Figure 1A

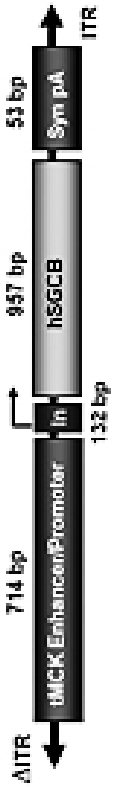


Figure 1B

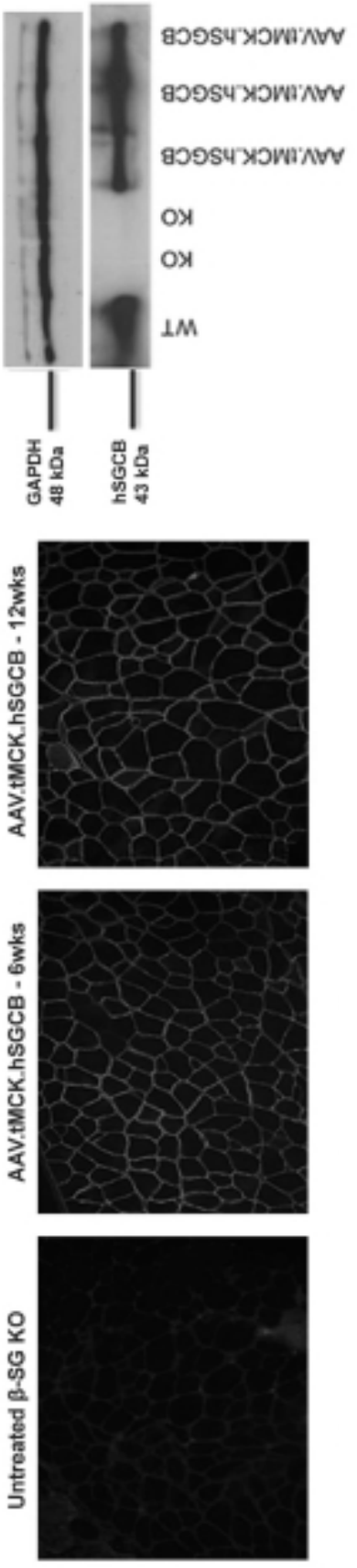


Figure 1C

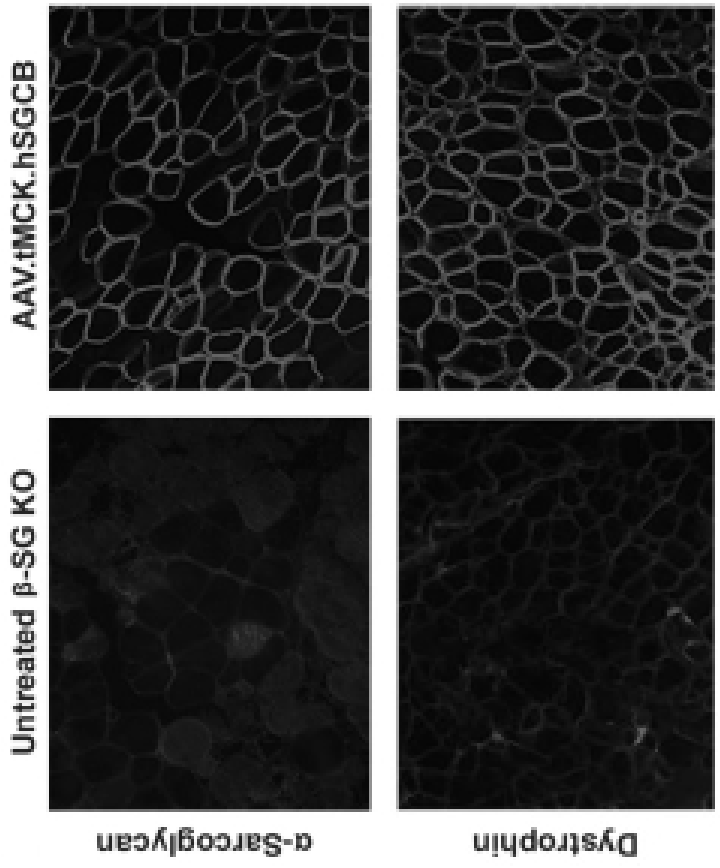


Figure 1D

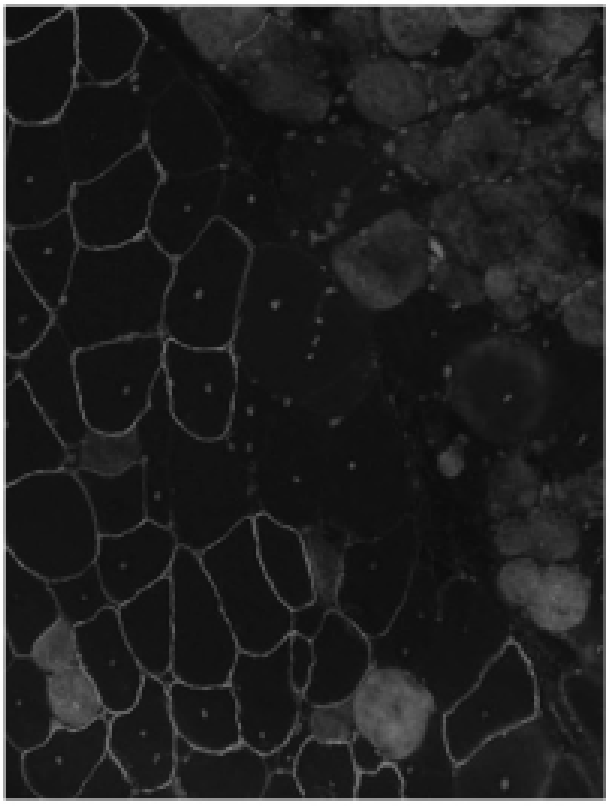


Figure 2A

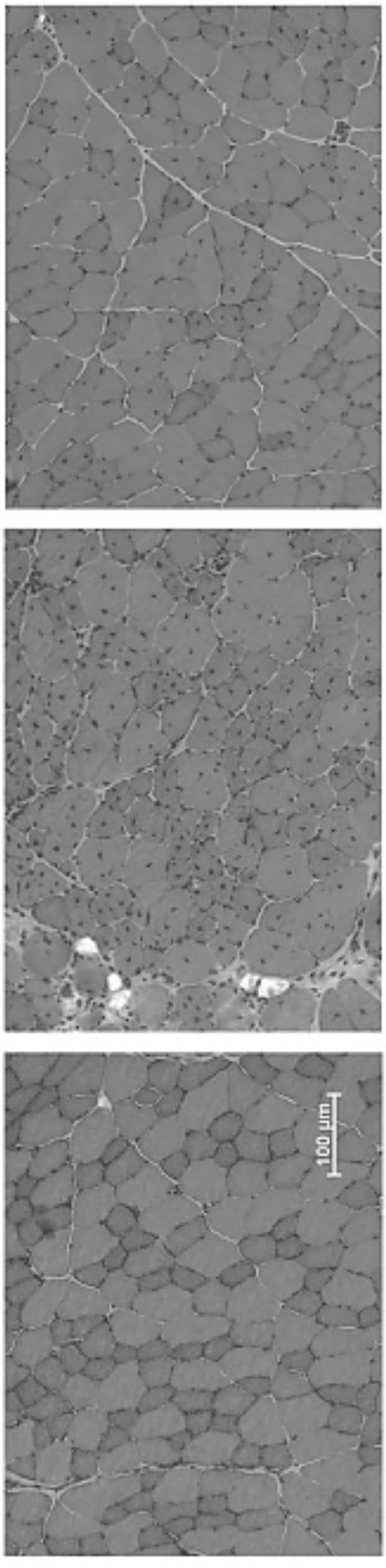


Figure 2B

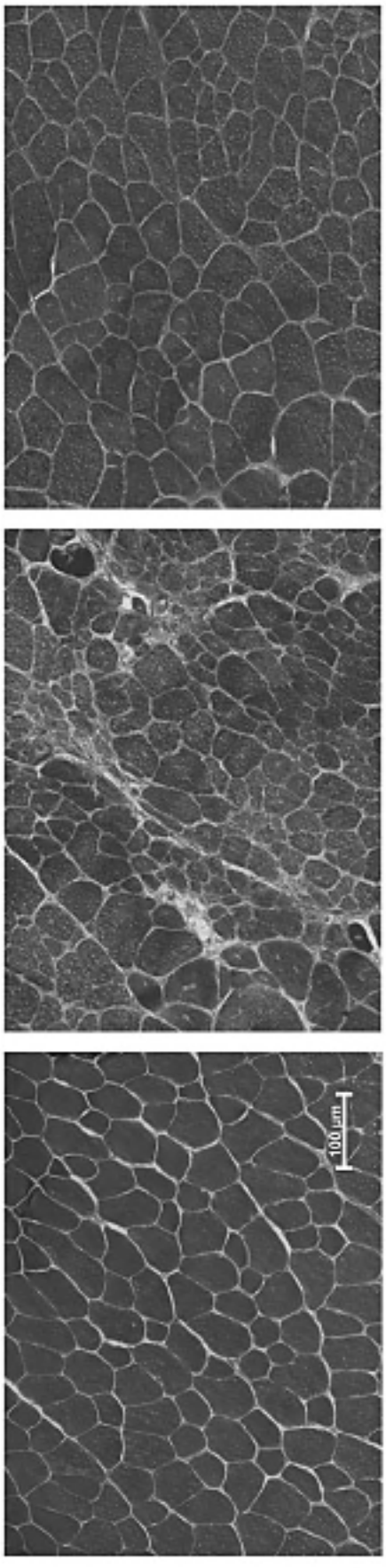


Figure 2C

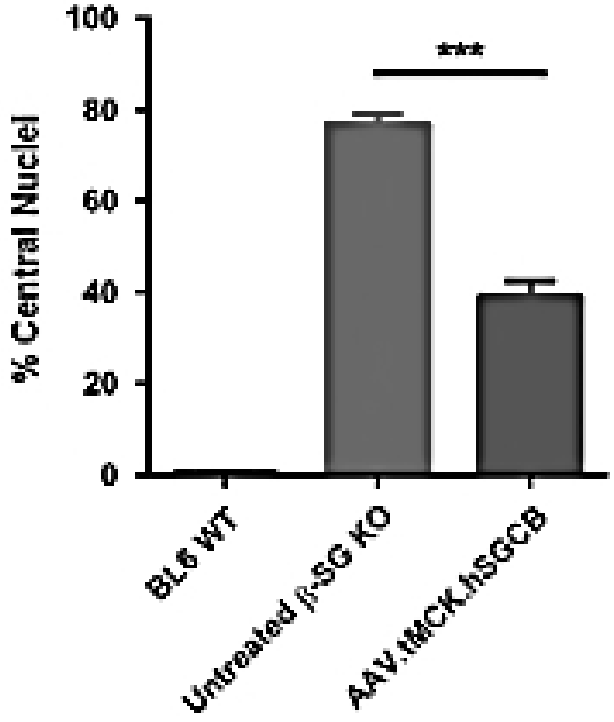


Figure 2D

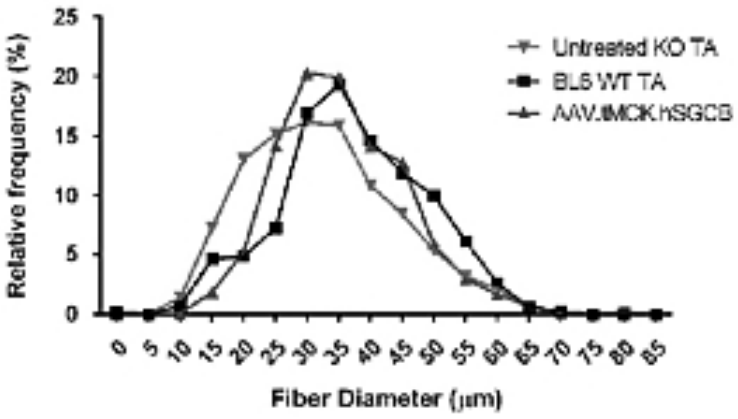


