

## Supporting Information

### **CB<sub>1</sub>R and iNOS are distinct players promoting pulmonary fibrosis in Hermansky-Pudlak syndrome**

Resat Cinar<sup>1\*</sup>, Joshua K. Park<sup>1</sup>, Charles N. Zawatsky<sup>1</sup>, Nathan J. Coffey<sup>1</sup>, Steven P. Bodine<sup>2</sup>, Jasmina Abdalla<sup>1</sup>, Tadafumi Yokoyama<sup>2†</sup>, Tony Jourdan<sup>1‡</sup>, Lindsey Jay<sup>1</sup>, Mei Xing G. Zuo<sup>1</sup>, Kevin J. O'Brien<sup>2</sup>, Junfeng Huang<sup>3</sup>, Ken Mackie<sup>4</sup>, Asaf Alimardanov<sup>3</sup>, Malliga R. Iyer<sup>1</sup>, William A. Gahl<sup>2,5</sup>, George Kunos<sup>1</sup>, Bernadette R. Gochuico<sup>2</sup>, May Christine V. Malicdan<sup>2,5</sup>

<sup>1</sup>Laboratory of Physiologic Studies, National Institute on Alcohol Abuse and Alcoholism, National Institutes of Health, Rockville, MD, 20852, USA.

<sup>2</sup>Section of Human Biochemical Genetics, Medical Genetics Branch, National Human Genome Research Institute, National Institutes of Health, Bethesda, MD, 20892, USA.

<sup>3</sup>Therapeutics Development Branch, Division of Preclinical Innovation, National Center for Advancing Translational Sciences, National Institutes of Health, Rockville, MD

<sup>4</sup>Department of Psychological and Brain Sciences, Indiana University, Bloomington, IN

<sup>5</sup>NIH Undiagnosed Diseases Program and Office of the Clinical Director, National Human Genome Research Institute, National Institutes of Health, Bethesda, MD 20892, USA.

\*To whom correspondence should be addressed: Resat Cinar, BPharm, PhD  
[resat.cinar@nih.gov](mailto:resat.cinar@nih.gov)

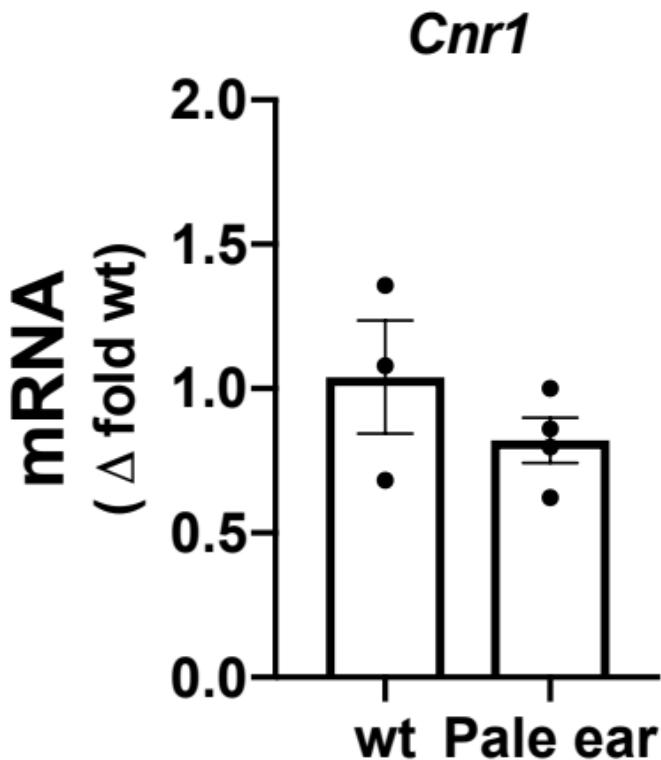
† Department of Pediatrics, Kanazawa University, Kanazawa Japan

‡ INSERM Lipids, Nutrition, Cancer UMR1231, University of Burgundy and Franche-Comté, Dijon, France

