Lab Dept: Urine/Stool

Test Name: CREATINE DISORDERS PANEL, URINE

General Information

Lab Order Codes: UCDP

Synonyms: Arginine: Glycine Amidinotransferase Deficiency (AGAT); Guanidinoacetate Methyltransferase Deficiency (GAMT); CrT1 Defect

CPT Codes: 82540 – Creatine
82570 – Creatinine
82542 – Column chromatography, includes mass spectrometry, if performed, non-drug analytes not elsewhere specified, qualitative or quantitative, each specimen

Test Includes: Urine Creatine, Creatinine & Guanidinoacetate levels reported in nmol/mL and Creatine/Creatinine Ratio.

Logistics

Test Indications: Useful for evaluation of patients with a clinical suspicion of inborn errors of creatine metabolism including arginine:glycine amidinotransferase deficiency (AGAT), guanidinoacetate methyltransferase deficiency (GAMT), and creatine transporter defect (SLC6A8).

Lab Testing Sections: Chemistry - Sendouts

Referred to: Mayo Medical Laboratories (Test# CRDPU)

Phone Numbers: MIN Lab: 612-813-6280
STP Lab: 651-220-6550

Test Availability: Daily, 24 hours

Turnaround Time: 3 – 18 days, performed Wednesdays at 12pm

Special Instructions: Immediately freeze specimen.

Specimen

Specimen Type: Random urine
Container: Plastic, 10-mL urine tube

Draw Volume: 1 mL urine from a random collection

Processed Volume: 1 mL (Minimum: 0.5 mL) from a random urine collection

Collection: Collect a clean random urine specimen

Special Processing: Lab Staff: Immediately freeze 1 mL (Minimum: 0.5 mL) random urine in 10 mL plastic tube. Forward promptly.

Note: If possible, Do Not send other tests ordered on the same vial of urine, otherwise other test turnaround time may increase.

Patient Preparation: None

Sample Rejection: Unlabeled or mislabeled specimen; specimens other than urine; warm specimens

### Interpretive

#### Reference Range:

<table>
<thead>
<tr>
<th>Age</th>
<th>Creatinine (nmol/mL)</th>
<th>Guanidinoacetate (nmol/mL)</th>
<th>Creatine (nmol/mL)</th>
<th>Creatine/Creatinine</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;or=31 days</td>
<td>430 - 5240</td>
<td>9 - 210</td>
<td>12 - 2930</td>
<td>0.02 – 0.93</td>
</tr>
<tr>
<td>32 days – 23 months</td>
<td>313 - 9040</td>
<td>16 - 860</td>
<td>18 - 10490</td>
<td>0.02 – 2.49</td>
</tr>
<tr>
<td>2 – 4 years</td>
<td>1140 - 12820</td>
<td>90 - 1260</td>
<td>200 - 9210</td>
<td>0.04 – 1.75</td>
</tr>
<tr>
<td>5 – 18 years</td>
<td>1190 - 25270</td>
<td>40 - 1190</td>
<td>60 - 9530</td>
<td>0.01 – 0.96</td>
</tr>
<tr>
<td>&gt;18 years Male</td>
<td>3854 - 23340</td>
<td>30 - 710</td>
<td>7 - 470</td>
<td>0.00 – 0.04</td>
</tr>
<tr>
<td>&gt;18 years Female</td>
<td>1540 - 18050</td>
<td>30 - 760</td>
<td>5 - 2810</td>
<td>0.00 – 0.46</td>
</tr>
</tbody>
</table>

Reports include concentrations of guanidinoacetate, creatine and creatinine, and a calculated creatine:creatinine ratio. When no significant abnormalities are detected, a simple descriptive interpretation is provided. When abnormal results are detected, a detailed interpretation is given. This interpretation includes an overview of the results and their significance, a correlation to available clinical information, elements of differential diagnosis, and recommendations for additional biochemical testing.
Critical Values: N/A

Limitations: Correct specimen collection and handling is crucial to achieve reliable results. Creatine supplementation will cause falsely-elevated results.

Methodology: Liquid Chromatography-Tandem Mass Spectrometry (LC-MS/MS): A random urine sample is combined with stable isotope-labeled internal standards and acetonitrile. After centrifugation, an aliquot of this diluted sample is analyzed by injection onto liquid chromatography columns that separate the analytes from the the bulk of the stable isotope dilution in the positive electrospray selected reaction monitoring mode using the applied Biosystems API 3000 MC/MC system with Analyst Software.

References: Mayo Medical Laboratories Web Page (August 2015)

Updates: 1/27/2016: CPT update