Clinically silent, right and left ventricular outflow tract obstructions in an adult patient (RCD code: IV-3A)

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Abstract

Right ventricular outflow tract obstruction is a narrowing of the way that conducts blood from the right ventricle to the pulmonary artery. Left ventricular outflow tract obstruction is a stenosis of the path that leads blood from the left ventricle to the aorta. Combination of right and left ventricular outflow tract obstruction is a very rare finding. We report a case of a 67-year-old male with asymptomatic biventricular outflow tract obstruction. We discuss the clinical presentation, diagnostic procedures, treatment opportunities for the patient based on review of literature and actual recommendations. JRCD 2017; 3 (2): 50–53

Key words: right ventricular outflow tract obstruction, left ventricular outflow tract obstruction, congenital heart disease, hypertrophic cardiomyopathy, rare disease

Background

Congenital heart defects (CHD) are often diagnosed in infancy, or even before birth, although there exists a population of adults whose CHD was not diagnosed during childhood.

We present the case of patient with biventricular outflow tract obstruction diagnosed at age ≥60 years. The complementary use of imaging techniques allowed to evaluate the anatomical, functional and structural characteristic of the heart and vessels.

Case presentation

A 67-year-old Caucasian man, with hypertension, working as a farmer was admitted to the Department of Cardiac and Vascular Diseases in October 2016, with suspicion of heart defects for cardiologic evaluation. Patient denied chest pain, exertional dyspnea and had good tolerance of physical effort. His everyday activity wasn't limited, he was able to work, walk up the stairs without restrictions. At the day of admission he was hemodynamically stable, with no ailments. His heart rate was 70 beats per minute, blood pressure was 160/80 mm Hg. On physical examination we found systolic ejection murmur grade 3/6 in Levine's scale over the left sternal border and apex.

Initial electrocardiogram showed right axis deviation, sinus rhythm 82/minute, P-cardiale, pathological Q waves in II, III, aVF leads and signs of right ventricular hypertrophy (Sokolow-Lyon Index = 14 mm). His oxygen saturation at rest was 96% on room air. Laboratory tests results were normal.

Echocardiographic examination revealed (Figure 1–3):
– preserved left ventricular ejection fraction
– concentric left ventricular hypertrophy (diastolic ventricular septum thickness was 13 mm, posterior wall thickness 12 mm, left ventricular mass index 125 g/m², relative wall thickness 0.46)
– right ventricular hypertrophy (thickness at infundibular level – 19 mm)
– severe infundibular right ventricular outflow tract obstruction with gradient of 116/68 mm Hg (max/mean) caused by hypertrophied muscle
– mild subvalvular left ventricular outflow tract obstruction with gradient of 25/12 mm Hg (max/mean) caused by subaortic fibrous ridge
– ascending aorta dilation
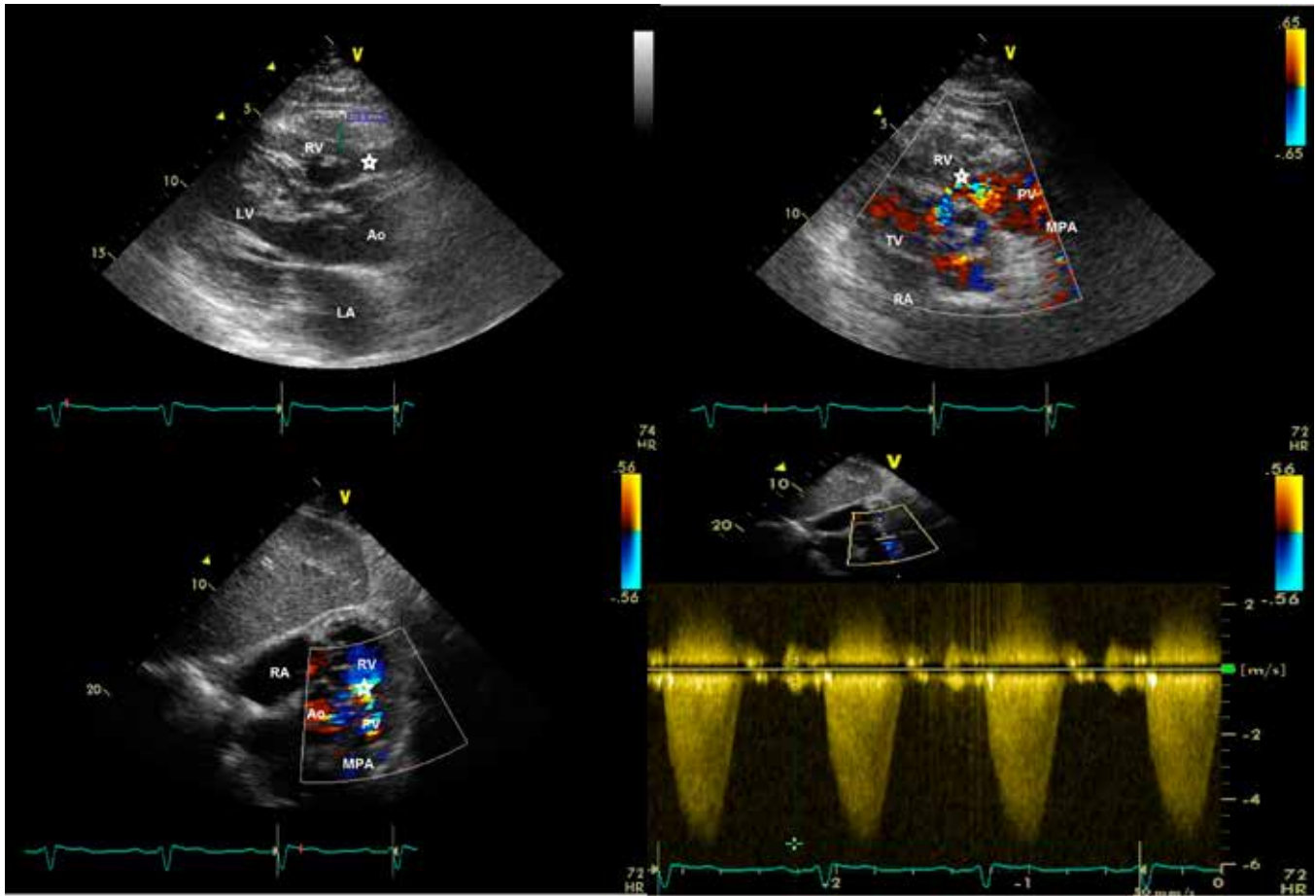– systolic jet suggesting membranous ventricular septal defect.

