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LETTER TO THE EDITOR

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Atrial flow regulator for severe drug resistant pulmonary arterial hypertension after congenital heart defect correction

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Pulmonary arterial hypertension (PAH) is a severe, progressive life-threatening condition. Inter-atrial shunt creation can lead to cardiac output increment at the expense of arterial blood desaturation [1, 2]. At long-term follow-up, however, spontaneous defect closure can occur [3, 4]. Lately, novel implantable atrial communication devices have been introduced, which may become a treatment option in end-stage PAH patients [5–7]. Atrial flow regulator (AFR) device delivered by Occlutech company is currently under European Community registration for compassionate use in patients with severe right ventricular (RV) failure due to pulmonary hypertension or left heart failure and under United States Food and Drug Administration's (FDA) emergency use guidance.

The present study describes the first AFR implantation in Poland in a 28-year-old male with drug resistant severe PAH, which developed 20 years subsequent to total correction of a congenital heart defect (double outlet right ventricle with ventricular septal defect) performed at the age of two. Cardiac magnetic resonance revealed no residual intracardiac shunt. No signs of PAH were evident until 2012 (age 22), when PAH-specific drug therapy was started. Since 2016 progressive deterioration started with one syncopal episode during exercise. Since March 2018 RV failure (World Health Organization [WHO] functional class IVa) with fluid retention (and up to 10 kg weight gain) had developed. The patient was on a waiting list for lung transplantation. In view of no other clinically meaningful treatment alternative, a decision to implant the AFR was made. Approval of the local ethics committee and informed consent from the patient was obtained. The patients' weight was 71 kg, height 172 cm, body surface area 1.85 m^2, blood pressure 110/80 mmHg, oxygen saturation (SpO_2) 94%, heart rate (sinus rhythm) 100 bpm. He had mild liver enlargement, as well as mild leg and abdominal edema. Right heart catheterization confirmed severe PAH with equalization of pulmonary and systemic pressure. The procedure was performed under general anesthesia, induced with etomidate, fentanyl and rocuronium for muscle relaxation and was maintained using volatile sevoflurane (0.7 to 1.1% administered in air/oxygen mixture). Muscle relaxation was reversed with sugammadex. Trans-septal puncture was performed under three-dimensional transoesophageal echocardiography guidance (**Suppl. Video 1**). Static septostomy was performed, followed by progressive balloon dilatation of atrial septum (balloon size 10–12 mm). Extra stiff wire was located in the left upper pulmonary vein. The AFR device (5 mm height/6 mm fenestration diameter) was inserted using 10 F introducing system and stabilized without complications. The

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Figure 1. Occlutech® atrial flow regulator.

AFR is a self-expandable double-disc nitinol wire mesh construction dedicated to create inter-atrial communication and allow blood flow across the interatrial septum (Fig. 1). The disc diameter ranges from 16 to 23 mm, fenestration from 4 to 10 mm, and connective waist between the two discs corresponding to atrial septum thickness from 2 to 10 mm. The device has very high flexibility and adaptability with unique braiding.

Six weeks after AFR implantation the patient's clinical status improved (WHO III) with no fluid retention on a reduced dose of diuretics. Signifi-

Table 1. Clinical, echocardiographic and hemodynamic parameters before and after atrial flow regulator (AFR) implantation.

	Before	Six weeks after AFR implantation
Functional class (WHO)	IVa	III
Six minute walking distance [m]	341	420
B-type natriuretic peptide [pg/mL]	250	90
Echocardiography		
RV/LV [mm]	59/39	54/46
RVEDvol [ml] 3D echo	265	250
RA area [cm^2]	28	25
RV EF [%] 3D echo	23	26
RVSP [mmHg]	105	98
LAESvol index [mL/m^2]	23	31
LVEDvol [mL]	88	93
LVEF [%]	52	56
VCI expiration/inspiration [mm]	20/14	17/11
Pericardial effusion	No	No
Right heart catheterization		
Arterial blood pressure (systolic/diastolic) [mmHg]	113/70	100/58
Cardiac index [$l/min/m^2$]	3.19	3.73
Central venous pressure [mmHg]	13	8
RA pressure (systolic/diastolic/mean) [mmHg]	17/10/13	10/7/8
RV pressure (systlic/diastolic/mean) [mmHg]	120/10/62	102/7/50
PA pressure (systlic/diastolic/mean) [mmHg]	128/85/100	100/64/76
Pulmonary capillary wedge pressure [mmHg]	9	11
Diastolic pressure gradient [mmHg]	76	53
Pulmonary vascular resistance [Wood units]	14.6	9.3
SaO_2 (PA/mixed/arterial) [%]	72/71/94	70/71/90
Drugs		
Furosemide *iv* [mg]	60	–
Torasemide *po* [mg]	200	100
Spironolactone *iv* [mg]	100	50
Chlortalidone *po* [mg]	50	50
Epoprostenol *iv* [mg/kg/min]	56	56
Sildenafil [mg]	60	60
Bosentan [mg]	250	250

3D echo — three-dimensional echocardiography; EF — ejection fraction; *iv* — intravenous; LAESvol — left atrial end-systolic volume; LV — left ventricle; LVEDvol — LV end-diastolic volume; PA — pulmonary artery; *po* — per os; RA — right atrial; RV — right ventricle; RVEDvol — RV end-diastolic volume; RVSP — right ventricular systolic pressure; SaO_2 — oxygen saturation; VCI — vena cava inferior; WHO — World Health Organization

cant amelioration of RV hemodynamics occurred, with decrement in pulmonary vascular resistance, increment of cardiac index, and slight reduction in SpO_2. Echocardiography revealed increase in left heart chambers dimensions with decrease in the right ones (Table 1).

The present researchers first experience with the Occlutech AFR device implementation was very promising. It provides a unique therapeutic option for decompensated RV failure in end-stage PAH patients. Future applications of the AFR device may be extended to other heart failure populations like severe diastolic left ventricular dysfunction with increased left atrial pressure.

Conflict of interest: None declared

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