Abcb4-KO(FVB)

Nomenclature	FVB-*Abcb4*^{em1Smoc}
Cat. NO.	NM-KO-200584
Strain State	Embryo cryopreservation

Gene Summary

Gene Symbol Abcb4	**Synonyms**	Mdr2; Pgy2; Pgy-2; mdr-2
	NCBI ID	18670
	MGI ID	97569
	Ensembl ID	ENSMUSG00000042476
	Human Ortholog	ABCB4

Model Description

Exon 3-4 of Abcb4 gene was deleted to generate Abcb4 knockout mice.

Research Application: Diseases associated with ABCB4 include Cholestasis, Progressive Familial Intrahepatic, 3 and Gallbladder Disease 1.
*Literature published using this strain should indicate: Abcb4-KO(FVB) mice (Cat. NO. NM-KO-200584) were purchased from Shanghai Model Organisms Center, Inc..

Disease Connection

Cholecystitis	**Phenotype(s)**	MGI:3840644
	Reference(s)	Lammert F, Wang DQ, Hillebrandt S, Geier A, Fickert P, Trauner M, Matern S, Paigen B, Carey MC, Spontaneous cholecysto- and hepatolithiasis in Mdr2-/- mice: a model for low phospholipid-associated cholelithiasis. Hepatology. 2004 Jan;39(1):117-28

Cholestasis	**Phenotype(s)**	MGI:5526018
	Reference(s)	Hochrath K, Ehnert S, Ackert-Bicknell CL, Lau Y, Schmid A, Krawczyk M, Hengstler JG, Dunn J, Hiththetiya K, Rathkolb B, Micklich K, Hans W, Fuchs H, Gailus-Durner V, Wolf E, de Angelis MH, Dooley S, Paigen B, Wildemann B, Lammert F, Nussler AK, Modeling hepatic osteodystrophy in Abcb4 deficient mice. Bone. 2013 Aug;55(2):501-11
Hepatocellular Carcinoma	**Phenotype(s)**	MGI:2653844
	Reference(s)	Mauad TH, van Nieuwkerk CM, Dingemans KP, Smit JJ, Schinkel AH, Notenboom RG, van den Bergh Weerman MA, Verkruisen RP, Groen AK, Oude Elferink RP, van der Valk MA, Borst P, Offerhaus GJ, Mice with homozygous disruption of the mdr2 P-glycoprotein gene. A novel animal model for studies of nonsuppurative inflammatory cholangitis and hepatocarcinogenesis. Am J Pathol. 1994 Nov;145(5):1237-45
Intrahepatic Cholestasis	**Phenotype(s)**	MGI:3694478
	Reference(s)	Li Z, Agellon LB, Vance DE, The role of phosphatidylethanolamine methyltransferase in a mouse model of intrahepatic cholestasis. Biochim Biophys Acta. 2011 Apr;1811(4):278-83
Primary Sclerosing Cholangitis	**Phenotype(s)**	MGI:5618431
	Reference(s)	Strack I, Schulte S, Varnholt H, Schievenbusch S, Tox U, Wendland K, Steffen HM, Drebber U, Dienes HP, Odenthal M, beta-Adrenoceptor blockade in sclerosing cholangitis of Mdr2 knockout mice: antifibrotic effects in a model of nonsinusoidal fibrosis. Lab Invest. 2011 Feb;91(2):252-61

