

# World Journal of *Gastroenterology*

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Editorial board member of *World Journal of Gastroenterology*, Yasemin H Balaban, MD, Doctor, Professor, Department of Gastroenterology, Hacettepe University, Ankara 06100, Turkey

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## Letter to the editor: Diagnosis of erythropoietic protoporphyria with severe liver injury - a case report

Debby Wensink, Margreet AEM Wagenmakers, JH Paul Wilson, Janneke G Langendonk

**ORCID number:** Debby Wensink (0000-0002-7249-5810); Margreet AEM Wagenmakers (0000-0003-2587-0283); JH Paul Wilson (0000-0001-6995-7044); Janneke G Langendonk (0000-0002-4899-7239).

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**Debby Wensink, Margreet AEM Wagenmakers, JH Paul Wilson, Janneke G Langendonk,** Porphyria Center Rotterdam, Center for lysosomal and metabolic disease, Department of Internal Medicine, Erasmus University Medical Center, Rotterdam 3015 GD, Netherlands

**Corresponding author:** Janneke G Langendonk, MD, PhD, Internist, Department of Internal Medicine, Erasmus University Medical Center, Dr. Molenwaterplein 40, Rotterdam 3015 GD, Netherlands. [j.langendonk@erasmusmc.nl](mailto:j.langendonk@erasmusmc.nl)

**Telephone:** +31-10-7035191

**Fax:** +31-10-7033639

### Abstract

Erythropoietic protoporphyria (EPP) is an extremely rare disease which is often unrecognized as diagnosis. In the recent article Lui *et al* describe a patient with a new diagnosis of EPP with severe liver injury. Approximately 5%-20% of patients with EPP develop liver manifestations. The most severe complication of EPP is an hepatic crisis, which is a medical emergency requiring urgent treatment. Intensive treatment should consist of (exchange) transfusions and preferably in a center that performs liver transplantations.

**Key words:** Erythropoietic protoporphyria; Liver disease; Blood transfusion; Protoporphyrin IX; Treatment

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**Core tip:** Erythropoietic protoporphyria (EPP) can be complicated by severe liver injury. The major highlight of this letter is to stress that an EPP hepatic crisis has to be recognized as it can be fatal, and the best therapeutic options are (exchange) transfusions and preferably in a center that performs liver transplantations.

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## TO THE EDITOR

With great interest we read the case report on the diagnosis of erythropoietic protoporphyria (EPP) with severe liver injury by Liu *et al*<sup>[1]</sup> in the February 2019 issue of *World Journal of Gastroenterology*. They report a case presenting with weight loss, jaundice, and hepatitis. After extensive investigation the diagnosis of EPP was made and the liver disease was explained as related to EPP. We greatly appreciate the dedication of the authors to improve awareness of EPP and EPP related liver injury in order to aid early recognition.

In addition to their paper, we like to propose treatment options for severely ill patients. Approximately 5%-20% of patients with EPP develop liver manifestations<sup>[2]</sup>. Hepatic manifestations of the disease are diverse, ranging from mildly disturbed liver enzymes, choledocholithiasis at early age to fatal hepatic failure. The case of Liu *et al* reports elevated transaminases, hyperbilirubinemia, enlargement of the liver and ascites on magnetic resonance imaging (MRI) suggesting severe liver disease. The most severe complication of EPP, which can rapidly result in fatal hepatic failure, is cholestatic liver failure, which we, by analogy with the hepatic crises of sickle cell disease, designate as an EPP hepatic crisis, to accentuate that this condition is a medical emergency requiring urgent treatment. These EPP crises present with severe abdominal pain, jaundice, and biochemically severe cholestatic hepatitis, accompanied by a significant increase in erythrocyte and plasma protoporphyrin IX over preexisting levels. During a hepatic crisis, which is often provoked when the liver is damaged by another cause such as alcohol consumption or viral hepatitis, the biliary excretion of protoporphyrin is reduced, leading to a rise of protoporphyrin levels which may further damage the canaliculi and biliary tract. This results in a vicious circle causing further liver damage and rapidly leads to acute liver failure if left untreated. In case of a hepatic crisis the suggested treatment of Liu *et al*, consisting of avoiding light, drinking glucose water, and a carrot diet including  $\beta$  carotene, resulted in recovery of their patient. We suggest a more intensive treatment in severely ill patients to prevent possible fatal outcome. The cornerstone of this therapy is to quickly lower protoporphyrin levels in order to provide an opportunity for the liver to recover. This can be achieved with exchange transfusions<sup>[3]</sup>. In addition, suppression of hematopoiesis and endogenous protoporphyrin production via blood transfusions to supraphysiological hemoglobin levels should be considered<sup>[4]</sup>. If this treatment fails, liver transplantation is the only treatment option left<sup>[5]</sup>. Because of the severity of EPP-related liver crises, prevention of liver damage should be attempted in all EPP patients, by advising against and restricting alcohol consumption, avoiding potentially hepatotoxic drugs and vaccinating against hepatitis A and B virus<sup>[6]</sup>.

To conclude, a severe EPP hepatic crisis can be fatal and should be treated intensively *via* (exchange) transfusions and preferably in a center that performs liver transplantations.

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