

Argininemia Care Plan (last updated 2/21/09)

<p>Clinical Considerations</p> <ul style="list-style-type: none"> Spasticity- can present with immobilization after orthopedic surgeries Usually asymptomatic in newborn but rarely can cause severe neonatal illness 	<p>Initial labs (diagnostic & baseline)</p> <ul style="list-style-type: none"> Quant plasma amino acids Ammonia Consider urine orotic acid Red blood cell arginase enzyme assay or sequence analysis 												
<p>Diet considerations/ treatment</p> <ul style="list-style-type: none"> Low-protein diet Targeted arginine level <ul style="list-style-type: none"> <300 µmol/L Formula (essential AA) Medication for hyperammonemia Avoidance of fasting Liver transplant 	<p>Monitoring</p> <ul style="list-style-type: none"> Quant plasma AA Targeted Arginine <300 µmol/L <table border="1" style="margin-left: 40px; 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;">0-6 months</td> <td style="padding: 2px 5px;">Every 2 weeks</td> </tr> <tr> <td style="padding: 2px 5px;">6-12 months</td> <td style="padding: 2px 5px;">Monthly</td> </tr> <tr> <td style="padding: 2px 5px;">1-6 years</td> <td style="padding: 2px 5px;">Every 3 months</td> </tr> <tr> <td style="padding: 2px 5px;">6-18 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;">Yearly</td> </tr> </tbody> </table> <p>***more frequent monitoring may be necessary</p>	Age	Frequency	0-6 months	Every 2 weeks	6-12 months	Monthly	1-6 years	Every 3 months	6-18 years	Every 6 months	>18 years	Yearly
Age	Frequency												
0-6 months	Every 2 weeks												
6-12 months	Monthly												
1-6 years	Every 3 months												
6-18 years	Every 6 months												
>18 years	Yearly												
<p>Frequency of visits</p> <table border="1" style="margin-left: 20px; 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;">0-6 months</td> <td style="padding: 2px 5px;">Every 2 months</td> </tr> <tr> <td style="padding: 2px 5px;">6-24 months</td> <td style="padding: 2px 5px;">Every 3 months</td> </tr> <tr> <td style="padding: 2px 5px;">2-18 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;">Yearly</td> </tr> </tbody> </table>	Age	Frequency	0-6 months	Every 2 months	6-24 months	Every 3 months	2-18 years	Every 6 months	>18 years	Yearly	<p>Clinic visit labs</p> <ul style="list-style-type: none"> Consider ammonia Consider orotic acid Consider prealbumin 		
Age	Frequency												
0-6 months	Every 2 months												
6-24 months	Every 3 months												
2-18 years	Every 6 months												
>18 years	Yearly												
<p>Emergency management</p> <ul style="list-style-type: none"> Immediate 10% (or >) dextrose with salts + intralipids (may need insulin) If ammonia high, IV meds or dialysis Track ammonia levels, IV meds (dose), dialysis, central line, intubation, ICU care, days in hospital, and coma score Transfer to peds center if necessary 	<p>Labs to obtain during illness</p> <ul style="list-style-type: none"> Comp metabolic panel Ammonia SAA 												
<p>Other evaluations</p> <ul style="list-style-type: none"> CT /MRI scan at time of Dx if neurological concerns Consider MRI in childhood if clinically warranted Neuro if clinically warranted Renal and Liver if clinically warranted Bone health <ul style="list-style-type: none"> DEXA-spine @ 9 y for non-weight bearing & 18y for all others Yearly developmental questionnaires (to be completed by parents) Developmental eval @ 3 & 6 y Neuropsych @ 9y Psychiatric screening @ 18 y Metabolic dietitian eval (at least yearly) 	<p>Yearly labs</p> <ul style="list-style-type: none"> Comp metabolic panel Consider ammonia Consider orotic acid Prealbumin / albumin Plasma Ferritin, transferrin, or iron studies Consider CBC, hemoglobin, and hematocrit Consider folate and vitamin B12 Consider other nutritional testing (Zinc, selenium, vit D, essential fatty acids, and lipid profile) 												

Argininemia Care Plan (last updated 2/21/09)

<p>Performance Measures</p> <ol style="list-style-type: none"> 1. Age of diagnosis (both positive NBS and confirmatory SAA) 2. Presence of illness at time of diagnosis (rare) 3. Days until ammonia is within treatment range 4. Initial lab studies <ol style="list-style-type: none"> a. NBS results b. Quant plasma amino acids c. Ammonia d. Urine orotic acid e. Molecular studies f. Enzyme studies 5. Monitoring lab studies <ol style="list-style-type: none"> a. Quant plasma amino acids b. CMP (w/ LFT's) c. Ammonia d. Urine orotic acid e. Nutrition labs 6. Presence or development of spasticity <ol style="list-style-type: none"> a. Use of BOTOX b. Other meds c. Surgery d. Rehab 7. Therapy during initial care <ol style="list-style-type: none"> a. Track IV meds (dose), dialysis, use of central line, intubation, ICU care, days in hospital, and Apache 2 score 8. Frequency of clinic contacts and visits (track compliance with visits) 9. Total decompensations and hospitalizations. <ol style="list-style-type: none"> a. Track ICU admissions b. # of days of hospitalization c. # of ER visits d. Track ammonia level, IV meds (dose), dialysis, use of central line, intubation, and coma score 10. Growth parameters (ht, wt, OFC, BMI) 11. DEXA results and number of fractures 12. Medications (track type and dosage) 	<ol style="list-style-type: none"> 13. Diet <ol style="list-style-type: none"> a. Frequency of Metabolic dietitian visits (phone and clinic visits) b. Frequency of dietary analysis (3 day diet records) c. Natural protein intake (tolerance) d. Medical foods (formula)- (Y/N) e. Modified low-protein products (Y/N) f. Mode (oral, G-tube, bolus/drip, meds only/meds and diet) 14. Neuropsychological evaluation 15. Psychiatric screening 16. Developmental services (PT, OT, & speech) 17. School performance <ol style="list-style-type: none"> a. Grade appropriate (Y/N) b. Special services (Y/N) c. IEP (Y/N) 18. Genetic Counseling (Y/N) <p>Outcome measures</p> <ol style="list-style-type: none"> 1. Mortality 2. Development <ol style="list-style-type: none"> a. IQ b. Level of functioning 3. History and/or presence of liver dysfunction/failure 4. History and/or presence of osteopenia 5. History and/or presence of abnormal MRI findings 6. History and/or presence of spasticity 7. History and/or presence of psychiatric illness 8. Growth <ol style="list-style-type: none"> a. Final adult parameters
--	---