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Name of Journal: *World Journal of Gastroenterology*

ESPS Manuscript NO: 28607

Manuscript Type: REVIEW

Current and future therapies for inherited cholestatic liver diseases

van der Woerd WL *et al.* Therapies for inherited cholestatic liver disease

Wendy L van der Woerd, Roderick HJ Houwen, Stan FJ van de Graaf

### Abstract

Familial intrahepatic cholestasis (FIC) comprises a group of rare cholestatic liver diseases associated with canalicular transport defects resulting predominantly from mutations in *ATP8B1*, *ABCB11* and *ABCB4*. Phenotypes range from benign recurrent intrahepatic cholestasis (BRIC), associated with recurrent cholestatic attacks, to progressive FIC (PFIC). Patients often suffer from severe pruritus and eventually progressive cholestasis results in liver failure. Currently, first-line treatment includes

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