 pack/box	Up to 175g

PLEASE NOTE: The above units are intended as a guide only as package size is variable. Any pack size greater than the suggested weights should be estimated as more than 1 unit.

LOW PROTEIN MILKS

Table 21

Product/Company	Unit Size	Age	Phe mg per 100mL	Leu mg per 100mL	Energy Kcal per 100mL per 100g	CHO g per 100mL/g	Fat g per 100mL/g	ACBS listed PIP code
COMPANY: TARANIS								
Dalia Liquid	24 x 200mL carton	1y- adult	6.4mg	14.1mg	50/100mL	6.41/100mL	2.6/100mL	401-5780
Dalia Powder	400g powder tin	3y- adult	10mg	20mg	428/100g	77.5/100g	12.3/100g	403-4971
COMPANY: MEVALIA								
Mevalia Lattis	500mL x 6	6m-adult	12mg	28mg	62/100mL	9.1/100mL	2.7/100mL	413-7196
COMPANY: NUTRICIA LTD								
Sno Pro	200mL	0- adult	8.7mg	23.7mg	89/100mL	11/100mL	4.7/100mL	022-0293
Loprofin Drink	200mL	1y-adult	10mg	20mg	40/100mL	5/100mL	2.0/100mL	043-0827
COMPANY: VITAFLO LTD								
Pro Zero	18 x 250mL	6m-adult	0	0	67/100mL	8.1/100mL	3.8/100mL	304-5065
Pro Zero	6 x 1L	6m-adult	0	0	67/100mL	8.1/100mL	3.8/100mL	340-5073

LOW PROTEIN PRODUCTS
Table 22

Product	Unit size	Age	Flavour	Energy Kcal per 100g per unit	CHO g per 100g per unit	Fat g per 100g per unit	ACBS listed PIP code
COMPANY: FATE SPECIAL FOODS							
FATE All Purpose Mix	500g	From weaning		352	88	0.2	279-8270
FATE Plain Cake Mix	2 x 250g	From weaning	Plain	367	92	0.1	279-8288
FATE chocolate Cake Mix	2 x 250g	From weaning	Chocolate	365	91	0.4	279-8296
COMPANY: JUVELA							
JUVELA Low Protein Egg Replacer	454g	From weaning		346 30/10g	82 8/10g	0.3 0/10g	032-1737
JUVELA Low protein Sliced Loaf	400g	From weaning		243	52.4	2.7	092-3961
JUVELA Low protein Bread Rolls	350g	From weaning		255	49.1	5.3	206-3485
JUVELA Low protein Mix	500g	From weaning		316	62.3	6.5	035-2765
JUVELA Low Protein Pizza Base	360g (2x180g)	From weaning		316	62.3	6.5	282-0538
COMPANY: METAX							
YoguMaxx (Yoghurt Substitute)	400g	From weaning		422	58	18	407-4159
COMPANY: MEVALIA LTD							

MEVALIA Low protein Bread Mix	500g	From weaning		348	84	0.4	381-9752
MEVALIA Low protein Pan Carre	400g	From weaning		214	39	3.4	381-7186
MEVALIA Low protein Pan Rustico	400g	From weaning		217	37	3.8	381-7194
MEVALIA Low protein Pan Casereccio	220g	From weaning		214	38	3.8	405-7857
MEVALIA Low protein Ciabattine	4 x 65g	From weaning		220	41	3.4	381-7202
MEVALIA Low protein Mini Baguette	2 x 100g	From weaning		235	43	3.9	402-8940
MEVALIA Low protein Grissini (breadsticks)	3 x50g	From weaning		434	76	13	415-2195
MEVALIA Low protein Pizza Base	2 x 150g	From weaning		263	49	4.2	381-7178
MEVALIA Low protein Ditali pasta	500g	From weaning		351	81	1.1	339-1114
MEVALIA Low protein Fusilli pasta	500g	From weaning		351	81	1.1	381-9778
MEVALIA Low protein Penne pasta	500g	From weaning		351	81	1.1	381-7152
MEVALIA Low protein Spaghetti	500g	From weaning		355	81	1.6	381-7145
MEVALIA Low protein Rice	400g	From weaning		343	79	1.3	339-1106

