




Complex percutaneous intervention on pulmonary arteries in an adult patient with a corrected Taussig–Bing anomaly

Złożona interwencja przezskórna na tętnicach płucnych u dorosłego pacjenta ze skorygowaną anomalią Taussig–Binga

Andrzej Tomasz Wittczak¹ , Marek Rybak¹, Paweł Dryżek², Tomasz Moszura² ,
Marek Maciejewski¹, Agata Bielecka-Dąbrowa^{1, 3} 

¹Department of Cardiology and Congenital Diseases of Adults, Polish Mother's Memorial Hospital Research Institute, Łódź, Poland

²Department of Cardiology, Polish Mother's Memorial Hospital Research Institute, Łódź, Poland

³Department of Hypertension, Chair of Nephrology and Hypertension, Medical University of Lodz, Łódź, Poland

Abstract

This study reports a case of a successful complex percutaneous intervention on pulmonary arteries in a 23-year-old adult patient with a corrected Taussig–Bing anomaly. The patient had a history of multiple surgeries, including an arterial switch operation, the Bentall procedure, and mitral valve replacement. On admission, the patient was asymptomatic, however significant stenosis of the pulmonary arteries was detected. The patient was qualified for cardiac catheterization. The complex, high-risk procedure with the implantation of three stents improved the morphology of the right pulmonary artery and consequently the function of the right ventricle. It is concluded that with the remarkable development of percutaneous techniques, more and more patients are receiving optimal, personalised treatment.

Key words: congenital heart disease, Taussig–Bing anomaly, double outlet right ventricle, percutaneous intervention, pulmonary artery stenting

Folia Cardiologica 2023; 18, 4: 176–181

Introduction

The Taussig–Bing anomaly (TBA) is a rare congenital heart disease (CHD), a subtype of double outlet right ventricle (DORV) [1]. In TBA the aorta originates entirely from the right ventricle (RV), the pulmonary artery (PA) arises from above the non-restrictive ventricular septal defect (VSD) and there is no pulmonary-mitral fibrous continuity [2]. DORV accounts for approximately 1% of all cases of CHD and its prevalence is reported to be 0.1 per 1000 live births; TBA is the third most common type of DORV [3, 4]. The arterial switch operation (ASO) with VSD closure is the

method of choice for the treatment of TBA [2]. Pulmonary arteries stenosis is a relatively frequent complication of the ASO [5, 6]. A percutaneous intervention on PA and stenting is an established method for the management of this complication [5, 6].

Case report

Patient presentation

A 23-year-old man with a corrected Taussig–Bing anomaly was admitted to the Department of Cardiology and Congenital Diseases of Adults for percutaneous intervention on the

Address for correspondence: Andrzej Tomasz Wittczak MD, Klinika Kardiologii i Wad Wrodzonych Doroslych, Instytut Centrum Zdrowia Matki Polki w Łodzi, ul. Rzgowska 281/289, 93–338 Łódź, Poland, e-mail: andrzejwitt2@gmail.com

This article is available in open access under Creative Common Attribution-Non-Commercial-No Derivatives 4.0 International (CC BY-NC-ND 4.0) license, allowing to download articles and share them with others as long as they credit the authors and the publisher, but without permission to change them in any way or use them commercially.

pulmonary arteries. He had a history of multiple surgeries. Three months after birth, he underwent pulmonary artery banding and patent ductus arteriosus ligation. One year later, he had an arterial switch operation with the LeCompte manoeuvre; the coexisting ventricular septal defect was closed with a Gore-Tex patch. At the age of 18, during a school lesson, he underwent cardiopulmonary arrest caused by ventricular fibrillation. He was first resuscitated by witnesses and then by paramedics. After the patient was discharged from the intensive care unit, he was admitted to the clinic, where significant regurgitation of neo-aortic and mitral valves was found. It was decided that the patient should receive an implantable cardioverter-defibrillator for secondary prevention of sudden cardiac death before being referred to cardiac surgery. The subcutaneous implantable cardioverter-defibrillator Boston Scientific Emblem was implanted. Four years later (at the age of 22) he was finally qualified for cardiac surgery in another centre. The operation included the Bentall procedure, mitral valve replacement, and right pulmonary arterioplasty (using a bovine pericardial patch). The follow-up hospitalization in the department showed good function of the mechanical valves, but stenosis of both pulmonary arteries was detected. The patient was referred to the cardiac surgery clinic for consultation and the date of the next hospitalization in the centre was set.

