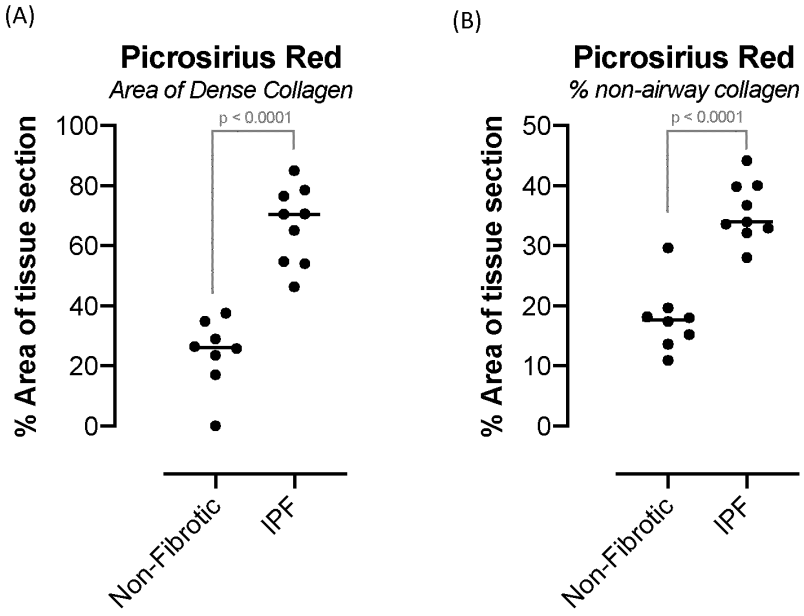
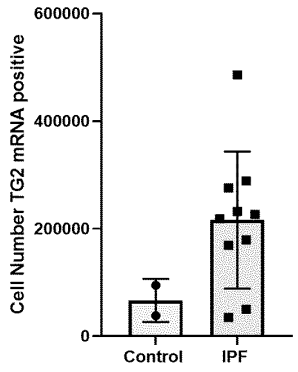

(A)
Picrosirius Red
Area of Dense Collagen
% Area of tissue section
p < 0.0001
100
80
60
40
20
0
Non-Fibrotic
IPF
(B)
Picrosirius Red
% non-airway collagen
% Area of tissue section
p < 0.0001
50
40
30
20
10
0
Non-Fibrotic
IPF

Figure 1

(A) : Total TG2 mRNA positive cells



(B) TG2 mRNA positive cells by staining intensity

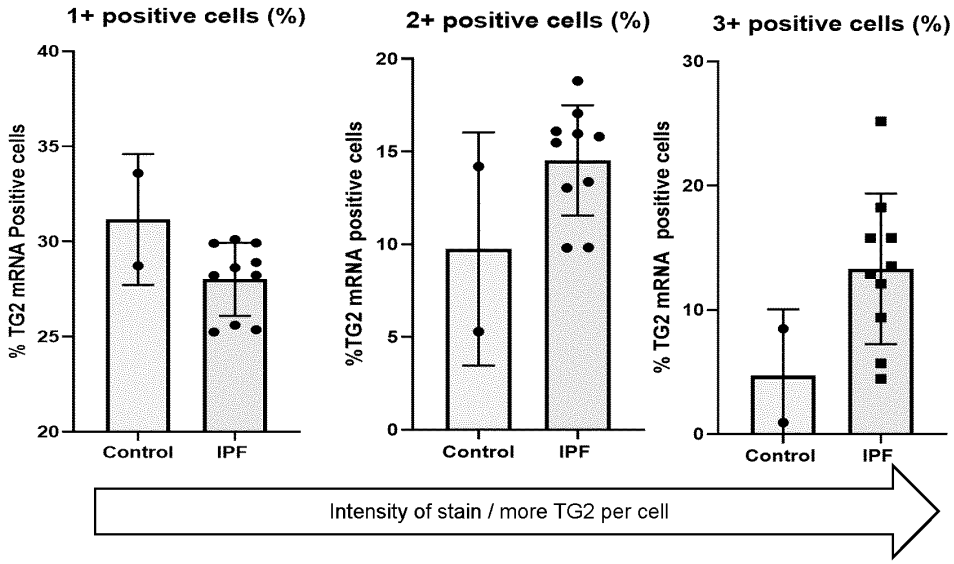
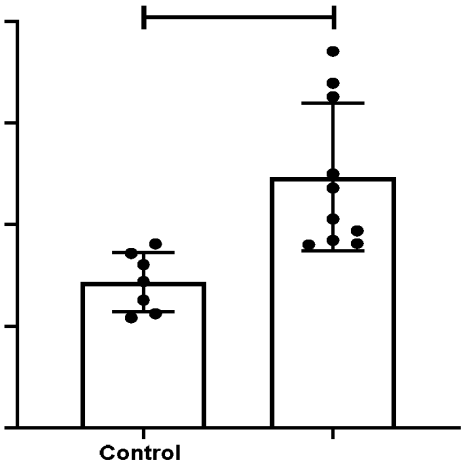


