

Partial External Biliary Diversion in BSEP deficiency: association between outco

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[ATP8B1 Deficiency - GeneReviews® - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK1297/)

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15 Oct 2001 ... In severe disease, **partial external biliary diversion** (PEBD) surgery may reduce pruritus. ... and length of disease-free intervals **between** episodes vary greatly. ... Clinical Biochemistry Test **Results** in ATP8B1 **Deficiency** Instances of paraneoplastic cholestasis **associated** with **mutation** of ATP8B1 are not ...

[Progressive Familial Intrahepatic Cholestasis](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4017198/)

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23 Nov 2013 ... PFIC1: It is also known as Byler disease and is **associated** with ... **deficiency** and the protein function is only partially impaired in them. ... PFIC2: This disease was previously known as Byler's syndrome6 and is a **result** of **mutation** in the **Partial external biliary diversion** (PEBD): this can be performed with ...

[Differences in presentation and progression between severe FIC1 ...](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3042805/)

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13 Apr 2010 ... **Mutations** in ATP8B1 or ABCB11 can **result** in hereditary cholestasis [1–3]. ... Our **results** also indicate that while **BSEP deficiency** is primarily a liver ... Given the high **correlation** (0.78) **between** AST and ALT measured in the same patient and short-term **outcome** after **partial external biliary diversion** in 13 ...

[\[DOC\] WJT-6-278.doc - World Journal of Gastroenterology](http://www.wjgnet.com/esps/DownloadFile.aspx?Type=InPress&FilePath...)

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24 Feb 2016 ... These **mutant** proteins **result** in the impairment of bile flow through the However, in cases of PFIC1, liver transplantation can be **associated** with an **Partial external biliary diversion** which was first described by Differences in presentation and progression **between** severe FIC1 and BSEP **deficiencies**.

[Genetics and Molecular Modeling of New **Mutations** of Familial ...](#)

Name of journal: *World Journal of Gastroenterology*

Manuscript NO: 32316

Manuscript Type: ORIGINAL ARTICLE

Basic Study

Partial external biliary diversion in bile salt export pump deficiency: Association between outcome and mutation

Philipp Ellinger, Jan Stindt, Carola Dröge, Katharina Sattler, Claudia Stross, Stefanie Kluge, Diran Herebian, Sander H J Smits, Martin Burdelski, Sebastian Schulz-Jürgensen, Antje Ballauff, Jan Schulte am Esch, Ertan Mayatepek, Dieter Häussinger, Ralf Kubitz, Lutz Schmitt

Abstract

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Partial external biliary diversion in bile salt export pump deficiency: Association b

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23 Nov 2013 ... The basic defect is impaired bile salt secretion in PFIC1/2 whereas in PFIC3, ... PBD, **partial biliary** drainage; PEBD, **partial external biliary** drainage; PFIC, ... PFIC1: It is also known as Byler disease and is **associated** with **bile salt export pump deficiency**: 82 different ABCB11 **mutations** in 109 families.

[Liver transplantation and the management of progressive familial ...](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4919732/)

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4919732/>

24 Jun 2016 ... **Mutation** of this protein significantly impairs bile salt secretion..... infants, is important since options for optimal **treatment** may differ **between** subtypes. **External biliary diversion** post-transplantation in patients with PFIC1 who are Recurrence of **bile salt export pump deficiency** after liver transplantation.

[Severe Bile Salt Export Pump Deficiency: 82 ... - Gastroenterology](http://www.gastrojournal.org/article/S0016-5085(08)00112-1/fulltext)

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Bile salt export pump (BSEP) **deficiency** is caused by **mutations** in ABCB11..... 20, 21, 22 **Partial external biliary diversion** and ileal exclusion can relieve pruritus ... In 7 families, clinical **outcome** and/or immunohistochemical results from a deceased aDenotes studies in which **mutations** are **associated** with mild disease.

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histochemistry and clinical **outcome**. Results: ... tified in 109 families (9 nonsense **mutations**, 10 small ... **Bile salt export pump** (BSEP) **deficiency** is caused by ... stage liver disease before adulthood.20 –22 **Partial external biliary diversion** and ileal exclusion can relieve pruritus sessed differences **between** groups.

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作者: A Srivastava - 2014 - 被引用次数: 45 - 相关文章

2013年11月23日 - The basic defect is impaired bile salt secretion in PFIC1/2 whereas in PFIC3, ... PBD, **partial biliary** drainage; PEBD, **partial external biliary** drainage; PFIC, ... PFIC1: It is also known as Byler disease and is **associated** with **bile salt export pump deficiency**: 82 different ABCB11 **mutations** in 109 families.

Liver transplantation and the management of progressive familial ...

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2016年6月24日 - **Mutation** of this protein significantly impairs bile salt secretion. infants, is important since options for optimal **treatment** may differ **between** subtypes. **External biliary diversion** post-transplantation in patients with PFIC1 who are Recurrence of **bile salt export pump deficiency** after liver transplantation.

Biliary atresia and other cholestatic childhood diseases: advances and ...

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作者: HJ Verkade - 2016 - 被引用次数: 14 - 相关文章

2016年4月27日 - Keywords: **biliary** atresia, Alagille syndrome, **biliary diversion**, and tracking BA **outcomes** and their predictors [25]. Table 1 ... **between** the **treatment** and control groups. When medical **treatment** fails **external partial biliary diversion** may be under five years of age with **bile salt export pump deficiency**.

Severe Bile Salt Export Pump Deficiency: 82 ... - Gastroenterology

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作者: SS Strautnieks - 2008 - 被引用次数: 233 - 相关文章