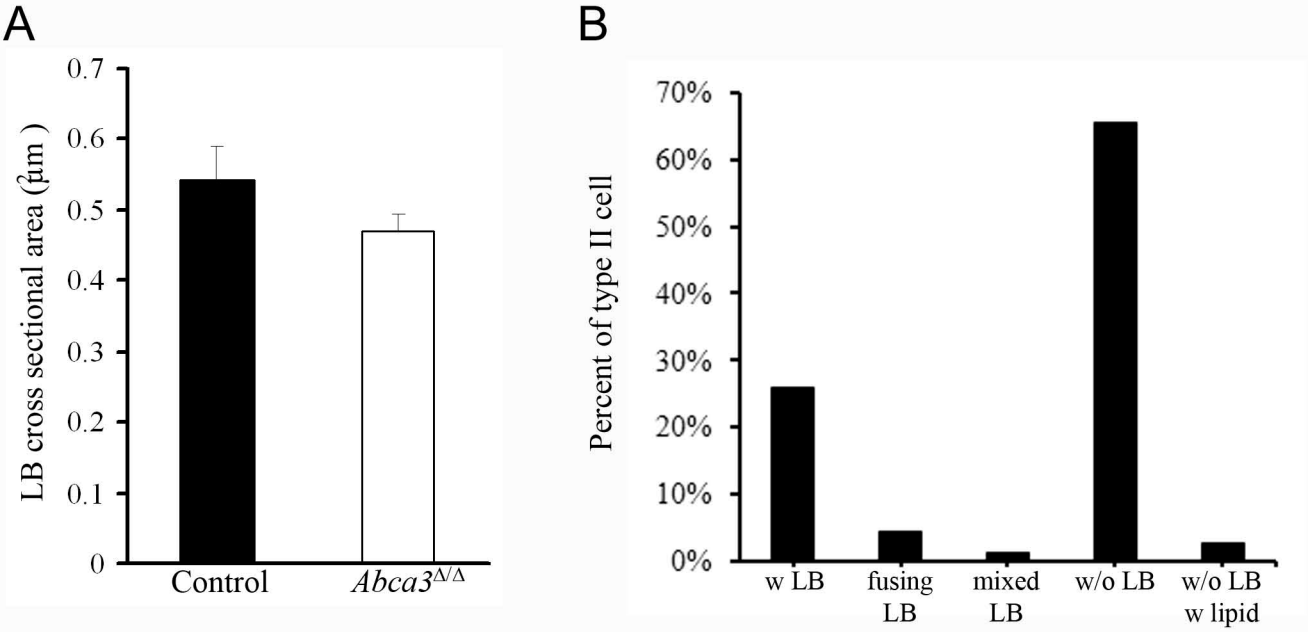
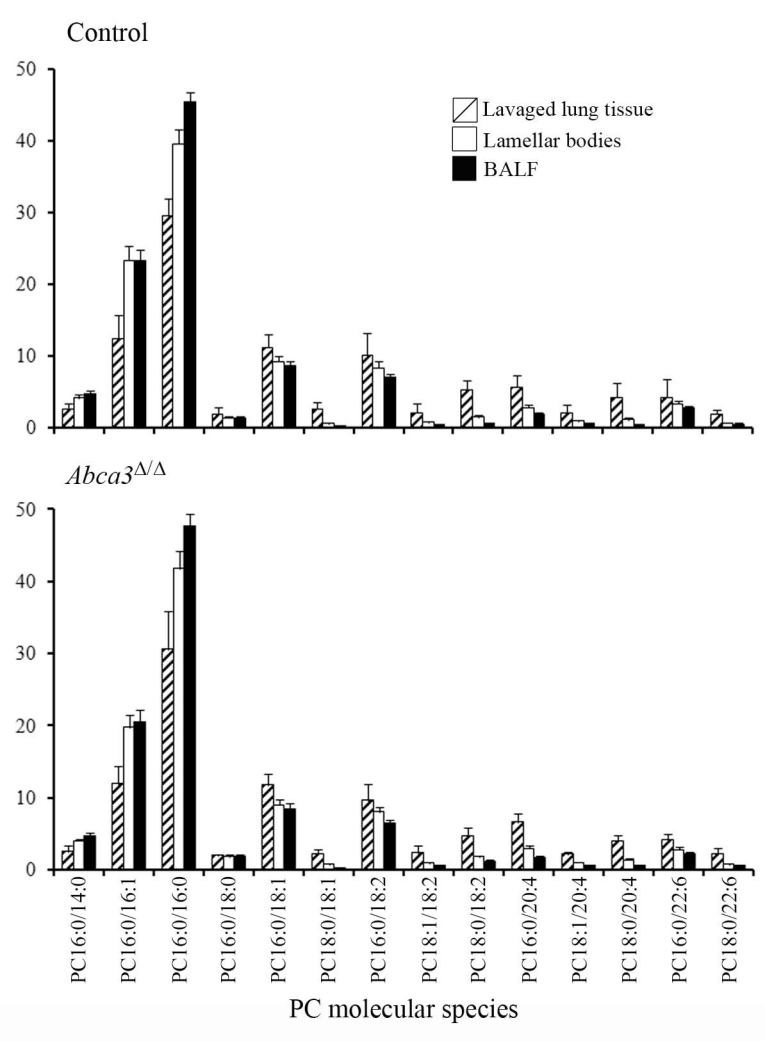
