

Tyrosinemia Type II and III Care Plan (last updated 2/21/09)

<p>Clinical Considerations</p> <ul style="list-style-type: none"> • Ocular and skin findings 	<p>Initial labs (diagnostic & baseline)</p> <ul style="list-style-type: none"> • SAA and UOA • Rule out liver dysfunction • Molecular currently not available in US (performed in Germany) 										
<p>Diet Considerations</p> <ul style="list-style-type: none"> • Phe/Tyr restricted diet 	<p>Monitoring labs</p> <ul style="list-style-type: none"> • Quant Phe/Tyr or plasma SAA <table border="1" style="margin-left: 20px; border-collapse: collapse; text-align: center;"> <thead> <tr> <th style="padding: 2px 5px;"><u>Age</u></th> <th style="padding: 2px 5px;"><u>Frequency</u></th> </tr> </thead> <tbody> <tr> <td style="padding: 2px 5px;">0-6 months</td> <td style="padding: 2px 5px;">Every week</td> </tr> <tr> <td style="padding: 2px 5px;">6-12 months</td> <td style="padding: 2px 5px;">Every 2 weeks</td> </tr> <tr> <td style="padding: 2px 5px;">1-3 years</td> <td style="padding: 2px 5px;">Monthly</td> </tr> <tr> <td style="padding: 2px 5px;">>3 years</td> <td style="padding: 2px 5px;">Every 3 months</td> </tr> </tbody> </table>	<u>Age</u>	<u>Frequency</u>	0-6 months	Every week	6-12 months	Every 2 weeks	1-3 years	Monthly	>3 years	Every 3 months
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<p>Frequency of metabolic visits</p> <table border="1" style="margin-left: 20px; border-collapse: collapse; text-align: center;"> <thead> <tr> <th style="padding: 2px 5px;"><u>Age</u></th> <th style="padding: 2px 5px;"><u>clinic visits</u></th> </tr> </thead> <tbody> <tr> <td style="padding: 2px 5px;">0-6 months</td> <td style="padding: 2px 5px;">Every 2 months</td> </tr> <tr> <td style="padding: 2px 5px;">6-12 months</td> <td style="padding: 2px 5px;">Every 3 months</td> </tr> <tr> <td style="padding: 2px 5px;">1-18 years</td> <td style="padding: 2px 5px;">Every 6 months</td> </tr> <tr> <td style="padding: 2px 5px;">>18 years</td> <td style="padding: 2px 5px;">Every year</td> </tr> </tbody> </table>	<u>Age</u>	<u>clinic visits</u>	0-6 months	Every 2 months	6-12 months	Every 3 months	1-18 years	Every 6 months	>18 years	Every year	<p>Clinic Visit labs</p> <ul style="list-style-type: none"> • See above
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<p>Other evaluations</p> <ul style="list-style-type: none"> • Ophthalmology at time of dx then yearly • Yearly developmental questionnaires (to be completed by parents) • Development eval @ 3 & 6 y • Neuropsych @ 9 y • Metabolic dietitian eval (at least yearly) 	<p>Yearly labs</p> <ul style="list-style-type: none"> • Prealbumin • Ferritin /transferritin • Folate and Vitamin B12 <ul style="list-style-type: none"> ○ If not compliant with formula 										

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Performance Indicators

1. Frequency of clinic visits and compliance with visits
2. Age of initiation of diet
3. Initial lab studies
 - a. NBS results
 - b. Quant plasma amino acids
 - c. Urine organic acids
 - d. LFTs
 - e. Molecular studies
 - f. Enzymatic studies
4. Monitoring lab studies
 - a. Phe and Tyr levels Or
 - b. Quant plasma amino acids
 - c. Nutrition labs
5. Eye findings (lacrimation, photophobia, redness, and pain) at time of diagnosis
 - a. Time of resolution of eye findings
6. Skin findings at time of diagnosis (if not diagnosed as newborn)
7. Ophthalmology evaluations
8. Growth parameters (ht, wt, OFC, BMI)
9. Diet
 - a. Frequency of Metabolic dietitian visits
 - b. Frequency of dietary analysis (3 day diet records)
 - c. Natural protein intake (tolerance)
 - d. Formula (Y/N)
 - e. Medical foods (Y/N)
 - f. Mode
10. Growth parameters (ht, wt, OFC, and BMI)
11. Neuropsychological evaluation results
12. Developmental services (PT, OT, & speech)
13. School performance
 - a. Grade appropriate (Y/N)
 - b. Special services (Y/N)
 - c. IEP (Y/N)
14. Genetic Counseling (Y/N)

Outcome Indicators

1. History and/or presence of eye findings (stellate corneal erosions and plaques)
2. History and/or presence of skin findings (persistent keratitis and hyperkeratosis)
3. Development
 - a. IQ
 - b. Level of functioning
 - c. Decline in IQ or level of function
4. Growth
 - a. Final adult parameters