

Glutaric Aciduria Type 1 (GA1) (New Diagnosis) Care Plan

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

Problem: Knowledge deficit due to new GA1 diagnosis	S/N Sig:	Date: _____ Planned by:	Problem No: 21
Goal: 1. To establish feeding and medication regime to prevent catabolism and promote growth and development. 2. To educate and support parents.			
Nursing Care	Self/ family care	Date and sign / countersign any changes	
<ol style="list-style-type: none"> Assess vital signs (T.P.R. / Blood Pressure / Oxygen saturation _____ hourly (<i>delete as appropriate</i>)). 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 glutarate level. Refer to Medical Guidelines for management of Metabolic Disorders for necessary laboratory investigations. Assess neurological status. Report and document signs of altered status e.g. irritable cry, abnormal movements (i.e. posturing, scissoring of legs, fisting, abnormal eye movements). Ensure adequate calorie intake to prevent catabolism. Refer to dietetic instruction sheet daily re: prescribed volume of feeds / calorie intake. Adjust protein and calorie intake as required due to changes in clinical status and as per medical guidance. Check urine daily for pH + ketones (preferably early morning sample). Report presence of ketones as this is a sign of catabolism and necessitates urgent management. Note: p H will be increased if there is energy deficiency of the organs esp. kidneys which can lead to a leak of phosphate & potassium). Record intake and output. Estimate volumes of vomitus. Replace volumes vomited using feeds or Intravenous dextrose or combination of Intravenous dextrose and feeds (delete as appropriate) to prevent loss of calories. Allow grace of _____mls (confirm same with Consultant). Thereafter replace ml for ml using _____ feeds (<i>insert name</i>) or iv fluids.. Liaise with Dietitian and Metabolic team regarding instructions. Ensure all feeds and volumes are checked by 2 staff members (one of whom is a RCN -refer to ward policy regarding same). Nasogastric feeding (use NG feeding care plan) and / or Intravenous therapy (use IV cannulation care plan) may be necessary to ensure delivery of recommended calories and fluids. 	<p>Parent(s) will report unusual or abnormal behaviour & movements to staff.</p> <p>Parent(s) will keep nappies for output estimation and will assist staff in estimation of loss through vomiting.</p>		

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<p>8. Assist in collection of specimens e.g. urine for glutarate, serum amino acids. Glutarate samples must be received in Laboratory by 9amThursday mornings. 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 laboratory by 9am). Liaise with team re skin biopsy to confirm diagnosis and screening of siblings. Date of Skin Biopsy Screening of Siblings Date.....</p> <p>9. Observe skin integrity as protein restricted diet may result in desquamation of skin / skin breakdown. Inform metabolic team as it may be necessary to increase protein intake. Apply barrier creams to affected areas.</p> <p>10. Administer medications as prescribed. Commence teaching re. medications, effects, side effects (use Drug Administration Parent Teaching Plan).</p> <p>11. Arrange cranial ultrasound and MRI scan on initial diagnosis and as indicated by metabolic consultant thereafter.</p> <p>12. Commence parental teaching programme regarding: diagnosis, genetic implications, diet, diet recipes, adjustment of dietary regimes, care of nasogastric tube (use Nasogastric Feeding Plan), central line (delete as appropriate). Liaise with Metabolic Nurse Specialist.</p> <p>13. Liaise with Multidisciplinary team (Psychology, Social Work etc.).</p> <p>14. Complete and document follow up care etc. in Patient Discharge Plan.</p>	<p>Parent(s) will assist with collection of urine specimens to develop competence for home sampling.</p> <p>Parent(s) will inform staff of skin breakdown noted during bathing, dressing or toileting.</p> <p>Parent(s) will assist with administration of medications to develop competence for home administration</p>	