On admission, the patient reported no symptoms. On physical examination, heart sounds were regular (HR 65 bpm), with mechanical valves click sounds and a systolic murmur heard best in the pulmonary valve auscultation area. His blood pressure was 102/66 mm Hg. His pharmacological treatment consisted of bisoprolol 3.75 mg/day, acenocoumarol (target INR: 2.5–3.5), potassium chloride 600 mg/day, and magnesium citrate (100 mg Mg²⁺/day).

Initial work up

Routine laboratory tests were normal. International normalized ratio was within the therapeutic range (3.31) and N-terminal pro-B-type natriuretic peptide was not elevated (76 pg/mL).

Transthoracic echocardiography (Figure 1) showed left ventricular hypertrophy with increased left ventricular internal diameter at end-diastole and end-systole (62 and 53 mm, respectively) and left atrial enlargement (left atrium diameter – 42 mm; left atrial volume index – 64.3 mL/m²). Dimensions of other cardiac chambers were normal. The Gore-Tex patch served as an interventricular septum (IVS), no shunt was visualised, however, this artificial IVS was dyskinetic (in the basal and mid segments). No other wall motion abnormalities were observed. Left ventricular ejection fraction was reduced to 44% – it was a consequence of abnormal IVS motion. In the aortic position, the mechanical aortic valve was observed: occluder motion was normal, the maximum pressure gradient (PG) was 12 mm Hg and

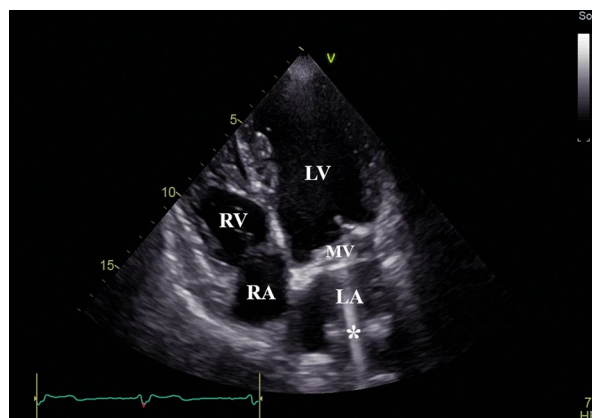


Figure 1. Transthoracic echocardiography, apical view. Visible increased left ventricular internal diameter, the mechanical mitral valve and the comet-tail artifact (marked as *); LA – left atrium; LV – left ventricle; MV – mitral valve; RA – right atrium; RV – right ventricle

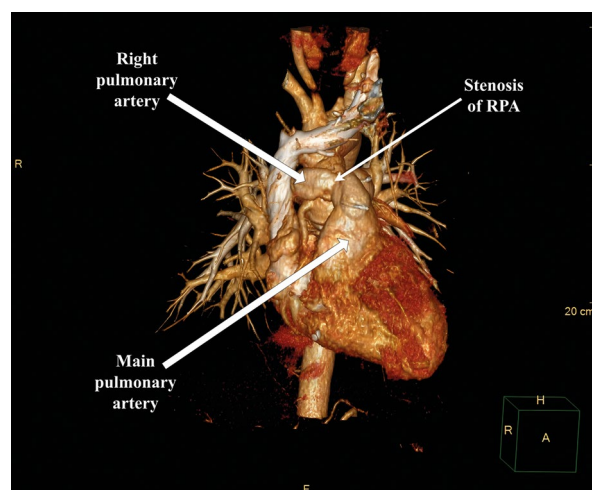


Figure 2. The computed tomography angiography with 3D reconstruction. Visible stenosis of the right pulmonary artery (RPA)

a small paravalvular leak was detected. The mechanical mitral valve was observed in the mitral position: occluder motion was normal and the maximum PG was 10 mm Hg. The tricuspid valve was normal. The PG of the pulmonary valve was 70/42 mm Hg (maximum and mean, respectively) and the peak velocity was 4.2 m/s. These measurements suggested significant stenosis of the PA. The visible part of the main PA measured 8 mm in the narrowest part. The systolic function of the RV was slightly reduced (tricuspid annular plane systolic excursion = 16 mm; peak lateral tricuspid annular systolic velocity [S'] = 8 cm/s).