MEVALIA Low protein Burger Mix	350g	From weaning		367	73	4.6	339-1098
MEVALIA Low protein Egg Replacer	400g	From weaning		340	84	0	401-7943
MEVALIA Low protein Chocotino	100g	From weaning		583	47	42	405-7592
MEVALIA Low protein Cookies	200g	From weaning		471	78	17	382-0602
MEVALIA Low protein Frollini Biscuits	200g	From weaning		476	84	15	382-0594
MEVALIA Low protein Fruit Bars	5 x 25g	From weaning		424	7	14	402-8957
COMPANY: NUTRICIA LTD							
LOPROFIN Part Baked Sliced Loaf	400g	From weaning		259	51	5	361-6067
LOPROFIN Part Baked Bread	260g (4x65grolls)	From weaning		259	51	5	361-6067
LOPROFIN Crackers	150g	3y- adult		450	78	15	039-7125
LOPROFIN Herb Crackers	150g	3y- adult		450	78	15	277-4446
LOPROFIN Cereal Loops	375g	1y- adult		385	94	1.1	266-0140
LOPROFIN Cereal Flakes	375g	1y- adult	Chocolate	374	91	0.9	328-5160
LOPROFIN Cereal Flakes	375g	1y- adult	Strawberry	380	93	0.7	328-5152
LOPROFIN Egg Replacer	500g	From weaning		346	82	0.3	022-8031
LOPROFIN Egg White Replacer	100g	From weaning		185	0	0	270-1456
LOPROFIN Mix	500g	From weaning		361	87	0.4	004-6607
LOPROFIN Cake Mix	375g	From weaning	Chocolate	366	88	1.0	332-7277

LOPROFIN Animal Pasta	500g	From weaning		363	87	1.2	335-6169
LOPROFIN Fusilli	500g	From weaning		363	87	1.2	331-5058
LOPROFIN Lasagne	250g	From weaning		363	87	1.2	298-1777
LOPROFIN Long Spaghetti	500g	From weaning		363	87	1.2	211-5251
LOPROFIN Macaroni	250g	From weaning		363	87	1.2	328-5194
LOPROFIN Penne	500g	From weaning		363	87	1.2	331-5041
LOPROFIN Rice	500g	From weaning		366	88	1.3	232-3376
LOPROFIN Tagliatelle	250g	From weaning		363	87	1.2	328-5210
COMPANY: PK FOODS							
PK foods Crispbread	75g	From weaning		406	89.7	5.1	280-3799
PK Foods Egg Replacer	200g	From weaning		347	85.7	0.3	280-3773
PK Foods Flour Mix	750g	From weaning		343	85.6	0	280-3765
PK foods Cherry Jelly Mix	320g	From weaning		356	88	0	280-3716
PK Foods Orange Jelly Mix	320g	From weaning		382	88	0	280-3724
PK Foods White Sliced Bread	300g	From weaning		263	47.2	6.5	280-3781
COMPANY: PROMIN							
PROMIN Low Protein All Purpose Baking Mix	1000g	From weaning		339	80	0.5	410-7611
PROMIN Low Protein Bread: Fresh White Sliced	4 x 800g	From weaning		257	51.8	4.3	394-3941
PROMIN Low Protein Bread: Fresh Brown Sliced	4 x 400g	From weaning		266	53.8	4.6	408-5528
PROMIN Low Protein Bread Buns Fresh Baked	4 x 75g	From weaning		257	51.8	4.3	405-6537
PROMIN Low Protein Breakfast Bars	6 x 40g	3y-adult	Apple & Cinnamon	487/100g	66/100g	24.4/100g	386-0830
PROMIN Low Protein Breakfast	6 x 40g	3y- adult	Banana	469/100g	65/100g	23/100g	386-0814