Figure 2E

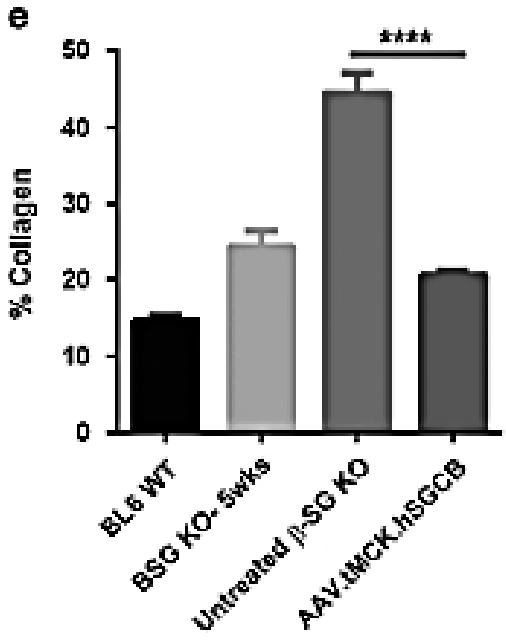


Figure 3A

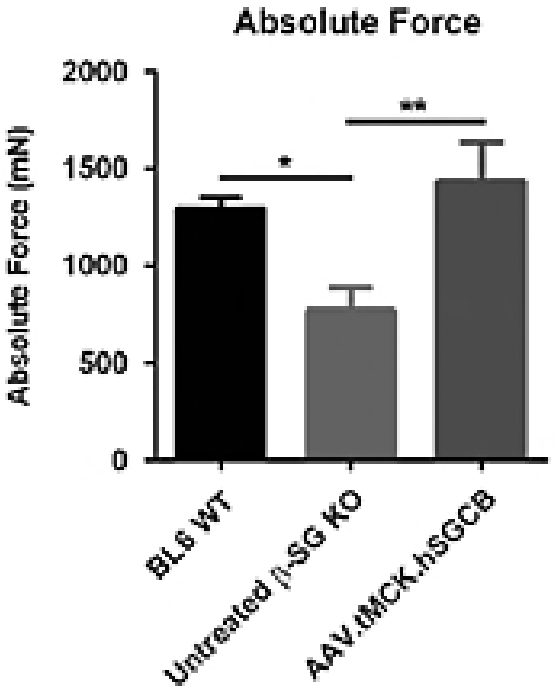


Figure 3B

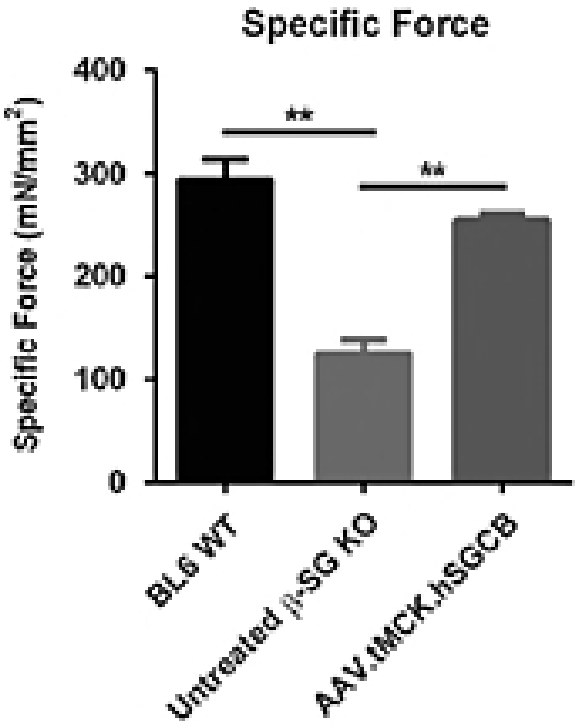


Figure 3C

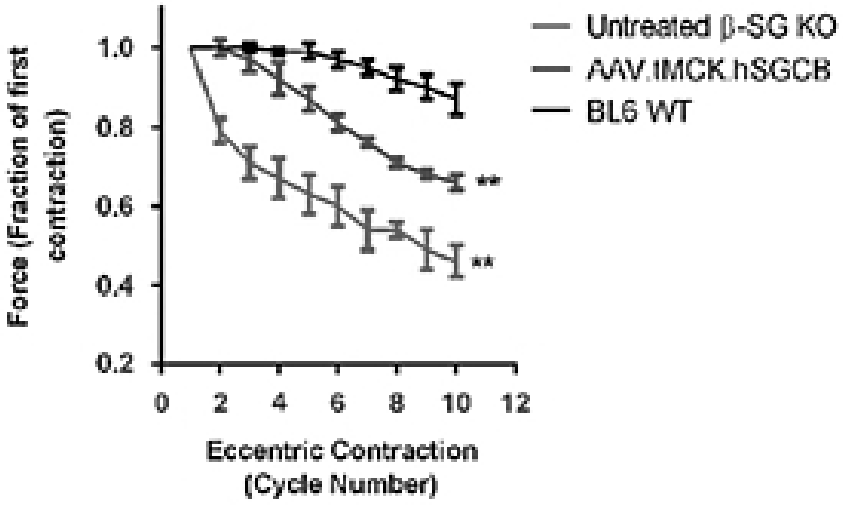


Figure 4A

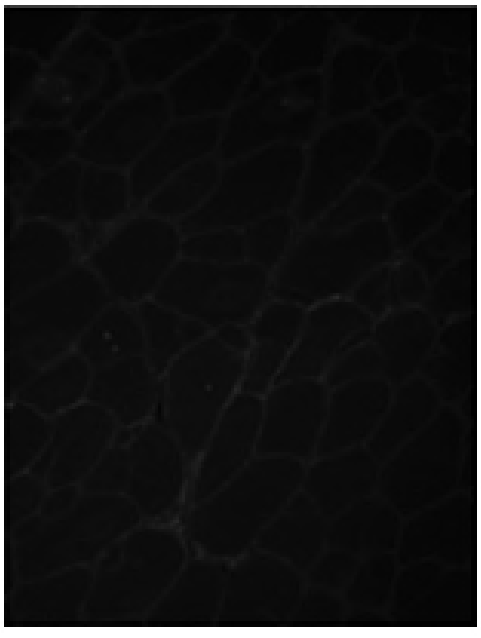


Figure 4B

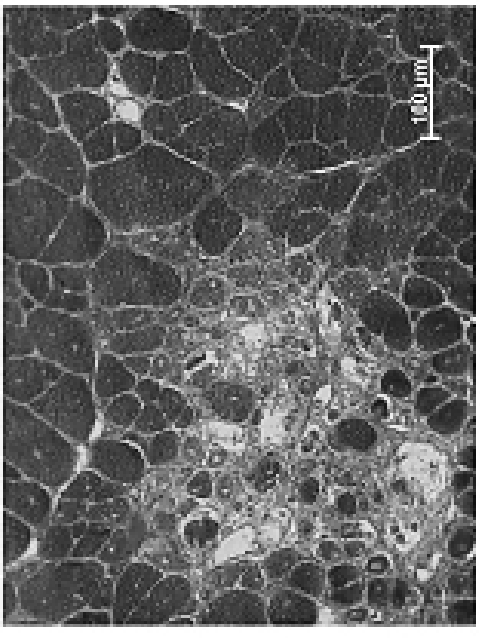


Figure 4C

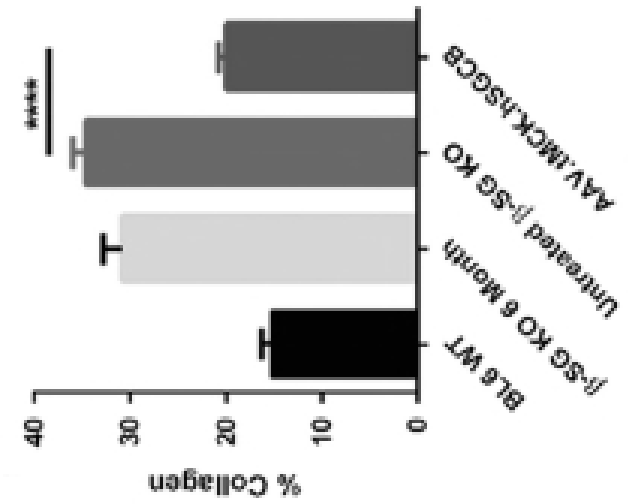
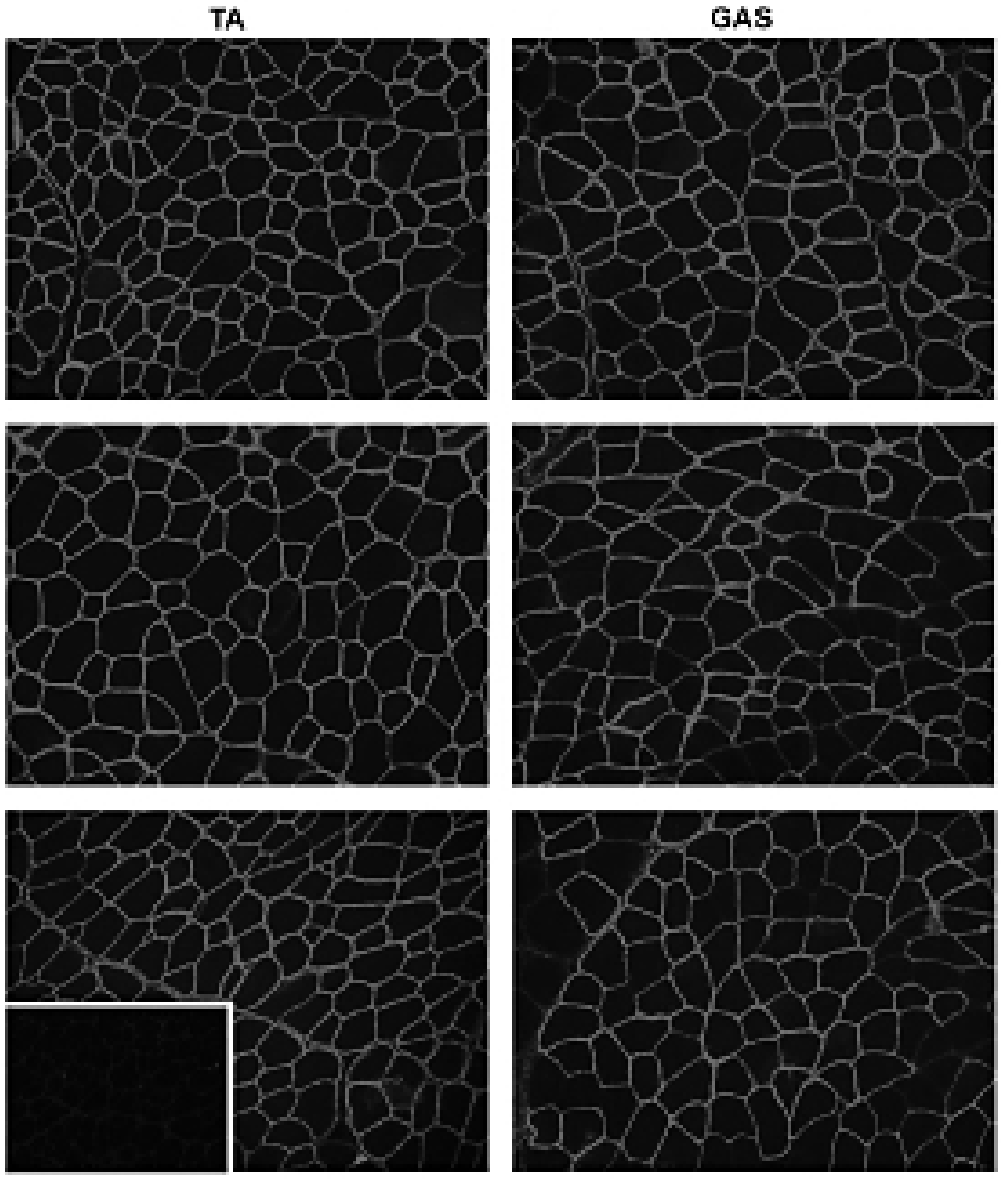