The computed tomography angiography (Figure 2) showed stenosis of the entire right PA (22 × 7 mm) and proximal part of the left PA (16 × 8 mm); the main PA was 33 × 17 mm.

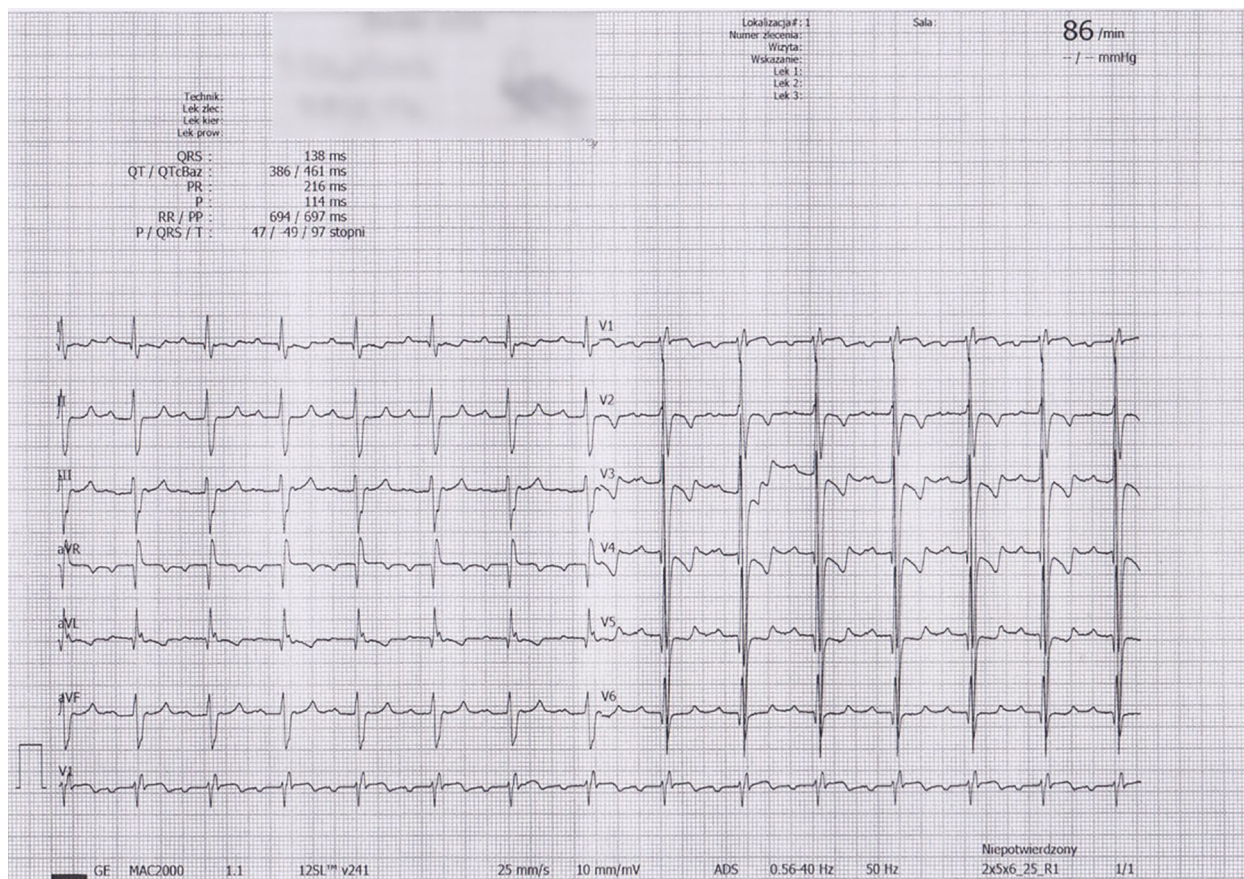


Figure 3. 12-lead electrocardiogram. Visible sinus rhythm, left axis deviation, right bundle branch block (QRS = 138 ms), left anterior hemiblock, features of left ventricular hypertrophy and 1st degree atrioventricular block (PQ = 210 ms)

The 12-lead electrocardiogram (Figure 3) showed sinus rhythm, left axis deviation, right bundle branch block (QRS = 138 ms), left anterior hemiblock, features of left ventricular hypertrophy and 1st degree atrioventricular block (PQ = 210 ms).

