

Dr M Champion, Consultant 020 7188 0848
 Dr H Mundy, Consultant 020 7188 0848
 Dr R Vara, Consultant 020 7188 4004
 Metabolic Registrar bleep 1460
 Metabolic Nurse Specialist 020 7188 0855
 Metabolic Dietitian 020 7188 4008

Metabolic Department
 Sky (6th Floor)
 Evelina Children's Hospital
 St Thomas' Hospital
 Lambeth Palace Road
 London SE1 7EH

Switchboard tel: 020 7188 7188

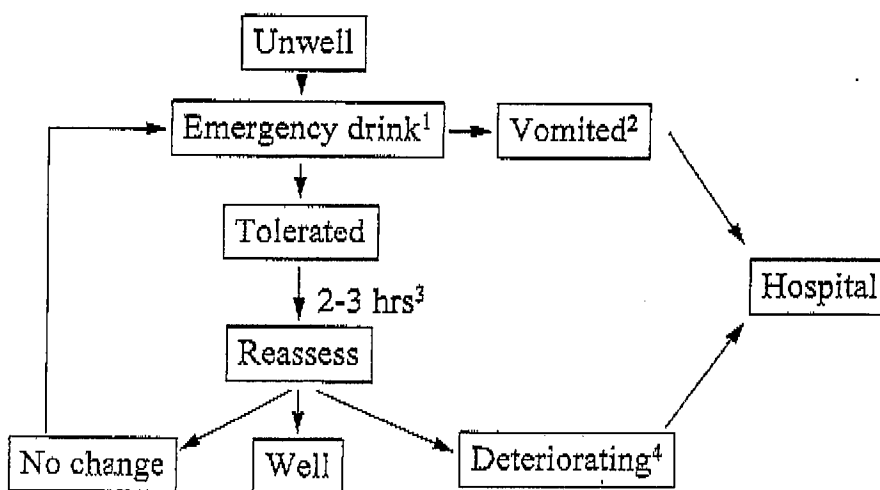
Guidelines for the management of patients with Glutaric Aciduria type I (GA1) during an acute illness

Background

Glutaric Aciduria is an inherited disorder that interferes with the break-down of certain amino acids. Infections, fasting or vomiting can lead to serious illness, with 'encephalopathy' - a reduced level of consciousness and other neurological abnormalities. Following these episodes, patients often have severe neurological problems, including an 'extrapyramidal' movement disorder. The damage results from the accumulation of glutaric acid and other toxic metabolites. For reasons that are unclear, episodes of encephalopathy and neurological damage are unusual except between the ages of 3 months and 4 years. Treatment aims to minimise the accumulation of toxic metabolites by preventing protein breakdown and to promote their excretion by use of carnitine (100 mg/kg/day).

Management at home

follow this procedure:



Notes:

Volumes and concentrations of carbohydrate-containing drinks vary with age - each child will have their own instructions. Guidelines are presented below, derived from Dixon & Leonard, 1992, Arch Dis Child 67:1387-91. Glucose polymer preparations include maxijul, polycal etc.

| Age (years) | Glucose polymer concentration (g/100ml) | Total daily volume* |
|-------------|-----------------------------------------|---------------------|
| 0-1 | 10 | 150-200 ml/kg |
| 1-2 | 15 | 95 ml/kg |
| 2-6 | 20 | 1200-1500 ml |
| 6-10 | 20 | 1500-2000 ml |
| >10 | 25 | 2000 ml |

*For each drink the volume will be this figure divided by 12 or 8 (depending on frequency).

1. In this condition, whenever a patient vomits, admission is needed for an intravenous infusion of glucose.
2. Patients should be reviewed & given carbohydrate-containing drinks every 2 hrs, day and night. This can sometimes be increased to 3 hrs in older children.
3. Under these circumstances, admission is URGENT.

Hospital management

1. Patients should always be admitted if the parent is sufficiently concerned to bring the child to hospital.
2. Ascertain the reason why the child has been brought to hospital (vomiting, refusing drinks etc) & assess the patient quickly. If the problem is refusal of drinks and there is no suggestion of encephalopathy (such a glazed look, lethargy or drowsiness), the child can be offered another drink orally or given it through a nasogastric tube. Otherwise an intravenous infusion should be started immediately.
3. If the child is unwell, check U&Es, glucose and blood gas but these are seldom abnormal even during episodes of severe encephalopathy. Management decisions should be based on the **clinical** status.
4. Most children will require an intravenous infusion of glucose, which must be started without delay. Start with an infusion of 10% glucose, at the rates suggested below. Check electrolytes and adjust the infusion as appropriate.

| Age (years) | Weight (kg) | Glucose to be provided | 10% glucose infusion rate |
|-------------|-------------|------------------------|---------------------------|
| 0-3 | | 10 mg/kg/min | 150 ml/kg/day |
| 3-6 | | 8 mg/kg/min | 120 ml/kg/day |
| >6 | <30 | 6 mg/kg/min | 90 ml/kg/day |
| >6 | 30-50 | 4.5 mg/kg/min | 67 ml/kg/day |
| >6 | >50 | 3 mg/kg/min | 45 ml/kg/day |

5. Give carnitine intravenously (100 mg/kg/day).
6. If hyperglycaemia develops, Contact Metabolic team for advice and consideration for transfer. (see contact details below)
7. If there is any hint of incipient encephalopathy, start neurological observations - at least hourly - & seek specialist help. If there is any concern about cerebral oedema, fluid volumes should be reduced and given centrally as concentrated solutions (eg 20% dextrose).
8. Monitor U&Es, glucose, blood gas, etc. up to 6 hourly according to the clinical state.

