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## ESPS PEER REVIEW REPORT

**Name of journal:** World Journal of Nephrology

**ESPS manuscript NO:** 11989

**Title:** Searching for a treatment for Alport syndrome using mouse models

**Reviewer code:** 00503255

**Science editor:** Ling-Ling Wen

**Date sent for review:** 2014-06-16 22:13

**Date reviewed:** 2014-06-18 06:52

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input 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"/> Existing	<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"/> No records	<input type="checkbox"/> Rejection
<input type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade D: Rejected	BPG Search:	<input type="checkbox"/> Minor revision
<input type="checkbox"/> Grade E: Poor		<input type="checkbox"/> Existing	<input type="checkbox"/> Major revision
		<input type="checkbox"/> No records	

### COMMENTS TO AUTHORS

Dr. Katayama reviewed studies about treatments for Alport syndrome using mouse model. It is well-written and has the latest and comprehensive information on this area. I have 3 comments.

- In the section of pharmacological intervention, the authors described MMP inhibitor cocktail showed dual effect on Alport mice from the paper of Zeiberg et al. It should be better to explain why MMP inhibitor cocktail induce these dual reactions in Alport mice.
- In the section of stem cell therapies, the authors showed that a sublethal dose of irradiation without subsequent BMT improved the survival of Alport mice from the authors' own study. How does irradiation improve the survival of Alport mice? Brief explanation should be added.
- The reference style the authors used is not appropriate for this journal. Please read the instructions to authors of this journal and correct references. For example, reference No 2, "Am J Hum Genet. 1983 Nov;35 (6):1241-51." should be changed to "Am J Hum Genet 1983;35:1241-1251"



**ESPS PEER REVIEW REPORT**

**Name of journal:** World Journal of Nephrology

**ESPS manuscript NO:** 11989

**Title:** Searching for a treatment for Alport syndrome using mouse models

**Reviewer code:** 00503179

**Science editor:** Ling-Ling Wen

**Date sent for review:** 2014-06-16 22:13

**Date reviewed:** 2014-06-25 23:43

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	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"/> Existing	<input type="checkbox"/> High priority for publication
<input checked="" type="checkbox"/> Grade C: Good	<input type="checkbox"/> Grade C: A great deal of language polishing	<input type="checkbox"/> No records	<input type="checkbox"/> Rejection
<input type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade D: Rejected	BPG Search:	<input type="checkbox"/> Minor revision
<input type="checkbox"/> Grade E: Poor		<input type="checkbox"/> Existing	<input type="checkbox"/> Major revision
		<input type="checkbox"/> No records	

**COMMENTS TO AUTHORS**

General comments: The paper is a review regarding Alport’s syndrome using a Mouse model. Genetic-, pathogenetic- and therapeutic aspects are reviewed. Results from animal studies are the central information. Little attention has been paid to Alport’s syndrome in humans. The paper is well written and contains a large list of relevant references. The review does not contain new information, but is useful since the present knowledge is updated regarding a mouse model. Specific comments:

Abstract: The abstract must reflect the content of the paper. It is the mouse model and the effect of intervention with therapies and genetics in mice, which is the purpose, not man. It must be rewritten.

Introduction: A paragraph should be included given information about the purpose of the review.

The five main sections in the paper: Pathogenesis, Mouse Model, Pharmacological Interventions, Genetic approaches and Stam Cell Therapies are short sections comprising the present knowledge with relevant references.

Conclusion: It is out of focus. The topic of the review is a mouse model. The conclusion is about treatment of patients. The content of the conclusion is not wrong. It is well-known facts regarding Alport’s syndrome. However, the conclusion must, of course, refer to the mouse model and the knowledge from animal experiments as dealt with in the paper.



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## ESPS PEER REVIEW REPORT

**Name of journal:** World Journal of Nephrology

**ESPS manuscript NO:** 11989

**Title:** Searching for a treatment for Alport syndrome using mouse models

**Reviewer code:** 00503252

**Science editor:** Ling-Ling Wen

**Date sent for review:** 2014-06-16 22:13

**Date reviewed:** 2014-06-27 14:27

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
<input type="checkbox"/> Grade A: Excellent	<input 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"/> Existing	<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"/> No records	<input type="checkbox"/> Rejection
<input type="checkbox"/> Grade D: Fair	<input type="checkbox"/> Grade D: Rejected	BPG Search:	<input type="checkbox"/> Minor revision
<input type="checkbox"/> Grade E: Poor		<input type="checkbox"/> Existing	<input type="checkbox"/> Major revision
		<input type="checkbox"/> No records	

### COMMENTS TO AUTHORS

Katayama et al. reviewed well the pathogenesis of Alport syndrome and its possible treatments in animals and men at present. Please explain the sentence, “the ACE inhibitor was much more effective than the ARB” in Col/4a3-/- mice more in detail. Are the effect of ACE inhibitors on Alport syndrome really class effect? Is there any difference of effects among ACE inhibitors? “the distal tubular membranes (TBMs)” should be “the distal tubular basement membrane (TBMs)”.