

- 3) **Do not administer IV lipids. Intralipid is contraindicated.**
- 4) Labs should be sent for glucose, electrolytes, blood ammonia, CBC, BUN, creatinine, uric acid, LFTs, CPK, blood gas for pH, and urine for myoglobin. Heparinized plasma for carnitine (1 ml) and urine organic acids (15 - 20cc) should be collected, frozen, and sent STAT on dry ice to the Southern California Kaiser Permanente Regional Reference Laboratories, 11668 Sherman Way, North Hollywood, CA 91605. (818) 502-5959 (within Kaiser 8-336-5959 or 8-336-4720).
- 5) A chest x-ray and Doppler echocardiography should be considered since cardiomyopathy is associated with this condition.
- 6) Treatment for any triggering infection should be started as appropriate.
- 7) Because of potentially severe metabolic stress caused by fever, temperatures in excess of 37.8° C should be treated aggressively with acetaminophen, and if necessary, ibuprofen. Use a cooling blanket if necessary.
- 8) Carnitine should be supplemented at her usual dose orally if tolerated. Do not give excess carnitine without consulting with the Metabolic/Genetics physician.
- 9) When enteral feeds are started they should be ordered every 3 - 4 hours. Her diet limits the use of long chain fats to 15 - 20% of total calories and recommends that MCT comprise 15 - 20 % of the calories. Daily calorie counts should be conducted to closely monitor her intake. Contact the Regional Metabolic Nutritionists, Joyce Wong, MS, RD, at (510) 596-7474 (within Kaiser 8-490-7474) or Amy Tung, MS, RD (510) 596-6918 for additional assistance with dietary management.
- 10) _____ should be continued on hydration therapy (1 ½ times maintenance) until her CPK level begins to decrease significantly, creatinine normalizes, and gross hematuria has resolved. She should also be clinically improved without significant myalgia. Please consult with genetics or nephrology service prior to hospital discharge.
- 11) Notify Dr. Joyce Kobori at (408) 972-3276 (within Kaiser 8-440-3276), or the Geneticist On Call through the Kaiser Santa Teresa page operator at (408) 972-7188 (within Kaiser 8-440-7188 on admission and prior to discharge).

CURRENT MEDICATIONS:

- phenobarbital (20mg/5 ml): 3 ml PO BID
- carnitine (100mg/ml): 1.75 ml PO BID

November 12, 1999

Page 2 of 2

Lisa Jaquez

Lisa.jaquez@gartner.com

Revised 05/29/00

M:\Metabolic ER Protocols\Metabolic ER Protocols\