

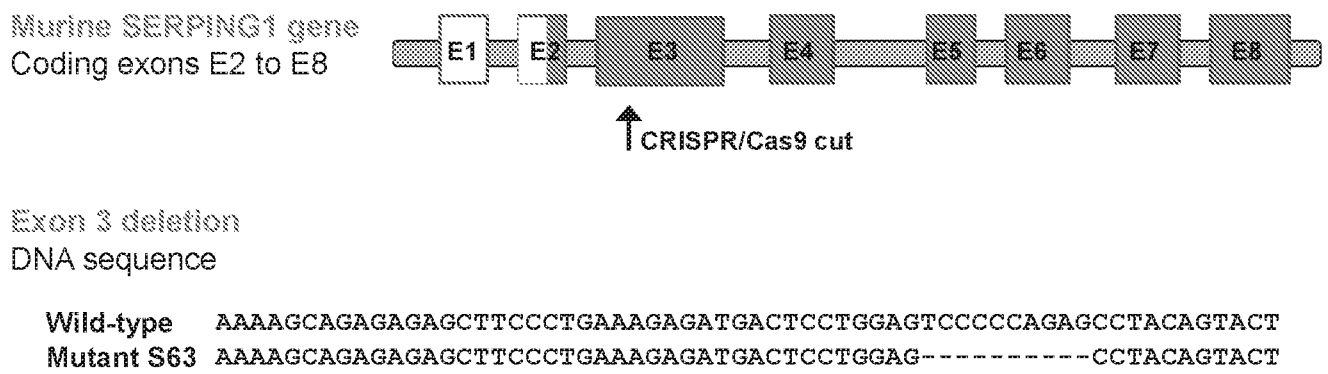
Figure 1

Figure 2A

Mouse C1EI (ELISA)

