A 52-year-old woman was seen at the outpatient clinic for adults with congenital heart disease. Cardiac auscultation revealed a single and loud S2 and a soft pansystolic murmur of tricuspid regurgitation. On clinical examination, clubbing of the toes was noted and cyanosis was present only in the lower half of the body, whereas her fingers did not present evidence of clubbing or cyanosis (Panel A). Using the same pulse oxymeter model simultaneously at her fingers and her toes while breathing room air, differential cyanosis (SO2 toes 80%, SO2 fingers 95%) could be confirmed (Panel B). On 10 L O2 via nasal cannula SO2 on the lower extremities remained unchanged, whereas SO2 increased in the upper extremities. Echocardiographically, a suprasystemic right ventricular systolic pressure was measured (estimated systolic pulmonary arterial pressure 120 mmHg) with an arm-cuff blood pressure measurement of 115/83 mmHg (Panel C). As clinically suspected, a large patent ductus arteriosus (PDA) was found (marked with asterisk in Panel D).

The clinical finding of differential cyanosis as outlined above is pathognomonic for a large untreated PDA associated with Eisenmenger syndrome (shunt reversal into a right-to-left shunt due to progressive pulmonary vascular disease). The right-to-left shunt occurs just distal to the origin of the left subclavian artery, and central cyanosis is not evident in the upper part of the body. The continuous murmur of a large PDA is usually absent as balanced pressure in the systemic and pulmonary circulation is present (making it often difficult to depict flow by colour Doppler).