

CPT I Deficiency Care Plan (last updated 2/21/09)

<p>Clinical Considerations</p> <ul style="list-style-type: none"> Maternal complications (acute fatty liver and HELLP) Hepatic dysfunction with elevated ammonia and enzymes is common. Creatine kinase elevation may be seen in acute episodes, Renal tubular acidosis Transient hypertriglyceridemia 	<p>Initial labs (diagnostic & baseline)</p> <ul style="list-style-type: none"> ACP Carnitine panel Comp metabolic panel Targeted mutational analysis (Inuit/Hutterite, otherwise private mutations, available at Oregon) OR Enzymatic asst on fibroblasts 								
<p>Diet considerations/ treatment</p> <ul style="list-style-type: none"> Avoid fasting Consider MCT oil supplementation 	<p>Monitoring</p> <ul style="list-style-type: none"> Comp metabolic panel at clinic visits 								
<p>Frequency of visits (non-diet)</p> <table border="1" style="width: 100%; border-collapse: collapse; margin-top: 5px;"> <thead> <tr> <th style="text-align: left; padding: 2px;">Age</th> <th style="text-align: left; padding: 2px;">Frequency of clinic visits</th> </tr> </thead> <tbody> <tr> <td style="padding: 2px;">0-4 years</td> <td style="padding: 2px;">Every 6 months</td> </tr> <tr> <td style="padding: 2px;">4-18 years</td> <td style="padding: 2px;">Once a year</td> </tr> <tr> <td style="padding: 2px;">>18 years</td> <td style="padding: 2px;">Every 3 years</td> </tr> </tbody> </table>	Age	Frequency of clinic visits	0-4 years	Every 6 months	4-18 years	Once a year	>18 years	Every 3 years	<p>Clinic visit labs</p> <ul style="list-style-type: none"> Comp metabolic panel
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0-4 years	Every 6 months								
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<p>Emergency management</p> <ul style="list-style-type: none"> Immediate IV 10% dextrose Provide emergency letter 	<p>Labs to obtain during illness</p> <ul style="list-style-type: none"> Comp metabolic panel Ammonia Serum phosphate Serum CK 								
<p>Other evaluations</p> <ul style="list-style-type: none"> Yearly developmental questionnaires (to be completed by parents) Developmental eval @ 3 & 6y Neuropsych @ 9y 	<p>Yearly labs</p> <ul style="list-style-type: none"> Comprehensive metabolic panel Triglycerides UA Phosphate (blood and urine) Urine creatinine 								

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Performance Measures	Outcome measures
<ol style="list-style-type: none"> 1. Age of diagnosis and diet initiation 2. Presence of illness at time of diagnosis including neonatal lethargy, acute liver failure, seizure or coma. 3. Presence of maternal illness (acute fatty liver, HELLP) 4. Initial lab studies <ol style="list-style-type: none"> a. Acylcarnitine profile b. Other abnormal laboratory findings (if obtained) including CMP, ammonia, and CK c. Genotype d. Enzymatic studies 5. Frequency of clinic contacts and visits (track compliance with visits) 6. Growth parameters (ht, wt, OFC, BMI) 7. Total decompensations and hospitalizations (including infections) <ol style="list-style-type: none"> a. # of days for hospitalizations b. # of ER visits c. Track labs including CMP, ammonia, serum phosphate, and CK d. Track rhabdomyolysis 8. Monitoring Laboratory studies <ol style="list-style-type: none"> a. Comp metabolic panel b. Triglycerides c. UA d. Phosphate (blood/urine) e. Urine creatinine 9. Neuropsychological evaluation results 10. Diet (Y/N) <ol style="list-style-type: none"> a. Frequency of Dietitian visits b. Frequency of dietary analysis (3 day diet records) c. Percent of MCT and LCFA in diet. <ol style="list-style-type: none"> i. Including if essential fatty acids are added d. Formula (Y/N) e. Mode (oral, G-tube, bolus/drip, meds only/meds and diet) 11. Developmental services (PT, OT, & speech) 12. School Performance <ol style="list-style-type: none"> a. Grade appropriate (Y/N) b. IEP (Y/N) c. Special services (Y/N) 13. Genetic Counseling (Y/N) 	<ol style="list-style-type: none"> 1. Mortality 2. History and/or presence of renal dysfunction 3. History and/or presence of liver dysfunction 4. History and/or presence of hypertriglyceridemia 5. Development <ol style="list-style-type: none"> a. IQ b. Level of functioning 6. Growth <ol style="list-style-type: none"> a. Final adult parameters