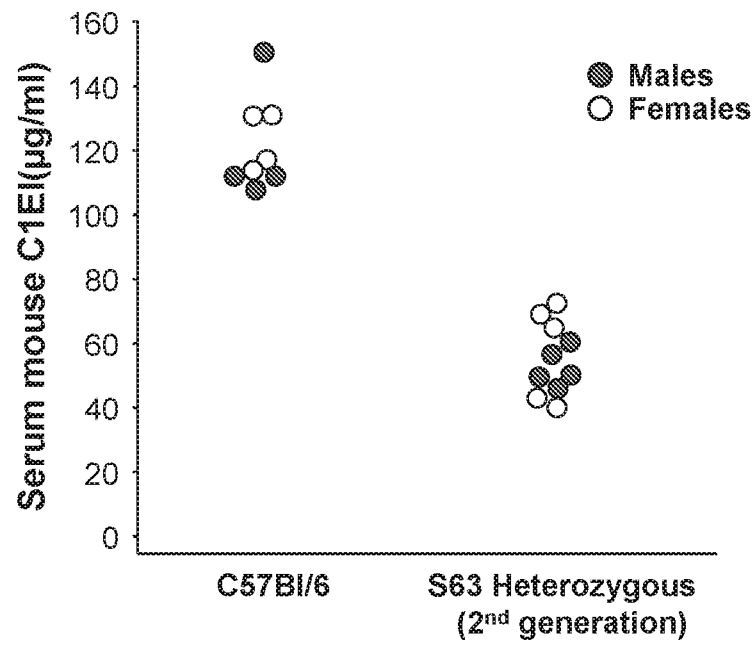


Figure 2B

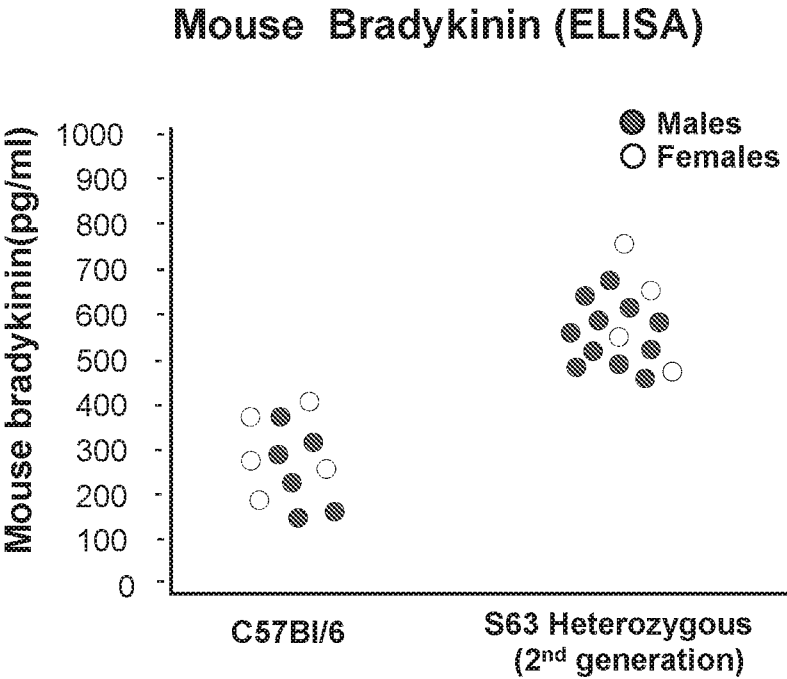


Figure 3A

Expression cassette

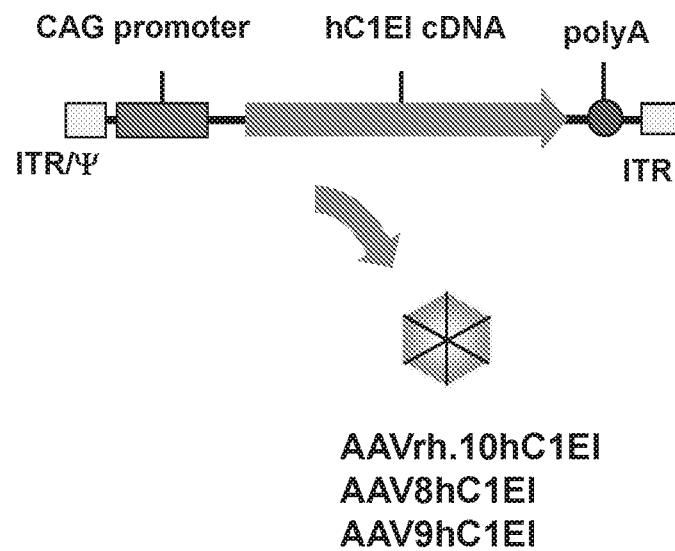


Figure 3B

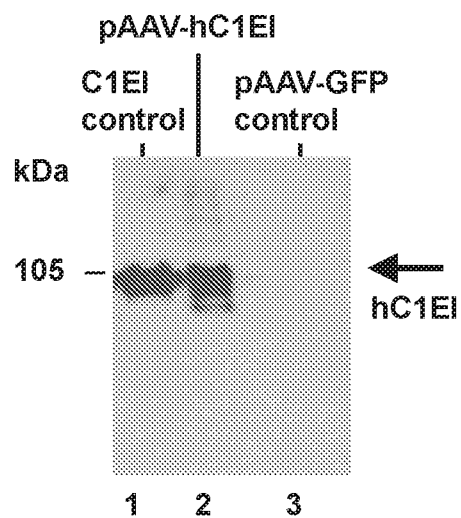
AAVrh.10hC1EI *in vitro* directed expression of C1EI

Figure 4A

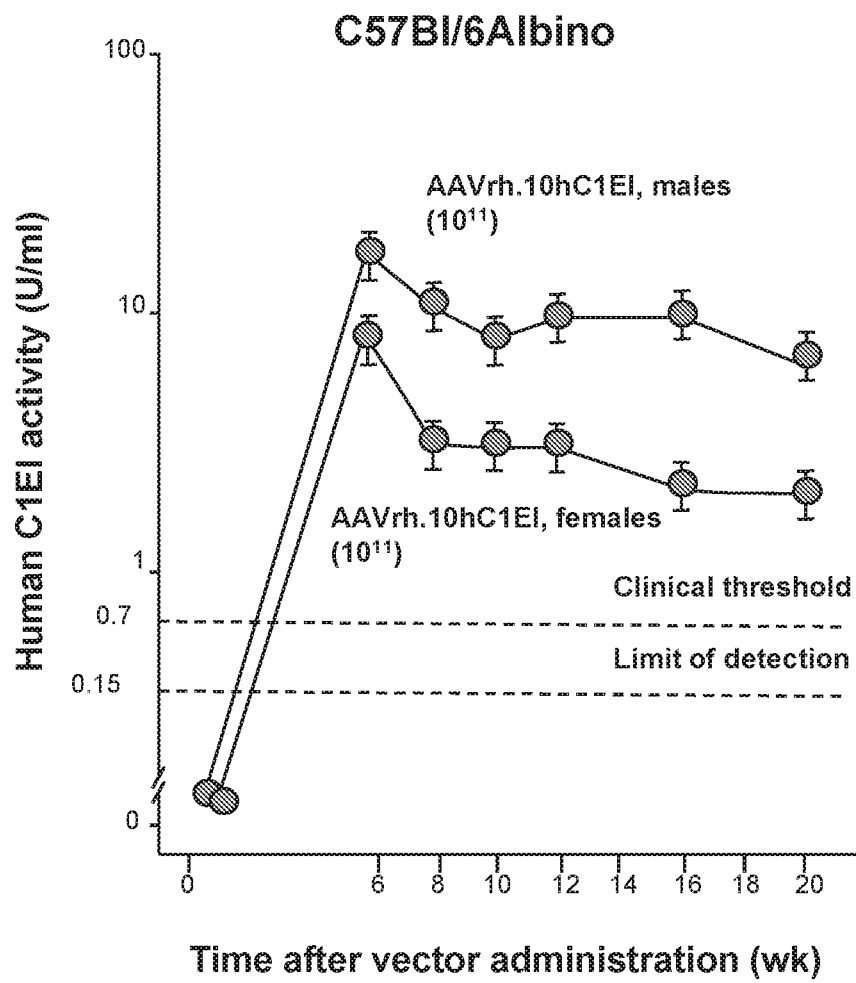


Figure 4B

C57Bl/6Albino (Dose Response)

