Guy's and St Thomas' NHS
NHS Foundation Trust

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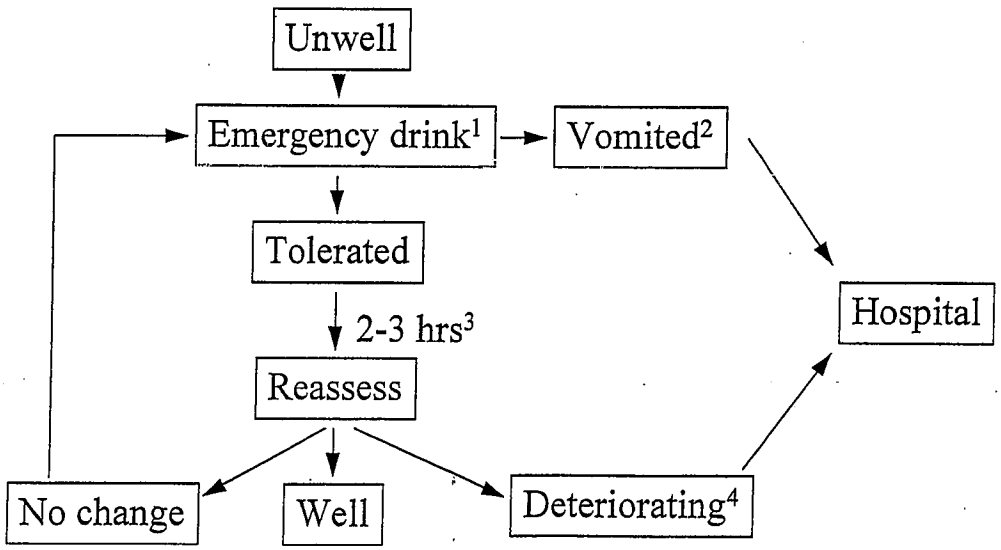
Guidelines for management of patients with VLCAD deficiency during acute decompensation

Background

VLCAD deficiency is a rare disorder of fat breakdown. Most of the time patients are healthy, though we do recommend a special low fat diet. Infections, fasting or vomiting can lead to serious illness, with encephalopathy and rhabdomyolysis (muscle breakdown). This results from the accumulation of toxic fatty acids; hypoglycaemia also occurs, but only at a relatively late stage: it is dangerous to base one's treatment on monitoring of blood glucose, particularly BM stix. Treatment aims to inhibit mobilisation of fat by providing ample glucose - enterally or intravenously.

Management at home

Patients are generally instructed to follow this procedure:



Notes:

1. 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 generally be this figure divided by 12.

2. If the parents are experienced & feel confident that their child is stable, they may try repeating the drink after a short interval but if this is still unsuccessful, admission is needed.

3. Patients should be reviewed & given carbohydrate-containing drinks every 2 hrs, day and night. May be increased to 3 hrs in older children.

4. Under these circumstances, admission is URGENT, particularly if the child has a glazed look.

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 or a single vomit, and there is no suggestion of incipient encephalopathy (such a glazed look or drowsiness), the child can be offered another drink orally or given it through a nasogastic tube.

3. If the child is unwell, check glucose, U&Es, blood gas, possibly NH_3 & other tests as appropriate.

4. Most children will require an intravenous infusion of glucose, which should be started without delay. Start with a small bolus (1 ml/kg of 25% glucose or 2.5 ml/kg of 10% glucose) and follow this with an infusion of 10% glucose, at the rates suggested below. Check electrolytes, but in the short term, it is usually unnecessary to add electrolytes to the infusion. Extra fluid should be given if the child is dehydrated or shocked, as for other patients.

Age (years)	**Weight (kg)**	**Glucose to be provided**	**10% glucose infusion rate**
0-2		10 mg/kg/min	150 ml/kg/day
2-6		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. If there is any hint of incipient encephalopathy, start neurological observations - at least hourly.

6. Despite its limitations (see Background), I suggest monitoring BM 4 hourly if the child's condition is stable. If unstable, monitor BM hourly with blood glucose, U&Es & blood gas 6 hourly.

7. If the patient is not improving, **please contact Evelina on: 020 7188 7188 and ask for the metabolic registrar on bleep 1460.**
If out of hours contact switch board and ask for the specialist registrar on call.

8. Rhabdomyolysis is often precipitated by infections in these children. This will lead to muscle pain and weakness with a high CK and, in severe cases, myoglobinuria (dark brown urine +ve for Hb on dipsticks). Myoglobinuria can occasionally lead to acute renal failure requiring dialysis for short periods.

9. Once the child is improving, allow him high carbohydrate drinks and subsequently low fat solids. Once he is taking drinks reliably, discontinue the intravenous infusion.

We operate a shared care policy so please inform us of any admissions.

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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 Emma Hall	020 7188 0848 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 specialities 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	