Figure 2

(A) TG2 Antigen staining is elevated in IPF Biopsy



(B) Lung function correlates with lung collagen levels

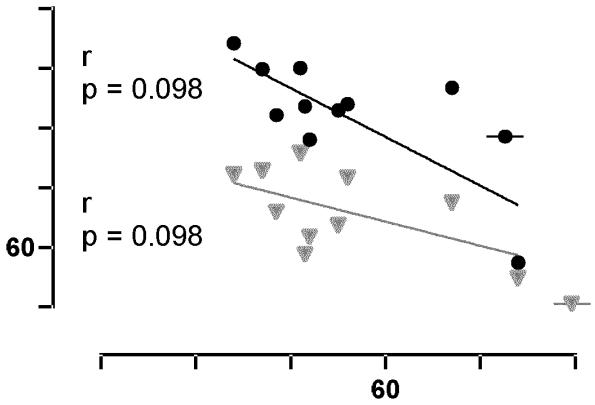


Figure 3

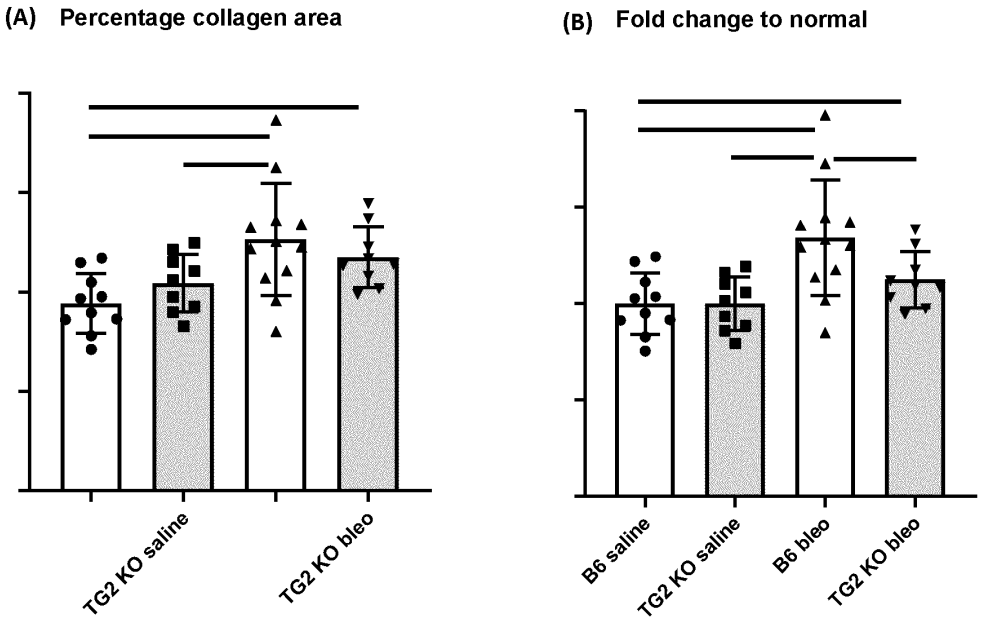

(A) Percentage collagen area
TG2 KO saline
TG2 KO bleo
(B) Fold change to normal
B6 saline
TG2 KO saline
B6 bleo
TG2 KO bleo