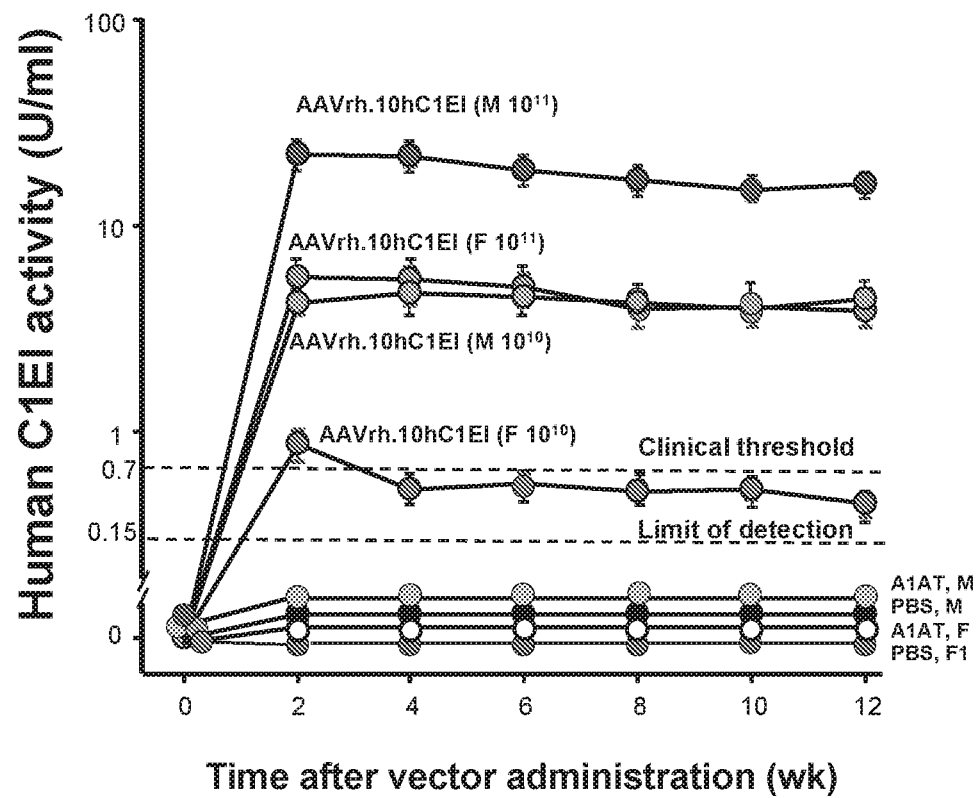


Figure 4C

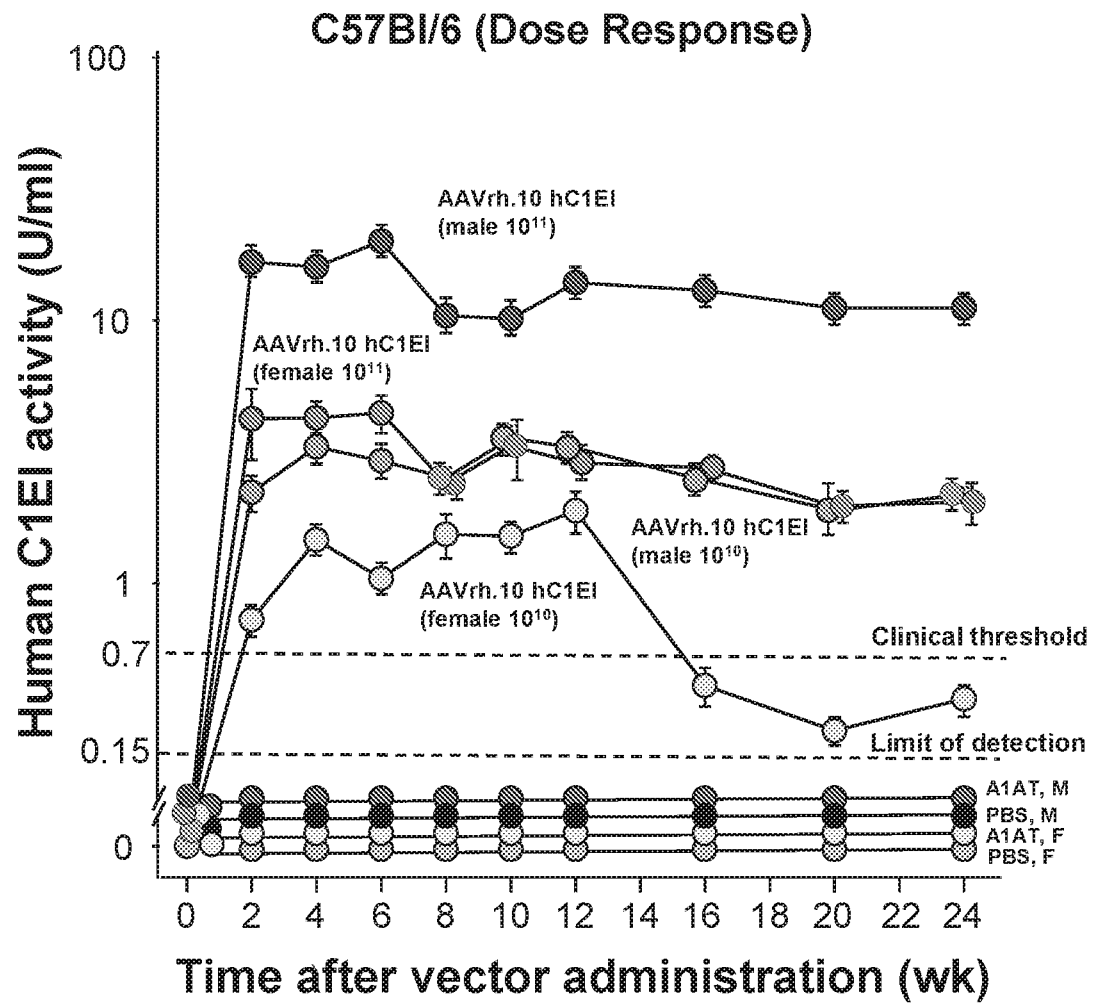
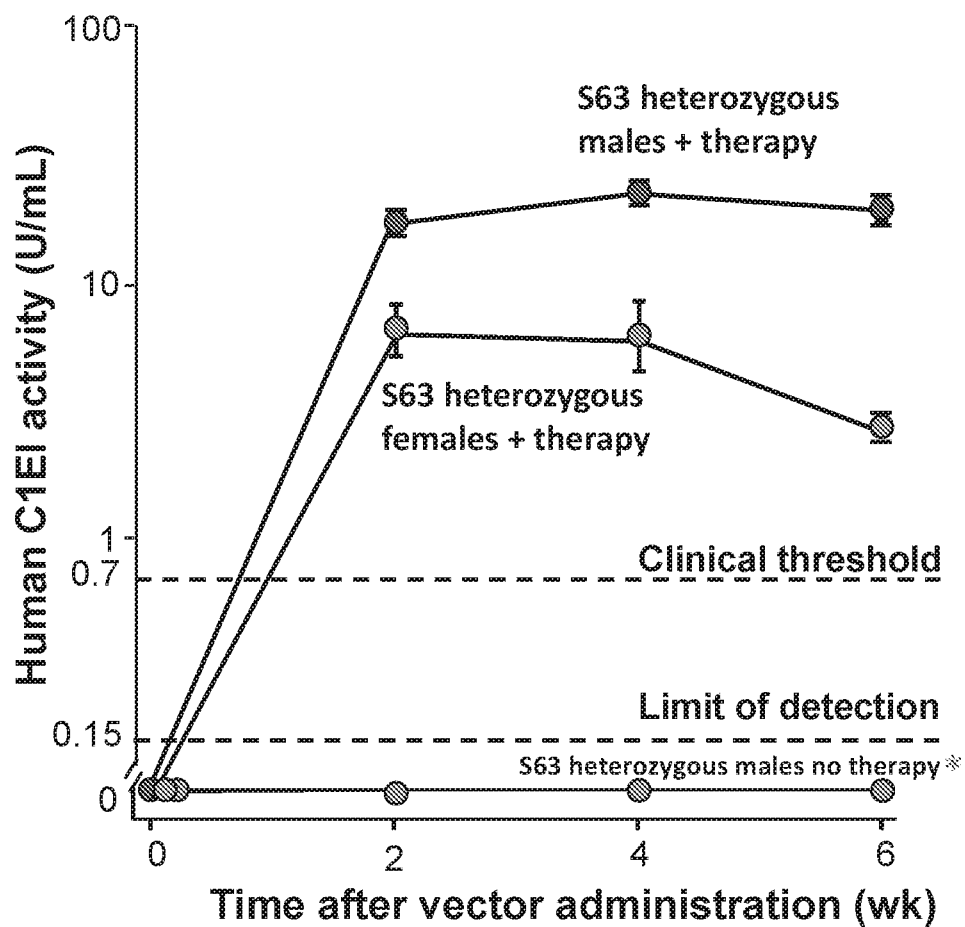


