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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Two-stage surgery for a large ventricular septal defect and patent ductus arteriosus associated with severe pulmonary arterial hypertension in an adult patient

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SUMMARY

Introduction Adult patients with pulmonary arterial hypertension (PAH) associated with congenital left-to-right shunting are often considered inoperable.

Case outline A 26-year-old man presented with effort intolerance and palpitations. The diagnosis of PAH in the presence of a large perimembranous ventricular septal defect (VSD) and patent ductus arteriosus (PDA) was established. The patient was managed with a two-stage surgical approach involving an initial ligation of PDA, followed by VSD closure using unidirectional valved patch. Treatment decisions were based on the results of both invasive and non-invasive investigations. At follow-up, the patient was asymptomatic with pulmonary arterial pressure and vascular resistance returning to normal levels.

Conclusion There is a possibility for an adult patient to have a congenital heart disease associated with marked pulmonary overcirculation that is still amenable to surgical repair. This implies that there is an individual response to a long-standing left-to-right shunt, and that the therapy should be considered on a case-by-case basis.

Keywords: congenital heart defects; pulmonary hypertension; cardiac surgical procedures; adults

INTRODUCTION

Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) comprises a heterogeneous group of disorders characterized by the presence of pulmonary arterial hypertension in the setting of repaired or unrepaired congenital systemic to pulmonary shunts [1, 2, 3]. PAH is fairly common in adult patients with CHD, and is associated with significant morbidity and mortality. The evidence regarding the management of PAH and CHD is scarce. The severity of PAH, the magnitude of the shunt, and the degree of vasoreactivity of the pulmonary circulation are key considerations in determining whether the condition is amenable to surgical treatment [4, 5, 6].

CASE REPORT

A 26-year-old man, 183 cm tall and weighing 65 kg, presented with effort intolerance and heart palpitations. His symptoms were consistent with NYHA functional class II. The patient was known for having a ventricular septal defect (VSD) and patent ductus arteriosus (PDA) diagnosed at birth. However, his parents did not consent to proposed surgical treatment at that time.

During a physical examination, there was an accentuated pulmonary component of the second heart sound, systolic murmur grade 2 over the precordium, and transcutaneous oxygen saturation of 96% on room air.

Chest X-ray showed significant cardiomegaly (cardiothoracic ratio of 0.62) with increased pulmonary vascular markings and clear lung fields.

Transthoracic echocardiogram revealed a large perimembranous-outlet VSD measuring 25 mm in diameter with no pressure gradient (PG) across it, and a widely opened PDA without transductal gradient. Left heart was dilated with a mildly impaired left ventricular systolic function (Table 1). There was an estimated pressure gradient of 78 mmHg across the tricuspid valve.

ECG showed extreme axis deviation, signs of biventricular hypertrophy, right bundle branch block, and biphasic T waves in the left-sided leads. A 24-hour Holter ECG monitoring revealed isolated ventricular extrasystoles.

Cardiac catheterization was performed to obtain hemodynamic data. Mean pulmonary arterial pressure (mPAP) was measured at 59 mmHg and there was a significant left-to-right shunt with a pulmonary to systemic blood flow ratio (Qp/Qs) of 2.1, pulmonary vascular resistance index (PVRI) of 6.8 WU/m^2, and pulmo-

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Table 1. Echocardiographic data

Echocardiographic parameter	at presentation	18 months after the PDA ligation	15 months after the VSD closure
LVEDD/LVESD (mm)	67/49	68/41	53/34
Left ventricular FS	0.27	0.40	0.36
Left atrial diameter (mm)	52	42	40
Tricuspid regurgitation gradient (mmHg)	78	65	N/A
Pressure gradient across the VSD (mmHg)	no gradient	45	68

LVEDD – left ventricular end-diastolic dimension; LVESD – left ventricular end-systolic dimension; FS – fractional shortening; TR – tricuspid regurgitation; VSD – ventricular septal defect

Table 2. Hemodynamic data obtained at cardiac catheterization

Catheterization data	at presentation	4 months after the PDA ligation	17 months after the VSD closure
Systolic/diastolic/mean PAP (mmHg)	93/30/59	73/29/48	34/11/19
Mean right atrial pressure (mmHg)	13	8	4
Systolic/diastolic/mean SAP (mmHg)	95/54/77	95/68/80	115/62/80
mPAP/mSAP	0.77	0.60	0.24
Qp/Qs	2.1	2.3	1.2
PVR/SVR	0.37	0.21	0.09
PVRI (WU/m^2)	6.8	5.1	1.9

PAP – pulmonary arterial pressure; SAP – systemic arterial pressure; mPAP/mSAP – the mean pulmonary arterial-to-systemic pressure ratio; Qp/Qs – pulmonary to systemic blood flow ratio; PVRI – pulmonary vascular resistance index; PVR/SVR – pulmonary to systemic vascular resistance ratio

nary to systemic vascular resistance ratio (PVR/SVR) of 0.37 (Table 2). Testing of pulmonary vasoreactivity to 100% oxygen inhalation bore no significant pressure reduction in the pulmonary artery.