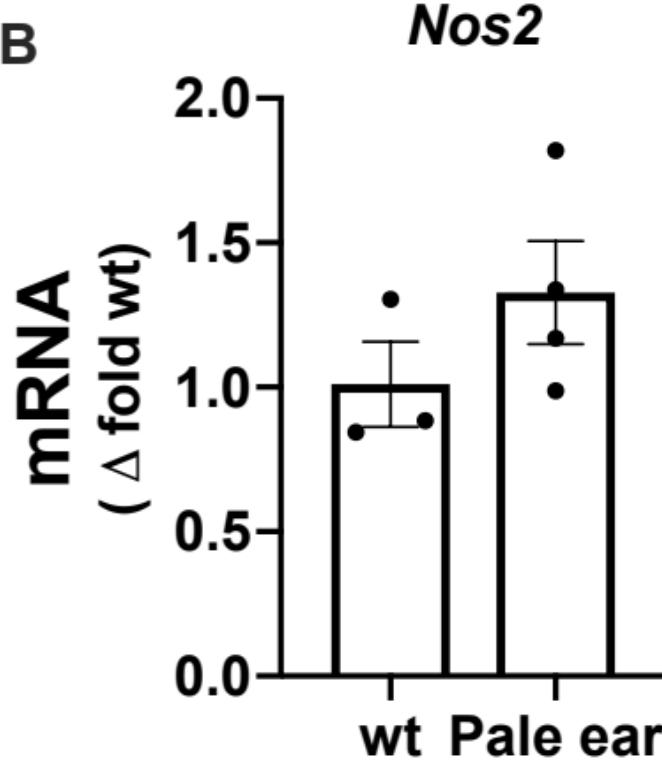
**Running Title:** Dual targeting of CB<sub>1</sub>R and iNOS in HPSPF