Figure 4D

S63 Heterozygous Mice



*S63 heterozygous females no therapy n=1, no activity

Figure 5A

AAV8hC1EI Vector

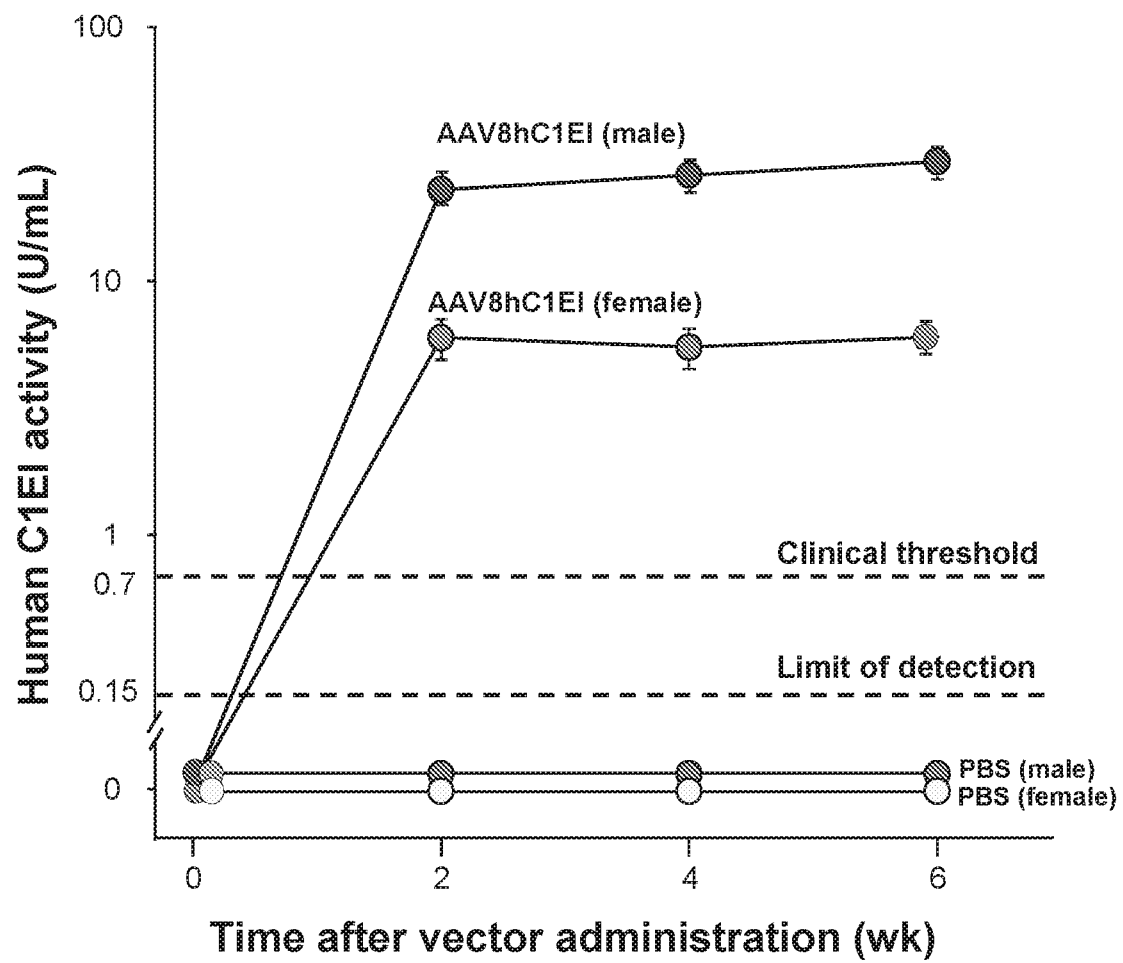


Figure 5B

