

## Supplementary Materials

**Table 1. Blood biochemical parameters in GSD patients.** Values are expressed as mean (standard deviation). Reference values are reported.

	<b>GSD Mean (SD)</b>	<b>GSD Ia Mean (SD)</b>	<b>GSD Ib Mean (SD)</b>	<b>Reference values</b>
<b>Glucose, fasting</b>	71.38 (10.28)	65.50 (7.05)	80.00 (10.79)	70–105 mg/dL
<b>Insulin, fasting</b>	5.88 (9.74)	2.45 (2.86)	8.96 (11.70)	0-25 uU/ml
<b>HOMA-IR</b>	1.10 (1.89)	0.42 (0.54)	1.76 (2.26)	
<b>QUICKI Index</b>	0.47 (0.12)	0.53 (0.13)	0.39 (0.07)	
<b>HOMA-<math>\beta</math></b>	1.64 (2.59)	0.76 (0.77)	2.37 (3.14)	
<b>Tyg-Index</b>	3.95 (0.42)	4.18 (0.39)	3.80 (0.35)	
<b>Cholesterol, total</b>	206.75 (124.68)	265.50 (152.17)	166.40 (68.02)	< 200 mg/dl
<b>Triglycerides</b>	300.25 (225.92)	422.50 (241.58)	204.60 (138.56)	<150 mg/dL
<b>Uric acid</b>	6.675 (1.03)	7.10 (0.50)	6.25 (3.02)	2.5–8 mg/dL
<b>Serum lactate</b>	3.00 (1.77)	3.85 (1.93)	2.14 (1.10)	0.7-2.1 mmol/L
<b>AST</b>	42.5 (23.84)	54.50 (28.35)	37.80 (19.25)	0–35 U/L
<b>ALT</b>	54.13 (43.44)	67.75 (47.13)	50.80 (42.36)	0–35 U/L

**Table 2. Fecal SCFA concentrations.** For each experimental group, mean and standard deviation from Mann-Whitney test are reported. Values are expressed as mean (standard deviation). P-values <0.05 were considered significant.

	<b>HC Mean (SD)</b>	<b>GSD Mean (SD)</b>	<b>p-value</b>
<b>Acetate</b>	1.84 (0.73)	3.54 (1.95)	0.0310*
<b>Propionate</b>	0.72 (0.24)	1.41 (1.07)	0.0381*
<b><i>iso</i>-butyrate</b>	0.29 (0.12)	0.28 (0.22)	0.8965
<b>Butyrate</b>	0.32 (0.12)	0.33 (0.22)	0.8381
<b><i>iso</i>-valerate</b>	0.24 (0.11)	0.19 (0.16)	0.3598
<b>Total SCFAs</b>	3.40 (0.85)	5.75 (2.88)	0.0159*