Figure 4

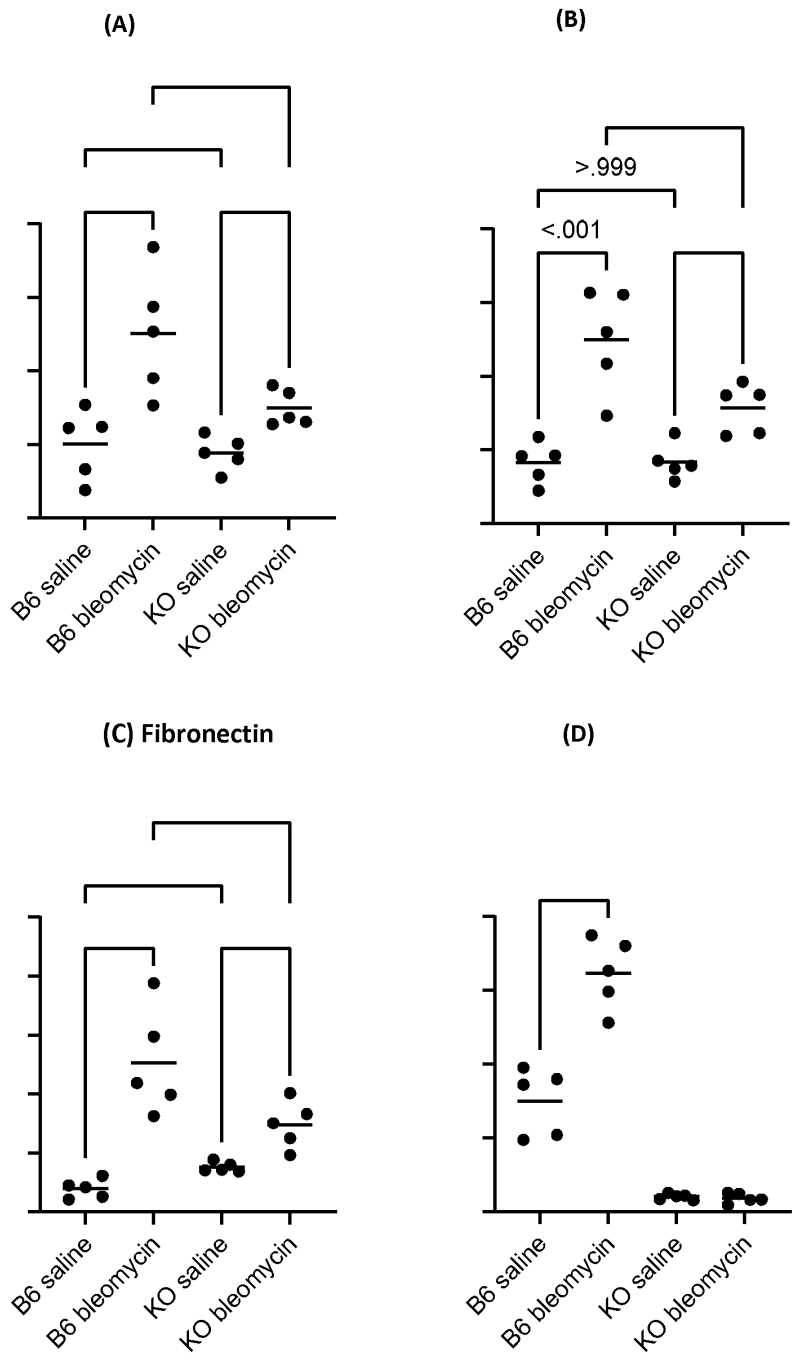

(A)
B6 saline
B6 bleomycin
KO saline
KO bleomycin
(B)
>.999
<.001
B6 saline
B6 bleomycin
KO saline
KO bleomycin
(C) Fibronectin
B6 saline
B6 bleomycin
KO saline
KO bleomycin
(D)
B6 saline
B6 bleomycin
KO saline
KO bleomycin

