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PEER-REVIEW REPORT

Name of journal: *World Journal of Hepatology*

Manuscript NO: 68421

Title: Recent updates on progressive familial intrahepatic cholestasis types 1, 2 and 3: Outcome and therapeutic strategies

Provenance and peer review: Invited manuscript; Externally peer reviewed

Peer-review model: Single blind

Reviewer's code: 03662585
Position: Peer Reviewer
Academic degree: MD

Professional title: Doctor

Reviewer's Country/Territory: Egypt

Author's Country/Territory: India

Manuscript submission date: 2021-05-22

Reviewer chosen by: AI Technique

Reviewer accepted review: 2021-05-29 06:26

Reviewer performed review: 2021-05-29 06:35

Review time: 1 Hour

Scientific quality	[ ] Grade A: Excellent [Y] Grade B: Very good [ ] Grade C: Good [ ] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[Y] Grade A: Priority publishing [ ] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[ ] Accept (High priority) [Y] Accept (General priority) [ ] Minor revision [ ] Major revision [ ] Rejection
Re-review	[ ] Yes [Y] No

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Peer-reviewer statements	Peer-Review: [Y] Anonymous [ ] Onymous Conflicts-of-Interest: [ ] Yes [Y] No

SPECIFIC COMMENTS TO AUTHORS

A nice review of progressive familial cholestasis which is well written

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Reviewer's code: 04736766
Position: Peer Reviewer
Academic degree: MBBS, MRCP

Professional title: Doctor

Reviewer's Country/Territory: United Kingdom

Author's Country/Territory: India

Manuscript submission date: 2021-05-22

Reviewer chosen by: AI Technique

Reviewer accepted review: 2021-05-31 21:08

Reviewer performed review: 2021-06-08 07:41

Review time: 7 Days and 10 Hours

Scientific quality	[ ] Grade A: Excellent [ ] Grade B: Very good [ ] Grade C: Good [Y] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[ ] Grade A: Priority publishing [Y] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[ ] Accept (High priority) [ ] Accept (General priority) [ ] Minor revision [Y] Major revision [ ] Rejection
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SPECIFIC COMMENTS TO AUTHORS

This article covers a wide remit of the genetic basis, natural history and clinical outcomes of PFIC 1,2,3 and finally the current and future treatment strategies. In the first section on ATP8B1 deficiency, the severe and mild phenotypes are described in turn. The subsection on 'clinical outcomes' is therefore an overlap and should be integrated into the subsections above. Similarly when describing BSEP and MDR3 there is overlap between clinical presentation and outcomes. Please structure these sections conventionally and distinctly - pathogenesis, presentation, natural history / prognosis etc. At present this information seems intermingled. Please avoid abbreviations in section headings (ICP,LPAC etc). This section is not well explained but is worthwhile to describe the more frequently encountered clinical scenarios of ICP, DILI in adults, the associations with genetic abnormalities of cholestasis genes, and future chronic liver disease. The term used in Table 1 "expanded role in childhood/ adulthood" is not clear - description such as 'associations with other cholestatic presentations" would be preferable. The article then comprehensively reviews different treatment strategies, including novel therapy targets. The outcomes of biliary diversion surgery in terms of survival and need for liver transplant should be described. The NAPPED cohort study is a key reference. Table 2 should be a summary and detailed description of the MOA of each drug which was already included in the main text is unnecessary. Table 4 could be supplemented by a diagram to show the therapeutic targets involved. Some of the included references are not central to the review, e.g. 34, 36, 39. Overall this article seems to lack focus and is not clear to read due to the current structuring. If the remit is recent updates on the understanding of the genetic variants and associated phenotype,

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the article should start with a succinct overview of the clinical presentation followed by the most clinically relevant updates. Regarding treatments, established strategies and anti-pruritic agents do not need extensive description but a summary table with an overview in the text would suffice, to focus on the newer agents. The article should end with a conclusion. There are some typographic / grammatical errors throughout.