

## Online Resource 1

- **Figure S1**
- **Figure S2**
- **Figure S3**

### **Glycosaminoglycan signatures in body fluids of Mucopolysaccharidosis type II mouse model under long-term enzyme replacement therapy**

Journal of Molecular Medicine

**Authors:** Maccari F, Rigon L, Mantovani V, Galeotti F, Salvalaio M, D'Avanzo F, Zanetti A, Capitani F, Gabrielli O, Tomanin R and Volpi N

Corresponding authors:

Nicola Volpi (e-mail address: [volpi@unimo.it](mailto:volpi@unimo.it)) - Department of Life Sciences, University of Modena and Reggio Emilia, Modena, Italy

Rosella Tomanin (e-mail address : [rosella.tomanin@unipd.it](mailto:rosella.tomanin@unipd.it))- Department of Women's and Children's Health, University of Padova, Italy

## Online Resource 1 - Figure S1

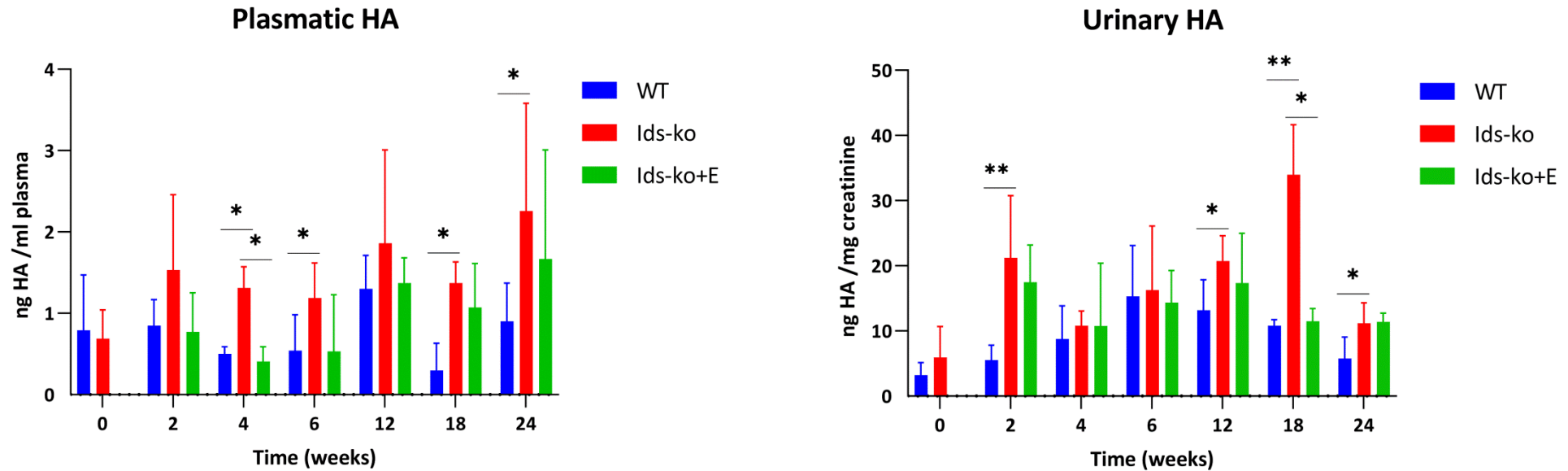


**Figure S1.** Quantitation of plasmatic and urinary HA by CE-LIF. HA concentration measured in plasma and urine of wild type, Ids-ko and Ids-ko mice treated with ERT (Ids-ko+E), during a time course of 24 weeks at T = 0, 2, 4, 6, 12, 18, 24 weeks. All values are represented as mean  $\pm$ SD. n = 6-7 for each group. Asterisks indicate a statistically significant difference between Ids-ko and WT and between Ids-ko and Ids-ko+E at the same time point (Mann–Whitney U test; \* 0.01  $\leq$  p-value < 0.05; \*\* 0.001  $\leq$  p-value < 0.01)

