

Figure S1: Expression of *Tll1* is not affected by *Bmp1* deletion in mouse fibroblasts. Primary mouse lung fibroblasts from *Bmp1* WT or cKO animals were treated with 4-OHT, then stimulated with TGF β for 24h. *Tll1*, *Acta2*, *Col1a1* and *Col1a2* expression were assessed by qPCR. Asterisks denote statistically significant differences (Student's unpaired t-test, *p < 0.05, **p < 0.01, ***p < 0.001).



Figure S2. Biochemical inhibition of BMP1 enzymatic activity inhibits procollagen cleavage in primary mouse lung fibroblasts. (A) Cell viability of primary mouse lung fibroblasts was measured in presence of increasing concentrations of UK383367 (vehicle: DMSO). (B) Primary mouse lung fibroblasts were stimulated with TGF β in the presence of 0 to 10 uM UK383337 or DMSO for 24h. BMP1 and CICP levels contained in supernatants were monitored by Western blot.



Figure S3: Validation of *Bmp1* and *Tll1* knockdown in mouse lung fibroblasts. CCL206 mouse lung fibroblasts transfected with *Bmp1* and/or *Tll1* siRNA were stimulated with TGF β . *Bmp1* and *Tll1* expression were assessed by RT-qPCR.



Figure S4. BMP1 depletion does not affect profibrotic gene expression in human lung fibroblasts. Human primary lung fibroblasts transfected with *BMP1* siRNA were stimulated with TGF β . Gene expression analysis of the TGF β -target genes *COL1A1*, *COL1A2*, *ACTA2* and *TNC* was performed. Asterisks denote statistically significant differences (Student's unpaired t-test, *p < 0.05, **p < 0.01, ***p < 0.001).



Figure S5. TLL1 is not required for CICP production in human primary lung fibroblasts. (A) Primary human lung fibroblasts were transfected with *BMP1* and/or *TLL1* siRNA and stimulated with TGF β for 24h. Supernatants were collected and analyzed for CICP content by Western blot. (B) Cells in (A) were collected for *BMP1* and *TLL1* expression analysis.







Medium TGF β + UK383367 (uM)

Figure S6. BMP1 biochemical inhibition does not affect profibrotic gene expression in human lung fibroblasts. (A) Human primary lung fibroblasts were incubated with increasing concentrations of UK383367 (or DMSO as vehicle) in presence of TGF β , and cell viability was assessed using the Cell Titer Glo assay. (B) Human primary lung fibroblasts treated with UK383367 were stimulated with TGF β , and gene expression analysis of the TGF β -target genes COL1A1, COL1A2, ACTA2 and FN1 was performed. Asterisks denote statistically significant differences (Student's unpaired t-test, **p < 0.01).

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Figure S7. BMP1 blocking antibodies has no toxic effect on primary mouse lung fibroblasts. Mouse lung fibroblasts CCL206 cells were stimulated with TGF β in presence of serial dilutions of BMP1 blocking antibodies or control IgG for 24h. Cell viability was assessed using the Cell-Titer Glo assay.



Figure S8. BMP1 blocking antibodies inhibit CICP production in activated primary mouse lung fibroblasts. Cells were stimulated with TGF β in presence of 1mg/ml of BMP1 blocking antibody (1E10, 6H10) or control (IgG) antibody for 24h. CICP and BMP1 levels contained in supernatants were analyzed by Western blot.