# Supplemental Figure 1

A

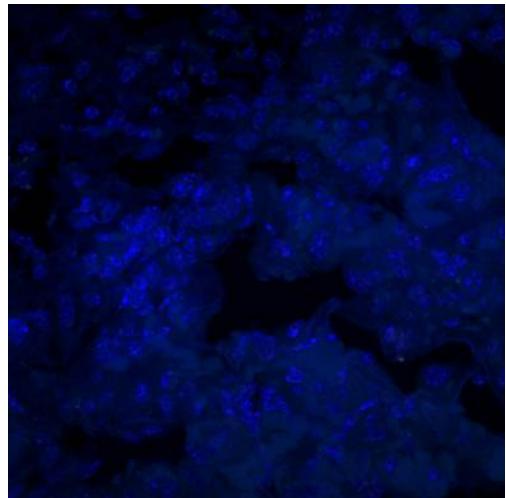


B

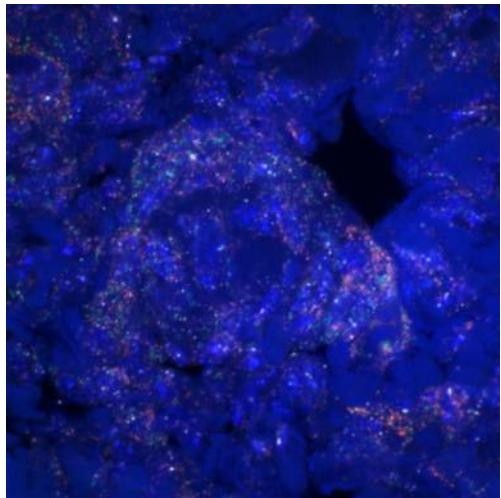


Supplemental Figure 2

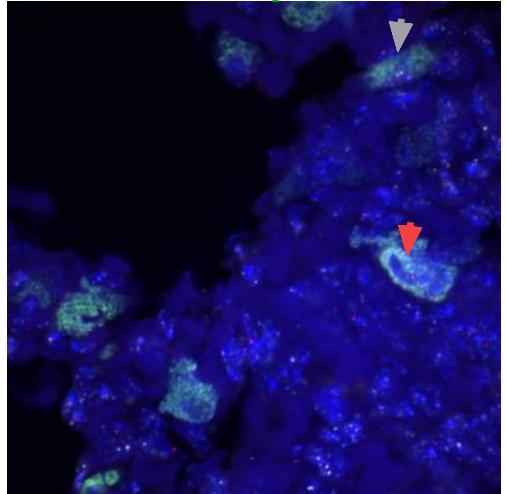
A Negative control



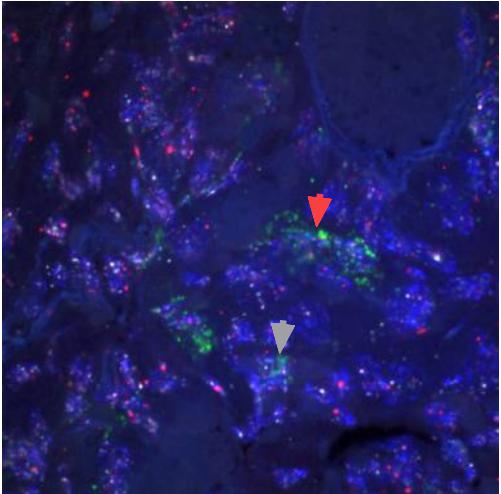
B Positive control



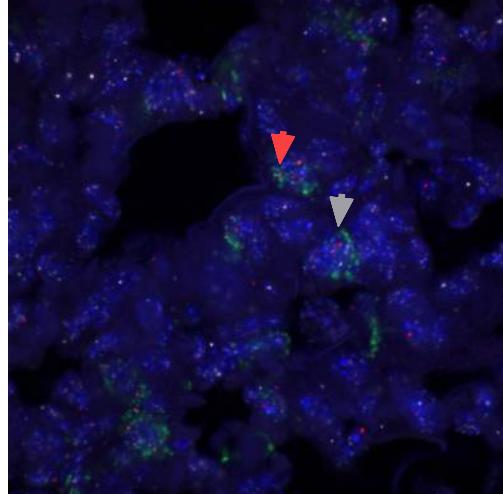
C *Nos2 / Sftpc / Cnr1*



