

IMAGE IN CARDIOLOGY

Complex congenital heart disease with absent pulmonary arteries[☆]



Cardiopatia congénita complexa com ausência de artérias pulmonares

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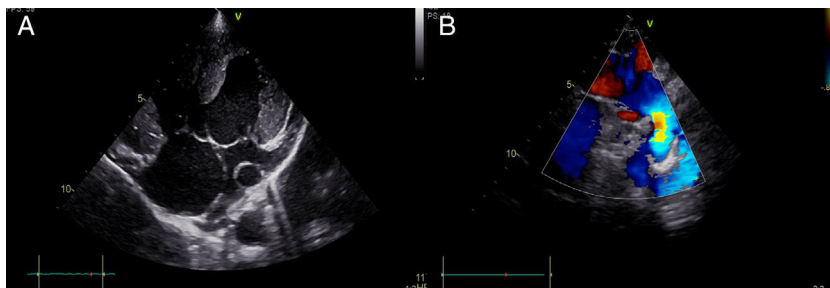


Figure 1 Echocardiogram. (A) Four-chamber view showing a large interventricular communication and severely dilated coronary sinus; (B) aortic arch, showing the origin of two major aortopulmonary collateral arteries.

A six-year-old girl, native of Guinea-Bissau, with a history of dyspnea on minimal exertion, recurrent respiratory infections and poor weight gain, was admitted to a reference center due to suspected tetralogy of Fallot with a view

to possible surgery. Physical examination revealed central cyanosis with digital clubbing; oxygen saturation was 30–50% on pulse oximetry and pulmonary auscultation showed no murmur and normal pulses.

The echocardiogram revealed complex cyanotic heart disease including transposition of the great arteries (TGA), pulmonary atresia, a large interventricular communication and systemic-pulmonary collateral arteries. No pulmonary arteries were visible but there was a persistent left superior vena cava with dilated coronary sinus (Figure 1).

Cardiac catheterization confirmed the existence of collaterals originating from the supra-aortic trunks, the

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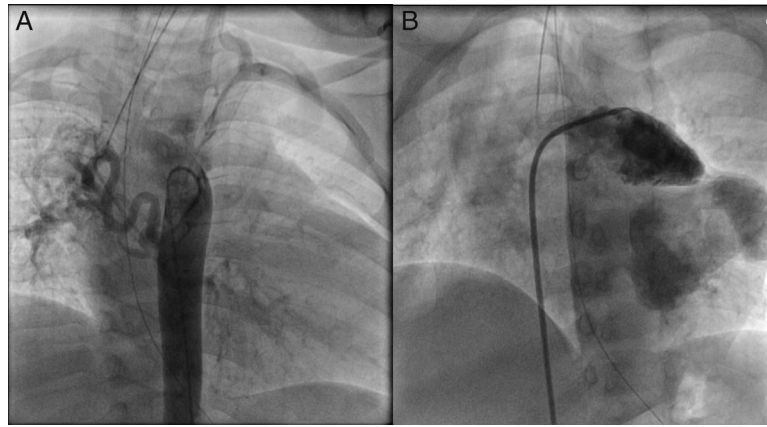


Figure 2 Cardiac angiography. (A) Emergence of a large and slightly tortuous major aortopulmonary collateral artery; (B) left juxtaposition of the atrial appendages.

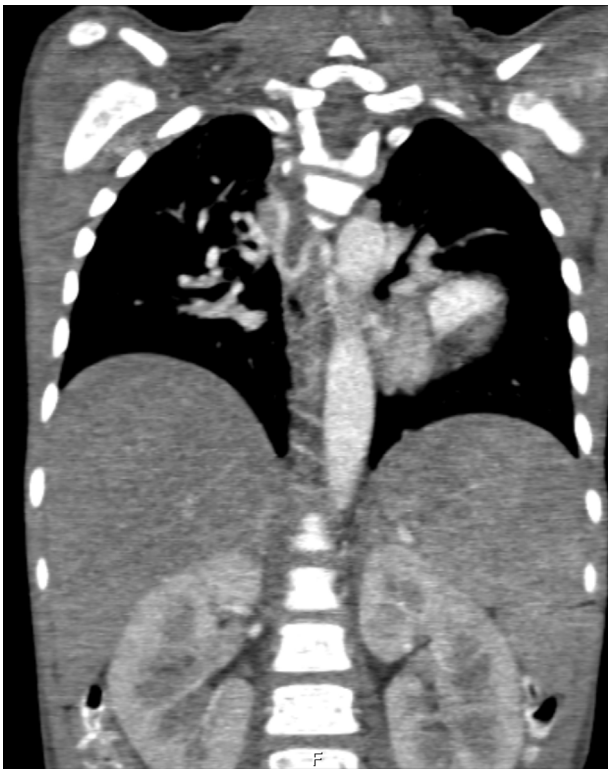


Figure 3 Computed tomography angiography showing a large major aortopulmonary collateral artery with stenosis of its proximal portion.

absence of central pulmonary arteries, and left juxtaposition of the atrial appendages (Figure 2). Computed tomography angiography revealed three major aortopulmonary collateral arteries (MAPCAs), one of which originated from the upper portion of the aortic arch, with severe stenosis at its origin and dilatation more proximally. It also showed absence of the pulmonary arteries, a 25-mm interventricular communication, and normal drainage of the pulmonary veins into the left atrium (Figure 3).

The authors present a rare case of complex heart disease, diagnosed late, which remains a common problem in less developed countries. The presence of MAPCAs allowed the patient to survive, but their natural evolution to stenosis led to progressive worsening of the patient's condition.

Surgical repair in this case will depend on unifocalization of the MAPCAs, which will be a highly complex operation due to the complete absence of the pulmonary arteries and the presence of TGA. The prognosis is accordingly reserved.

Conflicts of interest

The authors have no conflicts of interest to declare.