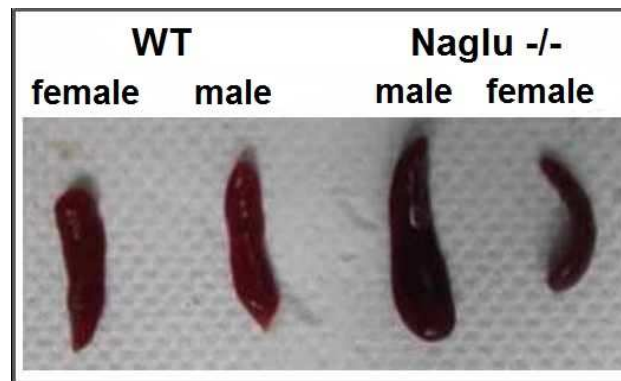


## Supplement

### Effect of glycosaminoglycans accumulation on the non-oxidative sulfur metabolism in mouse model of Sanfilippo syndrome, type B\*

Marta Kaczor-Kamińska, Kamil Kamiński, Krystyna Stalińska, Maria Wróbel, Arleta Feldman,

**Figure S1.** Comparison of spleen size and appearance depending on mouse genotype (individuals randomly selected).



**Table S1.** Level of sulfane sulfur and sulfurtransferases activity in selected tissues of mice, both female and male.

Values represent the arithmetic mean  $\pm$  SD of 3-4 animals, which each determination consisting of 4-20 assays.

\*p < 0.05; \*\*p < 0.01; \*\*\* p < 0.001 ( Student t-test). ND - not detected.

Experimental group		Sulfane sulfur [nmol/mg protein]	TST [nmol/mg protein·min]	MPST [nmol/mg protein·min]	CTH [nmol/mg protein·min]
<b>LIVER</b>					
WT	Female	120.99 $\pm$ 15.98	520.16 $\pm$ 47.72	345.37 $\pm$ 80.70	3.268 $\pm$ 0.524
	Male	109.79 $\pm$ 18.00	433.33 $\pm$ 28.91***	285.28 $\pm$ 112.64	2.945 $\pm$ 0.303*
Naglu <sup>-/-</sup>	Female	115.91 $\pm$ 8.22	548.80 $\pm$ 43.20	283.05 $\pm$ 73.24	2.026 $\pm$ 0.051
	Male	96.56 $\pm$ 7.26***	459.64 $\pm$ 58.44***	262.14 $\pm$ 68.19	1.961 $\pm$ 0.098*
<b>KIDNEY</b>					
WT	Female	142.15 $\pm$ 17.27	182.95 $\pm$ 5.60	339.83 $\pm$ 24.55	0.919 $\pm$ 0.182
	Male	145.20 $\pm$ 21.24	224.75 $\pm$ 30.00***	323.56 $\pm$ 88.76	0.718 $\pm$ 0.114***
Naglu <sup>-/-</sup>	Female	135.45 $\pm$ 12.59	191.03 $\pm$ 25.40	258.82 $\pm$ 49.46	1.080 $\pm$ 0.143
	Male	127.07 $\pm$ 13.21	214.65 $\pm$ 32.54*	244.34 $\pm$ 60.73	0.779 $\pm$ 0.124***
<b>HEART</b>					
WT	Female	193.19 $\pm$ 30.68	37.61 $\pm$ 7.55	78.75 $\pm$ 18.73	0.099 $\pm$ 0.073
	Male	204.32 $\pm$ 21.57	32.90 $\pm$ 4.24*	67.56 $\pm$ 15.49	0.156 $\pm$ 0.032
Naglu <sup>-/-</sup>	Female	199.26 $\pm$ 29.93	41.18 $\pm$ 3.51	89.93 $\pm$ 12.72	ND
	Male	214.23 $\pm$ 16.98	38.87 $\pm$ 2.33*	69.57 $\pm$ 19.45**	0.206 $\pm$ 0.040
<b>SPLEEN</b>					
WT	Female	77.16 $\pm$ 5.87	31.65 $\pm$ 1.12	52.10 $\pm$ 9.93	0.134 $\pm$ 0.036
	Male	60.39 $\pm$ 1.24***	13.09 $\pm$ 1.27***	59.56 $\pm$ 6.41	0.041 $\pm$ 0.000*
Naglu <sup>-/-</sup>	Female	67.03 $\pm$ 13.50	24.11 $\pm$ 2.36	34.99 $\pm$ 13.49	0.237 $\pm$ 0.048
	Male	70.94 $\pm$ 10.13	16.35 $\pm$ 0.00***	41.28 $\pm$ 7.91	ND