Figure 5

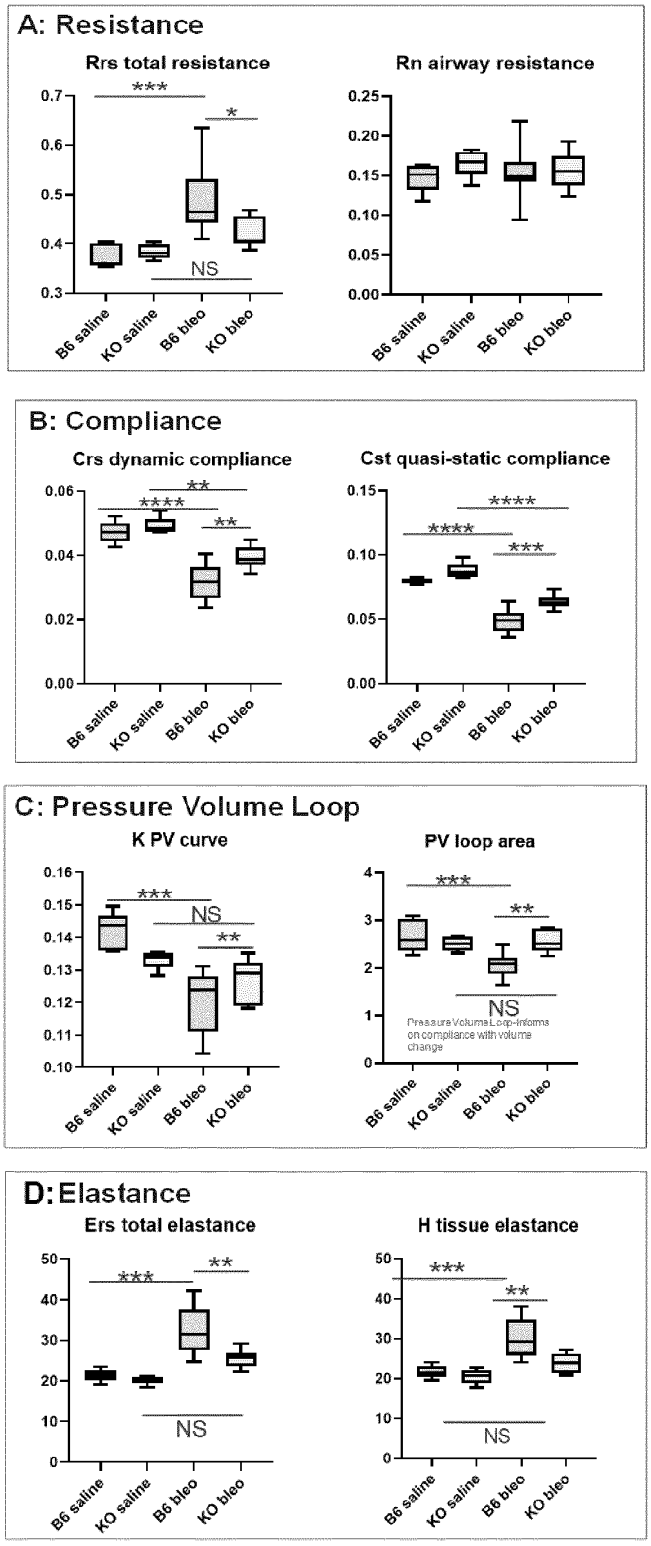
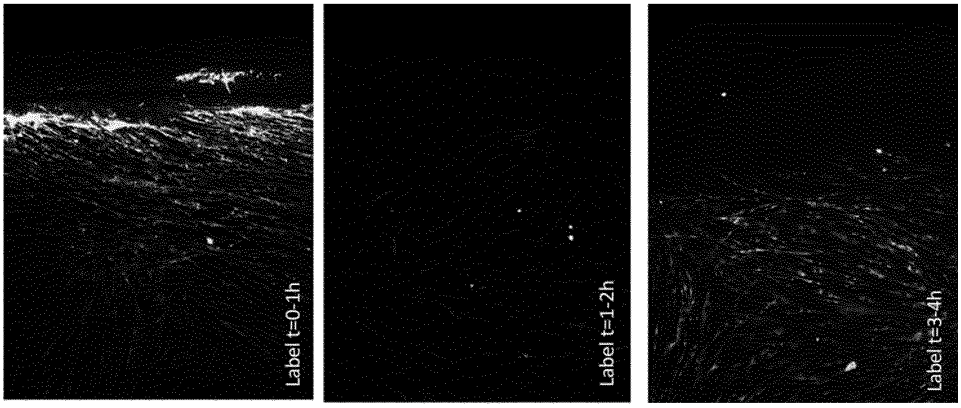
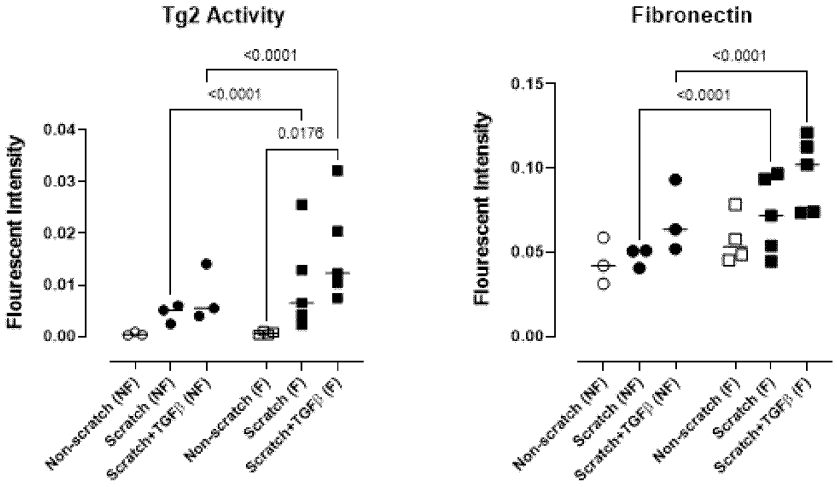