Diagnosis and management

The heart team, consisting of conservative cardiologists and interventional cardiologists, decided to qualify the patient for cardiac catheterization with the intention of implanting stents in the pulmonary arteries.

During catheterisation, the left pulmonary artery was measured to be 13.5 mm in the proximal segment and 16 mm in the distal segment. The proximal segment of the right pulmonary artery (RPA) was significantly stenosed with a diameter of only 4.1 mm (Figure 4 A–B). The further segment of the RPA was coiled spirally along with a bovine pericardial patch (from the right pulmonary arterioplasty); the diameter was 7.3 mm. The distal segment of the RPA was 14 mm in diameter. The pressure in the RV was about 50% of the systemic pressure (which was low at 82/43 mm Hg due to the reaction to the anaesthetics). Based on these

findings, an appropriate treatment plan was implemented. The BeGraft stent 14 × 29 mm was implanted in the proximal segment of the RPA. After implantation, the pressure difference in the area of the coiled artery was still significant (20 mm Hg). The 10 × 30 mm Formula stent was implanted distally (using a 14 × 39 mm balloon-in-balloon catheter). In the control angiography, the RPA wall outline was irregular but without significant pressure differences. It was decided to implant the third stent (BeGraft 14 × 39 mm), which connected two previously placed stents (Figure 4 C–D). At the end of the procedure, there were no significant differences in the RPA pressures and RV pressure was < 50% of systemic pressure.

Follow-up

There were no postprocedural complications and the patient was asymptomatic. Control transthoracic echocardiography showed a reduction in the maximum pulmonary valve pressure gradient to 40 mm Hg (from 70 mm Hg) and a reduction in peak velocity to 3.12 m/s (from 4.2 m/s). The function of the RV ventricle improved with tricuspid annular plane systolic excursion of 19 mm and S' of 9 cm/s (before

