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Reversible pulmonary hypertension associated with multivisceral Whipple's disease

Alice Camboulive^{1,2,3,9}, Etienne-Marie Jutant^{1,2,3,9}, Laurent Savale ^{1,2,3}, Xavier Jaïs^{1,2,3}, Olivier Sitbon ^{1,2,3}, Charlotte Mussini^{4,5}, Jérémie Bénichou^{5,6}, Jean-Christophe Lagier^{7,8}, Marc Humbert ^{1,2,3} and David Montani ^{1,2,3}

Affiliations: ¹Assistance Publique - Hôpitaux de Paris (AP-HP), Dept of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Center, Hôpital Bicêtre, Le Kremlin-Bicêtre, France. ²Université Paris-Saclay, School of Medicine, Le Kremlin-Bicêtre, France. ³INSERM UMR_S 999 "Pulmonary Hypertension: Pathophysiology and Novel Therapies", Hôpital Marie Lannelongue, Le Plessis-Robinson, France. ⁴AP-HP, Service d'Anatomie et de Cytologie Pathologiques, Hôpital de Bicêtre, Le Kremlin-Bicêtre, France. ⁵Hôpitaux Universitaires Paris-Saclay, Le Kremlin-Bicêtre, France. ⁶AP-HP, Service d'ophtalmologie, Hôpital de Bicêtre, Le Kremlin-Bicêtre, France. ⁷Aix Marseille University, IRD, AP-HM, MEPHI, Marseille, France. ⁸IHU-Méditerranée Infection, Marseille, France. ⁹Contributed equally.

Correspondence: David Montani, Service de Pneumologie, Hôpital Bicêtre, 78, Rue du général Leclerc, 94270 Le Kremlin-Bicêtre, France. E-mail: david.montani@daphp.fr



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A report of multivisceral Whipple's disease with severe precapillary pulmonary hypertension cured by doxycycline and hydroxychloroquine therapy associated with initial oral combination of bosentan and tadalafil. <https://bit.ly/2SECU3L>

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To the Editor:

Whipple's disease (WD) is a rare infectious disease developed through fecal–oral transmission and caused by *Tropheryma whipplei*, a ubiquitous gram bacillus [1]. The pathogenesis remains unclear, and several host factors seem to be implicated, including male sex, comorbidities and genetic susceptibility [1]. WD is a multivisceral disorder with frequent gastro-intestinal, joint and neurological involvement, as well as pulmonary, cardiovascular, mucocutaneous and ophthalmologic lesions [2]. The diagnosis is made *via* small bowel biopsy with periodic acid Schiff staining (PAS) positivity and by PCR on different biological samples [1]. Classical treatment requires prolonged antibiotic therapy and allows rapid improvement. Combination therapy with doxycycline and hydroxychloroquine over 1 year followed by lifetime treatment with doxycycline has been shown to lead to good clinical responses and fewer relapses [3]. Others have recommended initial use of intravenous antibiotics, followed by trimethoprim–sulfamethoxazole for 1 year [4]. Among lung complications of WD, pulmonary hypertension (PH) is very rare and remains poorly understood [5–8]. Here, we report the first well-documented case of a man with concomitant diagnosis of severe precapillary PH and multivisceral WD who had total reversibility in haemodynamics and clinical state after treatment of WD and PH.