

UREA CYCLE DEFECT (NEW DIAGNOSIS) Care Plan

Use in conjunction with Medical Guidelines for management of Metabolic Disorders for required investigation.

<p>Problem: _____ is at risk of neurological damage due to elevated serum ammonia and glutamine levels. Knowledge deficit due to new diagnosis.</p>	<p>S/N Sig:</p>	<p>Date: _____ Planned By:</p>	<p>Problem No: 78</p>
<p>Goal:</p> <ol style="list-style-type: none"> To prevent / reduce risk of neurological damage by reducing ammonia and glutamine levels to normal range. To educate parent(s) / child regarding diagnosis and management and provide support (where applicable). To initiate and maintain prescribed treatment. 			
<p>Nursing Care</p> <ol style="list-style-type: none"> Assess neurological status using Glasgow Coma Scale. Document and report evidence of deteriorating neurological function e.g. irritability, drowsiness, lethargy, ataxia, slurred speech, head-banging, seizures, coma etc. Assess and record vital signs (TPR, BP and Oxygen Saturation) _____ hourly. Report deviations from norm. Assist with laboratory investigations e.g. Ammonia, Amino Acids. Ammonia may be falsely elevated by Haemolysis, use of tourniquet, distress, traumatic sampling, proximity to wet nappy. Samples must be received in Laboratory within 10 minutes of sampling. Contact technician on call outside normal lab. hours and wait until arrival in hospital before proceeding with sampling. Administer ammonia reducing / alternative pathway drugs as prescribed. Adhere to manufacturer / metabolic protocols re. administration of Arginine, Sodium Benzoate and Sodium Phenylbutyrate. Monitor for side effects. Begin education programme with parents re. administration of medications. Administer Intravenous Dextrose / Intralipid as prescribed (use in conjunction with IV cannulation care plan). Liaise with Metabolic and Dietetic teams re. diet including protein and calorie intake. Diet: Refer to diet sheet for feeding regime. <ol style="list-style-type: none"> For _____ protein exchanges as _____ mls milk / formula _____ (<i>insert name and volumes prescribed</i>) (delete as appropriate) or as solids / food. For _____ <i>Dialamine</i> (Nitrogen depleted formula) For _____ calories / day. Negotiate with Mum / Dad re. feeding, feed preparation. Educate (in conjunction with Dietetic team) re. need for restricted protein. 	<p>Self / family care</p> <p>Parent(s) will give history of 'normal' or usual behaviour pattern (if applicable).</p> <p>Family will provide comfort and reassurance pre, inter and following procedures (where possible). Encourage parents to be present and assist during investigations.</p> <p>Parent(s) will assist in provision of oral hygiene.</p>	<p>Date and sign / countersign any changes</p>	

Nursing Care	Self/ family care	Date and sign / countersign any changes
<p>10. Use nasogastric / gastrostomy feeding (use in conjunction with relevant care plan) to supplement calorie and nutritional intake where unable to tolerate prescribed volumes orally. Attend to oral hygiene _____ hourly.</p> <p>11. Size N.G. Tube _____ FG.</p> <p>12. Commence teaching programme re. insertion and use of nasogastric tube (where applicable).</p> <p>13. Monitor blood glucose 4-6 hourly when on Intravenous regime.</p> <p>14. Refer to <i>Guidelines on Management of Metabolic patients</i> for instructions.</p> <p>15. Promote periods of uninterrupted rest to avoid stress and increased metabolic rate.</p> <p>16. Weigh daily / alternate days / twice weekly / weekly (delete as appropriate).</p> <p>17. Observe skin for decreased integrity due to restricted protein.</p> <p>18. Record Intake and Output. Refer to team re. replacement of lost calories (vomiting or diarrhoea)</p> <p>19. _____ is allowed a grace when vomits of _____mls (confirm same with Consultant); thereafter Replace ml for ml using _____ feeds (<i>insert name</i>). Liaise with Dietitian and Metabolic team re. instructions.</p> <p>20. Provide ongoing support and feedback to parents on procedures, investigations and results. Promote family-centred care. Allow time for questions. Liaise with Metabolic Nurse Specialist, Dietetic team and other members of the Multi-disciplinary team with regard to provision of information, support and education. Encourage parents to communicate worries and concerns</p> <p>21. Provide age appropriate explanation to _____. Involve play specialist where appropriate.</p> <p>22. Commence discharge planning (Use Patient Discharge plan and teaching plans for medication administration and NG feeding etc) to ensure support network is in place. Liaise with Dietetic team to ensure well and unwell dietary regimes are provided pre-discharge (1-2 days notification necessary).</p>	<p>Parent(s) will inform staff of skin breakdown noted during bathing, dressing or toileting.</p>	