• ……………………………………………………….. 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.

• Restart the usual oral diet as soon as possible. If this is not possible, initially give normal saline IV for rehydration and maintenance fluids (ie daily volume = 100ml/kg for 1st 10kg then 50 ml/kg for next 10kg then 20ml/kg thereafter).

• **DO NOT GIVE GLUCOSE ORALLY OR INTRAVENOUSLY (except for proven and symptomatic 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).

• Contact local metabolic unit for further advice about management as this is difficult.