9. If the child improves, allow him/her to eat and drink when he/she wants to (unless further vomiting seems likely).

Contact Details:

Evelina Children's Hospital 020 7188 7188

Metabolic Registrar bleep 1460 (Mon to Fri , 9-5)

Out of hours ask for "Specialities Registrar" or Metabolic Consultant on call

Dietitian bleep 1220 / 1331

Metabolic Nurse Specialist pager 08700 55 55 00 No: 838680 or 863656

Dr M Champion, Consultant 020 7188 4694
 Dr H Mundy, Consultant 020 7188 0848
 Metabolic Registrar bleep 1460
 Metabolic Nurse Specialist 020 7188 0855
 Metabolic Dietitian 020 7188 4008

Metabolic Department
 Sky (6th Floor)
 Evelina Children's Hospital
 St Thomas' Hospital
 Westminster Bridge Road
 London SE1 7EH

Switchboard tel: 020 7188 7188

Emergency Regimen (10% Carbohydrate)

The emergency regimen is 10% carbohydrate solution + GA Gel

This emergency regimen should be given if your child becomes unwell e.g. diarrhoea, vomiting, high temperature or any illness associated with loss of appetite.

- Give the emergency regimen of regular glucose polymer drinks.
- Drinks should be given every 2 hours (day & night)
- If your child does not tolerate the emergency regimen contact your doctor or the hospital as soon as possible.
- After 24 hours of being on the emergency regimen the usual diet can be reintroduced, taking extra glucose polymer drinks during the day until the normal diet is resumed.
- If your child is still unwell after 24 hours contact your local hospital.

*100mg/kg/d
 divided
 doses
 go up to
 200mg/kg/d
 add
 ammonia*

Feeding volumes

(The volume given will change at different ages as your child gains weight)

| | |
|-----------|----------------------------------|
| 3kg = | 40ml every 2 hours day & night |
| 3.5kg = | 45ml every 2 hours day & night |
| 4kg = | 50ml every 2 hours day & night |
| 4.5kg = | 55ml every 2 hours day & night |
| 5kg = | 60ml every 2 hours day & night |
| 5.5kg = | 65ml every 2 hours day & night |
| 6kg = | 70ml every 2 hours day & night |
| * 6.5kg = | 75ml every 2 hours day & night * |
| 8kg = | 80ml every 2 hours day & night |
| 9kg = | 90ml every 2 hours day & night |
| 13kg = | 100ml every 2 hours day & night |

Recipes

GA Gel

This must be given as part of the Emergency Regimen. Add a small amount to each of the Emergency Regimen feeds. The usual daily dose should be given.

Recipe using S.O.S - 10 powder

This is made up according to the instructions on the box.
1 sachet of S.O.S -10 is made up to 200ml with cooled, boiled water

Recipe using Maxijul* powder (always use level scoops)

Smaller volume recipe

5 yellow scoops of Maxijul
made up to 200ml using cooled,
boiled water.

Larger volume recipe

4 big blue scoops Maxijul
made up to 1000ml using cooled,
boiled water

or 132g sachet Maxijul
made up to 1200ml using cooled, boiled water.

Recipe using Dioralyte

(this might be prescribed for an illness where diarrhoea is present)

1 sachet Dioralyte
200ml cooled, boiled water
4 yellow scoops of Maxijul or 4 yellow scoops of powder from the SOS sachet

Give volumes according to weight (see previous page)

* Polycose, Caloreen, Vitajoule, Polycal are alternatives to Maxijul

Metabolic Consultant 020 7188 4694/0848
Metabolic Registrar bleep 1460
Metabolic Nurse Specialists 020 7188 0855
Metabolic Dietitians 020 7188 4008

Metabolic Department
Sky (6th Floor)
Evelina Children's Hospital
St Thomas' Hospital
Westminster Bridge Road
London SE1 7EH

Switchboard tel: 020 7188 7188

PAEDIATRIC METABOLIC SERVICE

To call the hospital switchboard please use **020 7188 7188**.
If you need to bleep, ask switchboard for the required bleep number.

Consultants in Paediatric Metabolic Medicine:

Dr Mike Champion
Dr Helen Mundy
Dr Roshni Vara

Consultant's Secretary: (Dr Champion/Dr Mundy)
(Dr Vara)

Valerie Williams 020 7188 0848
Emma Hall 020 7188 4004

Fax:

020 7188 8901

Metabolic Registrar:

Bleep no: 1460

Weekends, bank holidays and after 5pm call switch board and ask for the paediatric specialties registrar covering Evelina Children's Hospital

Metabolic Paediatric Dietitians:

020 7188 4008

Paula Hallam Bleep no: 1220 (Mon, Tue, Wed)
Jo Eardley (Wed, Thurs, Fri) Bleep no: 1220
Karen van Wyk Bleep no: 1331

Paediatric Metabolic Nurse Specialists:

020 7188 0855
Tanya Campbell
Mills Barnard

or via pager Mon - Fri 9am - 5pm
020 7188 7188 ask for pager no: 838680

Appointments: (to change or cancel)

020 7188 7188 ext: 50389/90/91/92