AAV9hC1EI Vector

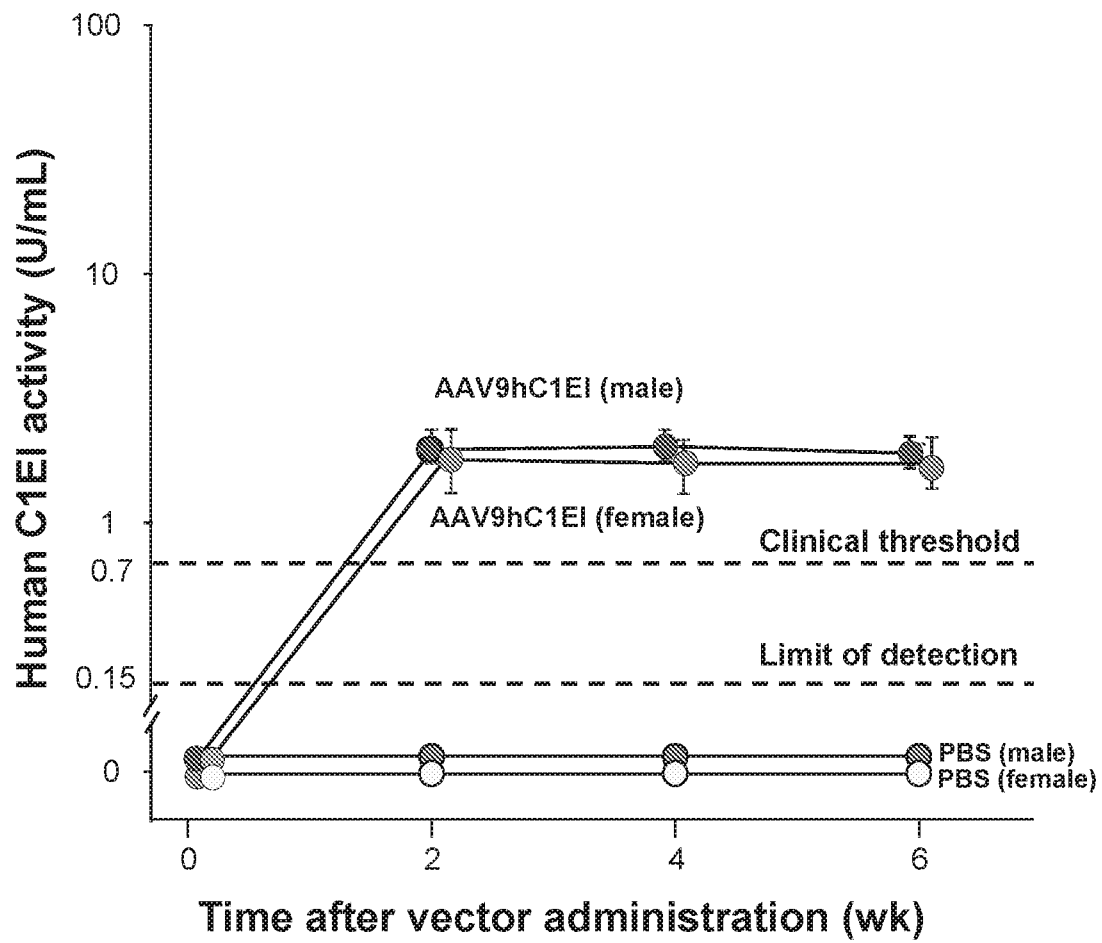


Figure 5C

AAVrh.10hC1EI Vector

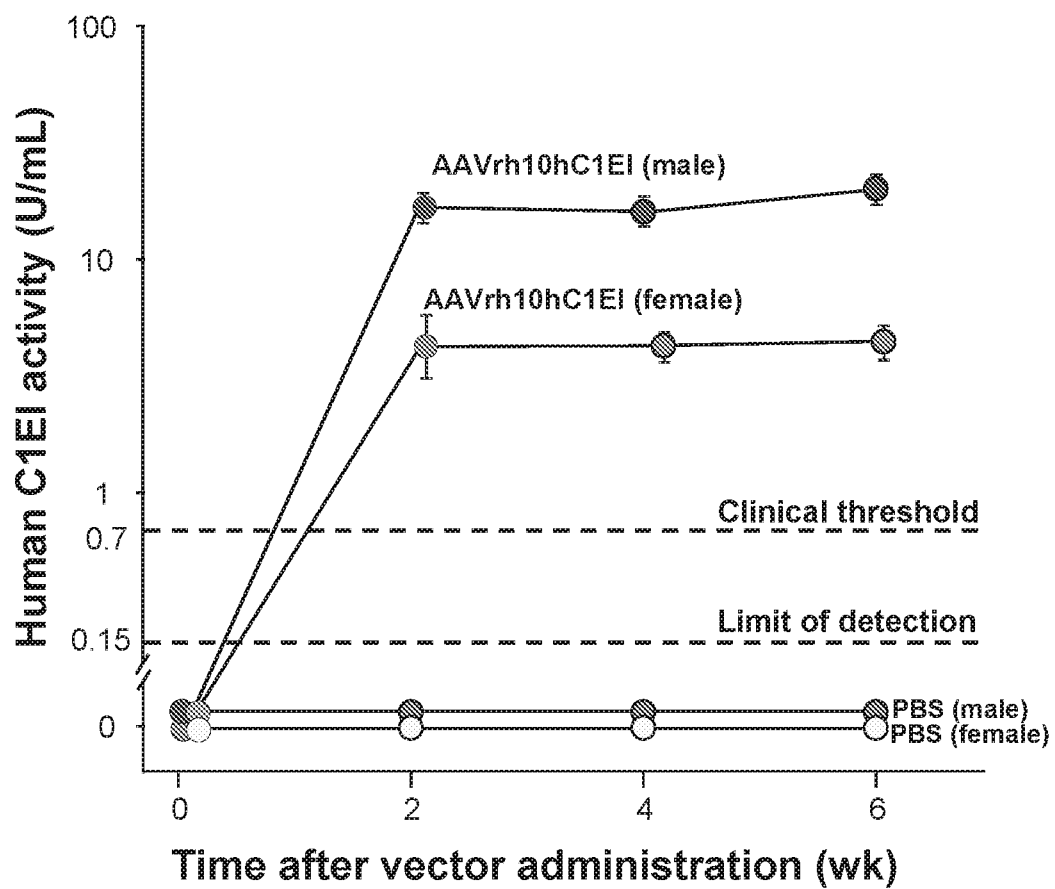


Figure 5D

AAV8hC1EI, AAV9hC1EI, or AAVrh.10hC1EI Vector