Figure 5A



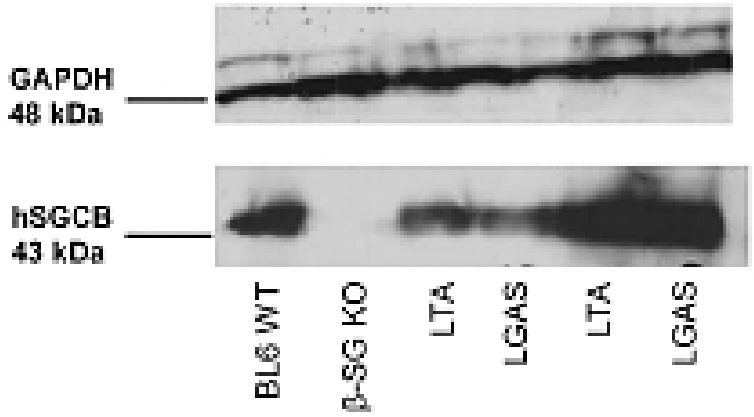


Figure 5B

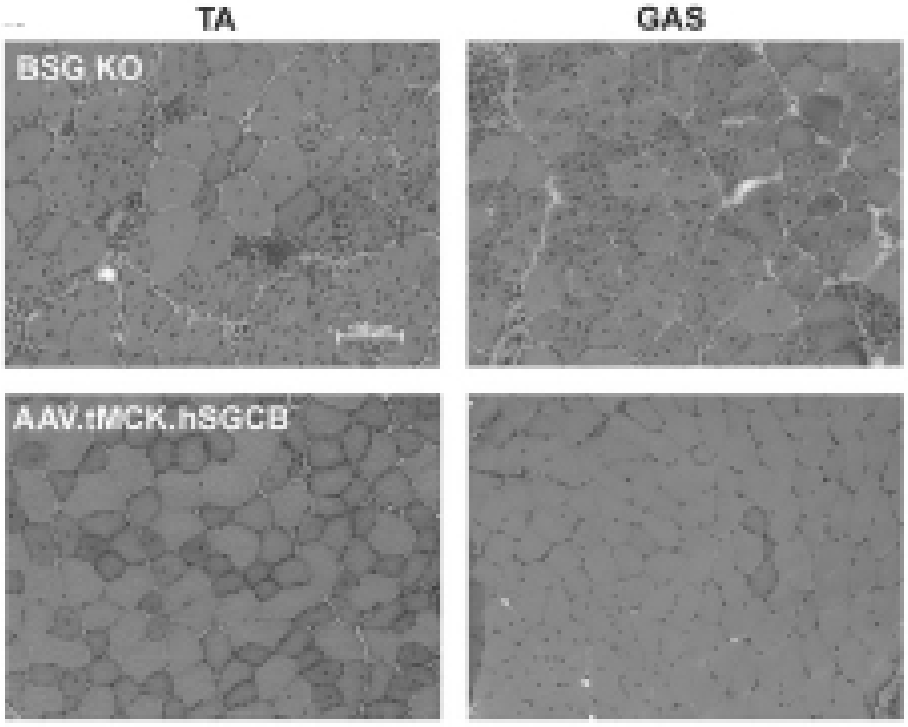


Figure 5C

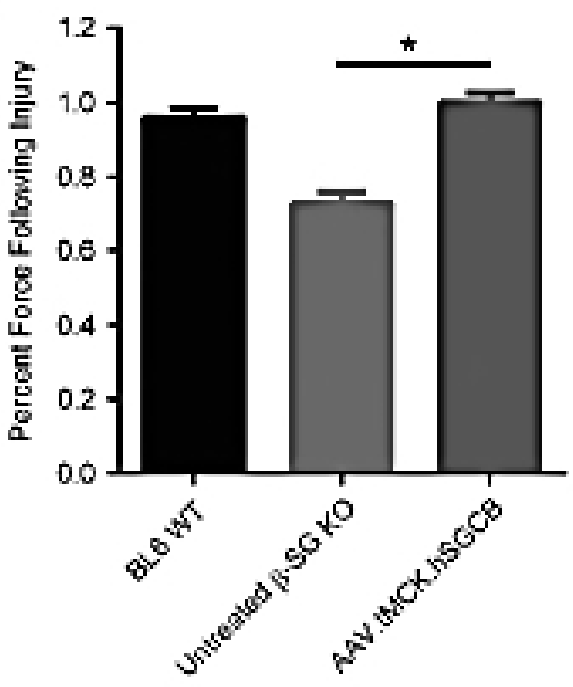


Figure 6A

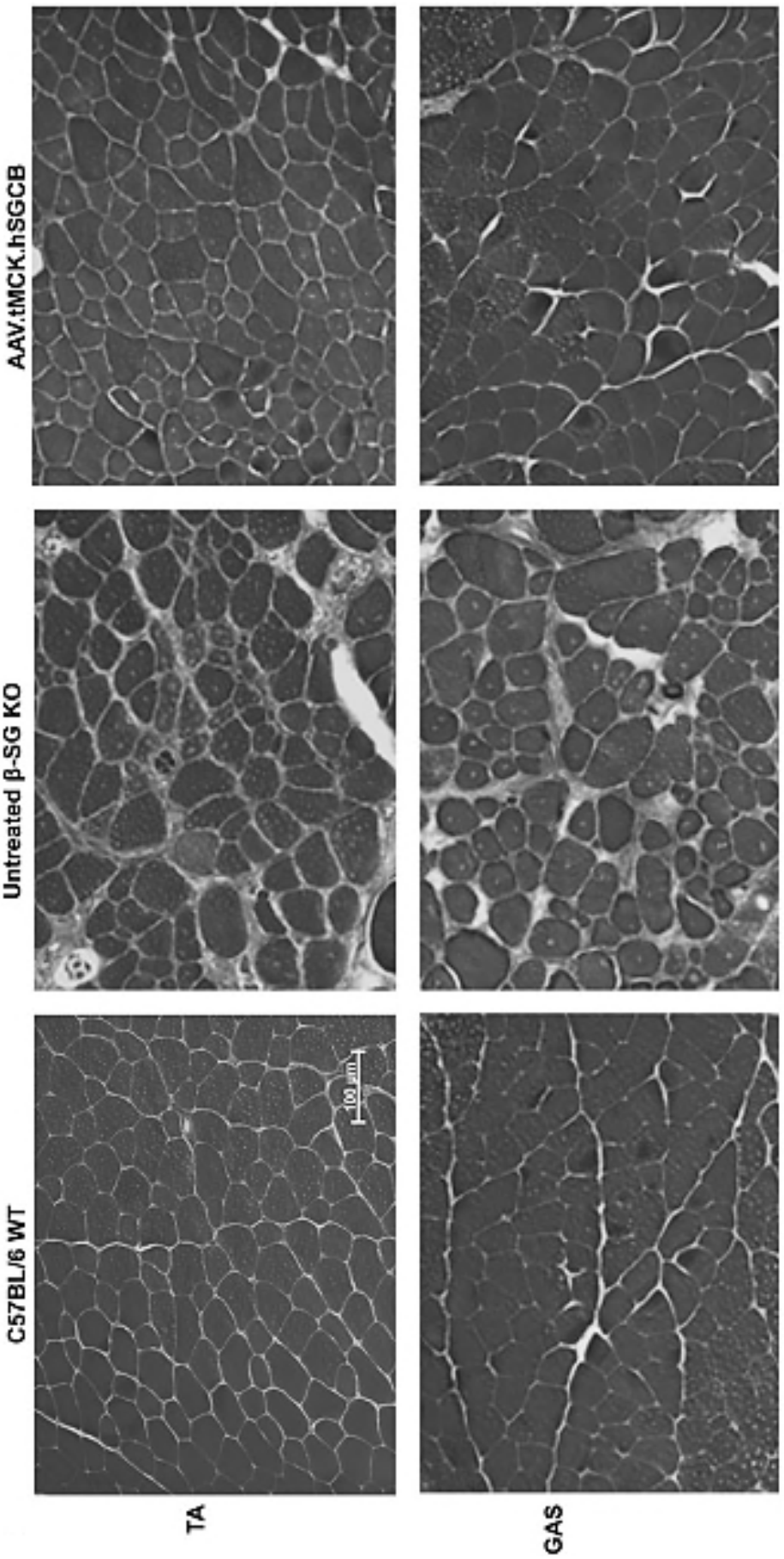


Figure 6B

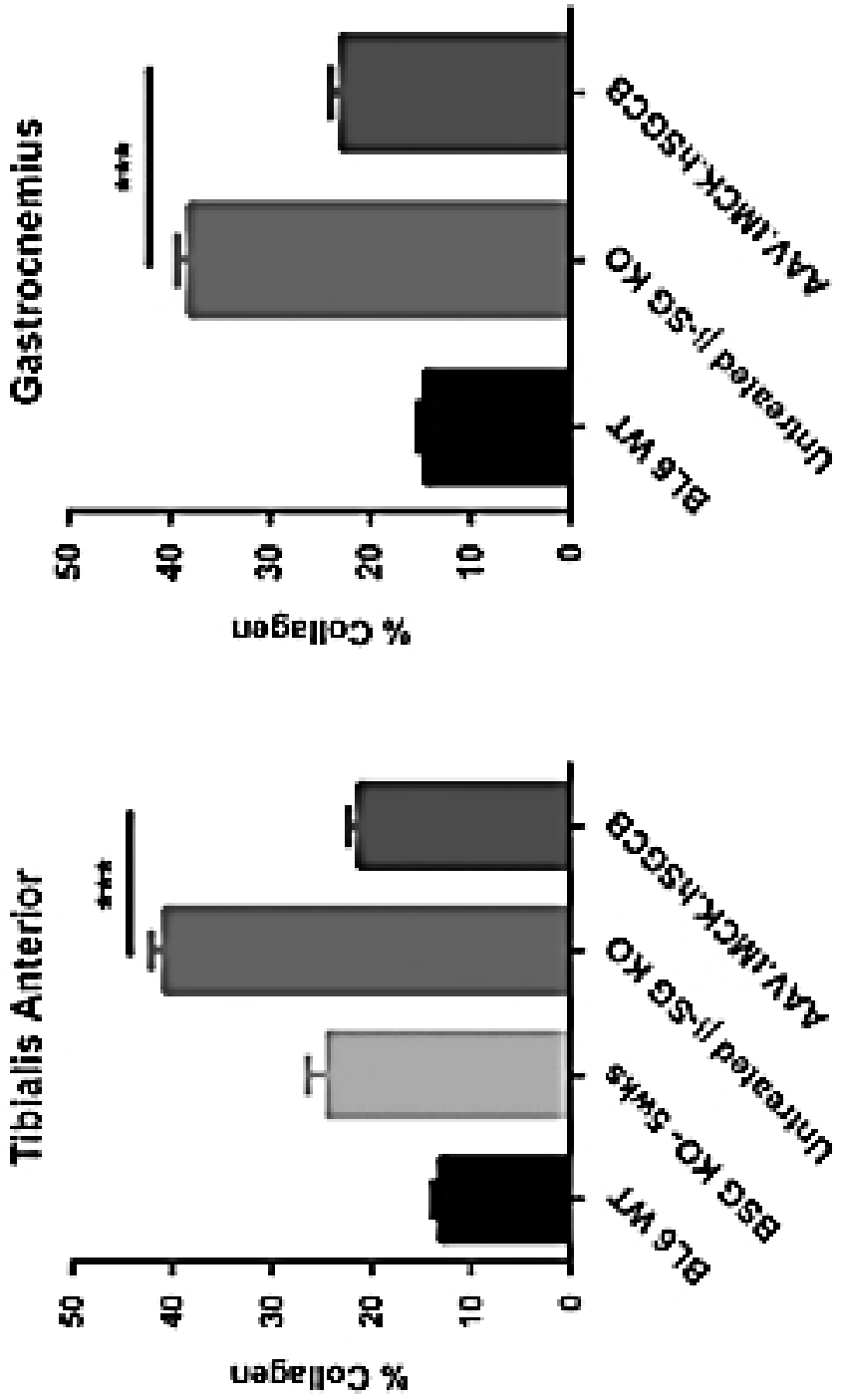

Tibialis Anterior
% Collagen
0
10
20
30
40
50
***
BL6 WT
BSG KO- 5wks
Untreated /i-SG KO
AAV.iMCK.hSGCB
Gastrocnemius
% Collagen
0
10
20
30
40
50
***
BL6 WT
Untreated /i-SG KO
AAV.iMCK.hSGCB

