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has CARNITINE TRANSPORTER DEFICIENCY

- Please read carefully. ASSESSMENT AND TREATMENT ARE URGENT. Treatment should be meticulous as there is a high risk of serious complications.
- The major complications are cardiomyopathy, muscle weakness and encephalopathy.
- **Start this treatment** if the patient is obviously unwell, vomiting, floppy or drowsy. Do not delay if you are uncertain.
- Give Glucose 200 mg/kg at once (2 ml/kg of 10% glucose or 1ml/kg of 20% glucose) over a few minutes.
- **Give normal saline 10 ml/kg** unless the peripheral circulation is poor or the patient is frankly shocked, 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.
- At the same time give carnitine intravenously 100 mg/kg over 30 minutes
- Continue with glucose 10% at 5 ml/kg/h until next solution, is ready glucose 10% / saline 0.45%. (For instructions to make this solution <u>click here</u>)
- If there is any doubt at all, the child must be admitted, even if only necessary for a short period of observation.
- This protocol is for the immediate management only.

More information can be found on BIMDG Emergency protocol for carnitine transporter deficiency.

http://www.bimdg.org.uk/protocols/ER-CTD-Lv1-2-128698-29-03-2008.pdf

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