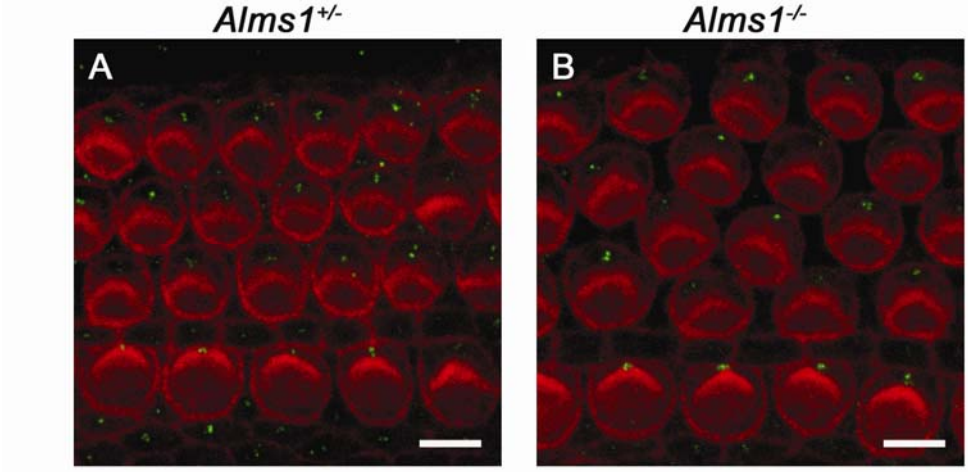
Supplementary Figure 1. ALMS1 localizes to basal bodies in neonatal *Alms1*$^{-/-}$ mice. (**A**) Mid-apical turn organ of Corti wholemount from a P2 *Alms1*$^{+/-}$ mouse stained with phalloidin (red) and an anti-ALMS1 antibody (green). ALMS1 localized to basal bodies of hair cells and supporting cells. (B) ALMS1 immunofluorescence was also detected in basal bodies of an *Alms1*$^{-/-}$ mouse littermate. Scale bars, 5 µm.

