Folia Cardiologica 2021
vol. 16, no. 5, pages 328–332
DOI: 10.5603/FC.2021.0047

ISSN 2353–7752
e-ISSN 2353–7760

Preoperative pharmacological management of a patient with ASD II and pulmonary hypertension before cardiac surgery

Magdalena Jachymek, Maciej Lewandowski, Marta Braksator, Małgorzata Peregud-Pogorzelska

Department of Cardiology, Pomeranian Medical University, Szczecin, Poland

Abstract

The article presents the case of a 36-year-old woman referred to the Clinic of Cardiology at the Pomeranian Medical University with exertional dyspnoea that had been increasing for 2 years. Upon examination, the patient was diagnosed with an atrial septal defect type II (ASD II) with a left-to-right shunt and pulmonary hypertension. Echocardiography revealed a defect of 4.3 cm in diameter, right ventricular systolic pressure (RSVP) 80 mm Hg, tricuspid regurgitation. Right heart catheterization (RHC) revealed: mean pulmonary arterial pressure (mPAP) 59 mm Hg, pulmonary vascular resistance 10.22 Wood units (WU) and a negative vasoreactivity test. Following a cardiosurgical consultation, the patient was qualified for preliminary pharmacological treatment and re-examination. Sildenafil was included in the treatment, followed by macitentan. Improvement in exercise tolerance was observed [in the 6-minute walk test — from 440 to 526 m; clinically from New York Heart Association (NYHA) III to NYHA I/II] as well as a decrease in N-terminal pro-B-type natriuretic peptide concentration (from 250 to 170 pg/mL). Echocardiography showed a decrease in RVSP to 60 mm Hg. In RHC performed after one year of treatment, mPAP decreased to 40 mm Hg, PVR decreased to 3.25 WU, and cardiac output increased from 5.57 to 10.44 L/min. Mixed venous oxygen saturation increased from 64.5% to 72.5%. After another cardiosurgical consultation, the patient was qualified for surgery. Closure of ASD II was performed with a pericardial patch and completed with tricuspid valve plasty. The peri-and postoperative period was uneventful, and the clinical and echocardiographic follow-up confirmed the positive effects of the treatment.

Key words: pulmonary hypertension, atrial septal defect, closure

Folia Cardiologica 2021; 16, 5: 328–332

Case report

The patient was referred to the Cardiology Department at 36 years of age. She had been experiencing symptoms in the form of progressive exertional dyspnoea for two years. There were no previous symptoms suggestive of a heart defect, even though the patient had delivered three children. Initially, she underwent diagnostic tests in a pulmonology unit for suspected sarcoidosis, but this was excluded, and what attracted attention was abnormal pulmonary vasculature on imaging tests [computed tomography (CT) of the thorax]. Transthoracic echocardiography (TTE) and transoesophageal echocardiography (TEE) were performed that revealed atrial septal defect type II (ASD II) with left-to-right shunting and signs suggestive of pulmonary hypertension (PH). Pulmonary perfusion scintigraphy showed changes consistent with pulmonary embolism and the patient was started on rivaroxaban. During further diagnostic tests at the Cardiology Department, coronary angiography and right heart catheterization (RHC) were performed, and after

Address for correspondence: Magdalena Jachymek MD, Klinika Kardiologii, Pomorski Uniwersytet Medyczny w Szczecinie, ul. Powstańców Wielkopolskich 72, 70–111 Szczecin, Poland, e-mail: magdajachymek@gmail.com

cardio-surgical consultation the patient was qualified for initial pharmacological management, and then for reassessment and qualification for further treatment.

Diagnostic test results

Electrocardiography

The sinus rhythm is regular, at 77 bpm. Intermediate heart axis. Incomplete right bundle branch block. Features of right atrial enlargement.

Echocardiography

An echocardiography study has confirmed significantly increased right ventricular systolic pressure (RSVP) at 80 mm Hg, severe tricuspid regurgitation, right ventricular systolic dysfunction with tricuspid annular plane systolic excursion (TAPSE) at 14 mm. The right atrium was enlarged (the right atrium area was 20 cm^2). TEE showed an atrial septal defect with a diameter of 4.3 cm, with a residual border and dominant left-to-right shunting (Figure 1). The results are presented in Table 1.

Coronary angiography

No abnormalities were found in coronary arteries.

Right heart catheterisation and pulmonary artery angiography

RHC has revealed significantly elevated mean pulmonary artery pressure (mPAP) at 59 mm Hg, reversibility testing with epoprostenol was negative. Pulmonary artery angiography has not revealed thrombotic changes — thromboembolic aetiology of PH was excluded. The exact measurements are presented in Table 1.

