Clinical vignette

Double-chambered right ventricle

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An asymptomatic 11-year old boy with a history of spontaneously regressed right ventricle (RV) rhabdomyoma was referred to our institution for cardiovascular magnetic resonance (CMR), because a recent transthoracic echocardiography (TTE) study raised the suspicion of double-chambered RV (DCRV). At the auscultation, a loud grade (4/6) systolic murmur was heard at precordium.

CMR revealed an aberrant fibro-muscular band (arrowheads) originating from the interventricular septum and dividing the RV cavity into a high-pressure chamber in the inlet part (IL-RV) and a low-pressure chamber in the outlet part (OL-RV) (Panels A and B). No other congenital defects have been visualized. The magnitude (Panel C) and phase-encoded flow (Panel D) images showed an accelerated blood flow at level of abnormal band and peak pressure gradient of 77 mmHg was measured. These findings confirmed the diagnosis of DCRV. This is principally a congenital abnormality, rare in the isolated form since it is often associated with ventricular septal defect or other congenital heart disorders. In our case, the muscular band was likely present at birth, although was probably concealed by tumour. It was minimally represented. Indeed, the obstruction may develop over time when progressive band hypertrophy occurs. The clinical presentation of DCRV differs from the absence of symptoms to syncpe or angina according to the degree of obstruction. Typically, DCRV is diagnosed in childhood or adolescence. TTE has shown good accuracy for diagnosis in infants but not in adolescence and adults, rendering the use of CMR desirable. Surgery is the treatment of choice and it is advisable before RV dysfunction supervenes.