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Senning operation as a palliative therapy for a girl with complex heart defect and Eisenmenger syndrome (RCD code: II-1A.4d)

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Abstract

Development of irreversible pulmonary hypertension in D-transposition of the great arteries (D-TGA) with ventricular septal defect (VSD) is a well-known phenomenon. Coexistence of left ventricular outflow tract obstruction (LVOTO) should theoretically protect the patient against pulmonary arterial hypertension (PAH). Application of the Blalock-Taussig shunt (B-TS) temporarily improves condition of the patient, but may contribute to irreversible PAH requiring palliative complex therapy. We present a case of a 18 year-old female with congenital complex heart defect: D-TGA with inflow VSD and subpulmonary stenosis, Eisenmenger syndrome, who was treated with several subsequent palliative cardiosurgical operations, including B-TS and Senning procedure and eventually, combined PAH-specific therapy, what resulted in stabilization of clinical status. JRCD 2016; 3 (1): 14–16

Key words: right heart catheterization, echocardiography, pulmonary hypertension, endothelin receptor antagonist, phosphodiesterase type 5 inhibitor, combination therapy

Case presentation

We present a case of an 18 year-old female with a complex congenital heart defect and Eisenmenger syndrome treated with palliative cardiosurgical operations and pharmacotherapy for pulmonary arterial hypertension (PAH).

The patient was delivered at term with body weight of 3700 g. Central cyanosis and soft systolic murmur with loudness of 2/6 in Levine scale were present at birth. Peripheral oxygen saturation (SpO_2) was very low (60–70%) with capillary blood partial oxygen pressure (pO_2) of 27 mm Hg. Transthoracic echocardiography (TTE) showed complex congenital heart defect: D-transposition of the great arteries (D-TGA), large inlet ventricular septal defect (VSD) with tricuspid valve (TV) straddling, patent foramen ovale (PFO) and sub-pulmonary stenosis (PS) due to the accessory connective tissue strands. Prostaglandin E1 (PGE1) infusion was started. Subsequent TTE study revealed moderate left ventricular outflow tract obstruction (LVOTO) with pressure gradient of 50 mmHg and no left-to-right shunt through the PFO.

The child was qualified for Blalock-Taussig shunt (B-TS) at the age of 4 weeks. After the operation SpO_2 increased to 75–80% with pO_2 of 36 mm Hg. The patient was discharged home in a satisfactory clinical condition.

At the age of 15 months cyanosis became intense with a drop of saturation to 60–70%. TTE showed D-TGA, large inlet VSD, mild LVOTO (pressure gradient of 17 mm Hg), extremely dilated pulmonary trunk and occluded B-TS. Cardiac catheterization revealed severe pulmonary hypertension (PH) with high pulmonary vascular resistance (PVR) of 10 Wood units (WU) and abnormal pulmonary-to-systemic resistance ratio (Rp/Rs) of 0.6 (Table 1, 2) unresponsive to 100% oxygen inhalation test. The patient was qualified for atrial septectomy to facilitate mixing of the blood at the atrial level. However, no improvement was observed after the operation. Therefore, the patient was qualified for palliative Senning procedure which was performed at the age of 29 months. During the operation a baffle within the atria was made in order to reroute

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Table 1. Pressures and saturations measured during heart catheterization revealed pulmonary arterial hypertension with normal pulmonary artery wedge pressure and significant desaturation in the superior vena cava

	SaO_2	PaO_2	Pressures		
			Systolic	Diastolic	Mean
SVC	39	24,9	-	-	-
RV	40	10	80	-	-
MPA	87	51,7	70	53	62
PAWP	-	-	-	-	6
LV	76	-	90	6	-
Ao asc	54	30,4	80	57	68

PAWP – pulmonary artery wedge pressure; SVC – superior vena cava; RV – right ventricle; MPA – main pulmonary artery; LV – left ventricle; Ao asc – ascending aorta; SaO2- oxygen saturation; PaO2- partial pressure of oxygen

the deoxygenated venous blood via mitral valve to pulmonary circulation. Oxygenated blood from the pulmonary veins was re-routed to the TV and systemic circulation. Due to PAH with Eisenmenger reaction VSD was left patent to provide bidirectional blood flow. After the operation SpO_2 increased significantly, up to 86% and the condition of the patient improved.

At the age of 10, haemodynamic evaluation revealed worsening of PAH with higher PVR of 25 WU and pathological Rp/Rs ratio of 0.8. Sildenafil therapy was applied with temporary relief of symptoms. During follow-up the patient complained of weakness and low exercise tolerance in functional class III by World Health Organization and worsening of cyanosis on exertion. The subsequent catheterization at the age of 17, disclosed PVR of 21 WU and Rp/Rs of 0.9 (Table 2). Angiocardiographic evaluation revealed tortuous tapering of peripheral pulmonary arteries (Figure 1) indicating progression of PAH. Therefore, combined treatment of sildenafil with bosentan was initiated, resulting in further improvement.

During ongoing follow-up, the patient remained stable in WHO functional class III. Her 6-minute walking-test distances ranged from 210 to 305 m and levels of N-terminal prohormone of brain natriuretic peptide from 137–233 pg/ml. At the age of 18 she was referred to the Centre of Rare Cardiovascular Diseases at the John Paul II Hospital in Krakow.

The combined therapy with bosentan and sildenafil was maintained and the patient did not report any significant side effects.

