BIMDG

British Inherited Metabolic Disease Group

Contact Details Name:
Hospital
Telephone:

This protocol has 4 pages

GLYCOGEN STORAGE DISEASE TYPE III– ACUTE DECOMPENSATION

(also called debrancher 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

Patients with glycogen storage disease type III may become hypoglycaemic during a fast, the length of which varies widely between patients. During any illness, patients have a greater tendency to become hypoglycaemic. The treatment aims to maintain normoglycaemia all the time either with frequent oral drinks, a continuous infusion via naso-gastric tube or gastrostomy or an intravenous infusion of glucose. This management should be started as soon as patients become unwell.

The early signs of decompensation may be subtle with changes in behaviour but some patients may remain asymptomatic despite being markedly hypoglycaemic. Others may present with a hypoglycaemic convulsion. Always listen to parents carefully as they probably know much more than you do.

2. Admission

Most patients who present to hospital will require admission as they are likely to have been having treatment already at 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 does not improve.

- **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.

The following blood tests should be done:

Glucose (laboratory and bedside strip test)
Urea & electrolytes
Full blood count
Blood culture

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

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.
- 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
- **It is safe practice to continue also a small volume of appropriate glucose polymer (emergency regimen) as a continuous nasogastric or gastrostomy feed** – this can be given as an *extra 10%* of the IV fluid rate (do not reduce the rate of IV fluids). This will help reduce the risk of hypoglycaemia if an intravenous cannula fails. Should the cannula fail, the glucose polymer rate can be increased until a new cannula is sited.
- 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.

For further information please refer to:
Saudubray J-M, van den Berghe G, Walter JH. (editors) Inborn Metabolic Diseases. Diagnosis and treatment. 5th Edition. Springer 2012