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Advanced Echocardiographic Assessment of Ebstein Anomaly in Children

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We describe the case of a 4-year-old female patient, diagnosed with Ebstein anomaly at the age of 1 year, who presented progressive deterioration of physical exercise capacity and failure to thrive. The physical examination revealed a 4/6 grade holosystolic murmur, the BMI was at 3.7 percentile, and SaO_2 was 90% on room air. The remainder of the physical examination was unimportant. The chest X-ray showed right sided chamber enlargement.

We have performed a complex echocardiographic assessment with an Epiq 7 (Philips Medical Systems) ultrasound system using an X5-1 xMatrix array transducer. The 2D echocardiogram revealed a dilated right atrium (RA) and right ventricle (RV) with dilation of the tricuspid annulus (35.9 mm, z = 2.92), and mild atrialization of the RV due to an abnormal tricuspid valve. The apical four-chamber image demonstrated tethering of the tricuspid septal leaflet to the ventricular septum, with a 24.3 mm/m^2 displacement index. The anterior leaflet was large, redundant, "sail-like", with dense attachments to the RV free wall. The left ventricle and atrium were slightly compressed by the volume overload in the right heart. There was a foramen ovale with a bidirectional shunt. We have traced the chamber areas (Figure 1A). According to the Carpentier classification, this was a type A Ebstein anomaly, with an adequate true RV volume.[1] We have also calculated the Great Ormond Street Echocardiogram (GOSE) score, defined as the ratio between the combined RA area (RA + atrialized RV) to that of the functional RV area + left atrium area + left ventricular area, at end-diastole, in apical four-chamber view.[2] The GOSE score was 0.85 (grade II) in our patient. Color Doppler examination revealed a severe tricuspid regurgitation (Figure 1B).

Speckle-tracking analysis of the RV free wall demonstrated a well-functioning RV, with a free wall strain of about –19.6%, with the following segmental strain values: basal –19%, mid –18%, apical –22% (Figure 1C).

We have also performed a 3D echocardiographic assessment of the tricuspid valve (Figure 2). The multiplanar reconstruction data and the 3D enface view of the tricuspid valve from ventricular perspective demonstrated a small, rudimentary septal leaflet(s) besides the interventricular septum (Figure 2-s) and a

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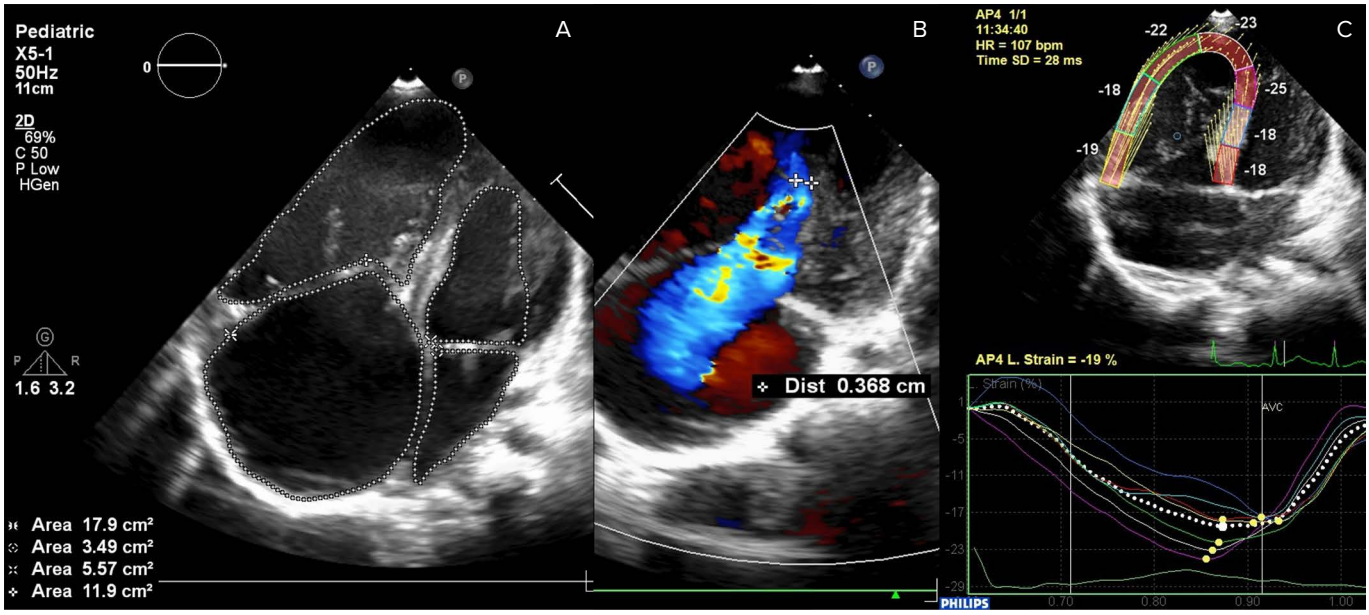

FIGURE 1. **A** – Apical four-chamber echocardiographic view with traced chamber areas, showing dilated right chambers. **B** – Color Doppler examination revealed a severe tricuspid regurgitation. **C** – Speckle-tracking analysis of the RV free wall demonstrated a well-functioning RV, with a free wall strain of about –19.6%.

