History and management of congenital aortic stenosis and coarctation of the aorta in a 38-year-old patient

Historia i leczenie wrodzonego zwężenia aorty i koarktacji aorty u 38-letniej pacjentki

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Medical Studies/Studia Medyczne 2019; 35 (4): 324–326
DOI: https://doi.org/10.5114/ms.2019.91251

Key words: congenital aortic stenosis, coarctation of aorta, balloon valvuloplasty, hybrid therapy.

Słowa kluczowe: wrodzone zwężenie aorty, koarktacja aorty, walwuloplastyka balonowa, terapia hybrydowa.

Abstract

We present the case of a 38-year-old woman with congenital aortic stenosis as well as coarctation of the aorta that was unrecognised in her childhood. The patient was treated by balloon valvuloplasty. Ten years after the procedure, clinical deterioration was observed with subsequent progression of valvular gradient, aortic regurgitation, and diagnosis of coarctation. At the age of 38 years, a routine echocardiographic test showed massive calcifications of aortic leaflets with a transvalvular gradient 77/41 mm Hg, and third-degree insufficiency with widening of the ascending aorta (42 mm) and normal left ventricle systolic function. Additionally, for the first time the test showed coarctation of the aorta – narrowing in the isthmus below the arch down to 15 mm, with a maximum flow gradient ca. 60 mm Hg. The management of the complex heart defects is currently being considered.

Streszczenie

Przedstawiamy przypadek 38-letniej kobiety z wrodzonym zwężeniem aorty oraz koarktacją aorty, której nie rozpoznano w dzieciństwie. Pacjentka była leczona metodą walwuloplastyki balonowej. Dziesięć lat po zabiegu zaobserwowano pogorszenie stanu klinicznego z późniejszym postępem gradientu zastawkowego, niedomykalnością aorty i rozpoznaniem koarktacji. W wieku 38 lat w rutynowym badaniu echokardiograficznym wykazano masywne zwapnienia płatków aorty z gradientem przezzastawkowym 77/41 mm Hg, niedomykalność trzeciego stopnia, poszerzenie aorty wstępującej (42 mm) z prawidłową funkcją skurczową lewej komory. Dodatkowo w badaniu po raz pierwszy stwierdzono koarktację aorty – zwężenie wymiarów aorty w cieśni poniżej łuku do 15 mm z maksymalnym gradientem przepływu około 60 mm Hg. Obecnie rozważane jest leczenie złożonej wady serca.

Introduction

Valvular heart diseases make up a large percentage of congenital cardiological diseases. As a result of the progress of early diagnostics and medical treatment, we are now witnessing longer lives in children after surgeries of congenital heart defects. At the same time, after years of observations, additional irregularities are being discovered, related to post-surgery re-progression of the disease or to revealing new, not previously diagnosed anomalies.

Case report

In a girl born naturally in the 38th week of pregnancy a mild tricuspid aortic valve stenosis was diagnosed in the second month of life. The heart defect was asymptomatic for 15 years. At the age of 15, due to a documented serious increase in a transvalvular gradient up to ca. 100/50 mm Hg accompanied by mild aortic insufficiency, a balloon valvuloplasty was performed, which decreased the maximum gradient down to ca. 65 mm Hg, and increased the aortic insufficiency up to a moderate degree. During the cardiac catheterisation, a decrease in the gradient of left ventricle – aorta was confirmed. In the following years no essential progression of the heart disease was observed, the transvalvular gradient amounted to ca. 60/30 mm Hg with moderate aortic insufficiency and a normal left ventricular contraction. In spite of stable echocardiographic parameters, 10 years after the bal-

loon valvuloplasty, intensified tiredness, breathlessness, palpitation, dizziness, faintness, and high blood pressure (160/80 mm Hg) appeared. A relative clinical stability was achieved as a result of applied β-blockers. At the age of 38 years, a routine echocardiographic examination showed massive calcifications of aortic leaflets with a transvalvular gradient 77/41 mm Hg, and third-degree insufficiency (Figure 1) with widening of the ascending aorta (42 mm) and normal left ventricle systolic function. Additionally, for the first time the test showed coarctation of the aorta – narrowing in the isthmus below the arch down to 15 mm, with a maximum flow gradient ca. 60 mm Hg (Figure 2). A physical examination of the patient showed a loud vascular murmur between scapulas, and a difference in pressure between upper and lower limbs of about 30 mm Hg. The presence of the coarctation was confirmed by angio-tomography of the chest. The patient currently shows continuing moderate clinical symptoms that impair everyday functioning (the patient works and studies).