Figure 1. Transthoracic echocardiography, systolic phase – left and right ventricular hypertrophy, severe right ventricular outflow tract obstruction (RVOTO-white star). Parasternal long axis view (upper-left), parasternal short axis view (upper-right) substernal short axis view (lower-right), substernal short axis view with RVOTO gradient measurement (lower-left). Ao – aortic valve, LA – left atrium, LV – left ventricle, RA – right atrium, RV – right ventricle, MPA – main pulmonary artery, PV – pulmonary valve, TV – tricuspid valve

Computed tomography revealed (Figure 2):
– infundibular right ventricular outflow tract obstruction (9 × 14 mm, diastole)
– enlarged pulmonary valve annulus (30 × 30 mm), pulmonary trunk (32 × 30 mm), right (30 × 28 mm) and left (35 × 32 mm) pulmonary artery
– ventricular septum hypertrophy (13–14 mm)
– mild left ventricular outflow tract obstruction (18 × 27 mm)
– aneurysm of the ascending aorta (49 × 48 mm)
– common inflow of superior and inferior left pulmonary veins to the left atrium (an anatomical variant)
– ventricular septal defect was excluded.

We did not perform exercise testing due to patient's unwillingness for further evaluation, therefore, we are unable to assess definitively if our a patient was truly asymptomatic.

Discussion

A combination of right and left ventricular outflow tract obstruction, as it occurred in our patient, is an unusual finding. A narrowing of the outflow tract is clinically detected at different stages of life. The more severe the obstruction, the earlier it is typically detected [1].

Right ventricular outflow tract obstruction (RVOTO) may occur at different levels with resultant variations in clinical presentation. It can be divided into subinfundibular, infunfibular, valvular and supravalvular obstruction [1,2]. Subinfundibular and infundibular form usually are accompanied with other lesions. Subinfundibular RVOTO tends to be combined with ventricular septal defect (VSD), while infundibular stenosis is often associated with tetralogy of Fallot (ToF) and VSD or it may result from reactive myocardial hypertrophy in valvular pulmonary stenosis [3,4]. We did not observe this abnormalities in our patient. There has been also reports of coexistence of infundibular RVOTO with VSD, dilation of the aorta and pulmonary artery, valvular insufficiency in Marfan syndrome [5,6]. Pulmonary valve stenosis constitutes 80 to 90% of all RVOTO cases. Typically it is a congenital, isolated defect. As a part of Noonan syndrome it coexists with other CHD including atrial septal defect, VSD, branch pulmonary stenosis and ToF or hypertrophic cardiomyopathy [7,8].

