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Cardiac imaging in a patient with differential clubbing

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Short title: Imaging in 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 (CHD) if large shunts are not closed on time [1]. Differential clubbing is a rare finding and could be 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 complaints 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 catheterisation), therefore ES developed. During physical examination significant clubbing of toes (drumstick toes and watch-glass nails) compared to fingers was observed (Figure 1 (Panel D)). Arterial blood gas tests showed hypoxemia in legs in comparison to hands (Figure 1 (Panel D)). 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 1 (Panel A)), signs of severe pulmonary hypertension (peak tricuspid regurgitation velocity 5.6 m/s, estimated systolic PA pressure 135 mmHg). The patient refused to repeat right heart catheterisation but consented for non-invasive imaging. Computed tomography pulmonary angiography and magnetic resonance angiography (MRA) were performed showing a large (diameter 23 x 14 mm) and short PDA (Figure 1 (Panel B, C)) with reversal shunt (flow of contrast from PA to ascending aorta, distal to the subclavian arteries (Figure 1 (Panel C)).

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 development of 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 instrumental examination, but precise diagnosis could be difficult to establish in patients with ES due to absent colour doppler flow in equal systemic and pulmonic pressures, anatomic variations of CHD [1]. Multi-modality 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 approved that PDA with reversed shunt is the cause of differential clubbing.
References:


Figure 1. A – patent ductus arteriosus (white arrow) in transthoracic echocardiography; B – patent ductus arteriosus in 3D reconstruction of a CTPA scan (black arrow); C – MRA of the chest shows early filling (during pulmonary arterial phase) of the descending aorta through patent ductus arteriosus suggesting more unsaturated blood going to the lower part of the body; D – significant clubbing of toes (drumstick toes and watch-glass nails) compared to fingers.