



British Inherited Metabolic Disease Group

Contact Details Name:

Hospital

Telephone:

GENERAL DIETARY INFORMATION FOR EMERGENCY REGIMENS

The emergency regimen drinks are a key part of the management of many inborn errors of metabolism. It is used try to prevent or ameliorate acute decompensation. This may be precipitated by any ‘metabolic stress’ although sometimes there is no obvious reason. Many different precipitants may be responsible for ‘metabolic stress’ including infection, fasting, dietary indiscretion and in some disorders exercise or constipation. Vomiting and diarrhoea are a potent cause of illness and should always be taken seriously.

The early signs of illness may be subtle. These may include exacerbation of pre-existing neurological problems, slight unsteadiness, an appetite that is even worse than usual or ‘just not right’. The family should be taught to recognise the symptoms and signs in each child so that treatment may start at the earliest possible stage.

The drinks are usually of soluble glucose polymer (soluble glucose polymer is marketed in the UK as SOS[®], Polycose[®], Maxijul[®], Polycal[®], Vitajoule[®], etc). The exact recipe/glucose polymer concentration recommended for each child varies but recipes for the standard concentrations can be found by clicking the links below:

[Click here for a 10% Carbohydrate Emergency Regimen suitable for children under 1yr](#)

[Click here for a 15% Carbohydrate Emergency Regimen suitable for children aged 1-2yrs](#)

[Click here for a 20% Carbohydrate Emergency Regimen suitable for children aged 2-9yrs](#)

[Click here for a 25% Carbohydrate Emergency Regimen suitable for children over 10yrs](#)

In some disorders other substances may be added (for example – oral rehydration salts in gastroenteritis, medicines in urea cycle disorders and aminoacid mixtures in Maple Syrup urine disease and glutaric aciduria type 1).

[Click here for Emergency Regimen recipes for children with Maple Syrup Urine Disease](#)

[Click here for Emergency Regimen recipes for children with Glutaric Aciduria Type 1](#)

The ‘classical’ emergency regimen has three stages, although many families develop their own strategies.

Stage 1. If the child is not quite right or may be at risk of illness (for example post immunisation) give regular oral drinks and reassess in 2-4 hours. If the child is better when re-assessed then go back to normal diet. If not well, go to stage 2.

Stage 2. Regular drinks to be given day and night. The frequency and composition will depend on the disorder. The volumes and concentration will vary with the age and weight of the child. If the child uses continuous feeds the emergency regimen may be given continuously via nasogastric tube or gastrostomy. This treatment should continue until the child improves. If the child does not improve go to stage 3.

Stage 3. If the child is obviously not well, not tolerating or not taking drinks or the family are worried contact or go to hospital.

It must be emphasised that early intervention is important and may prevent complications. Inborn errors are rare; if you are not sure about anything seek help. Detailed guidelines for individual conditions can be found elsewhere on this website.

References

Dixon MA, Leonard JV. Intercurrent illness in inborn errors of intermediary metabolism. *Arch Dis Child.* 1992 Nov;**67**(11):1387-91.

Morris AA, Leonard JV. Early recognition of metabolic decompensation. *Arch Dis Child.* 1997 Jun;**76**(6):555-6.

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