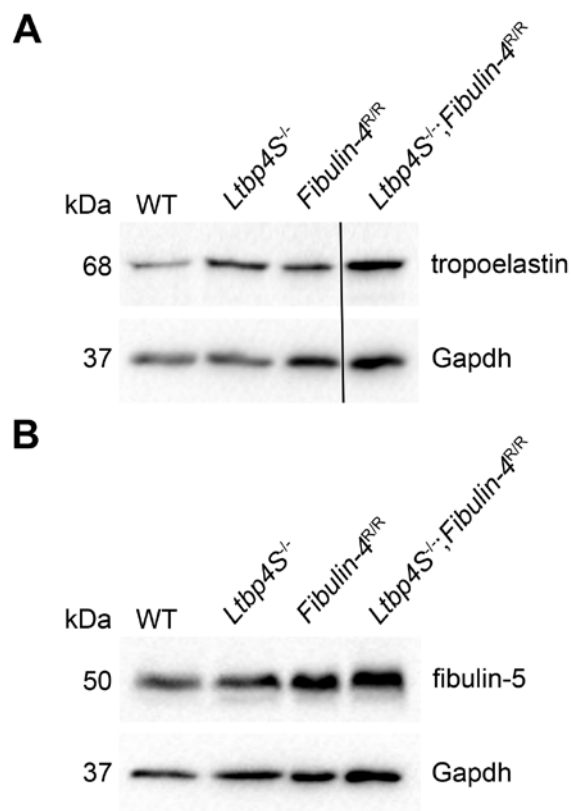


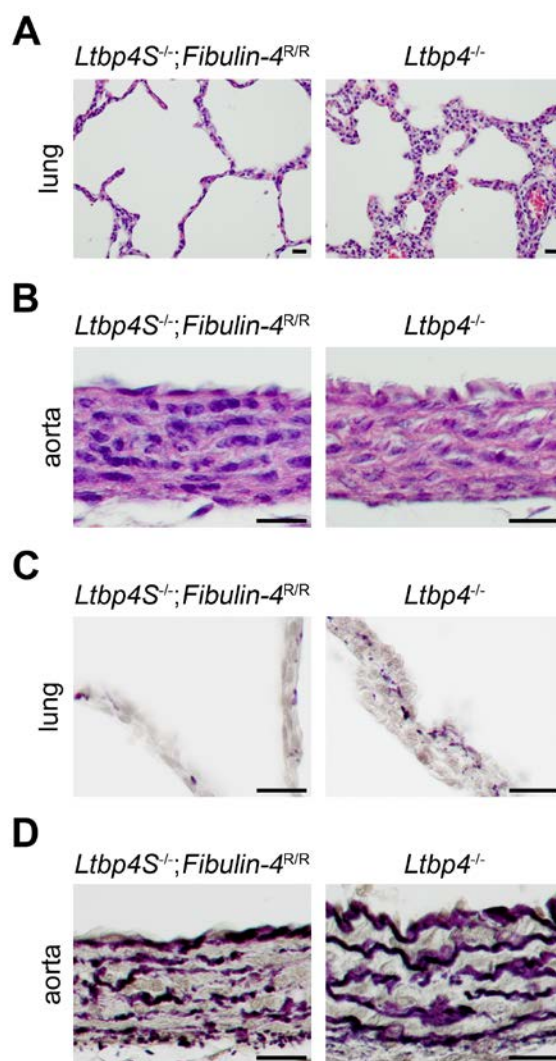
Supplementary material Fig. S1. Localization of Ltbp-4 and fibulin-4.

Representative images of Ltbp-4 immunoreactivity in (A) lungs and (B) abdominal aortas from WT, *Ltbp4S^{-/-}*, *Fibulin-4^{R/R}* and *Ltbp4S^{-/-};Fibulin-4^{R/R}* mice at P4. Representative images of fibulin-4 immunoreactivity in (C) lungs and (D) abdominal aortas from WT, *Ltbp4S^{-/-}*, *Fibulin-4^{R/R}* and *Ltbp4S^{-/-};Fibulin-4^{R/R}* mice (scale bars=20 μ m).