A: Resistance
Rrs total resistance
Rn airway resistance
B: Compliance
Crs dynamic compliance
Cst quasi-static compliance
C: Pressure Volume Loop
K PV curve
PV loop area
D: Elastance
Ers total elastance
H tissue elastance
B6 saline
KO saline
B6 bleo
KO bleo
NS

Figure 6

(A) Raised TG2 activity in a scratch wound in IPF fibroblasts is lost after 1 hour



(B) Quantification of elevated TG activity and Fibronectin in a scratch wound in IPF fibroblasts



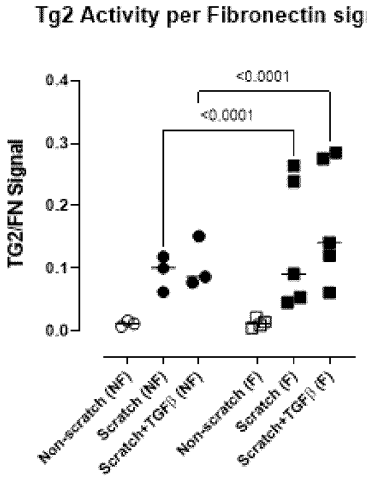


Figure 7

(C) TG activity in a lung fibroblast scratch wound is totally blocked by TG2 inhibitory antibody rbBB7

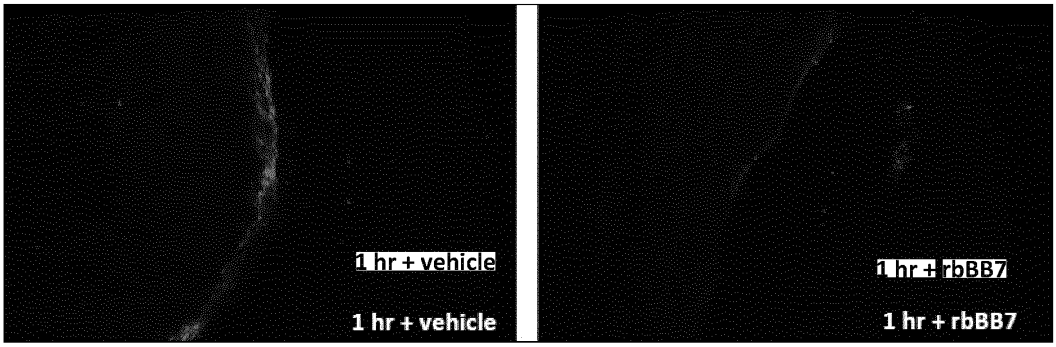


Figure 7 (cont')