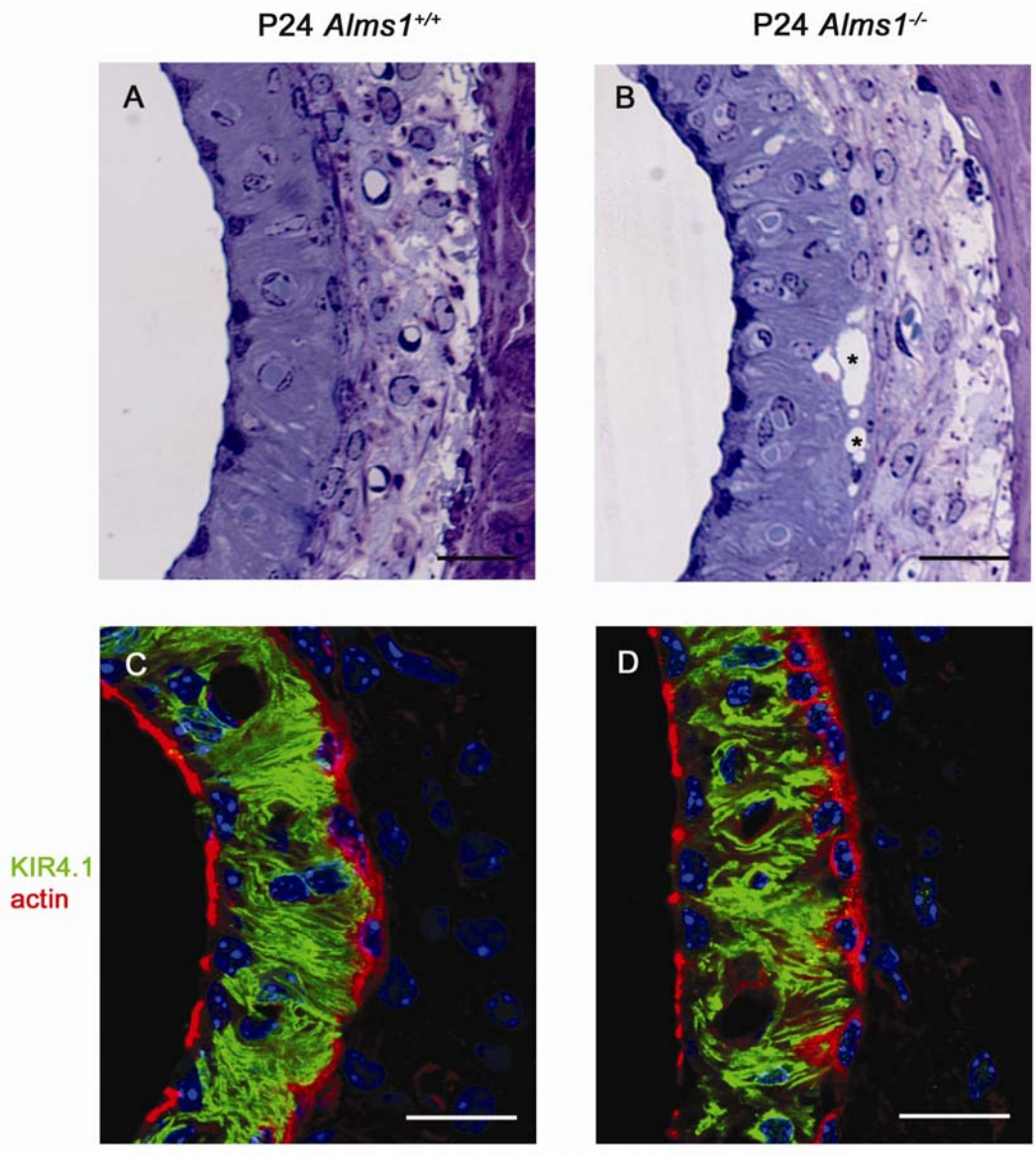
Supplementary Figure 2. Early ultrastructural changes in stria vascularis of *Alms1*$^{-/-}$ mice. (**A-B**) Photomicrographs of thin cochlear sections from P24 *Alms1*$^{+/+}$ and *Alms1*$^{-/-}$ mice. These figures show that the histopathology seen in older *Alms1*$^{-/-}$ mice was also present in these juvenile animals, and suggest that this degenerative process was progressive. There were no blebs on the apical (lumenal) membranes of marginal cells, which were seen in older animals. (**C-D**) Projections of confocal image stacks of cochlear cryo-sections from P24 *Alms1*$^{+/+}$ and *Alms1*$^{-/-}$ mice, labeled with an antibody against KIR4.1 (green) that labels intermediate cells, and phalloidin (red) that labels the basal cell layer and the apical region of marginal cells. Intermediate cells were present in the juvenile *Alms1*$^{-/-}$ mouse. Scale bars, 20 µm.

