

SUPPLEMENTARY MATERIALS

Supplementary Figure 1 Evaluation of epithelial and mesenchymal phenotypes in liver sections. **[a]** Immunofluorescence for S100a4 (in green) co-stained with CK-19 (in red). Nuclei are stained with DAPI. original magn. 40X, Scale bar = 20 μm . **[b-c]** Immunofluorescence for vimentin or E-cadherin (in green) co-stained with desmin (in red). Nuclei are stained with DAPI. original magn. 40X, Scale bar = 20 μm .

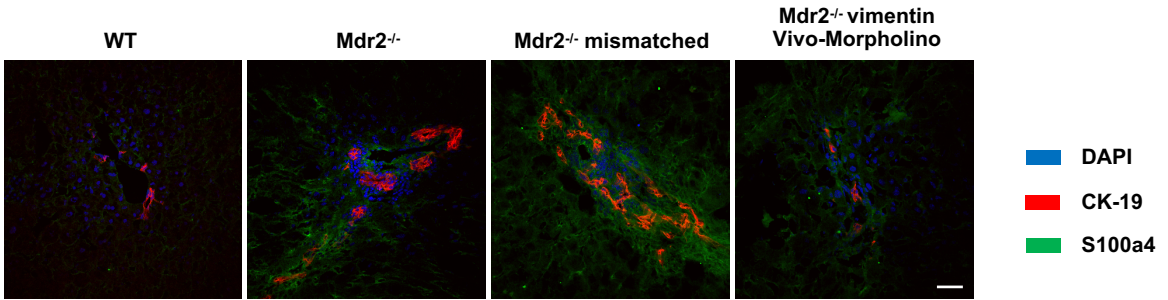
Supplementary Figure 2 Evaluation of hepatic inflammation in liver sections. Immunohistochemistry for F4/80 in liver sections, original magn. 20X, Scale bar = 100 μm . Percentage of F4/80-positive area. * $p < 0.05$ versus WT mice; # $p < 0.05$ versus Mdr2^{-/-} mice.

Supplementary Figure 3 Evaluation of angiogenesis in liver sections. Immunohistochemistry for VEGFA in liver sections, original magn. 20X, Scale bar = 100 μm . Percentage of VEGFA-positive area. * $p < 0.05$ versus WT mice; # $p < 0.05$ versus Mdr2^{-/-} mice.

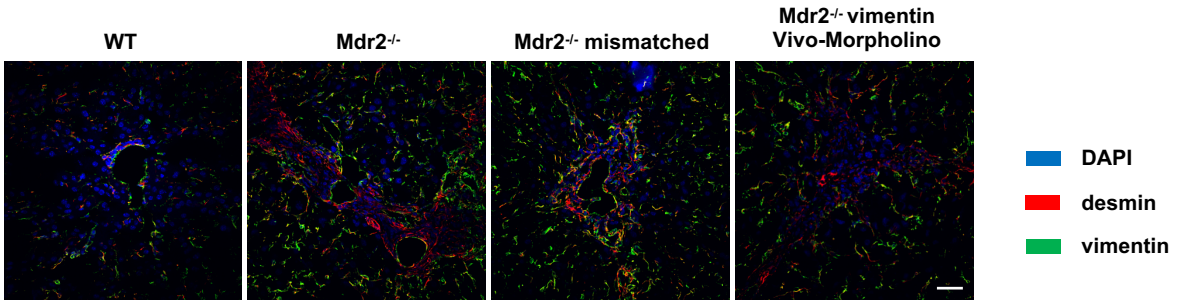
Supplementary Figure 4 Evaluation of E-cadherin expression in human PSC patients. Immunofluorescence for E-cadherin (in green) co-stained with CK-19 (in red). Nuclei are stained with DAPI. original magn. 20X, Scale bar = 10 μm .