Bars							
PROMIN Low Protein Breakfast Bars	6 x 40g	3y- adult	Chocolate & Cranberry	467/100g	69/100g	21/100g	386-0855
PROMIN Low Protein Breakfast Bars	6 x 40g	3y- adult	Cranberry	464/100g	68/100g	21/100g	386-0822
PROMIN Low Protein Burger Mix	4 x 62g	From weaning	Lamb & Mint	155 /100g	27/100g	4.9/100g	341-4927
PROMIN Low Protein Burger Mix	4 x 62g	From weaning	Original	155/100g	27/100g	4.9/100g	322-4862
PROMIN Low Protein Cheese Sauce Mix	225g	3y- adult		86/100g prepared product	18/100g prepared product	0.97/100g prepared product	383-7457
PROMIN Low Protein Couscous	500g	From weaning		353	86	0.8	290-7640
PROMIN Low Protein Croutons	4 x 40g	3y- adult		99/100g	84.2/100g	2.8	386-0798
PROMIN Low Protein Custard	6 x 36.5g	From weaning	Custard	108/100g prepared product	24/100g prepared product	1.3/100g prepared product	341-5031
PROMIN Low Protein Dessert	6 x 36.5g	From weaning	Caramel	109/100g prepared product	24/100g prepared product	1.4/100g prepared product	341-5049
PROMIN Low Protein Dessert	6 x 36.5g	From weaning	Chocolate & Banana	107/100g prepared product	24/100g prepared product	1.4/100g prepared product	327-9767
PROMIN Low Protein Dessert	6 x 36.5g	From weaning	Strawberry & Vanilla	109/100g prepared	26/100g prepared	0.6/100g prepared	327-9759

				product	product	product	
PROMIN Low Protein Hot Breakfast	6 x 57g	From weaning	Apple & Cinnamon	135/100g prepared product	31/100g prepared product	1/100g prepared product	317-2525
PROMIN Low Protein Hot Breakfast	6 x 57g	From weaning	Banana	130/100g prepared product	30/100g prepared product	1/100g prepared product	322-4854
PROMIN Low Protein Hot Breakfast	6 x 57g	From weaning	Chocolate	135/100g prepared product	31/100g prepared product	1/100g prepared product	317-2533
PROMIN Low Protein Hot Breakfast	6 x 56g	From weaning	Original	147/100g prepared product	34/100g prepared product	1.4/100g prepared product	317-2509
PROMIN Low Protein Lasagne Sheets	200g	From weaning		353	86	0.8	290-7582
PROMIN Low Protein MacPots	4 x 61g	From weaning	Macaroni Cheese	140/100g prepared product	24/100g prepared product	4.6/100g prepared product	377-9865
PROMIN Low Protein MacPots	4 x 61g	From weaning	Tomato Macaroni	140/100g prepared product	25/100g prepared product	4.4/100g prepared product	377-9873
PROMIN Low Protein Pasta: Alphabets	500g	From weaning		353	86	0.8	287-5466
PROMIN Low Protein Pasta: Elbows	500g	From weaning		353	86	0.8	290-7590
PROMIN Low Protein Pasta: Flat Noodles	500g	From weaning		353	86	0.8	356-1587