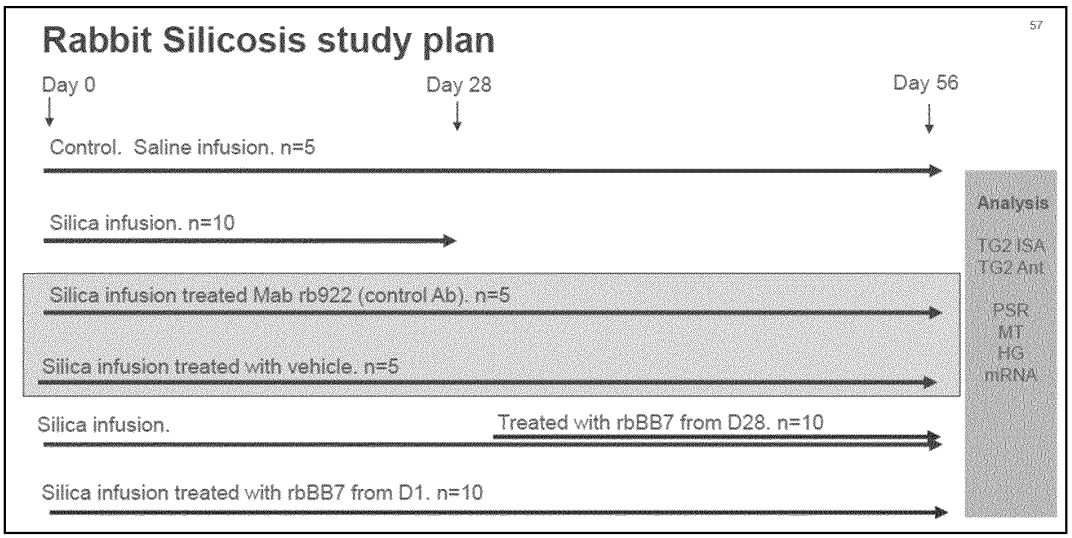


Figure 8

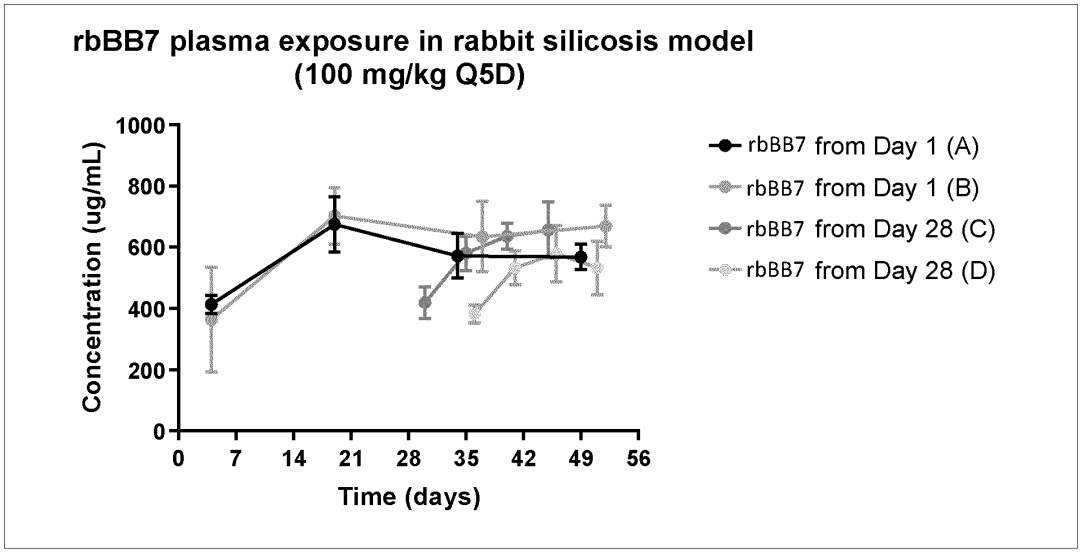


Figure 9

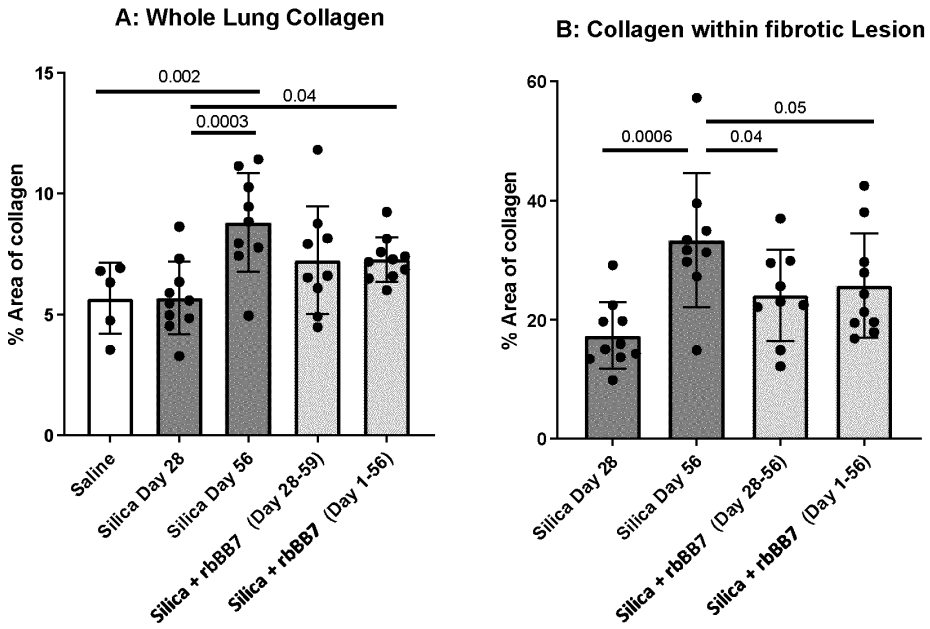