Supplementary Figure 1

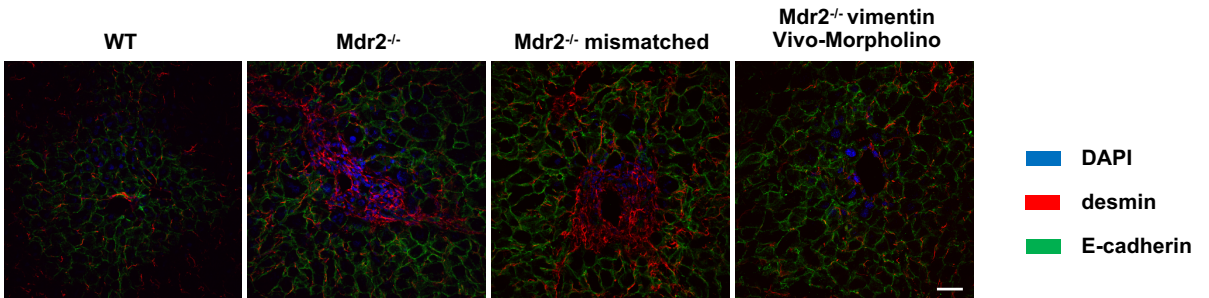
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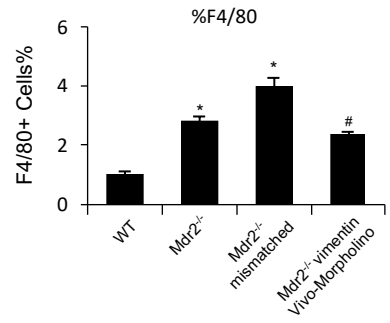
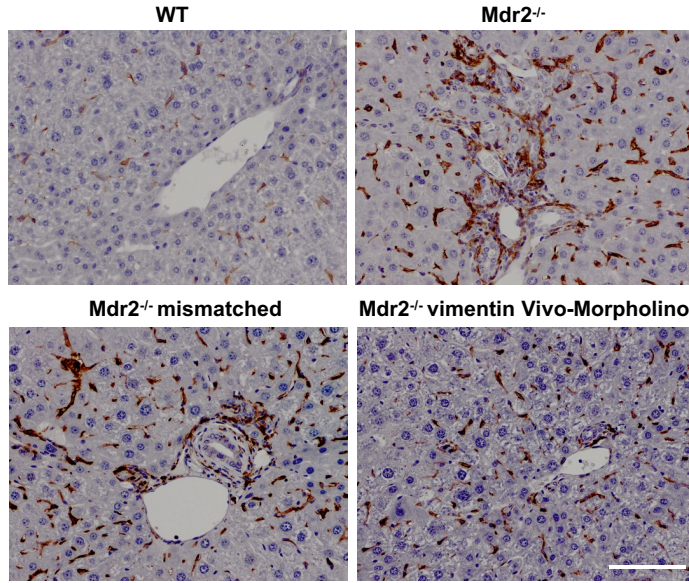
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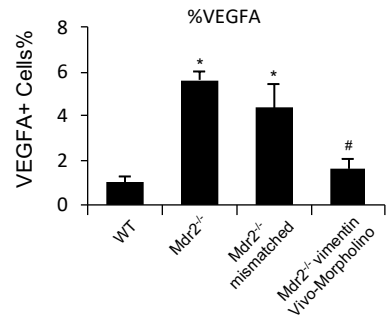
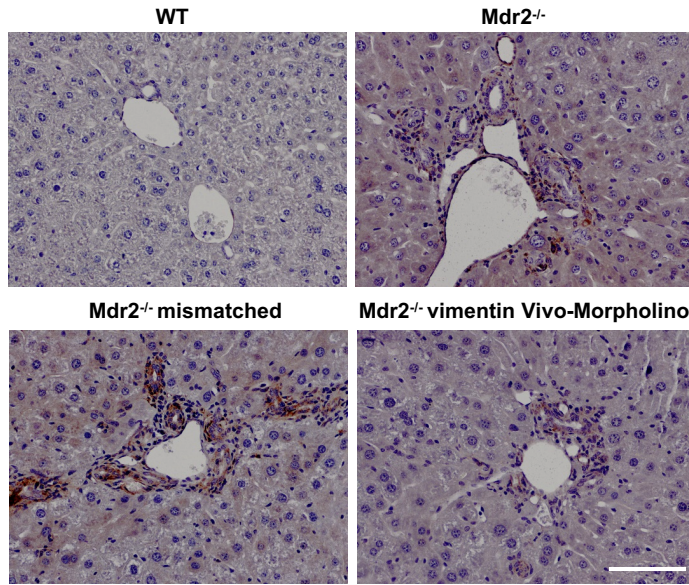
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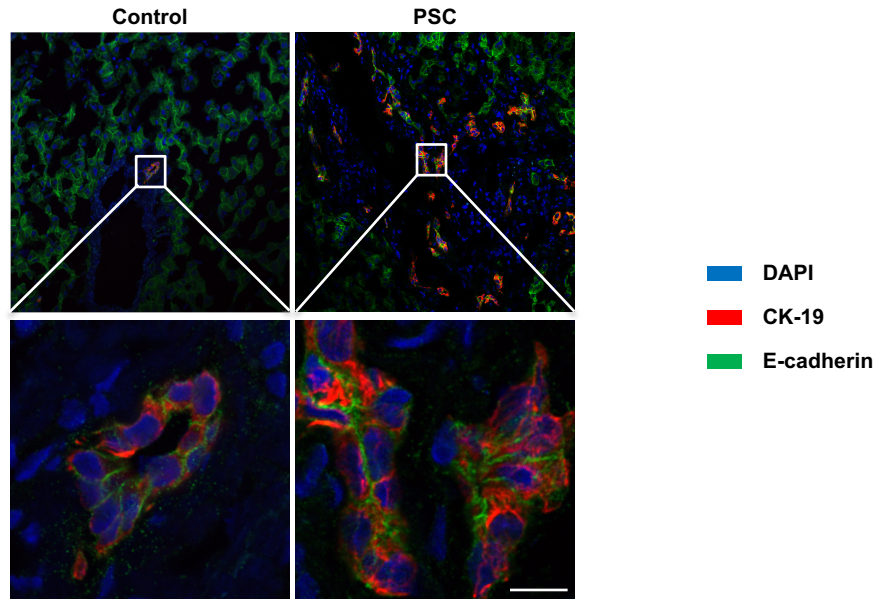
Supplementary Figure 2



