



British Inherited Metabolic Disease Group

## **ADULT EMERGENCY MANAGEMENT ORAL EMERGENCY REGIMEN (ER) FOR MAPLE SYRUP URINE DISEASE (MSUD)**

Emergency drinks are high sugar drinks, which should be taken during illness or if you become unwell e.g. nausea, vomiting, diarrhoea, high temperature or any illness resulting in loss of appetite and inability to take your normal diet. Emergency drinks should be **taken every 2 hours day and night**.

**FOR MORE SPECIFIC ADVICE PLEASE CONTACT YOUR LOCAL INHERITED METABOLIC DISEASE TEAM**

### **When to take the oral emergency regimen**

<b>Stage 1 – Possibly unwell</b>	<b>Stage 2 – Unwell but not drowsy and tolerating an oral intake</b>	<b>Stage 3 – Unwell and unable to take/ vomiting drinks or medicine or drowsy</b>
<p>At the first sign of feeling unwell/loss of appetite, take 200ml glucose polymer drink.</p> <p>If better within one hour return to a normal diet.</p> <p><b>If obviously unwell then follow stages 2 or 3 as Appropriate.</b></p>	<p>Commence full emergency regimen; 200mls of glucose polymer drink every 2 hours.</p> <p>If you continue to feel unwell stop normal diet and call your local inherited metabolic team for guidance.</p> <p>Continue to give normal medicines and amino acid supplement(s).</p>	<p>If unable to tolerate the emergency regimen or not taking medicines, if you do not improve, or if you become increasingly unwell contact your doctor or the hospital for admission. You will require 10% glucose to be given intravenously at 2ml/kg/hr <b>e.g. 140ml/hour for a 70kg adult.</b></p> <p><i>If you go into hospital, it is a good idea to take some of your glucose polymer and amino acids with you.</i></p>

## How to prepare the oral emergency regimen

***PLEASE CONTACT YOUR LOCAL INHERITED METABOLIC DISEASE TEAM TO INFORM THEM THAT YOU HAVE STARTED YOUR ORAL EMERGENCY REGIMEN AND TO DISCUSS YOUR DIET.***

### **You will need:**

1. Water.
2. Glass or shaker.
3. Glucose polymer.
4. A big blue scoop (if using Maxijul, Vitajoule or Polycal).

**Choose one of the glucose polymers listed in the table below and follow the recipe across.**

***Remember to continue to take your MSUD amino acid supplement(s) as well.***

## Types of Glucose Polymer available on prescription:



25 % Glucose Polymer Recipes				Oral rehydration solution recipe
Glucose Polymer	Weight of glucose polymer required for single dose	Recipe for 1 single dose (200ml)	Recipe for 24hr (2000ml)	
Maxijul Super Soluble	50g	2 big blue scoops (50g); made up to 200ml with water	20 scoops (500g); add water to 2000ml	Discuss recipe with your metabolic team if required
S.O.S. 25	Pre-measured sachet	Add 1 sachet; made up to 200ml with water	Add 10 sachets; add water to 2000ml	
Vitajoule	50g	2 big blue scoops (50g); made up to 200ml with water	20 scoops (500g); add water to 2000ml	
Polycal Powder	50g	2 big blue scoops (50g); made up to 200ml with water	20 scoops (500g); add water to 2000ml	

**STORE IN FRIDGE & USE WITHIN 24 HOURS**

## EXAMPLE: How to make up 200ml of a 25% glucose polymer drink



1. Add **TWO** level big blue scoops glucose polymer or **ONE** sachet S.O.S 25 – make up to 200ml with water.
2. Shake or stir well until dissolved.
3. Discuss with your metabolic team if you would like to add flavouring

## **Reintroduction of normal protein allowance**

When you have been on the above-mentioned emergency regimen for 24-48 hours, or as soon as you start to feel better, your normal protein allowance should be introduced over a period of a few days. Usually, this can be achieved over a period of 1-2 days, but occasionally your dietitian may suggest that you reintroduce your normal diet over a longer period of time. If in doubt, please contact your local inherited metabolic disease team for further advice.

## Note on the UK Government Sugar Tax

We have previously recommended that if adults do not tolerate the above-mentioned glucose polymers, to take alternative sources of energy from commercial drinks such as Lucozade and Ribena. However, due to the introduction of a “sugar tax” across the UK and Republic of Ireland these products have now been reformulated to contain significantly less sugar, impacting on the quantity of product to be consumed to meet the emergency guidelines.

We recommend that all individuals requiring an oral emergency regimen are prescribed a glucose polymer to prevent errors and potential catastrophic events. If you do not have one of the above on prescription please contact your metabolic dietitian.

If you cannot tolerate the above-mentioned glucose polymers, please contact your local inherited metabolic disease team for advice.

Low calorie drinks e.g. “Diet”, “Lite”, “No added sugar”, “Sugar free” drinks are not suitable. They are low in calories and artificial sweeteners, and do not provide sufficient calories for the emergency regimen