Figure 7B

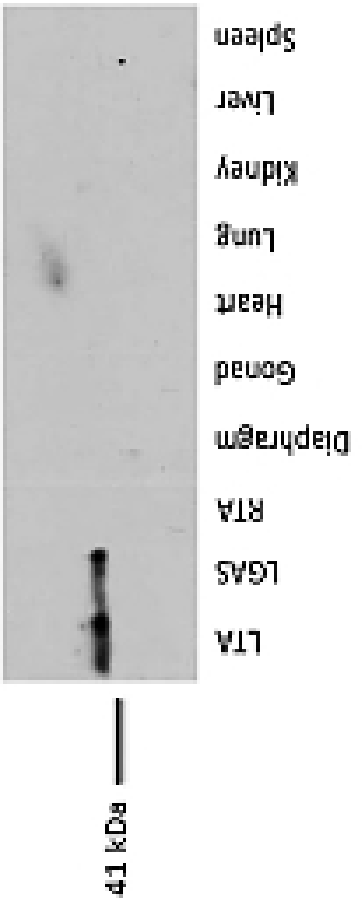


Figure 7A

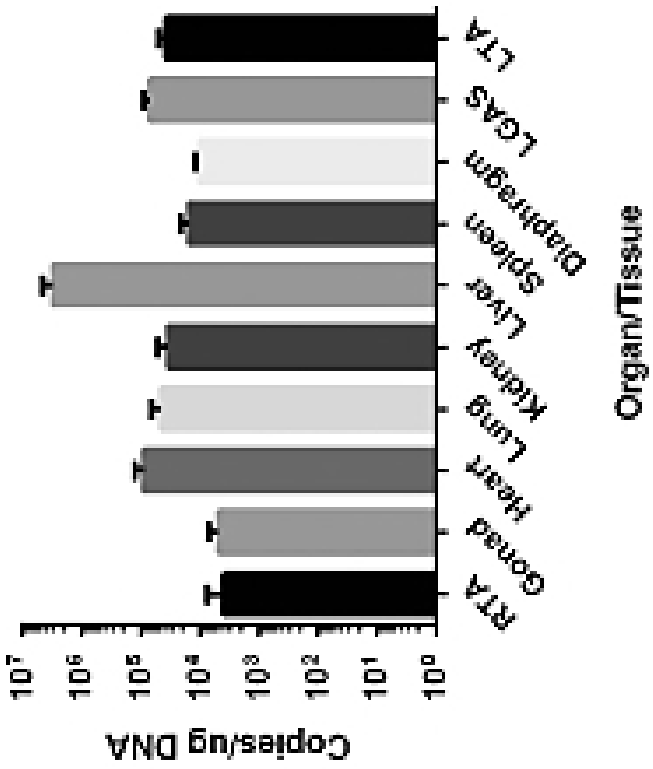


Figure 8A

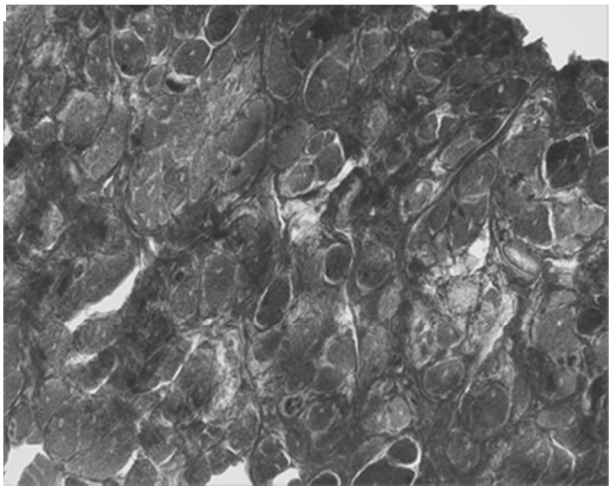


Figure 8B

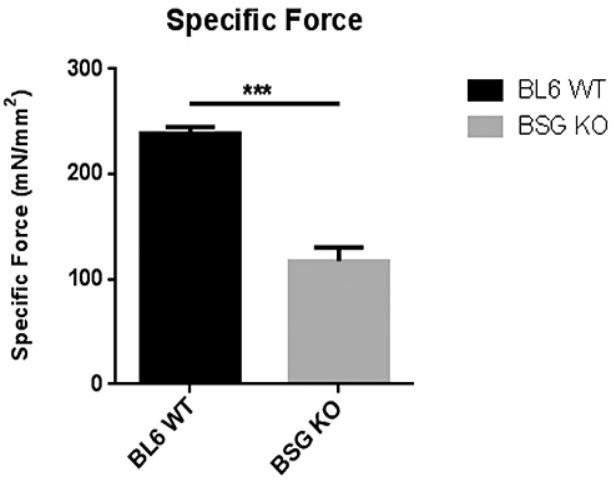


Figure 8C

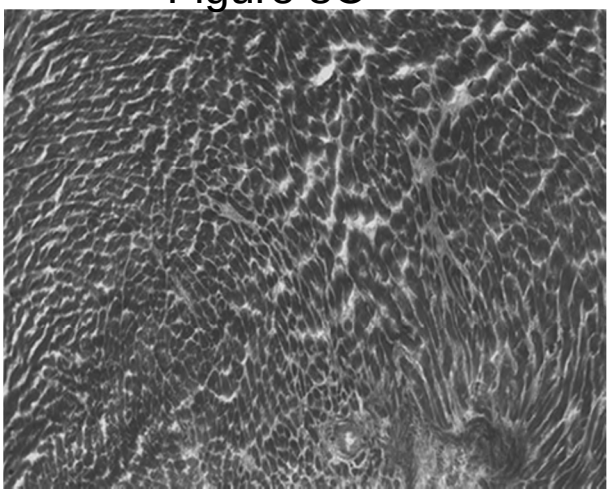


Figure 8D

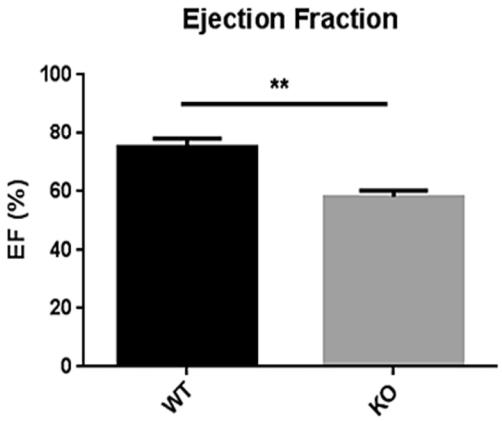


Figure 9



scAAV.MHCK7.hSGCB cassette

Figure 10

Delivery of scAAVrh.74.MHCK7.hSGCB via the tail vein of SGCB-/- mice results in nearly 100% transduction of muscle fibers in every muscle