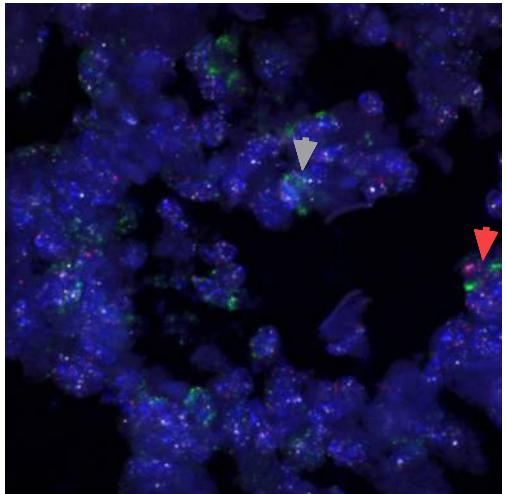
D *Nos2 / CD68 / Cnr1*



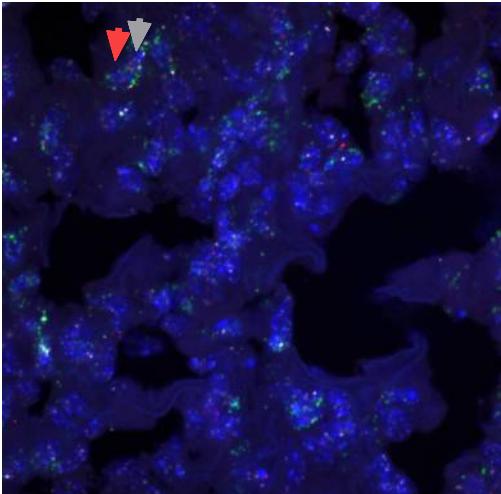
E *Nos2 / Col1a / Cnr1*



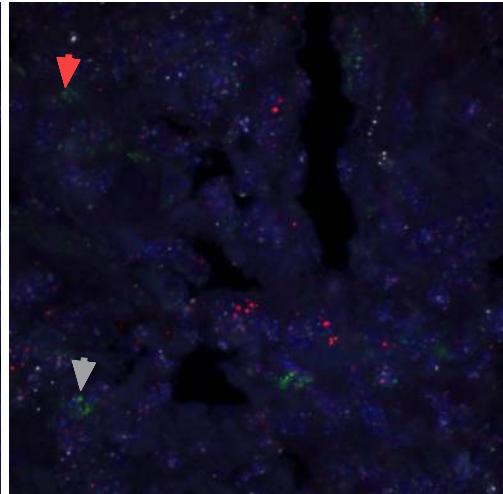
F *Nos2 / Pdpn / Cnr1*



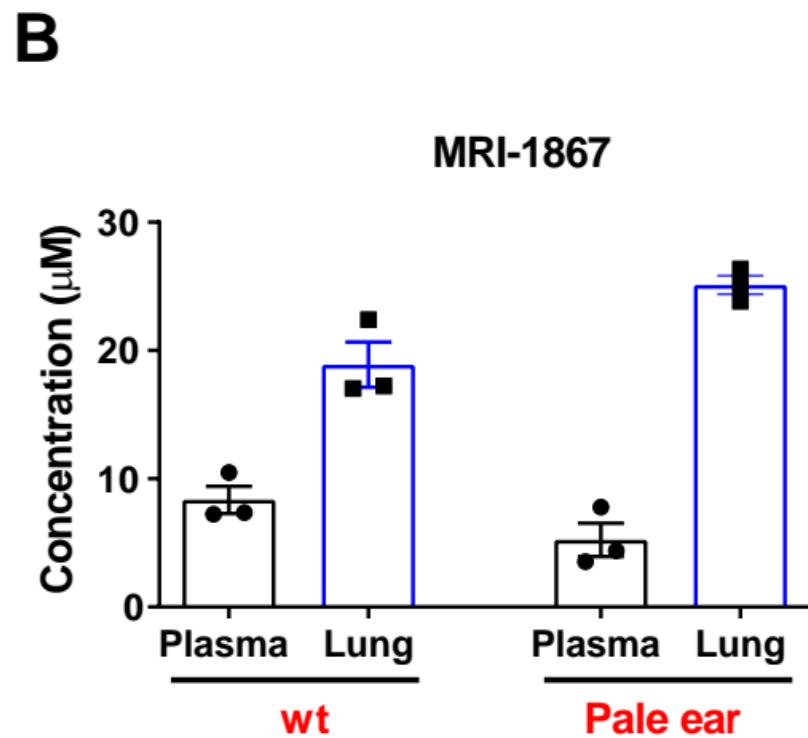
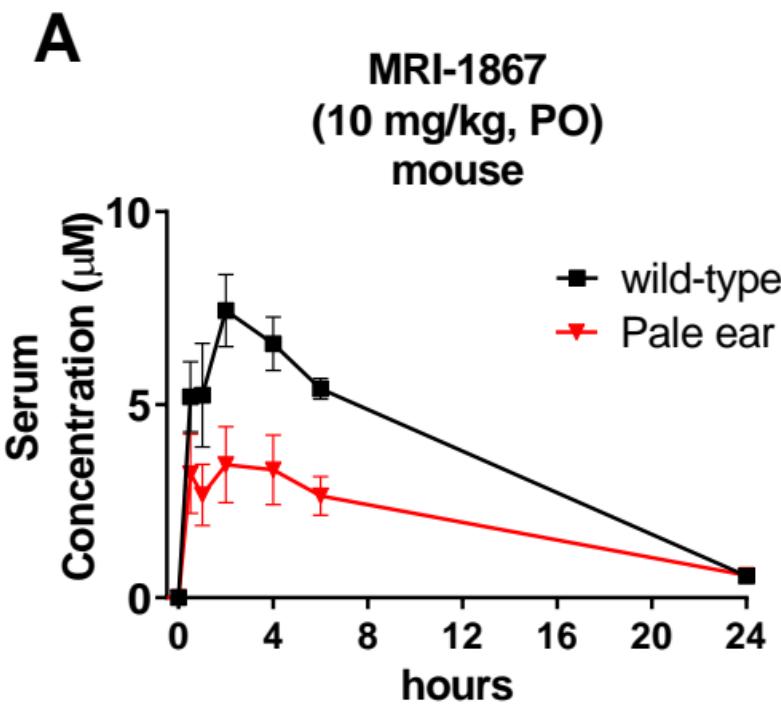
G *Nos2 / Pecam / Cnr1*



