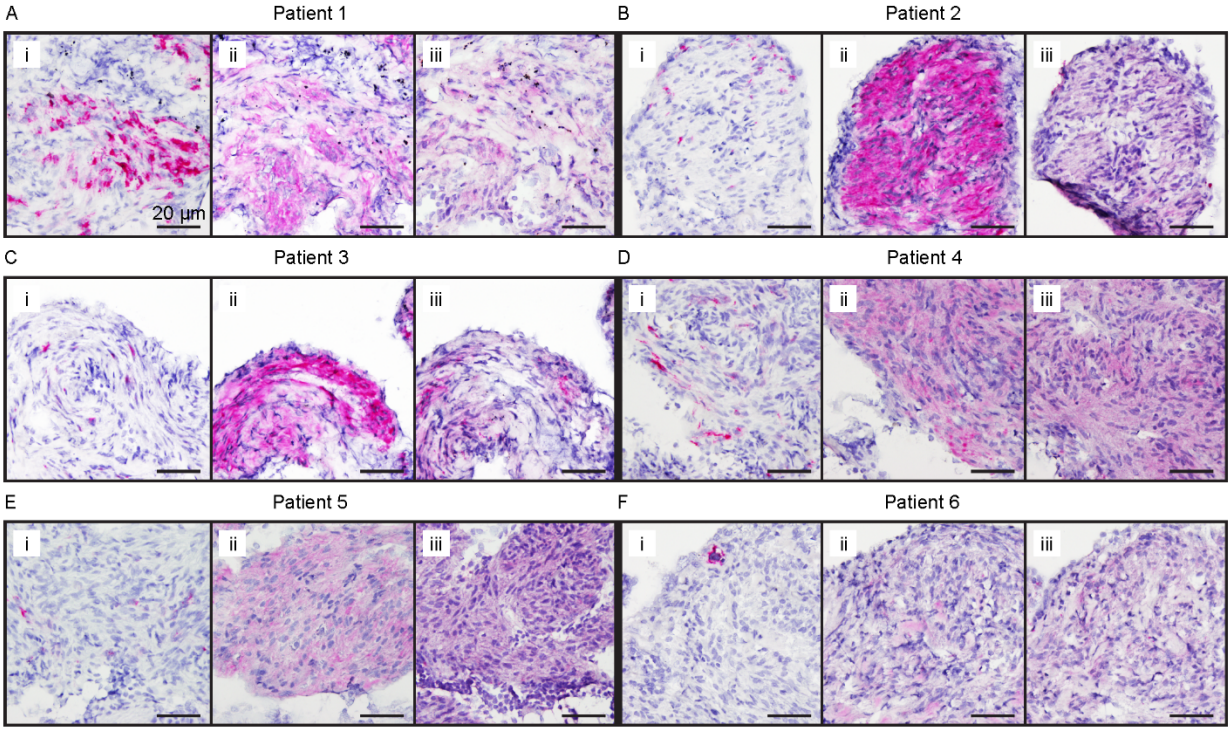
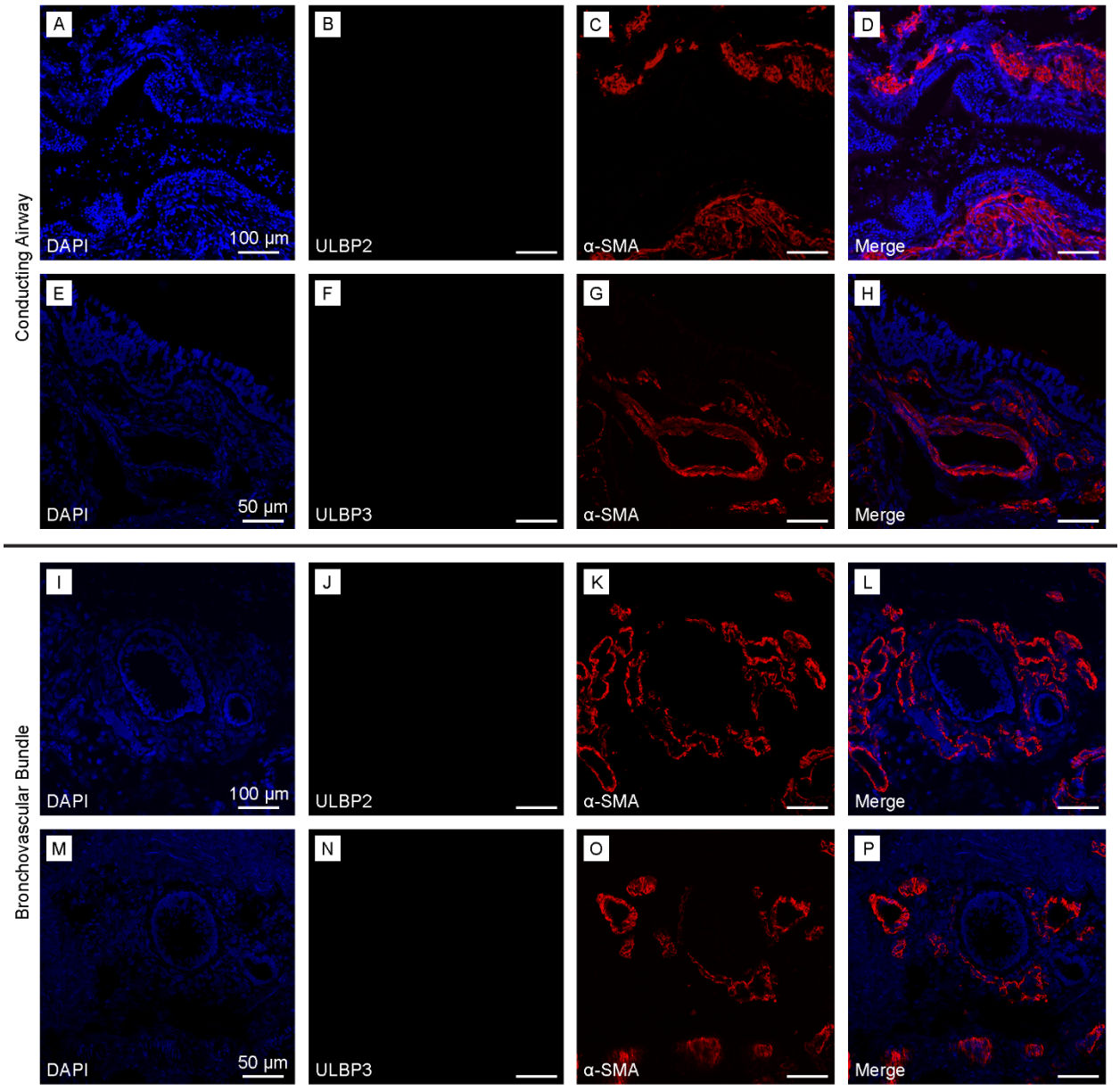


