



Figure S3 3UP8i increases FL-*SMN2* levels in skeletal muscle of Hemi-hybrid SMA mice. (*SMN2*^{tg/o}; *Smn*^{tm1Hung/tm1.1Jme}) were administered three doses (80 μ g/g body weight at P0, P5, P10) of 3UP8i or mismatch control (CON8i) by intraperitoneal (IP) injection. **(a)** *SMN2* exon-7 inclusion. Quadriceps muscle was harvested at P12 and analyzed via endpoint RT-PCR for full-length FL-*SMN2* and Δ 7-*SMN2*. **(b)** FL- and Δ 7-*SMN2* levels were plotted as ratio to total *SMN2* levels. Data presented as mean \pm SEM. (n=3). Student's *t*-test, ***P*<0.01 (FL). **(c)** Quadriceps muscle was harvested at P12, and subjected to immunoblot with α -SMN and α -tubulin antibodies. **(d)** SMN levels were normalized to β -tubulin. Data presented as mean \pm SEM. (n=3). Student's *t*-test, ***P*<0.01.