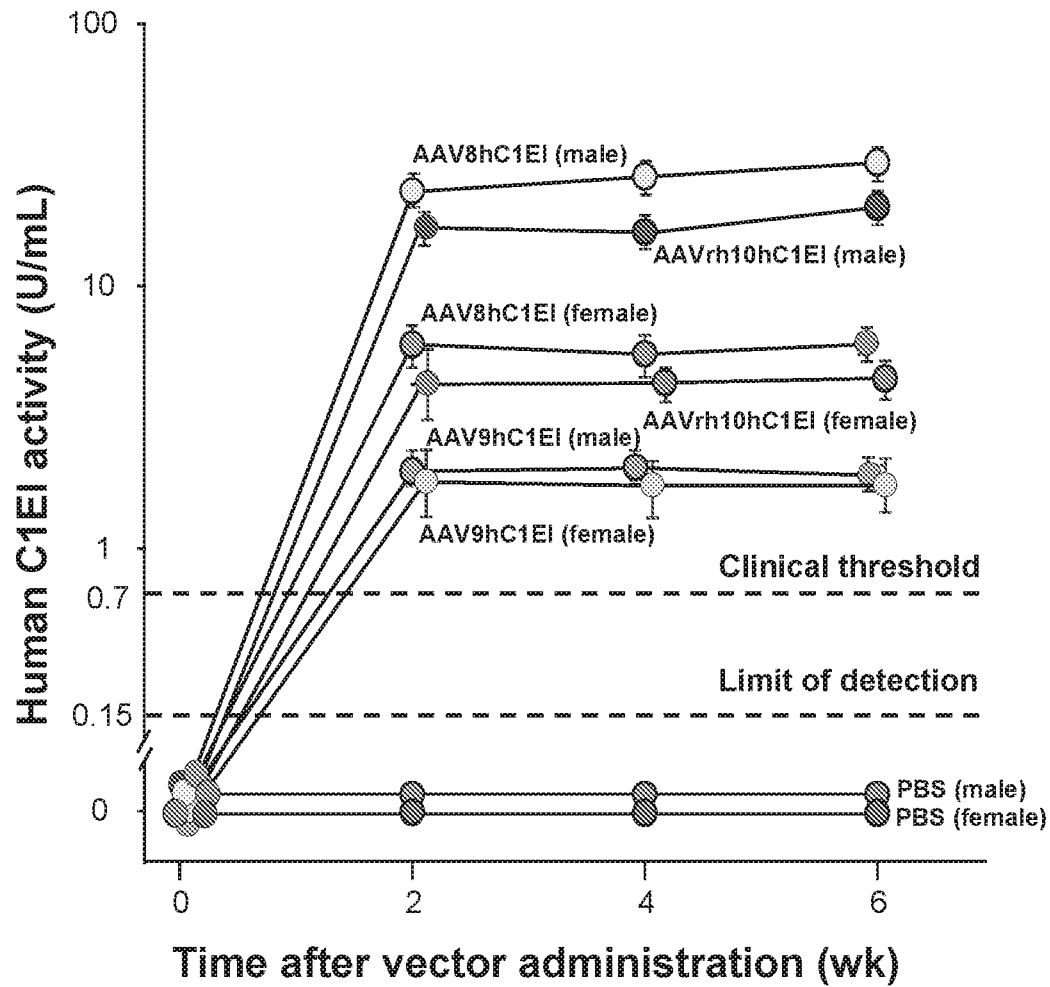


Figure 6

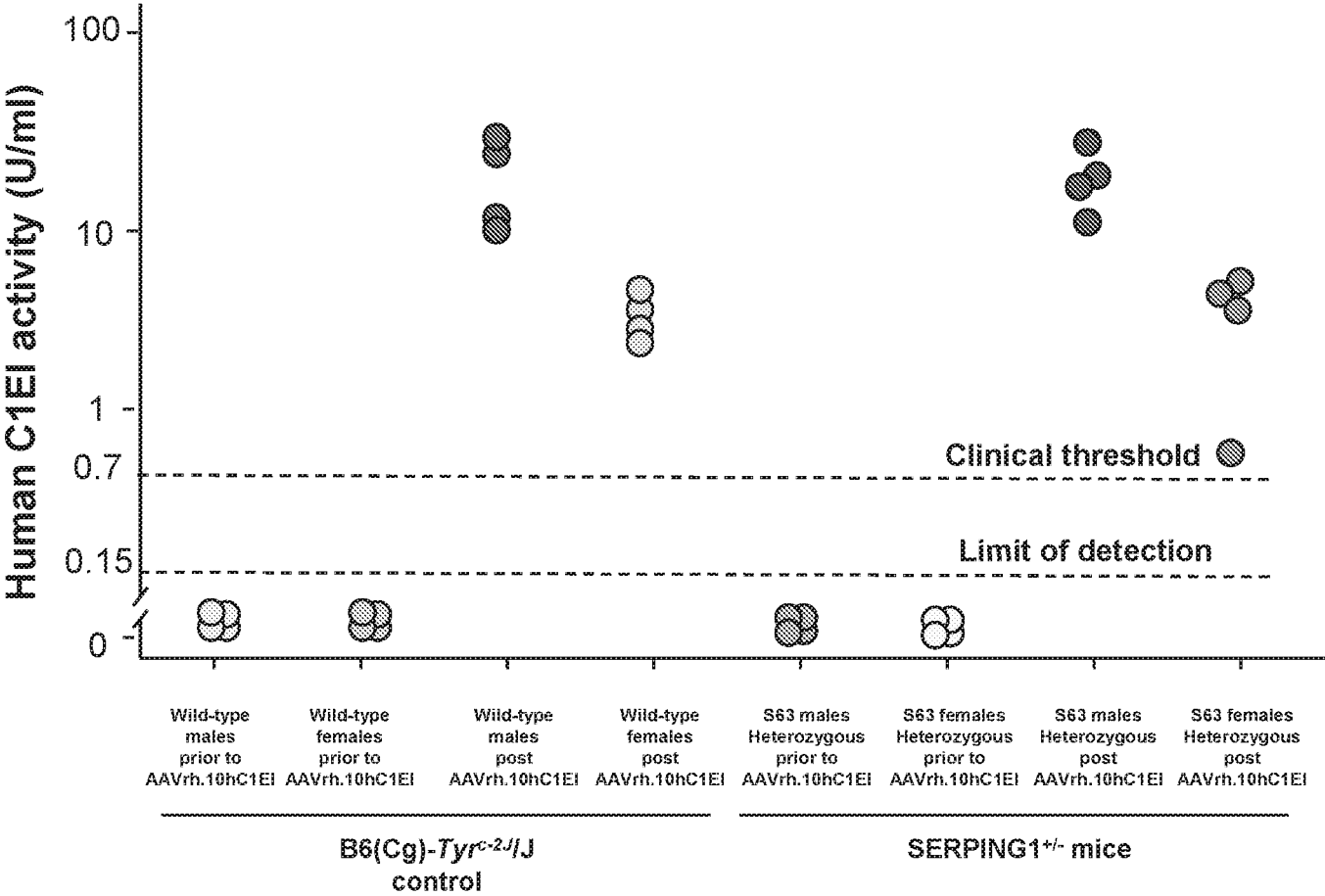
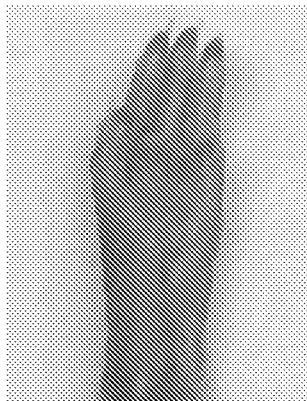
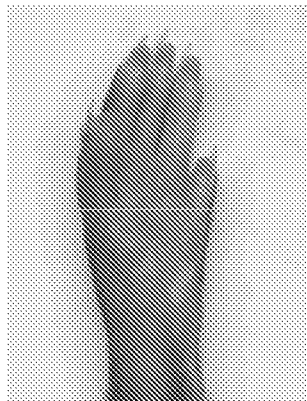


