

Participant	Gender	Age in yrs	History of HA	2 or more episodes of HA	Liver transplantation	Use of nitrogen-scavenging medications	Height in cm (Z-score)	Weight in kg (Z-score)	Prescribed dose of arginine in g/m <sup>2</sup> of BSA	Prescribed protein from diet in mg/kg	Prescribed protein per day from formula in mg/kg	Total protein prescribed from diet and supplemental formula per day as % of DRI	Pathogenic variants in ASL	LS aBMD Z-score	History of fracture prior to study enrollment (bone(s) fractured) <sup>b</sup>
1	M	11	+	+	-	+	129.5 (-2.3)	34 (-0.5)	1.9	0.67	0.22	100	Allele 1: p.Met256fs Allele 2: p.Ala398Asp	-1.7	Yes (cuboid and tibia)
2	F	8	+	-	-	+	128.7 (-0.2)	30 (0.6)	3.8	0.53	0.36	93	na	0.4	No
3	F	9	+	+	+	-	148.9 (2.3)	56 (2.5)	-	Unrestricted	-	na	Allele 1: p.Arg186Gln Allele 2: p.Arg193Gln	1.5	Yes (clavicle)
4	M	6	-	-	-	+	114.6 (-0.5)	22 (0.1)	4.5	0.6	0.6	91	Allele 1: p.Arg217Ter Allele 2: p.Ser447Asn	0.2	No
5	M	7	+	+	-	+	129.8 (1.1)	30.8 (1.4)	6.6	0.6	0.54	121	Allele 1: p.Leu419fs Allele 2: p.Ser447Asn	0.7	No
6	F	31	-	-	-	-	164.7 <sup>a</sup>	60.3 <sup>a</sup>	1.4	0.66	-	83	Allele 1: c.446+1G>A Allele 2: NA	-1.1	No
7	M	11	+	+	-	+	130.1 (-2.3)	31.7 (-1.1)	2.4	0.55	0.15	74	na	-4.7	No
8	M	9	-	-	-	+	129.4 (-0.5)	27.8 (0.1)	2.7	0.9	-	68	Allele 1: p.Arg12Gln Allele 2: p.Val349Cysfs*72	-0.6	No
9	M	8	-	-	-	-	125.7 (-1.0)	30.7 (0.6)	4.8	0.58	-	61	Allele 1: p.Val178Met Allele 2: p.Val178Met	0.7	No
10	M	6	-	-	-	+	112.5 (-0.7)	19.7 (-0.5)	3.8	0.6	0.6	124	Allele 1: p.Arg217Ter Allele 2: p.Ser447Asn	-2.4	No
11	M	21	+	+	+	-	178.9 <sup>a</sup>	65.2 <sup>a</sup>	1.1	Unrestricted	-	na	na	-0.7	Yes (skull bones)
12	M	20	+	+	+	-	171.8 <sup>a</sup>	77.72 <sup>a</sup>	-	Unrestricted	-	na	Allele 1: p.Gln286Arg Allele 2: na	0.5	No

**Supplemental Table 2: Characteristics of individuals with ASLD who were enrolled in the study.** Plus and minus signs denote the presence and absence of a characteristic, respectively. <sup>a</sup> Height and weight Z-scores are not calculable as the age of the participant is above 20 years. <sup>b</sup> Presence or absence of fracture was based on self- or parent-report. M – male, F-Female; HA hyperammonemia; DRI daily recommended intake; ASL Argininosuccinate lyase; aBMD areal bone mineral density; LS Lumbar spine; na – datum not available