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Curing HIV-associated pulmonary arterial hypertension

To the Editors:

Pulmonary arterial hypertension (PAH) results from chronic obstruction of small pulmonary arteries, leading to right ventricular failure and ultimately death. PAH is a severe complication, known to be related to HIV infection [1]. The frequency of PAH in HIV patients is strikingly higher than in the general population and the fourth-leading cause of PAH in the French registry [2]. The incidence of PAH related to HIV (PAH-HIV) has not decreased over time, despite efficient anti-retroviral therapy (ART) [3].

Since its description in 1987, PAH-HIV has been well characterised and multiple therapeutic strategies have been proposed, but no full recovery has yet been published. Here, we report a complete recovery from PAH in two HIV patients with a sustained and fully reversible PAH after a 4-yr discontinuation of bosentan.

Two females, a 36-yr-old with African origins and a 46-yr-old Caucasian intravenous drug abuser received ART 1 month and 25 yrs, respectively after the onset of HIV infection. Normal CD4+ cell count and undetectable HIV viral load were obtained rapidly (fig. 1). Other causes of PAH were ruled out.

For the first patient, bosentan was introduced 12 months after HIV diagnosis. After 5 yrs of persistent haemodynamics and normalisation of functional parameters, bosentan was withdrawn. 4 yrs after discontinuation, the patient remained asymptomatic with normal haemodynamics. For the second patient, bosentan was introduced 5 months after ART. Because of rapid normalisation of functional class and haemodynamic parameters, bosentan was withdrawn only 1 yr later. 2 yrs after bosentan discontinuation, the patient remained asymptomatic with normal haemodynamics.

While haemodynamic normalisation after bosentan has been described previously [1], there is a lack of data on long-term evaluation after specific therapy discontinuation. These patients are the first two cases with PAH-HIV and without other comorbidity in whom long-term vasodilator has been successfully withdrawn for 4 and 2 yrs, respectively.

Haemodynamic normalisation and long-term benefit in bosentan-treated PAH-HIV patients has recently been described, but not yet with full recovery [1] Considering that a complete recovery is a persistent remission over years, even after weaning from specific PAH treatment, there has been, to our knowledge, no description of cured PAH-HIV.

Inflammation may be one underlying mechanism of PAH, so according to the guidelines, we used ART, considering it may be beneficial when associated with another specific PAH treatment [4]. We continued ART therapy even after specific PAH treatment discontinuation and until this day.

As described for experimental inflammatory PAH [5] and other inflammation-related PAHs [6–9], PAH-HIV is more likely to reverse and cure. Consequently, we suggest that pulmonary vasodilators may be cautiously withdrawn in PAH-HIV patients when they fulfil two conditions: a 1-yr haemodynamic normalisation and controlled HIV disease. However, due to lack of data, we recommend long-term close monitoring, with serial systematic haemodynamic catheterisation after bosentan withdrawal.

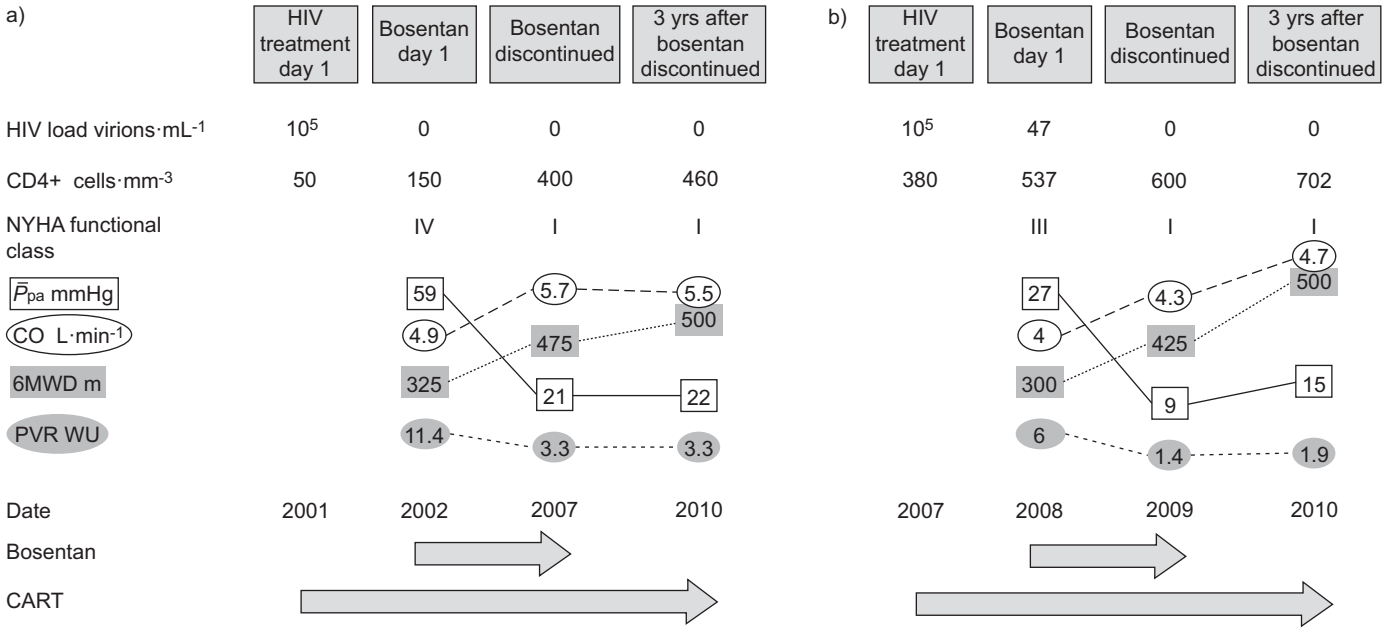

FIGURE 1. Time courses of two patients with pulmonary hypertension (PAH) associated with HIV. NYHA: New York Heart Association; $\bar{P}_{pa}$: mean pulmonary arterial pressure; CO: cardiac output; 6MWD: 6-min walking distance; PVR: pulmonary vascular resistance; WU: Wood units; CART: combination anti-retroviral therapy.

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