Management plan and treatment

The patient was consulted by cardiac surgeons in a Heart Team and qualified for initial pharmacotherapy, then the re-assessment and qualification for a further treatment. During her first hospital stay, the patient was started on sildenafil 60 mg/day and rivaroxaban was discontinued (after 5 months of treatment).

Monitoring the effects of treatment

One month after the first RHC, follow-up tests were performed and macitentan at 10 mg/day was included in the treatment. Subsequent follow-ups demonstrated improvement of exercise tolerance — the distance walked during the 6-minute walk test (6MWT) increased from 440 to 526 m (Figure 2), complaints were reduced [from New York Heart Association (NYHA) III to NYHA I/II] and N-terminal pro-B-type natriuretic peptide (NT-proBNP) blood levels were reduced from 250 to 170 pg/mL (Figure 3). The echocardiogram has revealed a reduction in RVSP to 60 mm Hg and an improvement in the systolic function of the right ventricle (TAPSE 20 mm). In a follow-up RHC performed after one year of pharmacotherapy, there was a significant reduction of pulmonary artery pressure (mPAP 40 mm Hg), reduced pulmonary vascular resistance [from 10.22 Woods units (WU) to 3.25 WU], increased left-to-right shunting (Qp/Qs increased from 1.67 to 1.89), and an increase in cardiac output (CO) from 5.57 to 10.44 L/min. Mixed venous oxygen saturation increased from 64.5% to 72.5% (Table 1).

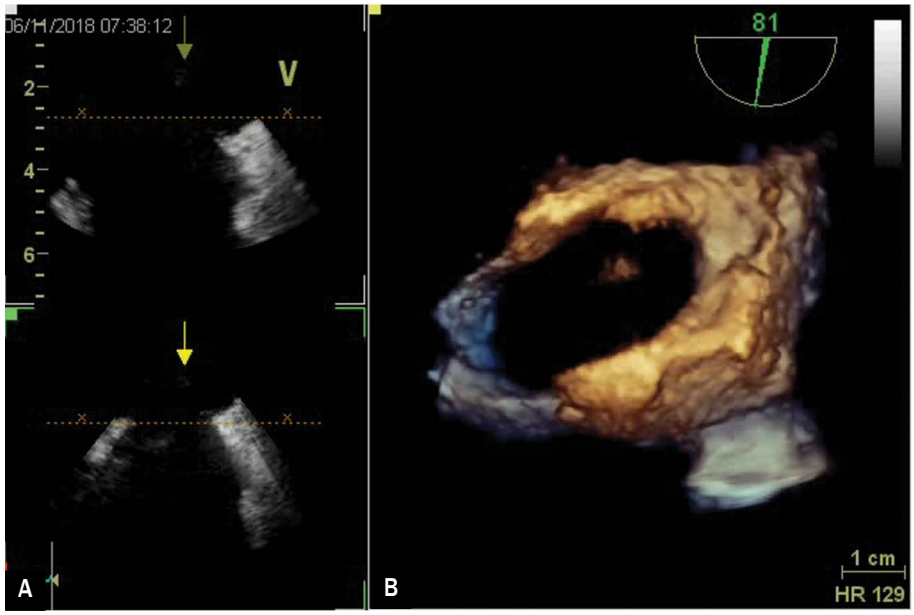

Figure 1A, B. A three-dimensional transoesophageal echocardiogram

Table 1. A comparison of measurements obtained during right heart catheterisation (RHC) and transthoracic echocardiography (TTE) before starting the treatment, and after a year of pharmacological management of pulmonary hypertension

Measurements obtained during RHC	Baseline RHC	Control RHC	Measurements obtained during TTE	Baseline TTE	Control TTE
mPAP [mm Hg]	59	40	RVSP [mm Hg]	80	60
RAP [mm Hg]	4	6	RAA [cm^2]	20	22.4
PW [mm Hg]	8	6	TAPSE [mm]	14	20
Qp/Qs	1.67	1.89	PAD [mm]	32	34
CO [L/min]	5.57	10.44	RVD [mm]	37	52
PVR [WU]	10.22	3.25	AcT [ms]	51	23
SatAo [%]	98	95	Qp/Qs	3	
Sat IVC [%]	45	72			
Sat SVC [%]	71	73			
Sat RA [%]	69	96			
Sat RV [%]	80	87			
Sat PA [%]	78	85			
MSV [%]	64.5	72.25			

mPAP — mean pulmonary arterial pressure; RAP — right atrial pressure; PW — pulmonary artery wedge pressure; Qp/Qs — pulmonary to systemic blood flow ratio; CO — cardiac output; PVR — pulmonary vascular resistance; WU — Wood units; Sat — saturation; Ao — aorta; IVC — inferior vena cava; SVC — superior vena cava; RA — right atrium; RV — right ventricle; PA — pulmonary artery; RVSP — right ventricular systolic pressure; RAA — right atrium area; TAPSE — tricuspid annular plane systolic excursion; PAD — pulmonary artery diameter; RVD — transverse right ventricular diameter; Act — pulmonary acceleration time

