

Supplemental Table 6 The uncorrected concentrations of SNAG- Δ^5 -CA, SNAG- Δ^5 -CG, SNAG- Δ^5 -CT, S7B- Δ^5 -CA and S7O- Δ^5 -CA in the urine of all subjects.

Sample	Age (Years)	Sex	SNAG- Δ^5 -CA (ng/mL)	SNAG- Δ^5 -CG (ng/mL)	SNAG- Δ^5 -CT (ng/mL)	S7B- Δ^5 -CA (ng/mL)	S7O- Δ^5 -CA (ng/mL)	Total (ng/mL)
NPC1	0.25	Male	11210	13900	16000	16800	18100	76200
NPC2	16	Female	611	3810	220	2300	378	7320
NPC3	13	Male	562	459	29.5	352	234	1640
NPC4	12	Female	1860	1730	228	818	557	5190
NPC5	5	Female	546	451	67.7	421	277	1760
NPC6	22	Female	253	419	67.0	699	176	1610
NPC7	15	Male	423	280	109	676	172	1660
NPC8	14	Female	1120	357	88.5	649	234	2450
NPC9	3	Male	4040	1800	755	4030	1850	12500
NPC10	8	Male	1.08	1.58	0.350	3260	493	3750
NPC11	36	Male	348	260	8.53	223	52.1	891
NPC12	24	Female	318	698	35.4	625	102	1780
NPC13	12	Female	529	1360	59.7	884	149	2980
NPC14	26	Female	934	979	137	1690	279	4020
NPC15	18	Female	890	1820	124	1448	225	4510
NPC16	11	Female	599	783	56.1	725	185	2350
NPC17	30	Female	N.Q.	0.780	N.Q.	169	27.5	197
NPC18	0.333	Male	5340	3940	2760	4010	17900	33900
NPC19	0	Male	3940	158	1140	2840	26700	34700
NPC20	11	Female	1090	1370	152	1180	185	3980
NPC21	4	Female	1350	864	55.7	1510	594	4380
NPC22	0.67	Male	235	283	19.0	342	88.9	968

NPC23	30	Female	223	278	42.4	395	99.5	1040
NPC24	0.6	Male	1870	1880	212	1190	279	5430
NPC25	6	Female	465	613	144	262	98.2	1580
NPC26	48	Male	544	607	43.6	328	69.3	1590
NPC27	0.5	Female	239	107	209	94.8	164	814
NPC28	9	Female	2820	2270	247	4060	758	10200
Healthy control1	0.25	Female	5.61	3.19	5.34	4.66	9.94	28.7
Healthy control2	0.33	Male	46.6	39.1	35.9	84.8	34.1	240
Healthy control3	0.33	Female	4.73	5.36	2.48	1.81	1.45	15.8
Healthy control4	0.5	Male	4.12	3.09	4.43	5.93	3.07	20.6
Healthy control5	0.5	Male	13.3	9.80	16.1	26.8	6.69	72.8
Healthy control6	0.5	Male	5.63	4.03	1.85	3.20	3.75	18.5
Healthy control7	1	Male	6.80	3.35	1.47	4.84	2.72	19.2
Healthy control8	1	Male	6.53	5.17	1.50	3.78	1.57	18.6
Healthy control9	1	Female	2.82	6.85	2.13	46.6	7.34	65.8
Healthy control10	2	Male	3.77	3.99	1.29	5.12	1.73	15.9
Healthy control11	2	Female	6.00	5.91	1.75	5.91	1.53	21.1
Healthy control12	2	Female	9.02	15.8	19.1	20.7	5.65	70.3
Healthy control13	2	Male	4.31	4.99	2.11	5.03	1.65	18.1
Healthy control14	2	Male	14.4	9.25	3.99	10.4	3.40	41.4
Healthy control15	2	Male	5.95	7.82	2.05	9.92	4.45	30.2
Healthy control16	35	Female	1.57	4.06	0.620	1.00	0.690	7.94
Healthy control17	28	Female	17.1	13.7	1.24	11.6	5.07	48.8
Healthy control18	25	Female	12.6	16.7	3.09	7.86	5.93	46.2
Healthy control19	23	Female	6.34	8.49	0.890	3.48	1.80	21.0
Healthy control20	24	Male	41.1	45.8	3.01	30.9	12.4	133
Healthy control21	30	Male	10.1	15.4	1.28	6.94	4.06	37.8

Healthy control22	30	Male	18.3	26.2	3.63	14.2	6.07	68.3
Healthy control23	36	Male	11.3	17.9	4.05	13.0	5.87	52.0
Healthy control24	21	Male	2.13	6.56	0.580	4.63	1.26	15.2
Healthy control25	22	Male	13.0	19.2	3.18	12.6	6.99	55.0
Healthy control26	23	Male	N.Q.	N.Q.	N.Q.	20.2	5.05	25.8
Healthy control27	44	Female	28.4	50.7	5.67	18.5	10.5	114
Healthy control28	29	Male	11.8	28.0	6.10	13.8	6.14	65.8
Healthy control29	47	Male	76.8	89.2	5.84	47.1	11.5	230
Healthy control30	31	Male	12.8	20.5	1.62	19.3	3.25	57.5
Healthy control31	36	Male	18.1	23.3	4.13	20.8	10.4	76.6
Healthy control32	36	Male	63.9	162	15.1	38.0	8.50	287
Healthy control33	34	Male	24.1	28.4	8.77	17.2	9.14	87.5
Healthy control34	22	Male	11.2	17.3	5.75	47.8	12.8	94.9
Healthy control35	23	Female	64.5	103	12.1	35.4	17.7	232
Healthy control36	21	Male	22.8	45.8	5.16	39.4	11.8	125
Healthy control37	21	Female	11.5	12.3	0.620	15.1	6.19	45.7
Healthy control38	26	Male	14.030	28.2	3.62	9.88	4.52	60.3
CTX01	42	Female	3.00	9.60	1.40	4.60	1.90	20.5
CTX02	56	Female	7.85	5.35	3.2	11.2	2.34	30.0
CTX03	50	Female	11.5	7.82	4.1	6.91	3.64	34.0
GAU01	18	Female	87.54	58.5	4.6	26.6	0.92	178
GAU02	20	Female	N.Q.	0.350	0.410	17.7	6.15	24.7
GAU03	15	Female	73.0	27.2	9.75	49.2	14.0	173
HUN01	18	Male	2.78	6.07	2.02	10.3	1.92	23.0
HUN02	8	Male	11.7	9.38	1.37	4.77	2.92	30.1
SLOS01	22	Male	25.7	33.0	12.8	126	29.5	227

N.Q., Quantitation was not carried out because it was below the lower limit of determination; NPC, Niemann–Pick disease type C; SNAG- Δ^5 -CA, 3 β -Sulfooxy-7 β -*N*-acetylglucosaminyl-5-cholenoic acid; SNAG- Δ^5 -CG, Glycine-amidated 3 β -sulfooxy-7 β -*N*-acetylglucosaminyl-5-cholenoic acid; SNAG- Δ^5 -CT, Taurine-amidated 3 β -sulfooxy-7 β -*N*-acetylglucosaminyl-5-cholenoic acid; S7B- Δ^5 -CA, 3 β -Sulfooxy-7 β -hydroxy-5-cholenoic acid; S7O- Δ^5 -CA, 3 β -Sulfooxy-7-oxo-5-cholenoic acid.