Cardiac imaging in a patient with differential clubbing

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Eisenmenger's syndrome (ES), a severe form of pulmonary arterial hypertension (PAH), occurs in patients with congenital heart defects (CHDs) if large shunts are not closed in time.[1] Differential clubbing is a rare finding and could be a characteristic of patent ductus arteriosus (PDA).[2] We present a rare case of differential clubbing in a patient with ES due to PDA.

A 36-year-old man with a history of CHD and PAH presented with complains of exertional dyspnea and heart palpitations. PDA was diagnosed at the age of 5 years and the defect was not closed due to severe PAH (confirmed by right heart catheterization), therefore ES developed.

On physical examination, significant clubbing of toes (drumstick toes and watch-glass nails) compared with fingers was observed (FIGURE 1A). Arterial blood gas tests showed hypoxemia in legs in comparison with hands (FIGURE 1A). An accentuated second heart sound in the second left intercostal space was audible.

Transthoracic echocardiography revealed hypertrophy and dilatation of the right ventricle with pressure overload (systolic leftward shift of the interventricular septum), enlarged pulmonary artery (PA), mild tricuspid regurgitation, PDA with undetectable shunt (FIGURE 1B), signs of severe pulmonary hypertension (peak tricuspid regurgitation velocity, 5.6 m/s; estimated systolic PA pressure, 135 mm Hg). The patient refused to repeat right heart catheterization but consented to noninvasive imaging. Computed tomography pulmonary angiography and magnetic resonance angiography (MRA) were performed showing a large (diameter, 23 × 14 mm) and short PDA (FIGURE 1C and 1D) with reversal shunt (flow of contrast from the PA to ascending aorta, distal to the subclavian arteries; FIGURE 1D).

Patient was managed with double combination targeted therapy of PAH with endothelin receptor antagonist and phosphodiesterase type 5 inhibitor.

When the arterial duct remains patent, it results in a left-to-right shunt because of blood flow from the high-resistance descending aorta into the low-resistance left PA.[3] Therefore, pulmonary blood flow becomes higher. CHD with left-to-right shunt and high pulmonary blood flow is characterized by severe PAH.[4] When pulmonary resistance becomes higher than systemic, the shunt changes direction from pulmonary to systemic and deoxygenated blood flows from the PA through the PDA to the proximal descending aorta.[3] Therefore, deoxygenated blood is delivered to the lower extremities; however, the upper extremities are supplied by oxygenated blood through branches of the aortic arch proximal to the PDA. This clinically results in differential clubbing and cyanosis.[5] It is an important diagnostic clue for PDA complicated with ES.[1]

Transthoracic echocardiography is the principal diagnostic examination, but a correct diagnosis can be difficult to establish in patients with ES due to absent color doppler flow in the presence of equal systemic and pulmonic pressures.[1] Multimodality imaging (computed tomography pulmonary angiography and MRA) are indicated for additional evaluation of PDA size and location, direction of flow across the PDA.[1] As in this case, MRA confirmed that PDA with reversed shunt is the cause of differential clubbing.

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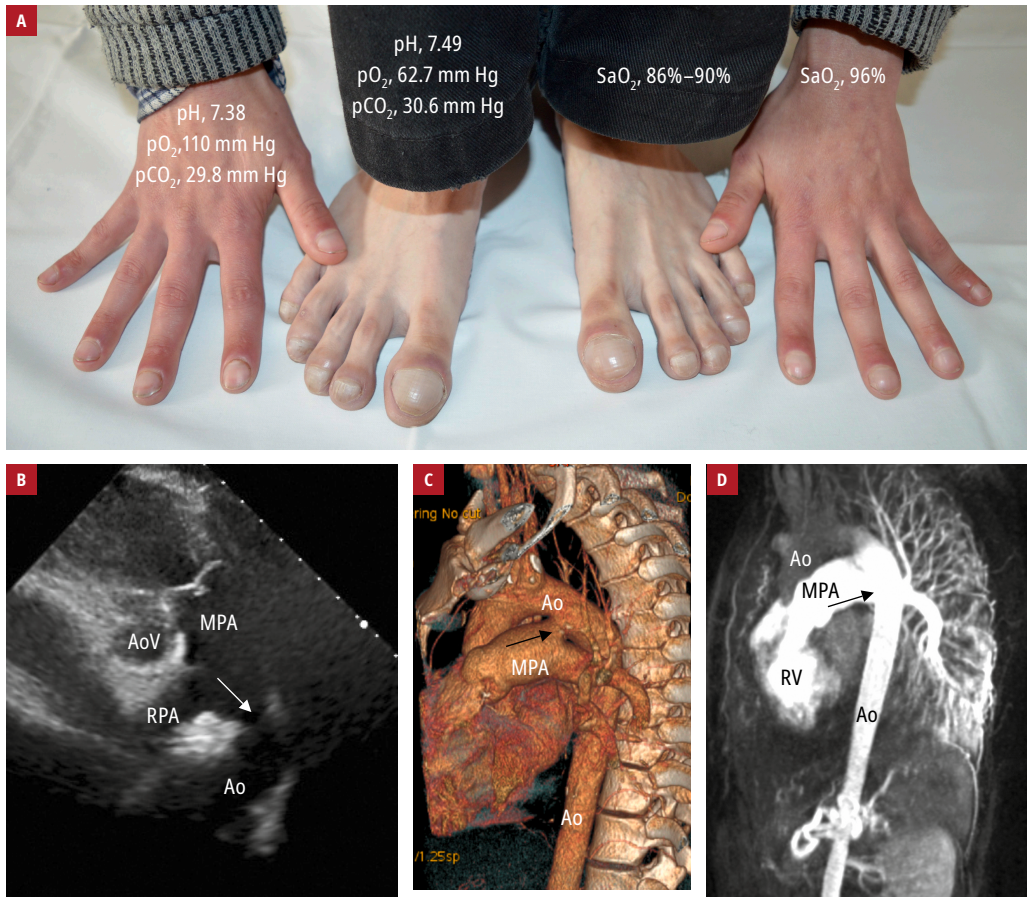

FIGURE 1 **A** – significant clubbing of toes (drumstick toes and watch-glass nails) compared with fingers; **B** – patent ductus arteriosus (arrow) on transthoracic echocardiography; **C** – patent ductus arteriosus in 3-dimensional reconstruction of a computed tomography pulmonary angiography scan (arrow); **D** – magnetic resonance angiography of the chest showing early filling (during pulmonary arterial phase) of the descending aorta through patent ductus arteriosus (arrow) suggesting more unsaturated blood going to the lower part of the body

Abbreviations: Ao, aorta; AoV, aortic valve; MPA, main pulmonary artery; pCO_2, partial pressure of carbon dioxide; pO_2, partial pressure of oxygen; RPA, right pulmonary artery; RV, right ventricular; SaO_2, oxygen saturation

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