



Contact Details Name:

Hospital

Telephone:

- has **CITRIN DEFICIENCY**
(also called **Citrullinaemia type 2**)
- Please read carefully. **ASSESSMENT IS URGENT**. Meticulous treatment is important as there is a high risk of serious complications.
- **Important note: The management of illness in CITRIN deficiency is quite different from other metabolic disorders. These patients have a special oral emergency regimen.**
- The major acute complications are encephalopathy and the patients are treated with a high protein, high fat, low carbohydrate diet. Patients may develop hypoglycaemia
- **Give normal saline 10 ml/kg** unless the peripheral circulation is poor or the patient is frankly shocked, and then give 20 ml/kg normal saline as a bolus immediately after the glucose. Repeat the saline bolus if the poor circulation persists as for a shocked non-metabolic patient.
- **DO NOT GIVE GLUCOSE ORALLY OR INTRAVENOUSLY (except for proven hypoglycaemia)**
- **If proven hypoglycaemia and able to take oral fluids safely give glucose or glucose polymer glucose 10% 3ml/kg (300mg/kg) or if not able to take glucose orally give a single bolus of glucose intravenously of glucose 10% 2ml /kg (200mg/kg).**
- **Continue with normal saline at 5 ml/kg/h**
- **Contact local metabolic unit for further advice about management as this is difficult.**
- *This protocol is for the immediate management only.*