

Maple Syrup Urine Disease Care Plan

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

Problem: _____ is at risk of neurological damage due to elevated branch chain amino acids (BCAA) levels caused by _____.	S/N Sig:	Date: _____ Planned By:	Problem No: 36
Goal: 1) Prevent neurological damage by reducing leucine to acceptable levels. 2) Assist Metabolic Team in ascertaining cause of elevated BCAAs			
Nursing Care <ol style="list-style-type: none"> Assess neurological status using Glasgow Coma Scale. Document evidence of altered neurological status i.e. disorientation, ataxia, slurred speech, abnormal limb movements, irritable cry etc. Obtain history of normal behaviour pattern from parents. Record vital signs (TPR, BP & Oxygen saturation) at 4 hourly intervals or more frequently if indicated. Assist in collection of specimens e.g. U&E / Serum amino acids etc. Administer Intravenous Dextrose and Intralipid solutions as prescribed to suppress catabolism and endogenous protein breakdown. (Use Intravenous Cannulation Care Plan). Monitor and record blood glucose levels 4-6 hourly when on intravenous therapy. Perform & record ward urinalysis on each void. Inform team if sample is positive for ketones and / or glucose. Liaise with Metabolic team and dietetic team regarding dietary regime (Protein exchanges, synthetic protein and calories). Collect blood for branch chain amino acid levels by finger prick _____ hourly (dictated by consultant). Micro tubes are available from St. Brigid's Ward and Laboratory stores. Pass nasogastric tube if patient is unable to meet required calories using oral and / or intravenous fluids (use NG feeding care plan). Negotiate with parents re. participation in insertion of nasogastric tube and feeding. Assess technique where this procedure has been taught previously. Record Intake and Output. Inform team of vomiting / diarrhoea. Estimate volumes of vomitus. Lost volumes will need to be replaced using oral / NG feeds / oral / NG feeds and intravenous dextrose / intravenous dextrose (delete as appropriate) to prevent loss of calories. If _____ vomits, s/he is allowed a grace of _____mls (confirm same with Consultant). Thereafter, replace ml for ml using _____ feeds (insert name) 	Self/ family care <p>Parent(s) will assist in assessment of neurological status by providing information on normal behaviour pattern.</p> <p>Family will provide comfort and reassurance pre, during and following procedures.</p> <p>Parents will inform staff if _____ appears to be in pain (may signify complications of Intravenous therapy).</p> <p>Parents will negotiate with staff regarding level of participation in feeding / preparation of feeds etc.</p>	Date and sign / countersign any changes	

Nursing Care	Self/ family care	Date and sign / countersign any changes
<p>11. Monitor closely for signs of fluid overload.</p> <p>12. Administer Isoleucine and Valine supplements as prescribed (may be delivered intravenously if unable to tolerate orally).</p> <p>13. Document leucine levels on appropriate flow sheet indicating where possible calorie / protein exchange intake at time of sampling Flow sheets available from St. Brigid's Ward.</p> <p>14. Weigh daily / alternate days / twice weekly / weekly (<i>delete as appropriate</i>) according to ward policy or clinical status.</p> <p>15. Assess skin integrity as may be impaired by protein restriction. Provide oral hygiene.</p> <p>16. Provide periods of rest as stress can increase metabolic rate.</p> <p>17. Keep _____ and family informed of all procedures / investigations / results. Encourage and promote family-centred care.</p>	<p>Parents will keep nappies for weighing by staff and will inform staff of episodes of vomiting / diarrhoea / voids.</p> <p>Parents will inform staff of skin breakdown noted during bathing, dressing or toileting and will assist if wished in provision of oral hygiene</p>	