

Biotinidase Deficiency Care Plan (last edited 2/21/09)

<p>Clinical Considerations</p> <ul style="list-style-type: none"> • Partials (see separate care plan) 	<p>Initial labs (diagnostic & baseline)</p> <ul style="list-style-type: none"> • Biotinidase enzyme assay • Consider mutational analysis • Consider UOA /ACP before initiation of biotin treatment 						
<p>Treatment Considerations</p> <ul style="list-style-type: none"> • Biotin 10 mg 	<p>Monitoring labs</p> <ul style="list-style-type: none"> • UOA or ACP yearly 						
<p>Frequency of metabolic visits</p> <table border="1" style="margin-left: auto; margin-right: auto; border-collapse: collapse; text-align: center;"> <thead> <tr> <th style="padding: 2px 5px;">Age</th> <th style="padding: 2px 5px;">Frequency</th> </tr> </thead> <tbody> <tr> <td style="padding: 2px 5px;">0-12 months</td> <td style="padding: 2px 5px;">Every 6 months</td> </tr> <tr> <td style="padding: 2px 5px;">>1 year</td> <td style="padding: 2px 5px;">Once a year</td> </tr> </tbody> </table>	Age	Frequency	0-12 months	Every 6 months	>1 year	Once a year	<p>Clinic Visit labs</p> <ul style="list-style-type: none"> • UOA or ACP yearly
Age	Frequency						
0-12 months	Every 6 months						
>1 year	Once a year						
<p>Other evaluations</p> <ul style="list-style-type: none"> • Audiology eval following Dx with f/u every 3 years • Ophthalmology eval within first year of life then f/u at 6 yrs • Yearly developmental questions (to be completed by parents) • Developmental eval at 3 & 6 yrs • Neuropsych at 9 yrs 	<p>Yearly labs</p> <ul style="list-style-type: none"> • UOA or ACP 						

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Performance Indicators	Outcome Indicators
<ol style="list-style-type: none">1. Age of biotin initiation2. Dosage of biotin3. Initial lab studies<ol style="list-style-type: none">a. Biotinidase activityb. Molecular results4. Monitoring lab studies<ol style="list-style-type: none">a. ACP or UOA results5. Frequency of clinic contact and visits (track compliance with visits)6. Growth parameters (ht, wt, OFC, BMI)7. Neuropsychological evaluation8. Developmental services (PT, OT, & speech)9. School performance<ol style="list-style-type: none">a. Grade appropriate (Y/N)b. Special services (Y/N)c. IEP (Y/N)10. Genetic Counseling (Y/N)	<ol style="list-style-type: none">1. Development (IQ)2. Presence of hearing loss3. History and/or presence of ophthalmologic problems4. History and/or presence of seizures5. History and/or presences of rashes6. Growth<ol style="list-style-type: none">a. Final adult parameters