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 schistosomal 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 periportal fibrosis and Symmers pipestem 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 schistosomal infection 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 portal fibrosis but several other factors, including the egg burden, chronicity, reinfection, and coinfections. 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

Dr Butrous receives an annual salary from Pfizer Global Research and Development. Dr Ghofrani has received public funding by the Deutsche Forschungsgemeinschaft (SFB 547) and European Union Framework 5 and 6. He has received pharmaceutical grant money from Schering AG, Pfizer, and Encysive; received travel accommodations and speakers’ honoraria from Schering, Actelion, Encysive, Pfizer, GSK, and Ergonex; and been a consultant to Bayer, Schering AG, Pfizer, GSK, Actelion, Encysive, Ergonex, and Aires. Professor Grimminger has received public funding by the Deutsche Forschungsgemeinschaft (SFB 547) and European Union Framework 5 and 6. He has received pharmaceutical grant money from Schering AG, Pfizer, and Altana Pharma; received travel accommodations and speakers’ honoraria from Schering, Actelion, Encysive, and Pfizer; and been a consultant to Bayer, Schering AG, and Pfizer.

Ghazwan Butrous, MB, ChB, PhD
Pulmonary Vascular Research Institute
Centre for Work and Learning
Division of Clinical Practice
University of Kent and Pfizer Global Research and Development
Kent, UK
Hossein Ardeshir Ghofrani, MD
Friedrich Grimminger, MD, PhD
Pulmonary Vascular Research Institute
Medical Clinic II
University Hospital Giessen
Giessen, Germany

References


(Circulation. 2009;120:e7.)
© 2009 American Heart Association, Inc.
Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.109.850800
Response to Letter Regarding Article, "Pulmonary Vascular Disease in the Developing World"
Ghazwan Butrous, Hossein Ardeschir Ghofrani and Friedrich Grimminger

Circulation. 2009;120:e7
doi: 10.1161/CIRCULATIONAHA.109.850800

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/120/1/e7

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/