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Images in Medicine: "Squatting"

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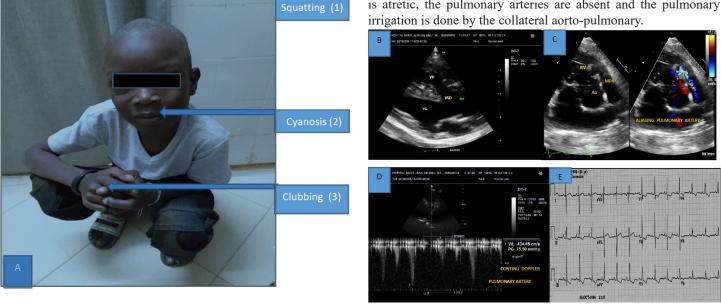
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"Squatting"

We receive in consultation N.N. a 12-years-old boy with shortness of breath and squatting after exertion. The squatting (Image A1) was very evocative of tetralogy of Fallot (ToF) seen late as is often the case in Africa [1,2]. La maladie bleue described in 1888 by Etienne –Louis Arthur Fallot is the most common cyanotic congenital heart disease. ToF embryological basis is the anterocephalad deviation of muscular outlet septum responsible of the combination of cardinal lesions: stenosis of the right ventricular out flow tract (RVOT), ventricular septum defect (VSD), over-riding aorta originating above the VSD, right ventricular hypertrophy parenthesis (RVH) [3,4].

The squatting, is a reflex attitude who increase systemic blood pressure in the limbs and aorta and promotes the passage of blood from the right ventricle into the pulmonary artery improving oxygenation of the body. Cyanosis and chronic hypoxia explain clubbing (A2 and A3). Blood oxygen saturation was 82%. Auscultation revealed a systolic murmur of the RVOT stenosis at the left third intercostal space. ECG shows a right axis deviation and R/S ratio greater than 1 in lead V1 characteristic of RVH (E). Transthoracic echocardiography establishes the diagnosis by the visualization in the long axis (B) of the VSD, the over-riding aorta and in small-axis (C) of a narrow infundibulum with a color Doppler aliasing suggestive of stenosis confirmed by continuous Doppler recording a high gradient at 75 mm Hg (D). ToF has in reality a broad anatomical spectrum. In the light forms the obstruction is located only at the level of the infundibulum of the right ventricle whereas in the severe forms the pulmonary valve is atretic, the pulmonary arteries are absent and the pulmonary



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The origin and initial course of the coronary arteries, the atrial septum and inter ventricular septum and the aortic arch should be carefully examined for associated abnormalities. Echocardiography may not be sufficient to provide all the necessary anatomical details for preoperative evaluation and cardiac catheterization or less invasive cardiac MRI will be required [5].

The treatment of ToF is eminently surgical ranging from early complete correction at the age of 4 to 6 months at one end to a multi-stage reconstruction of the pulmonary way at the other end [4]. The anatomical form presented by N.N was not very severe explaining late diagnosis and a couple months later he undergo surgical complete repair in Tübingen (Germany). He can go back now to school in his village and play soccer with his friends demonstrating that It was still necessary to operate children presenting with TOF irrespective of their age [1,2].

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