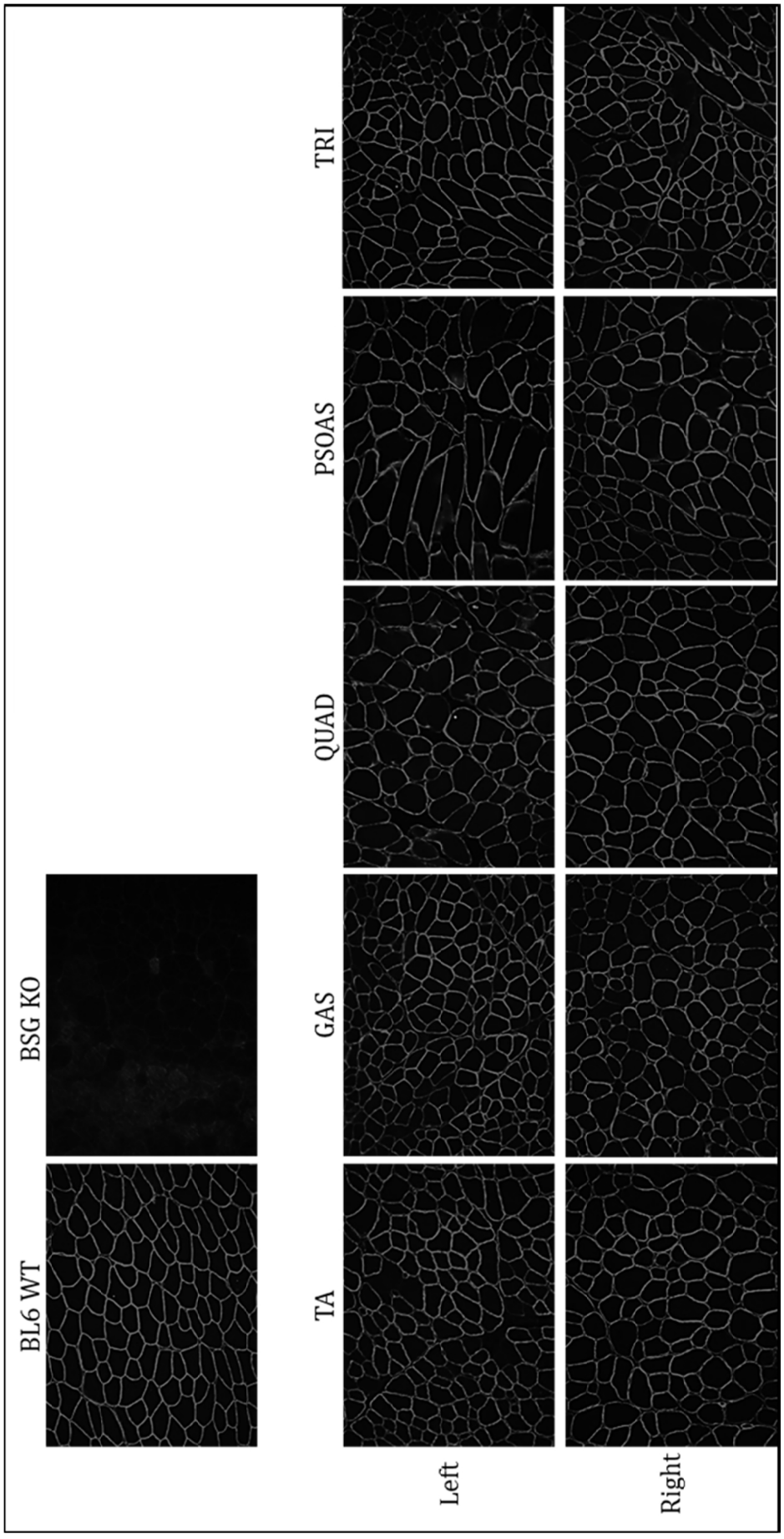


Figure 11

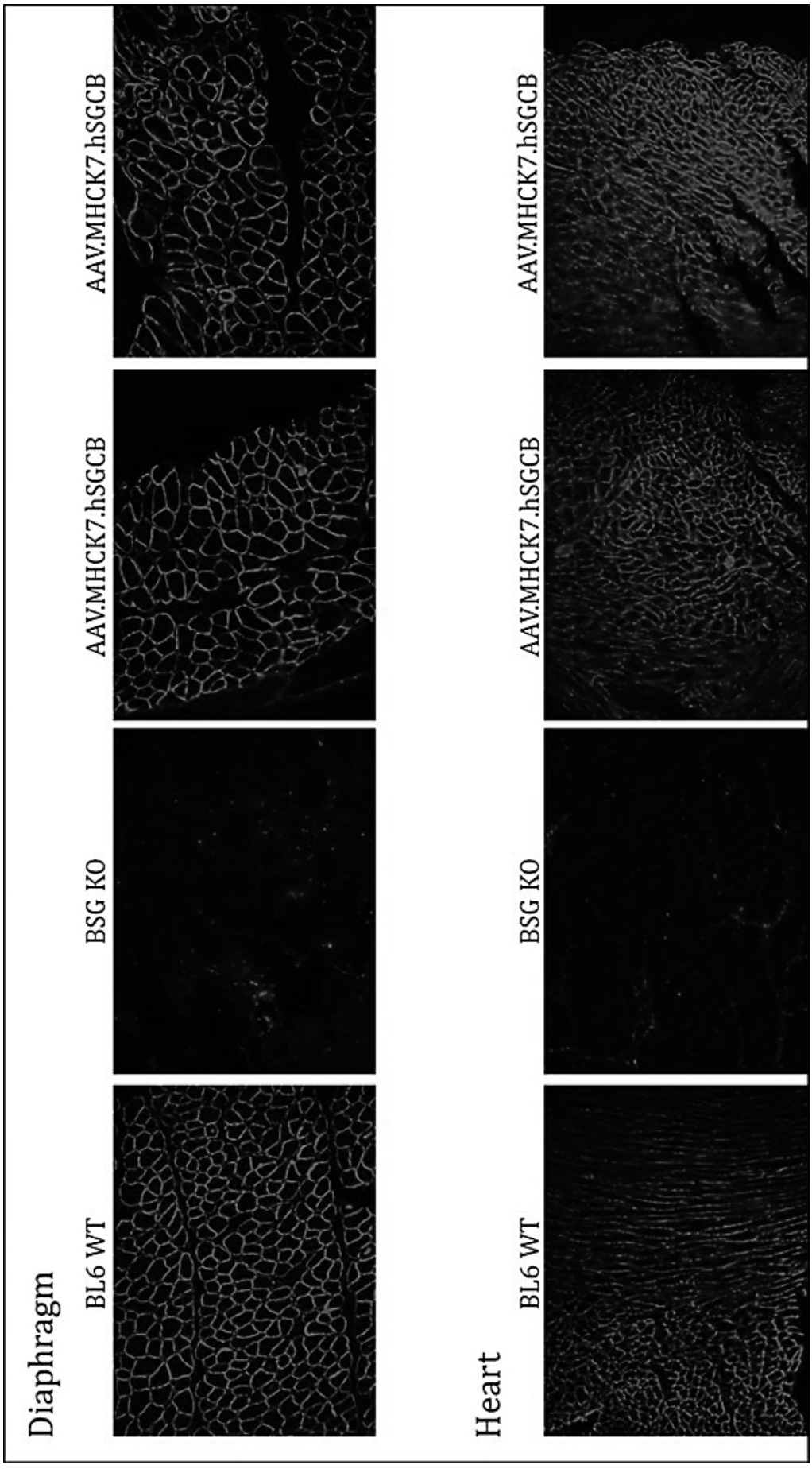

Diaphragm
BL6 WT
BSG KO
AAV.MHCK7.hSGCB
AAV.MHCK7.hSGCB
Heart
BL6 WT
BSG KO
AAV.MHCK7.hSGCB
AAV.MHCK7.hSGCB

