BIMDG

British Inherited Metabolic Disease Group

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This protocol has 4 pages

KETOTIC HYPOGLYCAEMIA

ALSO INCLUDES:

ISOLATED GLYCEROL KINASE DEFICIENCY AND GLYCOGEN SYNTHASE DEFICIENCY

(standard version)

- **Please read carefully. Meticulous treatment is important as there is a high risk of serious complications.**
- **If the instructions do not make sense or a problem is not addressed you must discuss your concerns with the consultant on call.**
- **If encephalopathic, give glucose immediately either intravenously or into the buccal cavity using Glucogel ®**

1. Background

Ketotic hypoglycaemia is a poorly understood condition that most commonly affects young children between the ages of one and five years. The history is characteristic. A toddler who is unwell with any intercurrent infection goes to bed having had little to eat in the evening. The next morning they are found encephalopathic or having a fit. The child recovers quickly on being given glucose orally or intravenously. Even if the presentation is typical, it is important to exclude other disorders. For details please refer to the protocol on hypoglycaemia (click here)

2. Admission

Most patients who come to hospital will require admission, for observation at least. Only allow the child home if you and the family are entirely happy and you have discussed the problems with the consultant on call. The family must have a clear management plan and be prepared to return if the child becomes ill again.

- **If there is any doubt at all, the child must be admitted, even if only necessary for a short period of observation.**

3. Initial plan and management in hospital

⇨ If the child is shocked or clearly very ill arrange for admission to ITU/High dependency.

⇨ If admitted to metabolic/general ward, make a careful clinical assessment including blood pressure and a Glasgow coma score (for details click here), even if the patient does not appear encephalopathic. This allows other staff to recognise if the child deteriorates, particularly around the time of a change of shifts.

If the child is still unwell the following additional investigations should be done:

pH and gases
blood glucose
Urea & electrolytes
Full blood count
Blood culture

If the child is febrile, has had a fit and is not improving consider appropriate investigation including lumbar puncture.

4. Management

Management decisions should be based primarily on the **clinical** status. The first decision about therapy is whether the child can be treated orally or will need intravenous therapy.

- Can the child tolerate oral fluids?
- If the child is relatively well - may be treated orally but assess very carefully.
- If the child is obviously unwell - must be treated with intravenous fluids

- **If there is any doubt at all, put up an intravenous line.**

A. ORAL.

If the child is relatively well and not vomiting, oral feeds may be given.

The emergency regimen should be used. This may be given as regular frequent drinks but if the patient is at risk of vomiting or is nauseated fluid should be given either continuously or as small boluses more frequently. For more information about the emergency oral management click here. This may only be necessary for a short time.

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

* If necessary, seek help from your local dietitian. In an emergency a heaped 5 ml medicine spoon holds approximately 7g of glucose polymer.

**For each drink the volume will generally be this figure divided by 12 and given 2 hourly but if the patient is nauseated or refuses try frequent smaller drinks or a continuous naso-gastric infusion.

Electrolytes should be added to the drinks if vomiting and/or diarrhoea is a problem using standard rehydration mixtures following manufacturer's instructions but substituting glucose polymer solution for water

B. INTRAVENOUS.

If the child is unwell

- Give Glucose 200 mg/kg **at once** (2 ml/kg of 10% glucose or 1ml/kg of 20% glucose) over a few minutes. If glucose cannot be given intravenously quickly give Glucogel® into the buccal cavity.
- Observe – if the child recovers quickly continue to observe and start oral feeds.
  - if the child does not recover completely continue with the treatment listed below.
- Give normal saline 10 ml/kg as a bolus immediately after the glucose unless the peripheral circulation is poor or the patient is frankly shocked, give 20 ml/kg normal saline instead of the 10 ml/kg.. Repeat the saline bolus if the poor circulation persists as for a shocked non-metabolic patient.
- Continue with glucose 10% at 5 ml/kg/h **ONLY until next solution is ready– do not leave on this high rate longer than necessary**. – see below
- Quickly calculate the deficit and maintenance and prepare the intravenous fluids
  - Deficit: estimate from clinical signs if no recent weight available
  - Maintenance: Formula for calculating daily maintenance fluid volume (BNF for children) 100ml/kg for 1st 10kg then 50 ml/kg for next 10kg then 20ml/kg thereafter, using calculated rehydrated weight. Deduct the fluid already given from the total for the first 24 hours.
  - Give 0.45% saline/10% glucose (for instructions to make this solution click here).

- Having calculated the deficit and the maintenance, administer the appropriate rate of 0.45% saline/10% glucose to correct the deficit within 24 hours
- Recheck the electrolytes every 24 hours if still on IV fluids.

-Potassium can be added, if appropriate, once urine flow is normal and the plasma potassium concentration is known.

-Treat any infection

5. Progress:

Monitoring: Reassess after 4-6 hours or earlier if there is any deterioration or no improvement
Clinical assessment should include Glasgow coma score (for details click here) and blood pressure.
Blood tests: Glucose (laboratory and bedside strip test)
Urea and electrolytes,

⇨ If improving, continue and for intravenous fluids after 6 hours, please refer to the previous section.

⇨If deteriorating, seek specialist help without delay.

6. Re-introduction of oral feeds: Restart oral feeds as soon as possible; once the child is alert and has stopped vomiting. If necessary, consult your local dietitian for more details.

7. Going Home: Only allow the child home if you and the family are entirely happy and you have discussed the problems with the consultant on call. The family must have a clear management plan and be prepared to return if the child deteriorates. They should also have a clear plan for subsequent intercurrent illness as it is usually straightforward preventing further episodes.

For further information please refer to:

Saudubray J-M, Baumgartner MR, Walter JH. (editors) Inborn Metabolic Diseases. Diagnosis and treatment. 6th Edition. Springer 2016