

**Name of Journal:** *World Journal of Gastroenterology*

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**Risk of malignancy** <sup>1</sup> **in Caroli disease and syndrome: A systematic review**

**Fahrner** <sup>1</sup> **R et al. Malignancy in Caroli disease and syndrome**

René **Fahrner**, Sandra G.C. Dennler, Daniel Inderbitzin

## Abstract

### BACKGROUND

<sup>1</sup> **Congenital intrahepatic bile duct dilatation** without fibrosis **is** called Caroli **disease** (CD), and is called Caroli syndrome (CS) when it has fibrotic and cirrhotic liver morphology. The development of intrahepatic carcinoma is described in both conditions, but the reported incidence varies extensively. Potential risk factors for the malignant transformation were not described. Furthermore, conservative or surgical

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### Systemic Sclerosis and Malignancy: A Review of Current Data

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4974831>

Systemic sclerosis (SSc) is associated with increased risk of malignancy. The organ systems most commonly affected are the lungs, the breasts and the hematological system. Risk factors predisposing a SSc patient for development of malignancy are not well defined, and the pathogenic basis of the association is yet to be explained.

Cited by: 19 Author: Nabil Zeineddine, Lara El Khoury, Joseph M...  
Publish Year: 2016

### Search Tools

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### Cancer increases risk of in-hospital death from COVID-19 ...

<https://www.nature.com/articles/s41375-020-0986-7>

Jul 20, 2020 · The impact of cancer on outcome of persons with coronavirus disease 2019 (COVID-19) after infectory syndrome coronavirus-2 ...

### Caroli disease | Genetic and Rare Diseases Information ...

<https://rarediseases.info.nih.gov/diseases/6002/caroli-disease>

Jul 19, 2017 · Caroli syndrome is often associated with an inherited condition called autosomal recessive polycystic kidney disease. People affected by Caroli disease and Caroli syndrome may also have an increased risk of developing cholangiocarcinoma.

#### PEOPLE ALSO ASK

- What is the prognosis of Caroli disease?
- Is Caroli disease genetic?
- Is metabolic syndrome associated with cancer?
- What are the symptoms of Caroli syndrome?

Feedback

### Polycystic ovary syndrome (PCOS) and the risk of coronary ...

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5085114>

Jun 07, 2016 · Polycystic ovary syndrome (PCOS) is one of the most common cause of infertility in women population . Around 4% of unselected population of reproductive age and 7% of the caucasian population is believed to have this syndrome . It is reported that PCOS patients had increased risk of many diseases, such as diabetes and infertility .

Cited by: 58 Author: Luqian Zhao, Zhigang Zhu, Huiling Lou, Gu...  
Publish Year: 2016

### Risk of metachronous colorectal cancer following colectomy ...

<https://onlinelibrary.wiley.com/doi/abs/10.1111/codi.13679>

Apr 13, 2017 · Lynch syndrome (LS) accounts for 2–4% of all colorectal cancer (CRC) cases, and is associated with an increased risk of developing metachronous colorectal cancer (mCRC). The role of extended colectomy in LS CRC is controversial. There are limited studies comparing the risk of mCRC following segmental colectomy and extended colectomy.

Cited by: 17 Author: C. C. Anele, S. O. Adegbola, A. Askari, A. Ra...  
Publish Year: 2017

### Caroli Disease - NORD (National Organization for Rare ...

<https://rarediseases.org/rare-diseases/caroli-disease>

Classification	Genetics	Purpose	Availability
Caroli disease is a rare inherited disorder characterized by abnormal widening (dilatation) of the ducts that carry bile from the liver (intrahepatic bile ducts). According to the medical literature, there are two forms of Caroli disease. In most cases, the isolated or simple form is characterized by widening of the bile ducts (dilatation or ectasia). A second, more complex form is often called Caroli syndrome. The complex form or syndrome is associated with the presence of bands of fibrous tissue in the liver (conge... <a href="https://rarediseases.org">See more on rarediseases.org</a>			

### High Cancer Risk in Peutz-Jeghers Syndrome: A Systematic ...

<https://pubmed.ncbi.nlm.nih.gov/20051941>

Objectives: Peutz-Jeghers syndrome (PJS) is an autosomal dominant inherited disorder associated with increased cancer risk. Surveillance and patient management are, however, hampered by a wide range in cancer risk estimates. We therefore performed a systematic review to assess cancer risks in PJS patients and used these data to develop a surveillance recommendation.

### Metabolic Syndrome and Cancer - PubMed Central (PMC)

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3191378>

Obesity, which is a component of metabolic syndrome, has been associated with high risks of cancers at the multiple sites. Accumulating epidemiologic evidence shows that excess body weight is associated with an increased risk of several common adult cancers.

### Metabolic Syndrome and Risk of Cancer: A systematic review ...

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3476894>

Oct 13, 2012 · Metabolic syndrome is associated with increased risk of common cancers; for some cancers, the risk differs between sexes, populations, and definitions of metabolic syndrome. The metabolic syndrome is a cluster of risk factors for cardiovascular disease and type 2 diabetes and constitutes a growing problem worldwide (1).

### Primary Sjögren's syndrome and malignancy risk: a ...

<https://ard.bmj.com/content/73/6/1151>

Objective To investigate the association between primary Sjögren's syndrome (pSS) and the risks of malignancy including overall malignancy and site-specific malignancies through a systematic review and meta-analysis. Methods We searched Pubmed before January 2013, with a restriction to English language publications. Studies were included if they met the following criteria: (1) a cohort or ...

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### [Systemic Sclerosis and Malignancy: A Review of Current Data](#)

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4974831>

**Systemic** sclerosis (SSc) is associated with increased **risk of malignancy**. The organ systems most commonly affected are the lungs, the breasts and the hematological system. **Risk** factors predisposing a SSc patient for development of **malignancy** are not well defined, and the pathogenic basis of the association is yet to be explained.

**Cited by:** 19      **Author:** Nabil Zeineddine, Lara El Khoury, Joseph...  
**Publish Year:** 2016

### [PDF] [Caroli's disease: Identification and treatment strategy](#)

[www.biliaryatresia.org/reference/Caroli's disease\\_ Identification and treatment...](http://www.biliaryatresia.org/reference/Caroli's_disease_Identification_and_treatment...)

**Caroli's disease** has been reported in associa-tion with Lawrence-Moon-Biedl **syndrome** [16]. **Systemic** amyloidosis has also been reported in patients with **Caroli's disease**, possibly related to **systemic** inflammation from recurrent episodes of cholangitis [17]. Diagnosis Cholangiography The gold standard for diagnosis of **Caroli's disease** has tra-

**Cited by:** 24      **Author:** Ashwin N. Ananthakrishnan, Kia Saeian  
**Publish Year:** 2007

### [Caroli disease | Genetic and Rare Diseases Information ...](#)

<https://rarediseases.info.nih.gov/diseases/6002/caroli-disease> ▾

Jul 19, 2017 - Caroli syndrome is often associated with an inherited condition called autosomal recessive polycystic kidney disease. People affected by Caroli disease and Caroli syndrome may also have an **increased risk of developing cholangiocarcinoma**.

#### PEOPLE ALSO ASK

What is the prognosis of Caroli disease? ▾

Is Caroli disease genetic? ▾

What is the treatment for Caroli disease? ▾

Is Caroli disease more common in males or females? ▾