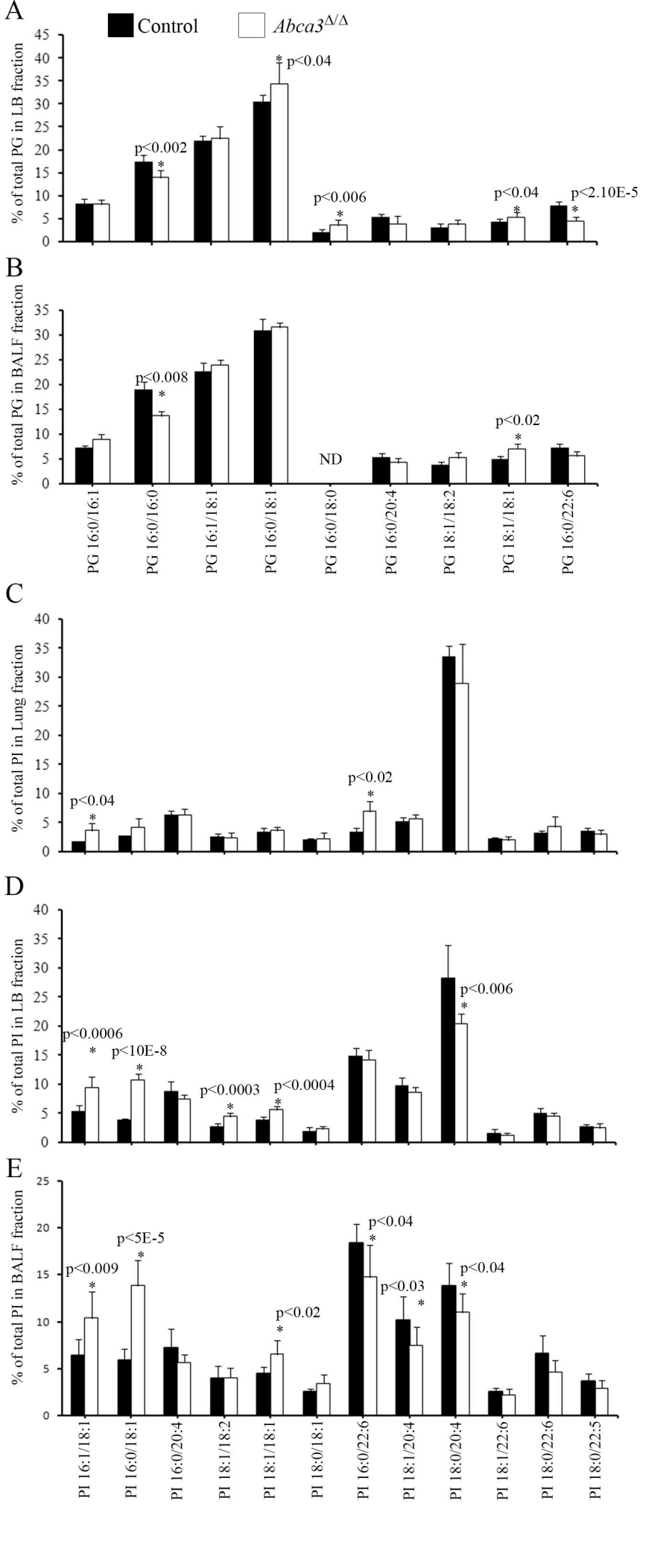

