A 46-year-old woman was referred to our centre for pulmonary hypertension assessment. She was in World Health Organization functional class 2. Physical examination revealed moderate digital erythema contrasting with severe clubbing of the feet (Fig. 1, panel A). Saturation was 93% in the upper arm and 81% in the lower limb (Fig. 1, panel AB). We evoked the diagnosis of Eisenmenger’s syndrome with right-to-left shunting through a large ductus arteriosus provoking desaturation only in the lower part of the body, in the absence of intracardiac shunting. Transthoracic echocardiography showed normal intracardiac anatomy, paradoxical septum, and a dilated and hypertrophic right ventricle with preserved systolic function (Fig. 2, panels A and B). Based on pulmonary regurgitation, mean pulmonary pressure was measured at 105 mmHg (Fig. 2, panels C and D). The flow through a ductus arteriosus was not visualized by colour Doppler. Intravenous agitated saline contrast injection made the descending aorta and the aortic cross opaque (Fig. 3, panels PA and B), confirming the presence of a large ductus arteriosus, which was evidenced by cardiac magnetic resonance (Fig. 3, panel AC). The haematocrit value was 46%, and the brain natriuretic peptide concentration was 50 pg/mL. Treatment with bosentan was initiated. After 3 months, the patient was in WHO functional class 2, the shunt in the ductus arteriosus was the same and the gradient of saturation between upper and lower limb did not differ. In the 6 min walking test, the patient walked a distance of 420 m; biochemical markers were not modified. The decrease in right ventricular afterload due to the ductus arteriosus may explain the preserved right ventricular function and functional tolerance, which are quite unusual in such severe cases of pulmonary hypertension.
Figure 1. Moderate digital erythema contrasting with severe clubbing of the feet (panel A). Gradient of saturation between the upper arm and the lower limb (panel B).
Figure 2. Transthoracic echocardiography showing normal intracardiac anatomy, paradoxical septum, and a dilated and hypertrophic right ventricle with preserved systolic function (panels A and B). Based on pulmonary regurgitation, mean pulmonary pressure was measured at 105 mmHg (Panels C and D).

Figure 3. Intravenous agitated saline contrast injection made the descending aorta and the aortic cross opaque (panels A and B), confirming the presence of a large ductus arteriosus, which was evidenced by cardiac magnetic resonance (panel C). Ao: descending aorta; DA: ductus arteriosus; PA: pulmonary artery.

Conflict of interest statement

None.