

ADULT EMERGENCY MANAGEMENT ORAL EMERGENCY REGIMEN (ER) FOR GLYCOGEN STORAGE DISORDERS (GSD)

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

| Stage 1 – Possibly unwell | Stage 2 – Unwell but not drowsy and tolerating some oral intake | Stage 3 – Unwell and unable to take/ vomiting drinks or medicine or drowsy |
|--|---|---|
| At the first sign of feeling unwell/loss of appetite, take 200ml of glucose polymer drink. | Start full emergency regimen; 200ml of glucose polymer drink every 2 hours. | If unable to tolerate the emergency regimen or medication, or if you do not improve or become increasingly unwell contact your metabolic team or your local hospital for admission. You will require 10% glucose to be given intravenously at 2ml/kg/hr e.g. 140ml/hr for a 70kg adult. |
| If better within one hour continue with normal diet. | Continue to eat and drink as able | |
| If obviously unwell then follow stages 2 or 3 as appropriate. | Continue to give normal medicines and cornstarch. | If you go to hospital, it is a good idea to take some glucose polymer and uncooked cornstarch with you |

How to prepare the oral emergency regimen

YOUR LOCAL INHERITED METABOLIC DISEASE TEAM WILL ADVISE YOU ON THE CORRECT GLUCOSE POLYMER CONCENTRATION AND RECIPE FOR YOU:

15% concentration

20% concentration

25% concentration

PLEASE CONTACT YOUR LOCAL INHERITED METABOLIC DISEASE TEAM TO INFORM THEM THAT YOU HAVE STARTED YOUR ORAL EMERGENCY REGIMEN

You will need:

1. Water
2. Glass, beaker or shaker
3. Glucose polymer
4. Scales or relevant scoops (big blue/yellow) if using Maxijul, Vitajoule or Polycal

Choose one of the glucose polymers listed on the next page and follow the recipe applicable to you (as advised by your metabolic team)

Please also continue to take your uncooked cornstarch as tolerated and check your blood glucose more regularly (if you have a glucose meter at home)

15% CONCENTRATION

Types of Glucose Polymer available on prescription:



| 15 % Glucose Polymer Recipes | | | | |
|--|---|---|--|---|
| Glucose Polymer | Weight of glucose polymer required per dose | Recipe for 1 single dose (200ml) | Recipe for 12hr (1200ml) | Oral rehydration solution recipe |
| Maxijul Super Soluble | 30g | 1 big blue scoop (25g*) & 1 yellow scoop (5g*); make up to 200ml with water | 7 big blue scoops (175g*) & 1 yellow scoop (5g*); make up to 1200ml with water | Discuss recipe with your metabolic team if required. This is not a substitute / replacement for your glucose polymer. |
| S.O.S. 15 | Pre-measured sachet | Add 1 sachet; make up to 200ml with water | Add 6 sachets; make up to 1200ml with water | |
| Vitajoule | 30g | 1 big blue scoop (25g*) & 1 yellow scoop (5g*); make up to 200ml with water | 7 big blue scoops (175g*) & 1 yellow scoop (5g*); make up to 1200ml with water | |
| Polycal Powder | 30g | 1 big blue scoop (25g*) & 1 yellow scoop (5g*); make up to 200ml with water | 7 big blue scoops (175g*) & 1 yellow scoop (5g*); make up to 1200ml with water | |
| STORE IN FRIDGE & USE WITHIN 24 HOURS | | | | |

* Scoop weights are approximated (as per Nutricia Product Scoop Chart 2016) and rounded up or down by 0.5g to simplify recipes. Weighing scales will provide greater accuracy

How to make up 200ml of a 15% glucose polymer drink



1. Measure **ONE** level big blue scoop and **ONE** level yellow scoop glucose polymer (**30g*** total weight) or **ONE** sachet S.O.S 15 and make up to a total volume of 200ml with water
2. Shake or stir well until dissolved
3. Discuss with your metabolic team if you would like to add flavouring

20% CONCENTRATION

Types of Glucose Polymer available on prescription:



| 20 % Glucose Polymer Recipes | | | | |
|--|---|---|--|---|
| Glucose Polymer | Weight of glucose polymer required per dose | Recipe for 1 single dose (200ml) | Recipe for 12hr (1200ml) | Oral rehydration solution recipe |
| Maxijul Super Soluble | 40g | 1 big blue scoop (25g*) & 3 yellow scoops (15g*); make up to 200ml with water | 9 big blue scoops (225g*) & 3 yellow scoops (15g*); make up to 1200ml with water | Discuss recipe with your metabolic team if required. This is not a substitute / replacement for your glucose polymer. |
| S.O.S. 20 | Pre-measured sachet | Add 1 sachet; made up to 200ml with water | Add 6 sachets; make up to 1200ml with water | |
| Vitajoule | 40g | 1 big blue scoop (25g*) & 3 yellow scoops (15g*); make up to 200ml with water | 9 big blue scoops (225g*) & 3 yellow scoops (15g*); make up to 1200ml with water | |
| Polycal Powder | 40g | 1 big blue scoop (25g*) & 3 yellow scoops (15g*); make up to 200ml with water | 9 big blue scoops (225g*) & 3 yellow scoops (15g*); make up to 1200ml with water | |
| STORE IN FRIDGE & USE WITHIN 24 HOURS | | | | |

* Scoop weights are approximated (as per Nutricia Product Scoop Chart 2016) and rounded up or down to nearest 0.5g to simplify recipes. Weighing scales will provide greater accuracy

How to make up 200ml of a 20% glucose polymer drink



1. Measure **ONE** level big blue scoop and **THREE** level yellow scoops glucose polymer (**40g*** total weight) or **ONE** sachet S.O.S 20 and make up to a total volume of 200ml with water
2. Shake or stir well until dissolved
3. Discuss with your metabolic team if you would like to add flavouring

25% CONCENTRATION

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 12hr (1200ml) | |
| Maxijul Super Soluble | 50g | 2 big blue scoops (50g*); make up to 200ml with water | 12 scoops (300g*); add water to 1200ml | Discuss recipe with your metabolic team if required. This is not a substitute / replacement for your glucose polymer. |
| S.O.S. 25 | Pre-measured sachet | Add 1 sachet; make up to 200ml with water | Add 6 sachets; add water to 1200ml | |
| Vitajoule | 50g | 2 big blue scoops (50g*); make up to 200ml with water | 12 scoops (300g*); add water to 2000ml | |
| Polycal Powder | 50g | 2 big blue scoops (50g*); make up to 200ml with water | 12 scoops (300g*); add water to 2000ml | |
| STORE IN FRIDGE & USE WITHIN 24 HOURS | | | | |

* Scoop weights are approximated (as per Nutricia Product Scoop Chart 2016) and rounded up or down to nearest 0.5g to simplify recipes. Weighing scales will provide greater accuracy

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



1. Measure **TWO** level big blue scoops glucose polymer (**50g*** total weight) or **ONE** sachet S.O.S 25 and make up to a total volume of 200ml with water
2. Shake or stir well until dissolved
3. Discuss with your metabolic team if you would like to add flavouring

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 glucose polymer on prescription (and a supply at home) 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. those labelled “Diet”, “Light”, “Lite”, “No Added Sugar” or “Sugar free” are not suitable. They are lower in sugar and often contain artificial sweeteners, and do not provide sufficient calories for the emergency regimen.