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PEER-REVIEW REPORT

Name of journal: World Journal of Medical Genetics

Manuscript NO: 55385

Title: A look at the clinical and molecular spectrum of Wiedemann-Steiner syndrome, an emerging member of the chromatinopathies family

Reviewer's code: 04022629
Position: Peer Reviewer
Academic degree: FRCS (Gen Surg), PhD
Professional title: Academic Fellow, Academic Research
Reviewer's Country/Territory: China
Author's Country/Territory: Italy

Manuscript submission date: 2020-03-14
Reviewer chosen by: AI Technique
Reviewer accepted review: 2020-03-15 03:38
Reviewer performed review: 2020-03-16 08:58
Review time: 1 Day and 5 Hours

Scientific quality	[ ] Grade A: Excellent [Y] Grade B: Very good [ ] Grade C: Good [ ] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[ ] Grade A: Priority publishing [Y] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[ ] Accept (High priority) [Y] Accept (General priority) [ ] Minor revision [ ] Major revision [ ] Rejection
Re-review	[ ] Yes [ ] No
Peer-reviewer statements	Peer-Review: [Y] Anonymous [ ] Onymous Conflicts-of-Interest: [ ] Yes [Y] No

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SPECIFIC COMMENTS TO AUTHORS

Fontana P summarized the clinical and molecular spectrum of Wiedemann-Steiner syndrome. The manuscript is generally well-writtern, however, there are one concern that should be addressed. 1. In Figure 1, the pictures should be manipulated to hide the information that can identify the patient.

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PEER-REVIEW REPORT

Name of journal: World Journal of Medical Genetics

Manuscript NO: 55385

Title: A look at the clinical and molecular spectrum of Wiedemann-Steiner syndrome, an emerging member of the chromatinopathies family

Reviewer's code: 00375480
Position: Peer Reviewer
Academic degree: MD, PhD
Professional title: Senior Scientist
Reviewer's Country/Territory: United States
Author's Country/Territory: Italy

Manuscript submission date: 2020-03-14
Reviewer chosen by: Jie Wang
Reviewer accepted review: 2020-03-25 19:05
Reviewer performed review: 2020-03-25 20:41
Review time: 1 Hour

Scientific quality	[ ] Grade A: Excellent [ ] Grade B: Very good [Y] Grade C: Good [ ] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[Y] Grade A: Priority publishing [ ] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[ ] Accept (High priority) [ ] Accept (General priority) [Y] Minor revision [ ] Major revision [ ] Rejection
Re-review	[ ] Yes [ ] No
Peer-reviewer statements	Peer-Review: [Y] Anonymous [ ] Onymous Conflicts-of-Interest: [ ] Yes [Y] No

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SPECIFIC COMMENTS TO AUTHORS

WJMG #55385 comments Wiedemann-Steiner Syndrome (WDSTS) is a rare autosomaldominant disorder caused by haploinsufficiency of the MLL gene (now known as KMT2A). KMT2A encodes a histone methyltransferase, and as a chromatin epigenetic "writer", it adds a lysine residue on histone H3 lysine 4 (H3K4). Mutations in this writer gene leads to a dysfunction of the epigenetic machinery. In this review, the authors summarized the clinical features of Wiedemann-Steiner syndrome and the mutation spectrum of KMT2A. The manuscript is well written. I have two suggestions for the authors. 1. Table 1 summarizes the location and clinical interpretation of KMT2A mutation. Figure 2 shows the enrichment of mutation in each exon of KMT2A. It would be more information to add a diagram that shows the protein structure of KMT2A overlaid with the reported mutations. 2. A discussion of the mutation hotspot in KMT2A function domains in relation to the clinical characteristics of WDSTS.

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PEER-REVIEW REPORT

Name of journal: World Journal of Medical Genetics

Manuscript NO: 55385

Title: A look at the clinical and molecular spectrum of Wiedemann-Steiner syndrome, an emerging member of the chromatinopathies family

Reviewer's code: 00699199
Position: Peer Reviewer
Academic degree: MD, PhD
Professional title: Assistant Professor
Reviewer's Country/Territory: United States
Author's Country/Territory: Italy

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Scientific quality	[ ] Grade A: Excellent [Y] Grade B: Very good [ ] Grade C: Good [ ] Grade D: Fair [ ] Grade E: Do not publish
Language quality	[ ] Grade A: Priority publishing [Y] Grade B: Minor language polishing [ ] Grade C: A great deal of language polishing [ ] Grade D: Rejection
Conclusion	[ ] Accept (High priority) [ ] Accept (General priority) [Y] Minor revision [ ] Major revision [ ] Rejection
Re-review	[ ] Yes [ ] No
Peer-reviewer statements	Peer-Review: [Y] Anonymous [ ] Onymous Conflicts-of-Interest: [ ] Yes [Y] No

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SPECIFIC COMMENTS TO AUTHORS

This is an informative and interesting review on chromatinopathies. It is generally well written. However, some English usage issues were noted. I have attached an edited document with corrections and comments. There were also some inaccuracies in the introduction regarding histone methyltransferases which I have fixed in the edited document. In general, the introduction could benefit from a little more background on histone methylation and KDMT2A. Here are some suggested papers to reference: Histone Lysine Methylation and Neurodevelopmental Disorders. Kim JH, Lee JH, Lee IS, Lee SB, Cho KS. Int J Mol Sci. 2017 Jun 30;18(7). pii: E1404. doi: 10.3390/ijms18071404. 1. Strom, S.P.; Lozano, R.; Lee, H.; Dorrani, N.; Mann, J.; O'Lague, P.F.; Mans, N.; Deignan, J.L.; Vilain, E.; Nelson, S.F. De Novo variants in the KMT2A (MLL) gene causing atypical Wiedemann-Steiner syndrome in two unrelated individuals identified by clinical exome sequencing. BMC Med. Genet. 2014, 15, 49. (already cited by the authors) 2. Gupta, S.; Kim, S.Y.; Artis, S.; Molfese, D.L.; Schumacher, A.; Sweatt, J.D.; Paylor, R.E.; Lubin, F.D. Histone methylation regulates memory formation. J. Neurosci. 2010, 30, 3589–3599. 3. Kim, S.Y.; Levenson, J.M.; Korsmeyer, S.; Sweatt, J.D.; Schumacher, A. Developmental regulation of Eed complex composition governs a switch in global histone modification in brain. J. Biol. Chem. 2007, 282, 9962–9972. 4. Jakovcevski, M.; Ruan, H.; Shen, E.Y.; Dincer, A.; Javidfar, B.; Ma, Q.; Peter, C.J.; Cheung, I.; Mitchell, A.C.; Jiang, Y. Neuronal Kmt2a/Mll1 histone methyltransferase is essential for prefrontal synaptic plasticity and working memory. J. Neurosci. 2015, 35, 5097–5108.