

## ***Response to the Reviewers***

MS #28343: Hematological Disorders and Pulmonary Hypertension

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The authors would like to thank the Reviewers for their kind and helpful comments and suggestions

### ***Reviewer #1 (Code 02446717)***

Thank you for your kind remarks. We have included a section on “Autoimmunity” and associated hematological diseases and PH. We have corrected typographical errors.

### ***Reviewer #2 (Code 02520738)***

Thank you for your helpful suggestions. We have included the suggested paper (Scicchitano P et al Endocrine March 2016) in the new section entitled “Autoimmunity”. As suggested we have added a figure depicting anemia, hemolysis and coagulopathy leading to EC damage and subsequently to PH. As far as the Table is concerned, we believe it will become complicated, because in some of the diseases such as in chronic myeloproliferative diseases, CTEPH occurs early during the course of the disease whereas during the late stages these patients have PAH. Similarly, some patients who received bone marrow transplantation and immunosuppressive therapy developed PVOD, whereas in a few the presentation was suggestive of PAH. These would require lengthy explanations. We hope the figure will give a simplified view of the relationship between hematological disorders and PH.

### ***Reviewer #3 (Code 002273750)***

Thank you very much for your kind remarks.

### ***Reviewer #4 (Code 00503221)***

Thank you for your comments. A statement about pulmonary artery pressure measurement has been included in the text.

### **Statement of Conflict of Interest**

None of the Authors (Rajamma Mathew, Jing Huang, Joseph M Wu, John T Fallon and Michael H Gewitz) of the manuscript entitled “Hematological disorders and pulmonary hypertension” have any conflict of interest.

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### *List of Corrections*

1. Comment #1: Page 1: In the revised version, the spelling corrections have been made. These appear in red.
2. Comment #2: Page 4: The reference numbers in bracket have been put in superscript.
3. Comment #3: Page 6: The reference # 6 has been correctly placed. The reference # 5 that appeared on page 6 was an error.
4. Comment #4; Page 6: I am enclosing the Fig 1 in a power-point slide. This should enable you to move the figure parts as desired. If there is a problem with the slide, please let me know.
5. Comment #5; Page 9: Reference number 33 has been correctly placed.
6. Comment #6; Page 32: The reference # 56 which appears to be a repeat of 34 has been removed. The references have been correctly numbered in the text and the reference list.
7. Comment #7; Page 36: The reference # 83, a repetition of #35 has been removed. The references have been renumbered in the text and the reference list.

**Other changes made:** 1. Page 10: a line has been added (it appears in red). Reference #44 (the original #41) has been moved to this section for clarity. The sentence is as follows: “**Morbidity and mortality in SCD are high**, and PH is a serious complication in SCD. Sudden death in patients with SCD and PH is not uncommon <sup>[44, 45]</sup>.” The reference numbers have been corrected in the reference list and the numbers appear in red. The references 42, 43 and 44 appear as #41, 42 and 43.

2. Page 11: A line has been added before the reference number 54. It emphasizes their experimental results. The sentence appears in red. “**Recent experimental studies in rodents reveal that it is the Hb-induced inflammation and to a lesser extent the Hb-induced oxidant injury leads to vascular injury** <sup>[54]</sup>.”

3. Page 17: The last sentence of the first paragraph under the heading of “Autoimmunity” has been slightly modified for better language expression. The original sentence “**Another patient with SLE with associated lupus anticoagulant and clotting disorder was described to have PH** <sup>[113]</sup>.” This sentence has been changed to “**Another patient with SLE and associated lupus anticoagulant and clotting disorder was described to have PH** <sup>[113]</sup>.”

4. Page 20: I have added a few articles such as “a” and “the” which were missing. They appear in red.