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IMAGE IN CARDIOLOGY

Tetralogy of Fallot with absent pulmonary valve☆

Tetralogia de Fallot com agenesia da válvula pulmonar

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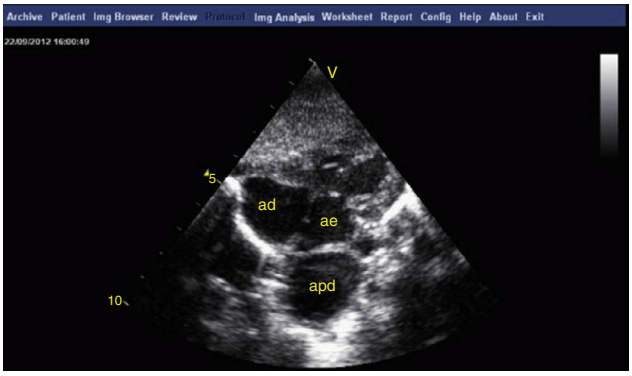

Figure 1 Transthoracic echocardiogram, subcostal 4-chamber view, showing the right pulmonary artery compressing the left atrium. ad: right atrium; ae: left atrium; apd: right pulmonary artery.

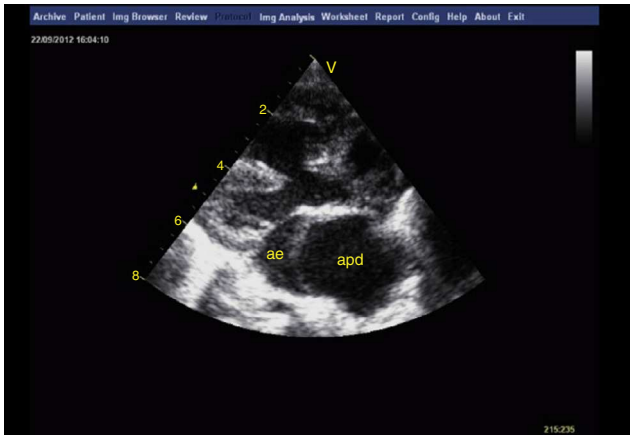

Figure 2 Transthoracic echocardiogram, parasternal long-axis view, showing perimembranous ventricular septal defect and overriding aorta (50%), and aneurysmal dilatation of the right pulmonary artery compressing the left atrium. ae: left atrium; apd: right pulmonary artery.

A 4-month-old female infant, normal karyotype, born in the Azores of non-consanguineous parents by uncomplicated eutocic delivery at full term after a monitored pregnancy without complications and Apgar score 5/9/10, had been admitted to the intensive care unit at two hours of life due to hypoxemia and grade IV/VI systolic murmur. She was diagnosed remotely with tetralogy of Fallot with absent pulmonary valve on the first day of life and was transferred to Hospital Santa Cruz with body weight of 3.695 kg (<5th percentile) for surgical correction of cyanotic heart disease. She was slightly pale and markedly polypneic, with systolic and diastolic murmurs on the left sternal border, but no other relevant alterations on physical

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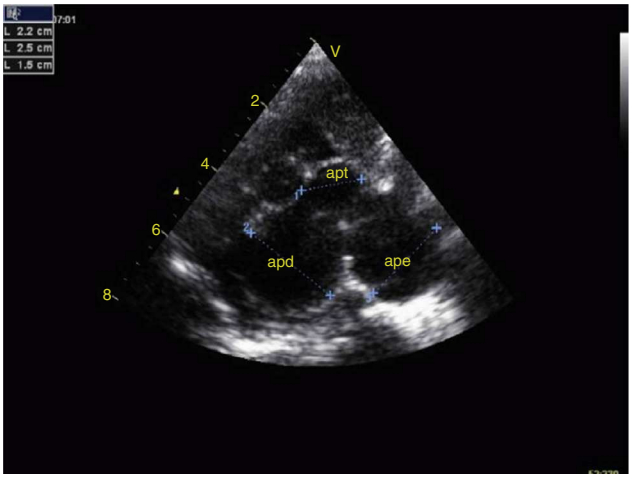

Figure 3 Transthoracic echocardiogram, parasternal short-axis view, showing aneurysmal dilatation of the pulmonary artery trunk (15 mm) and left (22 mm) and right (25 mm) branches. apd: right pulmonary artery; ape: left pulmonary artery; apt: pulmonary artery trunk.

examination. The echocardiogram confirmed tetralogy of Fallot and revealed non-restrictive perimembranous ventricular septal defect, overriding aorta (50%), infundibular stenosis due to anterocephalad deviation of the infundibular septum and muscle band, hypoplastic pulmonary annulus and absence of the pulmonary valve, the leaflets replaced by granulomatous masses causing severe obstruction (peak right ventricle-pulmonary artery gradient of 100 mmHg) and severe free pulmonary regurgitation. There was also aneurysmal dilatation of the pulmonary artery trunk (15 mm) and branches (right branch 25 mm and left branch 22 mm) (Figures 1–3).

These findings were compatible with tetralogy of Fallot with absent pulmonary valve. The patient underwent total surgical repair with reduction of both branches of the pulmonary artery and transposition of the great arteries (Lecompte maneuver). The operation was uneventful but the patient died 56 hours later from respiratory failure.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Conflicts of interest

The authors have no conflicts of interest to declare.