Figure 12A



Figure 12B

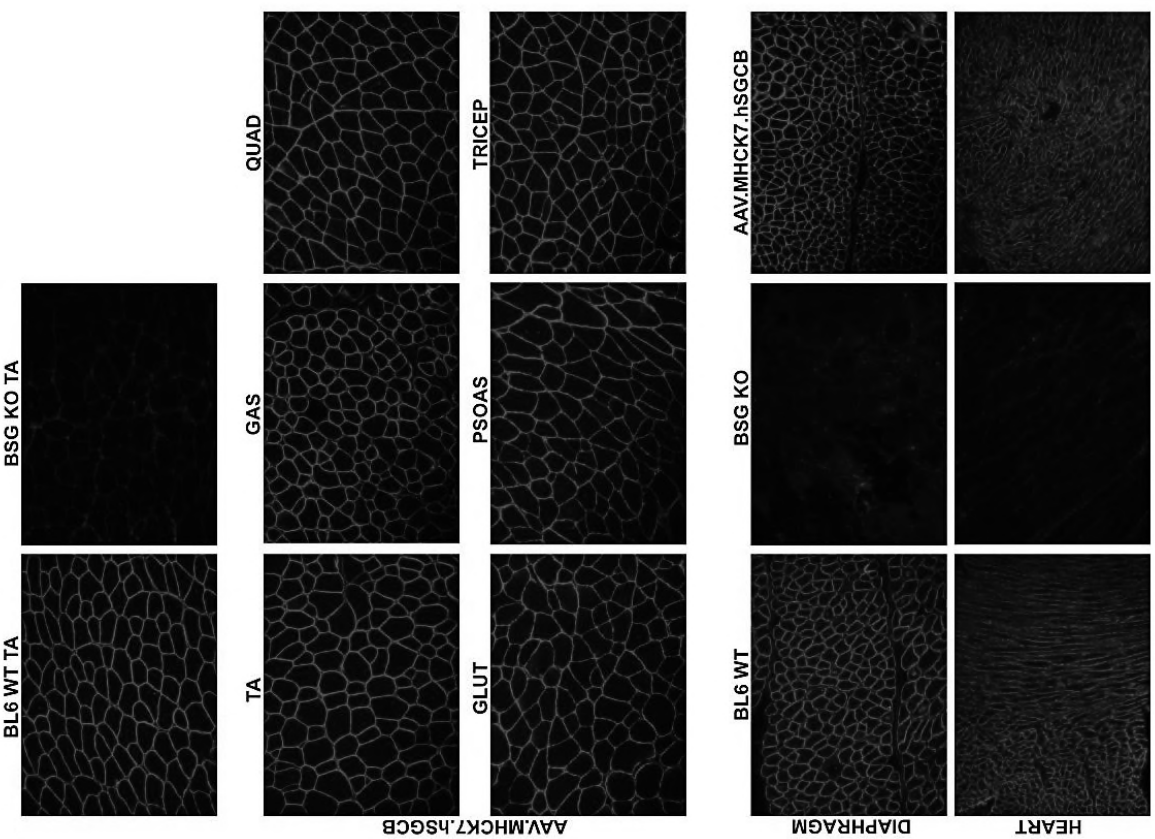


Figure 12C

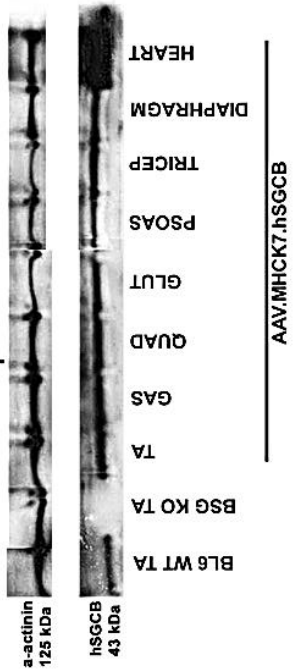


Figure 12D

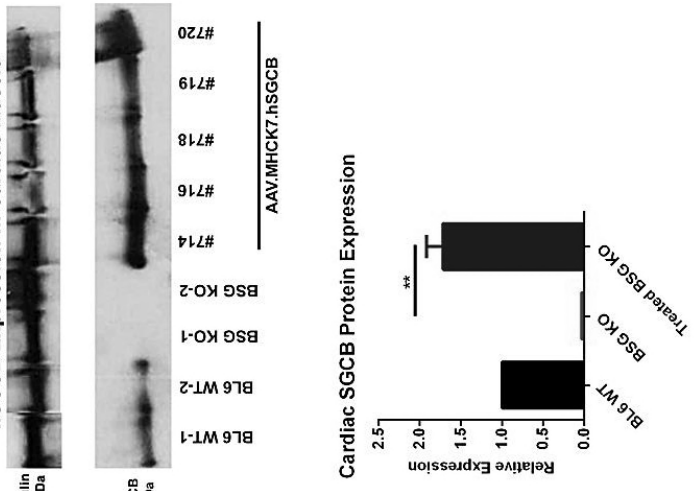


Figure 13A

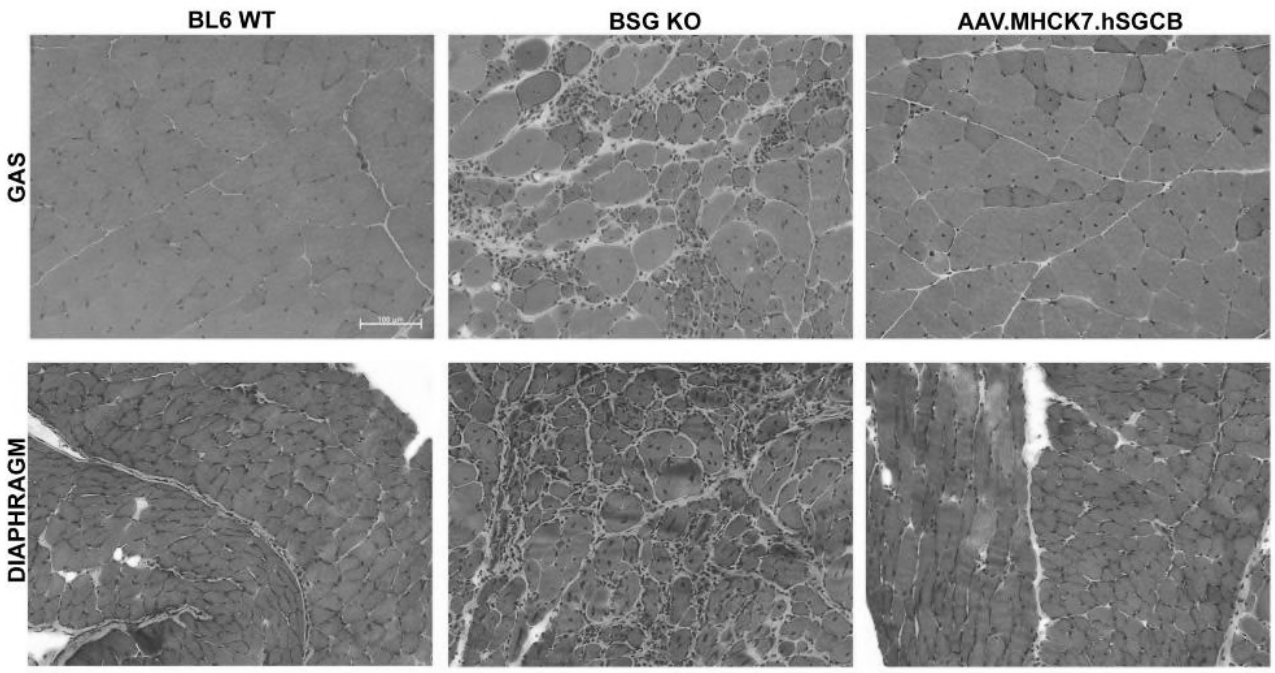


Figure 13B

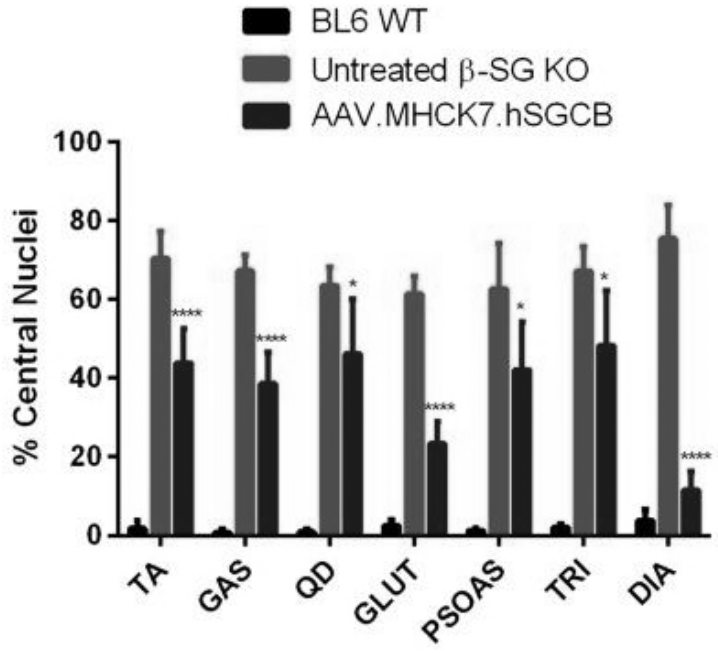


Figure 13C

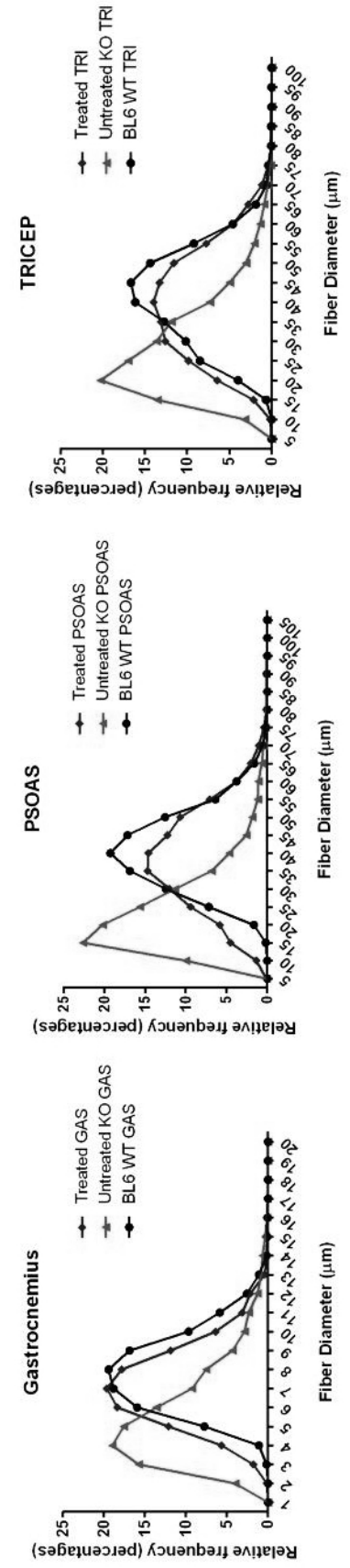


Figure 13D

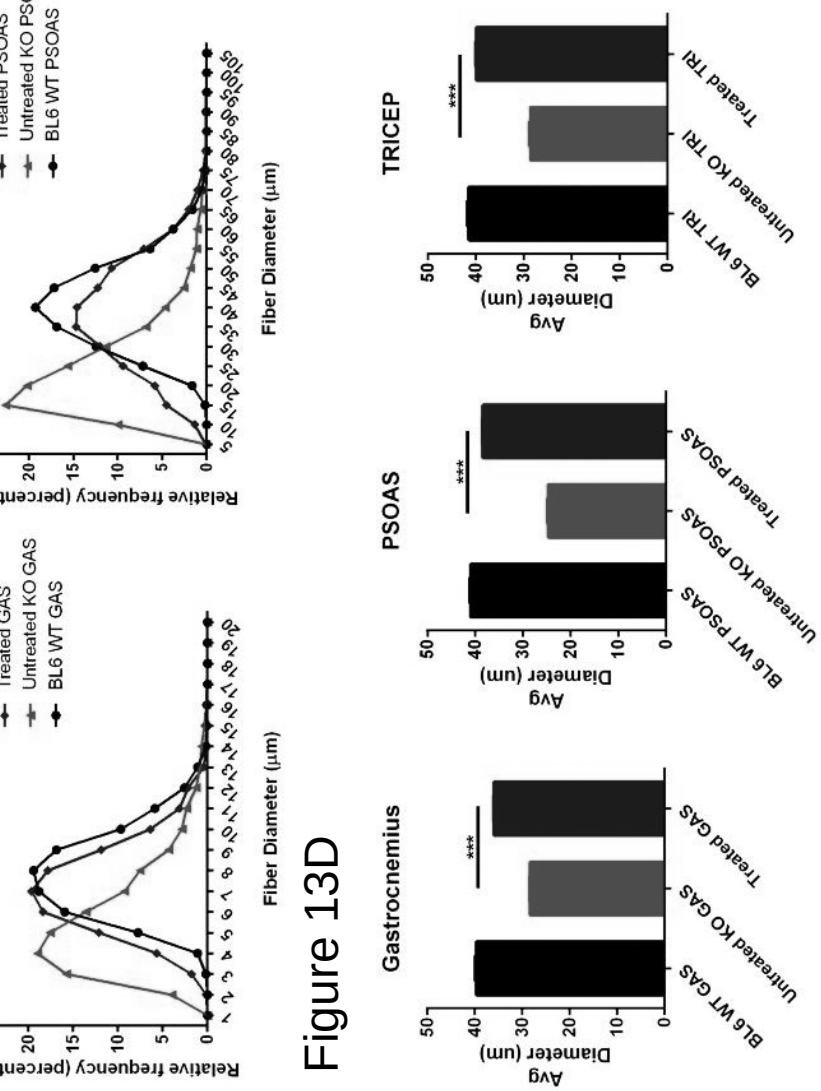


Figure 14A

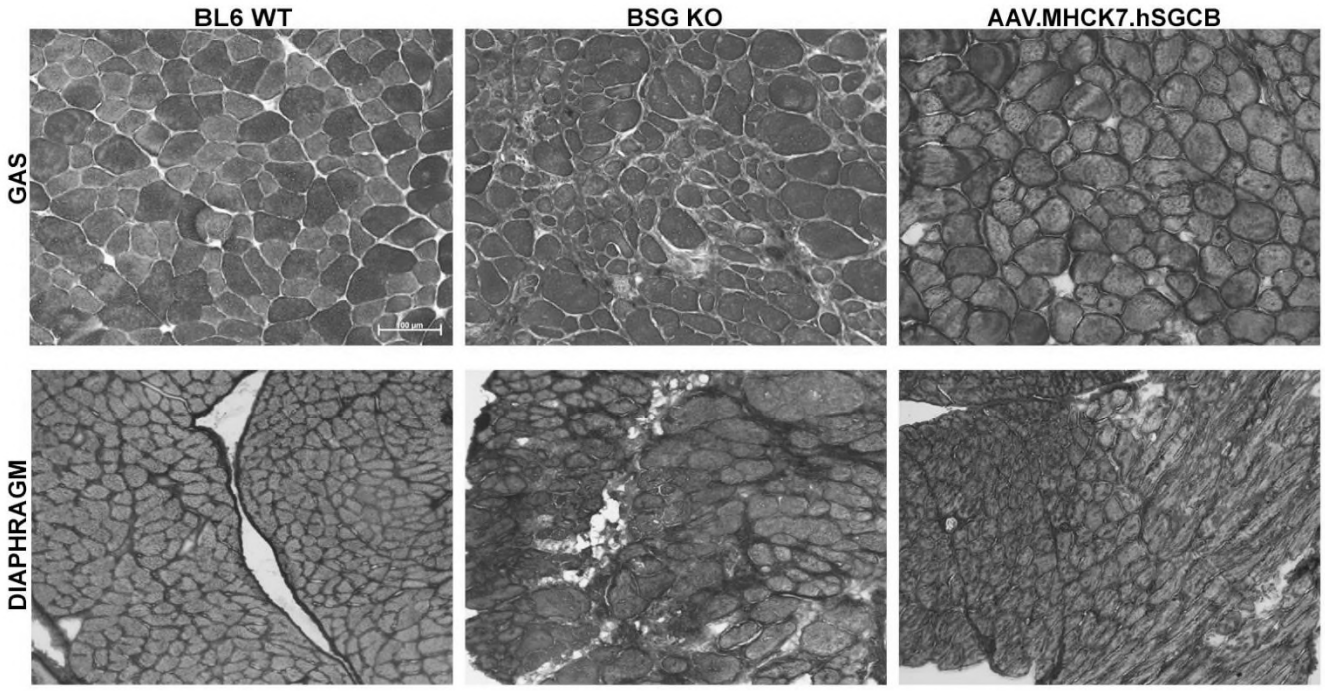


Figure 14B

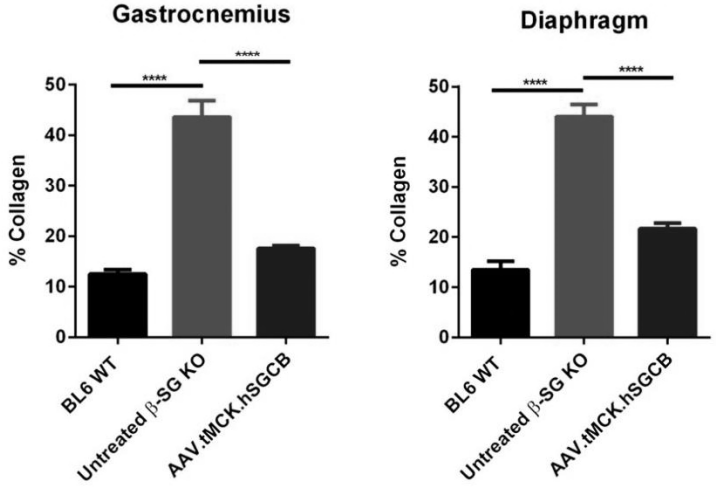


Figure 15

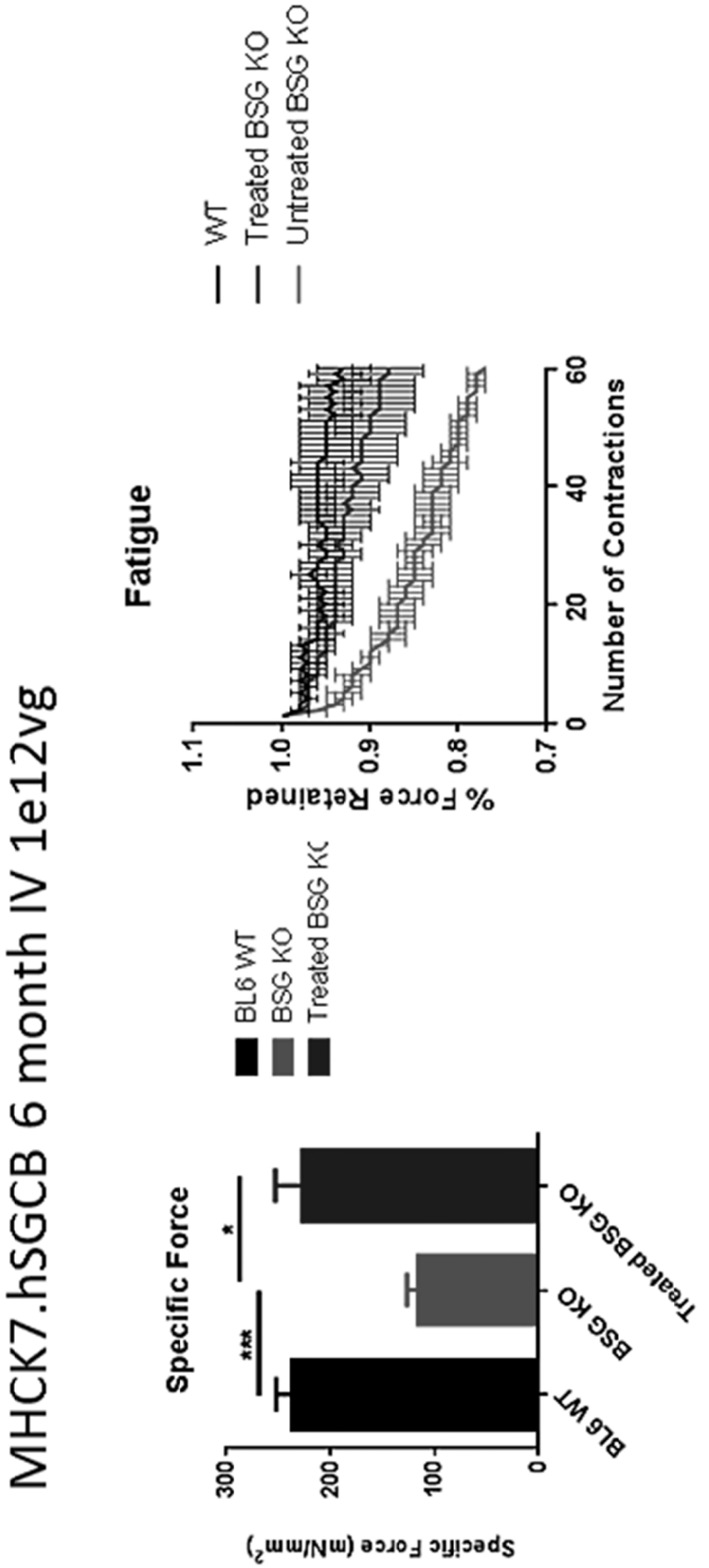
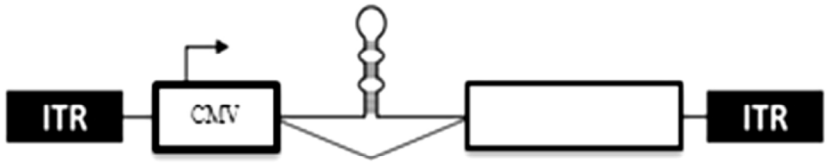

