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Altered Nutritional Requirements: Methylmalonic aciduria (MMA) / Propionic aciduria (PA) Care Plan Use in conjunction with Medical Guidelines for management of Metabolic Disorders for required investigation.

	S/N Sig:	Date:	Problem no:
Problem: is at risk of neurological damage secondary to catabolism due to		Planned by:	113
Goal: a) To prevent neurological deterioration and / damage secondary to catabolism and accumulation of toxic metabol 2) To assess parental knowledge base re. condition, implications etc.	ites.	1	
Nursing care:	Self / fa	mily care	Date and sign / countersign any changes
1. Assess vital signs (T.P.R. / Blood Pressure / Oxygen Saturation (delete as appropriate))hourly.			
Note: Any deviation from the normal should be promptly reported to the metabolic team. Septic work up (when pyrexial) should	ıld include		
serum amino acids and urinary organic acids. Refer to Medical Guidelines for Management of Metabolic Disorders for	r required		
investigations.			
2. Assess neurological status (record baseline GCS). Report and document signs of altered status e.g. irritable cry, abnormal movements		s) will report unusual or	
(i.e. posturing, scissoring of legs, fisting, abnormal eye movements). If blood Ammonia level is elevated refer to Hyperammona	iamia cara I	al behaviour & ents to staff.	
plan No. 29.			
3. Commence on 'Unwell Regime' (as per Consultant's instructions). Ensure adequate calorie intake to prevent catabolism. Calorie intake to prevent catabolism.	ories to be		
administered (insert number). Adjustments may be required due to changes in clinical status. Nasogastric feeding	g and / or		
intravenous therapy will be necessary if unable to supply necessary volume and calorie intake orally.			
Liaise with ward dietitian (bleep 834). Refer to Dietetic flow sheet / instructions regarding volumes of Synthetic Formula / protein ex	changes /		
calories.			
4. Check urine daily for pH and ketones (preferably early morning sample). Report & record presence of ketones as this is	a sign of		
catabolism.			
5. Record intake and output. Estimate volumes of vomitus. Lost volumes will need to be replaced using oral / NG feeds and in		s) will keep nappies for	
dextrose (delete as appropriate) to prevent loss of calories. If vomits, he/she is allowed a grace ofmls (cor		estimation and will assist estimation of loss through	
with Consultant), there after replace ml for ml using feeds (insert name). All feeds and volumes must be checked			
members (refer to ward policy re. same). Use nasogastric feeding and Intravenous cannula care plan were appropriate.			

6. Assist in collection of specimens as requested by Metabolic Team e.g. urine for MMA / PA (delete where appropriate), serum amino acids, ammonia etc. (refer to medical guidelines for sample queries) MMA / PA samples must be received in Lab by 9am on Thursday 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 lab by 9am). 7. 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. 8. Administer L-Carnitine and Metronidazole as prescribed by metabolic consultant. Vitamin B12 is administered to some patients with MMA – frequency of administration as per metabolic consultant's instructions.	Parent(s) will assist with collection of urine specimens Parent(s) will inform staff of skin breakdown noted during bathing, dressing or toileting. Parent(s) will assist with administration of medications	