

Figure S1. A. In genotype, the functional SMN is 800 bp length DNA, the mutated Smn is 500 bp length. B. The weights in specific muscles of tibialis (TA) $(52.10 \pm 5.738 \text{ mg N=4} \text{ vs } 55.625 \pm 2.616 \text{ mg N=8}, \text{P=0.997})$ gastrocnemius (Gas) $(134.15 \pm 20.935 \text{ mg N=4} \text{ vs } 138.9725 \pm 5.459268 \text{ mg 4=N}, \text{P=0.995})$ and quadriceps femoris (Quad) $(217.33 \pm 34.139 \text{ mg N=4} \text{ vs } 224.38 \pm 5.933 \text{ mg N=4}, \text{P=0.984})$ had no difference between WT and SMA. C. SMA mice had lower or even deleted expression level of SMN protein in GI tissues, such as colon, stomach and small intestine, compared to WT. But SMA mice were found having leaky expression from the *PrP92-SMN* transgene, outside of the neuronal compartment, especially in gastrocnemius (Gas) quadriceps femoris (Quad) and etc.