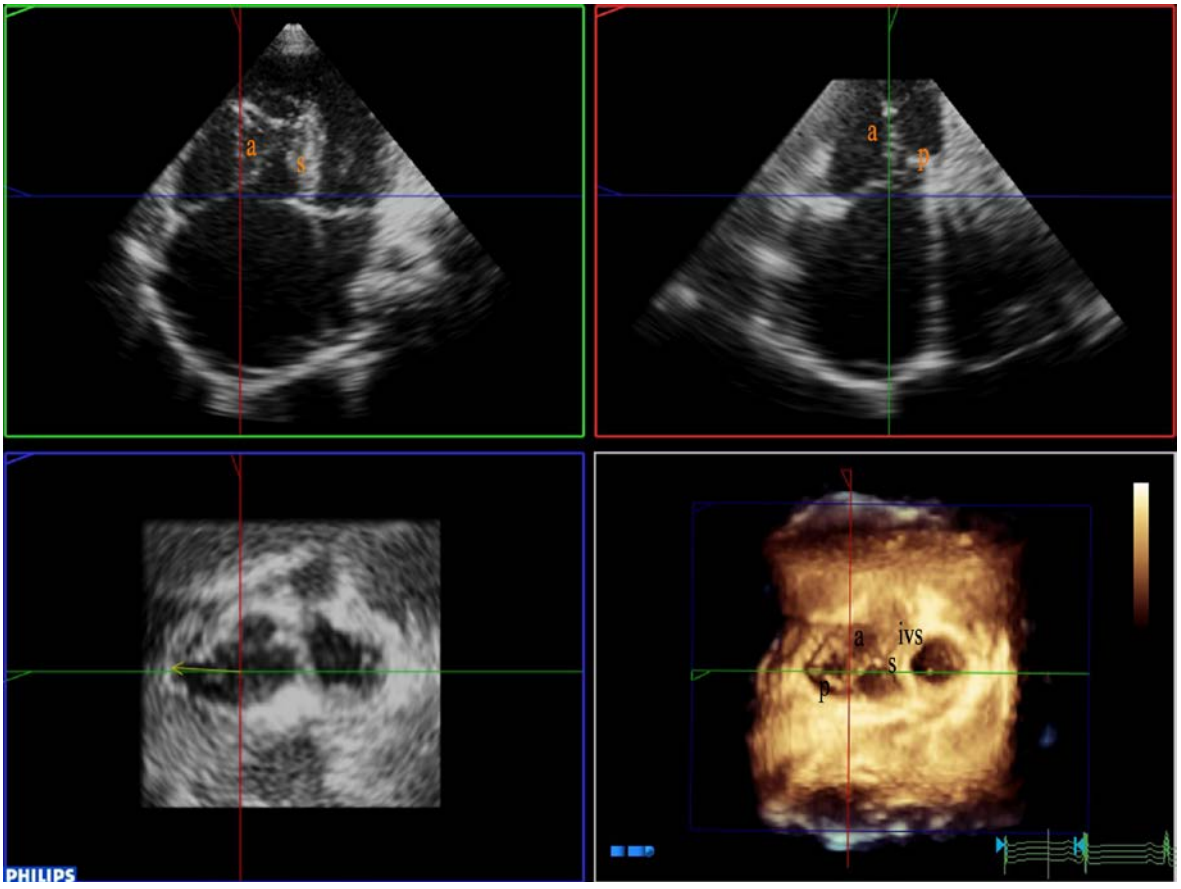

FIGURE 2. Multiplanar reconstruction data and the 3D enface view of the tricuspid valve from ventricular perspective with a rudimentary septal leaflet(s) besides the interventricular septum (s) and a large, sail-like anterior leaflet (a), with slight coaptation, visible on the corresponding four-chamber view, and also a small, tethered posterior leaflet (p), with poor coaptation between the posterior (p) and septal leaflet (s) on three dimensional display.

large, sail-like anterior leaflet (Figure 2-a), with slight coaptation, visible on the corresponding four-chamber view, and also a small, tethered posterior leaflet (Figure 2-p), with poor coaptation between the posterior (p) and septal leaflet (s) on 3D display.

Three-dimensional echocardiography complements the conventional echocardiographic (2D and color Doppler) assessment of the TV in Ebstein anomaly.[3–6] It allows a better description of leaflet morphology and spatial geometry of the tricuspid valve in this particular disease and thus can lead to a more effective management or surgical technique. Also, deformation imaging is useful in the assessment of the myocardial function of the RV.[7]

CONFLICT OF INTEREST

The authors declare no conflict of interests.

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