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ESPS PEER-REVIEW REPORT

Name of journal: World Journal of Hepatology

ESPS manuscript NO: 20636

Title: Wilson’s disease: A review of what we have learned

Reviewer’s code: 00051398

Reviewer’s country: Lebanon

Science editor: Fang-Fang Ji

Date sent for review: 2015-06-15 08:41

Date reviewed: 2015-06-26 21:45

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input type="checkbox"/> Grade A: Priority publishing	Google Search:	<input checked="" type="checkbox"/> Accept
<input checked="" type="checkbox"/> Grade B: Very good	<input checked="" type="checkbox"/> Grade B: Minor language polishing	<input type="checkbox"/> The same title	<input type="checkbox"/> High priority for publication
<input type="checkbox"/> Grade C: Good		<input type="checkbox"/> Duplicate publication	
<input type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade C: A great deal of language polishing	<input type="checkbox"/> Plagiarism	<input type="checkbox"/> Rejection
<input type="checkbox"/> Grade E: Poor	<input type="checkbox"/> Grade D: Rejected	<input checked="" type="checkbox"/> No	<input type="checkbox"/> Minor revision
		BPG Search:	<input type="checkbox"/> Major revision
		<input type="checkbox"/> The same title	
		<input type="checkbox"/> Duplicate publication	
		<input type="checkbox"/> Plagiarism	
		<input checked="" type="checkbox"/> No	

COMMENTS TO AUTHORS

this is well written paper reviewing WD patients diagnosis, manifestations, treatment including alternative treatment. there are minor editorial and typos. the stated sentence on page 2 regarding the Asn1270Ser being described only in scilian and Turkish is missing the fact it has been also reported in the homozygous state in lebanese patients. The authors elaborated on the different modalities in treating WD. it would be interesting if there are some data on the consequences of the long term treatment with copper chelators. copper is required in many enzymes and proteins, and in iron homeostasis.



ESPS PEER-REVIEW REPORT

Name of journal: World Journal of Hepatology

ESPS manuscript NO: 20636

Title: Wilson’s disease: A review of what we have learned

Reviewer’s code: 02444981

Reviewer’s country: Netherlands

Science editor: Fang-Fang Ji

Date sent for review: 2015-06-15 08:41

Date reviewed: 2015-06-27 19:15

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input checked="" type="checkbox"/> Grade A: Priority publishing	Google Search:	<input type="checkbox"/> Accept
<input type="checkbox"/> Grade B: Very good	<input type="checkbox"/> Grade B: Minor language polishing	<input type="checkbox"/> The same title	<input type="checkbox"/> High priority for publication
<input type="checkbox"/> Grade C: Good	<input type="checkbox"/> Grade C: A great deal of language polishing	<input type="checkbox"/> Duplicate publication	<input type="checkbox"/> Rejection
<input checked="" type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade D: Rejected	<input checked="" type="checkbox"/> Plagiarism	<input type="checkbox"/> Minor revision
<input type="checkbox"/> Grade E: Poor		[Y] No	<input type="checkbox"/> Major revision
		BPG Search:	
		<input type="checkbox"/> The same title	
		<input type="checkbox"/> Duplicate publication	
		<input type="checkbox"/> Plagiarism	
		[Y] No	

COMMENTS TO AUTHORS

To the authors: The manuscript entitled “Wilson's disease: What have we learned?” discusses different treatments in Wilson’s disease and their consequences. The interest of the authors in the subject is encouraging, and hopefully it will be continued to help doctors make better clinical decisions as well as patients with better treatments. However, none of those suggestions are based on robust evidence. So it should be explained in the introduction that the treatment is generally based on experts’ opinions and anecdotal experiences, and not on well-designed randomized comparative studies. Moreover, the title does not convey the main message and purpose of the article. Even more so, the article looks like a review article rather than a focused editorial. The keywords indicate that the main goal of the article would be about liver transplantation in Wilson’s disease. Finally, the introduction lacks the discussion over the paradigm shift that has taken place in the management of Wilson’s disease (Hoogenraad. Brain & Development 2006; Hoogenraad Encyclopedia of Movement Disorders 2010). Copper accumulation is not the cause of Wilson’s disease, but the raised free copper is the damaging element, which harms different organs. Thus, the treatment should aim at normalizing this alteration, which is only achieved by zinc therapy. Accordingly, the title would