MHCK7.hSGCB 6 month IV 1e12vg
Specific Force
Specific Force (mN/mm²)
300
200
100
0
***
*
BL6 WT
BSG KO
Treated BSG KO
BL6 WT
BSG KO
Treated BSG KO
Fatigue
% Force Retained
1.1
1.0
0.9
0.8
0.7
0
20
40
60
Number of Contractions
WT
Treated BSG KO
Untreated BSG KO

Figure 16

scAAVrh.74.CMV.miR29c



SEQ ID NO: 9: miR-29C IN A miR-30 BACKBONE

GGCCGGCC***tgtttgaatgaggcttcagtactttacagaat***CGTTGCCTGCACATCTTGGAAACACTTGCTGGGATTAC
TTCTTCAGGTTAACCCAACAGAAGGCTCGAGAAGGTATATTGCTGTTGACAGTGAGCGCAACC
GATTTCAAATGGTGCTAGAGTGAAGCCACAGATGTCTAGCACCATTTGAAATCGGTTATGCCTA
CTGCCTCGGAATTCAAGGGGCTACTTTAGGAGCAATTATCTTGTTTACTAAAACTGAATACCTT
GCTATCTCTTTGATACATTGGCCGGCC

Figure 17

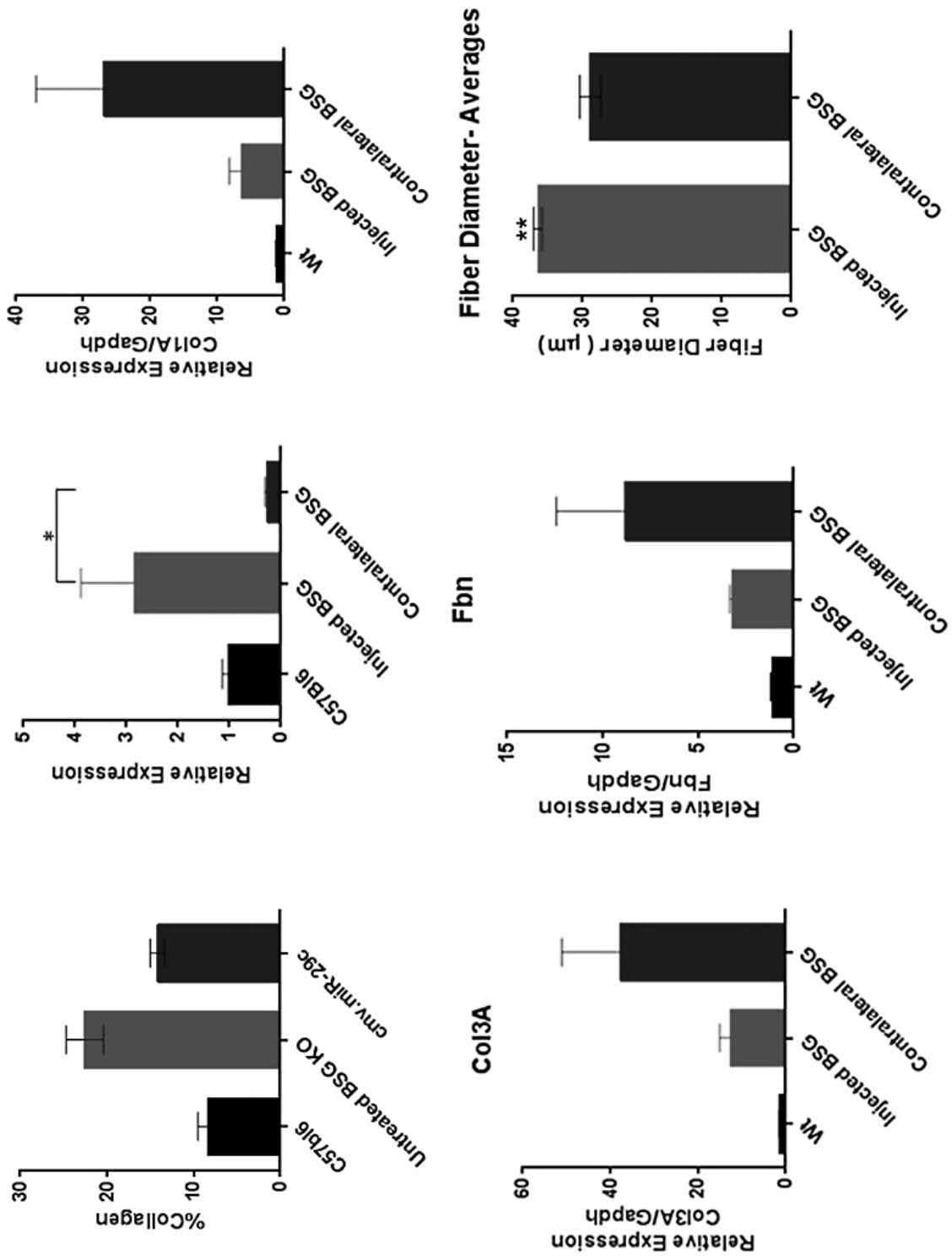
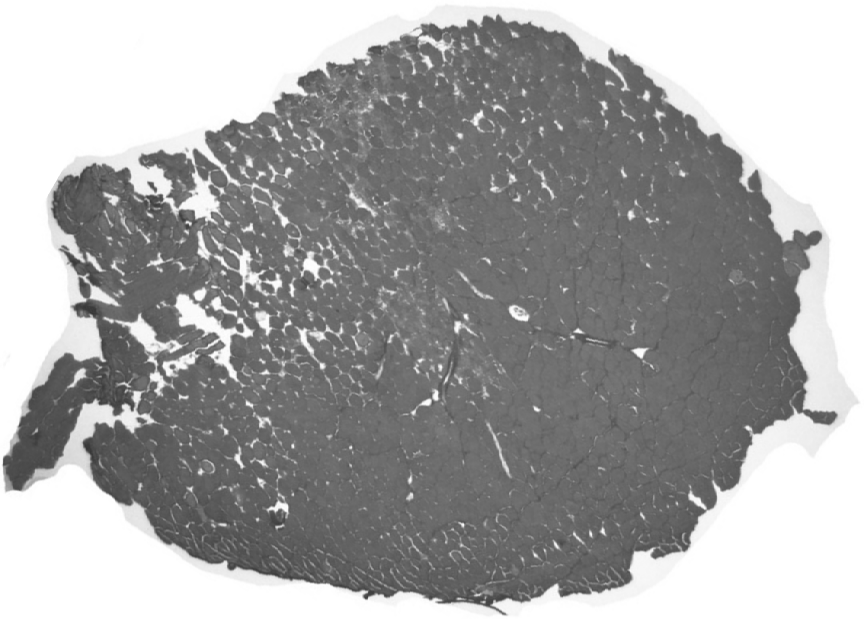

%Collagen
C57bl6
Untreated BSG KO
cmv.miR-29c
Mir levels
Relative Expression
C57Bl6
Injected BSG
Contralateral BSG
*
Col1A
Relative Expression
Col1A/Gapdh
Wt
Injected BSG
Contralateral BSG
Fiber Diameter- Averages
Fiber Diameter (µm)
Injected BSG
Contralateral BSG
**
Col3A
Relative Expression
Col3A/Gapdh
Wt
Injected BSG
Contralateral BSG
Fbn
Relative Expression
Fbn/Gapdh
Wt
Injected BSG
Contralateral BSG