Figure 7A

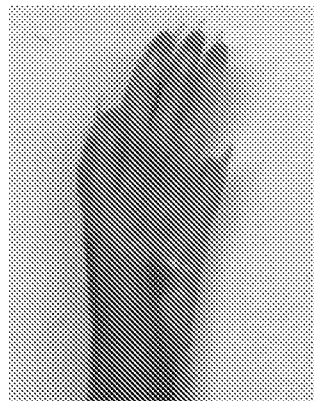
**Rear paw S63 male
no therapy**



**Rear paw S63 male
+ therapy**



**Rear paw S63 female
no therapy**



**Rear paw S63 female
+ therapy**

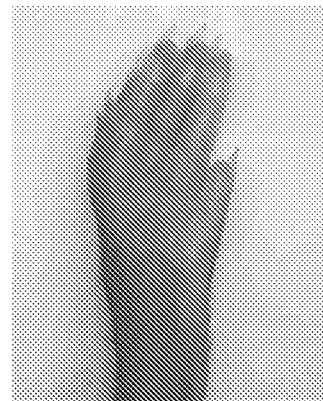
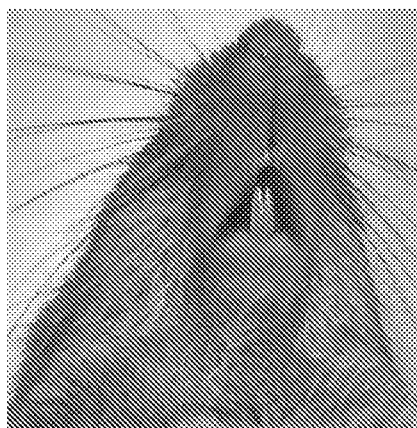
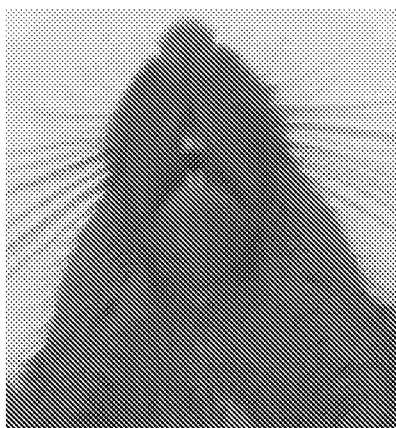


Figure 7B

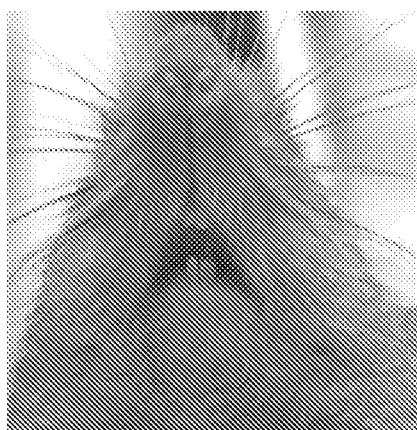
**Snout S63 male
no therapy**



**Snout S63 male
+ therapy**



**Snout S63 female
no therapy**

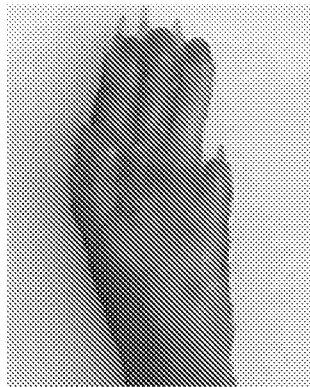


**Snout S63 female
+ therapy**

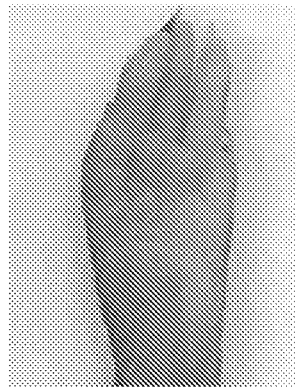


Figure 8A

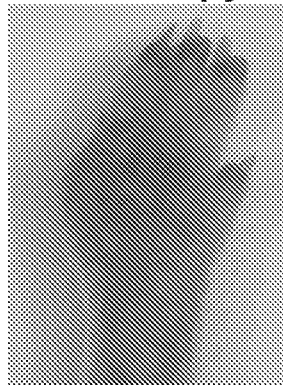
**Rear paw S63 male
no therapy**



**Rear paw S63 male
+ therapy**



**Rear paw S63 female
no therapy**



**Rear paw S63 female
+ therapy**

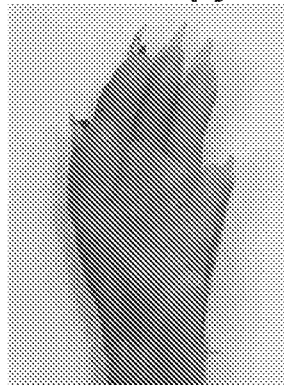
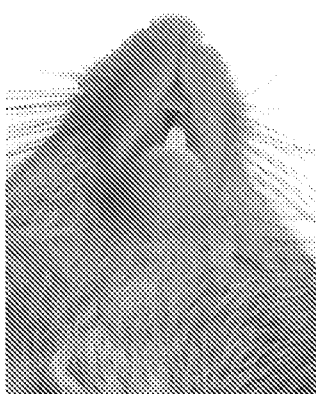
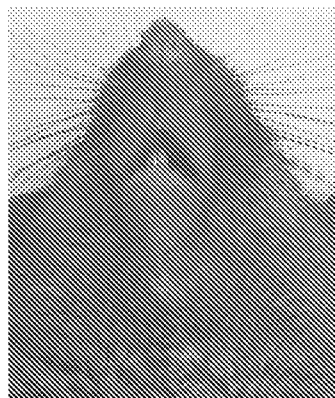