Given the presence of both VSD and PDA in association with near systemic PAH, there was an issue of appropriate management strategy. However, considering the increased pulmonary blood flow, dilated left cardiac chambers, and PVRI less than 8 WU/m^2, we opted for surgical repair. It was decided to perform a two-stage surgery involving PDA ligation as an initial procedure, followed by VSD closure in case of drop in pulmonary vascular resistance.

First, the large PDA of 15 mm in diameter was ligated. To note, the patient did not receive pulmonary vasodilators perioperatively because they were not available at that time. Echocardiographic evaluation was performed on postoperative day one and ten, three months, one year, and eighteen months (Table 1) after the PDA closure. Follow-up echocardiograms revealed marked reduction in the left atrial size and good left ventricular systolic function with mild decrease in pulmonary pressure. His condition improved as manifested by increased exercise capacity and disappearance of heart palpitations.

A repeated cardiac catheterization was performed four months after the PDA ligation and it demonstrated somewhat reduced pulmonary pressure (mPAP 48 mmHg), still large left-to-right shunting with Qp to Qs ratio of 2.3, PVR/SVR ratio < 0.3, and PVRI < 6 WU/m^2 (Table 2). Since these results were encouraging, it was decided to proceed with the complete repair, without the need for prior pulmonary vasoreactivity testing.

Surgical closure of the VSD under cardiopulmonary bypass was carried out using a unidirectional valved patch (UVP) as described by Novick et al. [7, 8]. A 5 mm fenestration was made in the primary VSD patch, and flap valve patch was created from bovine pericardial patch. Postoperative course was uneventful. The patient was started on mechanical ventilation with nitric oxide and oral Sildenafil on post-operative day one. He was weaned from mechanical ventilation on post-operative day two. Follow-up echocardiograms were performed on postoperative day one and ten, three months, one year, and 15 months after the VSD closure. Echocardiogram obtained on post-operative day one revealed normal left ventricular size and function. There was a minimal left-to-right shunting across the valved patch. At 15-month follow-up, there was a residual pressure gradient of 68 mmHg across the fenestrated patch (Table 1).

A repeat cardiac catheterization was undertaken 17 months following the last operation (Table 2). This verified that pulmonary vascular resistance and pressures returned to normal levels. Furthermore, it showed a small left-to-right shunt with the Qp to Qs ratio being markedly decreased.

The patient has been asymptomatic for eight years now. He has recently been reevaluated with echocardiography. The echocardiogram demonstrated normal both left and right ventricular function with no signs of pulmonary hypertension (Figure 1).

DISCUSSION

The estimated prevalence of PAH among adult patients with CHD is 4–28%, and the prevalence of Eisenmenger syndrome, the most severe form of PAH, approximately 1–6% [1].

The rate of progression of pulmonary vascular disease depends on the size and location of the underlying cardiac defect, the amount of left-to-right shunting, previous surgical repair, and genetic factors [9]. Ventricular septal defects are the single most common lesions causing PAH. It is estimated that 10% of all VSDs and 50% of large VSDs have the potential to cause Eisenmenger syndrome if not repaired by the age of two [10]. In addition, Eisenmenger syndrome is more likely to develop in patients with large aortopulmonary or interventricular shunts [11].

