



**DIETETIC NATIONAL FORMULARY FOR INHERITED METABOLIC DISORDERS
ADULTS AND PAEDIATRICS**

Acknowledgement:

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Chair of BIMDG-DG**

BIMDG DIETETIC METABOLIC FORMULARY

INTRODUCTION

www.bimdg.org.uk

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DISORDERS REQUIRING LOW AND MODIFIED FAT DIETS

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- _____

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CARBOHYDRATE DISORDERS

- _____
- _____

OTHER DISORDERS REQUIRING A MODIFIED CARBOHYDRATE DIET

- _____

EMERGENCY REGIMENS

NUTRITIONAL SUPPLEMENTATION/ SUPPORT

- _____
- _____
- _____
- _____
- _____

GLOSSARY OF TERMS

ACBS

BIMDG

AA

CHO

CGMP

DHA

EPA

FOS

GOS

LCPUFA

LCT

MCT

PHE

AMINO ACIDOPATHIES

HOMOCYSTINURIA (HCU)

Background information

Dietary treatment requires:

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-
- ---

Protein substitutes

Administration:

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential.

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												

COMPANY: VITAFLO

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MAPLE SYRUP URINE DISEASE (MSUD)

Background information

Dietary treatment requires:

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Protein substitutes

Dosage of protein substitute:

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-
-
-

Dosage of valine and isoleucine supplementation

Administration:

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential.

EMERGENCY MANAGEMENT.

Ingredients added to emergency regimens include:

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-
-
-
-

Dose:

Administration

[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												

	5g unit											
COMPANY: VITAFLO UK												

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PHENYLKETONURIA (PKU)

Background information

Dietary treatment requires:

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Role of protein substitute

Dosage of protein substitute

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Administration of protein substitutes:

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.

Additional notes

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													

neutral

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													

neutral

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													

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
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COMPANY: MEVALIA

neutral

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
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COMPANY: NUTRICIA

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neutral

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
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COMPANY: VITAFLO UK

			Red										
			White										
			Green										
			Gold										

			Yellow										
			Red White Green Gold Yellow										

neutral

TYROSINAEMIA

Background information

Dietary treatment for Tyrosinaemia type I and II requires:

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-
- ---

Dietary treatment for Tyrosinaemia type III

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Protein substitutes

Dosage of protein substitute

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-
-
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Protein substitutes

Administration:

Pregnancy

and the use of new dietary products may be essential.

Pregnancy is often a time of intensified dietary management

Alkaptonuria (AKU)

off label

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												
COMPANY: VITAFLO UK												

COMPANY: GALEN												

ORGANIC ACIDURIAS

GLUTARIC ACIDURIA TYPE 1

Background information

Dietary treatment requires:

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Protein substitutes

Dosage of protein substitute

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-

Protein substitute

Administration:

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential (particularly protein substitutes).

EMERGENCY MANAGEMENT

emergency regimen

-
- lysine-free, low tryptophan amino acid supplements
-

[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												
COMPANY: VITAFLO UK												

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METHYLMALONIC ACIDAEMIA AND PROPIONIC ACIDAEMIA (MMA/PA)

Background information

Dietary treatment

Protein substitutes

Infancy

Children/Adults/Pregnancy

energy supplements

Dosage of protein substitute/precursor free amino acids

Administration

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												
COMPANY: VITAFLO UK												

Energy supplements												

ISOVALERIC ACIDAEMIA (IVA)

Background information

Dietary treatment

Protein substitutes

Infancy:

Children/Adults/Pregnancy

energy supplements table 16

Dosage of protein substitute

Administration

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												
COMPANY: VITAFLO UK												
Energy supplements												

PYRIDOXINE DEPENDENT EPILEPSY (PDE)/ ANTIQUITIN DEFICIENCY

Background information

see GA1 dietary treatment).

Dietary treatment requires:

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Protein substitutes

Dosage of protein substitute

-
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-
-
-

UREA CYCLE DISORDERS

Background information

Dietary treatment requires:

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-
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-

Essential amino acids

Children/Adults/Pregnancy

Essential amino acids:

Essential amino acids are given at least

Administration:

Energy 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

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												
	Dose 5g											
COMPANY: VITAFLO UK												
Energy supplements												

OTHER CONDITIONS NEEDING PROTEIN RESTRICTION

3 HYDROXY 3 METHYLGLUTARYL HMG-COA LYASE DEFICIENCY

Background information

Dietary treatment requires:

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Administration

EMERGENCY MANAGEMENT

[See BIMDG emergency guidelines for further information.](#)

GYRATE ATROPHY / ORNITHINE AMINOTRANSFERASE DEFICIENCY

Background information

Dietary treatment requires:

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-
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-
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Essential amino acids

