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Images in Congenital Heart Disease

Late signs of cyanotic congenital cardiac disease

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A 17-YEAR-OLD BOY CAME TO OUR DEPARTMENT 4 months after the discovery of a congenital cardiac anomaly, and was found to have symmetrical clubbing of the fingers (Fig. 1a) and a blue tongue (Fig.1b). These are particularly good signs of cyanosis in dark people. He also had a loud systolic ejection murmur along the left parasternal area, discrete hepatomegaly, and a transcutaneous arterial saturation of oxygen of about 67%. The electrocardiogram showed sinus rhythm, with a superior QRS axis. The chest X-ray revealed a normal cardiac silhouette with oligaemic lung fields. His haemoglobin was 240 grams per litre, with a haematocrit of 75.8% and nearly 9 millions red cells per cubic millimetre (Fig. 1c). Transthoracic echocardiography confirmed the clinical diagnosis of absent right atrioventricular connection (*), with a functionally univentricular heart of left ventricular dominance, supravalvar pulmonary stenosis, and a large non-restrictive interatrial communication (Fig. 2). In view of poor ventricular function, and extremely hypoplastic pulmonary arteries, conservative palliation was not possible. Because cardiac transplantation in Mozambique was not possible either, his treatment was limited to optimization of medical management. The late discovery of such an advanced picture of cyanotic congenital cardiac disease is now exceptional in developed countries, but not so rare in sub-Saharan Africa.

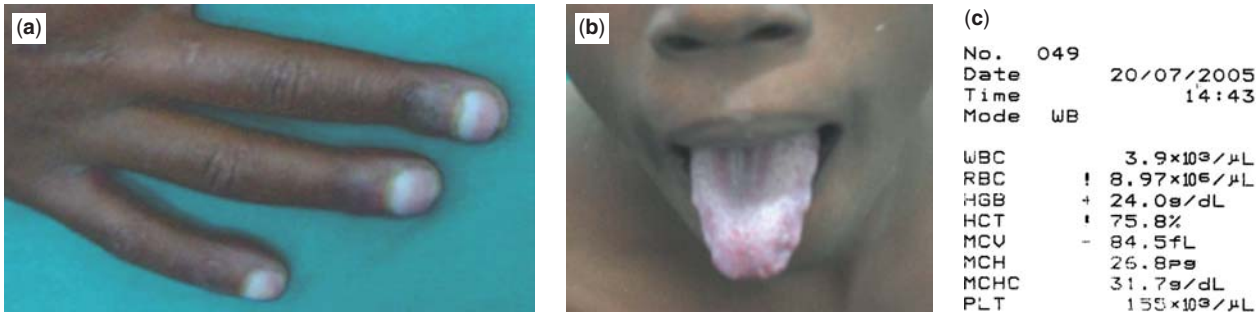

Figure 1.

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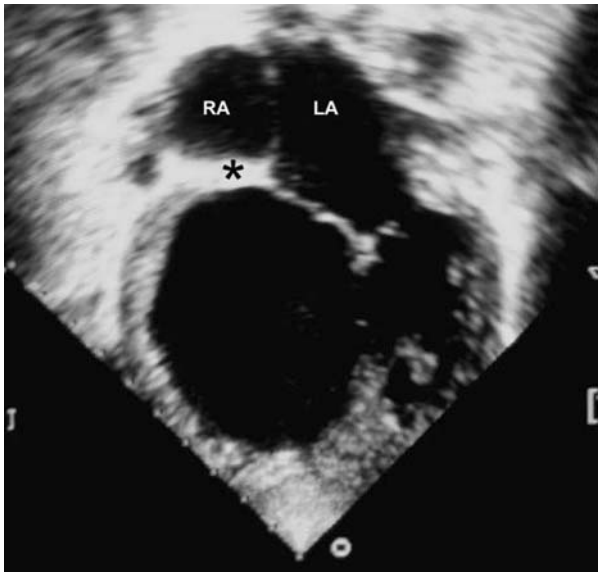

Figure 2.

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