

**Table 3. Urine Amino Acid Age-Specific Reference Ranges (mmol/mol creatinine)**

Amino Acid	≤30 days	31 days- 23.9 months	2-17.9 years	Adults (≥18 years)
Aspartic acid	≤8	≤13	≤2	≤2
Glutamic acid	5-22	4-35	≤12	≤4
Hydroxyproline	35-563	2-400	≤5	≤2
Serine	51-527	45-490	15-147	12-82
Asparagine	9-49	6-153	4-49	2-43
α-Aminoadipic acid	≤12	≤42	≤39	≤13
Glycine	249-2,382	122-2,309	27-479	≤383
Glutamine	≤412	48-459	21-218	24-211
Sarcosine	≤21	≤22	≤2	≤80
β-Alanine	≤10	≤17	≤6	≤12
Taurine	≤754	≤777	≤296	≤269
Histidine	46-349	65-630	10-493	20-309
Citrulline	≤5	≤15	≤5	≤2
Arginine	≤35	≤41	≤9	≤6
Threonine	≤130	10-183	5-70	5-53
Alanine	52-306	19-341	9-181	10-78
1-Methylhistidine	≤19	5-82	6-464	≤237
γ-Aminobutyric acid	≤1	≤2	≤2	≤2
3-Methylhistidine	10-52	16-41	13-46	12-41
β-Aminoisobutyric acid	≤312	≤358	≤154	≤102
Proline	≤254	≤251	≤13	≤2
Ethanolamine	101-568	63-204	31-132	24-75
α-Aminobutyric acid	≤8	≤8	≤6	≤2
Tyrosine	5-68	12-80	4-56	4-22
Valine	2-23	5-24	2-23	2-6
Methionine	≤8	≤8	≤6	≤2
Cystathionine	2-23	≤34	≤9	≤10
Isoleucine	≤10	≤14	≤6	≤4
Leucine	≤27	≤28	≤15	≤7
Homocystine	≤1	≤5	≤1	≤1
Phenylalanine	4-28	7-45	2-26	2-10
Tryptophan	2-24	6-53	2-31	2-16
Ornithine	≤45	≤13	≤6	≤5
Lysine	15-329	5-277	4-130	4-69
Cystine	17-56	7-32	4-23	4-15
Hydroxylysine	6-136	2-84	≤9	≤9

The order of amino acids in this table reflects laboratory considerations and matches the order presented in patient reports.