Supplementary Figure 3



Supplementary Figure 4



Supplemental Table 1. List of qPCR primers

Gene	Species	Detected transcript	Source
α -SMA	Mouse	NM_007392	QIAGEN
Col1a1	Mouse	NM_007742	QIAGEN
E-cadherin	Mouse	NM_009864	QIAGEN
Fn1	Mouse	NM_010233	QIAGEN
GAPDH	Mouse	NM_008084	QIAGEN
N-cadherin	Mouse	NM_007664	QIAGEN
p16	Mouse	NM_009877	QIAGEN
p21	Mouse	NM_007669	QIAGEN
S100a4	Mouse	NM_011311	QIAGEN
TGF- β 1	Mouse	NM_011577	QIAGEN
α -SMA	Human	NM_001141945	QIAGEN
Col1 α 1	Human	NM_000088	QIAGEN
E-cadherin	Human	NM_004360	QIAGEN
Fn-1	Human	NM_002026	QIAGEN
GAPDH	Human	NM_001256799	QIAGEN
N-cadherin	Human	NM_001042572	QIAGEN
p16	Human	NM_000077	QIAGEN
p21	Human	NM_000389	QIAGEN
S100a4	Human	NM_002961	QIAGEN
TGF- β 1	Human	NM_000660	QIAGEN
vimentin	Human	NM_003380	QIAGEN

Supplemental Table 2: Characteristics of human control and PSC samples.

Group	No.	Diagnosis	Age	Gender	Ethnicity	Cirrhosis	Sample Type
Control	S1	Normal	N/A	N/A	N/A	No	Total liver
Control	S2	Normal	N/A	N/A	N/A	No	Total liver
Control	S3	Normal	N/A	N/A	N/A	No	Total liver
Control	S4	Normal	N/A	N/A	N/A	No	Total liver
Control	S5	Normal	N/A	N/A	N/A	No	Total liver
Control	S6	Normal	N/A	N/A	N/A	No	FFPE liver sections
PSC	S1	Late stage PSC	60	Male	Caucasian	N/A	Total liver
PSC	S2	Early stage PSC	N/A	N/A	Caucasian	N/A	Total liver
PSC	S3	Early stage PSC	57	Male	Caucasian	N/A	Total liver
PSC	S4	Early stage PSC	33	Male	Caucasian	N/A	Total liver
PSC	S5	Early stage PSC	47	Female	Caucasian	N/A	Total liver

PSC	S6	Early stage PSC	42	Male	Caucasian	N/A	Total liver
PSC	S7	Early stage PSC	63	Male	Caucasian	N/A	Total liver
PSC	S8	Stage I PSC	N/A	N/A	N/A	N/A	FFPE liver sections
PSC	S9	Stage IV PSC	N/A	N/A	N/A	N/A	FFPE liver sections

FFPE = formalin fixed paraffin embedded; N/A = Not available; PSC = Primary Sclerosing Cholangitis. Table outlining the characteristics of human control and PSC samples.