

Supplementary Table 7 The creatinine-corrected concentration of SNAG- Δ^5 -CA, SNAG- Δ^5 -CG, SNAG- Δ^5 -CT, S7B- Δ^5 -CA, S7O- Δ^5 -CA and the total concentration in urine of healthy controls and NPC patients.

Healthy controls						
	SNAG- Δ^5 -CA (ng/mg Cr)	SNAG- Δ^5 -CG (ng/mg Cr)	SNAG- Δ^5 -CT (ng/mg Cr)	S7B- Δ^5 -CA (ng/mg Cr)	S7O- Δ^5 -CA (ng/mg Cr)	Total (ng/mg Cr)
Mean	23.7	25.1	12.6	24.5	13.9	98.2
SD	28.9	27.0	22.5	29.3	29.1	118
Minimum	3.33	6.06	0.305	5.41	1.50	9.78
Median	12.8	16.7	3.25	12.8	4.92	57.9
Maximum	145	165	92.7	125	172	499
Patients with NPC						
	SNAG- Δ^5 -CA (ng/mg Cr)	SNAG- Δ^5 -CG (ng/mg Cr)	SNAG- Δ^5 -CT (ng/mg Cr)	S7B- Δ^5 -CA (ng/mg Cr)	S7O- Δ^5 -CA (ng/mg Cr)	SNAG- Δ^5 -CAs (ng/mg Cr)
Mean	11100	11300	7270	9670	12100	47400
SD	26700	29200	3710	20000	32900	97000
Minimum	0.561	0.821	0.182	142	35.8	373
Median	897	879	85.7	887	231	2730
Maximum	131000	132000	42800	83400	148000	381000

Cr, Creatinine; 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.