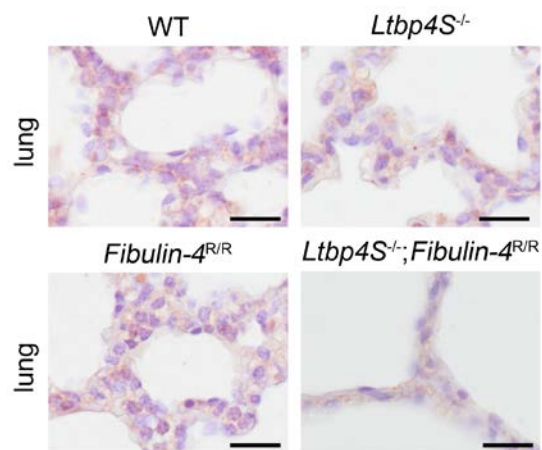
Supplementary material Fig. S2. Expression of tropoelastin and fibulin-5.

Representative immunoblots of lungs showed no differences in expression of (A) tropoelastin or (B) fibulin-5 in WT, *Ltp4S*^{-/-}, *Fibulin-4*^{RR} and *Ltp4S*^{-/-};*Fibulin-4*^{RR} mice at P4.



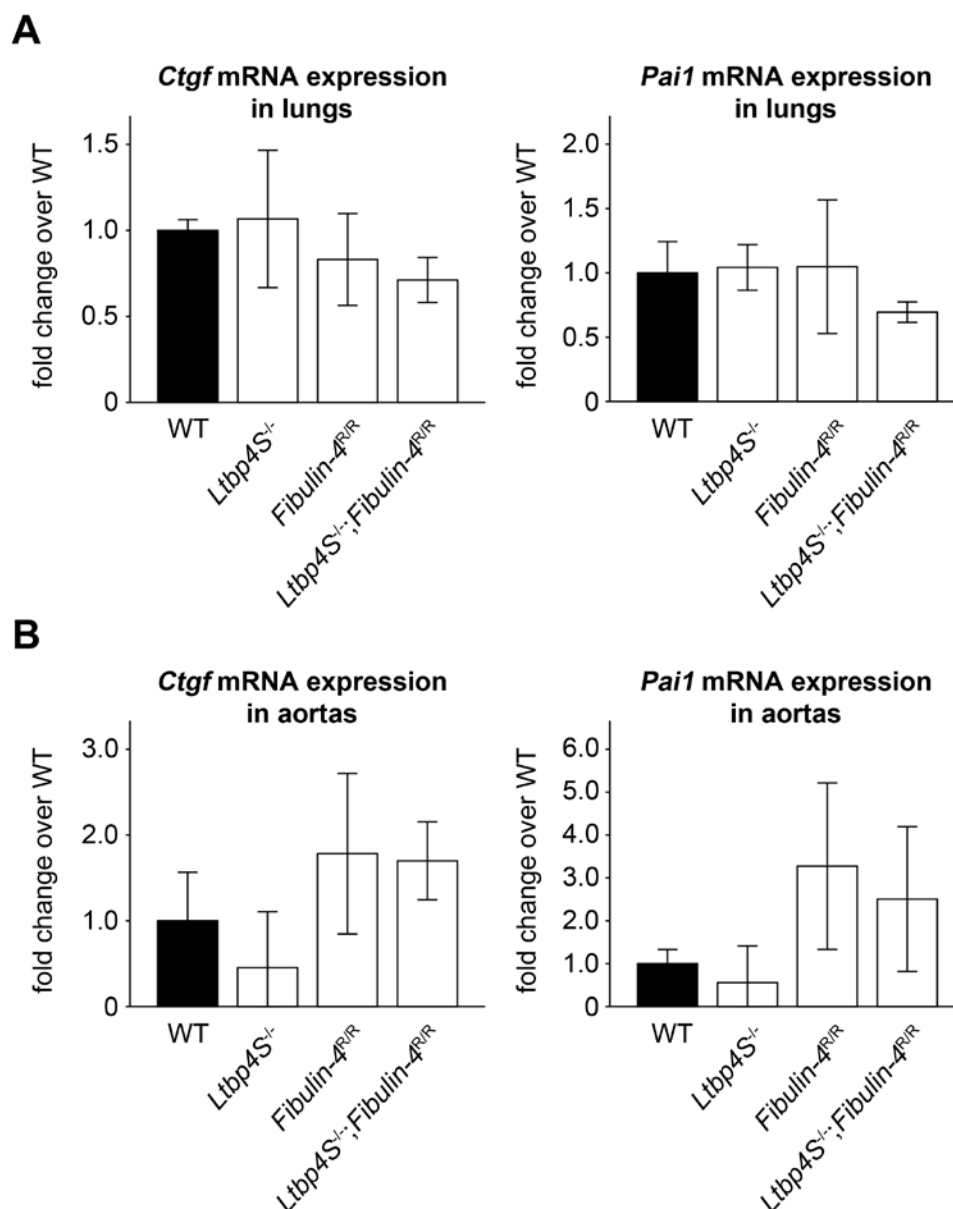
Supplementary material Fig. S3. Comparison of *Ltbp4S^{-/-};Fibulin-4^{R/R}* and *Ltbp4^{-/-}* mice at P4.