Figure 10

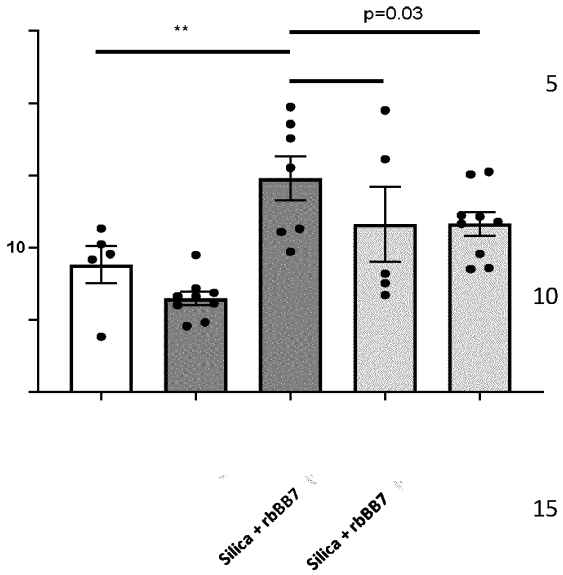


Figure 11

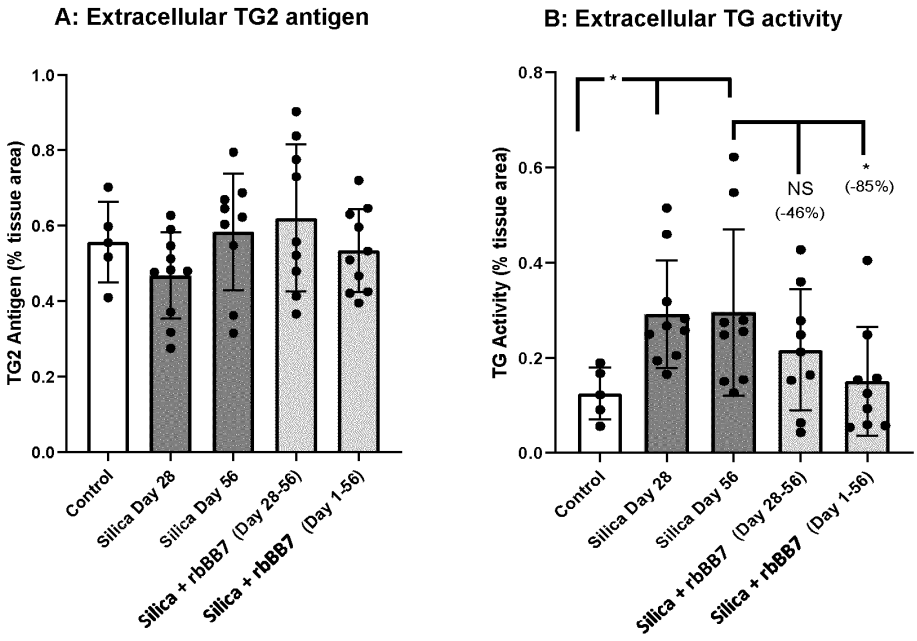


