

Name of Journal: *World Journal of Gastroenterology*

Manuscript NO: 48060

Manuscript Type: MINIREVIEWS

**Reduced lysosomal acid lipase activity: A new marker of liver disease severity across the clinical continuum of non-alcoholic fatty liver disease?**

Baratta F *et al.* Reduced lysosomal acid lipase activity

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# Abstract

Lysosomal acid lipase (LAL) plays a key role in intracellular lipid metabolism. <sup>2</sup>Reduced LAL activity promotes an increased multi-organ lysosomal cholesterol esters storage, as observed in two recessive autosomal genetic diseases, Wolman disease and Cholesterol ester storage disease. Severe liver steatosis and accelerated liver fibrosis are common features in patients with genetic LAL deficiency. By contrast, few reliable data are available on the modulation of LAL activity *in vivo* and on the epigenetic and metabolic factors capable of regulating its activity in subjects without homozygous mutations of the Lipase A gene. In the last years, a less severe and non-genetic reduction of <sup>8</sup>LAL activity was reported in children and adults with non-alcoholic fatty liver disease (NAFLD) suggesting a possible role of LAL reduction in the pathogenesis and progression of the

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**Reduced Lysosomal Acid Lipase Activity in Adult Patients With Non ...**<https://www.ncbi.nlm.nih.gov/pubmed/26288848> ▾ [翻译此页](#)

作者: F Baratta - 2015 - 被引用次数: 35 - 相关文章

Non-alcoholic fatty **liver disease** (NAFLD) is characterized by intra-hepatic fat accumulation and mechanisms involved in its pathogenesis are not fully explained. **Lysosomal Acid Lipase** (LAL) is a key enzyme in lipid metabolism. ... Our findings suggest a strong association between impaired LAL **activity** and **NAFLD**.

缺少字词: Marker Severity Across Continuum

**Low Serum Lysosomal Acid Lipase Activity Correlates with Advanced ...**<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4813175/> ▾ [翻译此页](#)

作者: E Shteyer - 2016 - 被引用次数: 13 - 相关文章

2016年2月27日 - **Medical** records of patients with microvesicular steatosis, cryptogenic cirrhosis ...

Serum LAL **activity** <0.5 indicates **severe** liver injury in patients with fatty liver and cirrhosis. ...

Keywords: **lysosomal acid lipase**, cholesteryl ester storage disease, ... The earliest stage of nonalcoholic fatty **liver disease** (NAFLD) ...

缺少字词: Continuum

**[PDF] abstract book - EASL**[www.easl.eu/naflid2018/wp-content/uploads/09/NAFLDSummit2018-Abstract-book.pdf](http://www.easl.eu/naflid2018/wp-content/uploads/09/NAFLDSummit2018-Abstract-book.pdf) ▾ [翻译此页](#)

2018年9月20日 - OP-01Effect of sebelipase alfa on liver parameters **over** 96 weeks in a .... P01-10

**Lysosomal acid lipase** deficit alters the response to therapy of .... **clinical** models of NASH and liver fibrosis .... with non-alcoholic fatty **liver disease** (NAFLD) Development in a ..... that might impact on fatty **liver disease severity**.

**Lysosomal Acid Lipase Deficiency Unmasked in Two Children With ...**[https://www.researchgate.net/publication/308084285\\_Lysosomal\\_Acid\\_Lipase\\_Deficiency\\_in\\_Two\\_Children\\_With\\_Non-Alcoholic\\_Fatty\\_Liver\\_Disease](https://www.researchgate.net/publication/308084285_Lysosomal_Acid_Lipase_Deficiency_in_Two_Children_With_Non-Alcoholic_Fatty_Liver_Disease) ▾ [翻译此页](#)

Therefore LAL **activity reduction** could be a contributing factor in progression of fatty liver ... Therefore we are far enough to speculate about potential **new** targets for the ... **Lysosomal Acid Lipase Activity** Deficiency in Children with **Liver Disease**:a ... also mimic the **clinical** picture of non alcoholic fat- ty **liver disease** ( NAFLD).

**Lysosomal acid lipase deficiency – An under-recognized cause of ...**<https://www.sciencedirect.com/science/article/pii/S0021915014002020> - [翻译此页](#)

作者: Ž Reiner - 2014 - 被引用次数: 167 - 相关文章

We review current understanding of **lysosomal acid lipase** deficiency (LAL-D). ... non-alcoholic fatty **liver disease** (NAFLD) or cryptogenic cirrhosis [4], [5], [6]. ... LAL-D is a heterogeneous disease that presents **along** a **clinical continuum**, with signs .... When LAL **activity** is absent or **reduced**, cholesteryl esters and triglycerides ...

**Lysosomal Acid Lipase Deficiency - ommbid - McGraw-Hill Medical**<https://ommbid.mhmedical.com/content.aspx?sectionid=62643187...> - [翻译此页](#)

Deficient **activity** of **lysosomal acid lipase** (LAL) leads to the accumulation of ... Deficiency of LAL (LALD) leads to a **continuum** of **diseases** historically divided ... Childhood/adult-onset LALD has a less **severe clinical** course that is highly variable. .... **over** time, accompanied by other indicators of progressive **liver** injury and ...

