

**Reporting Title:** Amino Acids, QN, Random, U**Performing Location:** Rochester**Additional Testing Requirements:**

Not all patients with homocystinuria will be detected by this assay. If homocystinuria is a concern, order HCYSP / Homocysteine, Total, Plasma or HCYSS / Homocysteine, Total, Serum in tandem with this test.

**Necessary Information:**

1. Patient's age is required.
2. Include family history, clinical condition (asymptomatic or acute episode), diet, and drug therapy information.
3. If prolidase deficiency is a concern, indicate on the amino acid order "Pretreat with acid hydrolysis prior to analysis". The acid hydrolysis will break up in vitro proline and hydroxyproline containing dipeptides, which are cleaved in vivo by prolidase.

**Specimen Requirements:**

Supplies: Urine Tubes, 10 mL (T068)

Specimen Volume: 2 mL

Collection Instructions: Collect a random urine specimen.

**Specimen Minimum Volume:**

1 mL

**Forms:**

If not ordering electronically, complete, print, and send a Biochemical Genetics Test Request (T798) with the specimen.

| Specimen Type | Temperature        | Time    | Special Container |
|---------------|--------------------|---------|-------------------|
| Urine         | Frozen (preferred) | 70 days |                   |
|               | Refrigerated       | 14 days |                   |

**Result Codes:**

| Result ID | Reporting Name      | Type    | Unit       | LOINC®  |
|-----------|---------------------|---------|------------|---------|
| 34466     | Phosphoserine       | Numeric | nmol/mg Cr | 28600-5 |
| 34467     | Phosphoethanolamine | Numeric | nmol/mg Cr | 28604-7 |

| Result ID | Reporting Name             | Type    | Unit       | LOINC®  |
|-----------|----------------------------|---------|------------|---------|
| 3452      | Taurine                    | Numeric | nmol/mg Cr | 28595-7 |
| 3456      | Asparagine                 | Numeric | nmol/mg Cr | 28603-9 |
| 3455      | Serine                     | Numeric | nmol/mg Cr | 30058-2 |
| 34477     | Hydroxyproline             | Numeric | nmol/mg Cr | 28601-3 |
| 3460      | Glycine                    | Numeric | nmol/mg Cr | 30066-5 |
| 3457      | Glutamine                  | Numeric | nmol/mg Cr | 30056-6 |
| 34478     | Aspartic Acid              | Numeric | nmol/mg Cr | 30061-6 |
| 34479     | Ethanolamine               | Numeric | nmol/mg Cr | 28605-4 |
| 3477      | Histidine                  | Numeric | nmol/mg Cr | 30047-5 |
| 3454      | Threonine                  | Numeric | nmol/mg Cr | 30057-4 |
| 3459      | Citrulline                 | Numeric | nmol/mg Cr | 30161-4 |
| 34480     | Sarcosine                  | Numeric | nmol/mg Cr | 28610-4 |
| 3472      | Beta-alanine               | Numeric | nmol/mg Cr | 28588-2 |
| 3461      | Alanine                    | Numeric | nmol/mg Cr | 30068-1 |
| 3458      | Glutamic Acid              | Numeric | nmol/mg Cr | 30059-0 |
| 3476      | 1-Methylhistidine          | Numeric | nmol/mg Cr | 28606-2 |
| 3478      | 3-Methylhistidine          | Numeric | nmol/mg Cr | 28594-0 |
| 34481     | Argininosuccinic Acid      | Numeric | nmol/mg Cr | 32229-7 |
| 3479      | Carnosine                  | Numeric | nmol/mg Cr | 28597-3 |
| 34482     | Anserine                   | Numeric | nmol/mg Cr | 28596-5 |
| 34483     | Homocitrulline             | Numeric | nmol/mg Cr | 32248-7 |
| 3480      | Arginine                   | Numeric | nmol/mg Cr | 30062-4 |
| 3462      | Alpha-aminoadipic Acid     | Numeric | nmol/mg Cr | 28598-1 |
| 34484     | Gamma-amino-n-butyric Acid | Numeric | nmol/mg Cr | 28593-2 |
| 3473      | Beta-aminoisobutyric Acid  | Numeric | nmol/mg Cr | 28602-1 |
| 3463      | Alpha-amino-n-butyric Acid | Numeric | nmol/mg Cr | 28590-8 |
| 34485     | Hydroxylysine              | Numeric | nmol/mg Cr | 30050-9 |
| 3483      | Proline                    | Numeric | nmol/mg Cr | 30067-3 |
| 3474      | Ornithine                  | Numeric | nmol/mg Cr | 30049-1 |
| 3466      | Cystathionine              | Numeric | nmol/mg Cr | 28599-9 |
| 3465      | Cystine                    | Numeric | nmol/mg Cr | 30065-7 |

| Result ID | Reporting Name        | Type         | Unit       | LOINC®  |
|-----------|-----------------------|--------------|------------|---------|
| 3475      | Lysine                | Numeric      | nmol/mg Cr | 30048-3 |
| 3467      | Methionine            | Numeric      | nmol/mg Cr | 30063-2 |
| 3464      | Valine                | Numeric      | nmol/mg Cr | 30064-0 |
| 3470      | Tyrosine              | Numeric      | nmol/mg Cr | 30054-1 |
| 3468      | Isoleucine            | Numeric      | nmol/mg Cr | 30052-5 |
| 3469      | Leucine               | Numeric      | nmol/mg Cr | 30053-3 |
| 3471      | Phenylalanine         | Numeric      | nmol/mg Cr | 30055-8 |
| 34486     | Tryptophan            | Numeric      | nmol/mg Cr | 28608-8 |
| 34487     | Allo-isoleucine       | Numeric      | nmol/mg Cr | 73908-6 |
| 3481      | Interpretation (AAPD) | Alphanumeric |            | 49248-8 |
| 113130    | Reviewed By           | Alphanumeric |            | 18771-6 |

LOINC and CPT codes are provided by the performing laboratory.