Validation Data

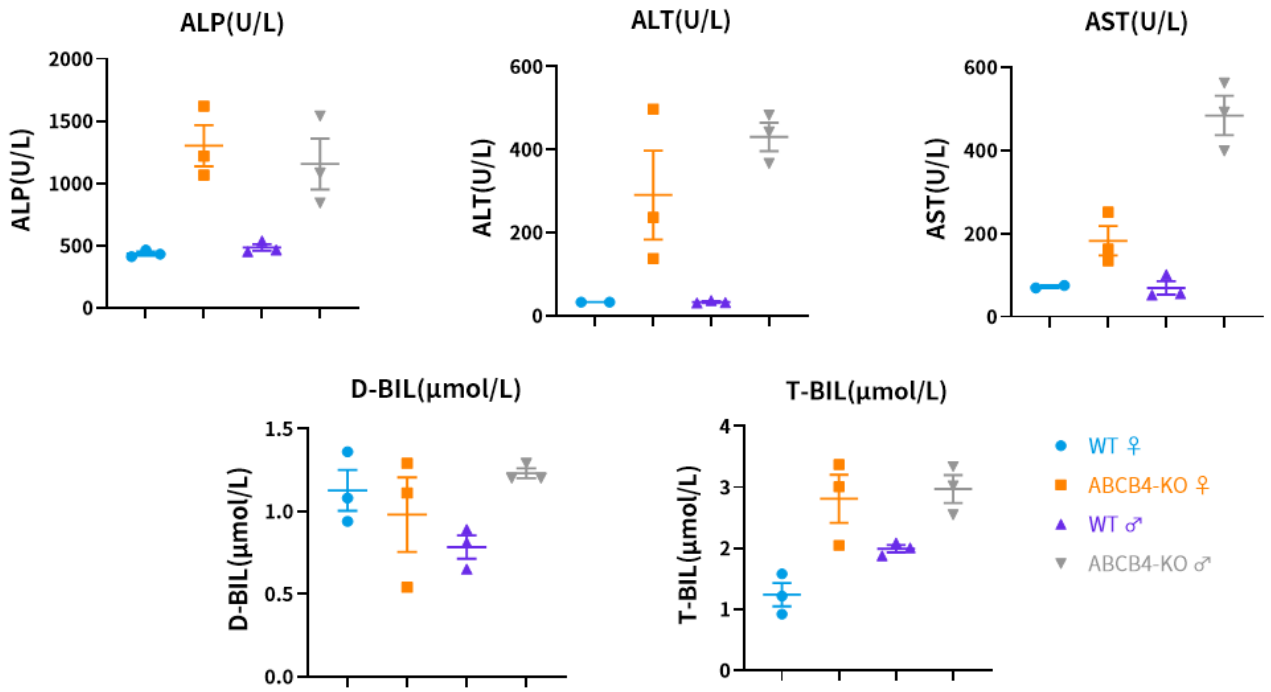
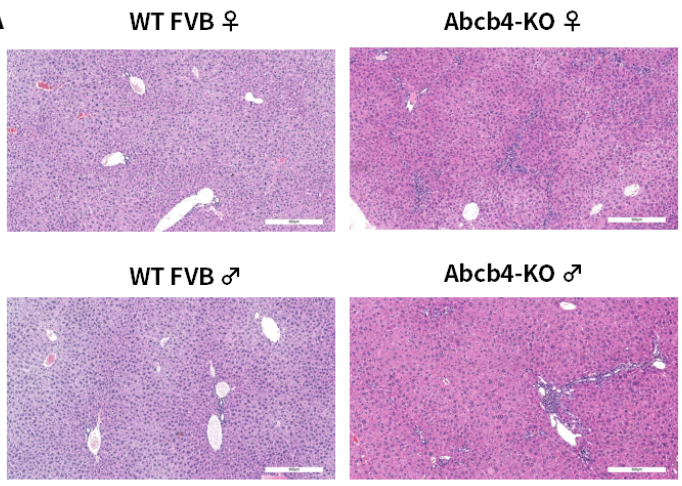


Fig1. Serum biochemistry of Abcb4-KO mice (n=3 female and 3 male).

A



B

Group	Bile duct hyperplasia	Interstitial focal inflammatory cell infiltration
WT FVB ♀	(-)	(-)
	(-)	(-)
	(-)	(-)
Abcb4-KO ♀	(+++)	(++)
	(+++)	(++)
	(+++)	(++)
WT FVB ♂	(-)	(-)
	(-)	(-)
	(-)	(-)
Abcb4-KO ♂	(+++)	(++)
	(++)	(+)
	(++)	(+)

Fig2. Significant bile duct hyperplasia and interstitial focal cell infiltration were observed in the liver of Abcb4-KO mice.

(A) Liver were collected from FVB wild-type (+/+) and Abcb4-KO mice, and analyzed by HE staining. (B) Pathological analysis of the liver in Abcb4-KO mice.

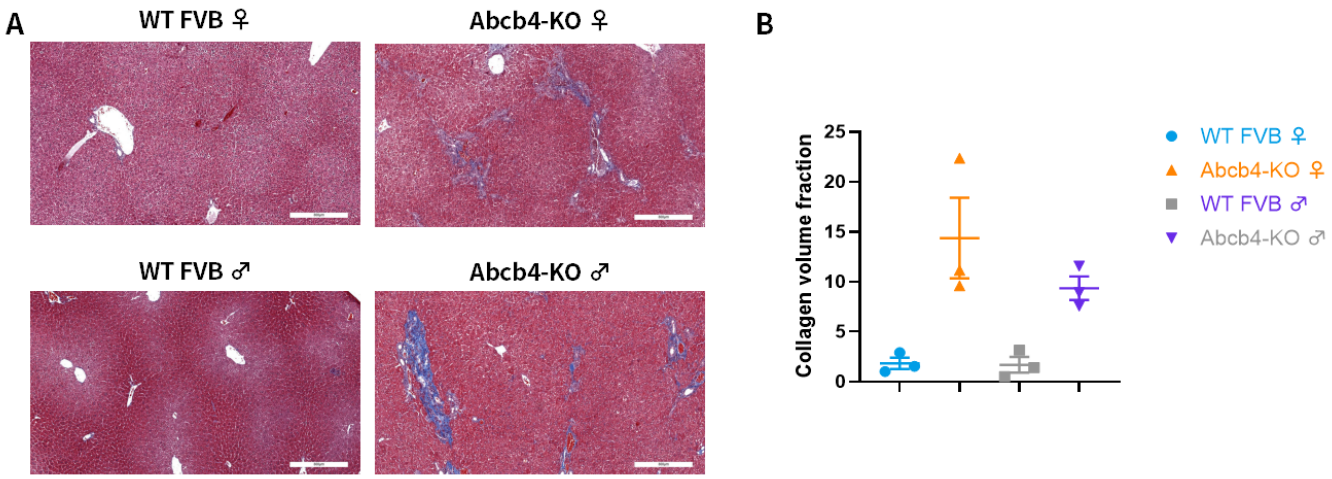


Fig3. Collagen fiber hyperplasia were observed in the liver of Abcb4-KO mice.

(A) Liver were collected from FVB wild-type (+/+) and Abcb4-KO mice, and analyzed by Masson staining. (B) Quantification of the Collagen volume fraction (CVF) .