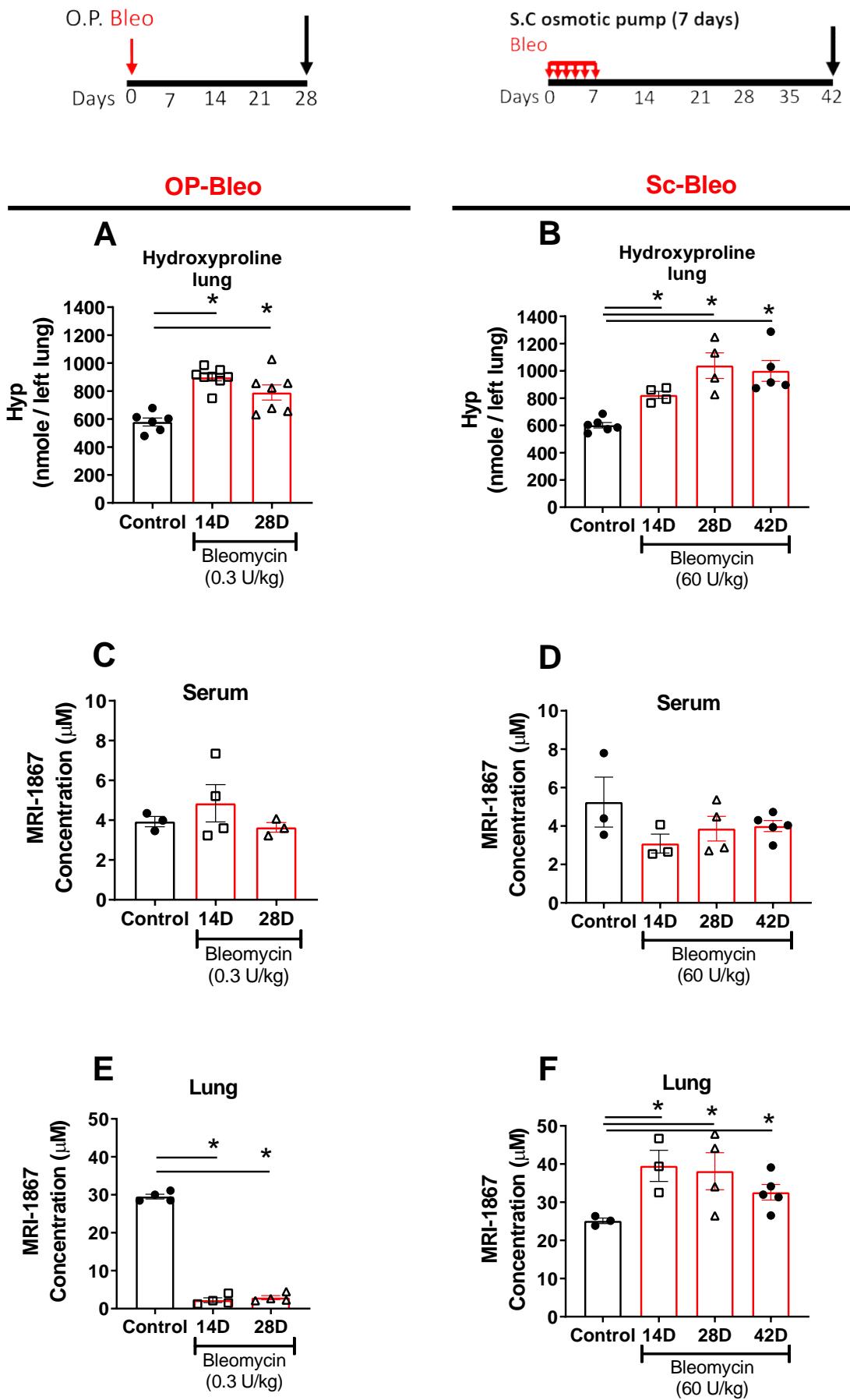
H *Nos2 / Cspg4 / Cnr1*



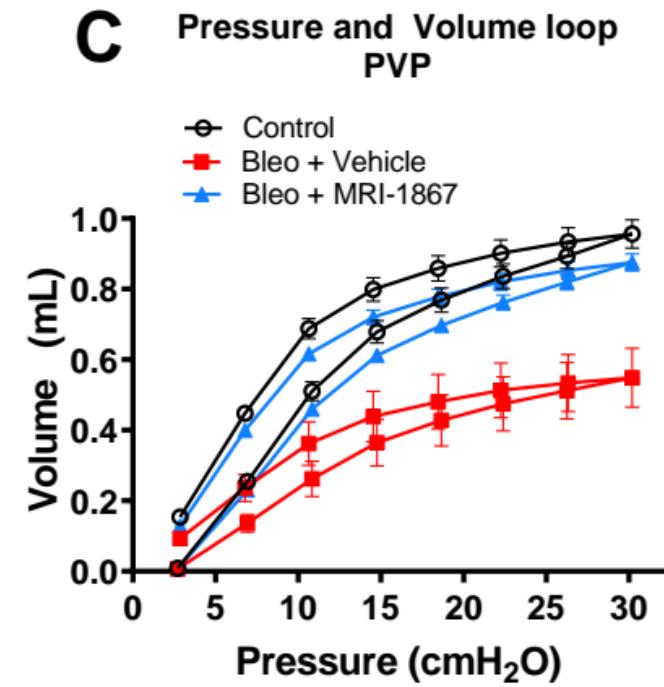