**Long term substrate reduction therapy with ezetimibe alone or**



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## Reduced lysosomal acid lipase activity - A potential role in the ... - NCBI

<https://www.ncbi.nlm.nih.gov/pubmed/27198736> ▼ 이 페이지 번역하기

PK Selvakumar 저술 - 2016 - 11회 인용 - 관련 학술자료

2016. 4. 29. - **Reduced lysosomal acid lipase activity** - A potential role in the pathogenesis of **non alcoholic fatty liver disease** in pediatric patients. ... (1)Department of Pediatric Gastroenterology and Hepatology, Cleveland Clinic, Cleveland, OH, USA. ... with NAFLD and assess its correlation with histological **severity**.

## Reduced Lysosomal Acid Lipase Activity in Adult Patients With Non ...

<https://www.ncbi.nlm.nih.gov/pubmed/26288848> ▼ 이 페이지 번역하기

F Baratta 저술 - 2015 - 37회 인용 - 관련 학술자료

**Reduced Lysosomal Acid Lipase Activity** in Adult Patients With Non-alcoholic ... ( 1)Department of Internal Medicine and **Medical Specialties**, Sapienza ... **Non- alcoholic fatty liver disease** (NAFLD) is characterized by intra-hepatic fat ... A better knowledge of the role of **LAL** may provide **new** insights in NAFLD pathogenesis.

## Lysosomal acid lipase deficiency - An under-recognized cause of ...

<https://www.sciencedirect.com/science/.../S0021915014002020> - 이 페이지 번역하기

Ž Reiner 저술 - 2014 - 176회 인용 - 관련 학술자료

Progressive **liver disease** is another characteristic feature of **LAL-D**, and patients ... non-alcoholic steatohepatitis (NASH), **non-alcoholic fatty liver disease** ... **LAL-D** is a heterogeneous disease that presents **along a clinical continuum**, with .... When **LAL activity** is absent or **reduced**, cholesteryl esters and triglycerides are not ...

## [PDF] abstract book - EASL

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2018. 9. 20. - P01-10 **Lysosomal acid lipase** deficit alters the response to therapy of ... **clinical** models of



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## Lysosomal Acid Lipase Deficiency - Endotext - NCBI Bookshelf

<https://www.ncbi.nlm.nih.gov/books/NBK395569>

**Lysosomal acid lipase** deficiency (LAL-D) is a rare, heterogeneous, autosomal recessive **disease** that presents along a **clinical continuum**. LAL-D is characterized by accumulation of cholesteryl esters and triglycerides primarily in the **liver** and spleen, but with other organ involvement as well.

Cited by: 3

Author: Nirav K Desai, Don P Wilson

Publish Year: 2016

Author: Nirav K Desai, Don P Wilson

## Reduced Lysosomal Acid Lipase Activity—A Potential role in ...

[https://www.researchgate.net/publication/301762284\\_Reduced\\_Lysosomal\\_Acid\\_Lipase...](https://www.researchgate.net/publication/301762284_Reduced_Lysosomal_Acid_Lipase...)

Request PDF on ResearchGate | **Reduced Lysosomal Acid Lipase** Activity—A Potential role in the Pathogenesis of **Non Alcoholic Fatty Liver Disease** in Pediatric patients | Background: Within the ...

## Lysosomal Acid Lipase Deficiency: Presentation, Diagnosis ...

[https://www.medscape.org/viewarticle/855755\\_transcript](https://www.medscape.org/viewarticle/855755_transcript) ▾

Bowden KL, Bilbey NJ, Bilawchuk LM, et al. **Lysosomal acid lipase** deficiency impairs regulation of ABCA1 gene and formation of high density lipoproteins in cholesteryl ester storage **disease**. ... American College of Gastroenterology The diagnosis and management of **non-alcoholic fatty liver disease**: practice guideline by the American ...

## Nonalcoholic fatty liver disease (NAFLD): Diagnosis ...

<https://www.sciencedirect.com/science/article/pii/S109291341830306X>

**Nonalcoholic fatty liver disease** (NAFLD) is increasingly prevalent and strongly associated with obesity, diabetes and the metabolic syndrome, not only in the Western societies, but also in most regions of the world in the 21st century.

Author: Lisa K. Koch, Matthew M. Yeh

Publish Year: 2018

## Lysosomal Acid Lipase: From Cellular Lipid Handler to ...

<https://www.sciencedirect.com/science/article/pii/S0165614718302311>

Moreover, hematopoietic stem cell transplantation, although limited by graft failure and **severity** of pretransplant **liver disease**, has been successfully used in patients with Wolman **disease** (WD) 32, 33 to restore LAL enzymatic **activity** in circulating cells and in resident macrophages, thus improving growth and survival rate.

Cited by: 1

Author: M. Gomaraschi, F. Bonacina, G.D. Norata

Publish Year: 2019