Figure 12

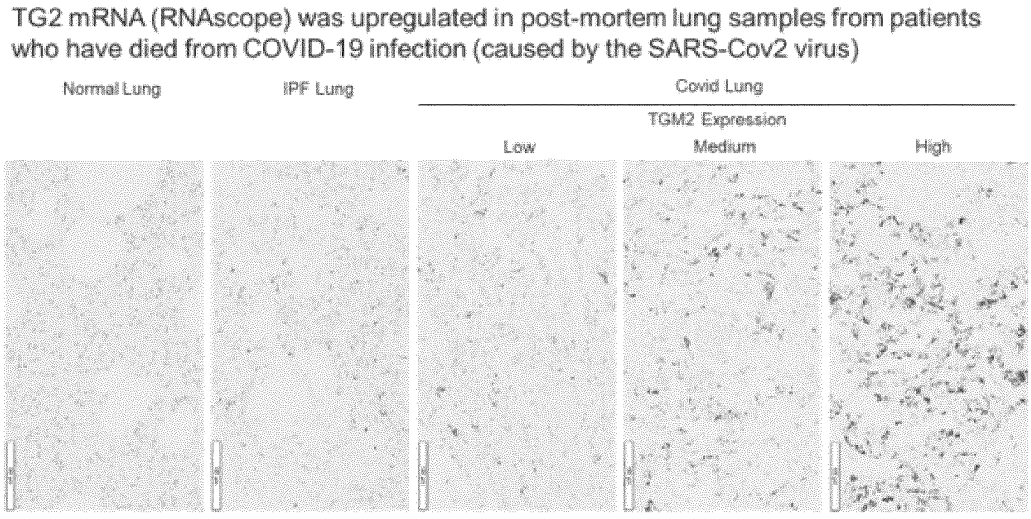
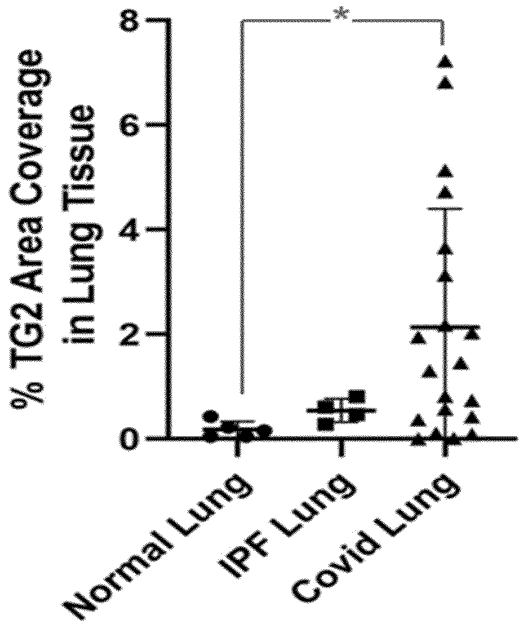


Figure 13



*P<0.05

Figure 14