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Author rebuttal to response regarding "Letter to the Editor regarding 'Could pulmonary arterial hypertension patients be at lower risk from severe COVID-19?'"

Evelyn Horn[1], Murali M. Chakinala[2], Ronald Oudiz[3], Elizabeth Joseloff[4] and Erika B. Rosenzweig[5]

[1]Weill Cornell Medicine, New York Presbyterian Hospital, New York, USA; [2]School of Medicine, Washington University in Saint Louis, St. Louis, MO, USA; [3]David Geffen School of Medicine at UCLA, UCLA Medical Center, Santa Monica, CA, USA; [4]Pulmonary Hypertension Association Silver Spring, MD, USA; [5]Departments of Pediatrics and Medicine, Columbia University Medical Center, New York, NY, USA

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In response to comments published by Timothy Fernandes et al. to our Letter to the Editor titled "Could pulmonary arterial hypertension patients be at a lower risk from severe COVID-19?,"[1] we believe it is critical to provide additional information and perspective. Fernandes et al. expressed concern that our survey data of pulmonary arterial hypertension (PAH) and CTEPH patients represented only a snapshot early in the COVID-19 pandemic when the U.S. was at the beginning stages of disease acceleration, and testing was limited. We agree with this observation and have extended our survey window through 6 May 2020, past the peak of COVID activity in much of the U.S. At the end of the survey period, we received a total of 86 responses by PH health care providers, with 66% estimating that their sites had more than 100 PAH and CTEPH patients strengthening our conclusions.

From our survey responses, 16 PAH/CTEPH patients were reported as testing positive for COVID-19, 11 (58%) were hospitalized, and 6 (55%) of those hospitalized required intubation. Given the timing of survey response, we acknowledge that these reports also reflect a time when PCR testing was limited in most regions to patients sick enough to require hospitalization. At the end of the survey period, 60% of responders who had hospitalized COVID-19-positive PAH/CTEPH patients indicated that they had patients discharged. Of the 16 PAH/CTEPH COVID-19-positive patients, three deaths were reported.

We agree with Fernandes et al.[1] that there are limitations to surveys such as ours and that exact incidence, hospitalization, and mortality numbers cannot be determined. However, with this broader reach of our survey that included centers with larger numbers of PAH patients, the fact that 16 PAH and CTEPH patients were reported to be COVID-19 positive suggests that severe infection requiring hospitalization is reassuringly similar to the general population and presumably lower than what was widely feared. Moreover, the overall mortality was 19% in this high-risk group with severe underlying cardiopulmonary co-morbidities.

Fernandes et al.[1] expressed concern that our Letter to the Editor may be misinterpreted by the PH community and could lead to less stringent adherence to precautions taken by PH patients, such as social distancing and wearing masks, to protect against SARS-CoV-2 infection. We, along with the Pulmonary Hypertension Association, strongly advocate that all patients and their families, caregivers, and friends follow strict social distancing and all other infection protection measures recommended by the Centers for Disease Control. Our patients are more accustomed to social distancing and excruciating attention to avoiding at high-risk situations. We specifically hypothesized in our Letter to the Editor that perhaps lower numbers of COVID-19-positive PAH/CTEPH patients may be a result of early implementation by the PH community of social distancing and other protective measures. Our preliminary observations are hypothesis generating and in no way minimize our recommendations to the PH community.

Corresponding author:
Erika B. Rosenzweig, Columbia University Medical Center, 3959 Broadway, New York 10032, USA.
Email: esb14@cumc.columbia.edu

There is still much unknown about COVID-19 and the impact on PH patients. Additional studies will need to test hypotheses to understand the science and optimize patient care. A recent *NEJM* article adds to this unusual endothelial pathology with further hypothesis generating ideas that could be pertinent to PAH.[2] Our initial letter was never intended to suggest that PAH patients lessen their self-discipline and intense protective measures, as we still consider them to be a high-risk cohort.

ORCID iD


Erika B. Rosenzweig https://orcid.org/0000-0003-4849-214X

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