

## Hyperphenylalaninemia Care Plan (last edited 2/21/09)

<p><b>Clinical Considerations</b></p> <ul style="list-style-type: none"> <li>• Diet</li> <li>• Females-Mat PKU</li> </ul>	<p><b>Initial labs (diagnostic &amp; baseline)</b></p> <ul style="list-style-type: none"> <li>• Quantitative Phe and Tyr</li> <li>• Urinary pteridine profile</li> <li>• Blood dihydropteridine reductase enzyme assay</li> </ul>								
<p><b>Diet Considerations</b></p> <ul style="list-style-type: none"> <li>• None as long as Phe remains &lt;6 mg/dL</li> <li>• Consider diet/therapy if Phe is 6-10 mg/dl</li> <li>• Diet initiation for Phe&gt; 10 mg/dl (see PKU care plan)</li> <li>• Avoid artificial sweeteners</li> </ul>	<p><b>Monitoring labs</b></p> <ul style="list-style-type: none"> <li>• Quant Phe</li> <li>• Recommended Treatment ranges               <ul style="list-style-type: none"> <li>○ &lt; 10 mg/dL</li> <li>○ Pregnant- 2-6 mg/dL</li> </ul> </li> </ul> <table border="1" style="margin-left: 20px; border-collapse: collapse; width: 80%;"> <thead> <tr> <th style="text-align: center; padding: 2px;">Age</th> <th style="text-align: center; padding: 2px;">Frequency</th> </tr> </thead> <tbody> <tr> <td style="padding: 2px;">0-12 m</td> <td style="padding: 2px;">Every 3 months if Phe &lt;5 mg/dl; monthly if Phe&gt; 5 mg/dL</td> </tr> <tr> <td style="padding: 2px;">12-24 m</td> <td style="padding: 2px;">Every 6 months</td> </tr> <tr> <td style="padding: 2px;">&gt; 24 mo</td> <td style="padding: 2px;">Yearly</td> </tr> </tbody> </table> <ul style="list-style-type: none"> <li>• If diet initiated at any point, switch to PKU care plan</li> </ul>	Age	Frequency	0-12 m	Every 3 months if Phe <5 mg/dl; monthly if Phe> 5 mg/dL	12-24 m	Every 6 months	> 24 mo	Yearly
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<p><b>Frequency of metabolic visits</b></p> <table border="1" style="margin-left: 20px; border-collapse: collapse; width: 80%;"> <thead> <tr> <th style="text-align: center; padding: 2px;">Age</th> <th style="text-align: center; padding: 2px;">clinic visits</th> </tr> </thead> <tbody> <tr> <td style="padding: 2px;">0-12 months</td> <td style="padding: 2px;">Yearly</td> </tr> <tr> <td style="padding: 2px;">&gt;12 months</td> <td style="padding: 2px;">Every 3 years</td> </tr> </tbody> </table> <ul style="list-style-type: none"> <li>• For males with Phe levels &lt;4 mg/dL at 2 years of age=discharged</li> </ul>	Age	clinic visits	0-12 months	Yearly	>12 months	Every 3 years	<p><b>Clinic Visit labs</b></p> <ul style="list-style-type: none"> <li>• Phe monitoring</li> </ul>		
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<p><b>Other evaluations</b></p> <ul style="list-style-type: none"> <li>• None</li> </ul>	<p><b>Yearly labs</b></p> <ul style="list-style-type: none"> <li>• Phe monitoring</li> </ul>								

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<b>Performance Indicators</b>	<b>Outcome Indicators</b>
<ol style="list-style-type: none"><li>1. Age of diagnosis</li><li>2. Initial lab studies<ol style="list-style-type: none"><li>a. Phe and Tyr levels</li><li>b. Urinary pteridine profile</li><li>c. Blood dihydropteridine reductase enzyme assay</li><li>d. Molecular results</li></ol></li><li>3. Monitoring lab studies<ol style="list-style-type: none"><li>a. Phe and Tyr levels<ol style="list-style-type: none"><li>i. Levels</li><li>ii. Frequency</li></ol></li></ol></li><li>4. Growth parameters (ht, wt, OFC, BMI)</li><li>5. Frequency of clinic contacts and visits (track compliance with visits)</li><li>6. Developmental services (PT, OT, &amp; speech)</li><li>7. School performance<ol style="list-style-type: none"><li>a. Grade appropriate (Y/N)</li><li>b. Special services (Y/N)</li><li>c. IEP (Y/N)</li></ol></li><li>8. Genetic Counseling (Y/N)</li></ol>	<ol style="list-style-type: none"><li>1. Development (parent questionnaires)</li><li>2. Growth</li><li>3. History and or presence of ADHD or psychiatric issues</li><li>4. Growth<ol style="list-style-type: none"><li>a. Final adult parameters</li></ol></li></ol>