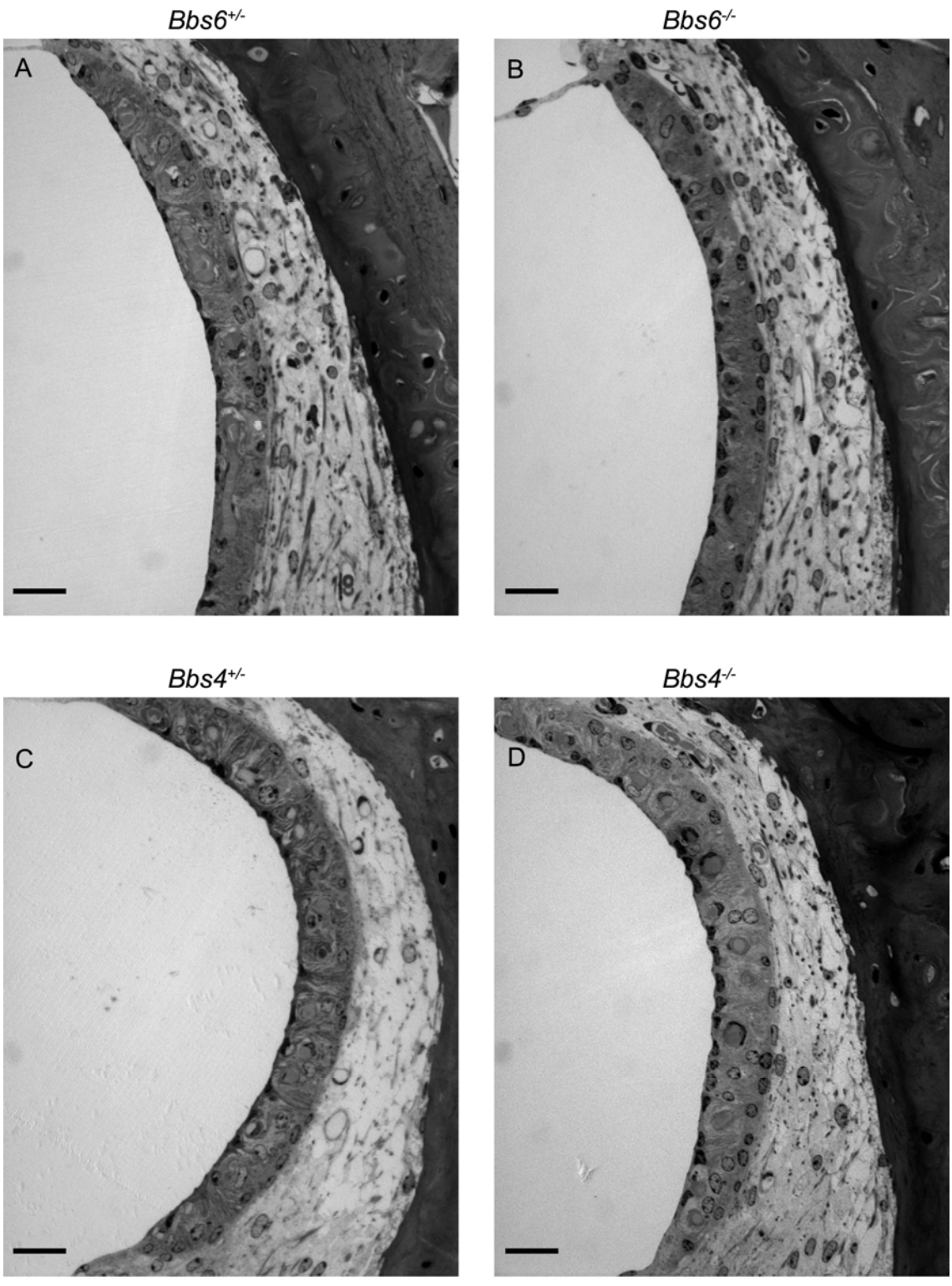
Supplementary Figure 3. Ultra-structure of stria vascularis in *Bbs* mutant mice was normal. (**A-B**) Photomicrographs of thin cochlear sections from 6-month old *Bbs6*$^{+/-}$ and *Bbs6*$^{-/-}$ mice, detailing the lateral wall. (**C-D**) Photomicrographs of thin cochlear sections from 6-month old *Bbs4*$^{+/-}$ and *Bbs4*$^{-/-}$ mice. Scale bars 20 µm.

