A new variety of double-chambered left ventricle

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An 11-year-old girl with aortic stenosis diagnosed at the age of 2 years presented with a 3-year history of increasing effort angina, syncope, and NYHA class IV. A transthoracic echocardiogram revealed a dysplastic trileaflet aortic valve with a peak gradient of 70 mmHg. The left ventricle (LV) was divided into two separate chambers by a thick non-contractile membrane traversing the LV cavity horizontally at the level of the papillary muscles.

Cardiovascular magnetic resonance (CMR) confirmed the presence of severe aortic valve stenosis (peak velocity 5 m/s). The LV was divided into contractile basal and severely hypokinetic apical halves by a complete membrane (Panels A–F; see Supplementary material online, Movie S1). The basal half received mitral inflow and ejected to the aorta; the apical half had no inlet or outlet, but nevertheless contained liquid blood. The apical half contributed minimally to ejection through basal displacement of part of the membrane in systole. Both papillary muscles appeared to pass through the membrane.

The patient underwent a Ross procedure and resection of the LV mid-cavity obstruction and made a good post-operative recovery. Intraoperative inspection demonstrated a fibrous shelf at the level of the papillary muscles in the mid-LV cavity, with no thrombus in the apical portion. Post-operative echocardiography and CMR showed no LV outflow tract obstruction and good LV systolic function, including good contractility of the apical portion.

Subdivision of the LV cavity is a rare cardiac anomaly. The case reported here is unique, in that the structure dividing the LV into basal and distal chambers was a non-muscular fibrous shelf, rather than muscular bands or accessory muscular pseudo-septa (resulting in side-by-side chambers), as in previously reported cases. A comparison of the images suggests pre-operative hibernation of myocardium of the apical half of the LV, which showed post-operative recovery of contractile function (see Supplementary material online, Movie S2).

Supplementary material is available at European Heart Journal online.

Late systolic frames of pre-operative (Panels A, C, and E) and corresponding post-operative CMR cine images (Panels B, D, and F). The obstructing mid-ventricular membrane is indicated by the white arrowheads. Panel A and Supplementary material online, Movie S1 show the jet through the stenosed aortic valve (black arrowhead). Panel B and Supplementary material online, Movie S2 show unobstructed flow through the replacement autograft valve.