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ESPS Peer-review Report
Name of Journal: World Journal of Gastroenterology
ESPS Manuscript NO: 7047
Title: A variant of ARC syndrome in a Chinese Han patient with high GGT cholestasis caused by VPS33B mutations
Reviewer code: 00006675
Science editor: Qi, Yuan
Date sent for review: 2013-11-02 19:57
Date reviewed: 2013-12-15 16:23

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
[ ] Grade A (Excellent)	[ ] Grade A: Priority Publishing	Google Search:	[ ] Accept
[ ] Grade B (Very good)	[Y] Grade B: minor language polishing	[ ] Existed	[ ] High priority for publication
[Y] Grade C (Good)	[ ] Grade C: a great deal of language polishing	[ ] No records	[ ]Rejection
[ ] Grade D (Fair)	[ ] Grade D: rejected	BPG Search:	[Y] Minor revision
[ ] Grade E (Poor)		[ ] Existed	[ ] Major revision
		[ ] No records	

COMMENTS TO AUTHORS
This is an interesting report by Wang et al., indicating that ARC syndrome cannot be excluded from the differential diagnosis of neonatal cholestasis based on data regarding serum levels of GGT activity. 1. An interesting data is the parallel increase in serum levels of GGT and total bile acids. This should be described in Results section and commented in the Discussion. 2. Can the authors provide any possible speculation regarding the mechanistic link among the mutation, the dysfunction in bile formation and the elevation of serum GGT? 3. Improve consistence regarding the use of GGT or gGt

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Name of Journal: World Journal of Gastroenterology
ESPS Manuscript NO: 7047
Title: A variant of ARC syndrome in a Chinese Han patient with high GGT cholestasis caused by VPS33B mutations
Reviewer code: 02860826
Science editor: Qi, Yuan
Date sent for review: 2013-11-02 19:57
Date reviewed: 2013-12-20 22:35

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
[ ] Grade A (Excellent)	[ ] Grade A: Priority Publishing	Google Search:	[ ] Accept
[ ] Grade B (Very good)	[Y] Grade B: minor language polishing	[ ] Existed	[ ] High priority for publication
[Y] Grade C (Good)	[ ] Grade C: a great deal of language polishing	[ ] No records	[ ]Rejection
[ ] Grade D (Fair)	[ ] Grade D: rejected	BPG Search:	[Y] Minor revision
[ ] Grade E (Poor)		[ ] Existed	[ ] Major revision
		[ ] No records	

COMMENTS TO AUTHORS
This case report concerns a second child of a non-consaguineous han couple with ARC syndrome. This is a severe autosomal recessive multisystem disorder. Germline mutations were found in VPS33B and in VIPAS39. A generally well-written case report with a few points that should modified: 1) In the abstract the authors illustrate only one case, but in the Cases reports they describe two cases. Please modified the "cases report" combining the two cases. I suppose that is more understandable if the second case is inserted into the medical history of the proband. 2) The authors use sometimes the term "patient" and sometimes "proband". Please use only one of the two terms. 3) In the manuscript I find GGT, gGT and γGT. Please use the first one (GGT), cited in the abstract. 4) Page 4 paragraph 2: "Epstein-Barr virus; blood cytomegalovirus DNA, and chromosome G bands." Please correct in : Epstein-Barr virus, blood cytomegalovirus DNA and chromosome G bands. 5) Page 3, paragraph 1: detailed premiers. I suppose that was detailed primers. 6) The authors use ", and" : please remove "," before "and".

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ESPS Peer-review Report
Name of Journal: World Journal of Gastroenterology
ESPS Manuscript NO: 7047
Title: A variant of ARC syndrome in a Chinese Han patient with high GGT cholestasis caused by VPS33B mutations
Reviewer code: 02860576
Science editor: Qi, Yuan
Date sent for review: 2013-11-02 19:57
Date reviewed: 2013-12-31 14:21

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
[ ] Grade A (Excellent)	[ ] Grade A: Priority Publishing	Google Search:	[Y] Accept
[Y] Grade B (Very good)	[Y] Grade B: minor language polishing	[ ] Existed	[ ] High priority for publication
[ ] Grade C (Good)	[ ] Grade C: a great deal of language polishing	[ ] No records	
[ ] Grade D (Fair)		BPG Search:	[ ]Rejection
[ ] Grade E (Poor)	[ ] Grade D: rejected	[ ] Existed	[ ] Minor revision
		[ ] No records	[ ] Major revision

COMMENTS TO AUTHORS
this is an intersting case report .please consider the followings: 1.blood ceruloplasmin is not related to this age ,should be deleted. 2.Ultrasound abdomen should be re written without repititions. 3.family history should follow history not physical examination and onvestigations

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ESPS Peer-review Report
Name of Journal: World Journal of Gastroenterology
ESPS Manuscript NO: 7047
Title: A variant of ARC syndrome in a Chinese Han patient with high GGT cholestasis caused by VPS33B mutations
Reviewer code: 02861186
Science editor: Qi, Yuan
Date sent for review: 2013-11-02 19:57
Date reviewed: 2014-01-08 01:18

CLASSIFICATION	LANGUAGE EVALUATION	RECOMMENDATION	CONCLUSION
[ ] Grade A (Excellent)	[ ] Grade A: Priority Publishing	Google Search:	[Y] Accept
[ ] Grade B (Very good)	[Y] Grade B: minor language polishing	[ ] Existed	[ ] High priority for publication
[Y] Grade C (Good)	[ ] Grade C: a great deal of language polishing	[ ] No records	
[ ] Grade D (Fair)		BPG Search:	[ ]Rejection
[ ] Grade E (Poor)	[ ] Grade D: rejected	[ ] Existed	[ ] Minor revision
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COMMENTS TO AUTHORS
Dear Editor,
this manuscript describes the problematic diagnosis of an rare syndrome in newborn, the ARC syndrome.
The proband (female child, age of >30 days) showed an increased gGT-activity cholestasis, which is rather a non-typical feature of the ARC syndrome. The residual clinical findings as well as the molecular results confirmed the diagnosis of the ARC syndrome. The authors mentioned that the sister of the patient already died of the severe complications of the ARC syndrome.

In the manuscript the authors gave a comprehensive state of the clinical, paraclinical facts and molecular results concerning the differential diagnosis for a high gGT cholestasis. In addition, the authors stated that the study protocol complied with the ethical guidelines (Helsinki 1975) and the local Ethics Committee on human research has approved the study protocol including the molecular and genetic analyses of the parents` DNA.

In review of the literature, no one has ever described the ARC syndrome with an increased gGT-activity. To my knowledge, it is highly questionable that the described increased gGT-activity reflects a "subtype" of the ARC syndrome. Moreover, in my eyes, it seems to be reasonable that the high gGT-activity reflects rather a more pronounced damage or reaction of the biliary endothel cells compared to the sister`s as well as to the other newborns` with the ARC syndrome.

In summary, I would recommend to publish this case report of an ARC syndrome with an increased gGT-activity, although it describes a very seldom feature of a rare disease.
But I do think, that this case report could help to keep the eyes open for and the focus on rare diseases.