

Uhl's disease

JORGE VINICIO ARAQUE RIVADENEIRA, FERNANDO L. HIDALGO O.

UHL'S DISEASE

A child of five years old, with several hospitalizations for severe right heart failure, without a definitive diagnosis. The echocardiogram shows severe dilatation and hypokinesis of the right ventricle. The tricuspid valve has normal morphology and implantation, with severe impairment. It draws the attention the presence of very thin walls in the right ventricle. Pericardial effusion. Left ventricle is anatomically and functionally normal (Figures 1 and 2). Cardiac magnetic resonance imaging (Figure 3) showed an important dilatation of the right ventricle and very thin walls, suggesting the absence of myocardium, without fatty or fibrous infiltration.

These changes suggest the presence of a right ventricular cardiomyopathy, secondary to arrhythmogenic dysplasia or Uhl's anomaly. The age of presentation, medical history and data from imaging studies tipped up the diagnosis to Uhl's anomaly.

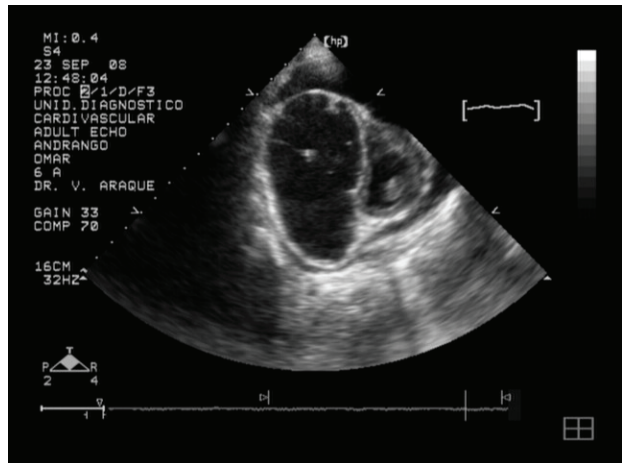


Fig. 1. Two-dimensional echocardiographic image (parasternal, short axis), important right ventricular dilatation, with very thin walls.

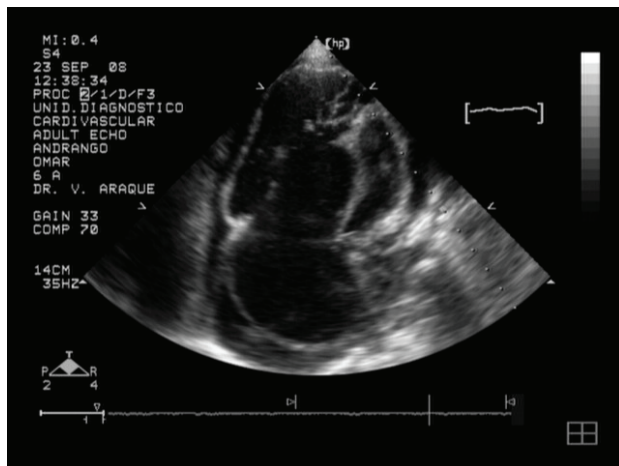


Fig. 2. Two-dimensional echocardiographic image (apical 4-chamber view), normal tricuspid valve implant (arrows).

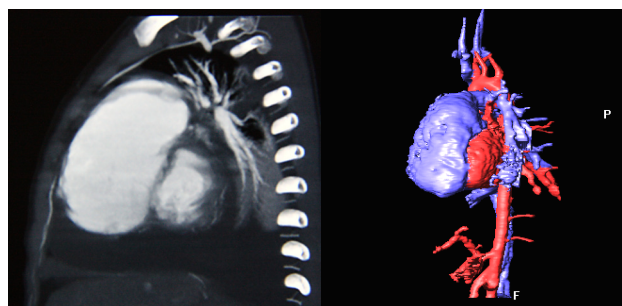


Fig. 3. Cardiac magnetic resonance imaging. **A.** Lack of fatty or fibrous infiltration. **B.** Volumetric reconstruction where is observed an important dilatation of the right ventricle (blue).