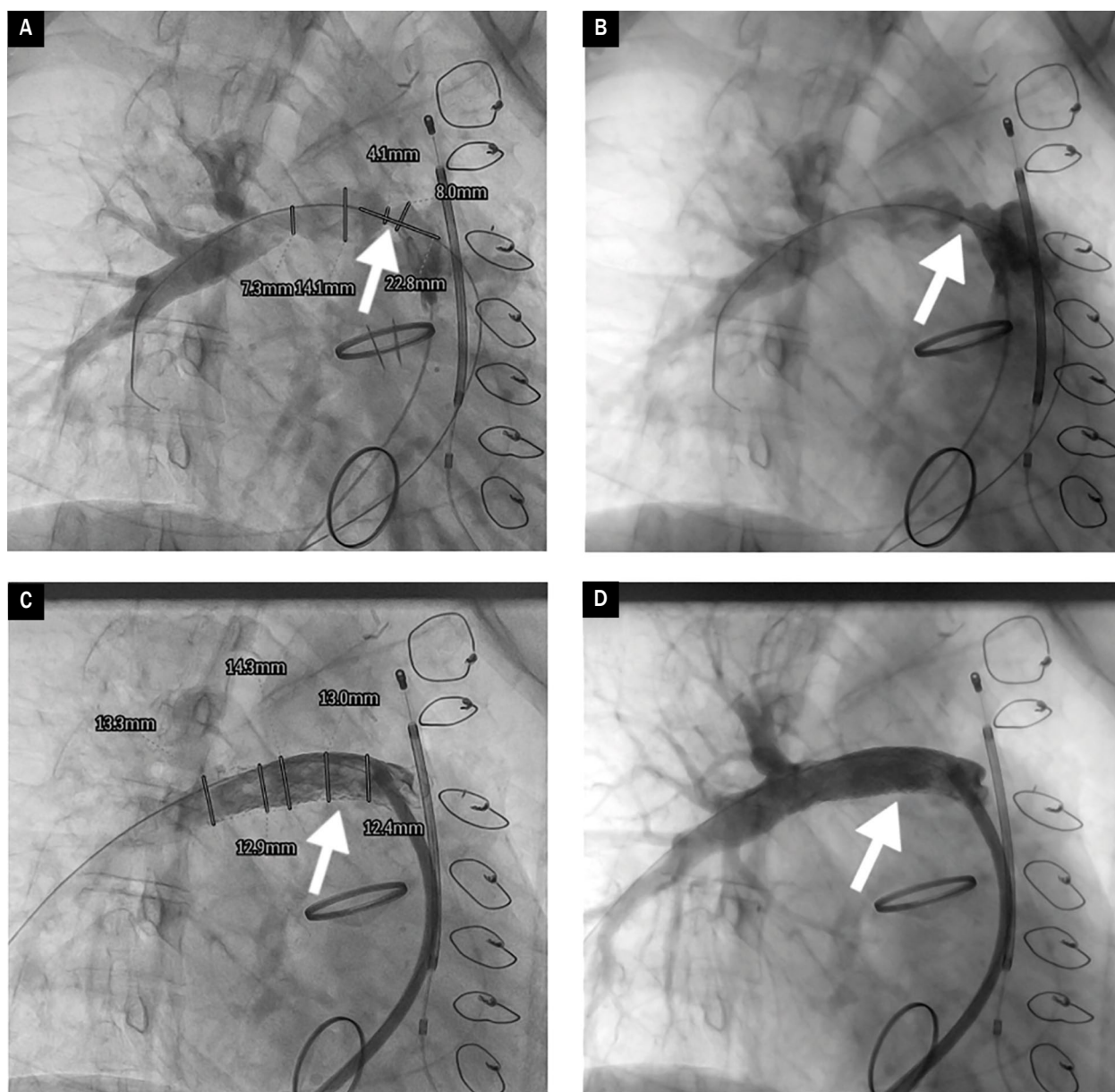


Figure 4. Pulmonary arteries angiography: **A, B.** Right pulmonary artery before stenting, the arrow points to the site of the stenosis; **C, D.** Right pulmonary artery angiography at the end of the procedure – implanted stents visible, the arrow points to the site of the former stenosis

the procedure it was 16 mm and 8 cm/s, respectively). Computed tomography angiography was performed one week after the procedure and showed three appropriately placed stents in the RPA with no evidence of mechanical damage. The postprocedural 24-hour Holter electrocardiography showed 2 supraventricular extrasystolic beats and 5 ventricular extrasystolic beats; no pauses were observed.

Eight days after the procedure, the patient was discharged from the department. His pharmacological treatment was slightly modified by reducing the dose of bisoprolol to 2.5 mg/day. He was instructed to report to the cardiology outpatient clinic at the centre in a month.

Discussion

The arterial switch operation with VSD closure is the method of choice for the treatment of TBA [2]. The operation is preferably performed early in life, using a primary one-stage approach [2]. According to the European Society of Cardiology CHD guidelines, the most common complications of the ASO include: 1) neo-aortic root dilatation, resulting in aortic regurgitation; 2) supra valvular pulmonary stenosis and pulmonary branch stenosis; 3) problems with the coronary arteries (which can cause LV dysfunction and ventricular arrhythmias); 4) acute angle of the aortic

arch arteries [5]. The LeCompte manoeuvre (frequently performed during ASO) may involve “stretching” of the pulmonary artery branches while moving the pulmonary artery bifurcation anterior to the proximal neo-ascending aorta [6]. This may predispose patients to develop branch pulmonary artery stenosis [6]. The CHD guidelines authors recommend that after ASO, stenting “should be considered for PA branch stenosis, regardless of symptoms, if > 50% diameter narrowing and right ventricular systolic pressure > 50 mm Hg and/or related reduced lung perfusion are present” [5].

In the described case, the patient underwent ASO with VSD closure in the second year of his life. The operation was complicated by significant neo-aortic valve regurgitation, which was treated with a Bentall procedure. Coronary artery pathology was excluded by computed tomography angiography performed before cardiac surgery. However, the problem of PA stenosis was serious. The complex, high-risk percutaneous procedure was the only treatment option. Fortunately, it was successful in improving the morphology of the RPA and consequently the function of the RV.

