

SUPPLEMENTARY INFORMATION

***In vitro* Rescue of the Bile Acid Transport Function of ABCB11 Variants by CFTR Potentiators**

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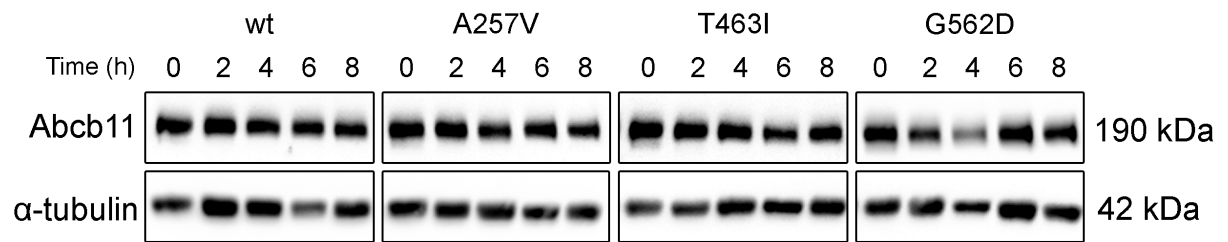
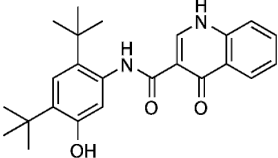
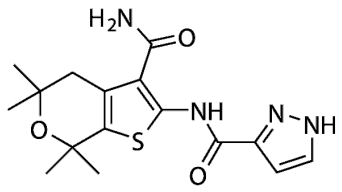
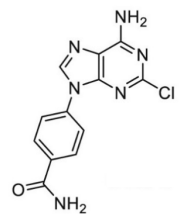
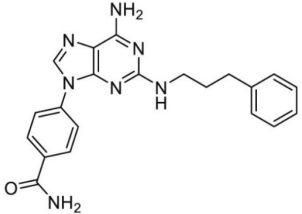


Figure S1: Stability of the Abcb11 variants. MDCK cells stably expressing Abcb11-GFP (wt or variants) were treated with cycloheximide (25 μ g/mL) to inhibit protein synthesis. Then, expression of Abcb11 (wt and variants) and α -tubulin was analysed by immunoblot using anti-GFP and anti-tubulin antibodies at the indicated time points.

Table S1: Chemical structures of CFTR potentiators tested in this study.

Compound	IUPAC Name	Chemical structure	PubChem CID	MW (Da)	References
Ivacaftor (VX-770)	N-(2,4-ditert-butyl-5-hydroxyphenyl)-4-oxo-1H-quinoline-3-carboxamide		16220172	392.49	[1]
GLPG1837 (ABBV-974)	N-(3-carbamoyl-5,5,7,7-tetramethyl-4H-thieno[2,3-c]pyran-2-yl)-1H-pyrazole-5-carboxamide		117857370	348.42	[2]
SBC040	4-(6-Amino-2-chloro-9H-purin-9-yl)benzamide		—	403.7	[3]
SBC219	4-(6-Amino-2-((3-phenylpropyl)amino)-9H-purin-9-yl)benzamide		—	387.4	[3]

Supplementary References

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