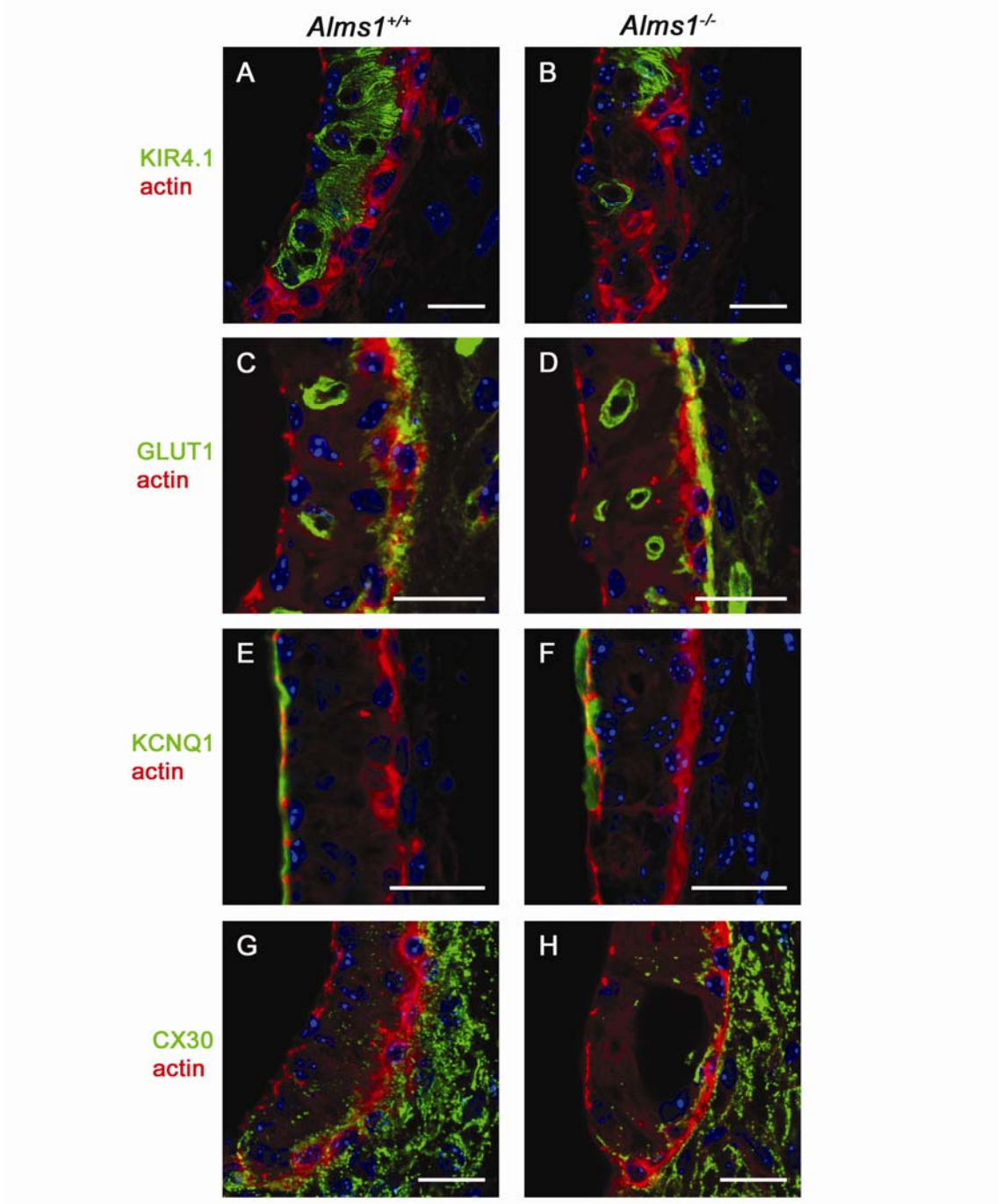
Supplementary Figure 4. Normal expression of proteins responsible for strial function in older *Alms1*$^{-/-}$ mice. (**A-H**) Projections of confocal image stacks of cochlear cryo-sections from 7-month old *Alms1*$^{+/+}$ and *Alms1*$^{-/-}$ mice, labeled with phalloidin (red) that labels actin

in the basal cell layer and the apical region of marginal cells, and antibodies against cell-specific proteins (green) as specified below. (**A**) An anti-KIR4.1 antibody labeled intermediate cells in *Alms1*$^{+/+}$ mouse stria vascularis. (**B**) Anti-KIR4.1 labeled cells were largely absent in the equivalent region of *Alms1*$^{-/-}$ mouse, though some comparably labeled intermediate cells were present at the top of the section. The basal cell layer and the marginal cell layer appeared intact. (**C**) In an *Alms1*$^{+/+}$ mouse an anti-GLUT1 antibody labeled the apical and basolateral membranes of basal cells, and vascular pericytes. (**D**) Comparable labeling was seen in *Alms1*$^{-/-}$ mouse basal cells and pericytes. (**E**) In an *Alms1*$^{+/+}$ mouse an anti-KCNQ1 antibody labeled the apical (lumenal) membrane of marginal cells. (**F**) There was comparable labeling in some marginal cells of an *Alms1*$^{-/-}$ mouse. KCNQ1 labeling was absent in the lower region of stria vascularis possibly denoting loss of marginal cells. (**G**) An anti-CX30 antibody labeled gap junctional plaques between spiral ligament fibrocytes, in the basal cell layer, and within the intermediate cell layer in an *Alms1*$^{+/+}$ mouse. (**H**) Comparable labeling was seen in an *Alms1*$^{-/-}$ mouse, even in a region where very large lesions were present. Scale bars 20 μm.


Alms1+/-
Alms1-/-
A
B


P24 Alms1+/+
P24 Alms1-/-
A
B
*
*
C
D
KIR4.1
actin


Bbs6+/-
Bbs6-/-
A
B
Bbs4+/-
Bbs4-/-
C
D


Alms1+/+
Alms1-/-
A
B
KIR4.1
actin
C
D
GLUT1
actin
E
F
KCNQ1
actin
G
H
CX30
actin