## Online Resource 1 - Figure S2

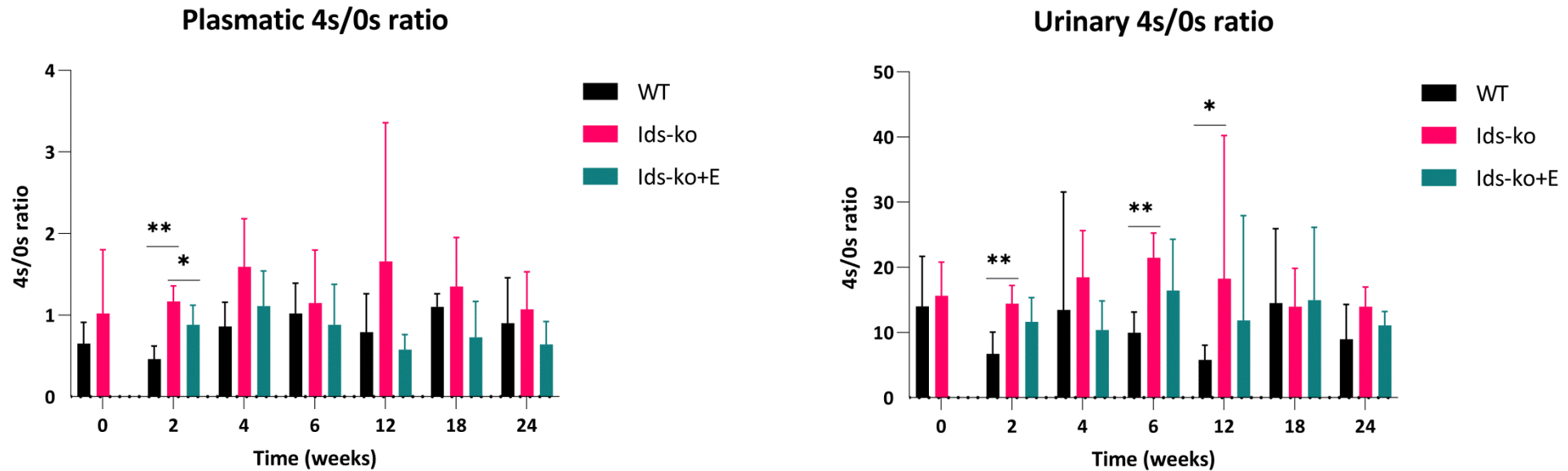


**Figure S2.** Plasmatic and urinary CS+DS 4s/0s disaccharides ratio. Ratio between the percentage of disaccharide 4s and the percentage of non-sulfated disaccharide (4s/0s) of CS+DS measured in plasma and urine of wild type, Ids-ko and Ids-ko mice treated with ERT (Ids-ko+E), during a time course of 24 weeks at T = 0, 2, 4, 6, 12, 18, 24 weeks. All values are represented as mean  $\pm$ SD. n = 6-7 for each group. Asterisks indicate a statistically significant difference between Ids-ko and WT and between Ids-ko and Ids-ko+E at the same time point (Mann–Whitney U test; \* 0.01  $\leq$  p-value < 0.05; \*\* 0.001  $\leq$  p-value < 0.01).

Online Resource 1 - Figure S3

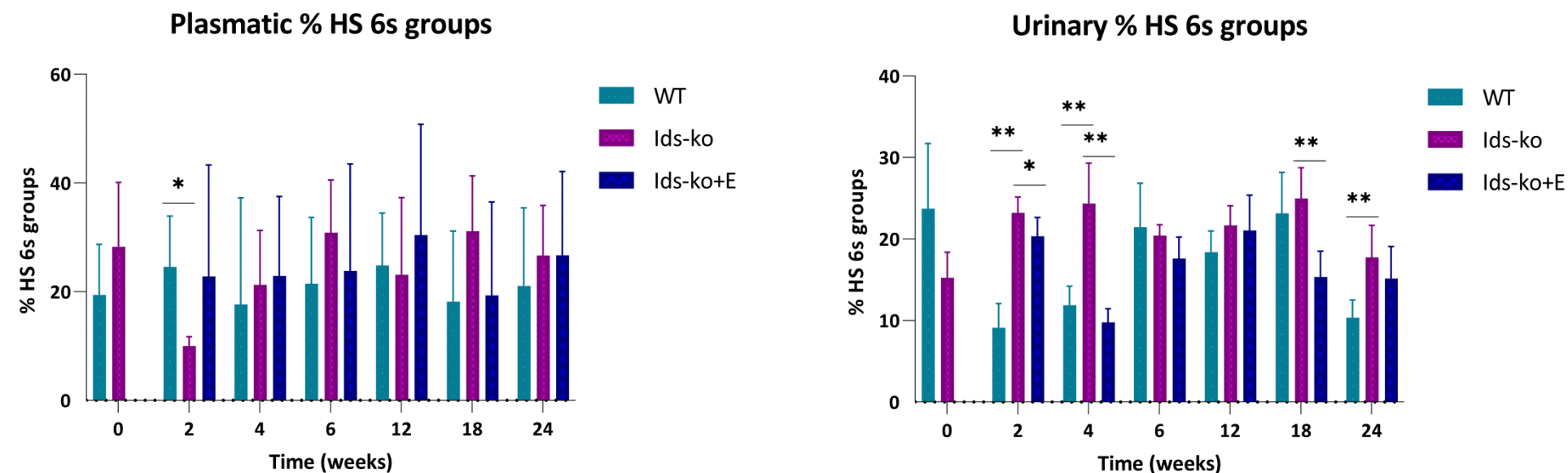


**Figure S3.** Percentage of plasmatic and urinary HS 6s disaccharides. Percentage of HS 6-sulfated disacaccharides measured in plasma and urine of wild type, Ids-ko and Ids-ko mice treated with ERT (Ids-ko+E), during a time course of 24 weeks at T = 0, 2, 4, 6, 12, 18, 24 weeks. All values are represented as mean  $\pm$ SD. n=6-7 for each group. Asterisks indicate a statistically significant difference between Ids-ko and WT and between Ids-ko and Ids-ko+E at the same time point (Mann–Whitney U test; \* 0.01 $\leq$ p-value<0.05; \*\* 0.001 $\leq$ p-value<0.01).