Supravalvular form is caused by stenosis of pulmonary trunk or peripheral pulmonary arteries. This CHD occurs seldom in isolation, rather it is found in combination with ToF, Noonan syndrome,
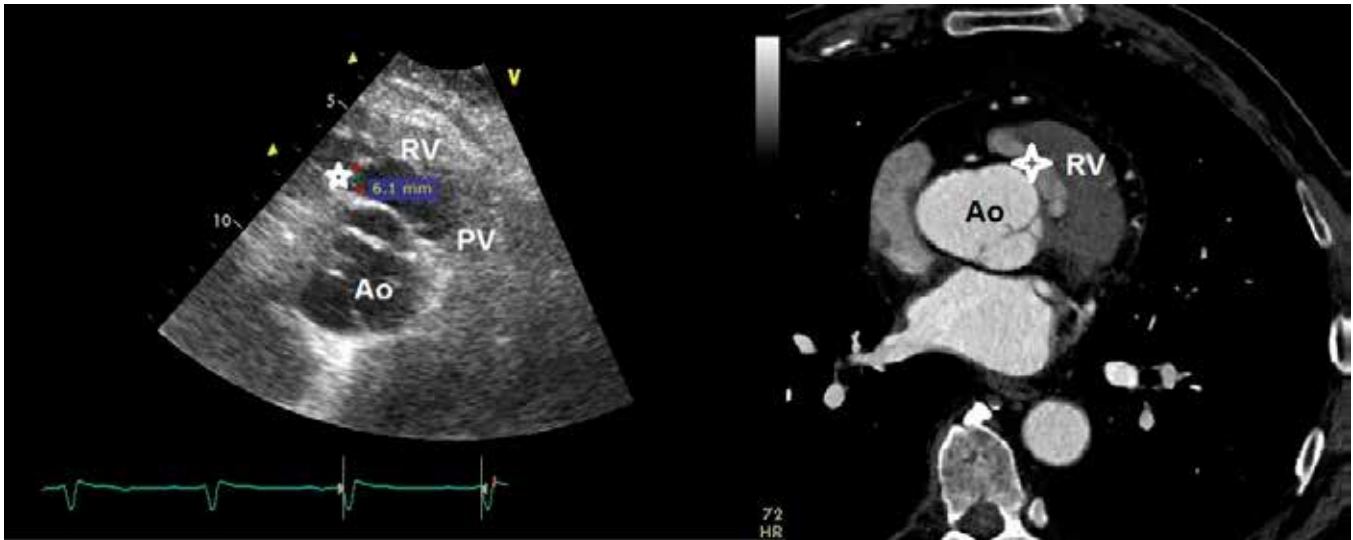
Figure 2. Right ventricular hypertrophy and right ventricular outflow tract obstruction (white star), assessed by echocardiography (left; systolic phase) and computed tomography (right; diastolic phase). Ao – aortic valve, RV – right ventricle, PV – pulmonary valve

Figure 3. Transthoracic echocardiography – apical four-chamber view, systolic phase. Mild subvalvular left ventricle outflow tract obstruction caused by subaortic fibrous ridge (white arrow). LA – left atrium, LV – left ventricle, RA – right atrium, RV – right ventricle

Williams-Beuren syndrome, Keutel syndrome, Alagille syndrome or congenital rubella syndrome. It can complicate previous surgical treatment of heart defects [9].

Left ventricular outflow tract obstruction may affect subvalvular, valvular, supravalvular level [1,2]. Subvalvular stenosis may result from hypertrophic cardiomyopathy, congenital subaortic fibromuscular membrane/fibrous ridge or previous cardiac surgery. It may coexist with VSD, atrioventricular septal defect and Shone syndrome [10]. Valvular stenosis is the most frequent (>75%) type of LVOTO [11,12]. Supravalvular form is caused by obstructive arteriopathy. It is usually associated with other congenital defects including hypoplastic aorta or Williams-Beuren syndrome. Coexistence of right and left ventricular tract obstruction seems to appear in hypertrophic cardiomyopathy (HCM). Previous reports involved small groups of patients or single cases [8,13]. In the right ventricle, obstruction is caused by hypertrophied muscle, whereas LVOTO is due to mitral valve systolic anterior motion and hypertrophy of

the interventricular septum [14]. Diagnosis of biventricular HCM in our patient is doubtful, considering only mild hypertrophy of the left ventricle and a mechanism of LVOTO.

Patients with ventricular outflow tract obstruction may be clinically silent. The obstruction tends to be progressive over time and patients become symptomatic. Symptoms include dyspnea, dizziness, angina or syncope. Complications include sudden deaths, heart failure, aortic dissection, endocarditis and ascending aorta aneurysm [1,2].

Clinical examination in patients with RVOTO reveals a harsh systolic murmur and a wide splitting of the second heart sound. In patients with valvular LVOTO a systolic murmur radiates to the carotids, whereas in those with subvalvuar form, it is heard only at the left sternal border and apex. Echocardiography is an invaluable tool in the diagnosis, prognosis, management strategy and follow-up of patients with suspected ventricular outflow tract obstruction [15,16]. Computed tomography or cardiovascular magnetic reso-

nance imaging are performed to confirm the diagnosis and deliver information about anatomy of other lesions. To reveal the level and the severity of the obstruction cardiac catherization may be considered [17,18]. Coronary angiography is required before qualification to surgical intervention when the patient is a male over 40 years of age, postmenopausal female or has the signs or risk factors of coronary artery disease [1,2,11].

In patients with severe infundibular RVOTO (peak gradient >64 mm Hg) or severe subaortic LVOTO (mean gradient ≥50 mm Hg) and in patients with concomitant other lesions surgical treatment is recommended [2].

Patient management and follow-up.

We planned to proceed cardiac catherization to confirm the severity of RVOTO and qualify the patient to surgical treatment but patient denied further diagnostic and therapeutic procedures. He received comprehensive information about the risk of potential consequences of his decision and was discharged on his own demand. Antihypertensive treatment was instituted as follows: ramipril 10mg/d, indapamide 1.5mg/d. The patient's condition was stable in a 6-month follow up.

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