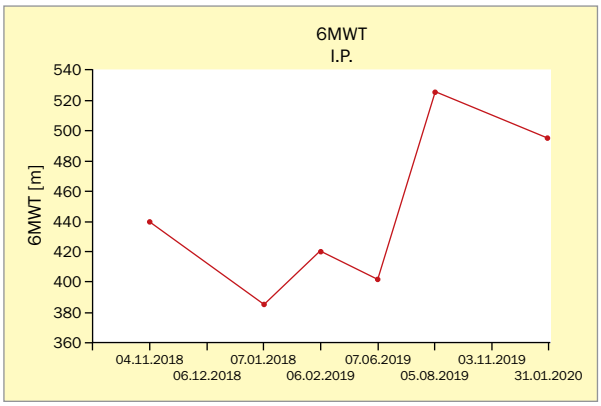

Figure 2. Distance walked in the 6-minute walk test (6MWT)

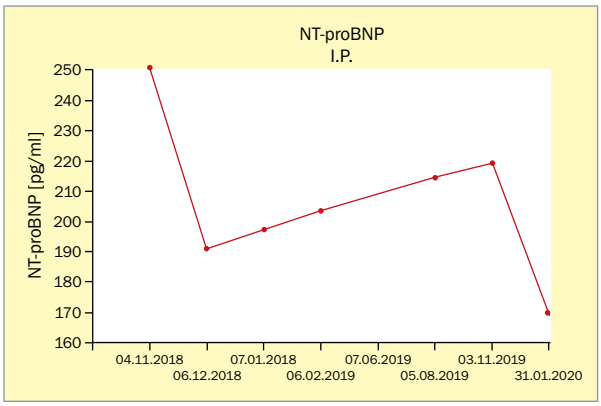

Figure 3. The blood level of N-terminal pro-B-type natriuretic peptide (NT-proBNP)

Another cardiosurgical consultation of the patient was held, as a result of which the patient was qualified for surgery. Atrial septal defect repair with a pericardial patch was performed with tricuspid valve repair. The peri- and postoperative course was uncomplicated, and the follow-up revealed that the good clinical and echocardiographic outcome of the applied treatment continues. An echocardiogram performed shortly after the procedure did not show a prominent wave of tricuspid regurgitation. Pharmacological treatment of PH was continued. One year after the surgery, the patient was able to walk 550 m in the 6MWT, complaints persisted at NYHA I, and NT-proBNP level dropped to 80 pg/mL.

Discussion

ASD is the most frequently diagnosed congenital heart defect in adults. The increased flow of blood through the pulmonary vascular bed caused by left-to-right shunting gradually leads to irreversible changes in pulmonary vessels, elevated tension and resistance in pulmonary arteries and, ultimately, to reversal of the shunt (Eisenmenger's syndrome). First symptoms most often develop in the third or fourth decade of life as reduced exercise tolerance, syncopes and palpitations. According to the European Society of Cardiology (ESC) Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension of 2015, PH caused by

a congenital heart defect belongs to group 1 — pulmonary arterial hypertension (PAH) [1].

Repair of the defect has been proven to have many advantages: improvement of physical capacity measured with cardiopulmonary exercise test, reduction of symptoms (NYHA class) and reduction of the right ventricle and right atrium sizes in echocardiography [2]. Positive changes are obtained even in patients who undergo correction after the age of 60, resulting in an improvement in their physical capacity and quality of life [3]. However, these data are for patients without a diagnosis of PH.

The decision on the repair of the defect, if an ultrasound has revealed PH, is taken based on cardiac catheterisation. Pulmonary vascular bed resistance higher than 5 WU and Eisenmenger's syndrome are contraindications for the repair of the defect [4]. Repair of defects in patients with significant PH worsens the prognosis and may result in acute right ventricular failure caused by a sudden reduction of right ventricle preload. The patient's pulmonary resistance was at 10.22 WU, which is why she was qualified for initial pharmacotherapy. Sequential treatment with sildenafil and macitentan was started (IB recommendation class) [1].

