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Severe aortic regurgitation and pulmonary hypertension in an 18-year-old patient after balloon aortic valvuloplasty (RCD code: IV-5.A2)

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Abstract

Aortic stenosis is the most common form of congenital left ventricle outflow tract obstruction. It may be a life threatening ductal-dependent condition in newborns and necessitating urgent percutaneous balloon aortic valvuloplasty (BAV). Although early results of valvuloplasty are usually satisfactory, long-term follow-up show frequent incidence of gradually progressing aortic insufficiency that requires valve replacement surgery. The case report presented below concerns a young patient with a history of BAV in childhood who developed severe aortic regurgitation accompanied by combined post- and pre-capillary pulmonary hypertension. JRCD 2016; 2 (6): 192–195

Key words: aortic valve replacement, bioprosthesis, echocardiography, heart catheterization, iliac artery occlusion, congestive heart failure, sildenafil

Case presentation

The report concerns an 18-year-old patient with severe aortic regurgitation (AR) and combined post-capillary and pre-capillary pulmonary hypertension (CpcPH). In infancy he was diagnosed with severe aortic stenosis (AS) and underwent urgent percutaneous balloon valvuloplasty (BAV) (1997). Since then he had remained under care of regional outpatient clinics due to obesity, allergic rhinitis and arterial hypertension. In 2011 he began to experience decrease in physical capacity. In June 2012 after developing atypical pneumonia he was admitted to Cardiology Department of University Children's Hospital in Krakow with signs of congestive heart failure. Transthoracic echocardiography (TTE) demonstrated cardiac enlargement, severe AR, significant tricuspid regurgitation (TR), estimated right ventricle systolic pressure (RVSP) of 88 mm Hg and preserved left ventricular ejection fraction (LVEF). Right heart catheterization (RHC) confirmed severe pulmonary hypertension (PH) in the course of left heart disease. Mean pulmonary artery pressure (mPAP) was 46 mm Hg, pulmonary wedge pressure (PWP) was 27 mm Hg, intracardiac shunt was ruled out. Angiography detected chronic occlusion of the right external iliac artery and common femoral artery with well-developed collateral circulation. Renal angiograms did not show important abnormalities. Magnetic resonance imaging revealed nonischemic myocardial damage of both ventricles in the form of subendocardial fibrosis. High-resolution computed tomography (HRCT) of the lungs disclosed diffused ground-glass opacities with air traps (prominent in the lower pulmonary lobes), bronchiectases and consolidations with tree-in-bud appearance in the right lower lobe. Spirometry results were within normal references. Pulmonology consultant pointed to PH as the most probable cause of HRCT findings. Due to high operative risk associated with severe PH he was disqualified from surgical aortic valve replacement (AVR). He was given sildenafil at the dose 30mg daily as a trial and was scheduled for reevaluation after 6 months. Cardiac catheterization performed in January 2013 showed no relevant hemodynamic improvement and the drug was discontinued. Additional hemodynamic assessment confirmed severe AR. No stenotic lesions were found on coronary angiograms.

In July 2014 upon reaching his adolescence the patient was referred to our Department to continue follow-up. He complained of dyspnea on exertion in class II/III by New York Heart Association (NYHA) and recurrent lower limbs oedema. Physical examination revealed obesity, heart rate of 100 bpm, blood

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Table 1. Comparison of transthoracic echocardiography results

Parameter	JUNE 2012	JULY 2014
LVEDd/ LVESd [mm]	57/39	58/37
IVSd/IVSs [mm]	10/14,5	12/18
PWd/PWs [mm]	10/16	10/15
LVEF [%]	58	55
Ascending aorta [mm]	30	41
AR	significant	significant
LA area in 4AC view [cm^2]		26
MV E/A ratio		2,5
EDMV by TDI [m/s]		0,05
MPA [mm]		32
PV- AccT [ms]	80	72
RVOT in PLAX [mm]	30	36
RV in A4C [mm]		58
RV-FW s/d [mm]		8/12
RA area in 4AC [cm^2]		29
TV annulus [mm]	36	57
TR	significant	significant
RVSP [mm Hg]	88	120
TAPSE [mm]	17	17
TASV by TDI [m/s]		0,18
IVC e/i [mm]		30/24

