Temple Street
CHILDREN'S UNIVERSITY HOSPITAL

Addressograph

Homocystinuria (HCU) New Diagnosis Care Plan

PROBLEM: Parental Knowledge deficit due to new diagnosis.	S/N Sig:	Date:_____ Planned by:_____	Problem no: 45

GOAL: 1) To establish recommended dietary regime for management of Homocystinuria (due to non-responsive cystathionine beta synthase deficiency)
2) To educate parent (s) on HCU, management, complications of non-treatment / non-compliance with recommended management

Nursing Care	Self/ Family Care	Date & Sign / Countersign Any Changes
Initial Assessment: A. Assess vital signs _____hourly. Record on Paediatric Observation Chart (PEWS score). B. _____ will continue on existing feeding regime until baseline blood specimens have been obtained and pyridoxine responsiveness has been established. C. Assist in obtaining baseline bloods for: Quantitative Amino Acids, Total and Free Homocysteine, DNA mutation, B_{12} & Folate, CBS Enzyme (EDTA). **D. Liaise with Metabolic Consultant regarding trial of Pyridoxine (to confirm / out rule Pyridoxine responsiveness).** **Following diagnosis of non- responsive HCU:** 1. Establish dietary and medication management as prescribed by Metabolic Consultant. 2. Take blood for Total & Free Homocysteine sample (daily / alternate days / or other _____ (insert frequency) as instructed by Metabolic Consultant. ***Ensure rapid delivery of amino acids specimens to Metabolic Laboratory (require deproteinising within 10 mins of collection).*** 3. Liaise with Metabolic Team and Dietetic Team re. commencement of special diet. 4. Administer Vit B_{12} and Folate and B6 as prescribed. 5. Commence parental education programme re. condition, diet, implications of non-compliance and non-treatment in liaison with Metabolic Nurse Specialist. Give HCU Handbook to parents to provide reference material and learning resource. Encourage to read and ask questions. 6. Assess parent's level of understanding of the condition/management throughout _____'s admission and prior to discharge. 7. Monitor fluid and calorie intake. 8. Weigh _____ (insert frequency) 9. Involve Psychology and Social Work teams in ongoing care and establishing support networks for discharge. 10. Liaise with Metabolic team re. scheduling of consults to other disciplines and screening of siblings. Cardiology date _____, Ophthalmology date _____, Screening of Sibling(s) date_____. 11. Liaise with PHN and provide him/her with information re HCU. 12. Establish, if possible, the facility to take bloods at local hospital. Reference: Management Protocol for Patients diagnosed with homocystinuria (HCU) due to Cystathionine β-syntase deficiency.		