Figure 18



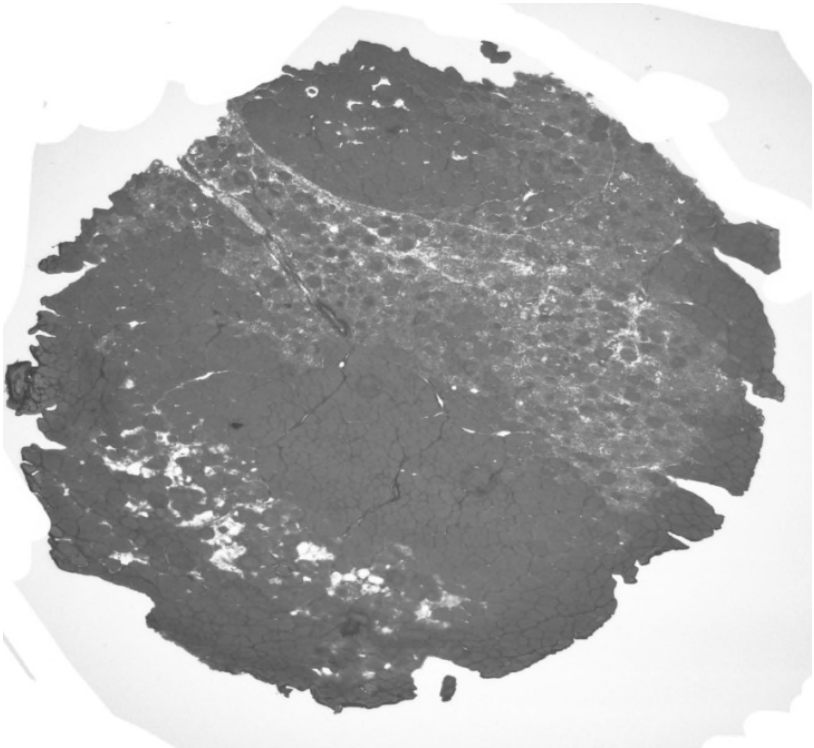


Figure 19A

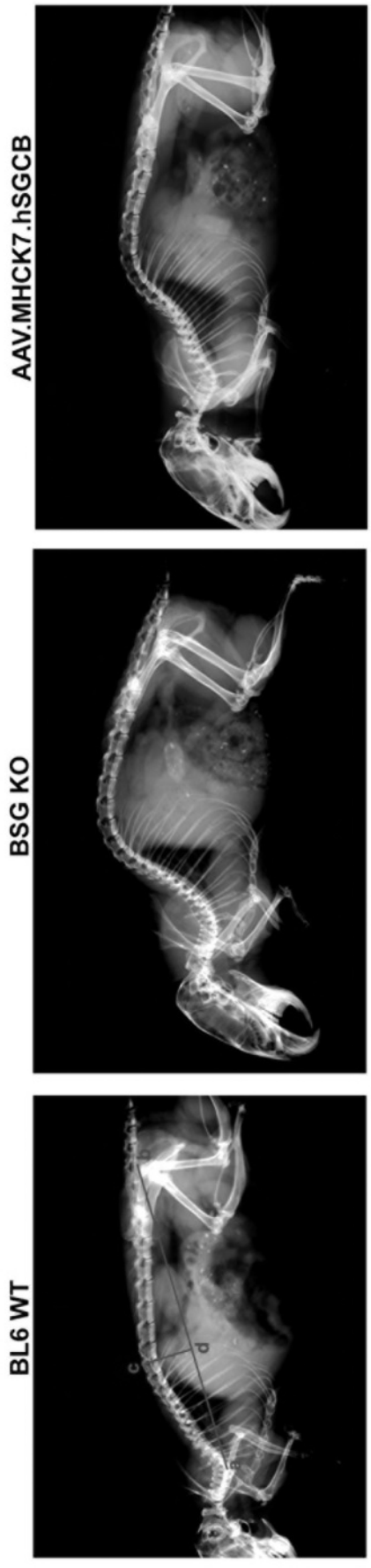


Figure 19B

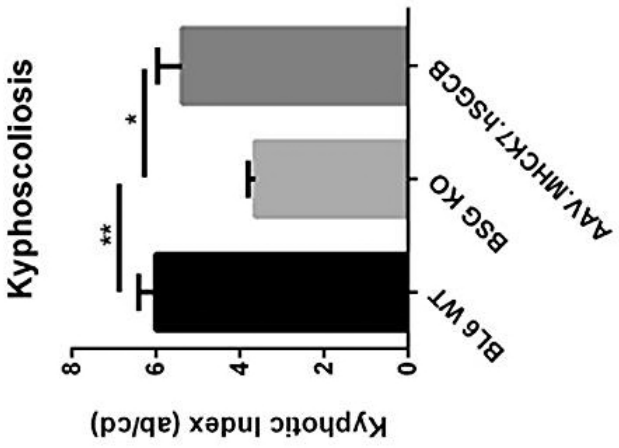


Figure 20A

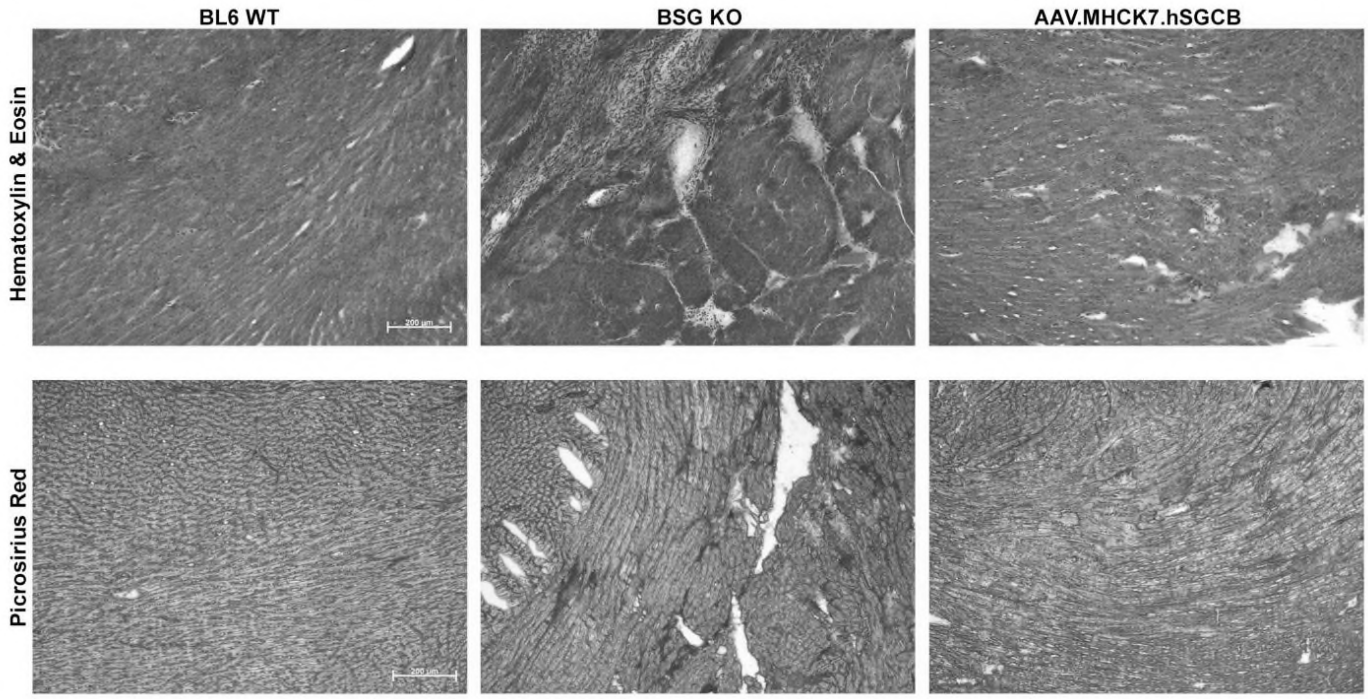


Figure 20B

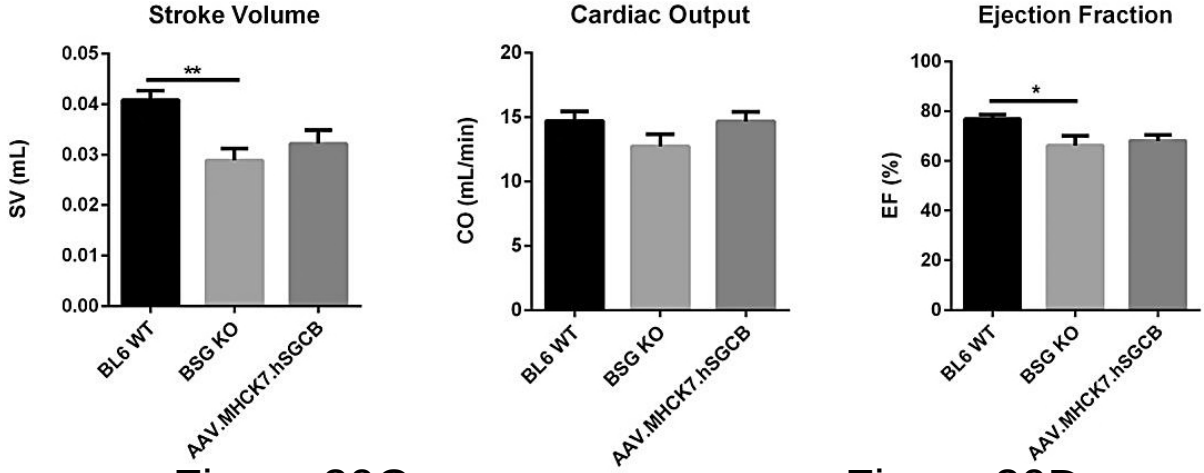


Figure 20C

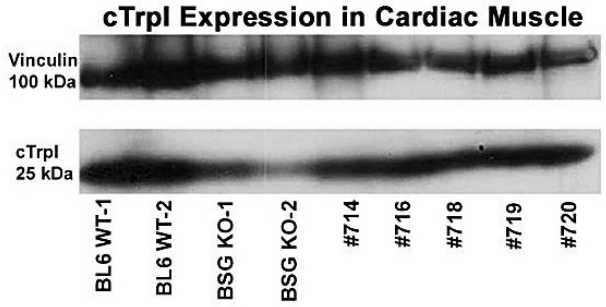


Figure 20D

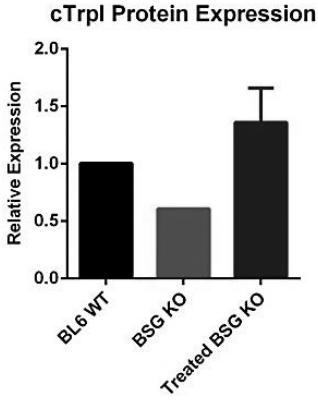


Figure 21A

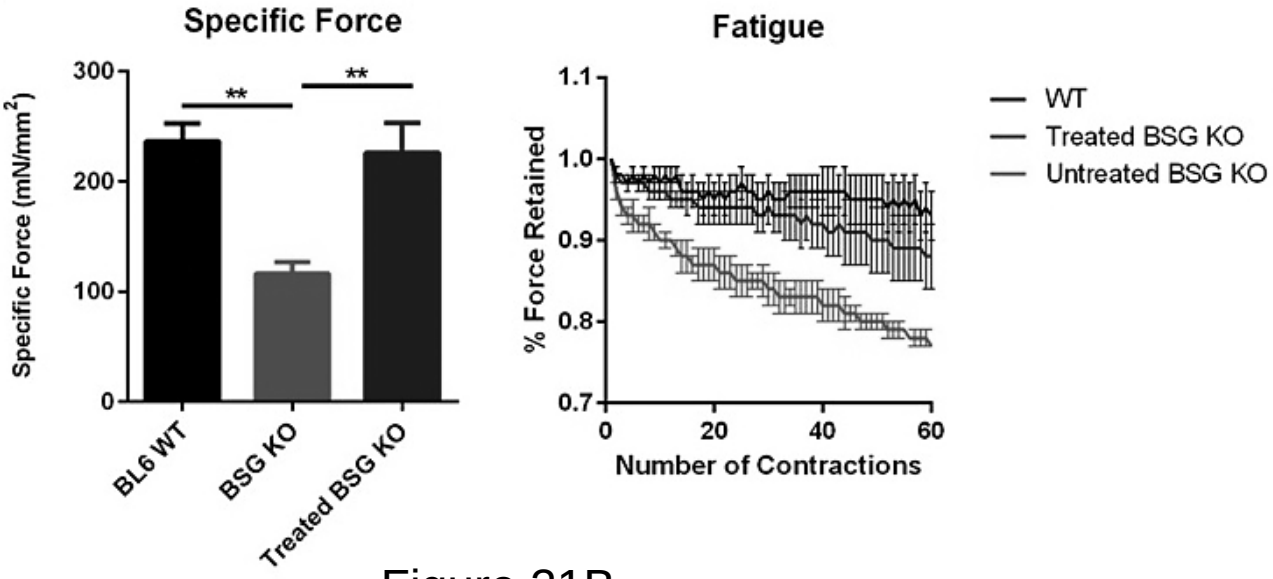


Figure 21B

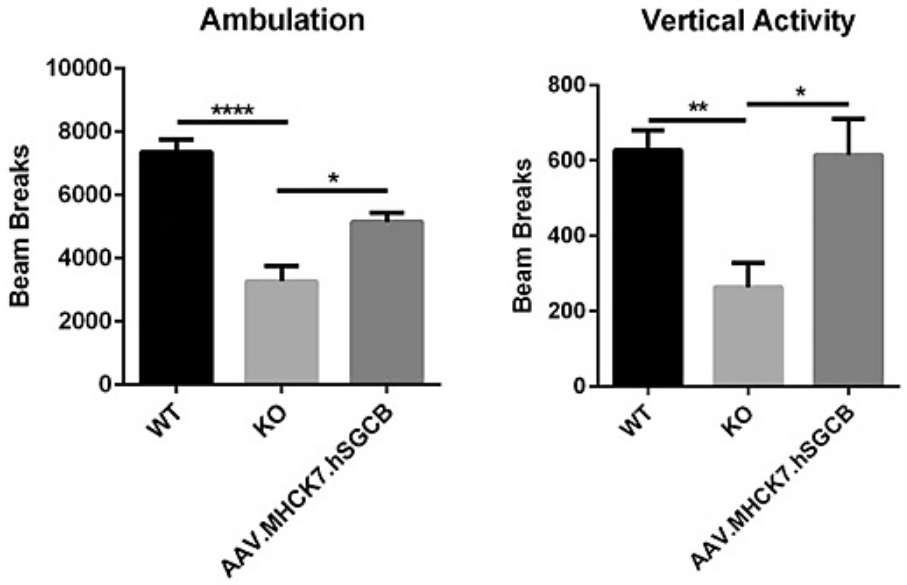


Figure 22B



Figure 22A