Conclusions

In conclusion, the described case illustrates that with the remarkable development of percutaneous techniques, more and more patients are receiving optimal, personalised

treatment for their condition. As the prevalence of CHD increases in the community, it is important to emphasise the need for specialised adult CHD centres to be available to all CHD patients. Lifelong and regular follow-up in such a centre is crucial for all patients in this group.

Article information

Author contributions

ATW – prepared the manuscript; MR, PD, TM, MM, ABD – took part in the clinical decision-making process; MR and ABD – provided medical care for patient; PD – performed the percutaneous procedure; ABD – conceived this study, reviewed and improved the manuscript.

Conflict of interest

The authors declare no conflict of interest.

Ethics statement

Patient consent has been signed and collected in accordance with the journal’s patient consent policy.

Funding

None declared.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/fovia_cardiologica.

Streszczenie

W niniejszej pracy przedstawiono przypadek udanej złożonej interwencji przezskórnej na tętnicach płucnych u 23-letniego pacjenta ze skorygowaną anomalią Taussig–Binga. Wada skorygowana była zabiegiem typu *arterial switch*; ponadto w wywiadzie odnotowano przebytą operację Bentalla i wymianę zastawki mitralnej. Przy przyjęciu pacjent był bezobjawowy, jednak wykryto istotne zwężenie tętnic płucnych. Pacjent został zakwalifikowany do cewnikowania serca. Złożony zabieg wysokiego ryzyka z implantacją trzech stentów poprawił morfologię prawej tętnicy płucnej, a w konsekwencji funkcję prawej komory. Podsumowując, dzięki niezwykłemu rozwojowi technik przezskórnych, coraz więcej pacjentów otrzymuje optymalne, spersonalizowane leczenie.

Słowa kluczowe: wrodzona wada serca, anomalia Taussig–Binga, dwuuściowa prawa komora, interwencja przezskórna, stentowanie tętnicy płucnej

Folia Cardiologica 2023; 18, 4: 176–181

References

1. Konstantinov IE. Taussig-Bing anomaly: from original description to the current era. *Tex Heart Inst J*. 2009; 36(6): 580–585, indexed in Pubmed: [20069085](https://pubmed.ncbi.nlm.nih.gov/20069085/).
2. Schwarz F, Blaszczyk HC, Sinzobahamya N, et al. The Taussig-Bing anomaly: long-term results. *Eur J Cardiothorac Surg*. 2013; 44(5): 821–827, doi: [10.1093/ejcts/ezt148](https://doi.org/10.1093/ejcts/ezt148), indexed in Pubmed: [23644700](https://pubmed.ncbi.nlm.nih.gov/23644700/).

3. Liu Y, Chen S, Zühlke L, et al. Global birth prevalence of congenital heart defects 1970-2017: updated systematic review and meta-analysis of 260 studies. *Int J Epidemiol.* 2019; 48(2): 455–463, doi: [10.1093/ije/dyz009](https://doi.org/10.1093/ije/dyz009), indexed in Pubmed: [30783674](https://pubmed.ncbi.nlm.nih.gov/30783674/).
4. Vergnat M, Baruteau AE, Houyel L, et al. Late outcomes after arterial switch operation for Taussig-Bing anomaly. *J Thorac Cardiovasc Surg.* 2015; 149(4): 1124–30; discussion 1130, doi: [10.1016/j.jtcvs.2014.10.082](https://doi.org/10.1016/j.jtcvs.2014.10.082), indexed in Pubmed: [25466859](https://pubmed.ncbi.nlm.nih.gov/25466859/).
5. 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](https://doi.org/10.1093/eurheartj/ehaa554), indexed in Pubmed: [32860028](https://pubmed.ncbi.nlm.nih.gov/32860028/).
6. Lee J, Abdullah Shahbah D, El-Said H, et al. Pulmonary artery interventions after the arterial switch operation: Unique and significant risks. *Congenit Heart Dis.* 2019; 14(2): 288–296, doi: [10.1111/chd.12726](https://doi.org/10.1111/chd.12726), indexed in Pubmed: [30620141](https://pubmed.ncbi.nlm.nih.gov/30620141/).