



**Figure S1** Taxonomic characterization of gut microbiota composition at (A) phylum, (B) family, and (C) genus level for the control and GSD groups. GSD, glycogen storage disease.

**Table S1** Comparison of fecal BAs profiles between control and GSD groups

Class	BAs	Control, mean ( $\mu\text{g/g}$ )	GSD, mean ( $\mu\text{g/g}$ )	P value <sup>#</sup>
Primary BAs	CA	11.2801969	20.61990685	0.020759
	CDCA	10.17369661	3.683456148	0.0025013
	UCA	2.151716956	3.793816616	0.2983419
	12-KCDCA	1.506935744	2.546713802	0.0457812
	3 $\beta$ -UDCA	0.858643292	1.772437956	0.2425835
	$\beta$ -MCA	0.769340721	0.848135194	0.9180704
	GCA	0.367720148	0.901878028	0.4872060
	3-DHCA	0.297299973	0.577481671	0.0278792
	GCDCA	0.094808005	0.10204944	0.0383687
	HCA	0.002386241	0.002892227	0.3212003
	TCA	0.291772874	0.560007226	0.6491462
	TCDCA	0.217503262	0.544295142	0.5311629
	AlloCA	0.207295349	0.257712334	0.7294114
	$\omega$ -MCA	0.176985013	0.24962383	0.4777140
	NorCA	0.077979959	0.075228236	0.6524580
	$\alpha$ -MCA	0.041208593	0.0739221	0.4470741
	TDCA	0.051464697	0.004181661	0.0019313
	MDCA	0.025885312	0.011062938	0.0129266
	DHCA	0.016696444	0.003379991	0.2773761
	GHCA	0.002688107	0.008082932	0.5143057
	T- $\omega$ -MCA	0.00196323	0.000486102	0.0752088
	T- $\alpha$ -MCA	0.004935654	0.001260246	0.0207982
	T- $\beta$ -MCA	0.176985013	0.24962383	0.1551362
	isoDCA	0.000643001	0	0.2462265
	CDCA-3Gln	0.010842527	0.198311731	0.0088916
	Secondary BAs	DCA	13.06607922	4.743537876
7-DHCA		5.553409697	10.24216264	0.1394081
LCA		11.11933421	3.92771393	0.0041409
UDCA&HDCA		4.662080457	7.035059654	0.2485370
7-KLCA		4.63394691	10.24216264	0.0309491
12-KLCA		5.321889503	0.979024503	0.0001670
isoLCA		4.198593239	0.838093409	0.0024490
LCA-3S		0.359822515	0.345085511	0.8634095
DHLCA		0.478861258	0.049911224	0.0002185
7,12-DKLCA		0.186715385	0.063402632	0.1445863

Table S1 (continued)

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Class	BAs	Control, mean ( $\mu\text{g/g}$ )	GSD, mean ( $\mu\text{g/g}$ )	P value <sup>#</sup>
	CDCA-24A- $\beta$ Glu	0.003608031	0.149087123	0.0007537
	TUDCA&THDCA	0.030558244	0.024038676	0.6693103
	GUDCA	0.030890911	0.022523335	0.6046564
	GDCA	0.025618494	0.004770455	0.0005729
	ApoCA	0.007262285	0.008993431	0.5648654
	NorDCA	0.010398932	0.001456793	0.0001890
	GLCA	0.008411213	0.001430293	0.0060408
	TLCA	0.006897288	0.001003956	0.0084098
	THCA	0.002386241	0.002892227	0.7967290

<sup>#</sup>, Mann-Whitney test. BAs, bile acids; GSD, glycogen storage disease.