Review of literature

D-TGA is the most frequent cyanotic heart defect. It is present in 5 to 7% of patients with congenital heart diseases [1]. Clinical manifestation depends on complexity of D-TGA and coexisting other heart defects. Major symptoms include cyanosis and heart murmur. Cardinal feature of D-TGA is ventriculo-arterial discordance with the aorta arising from morphological right ventricle and the pulmonary trunk from morphological left ventricle. Pulmonary and systemic circulations in D-TGA are parallel. Deoxygenated blood comes back from the systemic veins into the right atrium and through the right ventricle is pumped directly into the aorta bypassing the lungs. On the opposite, highly oxygenated blood returns from pulmonary circulation via pulmonary veins into left atrium, left ventricle and is pumped into the lungs back again. Shunts between both circulations (e.g. PFO, VSD or patent arterial duct) enable blood mixing.

Due to the complex anatomy and variable clinical manifestations D-TGA may be divided into 4 groups: 1) simple D-TGA, 2) D-TGA with VSD, 3) D-TGA with VSD and LVOTO, as in our case and 4) D-TGA with VSD and PAH. LVOTO may be of a different type. The most common causes include pulmonary valve stenosis or sub-pulmonary stenosis due to conal septum posterior deviation, mitral valve anomaly with additional tissues or strands attached to the interventricular septum and TV anomaly with septal leaflet prolapse [2]. In our patient LVOTO was the result of multiple sub-pulmonary strands from the mitral valve, which did not prevent from the development of PAH.

The initial treatment of choice in D-TGA with hypoxia is PGE1 infusion providing the patency of arterial duct. In case of restrictive PFO balloon septostomy (BAS) should be performed. Wide interatrial communication provides left-to-right shunt of highly oxygenated blood into the systemic circulation.

Balloon septostomy was not performed in our patient probably due to adequate blood mixing through a large VSD. The correction – arterial switch (Jatene operation) with VSD closure was not possible because of coexisting sub-pulmonary stenosis and TV straddling. Therefore, only palliative procedures were considered. Surgical management strategies in D-TGA with VSD and signifi-

Table 2. Comparison of haemodynamic evaluations from subsequent catheterizations

Heart catheterization	age (years)	MPA (mmHg)	PVR (Wood units)	Rp/Rs	PAWP (mmHg)
1- before Senning operation	1,5	62	10	0,6	6
2 – before sildenafil therapy	8	69	25	0,8	4,5
3 – before bosentan therapy	17	51	21	0,9	8

MPA – main pulmonary artery; PVR – pulmonary vascular resistance; Rp/Rs – pulmonary / systemic resistance ratio; PAWP – pulmonary artery wedge pressure

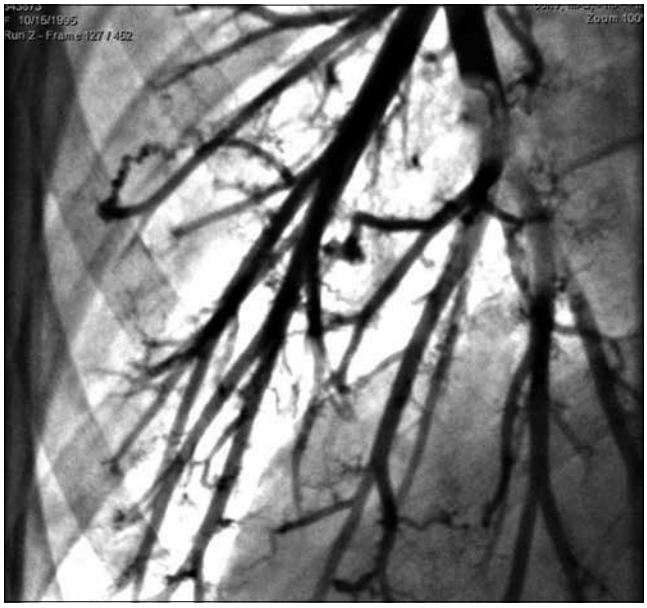

Figure 1. Right pulmonary artery angiography. Pathological tortuous tapering arteries resembling „dead tree"

cant LVOTO include B-TS while in mild LVOTO pulmonary artery banding would be the best option. Rastelli operation could be an option for correction in the future. However, TV straddling in our patient was a serious obstacle for VSD closure. In our case, only palliative procedures typical for treatment of single ventricle should have been considered such as hemi-Fontan or Glenn with eventual total cavopulmonary circulation (Fontan circulation).

In our patient mild LVOTO and the application of B-TS might have contributed to development of PAH. The presence of Eisenmenger syndrome affected therapeutic options. Most suitable solution was to connect pulmonary and systemic circulations at the level of atria (palliative atrial redirection – Senning operation) or arteries (palliative arterial switch) without closing interventricular communication.

PAH may develop in congenital heart defects with increased pulmonary flow (especially VSD, complete atrioventricular septal defect, aortopulmonary window) when high systemic pressure is transmitted to the pulmonary circulation [3]. D-TGA with VSD is a special condition predisposing to obstructive pulmonary vascular disease. Initially, PAH may be reversible, but as soon as arteriopathy develops pulmonary resistance increases, leading to Eisenmenger syndrome [4], as it happened in our patient.

The availability of pharmacological PAH – specific therapies has increased during the last decade [5]. In our patient, we initiated the treatment with sildenafil and eventually added bosentan. Upon reaching adolescence, the patient was referred to the PH adult unit, which cooperates with our pediatric department for further management and follow-up.

During the last decade the number of new patients with Eisenmenger syndrome has decreased due to early screening echocardiography as well as the progress in cardiac surgery and postoperative care.

Early detection of congenital heart defects and their efficient surgical treatment are essential components preventing from development of PAH and Eisenmenger syndrome in this group of young patients. Once PAH develops, a number of pharmaceutical agents may be used to ameliorate symptoms and improve prognosis.

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