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Right and Left Ventricular Outflow Tract Obstruction in Hypertrophic Cardiomyopathy

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Hypertrophic obstructive cardiomyopathy (HOCM) affecting both the right and left ventricular (RV and LV) outflow tracts is rare, and the mechanism of obstruction is different in each ventricle. A 21-year-old man with a family history of HOCM and sudden death was referred with breathlessness (New York Heart Association grade II) and occasional presyncope. There had been no discharges of an implantable defibrillator inserted 2 years earlier, and the sole medication was a β -blocker. A transthoracic echocardiogram revealed marked, bilateral ventricular hypertrophy. Doppler

revealed a 70 mm Hg LV outflow gradient and obstruction of the right outflow tract at rest. Right and left heart catheterization, with simultaneous central aortic and LV pressure recording, revealed a 100 mm Hg subaortic gradient (Figure, a) and a 75 mm Hg subpulmonic gradient at rest (Figure, b and c). Ventriculography confirmed that the obstruction of the LV outflow was due to systolic anterior mitral valve movement (Figure, d and e), whereas RV outflow obstruction was due to massive hypertrophy of the crista supraventricularis (Figure, f).

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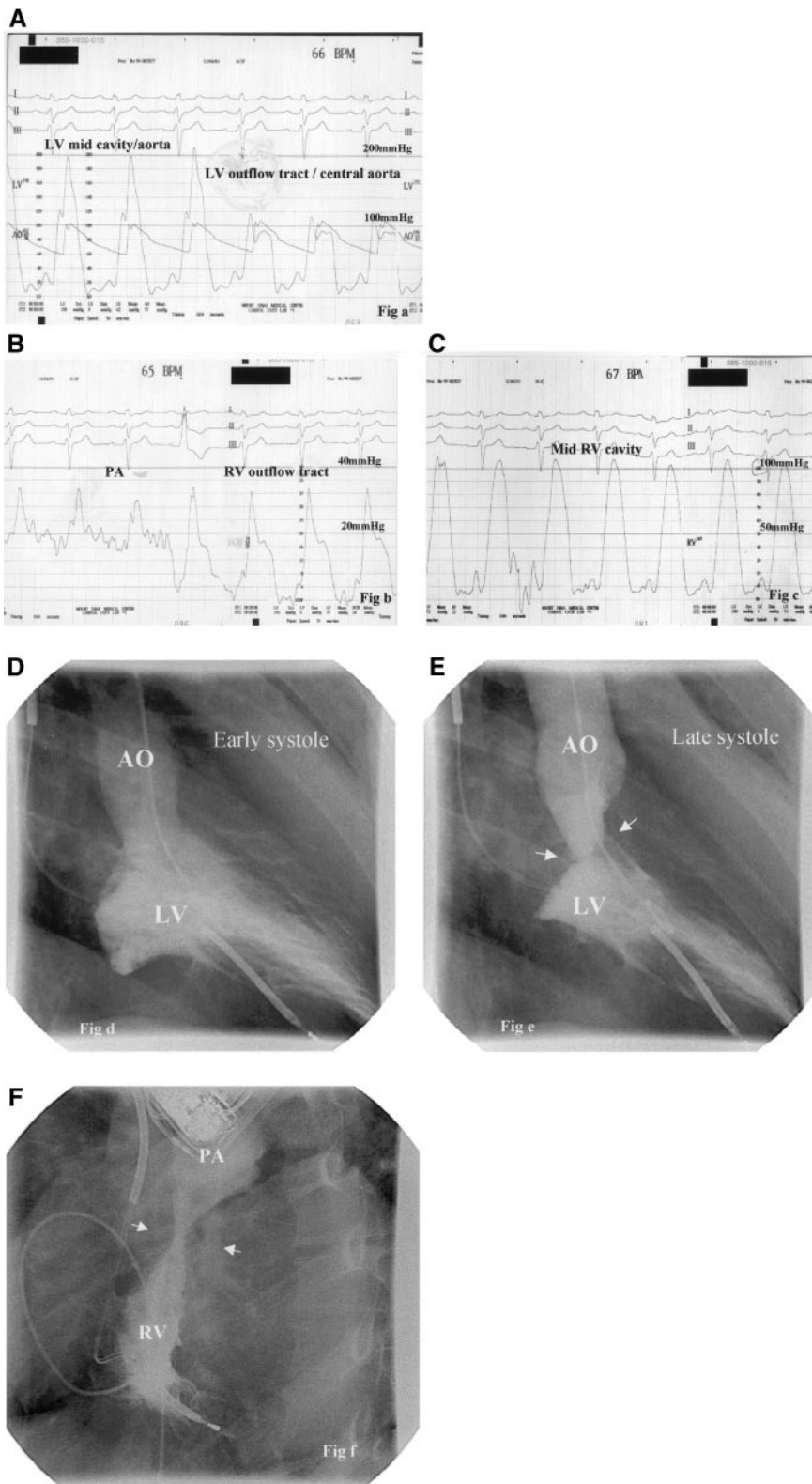
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a, Simultaneous pressure recordings from the mid LV cavity and central aorta show a 100 mm Hg gradient. The aortic trace shows a “spike and dome” pattern due to systolic anterior mitral valve movement, which impedes late systolic ejection. Pullback from the central LV cavity confirms that the gradient is sub-aortic. b and c, Pullback from the pulmonary artery (PA) reveals a 75 mm Hg subpulmonic gradient. d and e, Left ventriculogram (right anterior oblique projection). Arrows show systolic anterior mitral valve movement obstructing outflow in late systole. AO indicates aorta. f, Right ventriculogram (left lateral projection) demonstrates massive hypertrophy of the RV outflow (arrows) with an “hour-glass” deformity.