

CLINICAL IMAGES

Differential clubbing and cyanosis in a patient with pulmonary hypertension

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A 41-year-old man was admitted with hemoptysis and a 10-year history of progressive exertional decline. He had a patent ductus arteriosus diagnosed in adult life that was unrepaired because of concomitant pulmonary hypertension. On examination, he had a sustained right ventricular impulse with a palpable second heart sound. The continuous murmur usually heard in patent ductus arteriosus was not audible. Cyanosis and clubbing were more profound in the toes compared with the fingers (Figure 1). Oxygen saturation in the toes on room air was 88% compared with 96% in the fingers.

Echocardiography showed elevated pulmonary artery pressures with bidirectional flow through a large patent ductus arteriosus (Appendix 1, available at www.cmaj.ca/cgi/content/full/cmaj.091003/DC1). Cardiac catheterization was concordant, with a main pulmonary artery systolic pressure of 91 mm Hg, versus 93 mm Hg in the ascending aorta. The presence of shunting was confirmed with an increase in oxygen saturation from 75% in the right ventricle to 90% in the main pulmonary artery. Cardiac computed tomography confirmed the presence of a large patent ductus arteriosus (Appendix 1), thus establishing the cause of the Eisenmenger syndrome in this patient.

A patent ductus arteriosus is a normal neonatal finding, which generally disappears within the first few days of life. If a significant patent ductus arteriosus persists, flow is left-to-right throughout the cardiac cycle during childhood, which may result in pulmonary vascular remodelling, bidirectional flow and Eisenmenger syndrome (increased pulmonary vascular resistance and pulmonary artery hypertension). Blood can bypass the lungs via the patent ductus arteriosus, resulting in direct perfusion of the lower limbs with desaturated blood.

The diagnosis of patent ductus arteriosus may be challenging in the patient with Eisenmenger syndrome, when systemic level pulmonary artery pressure effectively abolishes



Figure 1: Toes and fingers of a 41-year-old man admitted with hemoptysis. The toes show more profound cyanosis and clubbing than the fingers.

the usual “machinery” murmur. Differential clubbing and cyanosis is a clue to the diagnosis of patent ductus arteriosus related to Eisenmenger syndrome.

The development of significant pulmonary hypertension precludes surgical or interventional ductal closure. In our patient, the hemoptysis settled with conservative management. In the longer term, progressive decline in exercise capacity or symptomatic right heart failure can only be treated with heart and lung transplantation.¹

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REFERENCE

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