



British Inherited Metabolic Disease Group

PATIENT NAME

HOSPITAL

DATE OF BIRTH

EMERGENCY CONTACT

**9-5pm Monday to Friday
Out of hours**

ADULT EMERGENCY MANAGEMENT

**Fructose 1,6-Bisphosphatase Deficiency
(also known as Fructose 1,6-Diphosphatase deficiency)**

IMMEDIATE ACTIONS

Triage to high priority

Maintain glucose 6-10 mmol/L.

These guidelines are intended for immediate emergency management only. Please contact your local metabolic team early for specific advice on individual patients.

BACKGROUND

Fructose 1,6-bisphosphatase deficiency is a disorder of gluconeogenesis. Most of the time patients are healthy and do not require a special diet. Intercurrent infection, prolonged fasting, alcohol excess, diarrhoea or vomiting can lead to serious illness with drowsiness, tachypnoea, marked acidosis and hypoglycaemia. Hypoglycaemia only occurs at a relatively late stage so that blood glucose / BMstix should not be relied on. Do not delay treatment just because the blood glucose is not low. **The aim should always be to intervene whilst the blood glucose is normal.** Treatment aims to prevent decompensation by giving glucose by mouth or intravenously. Bicarbonate is rarely needed to correct acidosis.

If there is any doubt at all, the patient should be admitted, even if only for a short period of observation.

INITIAL ASSESSMENT AND MANAGEMENT IN HOSPITAL

If the patient is shocked or clearly very ill arrange for admission to ITU / HDU.

Management decisions should be based primarily on the **clinical** status. If the patient is relatively well – they may be treated orally using their [oral emergency regimen \(click here\)](#) (generally give 200ml of a 25% glucose polymer solution every 2 hours) but assess very carefully. If the patient is obviously unwell – they must be treated with intravenous fluids.

Warning: Fructose or sucrose should not be given during acute illness. As oral medications may contain sucrose and sorbitol, only sugar-free medicines should be given (check constituents).

INITIAL INVESTIGATIONS

Blood pH and gases

Glucose

Urea & electrolytes

Full blood count

Liver function tests

Lactate

Other tests as clinically indicated (eg CRP, blood & urine cultures)

TREATMENT

1. Correct dehydration initially with 0.9% NaCl. Correct hypoglycaemia initially with 50ml of 50% dextrose over 30 minutes.
2. Start intravenous 10% dextrose as soon as possible at a rate of 2mls/kg/hr, (e.g. 140 mls/hr in a 70 kg person).
3. Treat any underlying infection or other clinical problem.
4. Give analgesia, anti-pyretic or an anti-emetic as required.

NOTE: Patients can quickly become hypoglycaemic if intravenous access fails / tissues – ensure iv access is secure.

Sodium bicarbonate is not routinely given as any acidosis usually corrects quickly, but sodium bicarbonate may be needed if, after the initial bolus of glucose, the pH is still <7.1 or the pH is deteriorating rapidly or the base deficit is still greater than 15 mmol/L.

Hyperglycaemia can be a problem. If the blood glucose exceeds 10 mmol/L, start an insulin infusion using the local diabetic protocol rather than reducing the glucose intake. **Strict supervision is essential.** National guidelines are available at:

[\(http://www.diabetes.org.uk/About_us/Our_Views/Care_recommendations/The-Management-of-Diabetic-Ketoacidosis-in-Adults/\)](http://www.diabetes.org.uk/About_us/Our_Views/Care_recommendations/The-Management-of-Diabetic-Ketoacidosis-in-Adults/).

COMPLICATIONS

Hepatic or renal injury has been reported from the metabolic crisis associated with fructose 1,6-bisphosphatase deficiency.

MONITORING

Reassess regularly and if there is a change for the worse repeat the clinical assessment and blood tests:

Blood pH and gases

Glucose

Urea & electrolytes

Potassium: Hypokalaemia may occur so plasma potassium concentration should be monitored and corrected appropriately.

Clinical assessment should include the Glasgow Coma Scale and blood pressure. Patients should remain on iv dextrose until tolerating oral food normally. See the BIMDG [oral emergency regimen \(click here\)](#) for more details.

MORE USEFUL INFORMATION

<http://www.bimdg.org.uk/> and click on the red tab for emergency guidelines.

Pubmed: <http://www.ncbi.nlm.nih.gov/pubmed/>