Response to Letter Regarding Article, “Pulmonary Vascular Disease in the Developing World”

We thank Drs Lambertucci and Antunes for their useful and constructive remarks about our article.1 We also appreciate the work of Dr Lambertucci’s team in this field, which has increased our knowledge over the last 2 decades.

We agree about how the term Katayama syndrome should be used. However, we think that the issue of resolution of chest radiological changes in their article needs some consideration. They said that these patients did not suffer from pulmonary hypertension; however, there was no attempt to investigate that possibility. Furthermore, our current experimental observations suggest that the changes in the pulmonary vasculature after schistosoma infection are far more common but may not always be associated with significant increases in the total vascular resistance. However, this notion needs further investigation.

As for the relationship between perportal fibrosis and Symmers plestem fibrosis, we believe that Drs Lambertucci and Antunes highlighted several important points that need revisiting. First, our knowledge of the incidence and prevalence of pulmonary vascular diseases due to schistosomiasis is far from accurate. Most previously published works (the majority of which came from Brazilian investigations) have been anecdotal observational studies. We have no studies yet from sub-Saharan Africa where 80% of the world’s schistosoma infection is present. Second, our main hypothesis is that pulmonary vascular disease is caused by the inflammatory response, most probably to the highly antigenic eggs. Portal hypertension and the development of the collateral circulation probably facilitate the diversion of the eggs to the pulmonary compartment.2

Drs Lambertucci and Antunes state, “Without Symmers fibrosis, portal hypertension related to schistosomiasis has not yet been reported.” This statement is correct mainly when the infection is due to Schistosoma mansoni, which is the most prevalent species in Brazil. This species apparently has a higher incidence of pulmonary hypertension than other species, mainly Schistosoma japonicum,3,4 that affect the intestinal and hepatic circulatory system, because of the different antigenicity of the eggs. Furthermore, pulmonary hypertension has been reported in other species that affect the urinary system, such as Schistosoma hematobium.5 It seems that the pathological changes are not only related to perportal fibrosis but several other factors, including the egg burden, chronicity, reinfection, and co-infections. Genetic factors also predispose certain individuals to develop other conditions depending on gender, nutrition, immunological status, and environment.6

Once again, we thank Drs Lambertucci and Antunes for their excellent and important comments, giving us the opportunity to revisit some controversial issues that are still not fully understood. It is clear that further studies are necessary to better understand the impact of this complex infection on pulmonary vascular diseases.

Disclosures

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