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Name of Journal: *World Journal of Gastroenterology***Manuscript NO:** 53539**Manuscript Type:** REVIEW**Review of primary sclerosing cholangitis with increased IgG4 levels**Manganis CD *et al.* PSC and high IgG4

Charis D Manganis, Roger W Chapman, Emma L Culver

Abstract

Primary sclerosing cholangitis (PSC) is a chronic progressive liver disease. Subtypes of PSC have been described, most recently PSC with elevated serum and/or tissue IgG4 subclass. We aim to summarise the clinical phenotype, disease associations, differential diagnosis, response to therapy and pathogenic mechanisms underlying PSC-high IgG4 subtype. We reviewed PubMed, MEDLINE and Embase with the search terms "primary sclerosing cholangitis", "IgG4", and "IgG4-related sclerosing cholangitis (IgG4-SC)". Elevated serum IgG4

Match Overview

1	Crossref 56 words "Biliary Disease", Springer Science and Business Media LLC, 2017	1%
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Primary sclerosing cholangitis with increased IgG4 levels: a dis



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Diagnosis of IgG4-related sclerosing cholangitis

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3837265>

Nov 21, 2013 · **IgG4-related sclerosing cholangitis (IgG4-SC)** is a characteristic type of **sclerosing cholangitis**, with an unknown pathogenic mechanism. Patients with **IgG4-SC** display **increased serum IgG4 levels**[1] and dense infiltration of **IgG4-positive** plasma cells ...

Cited by: 81

Author: Takahiro Nakazawa, Itaru Naitoh, Kazuki ...

Publish Year: 2013

Primary sclerosing cholangitis: a clinical update ...

<https://academic.oup.com/bmb/article/114/1/53/246353> ▼

May 16, 2015 · **Primary sclerosing cholangitis (PSC)** is a progressive **cholestatic disorder** that ultimately can lead to cirrhosis, liver failure, malignancy and death. It is strongly associated with inflammatory bowel disease (IBD), and though a rare disease, its **incidence is increasing**. There are no proven medical therapies for PSC.

Cited by: 21

Author: Kate D. Williamson, Kate D. Williamson, ...

Publish Year: 2015

IgG4-related sclerosing cholangitis Culver 2017



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IgG4-Related Sclerosing Cholangitis and Primary Sclerosing ...

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6529173>

Sep 13, 2018 - As described, **serum IgG4 level** was elevated in 12.5% of patients with **PSC** (Fig. 2B). Histopathologically, an obliterative, **non-suppurative cholangitis** with substantial periductular fibrosis, referred as "**onion-skin fibrosis**," is a typical finding, although ...

Cited by: 1 Author: Atsushi Tanaka

Publish Year: 2019

Diagnosis of IgG4-related sclerosing cholangitis

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3837265>

Nov 21, 2013 - The patients with elevated IgG4 levels had higher levels of **total bilirubin** and **alkaline phosphatase**, higher **PSC Mayo risk scores**, and lower incidence of **IBD**. It is important to note that the time to **liver transplantation** was shorter in the patients with elevated IgG4 levels (1.7 years vs 6.5 years, $P = 0.0009$). As only one of the patients in their series had an abnormal pancreatogram, the ...

Cited by: 81 Author: Takahiro Nakazawa, Itaru Naitoh, Kazuki ...

Publish Year: 2013

Primary sclerosing cholangitis with increased ...

<https://www.ncbi.nlm.nih.gov/pubmed/31852163>

RATIONALE: **Primary sclerosing cholangitis** (PSC) is recognized as an autoimmune-mediated liver disease characterized by progressive biliary inflammation and fibrosis. Some PSC cases **with elevated immunoglobulin G4 (IgG4) levels** are likely to be misdiagnosed with immunoglobulin G4-related **sclerosing cholangitis (IgG4-SC)**.

Author: Qing-Ling Chen, Rui Zhong, Xiao-Xue... Publish Year: 2019

Primary sclerosing cholangitis: A review and update

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6028044>

Abbreviations: **AIDS**, acquired immune **deficiency syndrome**; **CMV**, **cytomegalovirus**; **IgG4**, immunoglobulin G **subclass 4**; **PSC**, **primary sclerosing cholangitis**. A small proportion of patients with **PSC** present with normal serum alkaline phosphatase (ALP); 40 therefore, normal ALP should not diagnosis of **PSC** or discourage further investigation if other clinical data suggest it

Primary Sclerosing Cholangitis

Medical Condition

Hardening and narrowing of bile ducts



Chronic liver disease involving inflammation, scarring and narrowing of bile ducts.

📅 Very rare (Fewer than 20,000 cases per year in US)

🧪 Requires lab test or imaging

👨‍⚕️ Treatment from medical professional advised

🕒 Can last several months or years

The cause is not clearly understood. These can include: fatigue, pain in the upper right abdomen, night sweats, enlarged spleen, enlarged liver, chills, weight loss, jaundice and itching. The known cure is liver transplant. It can be treated by managing the symptoms using drugs such as antibiotics, opioid antagonists, bile acid sequestrants and ursodeoxycholic acid (UDCA).

Symptoms



IgG4-Related Sclerosing Cholangitis and Primary Sclerosing ...

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Primary sclerosing cholangitis with increased immunoglobulin G4 levels: A case report. **Chen QL**(1), **Zhong R**(2), **Zhang XX**(1), **Feng LN**(1), **Wen XY**(1), Jin QL(1). Author information: (1)Department of Hepatology. (2)Department of Neurology, The First Hospital of ...

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Cited by: 81

Author: Takahiro Nakazawa, Itaru Naitoh, Kazuki ...

Publish Year: 2013

PEOPLE ALSO ASK

What is abstract IgG4 related sclerosing cholangitis? ▾

What is the IgG4 level in the pancreas? ▾

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