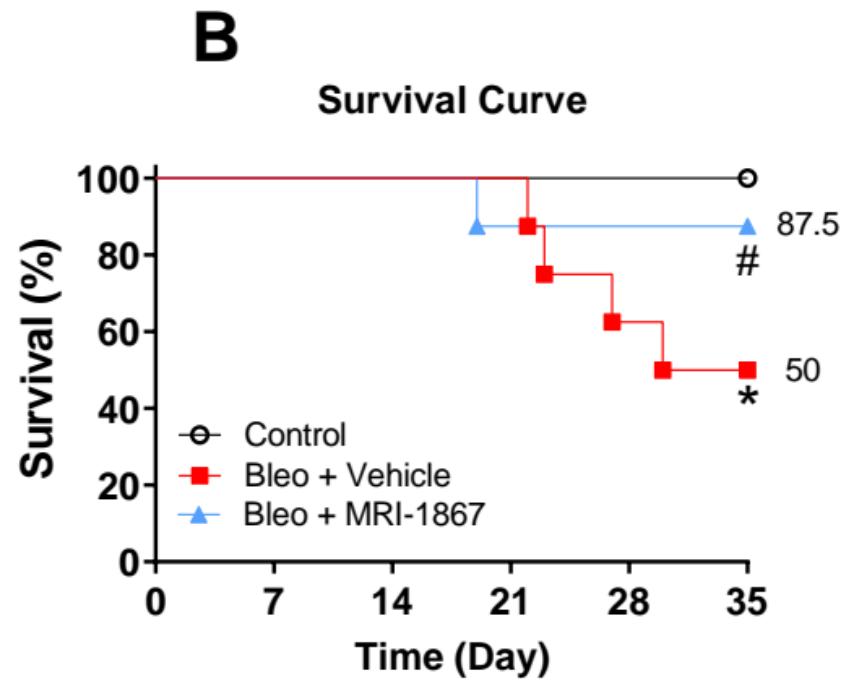
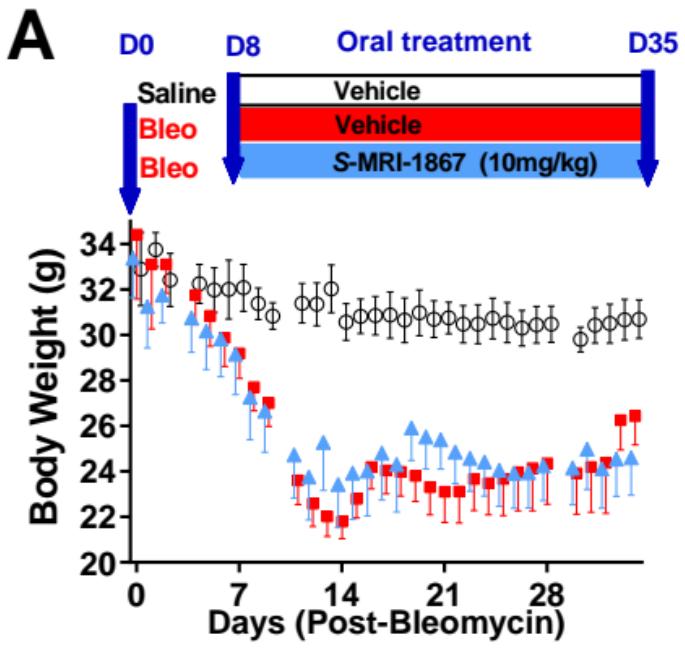
## Supplemental Figure 3