Figure 8B

**Snout S63 male
no therapy**



**Snout S63 male
+ therapy**



**Snout S63 female
no therapy**



**Snout S63 female
+ therapy**

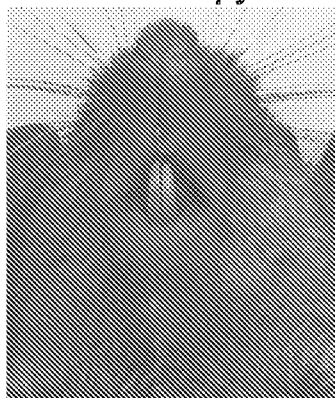


Figure 9A. Hind Paw

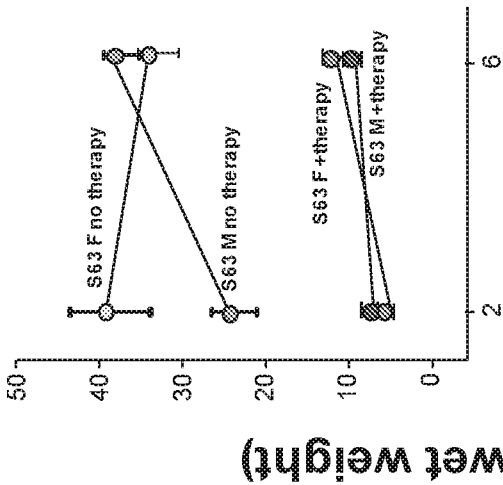


Figure 9B. Kidney

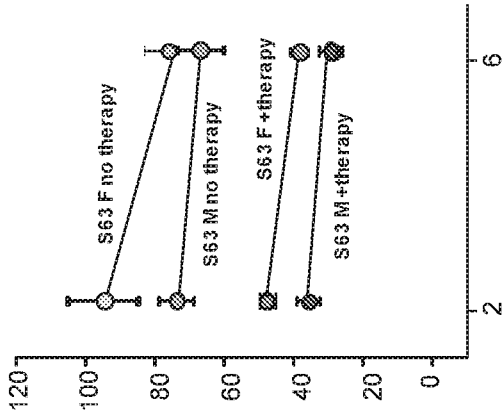


Figure 9C. Intestines

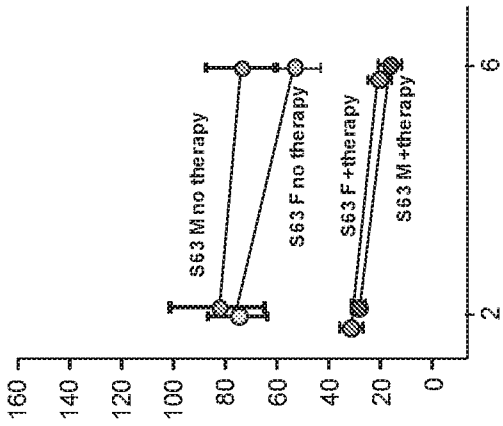


Figure 9D. Lung

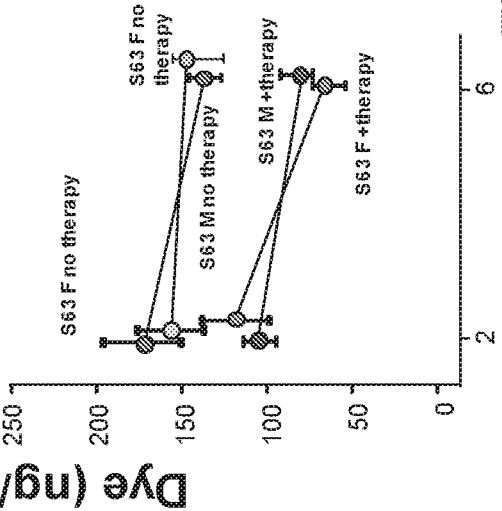


Figure 9E. Spleen

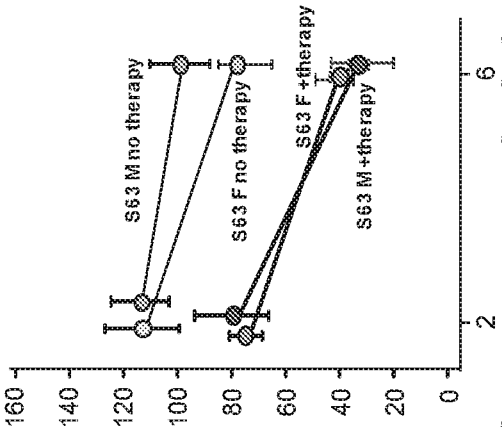
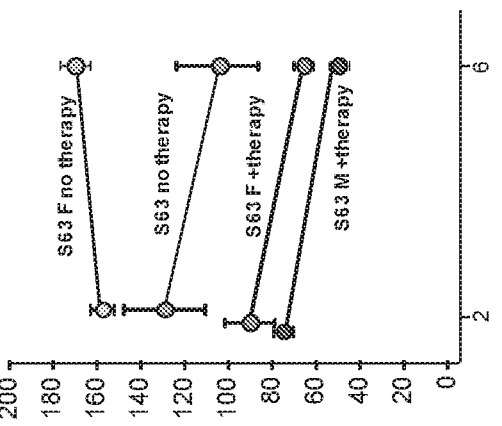


Figure 9F. Heart



Time after vector administration (wk)

Figure 10