## Supplemental Report:

No

## CPT Code Information:

82139

## Reference Values:

| Amino Acid          |      | Age groups      |              |           |           |            |                |
|---------------------|------|-----------------|--------------|-----------|-----------|------------|----------------|
|                     |      | < or =12 months | 13-35 months | 3-6 years | 7-8 years | 9-17 years | > or =18 years |
| Phosphoserine       | PSer | <1              | <1           | <1        | <1        | <1         | <1             |
| Phosphoethanolamine | PEtN | 15-341          | 33-342       | 19-164    | 12-118    | <88        | <48            |
| Taurine             | Tau  | 37-8300         | 64-3255      | 76-3519   | 50-2051   | 57-2235    | 24-1531        |
| Asparagine          | Asn  | 25-1000         | 62-884       | 28-412    | 38-396    | 22-283     | 25-238         |
| Serine              | Ser  | 18-4483         | 284-1959     | 179-1285  | 153-765   | 105-846    | 97-540         |
| Hydroxyproline      | Hyp  | <2536           | <89          | <46       | <19       | <22        | <15            |

|                            |       |           |          |          |          |          |          |
|----------------------------|-------|-----------|----------|----------|----------|----------|----------|
| Glycine                    | Gly   | 362-18614 | 627-6914 | 412-5705 | 449-4492 | 316-4249 | 229-2989 |
| Glutamine                  | Gln   | 139-2985  | 263-2979 | 152-1325 | 164-1125 | 188-1365 | 93-686   |
| Aspartic Acid              | Asp   | <64       | <56      | <30      | <9       | <11      | <10      |
| Ethanolamine               | EtN   | 282-3782  | 256-947  | 193-643  | 137-564  | 158-596  | 95-471   |
| Histidine                  | His   | 145-3833  | 427-3398 | 230-2635 | 268-2147 | 134-1983 | 81-1128  |
| Threonine                  | Thr   | 25-1217   | 55-763   | 30-554   | 25-456   | 37-418   | 31-278   |
| Citrulline                 | Cit   | <72       | <57      | <14      | <9       | <14      | <12      |
| Sarcosine                  | Sar   | <75       | <12      | <9       | <2       | <3       | <3       |
| Beta-Alanine               | bAla  | <219      | <92      | <25      | <25      | <49      | <52      |
| Alanine                    | Ala   | 93-3007   | 101-1500 | 64-1299  | 44-814   | 51-696   | 56-518   |
| Glutamic Acid              | Glu   | <243      | 12-128   | <76      | <39      | <62      | <34      |
| 1-Methylhistidine          | 1MHis | 17-419    | 18-1629  | 10-1476  | 19-1435  | 12-1549  | 23-1339  |
| 3-Methylhistidine          | 3MHis | 88-350    | 86-330   | 56-316   | 77-260   | 47-262   | 70-246   |
| Argininosuccinic Acid      | Asa   | <77       | <48      | <37      | <24      | <69      | <15      |
| Carnosine                  | Car   | 27-1021   | 16-616   | 18-319   | <161     | <109     | <35      |
| Anserine                   | Ans   | <277      | <820     | <398     | <141     | <369     | <38      |
| Homocitrulline             | Hcit  | <295      | 11-158   | <71      | <62      | <33      | <30      |
| Arginine                   | Arg   | 10-560    | 20-395   | 14-240   | <134     | <153     | <114     |
| Alpha-aminoadipic Acid     | Aad   | 10-275    | 15-324   | 10-135   | <84      | <76      | <47      |
| Gamma Amino-n-butyric Acid | GABA  | <25       | <13      | <11      | <6       | <5       | <5       |
| Beta-aminoisobutyric Acid  | bAib  | 18-3137   | <980     | 15-1039  | 24-511   | 11-286   | <301     |
| Alpha-amino-n-butyric Acid | Abu   | <63       | <56      | <38      | <30      | <31      | <19      |
| Hydroxylysine              | Hyl   | <150      | <57      | <34      | <26      | <31      | <12      |
| Proline                    | Pro   | 28-2029   | <119     | <78      | <20      | <28      | <26      |
| Ornithine                  | Orn   | <265      | <70      | <44      | <17      | <18      | <25      |
| Cystathionine              | Cth   | <302      | <56      | <26      | <18      | <44      | <30      |
| Cystine                    | Cys   | 12-504    | 11-133   | <130     | <56      | <104     | 10-98    |
| Lysine                     | Lys   | 19-1988   | 25-743   | 14-307   | 17-276   | 10-240   | 15-271   |
| Methionine                 | Met   | <41       | <41      | <25      | <23      | <20      | <16      |
| Valine                     | Val   | 11-211    | 11-211   | <139     | 16-91    | <75      | 11-61    |
| Tyrosine                   | Tyr   | 39-685    | 38-479   | 23-254   | 22-245   | 12-208   | 15-115   |

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|                 |         |        |        |        |        |        |        |
|-----------------|---------|--------|--------|--------|--------|--------|--------|
| Isoleucine      | Ile     | <86    | <78    | <62    | <34    | <28    | <22    |
| Leucine         | Leu     | <200   | 15-167 | 12-100 | 13-73  | <62    | <51    |
| Phenylalanine   | Phe     | 14-280 | 34-254 | 20-150 | 21-106 | 11-111 | 13-70  |
| Tryptophan      | Trp     | 14-315 | 14-315 | 10-303 | 10-303 | 15-229 | 18-114 |
| Allo-isoleucine | Allolle | <29    | <10    | <8     | <8     | <8     | <7     |

All results reported as nmol/mg creatinine.