

**Altered Nutritional Requirements: Glutaric Aciduria Type 1 (GA1) Care Plan**  
Use in conjunction with Medical Guidelines for management of Metabolic Disorders for required investigation.

<b>PROBLEM:</b> _____ is at risk of neurological damage secondary to catabolism due to _____	<b>S/N Signature:</b> _____	<b>Date:</b> _____ <b>Planned by:</b> _____	<b>Problem no:</b> <b>20</b>
<b>GOAL: 1)</b> To prevent neurological deterioration and / damage secondary to catabolism and accumulation of toxic metabolites. <b>2)</b> To assess parental and patient knowledge base re. condition, implications etc.			
<b>Nursing Care</b>	<b>Self / Family Care</b>	<b>Date and Sign Any Changes</b>	
<ol style="list-style-type: none"> <li>Assess vital signs _____ hourly. Record on Paediatric Observation Chart (PEWS score). Note: Any deviation from the normal should be promptly reported to the metabolic team. Septic work up (when pyrexial) should include serum amino acids and urinary organic acids. Refer to Medical Guidelines for Management of Metabolic Disorders for required investigations.</li> <li>Assess neurological status (record baseline GCS). Report and document signs of altered status e.g. irritable cry, abnormal movements (i.e. posturing, scissoring of legs, fisting, abnormal eye movements). Continue GCS monitoring _____ Hourly if altered status noted (frequency is dictated as per metabolic consultant).</li> <li>Commence on <b>'Unwell Regime' (as per Consultant's instructions)</b>.           <ul style="list-style-type: none"> <li>Ensure adequate calorie intake to prevent catabolism. Calories to be administered _____ (insert number). Adjustments may be required due to changes in clinical status.</li> <li>Nasogastric feeding and / or intravenous therapy will be necessary if unable to supply necessary volume and calorie intake orally.</li> <li>Liaise with ward dietitian (bleep 834). Refer to Dietetic flow sheet / instructions regarding volumes of Synthetic Formula / protein exchanges / calories.</li> <li>Record Exchanges/Calorie content of IV and oral intake on Calorie Intake Flow Sheet.</li> </ul> </li> <li>Check urine daily for pH and ketones (preferably early morning sample). <b>Report &amp; record presence of ketones as this is a sign of catabolism.</b> Note: pH will be increased if there is renal tubular leak of phosphate &amp; potassium.</li> <li>Record intake and output. Estimate volumes of vomitus. Lost volumes will need to be replaced using oral / NG feeds <b>and</b> intravenous dextrose (delete as appropriate) to prevent loss of calories. If _____ vomits, he/she is allowed a grace of _____mls (confirm same with Consultant). Replace vomitus ml for ml using _____ feeds (insert name). All feeds and volumes must be checked by 2 staff members (refer to ward policy re. same). Use nasogastric feeding and Intravenous cannula care plan were appropriate.</li> <li>Assist in collection of specimens as requested by Metabolic Team e.g. urine for glutarate, serum amino acids.</li> </ol>	<p>Parent(s) will report unusual or abnormal behaviour &amp; movements to staff.</p> <p>Family will decide level of involvement in changing of nasogastric tube, preparation and administration of feeds while in hospital and when at home.</p> <p>Parent(s) will keep nappies for output estimation and will assist staff in estimation of loss through vomiting.</p> <p>Parent(s) will assist with collection of urine specimens to develop</p>		

<p>Glutarate samples must be frozen if not sent directly to laboratory. Plasma Amino Acids are performed daily (for same day results samples must be received in lab by 9am).</p> <p>Assess parents' correct technique for urine sample collection. Urine samples are frequently requested on an out patient basis.</p> <ol style="list-style-type: none"> <li>7. Observe skin integrity as protein restricted diet may result in desquamation of skin / skin breakdown. Inform metabolic team as protein intake may need to be increased.</li> <li>8. Administer medications as prescribed. Monitor for side effects. (Note If carnitine dose is increased this may cause loose stools – medical team must be notified and IV administration may be required.)</li> <li>9. Liaise with Metabolic Multidisciplinary team (Psychology, Social Work etc.).</li> <li>10. Complete and document follow up care etc. in Patient Discharge Plan.</li> </ol>	<p>competence for home sampling.</p> <p>Parent(s) will inform staff of skin breakdown noted during bathing, dressing or toileting</p>	
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