Supplemental figure 1. Immunohistochemical analysis of the markers Human melanoma black (HMB-45), alpha-smooth muscle actin (α-SMA), and the NKG2D ligand ULBP2 in multiple lymphangioleiomyomatosis (LAM) patients. (A-F) For each patient, (i) HMB-45, (ii) α-SMA, and (iii) ULBP2. Scale bars: 20 µm. This figure illustrates immunohistochemical staining of the remaining LAM patients not shown in Figure 1 (n=6).


Conducting Airway
A
DAPI
100 μm
B
ULBP2
C
α-SMA
D
Merge
E
DAPI
50 μm
F
ULBP3
G
α-SMA
H
Merge
Bronchovascular Bundle
I
DAPI
100 μm
J
ULBP2
K
α-SMA
L
Merge
M
DAPI
50 μm
N
ULBP3
O
α-SMA
P
Merge

Supplemental figure 2. Control staining for immunofluorescent analysis of NKG2D ligands and alpha-smooth muscle actin (α-SMA) in lymphangioleiomyomatosis (LAM) lung tissue. (A-P) LAM lung tissue sections (5 μm thickness) were stained immunofluorescently for the NKG2D ligands ULBP2 and ULBP3, as well as for the marker α-SMA. (A-H) Confocal microscopy images of conducting airways and (I-P) bronchovascular bundles located within LAM lung tissue sections. Bronchiolar smooth muscle (C, G) and vascular smooth muscle (K, O) express α-SMA (red fluorescence). No staining for the NKG2D ligands ULBP2 and ULBP3 was observed in the conducting airway (B, F) or bronchovascular bundle (J, N) tissue sections. (D, H, L, P) Merged images illustrate histopathological localization of DAPI, NKG2D ligands, and α-SMA in bronchiolar and vascular smooth muscle within tissue sections. (A-D, I-L) Scale bars: 100 μm. (E-H, M-P) Scale bars: 50 μm.

Supplemental Table 1: LAM patient demographics

Patient	FEV_1 (L)	% FEV_1 predicted	FVC (L)	% FVC predicted	Age	mTOR inhibitors
1	0.9	34%	2.84	82%	44	Sirolimus
2	1.5	53%	2.51	69%	52	Sirolimus
3	1.92	70%	3.0	85%	62	None
4	2.95	115%	3.74	114%	41	None
5	1.48	58%	2.27	68%	48	None
6	2.11	62%	3.54	82%	49	Sirolimus
7	2.28	84%	3.64	102%	52	Everolimus
Average	1.88 ± 0.66	0.68 ± 0.26	3.08 ± 0.58	0.86 ± 0.17	49.7± 6.7	