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British Inherited Metabolic Disease Group

EMERGENCY FEEDS FOR GLUTARIC ACIDURIA TYPE 1 (GA1)

CLICK HERE FOR EMERGENCY FEEDS SUITABLE FOR FULL ENTERAL FEEDING

CLICK HERE FOR CONCENTRATED BRANCHED CHAIN FREE L-AMINO ACIDS TO RUN WITH INTRAVENOUS FLUIDS

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EMERGENCY FEEDS FOR GLUTARIC ACIDURIA TYPE 1

FULL ENTERAL EMERGENCY FEED

Lysine-free, low tryptophan L-amino acids and glucose polymer

Use patients own emergency regimen (ER) recipe

Use aged based ER recipes below if not available

If emergency feed ingredients are not available use IV guidelines

Age ≤ 1year	Age 1 to 6 years	Age ≥7years
CLICK HERE FOR RECIPE	CLICK HERE FOR RECIPE	CLICK HERE FOR RECIPE (glucose polymer only)

EMERGENCY FEED ADMINISTRATION

- Stop natural protein (from infant formula/food)
- Give volume of emergency feed based on body weight
- Give emergency feed orally every 2 hours day and night
- If emergency feed is not tolerated or fluid requirement not met, administer by bolus or continuous tube feed, without delay
- Use emergency feed for a maximum of 24-36 hours
- Introduce usual diet/feeds as soon as clinically stable

MEDICATIONS

- L-carnitine: continue as prescribed for illness
- Antipyretics: as clinically indicated

Contact the child's specialist metabolic team and Dietitian for further advice on the ER and introduction of usual diet/feeds

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ENTERAL FEEDING ONLY EMERGENCY REGIMEN FOR CHILDREN WITH GA1 UNDER 1 YEAR OF AGE

Aim: to provide 1-1.5 g/kg/day protein equivalent from lysine-free/low tryptophan L-amino acids with glucose polymer added to 10% carbohydrate concentration.

Example based on: GA1 Anamix Infant (Nutricia), lysine-free, low tryptophan L-amino acids and **glucose polymer**

How to make	Ingredients	Amounts
Step 1	**GA1 Anamix Infant** 	• measure 3 level scoops of GA1 Anamix Infant (use GA1 Anamix scoop) into a baby bottle
Step 2	**Glucose polymer powders** e.g. Maxijul, Polycal, SOS powder, Vitajoule	• add 15g glucose polymer powder For weight of glucose polymer powder in scoop – click here for details.
Step 3	**Sterile water**	• make up to 200ml with sterile water
Analysis per 100ml of feed recipe: 63 kcals, 1.0g protein equivalent, 10g carbohydrate.		

Guide for feed volume to be administered: based on infant weight

Wt (kg)	ml per kg/bodyweight/day	Feed rate: ml/hour (oral or via nasogastric tube)
3	150	20 ml
4	150	25 ml
5	150	30 ml
6	150	40 ml
7	150	45 ml
8	150	50 ml
9	130	50 ml
10	120	50 ml

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ENTERAL FEEDING ONLY EMERGENCY REGIMEN FOR CHILDREN WITH GA1 >1-6 YEARS OF AGE

Aim: to provide 1g/kg/day protein equivalent from lysine-free/low tryptophan L-amino acids with glucose polymer added to 15% or 20% carbohydrate concentration based on age.

Example based on: GA Amino5 (Vitaflo International), lysine-free, low tryptophan L-amino acids + **glucose polymer**.

Age years	**Weight (kg)** 50th centile for boys	**Feed rate: ml/hour (oral or NG tube)**	**Recipe for GA Amino5 + glucose polymer*** 		
1 to 2	10-13	50 - 55	1 sachet (6g) GA Amino 5	60g glucose polymer	made up to 400ml with sterile water
Analysis **per 100ml** of feed recipe: 1.3g lysine-free/low tryptophan protein equivalent, 15g carbohydrate and 65 kcals					
2 to 4	13-16	50 - 55	1 sachet (6g) GA Amino 5	80g glucose polymer	made up to 400ml with sterile water
5 to 6	17-20	60 - 65			
Analysis **per 100ml** of feed recipe: 1.3g lysine-free/low tryptophan protein equivalent, 20g carbohydrate and 85 kcals					

*Any glucose polymer powder (Maxijul, Polycal, SOS powder, Vitajoule) is suitable. For weight of glucose polymer powder in scoop – click here for details.
Fluid volume calculations are also detailed here.

ALTERNATIVE LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACID PRODUCT

The quantity of each powder contains same protein equivalent as 1 sachet of GA Amino5 and can be substituted for this. If the L-amino acid supplement contains carbohydrate, reduce glucose polymer in the recipe by the amount it provides.

Product Nutritional analysis	**Amount of powder**	**Protein Equivalent (g)**	**Kcals**	**Carbohydrate (g)**
GA Amino 5 (Vitaflo) 	**1 sachet (6g)**	**5**	**20**	**0**
GA Gel (Vitaflo) 1 sachet weighs 24g 	**12g**	**5**	**41**	**5**
XLYS, TRY Glutaridon (Nutricia) Scoop weights: click here 	**6g**	**5**	**20**	**0**
XLYS, LOW TRY Maxamaid (Nutricia) Scoop weights: click here 	**20g**	**5**	**62**	**10**
GA1 ANAMIX JUNIOR (Nutricia) 	**1 sachet = 18g**	**5**	**66**	**5**

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ENTERAL FEEDING ONLY EMERGENCY REGIMEN FOR CHILDREN WITH GA1 AGED ≥ 7 YEARS

From age 7 years, there is no recommendation to give lysine-free/low tryptophan L-amino acids in the emergency feeds.

