A 18 year-old man presented with gradually progressive dyspnea. Examination revealed grade 3 clubbing and cyanosis of all extremities except the right upper limb (A). Left parasternal heave and palpable second heart sound, but no murmur, were observed.

Electrocardiogram showed right ventricular (RV) hypertrophy (B). Chest radiograph showed dilated pulmonary artery and decreased lung vascularity (C). Contrast echocardiography (D and E) (Online Videos 1, 2, and 3) and computed tomography imaging (F and G) revealed large (18 mm) type B patent ductus arteriosus (PDA) with pulmonary-to-systemic shunting. The RV and pulmonary arteries were dilated with severe pulmonary hypertension (Online Figs. 1, 2, and 3). The patient was managed medically.

Differential cyanosis and clubbing is typical of PDA with Eisenmenger’s syndrome. Shunt reversal causes deoxygenated blood from the RV to be shunted to the aorta (Ao) distal to left subclavian artery (LSCA). This leads to selective affection of lower extremities, sparing both upper extremities. The left upper limb might rarely be involved if the LSCA originates distal to PDA. However, in this case, very large-sized PDA probably caused a jet effect with selective streaming of deoxygenated blood to LSCA, descending Ao, and resultant affection of left upper extremity.

LPA = left pulmonary artery.