LVEDd/LVESd – left ventricle end-diastolic/end-systolic diameter, IVSd/IVSs – interventricular septum diastolic/systolic thickness, PWd/s –left ventricular posterior wall systolic/diastolic thickness, LVEF – left ventricular ejection fraction, LA – left atrium, RA- right atrium, AR – aortic regurgitation, PHT – pressure half-time, LVOTd- left ventricular outflow tract diameter, MV – mitral valve, EDMV- early diastolic mitral annulus velocity, MPA – main pulmonary artery, PV-Acct – pulmonary acceleration time, RVOT – right ventricular outflow tract, RV – right ventricle, RV-FW s/d – systolic/diastolic right ventricular wall thickness, TV – tricuspid valve, TR – tricuspid regurgitation, RVSP – right ventricular systolic pressure, TAPSE – tricuspid annular plane systolic excursion, TASV- tricuspid annular systolic velocity, IVC e/i- inferior vena cava (inspiration/expiration diameter), 4AC- apical four chamver view, PLAX – parasternal long-axis view, TDI – tissue Doppler imaging

Table 2. Abnormal laboratory findings

		Reference range
NT-proBNP [pg/mL]	799	< 125,0
AST [U/L]	46	10 – 40
ALT [U/L]	51	10 – 40
GGT [U/L]	105	< 60
TG [mmol/L]	2,75	≤ 1,70
TSH [uIU/mL]	5,08	0,3 – 4,0
fT3 [pmol/L]	6,18	3,10 – 9,20
fT4 [pmol/L]	20,33	10,3 – 20,3

ALT- alanine aminotransferase, AST- aspartate transaminase, GGT- gamma-glutamyl transferase, TG- triglierydes, TSH- thyroid-stimulating hormone, fT4- thyroxine, fT3-triiodothyronine, NT-proBNP-N-terminal pro b-type natriuretic peptide

pressure of 180/90 mm Hg. Electrocardiogram (ECG) demonstrated sinus rhythm, right ventricular hypertrophy and biatrial enlargement. Laboratory abnormalities included hypertriglyceridemia, elevated natriuretic peptide and liver enzymes. Thyroid panel was typical for subclinical hypothyroidism (Table 2). Transthoracic echocardiography (TTE) disclosed enlarged heart chambers, hypertrophied ventricles, preserved LVEF and LV diastolic dysfunction, severe AR and TR with RVSP of 120 mm Hg (Table 1, Figure 1 and 2). Aortic valve was trileaflet with rudimentary left coronary cusp (Figure 3). Abdominal ultrasound showed enlarged liver with increased echogenicity and moderately dilated hepatic veins, no renal arterial pathology. 24-hour Holter ECG monitoring registered two short episodes of non-sustained ventricular tachycardia. Cardiopulmonary exercise test was terminated due to dyspnea. Peak oxygen uptake was 2.9 ml/kg/min. Distance in 6-minute walking test was 480 meters. Results of pulmonary functional tests were within normal renges. Heart catheterization detected significant postcapillary PH, decreased cardiac output and high LV filling pressure (Table 3). Hemodynamic assessment also reported elevated diastolic pressure gradient (DPG) of 29 mm Hg and transpulmonary pressure gradient (TPG) of 49 mm Hg denoting a precapillary component of PH. Taking into consideration previous unsuccessful sildenafil therapy it was decided to address the worsening AR in the first place. The patient was consulted by a multidisciplinary group of experts during several local Center for Rare Cardiovascular Diseases (CRCD) sessions and was eventually qualified for AVR. He underwent surgical implantation of Carpentier-Edwards Perimount 23A bovine pericardial valve in February 2015. The procedure and recovery were uneventful. Post-operative TTE showed proper seating and function of the bioprosthetic valve, mild TR and twofold reduction of RVSP (60 mm Hg). Ten months after surgery the patient was in functional class I by NYHA without diuretics. Echocardiogram demonstrated mild TR with RVSP of 30 mm Hg. He currently remains under surveillance of outpatient cardiothoracic clinic.