Dose of essential amino acids:

Administration

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

<i>Energy supplements</i> _____	
<i>Vitamin and mineral/ essential fatty acid supplements -</i> _____	Page 52 of 99
<i>Essential amino acid supplement</i> _____	Return to CONTENTS

SINGLE DOSE L- AMINO ACIDS

Background information

-
-
-

Administration:

Emergency management:

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												

COMPANY: VITAFLO UK

DISORDERS OF FAT METABOLISM

FATTY ACID OXIDATION DISORDERS

DISORDERS OF LONG CHAIN FATTY ACID OXIDATION

Defects of the carnitine cycle

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-

Defects of β -oxidation

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-
-

Background information

Treatment requires:

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-

MCT products for long chain fatty acid oxidation disorders

Dose:

Administration:

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.

EMERGENCY MANAGEMENT

[See BIMDG emergency guidelines for further information.](#)

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											
COMPANY: VITAFLO UK											

Medium chain acyl CoA dehydrogenase (MCADD) deficiency

Background information

MCT products are contraindicated.

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

KETOGENIC DIETS

Background information

Type of ketogenic diets

-
-
-

GLUT 1 DEFICIENCY

Background information

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											

COMPANY: VITAFLO LTD											

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

Background information

Dietary treatment requires:

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-
-
-
-
-

Dose of Lorenzo/GTO oil

Administration

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												

DISORDERS REQUIRING A MODIFIED CARBOHYDRATE DIET

GLYCOGEN STORAGE DISEASES (GSD)

Background information

Dietary treatment requires:

- *Glycosade*
- */ Glycosade*

Dose: Glucose polymer)

Slow release carbohydrate (cornstarch/ *Glycosade*)

Administration

Protein supplementation

Pregnancy

Pregnancy is often a time of intensified dietary management and the use of new dietary products may be essential.

EMERGENCY MANAGEMENT

[See BIMDG emergency guidelines for further information.](#)

Table 13

COMPANY: NUTRICIA LTD											

COMPANY: VITAFLO UK											

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											
COMPANY: SUPERMARKETS											

GALACTOSAEMIA

Background information

Dietary treatment requires:

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-
-

Dose of lactose free infant formula:

Administration:

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										
COMPANY: MEAD JOHNSON										
COMPANY: NUTRICIA LTD										

CITRIN DEFICIENCY OR CITRULLINEMIA TYPE II (CTLN2)

Background information

Dietary treatment requires:

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-
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-

Protein and fat supplements

Dose:

EMERGENCY REGIMEN

[See BIMDG emergency guidelines for further information.](#)

Table 15

COMPANY: NUTRICIA LTD											
COMPANY: VITAFLO LTD											
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											

EMERGENCY REGIMENS reference to energy supplements [table 16](#)

Background information

Administration

Dosage

[See BIMDG emergency guidelines for further information.](#)

KETOTIC HYPOGLYCAEMIA (KH)

Background

EMERGENCY REGIMEN table 16

[See BIMDG emergency guidelines for further information.](#)

ENERGY SUPPLEMENTS

Background information

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**Fat emulsions may be contraindicated in some metabolic conditions*

Dose of energy supplements

Administration:

PROTEIN SUPPLEMENTS

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											
Fat energy supplements											

Carbohydrate and fat energy supplements

Protein energy supplements

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											
											353 10 15 20 25
Fat energy supplements											
Carbohydrate and fat energy supplements											

Protein energy supplements

COMPANY: Nutrinovo

Protein supplement

ProSource TF	45mL sachet powder	3y-adult	Neutral	Protein hydrolysed collagen	44	1	11	0	No	None	348-9622
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VITAMIN AND MINERAL / ESSENTIAL FATTY ACID SUPPLEMENTS

Background information

Dose

Administration

Essential fatty acids (EFA)

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												
COMPANY: VITAFLO UK												

COMPANY: ALLIANCE PHARMACEUTICALS												
COMPANY: ESSENTIAL PHARMECEUTICALS LTD												

FLAVOUR SACHETS

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												
COMPANY: VITAFLO UK												
												-

SPECIAL LOW PROTEIN FOODS FOR USE WITH LOW PROTEIN DIETS

Background information

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Table 19

monthly

PLEASE NOTE:

Table 20

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

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								
COMPANY: MEVALIA								
COMPANY: NUTRICIA LTD								
COMPANY: VITAFLO LTD								

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							
COMPANY: JUVELA							
COMPANY: METAX							
COMPANY: MEVALIA LTD							

COMPANY: NUTRICIA LTD							

COMPANY: PK FOODS							
COMPANY: PROMIN							

COMPANY: TARANIS							

t							

COMPANY: VITAFLO LTD							

References

WHO/FAO/UNU 2007.

United Nations University.