PROMIN Low Protein Pasta: Macaroni	500g	From weaning		353	86	0.8	290-7624
PROMIN Low Protein Pasta: Shells	500g	From weaning		353	86	0.8	290-7616
PROMIN Low Protein Pasta: Short Cut Spaghetti	500g	From weaning		353	86	0.8	290-7632
PROMIN Low Protein Pasta: Spirals	500g	From weaning		353	86	0.8	290-7608
PROMIN Low Protein Pastameal	500g	From weaning		353	86	0.8	240-5181
PROMIN Low Protein Pasta in Sauce	4 x 66g	From weaning	Cheese & Broccoli	106/100g prepared product	26/100g prepared product	0.13/100g prepared product	290-7657
PROMIN Low Protein Pasta in Sauce	4 x 72g	From weaning	Moroccan	129/100g prepared product	31/100g prepared product	0.9/100g prepared product	327-9734
PROMIN Low Protein Pasta in Sauce	4 x 72g	From weaning	Tomato Pepper & Herb	99/100g prepared product	24/100g prepared product	0.1/100g prepared product	290-7665
PROMIN Low Protein Promin Plus Fibre Pasta: Flat Noodles	500g	From weaning		363	88	0.7	384-5310
PROMIN Low Protein Promin Plus Fibre Pasta: Macaroni	500g	From weaning		363	88	0.7	384-5336
PROMIN Low Protein Promin Plus Fibre Pasta: Spaghetti	500g	From weaning		363	88	0.7	384-7465
PROMIN Low Protein Promin	500g	From weaning		363	88	0.7	384-5302

Plus Fibre Pasta: Spirals							
PROMIN Low Protein Pasta (Tricolour) Alphabets	500g	From weaning		351	86	0.8	240-5157
PROMIN Low Protein Pasta (Tricolour) Elbows	500g	From weaning		351	86	0.8	286-8487
PROMIN Low Protein Pasta (Tricolour) Shells	500g	From weaning		351	86	0.8	240-5132
PROMIN Low Protein Pasta (Tricolour) Spirals	500g	From weaning		351	86	0.8	240-5140
PROMIN Low Protein Potato Pots	4 x 50g	From weaning	Cabbage, Bacon & Croutons	111/100g prepared product	23/100g prepared product	1.6/100g prepared product	378-2026
PROMIN Low Protein Potato Pots	4 x 50g	From weaning	Onion & Croutons	115/100g prepared product	23/100g prepared product	2.1/100g prepared product	378-2000
PROMIN Low Protein Potato Pots	4 x 50g	From weaning	Sausage & Croutons	111/100g prepared product	23/100g prepared product	1.7/100g prepared product	378-2018
PROMIN Low Protein Potato Cake Mix	300g	From weaning		165/100g prepared product	31/100g prepared product	3.7/100g prepared product	395-0292
PROMIN Low Protein Rice	500g	From weaning		353/100g prepared product	86/100g prepared product	0.8/100g prepared product	240-5165

PROMIN Low Protein Rice Pudding	4 x 69g	From weaning	Apple	119/100g prepared product	27/100g prepared product	1.4/100g prepared product	326-3910
PROMIN Low Protein Rice Pudding	4 x 69g	From weaning	Banana	122/100g prepared product	26/100g prepared product	1.8/100g prepared product	326-3902
PROMIN Low Protein Rice Pudding	4 x 69g	From weaning	Original	122/100g prepared product	26/100g prepared product	1.8/100g prepared product	324-6873
PROMIN Low Protein Rice Pudding	4 x 69g	From weaning	Strawberry	119/100g prepared product	26/100g prepared product	1.4/100g prepared product	326-3928
PROMIN Low Protein Sausage Mix	4 x 30g	From weaning	Apple & Sage	148/100g prepared product	28/100g prepared product	4.2/100g prepared product	341-4968
PROMIN Low Protein Sausage Mix	4 x 30g	From weaning	Original	150/100g prepared product	27/100g prepared product	4.6/100g prepared product	341-4984
PROMIN Low Protein Sausage Mix	4 x 30g	From weaning	Tomato & Basil	135/100g prepared product	31/100g prepared product	1.1/100g prepared product	341-4950
PROMIN Low Protein Snax	12 x 25g	3y- adult	Cheese & Onion	369	78	22	402-4188
PROMIN Low Protein Snax	12 x 25g	3y- adult	Jalapeno	457	78	16	402-4170
PROMIN Low Protein Snax	12 x 25g	3y- adult	Mixed	465	78	17	356-1586
PROMIN Low Protein Snax	12 x 25g	3y- adult	Ready Salted	465	78	17	402-444

