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| **Nursing Care Plan for Children with Maple Syrup Urine Disease** **Addressograph****Use in conjunction with Medical Guidelines for Management of Metabolic Disorders for required investigation and the Nursing Guidelines for Children with Maple Syrup Urine Disease (PP-CLIN-NUR-104)**  |
| **Problem: a**) \_\_\_\_\_\_\_\_\_\_\_ is at risk of neurological damage due to elevated branch chain amino acids (BCAA) levels caused by \_\_\_\_\_\_\_\_\_\_\_. | **S/N Sig:\_\_\_\_\_\_\_\_\_\_\_****NMBI\_\_\_\_\_\_\_\_\_\_\_\_\_** | **Date:\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_****Planned By:\_\_\_\_\_\_\_\_\_\_\_\_****Grade:\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_** | **Problem no:****36** |
| **Goal:** a) Prevent neurological damage by reducing leucine to acceptable levels.  b) Assist Metabolic Team in ascertaining cause of elevated BCAAs.  |
| **Nursing care:** | **Self / family care** | **Date/Signature/Grade/NMBI no. for any changes made to care** |
| 1. **Patient Assessment**
2. Assess neurological status using Glasgow Coma Scale at baseline and record \_\_\_\_\_\_\_\_\_ hourly. Document evidence of altered neurological status i.e. disorientation, ataxia, slurred speech, abnormal limb movements, irritable cry etc. (Bouchereau et al. 2017). Obtain history of normal behaviour pattern from parent(s).
3. Record vital signs and document on PEWS chart at \_\_\_\_\_\_ hourly intervals or more frequently as condition indicates.
4. **EMERGENCY MANAGEMENT**
5. Assist in collection of specimens e.g. BCAA, U&E / Serum amino acids, etc.
6. Administer Intravenous NaCl / Glucose and Smoflipid solutions as prescribed to suppress catabolism and endogenous protein breakdown (Frazier et al, 2014). Assess PIVC site regularly and document VIP score in PEWS chart. Use in conjunction with Intravenous Cannulation Care Plan (RF-NUR-019).
7. Monitor and record blood glucose levels 4-6 hourly when on intravenous therapy and report any abnormalities to the medical team.
8. Perform & record ward urinalysis on admission and on each subsequent void. Inform team if sample is positive for ketones and / or glucose as this requires immediate intervention.
9. **BRANCH CHAIN AMINO ACIDS**
10. Measure frequently during periods of acute illness (usually 4-8 hourly) to evaluate effectiveness of prescribed dietary and intravenous regime and monitor metabolic status.
11. Collect blood for branch chain amino acid levels by finger prick \_\_\_\_\_\_\_\_\_\_ hourly (dictated by Consultant). Microvette tubes are available from St. Brigid’s Ward and Laboratory stores.
12. Document BCAA levels on appropriate flow sheet indicating where possible calorie / protein exchange intake at time of sampling.
 | Parent (s) will assist in assessment of neurological status by providing information on normal behaviour pattern.Family will provide comfort and reassurance pre, during and following procedures.Parent(s) will inform staff if \_\_\_\_\_\_\_\_\_ appears to be in pain (may signify complications of Intravenous therapy). |  |
| **Nursing Care Plan for Children with Maple Syrup Urine Disease continued** |
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| 1. **Dietary Interventions**
2. Liaise with Metabolic team and dietetic team regarding dietary regime (Protein exchanges, synthetic protein and calories). Refer to patient specific Unwell Plan / Daily Dietary Flow Sheet
3. Insert nasogastric tube if patient is unable to meet required calories using oral (use NG feeding care plan RF-NUR-024). Negotiate with parents regarding participation in insertion of nasogastric tube and feeding. Assess parent(s) technique where this procedure has been taught previously.
4. **INTAKE & OUTPUT**
5. Record Intake and Output to include strict monitoring of positive / negative fluid balances. Inform team of vomiting / diarrhoea. Estimate volumes of vomitus.
6. Replace gastric or intestinal losses using oral / NG feeds and intravenous glucose / Smoflipid (delete as appropriate) to prevent loss of calories. If \_\_\_\_\_\_\_\_\_\_ vomits, allow grace of \_\_\_\_\_\_\_mls (confirm same with Consultant). Thereafter, replace ml for ml using \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ feeds (insert name).
7. Record daily calorie intake/IV administration on Oral / Nasogastric Calorie Intake Flow Sheet.
8. Monitor closely for signs of fluid overload. Consider cumulative fluid balances after each 24 hour recorded balance
9. Administer Isoleucine and Valine supplements as prescribed (Simon et al. 2006). These may be delivered intravenously if unable to tolerate orally.
10. Weigh daily / alternate days / twice weekly / weekly (delete as appropriate) according to ward policy or clinical status and record in healthcare record.

 1. **GENERAL OBSERVATIONS / INTERVENTIONS:**
2. Assess skin integrity as may be impaired by protein restriction. The nappy area can be especially red and if so needs to be reviewed by metabolic team and consultant informed as this may influence the decision to increase protein intake.
3. Provide oral hygiene, especially if vomiting or if feeding is being provided via Nasogastric tube.
4. Provide periods of rest as stress can increase metabolic rate.
5. **COMMUNICATION:**
6. Keep \_\_\_\_\_\_\_\_\_\_\_\_and family informed of all procedures / investigations / results.

  | Parent(s) will negotiate with staff regarding level of participation in feeding / preparation of feeds etc.Parent(s) will retain nappies for weighing by staff and will inform staff of episodes of vomiting / diarrhoea / voidsskin breakdown noted during bathing, dressing or toileting and will assist if wished in provision of oral hygiene |  |

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| **REFERENCES:**Bouchereau J., Leduc-Leballeur J., Pichard S., Imbard A., Benoist J.F., Warde M.T.A., Arnoux J.B., Barbier V., Brassier A., Broue P., Cano A., Chabrol B., Damon G., Gay C., Guillain I., Habarou F., Lamireau D., Ottolenghi C., Paermentier L., Sabourdy F., Touati G., Ogier de Baulny H., deLonlay P. & Schiff M. (2017) Neurocognitive profiles in MSUD school-age patients. *Journal of Inherited Metabolic Disease.* **40**, 377-383.Frazier D.M., Allgeier C., Homer C., Marriage B.J., Ogata B., Rohr F., Splett P.L. Stembridge A. & Singh R.H. (2014) Nutrition management guideline for maple syrup urine disease: An evidence- and consensus-based approach. *Mollecular Genetics and Metabolism.* **112**. 210-217. Simon E., Fingerhut R., Baumkotter J., Konstantopoulou V., Ratschmann R. & Wendel U. (2006) Maple Syrup urine disease: Favourable effect of early diagnosis by newborn screening on the neonatal course of the disease. *Journal of Inherited Metabolic Disease.* **29**, 532-537.   |  |  |