

Reporting Title: Guanidinoacetate and Creatine, U**Performing Location:** Kennedy Krieger Ins**Specimen Requirements:**

Collect 2 mL urine from fasting patient. Urine should be collected without preservative. Ship frozen.

Specimen Type	Temperature	Time
Urine	Frozen	

Result Codes:

Result ID	Reporting Name	Type	Unit	LOINC®
Z2990	Creatine/Creatinine ratio	Alphanumeric		
Z2991	Guanidinoacetate	Alphanumeric	mmol/mol Cr	
Z2992	Interpretation	Alphanumeric		

CPT Code Information:

82544

Reference Values:

Creatine/Creatinine ratio

Normal Range:

<4y: 0.01 - 1.19 (SD), N=51

4-12y: 0.01 - 1.51 (SD), N=41

>12y: 0.01 - 0.40 (SD), N=43

Affected Range:

X-linked creatine transporter deficiency:

range = 2.0 - 4.8, n=6

Guanidinoacetate

Normal Range:

<4y: 9 - 200 mmol/mol creatinine, N = 51

4-12y: 12 - 127 mmol/mol creatinine, N = 41

>12y: 8 - 68 mmol/mol creatinine, N = 43

Affected Range:

guanidinoacetate methyltransferase deficiency: 458 - 777, n = 3

arginine: glycine amidinotransferase deficiency: <1, n = 2

Test Performed by: Kennedy Krieger Institute
707 North Broadway
Baltimore, MD 21205