PROMIN Low Protein Snax	12 x 25g	3y- adult	Salt & Vinegar	458	78	16	402-4162
PROMIN Low Protein Soups	4 x 23g	3y- adult	Creamy Chicken Soup & Croutons	40/100g prepared product	8.5/100g prepared product	0.5/100g prepared product	384-5294
PROMIN Low Protein Soups	4 x 23g	3y- adult	Creamy Tomato & Croutons	35/100g prepared product	6.9/100g prepared product	0.7/100g prepared product	383-7465
PROMIN Low Protein Soups	4 x 23g	3y- adult	Minestrone & Croutons	38/100g prepared product	8.2/100g prepared product	0.3/100g prepared product	383-7481
PROMIN Low Protein Soups	4 x 23g	3y- adult	Pea & Mint Soup & Croutons	35/100g prepared product	6.6/100g prepared product	0.2/100g prepared product	383-7473
PROMIN Low Protein XPots	4 x 60g	3y- adult	All Day Scramble	140/100g prepared product	23/100g prepared product	5.1/100g prepared product	369-3116
PROMIN Low Protein XPots	4 x 60g	3y- adult	Beef & Tomato	140/100g prepared product	24/100g prepared product	4.8/100g prepared product	369-3108
PROMIN Low Protein XPots	4 x 60g	3y- adult	Chip Shop Curry	137/100g prepared product	23/100g prepared product	4.7/100g prepared product	369-3082
PROMIN Low Protein XPots	4 x 60g	3y- adult	Rogan Style Curry	136/100g prepared product	23/100g prepared product	4.5/100g prepared product	369-3074
COMPANY: TARANIS							

TARANIS Low Protein Biscuits with Caramel Shards	130g	3y- adult		483	79.5	18.2	401-5731
TARANIS Low Protein Cake Bars	6 x 40g	3y- adult	Apricot	372	58	15.2	357-9471
TARANIS Low Protein Cake Bars	6 x 40g	3y- adult	Lemon	372	58	15.2	341-5064
TARANIS Low Protein Cake Bars	6 x 40g	3y- adult	Pear	372	58	15.2	357-9448
TARANIS Low Protein Chocolate Chip Biscuits	120g	3y- adult		491	75.7	18.6	401-5772
TARANIS Low Protein Chocolate Chip Cookies	135g	3y- adult		484	75.9	19.6	401-5772
TARANIS Low Protein Fish Substitute	4 x 62g	3y- adult		360	72.7	0.4	401-7695
TARANIS Low Protein Hazelnut Spread	230g	3y- adult	Chocolate	347	90	1.7	387-0225
TARANIS Low Protein Mix for Pancakes & Waffles	300g	3y- adult		353	86.5	0.4	403-5085
TARANIS Low Protein Natural Cake Mix	300g	3y- adult		366	84.6	1.3	403-4963
TARANIS Low Protein Pause Dessert	4 x 125g	3y- adult	Caramel	181	26.9	8.2	401-7687
TARANIS Low Protein Pause Dessert	4 x 125g	3y- adult	Strawberry	181	27.1	8.1	401-7679
TARANIS Low Protein Raspberry Shortbread Biscuits	120g	3y- adult	Raspberry	476	76.6	18.6	401-5756
TARANIS Low Protein Risotto	4x300g	3y- adult		103	14.0	5	407-1965

Substitute							
TARANIS Low Protein Rusks	250g	3y- adult		413	76.3	10.0	401-5723
TARANIS Low Protein Shortbread Biscuits	120g	3y- adult		484	77.6	19.1	401-5749
COMPANY: VITAFLO LTD							
VITAFLO Mini Crackers	15 x 40g	3y- adult		444	77	15	381-5727
VITAFLO Vitabite	7 x 25g	1y- adult		549	61	33	277-1053

References

WHO/FAO/UNU 2007. World Health Organisation 2007. Protein and amino acid requirements in human nutrition. Report of a joint WHO/FAO/UNU expert consultation (WHO Technical Report Series 935). *United Nations University.*