## Supplemental Figure 4



## Supplemental Figure 5



**Supplemental Figure 6**

**A** Control

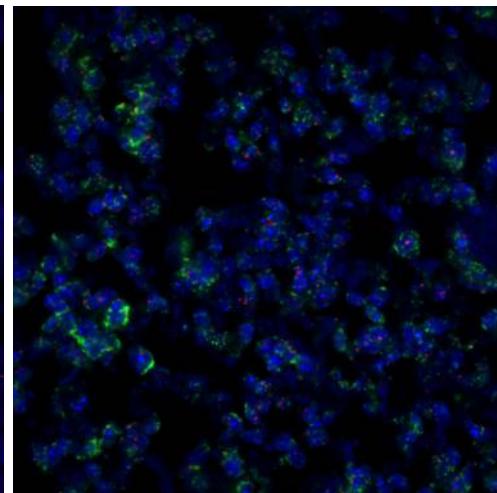
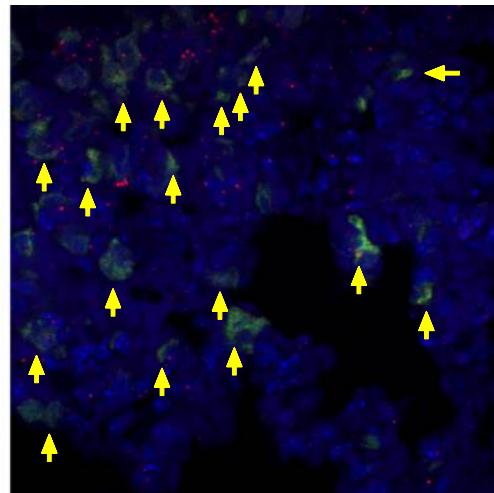
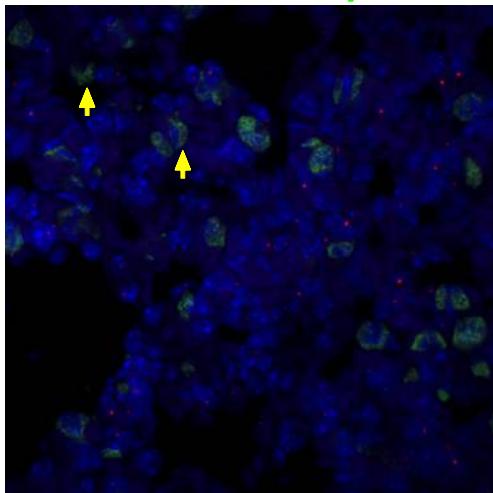
Bleomycin

Positive Control

*Cnr1 / Sftpc*

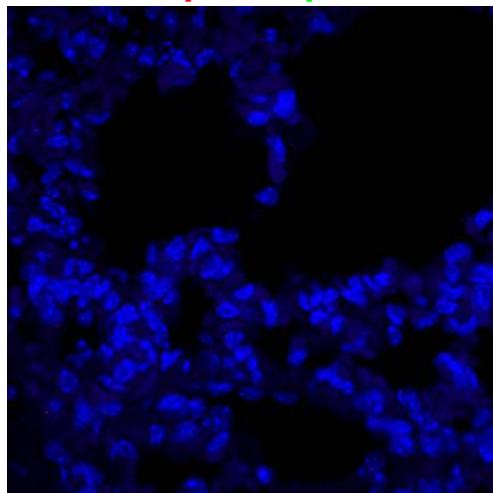
*Cnr1 / Sftpc*

*Polr2A / Ubc*



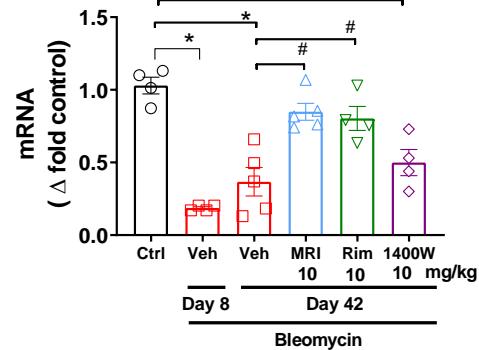
Negative control

*dapB / dapB*



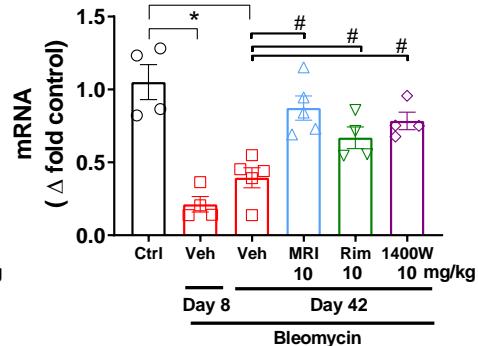
**B**

*Ppargc1a*  
(PGC1 $\alpha$ )



**C**

*Pink1*



**D**

*Tgfβ1*