A
0.7
0.6
0.5
0.4
0.3
0.2
0.1
0
LB cross sectional area (μm)
Control
Abca3Δ/Δ
B
70%
60%
50%
40%
30%
20%
10%
0%
Percent of type II cell
w LB
fusing LB
mixed LB
w/o LB
w/o LB w lipid


Control
Lavaged lung tissue
Lamellar bodies
BALF
Abca3Δ/Δ
PC16:0/14:0
PC16:0/16:1
PC16:0/16:0
PC16:0/18:0
PC16:0/18:1
PC18:0/18:1
PC16:0/18:2
PC18:1/18:2
PC18:0/18:2
PC16:0/20:4
PC18:1/20:4
PC18:0/20:4
PC16:0/22:6
PC18:0/22:6
PC molecular species


A
Control
Abca3Δ/Δ
% of total PG in LB fraction
p<0.002
p<0.04
p<0.006
p<0.04
p<2.10E-5
B
% of total PG in BALF fraction
p<0.008
p<0.02
ND
PG 16:0/16:1
PG 16:0/16:0
PG 16:1/18:1
PG 16:0/18:1
PG 16:0/18:0
PG 16:0/20:4
PG 18:1/18:2
PG 18:1/18:1
PG 16:0/22:6
C
% of total PI in Lung fraction
p<0.04
p<0.02
D
% of total PI in LB fraction
p<0.0006
p<10E-8
p<0.0003
p<0.0004
p<0.006
E
% of total PI in BALF fraction
p<0.009
p<5E-5
p<0.02
p<0.04
p<0.03
p<0.04
PI 16:1/18:1
PI 16:0/18:1
PI 16:0/20:4
PI 18:1/18:2
PI 18:1/18:1
PI 18:0/18:1
PI 16:0/22:6
PI 18:1/20:4
PI 18:0/20:4
PI 18:1/22:6
PI 18:0/22:6
PI 18:0/22:5

Supplemental Figure 1 Analysis of lamellar bodies in adult *Abca3*$^{\Delta/\Delta}$ mice.

(**A**) To determine whether lamellar bodies (LBs) present in type II cells of both control and *Abca3*$^{\Delta/\Delta}$ were similar in size, LB area was determined as described in *Material and Methods*. There were no differences in LB cross-sectional area between control and *Abca3*$^{\Delta/\Delta}$ type II cells. (**B**) While alveolar type II cells contained normal LBs in control mice, *Abca3*$^{\Delta/\Delta}$ contained various populations of type II cells: cells with LBs (w LB), cells with fusing LBs (fusing LB), cells with both normal and fusing LBs (mixed), cells without LBs (w/o LB) and cells without LBs that contained lipid droplets (w/o LB, w lipid). Results are representative of n ≥ 3 *Abca3*$^{\Delta/\Delta}$ mice.

Supplemental Figure 2 The molecular species compositions of PC in lavaged lung tissue, lamellar bodies and BALF from control and adult *Abca3*$^{\Delta/\Delta}$ mice were determined by electrospray ionization mass spectrometry and expressed as a percentage of total PC in that tissue fraction. There was no difference to PC composition of any of these fractions between the two groups of mice, indicating that the mechanisms for molecular selectivity and acyl remodeling were unaltered in the adult *Abca3*$^{\Delta/\Delta}$ mice.

Supplemental Figure 3 PG and PI species and content variation in adult *Abca3*$^{\Delta/\Delta}$ mice.

PG and PI species were determined in lung tissue (after lavage), isolated lamellar bodies (LBs), and BALF from *Abca3*$^{\Delta/\Delta}$ and control littermates by electrospray ionization mass spectrometry (ESI-MS) analysis as described in *Materials and Methods*. Composition of individual PG molecular species in lamellar bodies (**A**) and bronchoalveolar lavage fluid (BALF) (**B**) from *Abca3*$^{\Delta/\Delta}$ and control littermates. Composition of individual PI molecular species in lavaged

lung (**C**), lamellar bodies (**D**) and BALF (**E**) from *Abca3*$^{\Delta/\Delta}$ and control littermates. Individual molecular species from each phospholipid class are expressed as means ± SE of 6-8 animals per group.