(A) Lungs from of *Ltbp4S^{-/-};Fibulin-4^{R/R}* and *Ltbp4^{-/-}* mice showed high degree of severely enlarged alveolar spaces with multifocal areas of atelectasis and a lack of alveolar and lobular architecture. (B) Abdominal aortas of *Ltbp4S^{-/-};Fibulin-4^{R/R}* and *Ltbp4^{-/-}* mice showed marked thickening of the aortic wall. Elastic fibers were completely fragmented in (C) lungs and (D) abdominal aortas of *Ltbp4S^{-/-};Fibulin-4^{R/R}* and *Ltbp4^{-/-}* mice. The pathological alterations were more severe *Ltbp4S^{-/-};Fibulin-4^{R/R}* compared to *Ltbp4^{-/-}* mice (scale bars=20 μ m).



Supplementary material Fig. S4. Localization of fibrillin-1.

Representative images of fibrillin-1 immunoreactivity in lungs from WT, *Ltp4S*^{-/-}, *Fibulin-4*^{R/R} and *Ltp4S*^{-/-}; *Fibulin-4*^{R/R} mice at P4 (scale bars=20 μ m).



Supplementary material Fig. S5. *Ctgf* and *Pai1* mRNA expression.

Ctgf and *Pai1* mRNA expression showed no differences in (A) lungs and (B) abdominal aortas from WT, *Ltbp4S*^{-/-}, *Fibulin-4*^{R/R} and *Ltbp4S*^{-/-};*Fibulin-4*^{R/R} mice (n=3-4) at P4. Data were presented as mean ± s.d.; indicated the number of analyzed tissue of individual mice. Differences between groups were analyzed by two-way ANOVA, followed by Bonferroni correction (p=n.s.).

Supplementary material Table S1. Primers.

3C7WT	5'-GGCTCATGCTTGAATGTTTCAG-3'
3C7P3	5'-CCAATCTTGCTTCTTTGCTGAGC-3'
3C7TG	5'-ATCATGCAAGCTGGTGGCTG-3'
5'Fibulin-4(mus81)	5'-TAAGTATACCCGGTGCCCA-3'
3'Fibulin-4(mus81)	5'-CGTACGCAAGCTACACGTTG-3'
NEO-Fbln4	5'-GCTATCAGGACATAGCGTTG-3'
Ltbp-4S (Primerdesign, Row nhams, Southampton, UK)	Forward 5'-GGGACCCGGCTTCCGC-3' Reverse 5'-GCAGCGATCAGGCTTCAACA-3' Probe 5'-CCTTCCTATGTCCCTTGATCTGTCAACAACGtgaagg-3'
Ltbp-4L (Primerdesign, Row nhams, Southampton, UK)	Forward 5'-GTTGCTGCCTGTCACCTGC-3' Reverse 5'-GCACCCCTCTAGACCTGTG-3' Probe 5'-CCTAGACCAAGACACCTAAGAGTAGCCGCgtctagg-3'
Efemp2	Forward 5'-TGGTGCCTACAATGCCTTTC-3' Reverse 5'-GGCGCTGACATTGTTGATTT-3'
Ctgf	Forward 5'-AGCTGACCTGGAGGAAAACA-3' Reverse 5'-GACAGGCTTGGCGATTTTAG-3'
Pai1	Forward 5'-CCCCACGGAGATGGTTATAG-3' Reverse 5'-ATCACTTGGCCCATGAAGAG-3'
Gapdh	Forward 5'-ATGTGTCCGTCGTGGATCTGA-3' Reverse 5'-TGCCTGCTTACCACCTTCT-3'

Supplementary material Table S2. Primary antibodies.

		Immunoblot	Immunohistochemistry
Ltbp-4	AF2885; R&D Systems, Germany	1:1000	1:100
Fibrillin-1	#9543, kind gift from Lynn Sakai, Shriners Hospital for Children, Portland, OR, USA	-	1:100
Fibulin-4	kind gift from Takako Sasaki, Oita University, Japan	1:2000	1:400
Fibulin-5	#3775; Epitomics, USA	1:5000	1:500
tropoelastin	PR385, EPC, USA	1:1000	-