There are two possibilities of ASD repair. The surgical method is safe and has a low mortality rate (< 1%). The defect may also be repaired percutaneously, providing that its diameter is 38 mm or less and that there is at least a 5 mm border (in addition to the peri-aortic part) [4]. Both methods have similar efficacy but percutaneous repair is associated with a shorter hospital stay and lower perioperative morbidity. Currently, the percutaneous method is the treatment of choice, and surgical treatment is reserved for patients who do not qualify for a percutaneous procedure. In the patient described in this report, the septal defect was too large, which is why its repair with a pericardial patch through lateral mini-thoracotomy access was performed with tricuspid valve repair.

The concept of pharmacological treatment aiming at a safe repair of a defect was not formulated precisely in ESC guidelines on PH of 2015, as there was not enough scientific evidence to prove its efficacy and safety at that time. Guidelines on the treatment of congenital heart defects in adults of 2010 [5] permitted repair of a defect with associated PH after initial targeted treatment. New ESC guidelines on congenital heart defects in adults were published in 2020. They allow the possibility of ASD repair following pharmacological treatment of PH if the applied treatment results in a reduction in pulmonary resistance to < 5 WU, and Qp/Qs continues at > 1.5 and no desaturation is observed on exertion (IIb class recommendations) [4]. Currently, in such cases, percutaneous fenestrated ASD closure is recommended. In the case described in this study, surgical repair of the defect was performed without a residual shunt. Of note, the treatment was started before the 2020 guidelines were published. The recommendation for leaving a shunt is based on expert opinions (recommendation C) and not evidenced by randomised control trials. Moreover, surgical repair of the defect assumes its complete removal. In addition to the case presented here, the literature describes many examples of effective treatment with targeted medications used in PH for reducing pulmonary tension before a defect repair surgery [6–8]. Such an approach seems to be advisable for significant improvement of haemodynamic parameters obtained with the applied treatment. This report is another voice in the discussion on this difficult situation that requires special attention.

Conclusions

The case reported here demonstrates the efficacy of pharmacological treatment of PH in the course of a congenital heart defect. A significant improvement of the haemodynamic parameters was obtained in the pulmonary circulation. This allowed to safely perform heart surgery removing the primary cause of the dysfunctions detected.

Conflict of interest

The authors declare no conflict of interest during the preparation of the article.

References

1. Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. Kardiol Pol. 2015; 73(12): 1127–1206, doi: 10.5603/kp.2015.0242.
2. Prochownik P, Przewłocki T, Podolec P, et al. Improvement of physical capacity in patients undergoing transcatheter closure of atrial septal defects. Post Kardiol Interw. 2018; 14(1): 90–94, doi: 10.5114/aic.2018.74360, indexed in Pubmed: 29743909.
3. Komar M, Przewlocki T, Olszowska M, et al. The benefit of atrial septal defect closure in elderly patients. Clin Interv Aging. 2014; 9: 1101–1107, doi: 10.2147/CIA.S62313, indexed in Pubmed: 25075179.
4. Baumgartner H, De Backer J, Babu-Narayan SV, et al. ESC Scientific Document Group. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021; 42(6): 563–645, doi: 10.1093/eurheartj/ehaa554, indexed in Pubmed: 32860028.
5. Baumgartner H, Bonhoeffer P, De Groot NMS, et al. Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC), Association for European Paediatric Cardiology (AEPC), ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010; 31(23): 2915–2957, doi: 10.1093/eurheartj/ehq249, indexed in Pubmed: 20801927.
6. Taniguchi Yu, Emoto N, Miyagawa K, et al. Subsequent shunt closure after targeted medical therapy can be an effective strategy for secundum atrial septal defect with severe pulmonary arterial hypertension: two case reports : strategy for ASD with severe PAH. Heart Vessels. 2014; 29(2): 282–285, doi: 10.1007/s00380-013-0351-0, indexed in Pubmed: 23595779.

7. Park YK, Park JH, Yu JH, et al. Transient use of oral bosentan can be an additional option to reduce pulmonary arterial hypertension in a patient with severe pulmonary arterial hypertension associated with atrial septal defect. J Cardiovasc Ultrasound. 2011; 19(3): 159–162, doi: 10.4250/jcu.2011.19.3.159, indexed in Pubmed: 22073329.

8. Kim YH, Yu JJ, Yun TJ, et al. Repair of atrial septal defect with Eisenmenger syndrome after long-term sildenafil therapy. Ann Thorac Surg. 2010; 89(5): 1629–1630, doi: 10.1016/j.athoracsur.2009.10.027, indexed in Pubmed: 20417794.