

FRUCTOSAEMIA (Hereditary Fructose Intolerance)**Priority patient: must not wait in A&E / ED**

Patient label

RISK OF HYPOGLYCAEMIA and LIVER FAILURE

Do not wait for signs of decompensation, in all cases initiate management as set out below.

1 EMERGENCY WORKUP**Capillary and venous blood glucose, electrolytes, liver enzymes, PT, Factor V.** Must not delay treatment.**2 TREATMENT TO BE STARTED URGENTLY, without waiting for lab results**

- If hypoglycaemia < 60 mg/dL (3.3 mmol/L):**
 - Administer 1mL/kg of body weight of 30% glucose (max. 30 mL) orally or enterally if conscious.
 - If unconscious, administer 3mL/kg of 10% glucose IV (30% glucose possible by central line or intraosseous route, some teams allow injection of 30% glucose via a peripheral line in cases of refractory hypoglycaemia).
- Check capillary blood glucose 5 minutes later. If still hypoglycaemic, administer a second dose of glucose and check 5 min later.
- Immediately start an infusion even if blood glucose levels have been corrected: Infusion using 10% glucose (=dextrose) in water with standard electrolyte additions* (never pure 10% glucose)

Age	0-24 months	2-4 years	4-14 years	> 14 years / adult	MAX INITIAL RATE
Polyionic 10% glucose (glucose infusion rate)	5mL/kg/h (or 8mg/kg/min)	4mL/kg/h (7 mg/kg/min)	3.5mL/kg/h (6mg/kg/min)	2.5mL/kg/h (4mg/kg/min)	120mL/h (3L/24h)

*e.g. : Bionolyte®, Glucidion®, etc. if no pre-made solution available, use 10% glucose in water + 4g/L NaCl (70 mEq/L) + 2g/L KCl (27 mEq/L)

If IV line is impossible => Nasogastric tube or gastrostomy: prepare the IV fluids listed above and pass them through the tube at the same rates.

**Stop intake of fructose / sucrose / sorbitol. Warning: many oral rehydration solutions contain these!****3 SEVERITY SIGNS = Consult / Transfer to Intensive Care**

- Comatose**
- Signs of severe hepatic failure:** Prothrombin ratio < 30%, factor V < 50%

4 DRUG CONTRAINDICATIONS / GENERAL ADVICE**Absolute contraindication for all drugs containing fructose, sucrose or sorbitol** (check the excipients and the composition of rehydration solutions). By way of example and not an exhaustive list:

- Oral rehydration solutions allowed: Fanolyte®, Novalac Hydranova®, Physiosalt®, Viatol®
 - Oral rehydration solutions forbidden: Adiaril®, Picolite®, Ydrovit®
- } Always check

PATHOPHYSIOLOGY:Hereditary fructose intolerance is an autosomal recessive genetic disease caused by deficiency of the enzyme Aldolase B, which prevents utilisation of fructose by cells. **If even tiny amounts of fructose/sucrose/sorbitol are ingested, the accumulation of fructose in the liver leads to metabolic hepatitis or even acute liver failure (which may be severe) and severe hypoglycaemia.** Treatment consists of:

- a diet free from fructose/sucrose/sorbitol. All fruits and many vegetables, and all sugary foods and drinks are therefore prohibited (fruit juice, cakes, etc.), as well as the majority of ready-made dishes from supermarkets or the catering industry. Hereditary fructose intolerance is asymptomatic as long as the diet is followed.
- Supplementation with vitamin C compensates for the lack of fruit ingestion. Commercially-available oral vitamin C formulations all contain fructose/sucrose/sorbitol. Patients are therefore treated either with a pharmacist-prepared oral capsule, or by a commercially-available intravenous preparation taken orally.

REFERENCE PHYSICIANS AND CONTACT DETAILS

On-call telephone numbers for metabolic emergencies of:

At night, only the medical teams can call in emergency situations and only if the emergency certificate has not been understood or if the clinical state or test results are worrying. As far as possible make calls before night-time.

Secretarial issues must be dealt with the outpatient office during the week or by email addressed to the patient's referring metabolic physician.

Certificate issued on :

Dr



To be completed by each department

Secretarial issues must be dealt with via the medical secretariat during the week or by email addressed to the patient's referring metabolic doctor.

Certificate issued le

Dr

REFERENCE DOCTORS AND CONTACT DETAILS

On-call telephone numbers for metabolic emergencies of:

At night, only the medical teams can call in emergency situations and only if the emergency certificate has not been understood or if the clinical state or test results are worrying. As far as possible make calls before night time.

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Certificate issued on

Dr