



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


<b>FULL ENTERAL EMERGENCY FEED</b> <b>Lysine-free, low tryptophan L-amino acids and glucose polymer</b>  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		
<b>Age <math>\leq</math> 1year</b>  <a href="#"><u>CLICK HERE FOR RECIPE</u></a>	<b>Age 1 to 6 years</b>  <a href="#"><u>CLICK HERE FOR RECIPE</u></a>	<b>Age <math>\geq</math> 7years</b>  <a href="#"><u>CLICK HERE FOR RECIPE</u></a> <b>(glucose polymer only)</b>
<b>EMERGENCY FEED ADMINISTRATION</b> <ul style="list-style-type: none"><li>• Stop natural protein (from infant formula/food)</li><li>• Give volume of emergency feed based on body weight</li><li>• Give emergency feed orally every 2 hours day and night</li><li>• If emergency feed is not tolerated or fluid requirement not met, administer by bolus or continuous tube feed, without delay</li><li>• Use emergency feed for a maximum of 24-36 hours</li><li>• Introduce usual diet/feeds as soon as clinically stable</li></ul>		
<b>MEDICATIONS</b> <ul style="list-style-type: none"><li>• L-carnitine: continue as prescribed for illness</li><li>• Antipyretics: as clinically indicated</li></ul>		
<b>Contact the child's specialist metabolic team and Dietitian for further advice on the ER and introduction of usual diet/feeds</b>		

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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
<b>Step 1</b>	<b>GA1 Anamix Infant</b> 	<ul style="list-style-type: none"> <li>• measure 3 level scoops of GA1 Anamix Infant (use GA1 Anamix scoop) into a baby bottle</li> </ul>
<b>Step 2</b>	<b>Glucose polymer powders</b> e.g. Maxijul, Polycal, SOS powder, Vitajoule	<ul style="list-style-type: none"> <li>• add 15g glucose polymer powder</li> </ul> <p><a href="#">For weight of glucose polymer powder in scoop – click here for details.</a></p>
<b>Step 3</b>	<b>Sterile water</b>	<ul style="list-style-type: none"> <li>• make up to 200ml with sterile water</li> </ul>
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) 50 <sup>th</sup> 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 <b>per 100ml</b> 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 <b>per 100ml</b> 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) <a href="#">Scoop weights: click here</a> 	6g	5	20	0
XLYS, LOW TRY Maxamaid (Nutricia) <a href="#">Scoop weights: click here</a> 	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  $\geq 7$  YEARS**

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

However in practice individual units may vary and all such patients should be discussed with their specialist metabolic unit regarding whether or not to give lysine-free/low tryptophan L-amino acids in the emergency feeds.

If glucose polymer only is recommended, click below for recipe details and fluid volumes based on weight.

[For 7 to 9 years of age: use 20% glucose polymer](#)

[For  \$\geq 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  $\leq$  1year**

**[CLICK HERE FOR  
AMINO ACID  
RECIPE](#)**

**Age 1 to 6 years**

**[CLICK HERE FOR  
AMINO ACID  
RECIPE](#)**

**Age  $\geq$  7 years**

**[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	<div>2 sachet (12g) GA Amino 5</div> <div></div>	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 <b>100ml</b> of feed recipe: <b>5g</b> lysine-free/low tryptophan protein equivalent, 10g carbohydrate and 60kcal. <b>20ml</b> feed recipe provides <b>1g</b> 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 <a href="#">Scoop weights: click here</a>	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)



Age years	Weight (kg) 50 <sup>th</sup> centile wt for boys	Feed rate: ml/hour continuous via NG tube (plus IV fluids)	Recipe for GA Amino5 + glucose polymer*		
1 to 2	10-13	10-13	2 sachets GA Amino 5	30g glucose polymer	made up to 200 ml with sterile water
Nutritional analysis per <b>100ml</b> of feed recipe: 5g lysine-free/low tryptophan protein equivalent, 15g carbohydrate and 80kcal <b>20ml</b> of feed recipe provides <b>1g</b> lysine-free/low tryptophan protein equivalent					
2 to 4	13-16	13-15	2 sachets GA Amino 5	40g glucose polymer	made up to 200 ml with sterile water
5 to 6	17-20	16-18			
Nutritional analysis per <b>100ml</b> of feed recipe: <b>5g</b> lysine-free/low tryptophan protein equivalent, 20g carbohydrate and 100kcal <b>20ml</b> of feed recipe provides <b>1g</b> 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 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	Amount of powder	Protein g	Kcals	Carbohydrate g
<b>Nutritional analysis</b> GA Amino 5 (Vitaflo) 	1 sachet (6g)	5	20	0
XLYS LOW TRY Glutaridon (Nutricia) 	6g <a href="#">Scoop weights: click here</a>	5	20	0

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**FOR CHILDREN WITH GA1 AGED  $\geq 7$  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.

However in practice individual units may vary and all such patients should be discussed with their specialist metabolic unit regarding whether or not to give enteral lysine-free/low tryptophan L-amino acids alongside IV fluids.

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