

Images in Congenital Heart Disease

Pericardial effusion, arrhythmogenic right ventricular dysplasia and apical diverticulum: an uncommon association

Cláudia Moura, Jose C. Areias, Jorge Moreira

Paediatric Cardiology Center, Hospital of S. João, Oporto, Portugal

A 14-YEAR-OLD ASYMPTOMATIC BOY WAS admitted because of cardiomegaly on his chest X-ray. The echocardiogram (Fig. 1) disclosed a circumferential pericardial effusion and a saccular image within the apical region of the right ventricle. Hypokinesia of the anterior and inferior segments of the right ventricle was evident. His electrocardiogram showed right bundle branch block and no arrhythmias. A pericardiocentesis revealed exudate, but no etiologic agent was identified.

In the following 6 months the effusion recurred twice with similar findings on drainage of the fluid, and therefore cardiac catheterization was performed. Angiography in the right ventricle showed a diverticulum near its apex and an enlarged cavity with

depressed regional motion. The trabeculations in the anterior wall were clearly abnormal in systole and diastole (Figs 2 and 3 respectively), suggesting arrhythmogenic right ventricular dysplasia. Magnetic

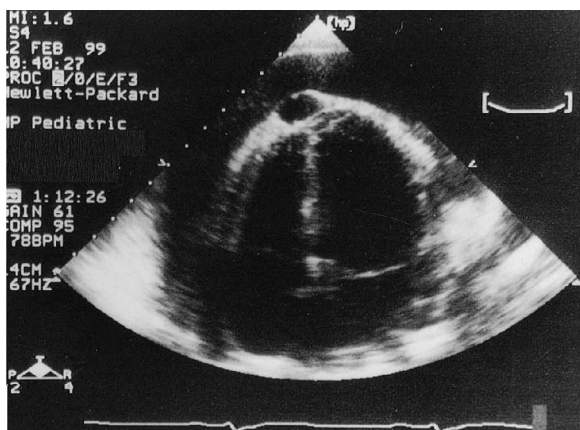


Figure 1.



Figure 2.

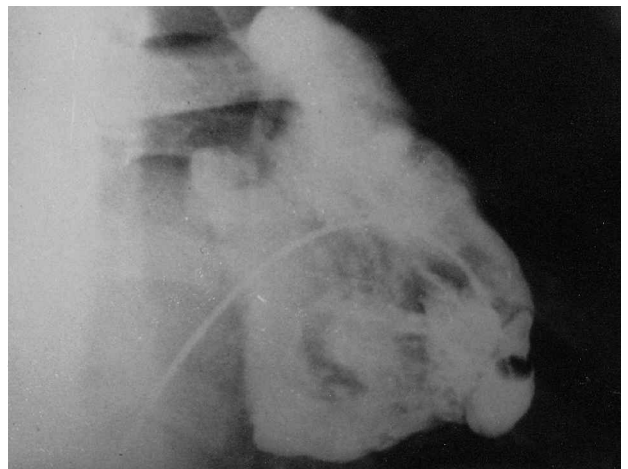


Figure 3.

Correspondence to: Dr Cláudia Moura, Paediatric Cardiology Center – Hospital of S. João, Alameda Professor Hernani Monteiro, 4202-451 Porto, Portugal. Fax: 225505919; E-mail: cmoura@hsojoao.min-saude.pt; claudiammoura@yahoo.com

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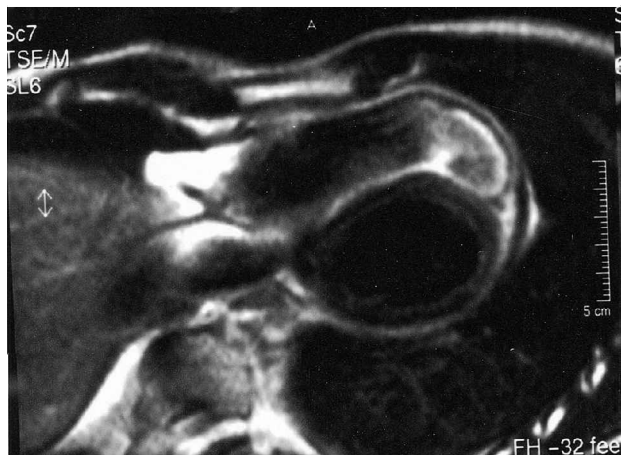


Figure 4.

resonance imaging (Fig. 4) disclosed myocardial thinning in the anterior wall of the right ventricle.

Ten months after he was first seen, creation of a pleuro-pericardial window was done to control the huge effusions, which recurred despite oral anti-inflammatory treatment.

The clinical evolution of this adolescent thus far has been good, and at the age of 16, he remains well, with no further effusions and no records of arrhythmia. As pressures in the right heart are normal, resection of the diverticulum has not been considered, although we recognize that rupture is one of the potential complications.