

Supplemental Table S1. Amino acid profile of plasma in the NICCD period of the examined 20 affected children

<i>Urea cycle related amino acids</i> (μM)		<i>Ketogenic amino acids</i> (μM)	
Urea	1765.7 ± 300.2	Tyrosine	89.9 ± 20.9
Citrulline	130.2 ± 39.9	Phenylalanine	77.5 ± 24.8
Arginine	127.4 ± 25.5	Lysine	283.3 ± 29.6
Ornithine	112.7 ± 27.4	Tryptophan	34.7 ± 10.5
Aspartate	4.2 ± 2.2	AAAs	167.4 ± 41.0
Asparagine	47.9 ± 9.1		
Glutamate	58.8 ± 13.4	<i>Branched-chain amino acids</i> (μM)	
Glutamine	284.0 ± 40.6	Leucine	74.4 ± 11.4
		Isoleucine	46.9 ± 8.7
<i>Glucogenic amino acids</i> (μM)		Valine	140.7 ± 32.7
Glycine	187.8 ± 38.0	BCAAs	262.0 ± 37.6
Serine	148.7 ± 29.6		
Alanine	248.4 ± 45.2	<i>Ratios</i>	
Cystine	42.1 ± 6.9	Fisher ratio	1.6 ± 0.4
Methionine	95.8 ± 55.7	Thr/Ser	2.0 ± 0.2
Threonine	289.6 ± 42.7	Cit/Ser	0.9 ± 0.3
Histidine	76.5 ± 19.8	Cit/(Ile+Leu)	1.1 ± 0.4

Data are presented as mean ± SD. BCAAs, the sum of valine, leucine and isoleucine; AAAs, the sum of phenylalanine and tyrosine; Fisher ratio, BCAAs/AAAs; Thr/Ser, the ratio of threonine to serine; Cit/Ser, the ratio of citrulline to serine; Cit/Ser, the ratio of citrulline to the sum of isoleucine and leucine.