Use standard emergency feeds based on glucose polymer only. Click below for recipe details and fluid volumes based on weight.

For 7 to 9 years of age: use 20% glucose polymer

For ≥10 year of age: use 25% glucose polymer

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CONCENTRATED LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACIDS ENTERAL FEEDS PLUS IV FLUIDS

(On IV management page)

CONCENTRATED LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACIDS PLUS IV FLUIDS	
IV 10% dextrose/0.45% saline fluids (see medical management – **CLICK HERE for instructions**)	
Age ≤ 1year **CLICK HERE FOR AMINO ACID RECIPE**	**Age 1 to 6years** **CLICK HERE FOR AMINO ACID RECIPE**
MEDICATIONS L-carnitine: continue IV as prescribed Antipyretics: as clinically indicated	
Once clinically stable, Titrate onto usual low protein diet and lysine free/low tryptophan L-amino acid supplement	

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ENTERAL FEEDS TO GIVE WITH IV FLUIDS FOR INFANTS WITH GA1, UNDER 1 YEAR OF AGE:

CONCENTRATED LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACIDS

Aim: to provide 1g/kg/day protein equivalent from lysine-free/low tryptophan L-amino acid with glucose polymer added to 10% carbohydrate concentration in a small volume recipe.

Example based on: GA Amino5 (Vitaflo International), lysine-free, low tryptophan amino acids + **glucose polymer** (e.g. Maxijul, Polycal, SOS powder, Vitajoule)

Wt (kg) of infant	**Feed rate: ml/hour continuous via NG tube (plus IV fluids)**	**Recipe for: GA Amino5 + glucose polymer***		
3	3	2 sachet (12g) GA Amino 5 	20g glucose polymer	made up to 200ml with sterile water
4	4			
5	5			
6	6			
7	7			
8	8			
9	8			
10	9			
Nutritional analysis per **100ml** of feed recipe: **5g** lysine-free/low tryptophan protein equivalent, 10g carbohydrate and 60kcals. **20ml** feed recipe provides **1g** lysine-free/low tryptophan protein equivalent.				

*Any glucose polymer powder (Maxijul, Polycal, SOS powder, Vitajoule) is suitable. For weight of glucose polymer powder in scoop – click here for details.

This volume is given in addition to IV maintenance fluids to maintain hydration

ALTERNATIVE LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACID PRODUCT

Amount of powder contains same protein equivalent as 1 sachet of GA Amino 5.

Product Nutritional analysis	**Amount of powder**	**Protein equivalent g**	**Kcals**	**Carbohydrate g**
GA Amino5 (Vitaflo) 	**1 sachet (6g)**	**5**	**20**	**0**
XLYS LOW TRY Glutaridon (Nutricia) 	**6g** Scoop weights: click here	**5**	**20**	**0**

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ENTERAL FEEDS TO GIVE WITH IV FLUIDS FOR CHILDREN WITH GA1 >1 TO ≤ 6 YEARS OF AGE:

CONCENTRATED LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACIDS

Aim: to provide 1g/kg/day protein equivalent from lysine-free/low tryptophan L-amino acids with glucose polymer added to 15 or 20% carbohydrate concentrations (based on age) in a small volume recipe.

Example based on: GA Amino5 (Vitaflo International), lysine-free, low tryptophan L-amino acids + **glucose polymer** (e.g. Maxijul, Polycal, SOS powder, Vitajoule)

<table>
<tr><th>Age
years</th><th>Weight
(kg)
50th
centile wt
for boys</th><th>Feed rate:
ml/hour
continuous via
NG tube
(plus IV fluids)</th><th colspan="3">Recipe for GA Amino5 + glucose polymer*</th></tr>
<tr><td>1 to 2</td><td>10-13</td><td>10-13</td><td>2 sachets
GA Amino 5</td><td>30g
glucose polymer</td><td>made up to 200 ml with
sterile water</td></tr>
<tr><td colspan="6">Nutritional analysis per 100ml of feed recipe:
5g lysine-free/low tryptophan protein equivalent, 15g carbohydrate and 80kcals
20ml of feed recipe provides 1g lysine-free/low tryptophan protein equivalent</td></tr>
<tr><td>2 to 4</td><td>13-16</td><td>13-15</td><td rowspan="2">2 sachets
GA Amino 5</td><td rowspan="2">40g
glucose polymer</td><td rowspan="2">made up to 200 ml with
sterile water</td></tr>
<tr><td>5 to 6</td><td>17-20</td><td>16-18</td></tr>
<tr><td colspan="6">Nutritional analysis per 100ml of feed recipe:
5g lysine-free/low tryptophan protein equivalent, 20g carbohydrate and 100kcals
20ml of feed recipe provides 1g lysine-free/low tryptophan protein equivalent.</td></tr>
</table>

*Any glucose polymer powder (Maxijul, Polycal, SOS powder, Vitajoule) is suitable. For weight of glucose polymer powder in scoop – click here for details.

This volume is in addition to IV maintenance fluids to maintain hydration

ALTERNATIVE LYSINE FREE/LOW TRYPTOPHAN L-AMINO ACIDS

The amount of powder contains same protein equivalent as 1 sachet of GA Amino5 and can be substituted for this.

Product Nutritional analysis	Amount of powder	Protein g	Kcals	Carbohydrate g
GA Amino 5 (Vitaflo) 	**1 sachet (6g)**	**5**	**20**	**0**
XLYS LOW TRY Glutaridon (Nutricia) 	**6g** Scoop weights: click here	**5**	**20**	**0**

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FOR CHILDREN WITH GA1 AGED ≥7 YEARS

From age 7 years, there is currently no recommendation to give enteral lysine-free/low tryptophan L-amino acids with IV 10% dextrose /0.45% saline fluids.