Discussion

Congenital left ventricle outflow tract obstruction (LVOTO) is most often localized at the level of the aortic valve. It comprises 8% of all congenital heart defects. Bicuspid aortic valve is responsible for 60–80% of congenital AS, with 1–2% incidence in general population [1]. Approximately 10% of newborns with congenital AS require aortic valvuloplasty due to severe LVOTO. In this

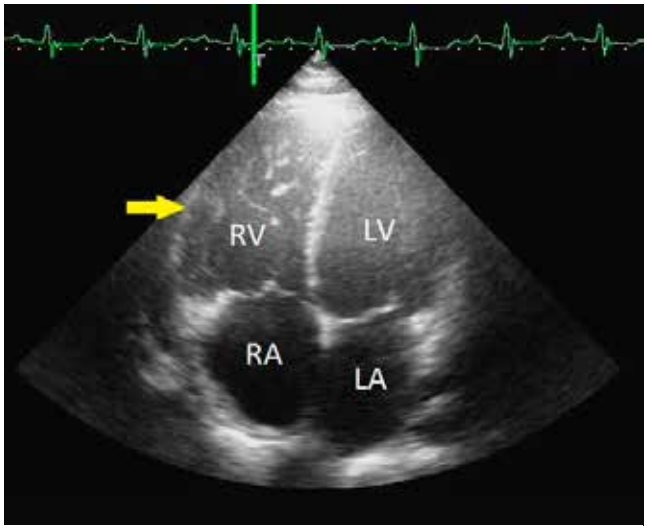

Figure 1. Transthoracic echocardiography. Apical four chamber view. Enalargement of all heart chambers. Ventricular hypertrophy, especially of the right ventricle wall (arrow). RA – right atrium, RV-right ventricle, LA – left atrium, LV – left ventricle

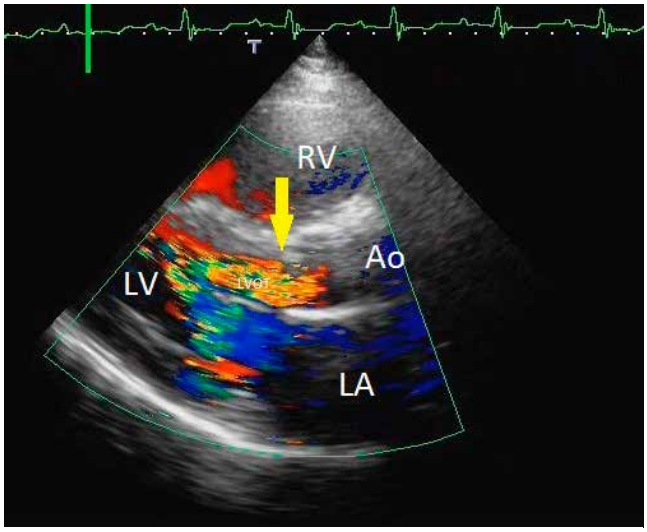

Figure 2. Transthoracic echocardiography. Parasternal long-axis view. Aortic regurgitation – the jet's width equals LVOT diameter (arrow). RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle, LVOT – left ventricular outflow tract

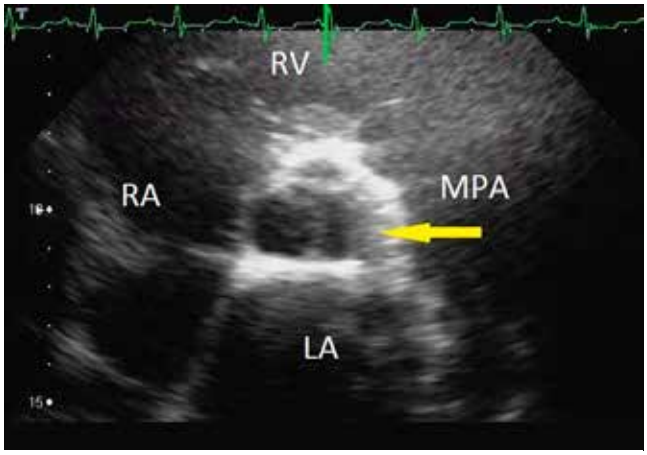

Figure 3. Transthoracic echocardiography. Parasternal short-axis view. Diastolic image of the aortic valve – the left coronary cusp is rudimentary (arrow). RA – right atrium, RV – right ventricle, LA – left atrium, MPA – main pulmonary artery

group of patients it is usually a palliative strategy and the outcome depends on morphology of the valve and severity of LV dysfunction. Early results of these procedures are mostly satisfactory and severe post-operative AR is infrequent and rarely requires surgery (e.g. Ross procedure). However, in long-term follow-up the frequency of AR tends to increase. Approximately 30% of these patients will develop significant AR within 5–14 years [2]. In adult population BAV is generally reserved for patients with advanced and hemodynamically unstable AS as palliative therapy or bridge to non-cardiac surgery [3].