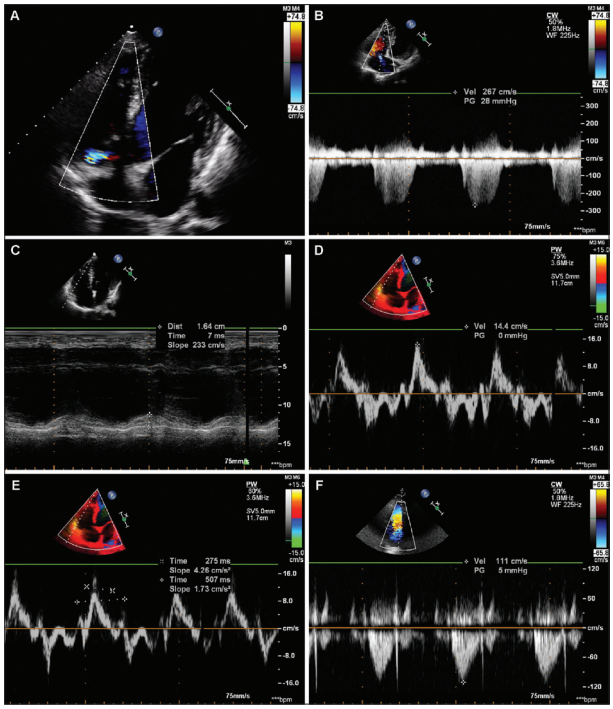

Figure 1. (A) An apical four-chamber view demonstrating left-to-right shunting through the residual ventricular septal defect; (B) This apical four-chamber view shows a peak tricuspid regurgitation jet velocity of 2.67 m/s, which gives an estimated pressure gradient of 28 mmHg across the tricuspid valve, indicating a normal right ventricular systolic pressure; (C) The normal tricuspid annular plane systolic excursion (TAPSE) as assessed with M-mode, indicating a good right ventricular longitudinal systolic function; (D) The normal tricuspid annular peak systolic velocity as assessed with right ventricular tissue Doppler imaging; (E) The right ventricular myocardial performance index (Tei index) is calculated to be 0.84, demonstrating a preserved global ventricular function; (F) A parasternal short-axis view showing a normal Doppler flow pattern in the main pulmonary artery

While there are some data regarding surgical repair for PAH-CHD in adults, very little is known about the long-term outcome of defect closure in this patient group [12, 13, 14].

In this report, we aimed to describe a management strategy for a symptomatic adult patient with severe PAH in the setting of a significant pulmonary overcirculation. The main rationale for considering the suitability for surgical repair was the presence of dilated left heart chambers, significant left-to-right shunt (Qp/Qs of 2.1), and absence of cyanosis [4]. There was, however, a high pulmonary vascular resistance (PVR/SVR ratio of 0.37, PVRI 6.8 WU/m^2). According to the now available guidelines, the patient was in the, so-called, grey zone of operability. In such cases the value of pulmonary arterial pressure and the PVR/SVR ratio, as well as the degree of pulmonary vasoreactivity, may guide clinicians in choosing patients who would benefit from surgical treatment [2, 4, 5, 12, 15].

The INOP test I was a multicenter study that gathered data on preoperative hemodynamics, including pulmonary vascular response to oxygen and NO, in patients with PAH-CHD (PVR/SVR ≥ 0.33). A PVR/SVR ratio of less than 0.42 and 0.27 with the use of oxygen alone and oxygen plus nitrogen oxide, respectively, was identified as an optimal cut-off value for determining operability (reduced risk of death or right ventricular failure after surgery) [6]. Some authors recommend that a course of selective pulmonary vasodilators be used for a sufficient period of time with subsequent assessment of response to this therapy before carrying out surgical repair for CHD [16].

There are several reports on partial closure of VSD in patients with severe PAH using a UVP [7, 8, 17–20]. During periods of acute elevation of pulmonary arterial pressure, valve opening allows blood flow from right to left. This right-to-left shunt prevents acute right ventricular failure that could be caused by refractory PAH and helps in maintaining adequate cardiac output, thus reducing the risk of early postoperative death. Therefore, unidirectional flap valve patch should enable low-risk VSD closure in the presence of pronounced, but potentially reversible, PAH [7, 8, 12].

In conclusion, evidence-based data on the management of pulmonary hypertension in adult patients with unrepaired CHD and significant left-to-right shunting are scarce. The literature, moreover, is lacking with regard to long-term outcome. The treatment approach should be individualized based on clinical presentation and the results of non-invasive and invasive investigations, including cardiac catheterization and pulmonary vasoreactivity testing.

Our report shows that normal pulmonary hemodynamics can be established in a patient with a long-standing PAH-CHD with a stepwise surgical repair. Furthermore, both short-term and long-term postoperative outcome might be favorable.

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Хируршко лечење у две етапе одраслог болесника са великим дефектом међукоморске преграде, отвореним артеријским каналом и придруженом тешком плућном артеријском хипертензијом

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САЖЕТАК

Увод Одрасли болесници са урођеним срчаним манама са лево-десним шантом и придруженом плућном артеријском хипертензијом (ПАХ) често се сматрају инoперабилним.

Приказ болесника Двадесетшестогодишњи мушкарац долази на преглед због појачаног замарања при напору и осећаја лупања срца. Постављена му је дијагноза великог перимембранозног дефекта међукоморске преграде, отвореног артеријског канала и ПАХ. Срчана мана је хируршки коригована у две етапе. Прво је учињено подвезивање артеријског канала, а потом и затварање дефекта међукоморске преграде коришћењем закрпе која функционише као једносмерни вентил. Одлуке о лечењу су биле засноване на налазима неинвазивних и инвазивних испитивања. Током праћења притисак у плућној артерији и плућна васкуларна резистенција су се вратили на нормалан ниво, а болесник није имао тегоба.

Закључак Постоји могућност да одрасла особа има урођену срчануману са значајно повећаним протоком крви кроз плућа, која се још увек може успешно оперисати. То значи да постоји индивидуалан одговор на дуготрајан лево-десни шант и да лечење треба разматрати од случаја до случаја.

Кључне речи: урођене срчане мане; плућна хипертензија; кардиохируршке операције; одрасли