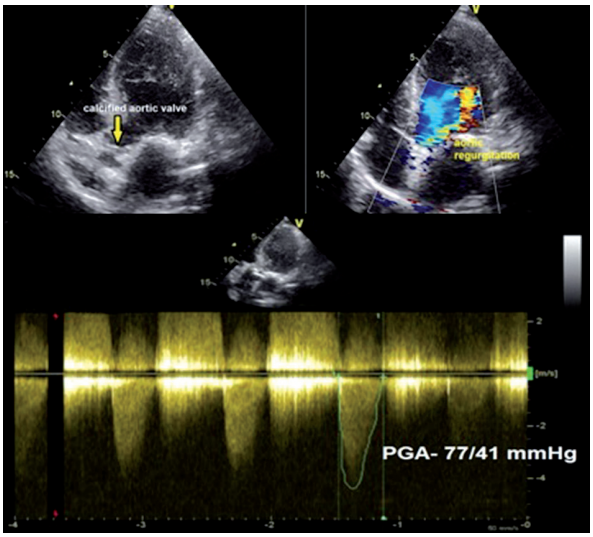

Figure 1. Transthoracic echocardiography, four chamber view. Aortic valve stenosis and regurgitation

Discussion

Congenital aortic stenosis constitutes 2–4% of all congenital heart defects [1]. In newborns with a small aortic stenosis, no clinical symptoms are observed, the defect is discovered on the basis of auscultation. The natural development of the disease is also characterised by its long symptomless course [1]. In the studied clinical case, the patient underwent an interventional therapy due to the progression of stenosis after 15 years of observation. The basic method of treatment of congenital aortic stenosis is balloon valvuloplasty. According to long-term observations, this procedure is very effective and essentially delays performing a necessary cardiosurgery. The observation of the patients after the balloon valvuloplasty showed 10-year survival without the necessity of a surgery in 67–83% of patients [2, 3]. In our patient, 10 years after the valvuloplasty, some clinical complaints appeared; however, a substantial increase in echocardiographic parameters was documented 16 years after the treatment. One of the complications of the valvuloplasty is the aggravation of aortic regurgitation. According to presented observations, a moderate and severe aortic insufficiency was found in 19–25% of patients after the procedure [2] and in 34% 4.8 years after the treatment on average and 51% after 10 years [3]. In the described case, the clinical deterioration was probably caused by the intensifying aortic insufficiency, which confirms the existence of betterment as a result of applied pharmacotherapy.

Coarctation of the aorta accounts for 5–8% of all congenital heart defects [4] and is defined as a localised narrowing of the aortic lumen by a ridge, composed of medial wall thickening and infolding of aortic wall tissue [5]. Coarctation of the aorta may occur

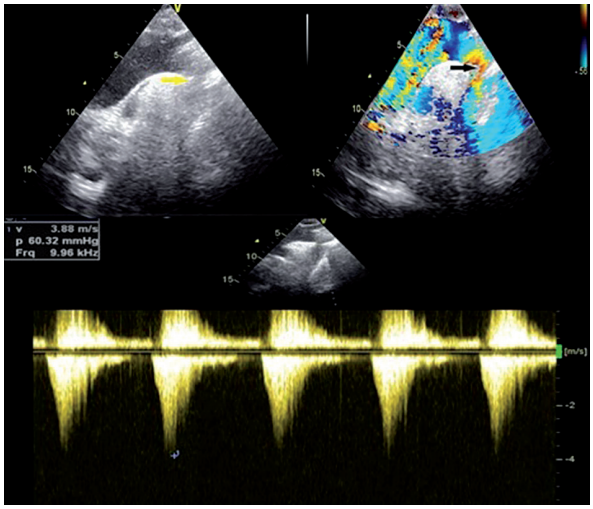

Figure 2. Transthoracic echocardiography, suprasternal view. Coarctation of aortae

as an isolated defect or co-exist with other anomalies: bicuspid aortic valve, hypoplasia of the aortic arch, subvalvular stenosis, mitral valve defects, atrial and ventricular septal defects, and persistent Botall's duct [6]. Co-existence of congenital aortic stenosis and coarctation of the aorta is rare and occurs only in ca. 5–10% of cases of left ventricular outflow tract obstruction [7]. Because clinical symptoms of both anomalies appear late, an additional increase in the transvalvular gradient makes it difficult to evaluate the function of flow in the coarctation; therefore, the diagnostics of this anomaly, with nearly 40-year-old limited diagnostic capabilities, was not obvious. Treating patients with coarctation of the aorta essentially differs depending on age. In newborns, infants, and

small children, a surgical resection of the coarctation is selected by means of various surgical techniques [8]. In older children (with body weight over 25 kg) and in adults coarctation stenting is the preferred form of therapy. This treatment is less invasive and bears little risk of restenosis and post-surgical arterial dilatation. In younger children balloon angioplasty is applied because the implantation of stent in a child with a growing aorta in the developmental period involves the problem of selection of the appropriate stent size, frequent necessity of re-dilatation, high risk of neointimal hyperplasia with restenosis, and development of aneurysm [8]. Therapeutic procedures in patients with double aortic anomaly is more complex. In current reports we can come across a hybrid therapy, which is a combination of a cardiosurgical excision of the site of coarctation with end-to-end anastomosis through lateral thoracotomy and a simultaneous balloon valvuloplasty of the aortic valve via the carotid artery [9]. Obviously, such a procedure is recommended in patients without advanced cusp calcification and substantial aortic insufficiency. In the described case, replacement of the aortic valve is necessary as well as the selection of an appropriate coarctation repair strategy. In the literature we can come across single reports of one-stage aortic valve replacement and transpericardial ascending aorta-descending aorta bypass [10]. In our patient, the replacement of the aortic valve with the surgical repair of coarctation or replacement of the valve combined with the stent therapy for aortic coarctation is considered.

Conflict of interest

The authors declare no conflict of interest.

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