AR may be caused by valvular pathology (e.g. destruction of cusps in the course of infective endocarditis) or changed geometry of the aortic bulb. If left untreated, severe regurgitation is of unfavorable prognosis. In symptomatic patients early death rate may reach up to 20%. On the other hand, asymptomatic patients with chronic severe AR without impaired LV function have low probability of disadvantageous events. In case of LV end-diastolic diameter > 50 mm, the estimated probability of death, occurence of symptoms or LV dysfunction is around 19% per year. At present, AVR is performed most often, however in specialized clinical centers the number of repair procedures continues to increase. According to the current European Society of Cardiology (ESC) guidelines indications for surgical management of chronic, severe AR are: development of symptoms, resting LVEF ≤50%, simultaneous need for coronary artery bypass grafting or surgery of the ascending aorta (Class I, Level B), and severe LV enlargement (Class IIa, Level C) [4]. Distinct criteria were established for patients with Marfan syndrome [4].

PH is a common complication of left heart diseases (LHD) including valvular disorders, and results in more severe symptoms and worse exercise tolerance. The primary goal of therapy in PH-LHD is to improve management of the underlying condition before considering specific PH treatment, e.g. repair of valvular heart disease. In this group of patients the 2015 ESC guidelines recommend the use of TPG, DPG and pulmonary vascular resistance to differentiate the type of this condition, i.e. isolated post-capillary pulmonary hypertension and CpcPH [5]. The changed compliance of pulmonary arteries may be responsible for the latter [6]. Patients with PH-LHD and identified pre-capillary component should be referred to an expert PH center for a complete diagnostic workup and individualized treatment [7]. At present there is little evidence supporting the use of therapies approved for pulmonary arterial hypertension in CpcPH patients [5].

In young patients undergoing AVR the decision concerning optimal timing for surgery and its type is complex. Considerable probability of reoperation, lifelong anticoagulation in case of mechanical prostheses, frequent non-compliance associated with lifestyle need to be taken into account [7]. Many studies have proven that

Table 3. Results of cardiac catheterization performed in July 2014

PRESSURE [mm Hg]	REST	AFTER NO INHALATION
RA		18/10/10
RV	121/5/14	116/2/12
PA	119/56/76	110/53/73
PCWP	33/22/27	35/29/32
LV		140/2/25
AORTA	144/87/106	124/79/98
OXYGEN SATURATION [%]		
IVC		63,8
SVC		58,5
RA		57,5
RV	59,6	56,4
PA	62,9	64,3
AORTA	95	87,7
Cardiac Output [l/min]	4,08	6,19
Cardiac Index [l/min/m^2]	1,87	2,84
Qp/Qs	1,06	1,06
PVR [ARU]	995,4 (12 WU)	529,9 (6,6 WU)
TPR [ARU]	1400,9 (17,5 WU)	943,5 (11,8 WU)
SVR [ARU]	2019,6 (25,2 WU)	1137,3 (14,2 WU)

NO- nitric oxide, RA- right atrium, RV-right ventricle, PA-pulmonary artery, PCWP-pulmonary capillary wedge pressure, LV-left ventricle, VCI-vena cava inferior, VCS-vena cava superior, Qp/Qs- pulmonary to systemic flow ratio, PVR- pulmonary vascular resistance, TPR-total pulmonary resistance, SVR- systemic vascular resistance, ARU – absolute resistance unit, WU – Wood unit

in the young, bioprostheses are prone to accelerated degeneration [8]. Follow-up of young patients with Carpentier-Edwards pericardial bovine valve implants show their long-term durability with low rates of structural failure [9]. It should be emphasized that rigorous post-operative surveillance is critical for early detection of possible complications and their prompt management.

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