better change to something that explains the main message of the article; and the same for the keywords. Moreover, the paper in the Encyclopedia of Movement Disorders can be used as a guidance to the revision. Minor suggestions: - Introduction - line 6- what do you mean by “abnormal copper accumulation”? It is better change to impaired copper metabolism and consequent copper intoxication - Page 5-6; the penicillamine paragraph would better divide into 2-3 paragraphs. - Page 6 - lines 2-3 - What is the evidence indicating that penicillamine remains the treatment of choice? No comparative data exist. - Page 6 - Trientine section - line 3-4 - What is the evidence in support of “a good safety profile” for trientine? There is no controlled study. - Page 7 - line 2 - What is the reference for the less neurotoxicity of trientine compared to penicillamine? - Page 7 - Zinc section - first sentence: The authors would better add “; however, it was kept unrecognized until 1978. Moreover, reference 60 would better be deleted. - Page 7 - Zinc section - second sentence would better change to: “Zinc acetate is regarded to have a better gastric tolerance. However, in terms of efficacy there is no difference between zinc salts.” Moreover, in the cited reference (Hoogenraad et al. 1978) the authors used zinc sulphate, and not zinc acetate. - Page 9 - line 3-4 - Correcting the underlying hepatic metabolic defect in WD is not at all an indication for doing such a dangerous operation, which needs a lifelong immunosuppressive treatment, when there is a safe oral treatment. - Page 9 - the sentence after reference 99 (Improvement in neurological symptoms, without a negative effect on survival, has been reported however, in smaller series.) needs a reference. - Page 10 - The sentence “Nevertheless, no randomized controlled studies offer solid evidence for its use” actually applies to all the treatments and suggestion. - Page 10 - Fulminant hepatic failure- second paragraph: What is the reference for the sentence “Treatment in this setting is life-saving, and the best option is offered by liver transplantation?” - Page 10 - Fulminant hepatic failure- second paragraph; last sentence: The authors should propose at least 2 references to suggest the validity of the RWPI in predicting mortality without liver transplantation. - Page 10 - Pregnancy - lines 6, 7: With regard to the sentence “Although there have been reports of birth defects during treatment for WD, the rarity of this disease makes it difficult to establish a true increased risk in this population,” even few cases are enough to conc



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ESPS PEER-REVIEW REPORT

Name of journal: World Journal of Hepatology

ESPS manuscript NO: 20636

Title: Wilson's disease: A review of what we have learned

Reviewer's code: 00502871

Reviewer's country: United States

Science editor: Fang-Fang Ji

Date sent for review: 2015-06-15 08:41

Date reviewed: 2015-06-18 01:08

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input checked="" type="checkbox"/> Grade A: Excellent	<input checked="" type="checkbox"/> Grade A: Priority publishing	Google Search:	<input checked="" type="checkbox"/> Accept
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COMMENTS TO AUTHORS

This is a well written overview of Wilson's disease. The section on pharmacologic therapy is useful.



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ESPS PEER-REVIEW REPORT

Name of journal: World Journal of Hepatology

ESPS manuscript NO: 20636

Title: Wilson’s disease: A review of what we have learned

Reviewer’s code: 00053746

Reviewer’s country: Czech Repoublic

Science editor: Fang-Fang Ji

Date sent for review: 2015-06-15 08:41

Date reviewed: 2015-07-03 23:53

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
<input checked="" type="checkbox"/> Grade A: Excellent	<input checked="" type="checkbox"/> Grade A: Priority publishing	Google Search:	<input type="checkbox"/> Accept
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COMMENTS TO AUTHORS

The authors present a review on Wilson disease. The review is well written and gives good overview on Wilson disease. I have these remarks: In diagnosis: 6-hour challenge test with penicillamine is not a standard method and should not be recommended as a routine technique. Genetic testing is a very useful tool in diagnosis today. With the use of routine molecular techniques, the diagnosis could be done in short time in substantial part of patients with prevalent mutations. In family screening is the genetic testing irreplaceable. In treatment: The combination of zinc and penicillamine is not a standard method; it is used in specific situations



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Title: Wilson’s disease: A review of what we have learned

Reviewer’s code: 00053746

Reviewer’s country: Czech Republic

Science editor: Fang-Fang Ji

Date sent for review: 2015-06-15 08:41

Date reviewed: 2015-07-03 23:56

CLASSIFICATION	LANGUAGE EVALUATION	SCIENTIFIC MISCONDUCT	CONCLUSION
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