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Short Report

3-year Follow-up Results of Abant Izzet Baysal University Training and Research Hospital Patients with Pulmonary Hypertension

Aslı KURTAR MANSIROĞLU[1]

[1] Abant İzzet Baysal University Training and Research Hospital, Bolu, Turkey

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Pulmonary hypertension (PH) is defined as the mean pulmonary artery pressure (mPAB) measured by right heart catheterization (RHC) 25 mmHg or higher at rest. Different hemodynamic PH definitions were made according to the combinations of pulmonary artery pressure measured by right heart catheterization, pulmonary artery end pressure (PCWP), cardiac output (CO), diastolic pressure gradient, and pulmonary vascular resistance (PVR).[1] The differential diagnosis of PH is at least as important as its treatment, as it is a condition that can cause many diseases. Symptoms and signs include shortness of breath, lightheadedness, syncope, tiredness, chest pain, swelling of the legs, poor appetite, chest pain, right-sided abdominal pain, palpitations, cyanosis and rarely non-productive cough, exercise-induced nausea, and vomiting. We herein present 3-year follow-up results of patients with PH.

In symptomatic patients with suspected PH, those with the possibility of PH echocardiographically were evaluated in our clinic with a multidisciplinary approach. Left heart diseases and lung diseases were excluded as a result of the evaluations of Chest Diseases, Internal medicine, and Rheumatology departments. RHC was performed in our clinic between January 2018 and January 2021 to confirm the diagnosis of group 1 PH in 30 patients whose chronic thromboembolic PH (CTEPH) diagnosis was excluded by negative ventilation-perfusion scintigraphy and/or pulmonary CT angiography. Considering the PH etiology of the patients; Eisenmenger syndrome due to congenital diseases in 2 patients; Group 4 PH in 7 patients; Group 2 PH in 3 patients; idiopathic PH diagnosis in 5 patients, and 5 patients were diagnosed with PH secondary to connective tissue diseases, and thus, pulmonary arterial hypertension (PAH)-specific treatment was initiated in a total of 22 patients (Table 1). In our clinic, vasoreactivity test is performed with adenosine. There was no vasoreactivity response in any of the patients who underwent the vasoreactivity test.

PAH was followed-up before specific treatment

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Address for Correspondence:
Aslı KURTAR MANSIROGLU
Abant İzzet Baysal University Training and Research Hospital, Bolu, Turkey
E-mail: dr.asli.kurtar@gmail.com

Table 1. Clinical classification of 22 patients with pulmonary hypertension

PH subtypes	Patient number
Group 1 PAH	Idiopathic PAH (n=5)
	Connective tissue disease (n=5)
	Congenital heart disease (n=2)
Group 2 PH	PH due to left heart disease (n=3)
Group 4 PH (CTEPH and other pulmonary artery obstructions)	CTEPH (n=3)
	Operated due to CTEPH and residue PH (n=2)
	Operated due to hydatid cyst and residue PH (n=1)
	Arteritis (n=1)

PAH: pulmonary arterial hypertension, PH: pulmonary hypertension, CTEPH: chronic thromboembolic pulmonary hypertension.

was initiated because of isolated postcapillary PH or the average PAP <25 mmHg in 8 patients according to RHC findings. When the treatments received by the patients were examined, 5 patients were referred to as single PH specific therapy; 14 patients dual combination therapy; 3 patients received triple combination therapy (Table 2). Since there was no vasodilator response in patients who underwent vasoreactivity test during right heart catheterization, no patient was followed up with high-dose calcium channel blocker. During their follow-up, 9 of our patients were followed up in the NHYA 1, 10 in the NHYA 2, 1 in the NHYA 3, and 2 in the NHYA 4 clinic, and one of these patients started subcutaneous prostanoid treatment in an external center; 1 patient in group 2 with precapillary and postcapillary PHT died in our hospital.

A diagnosis of PH requires clinical suspicion based on symptoms and physical examination. A series of examinations are required to determine the compliance of this suspicion with hemodynamic criteria, the etiology of the disease, and the functional and hemodynamic severity. These examinations should at least be interpreted by a multidisciplinary team of Cardiology, Rheumatology, Radiology, and Chest Diseases specialists. In our daily practice, it is necessary to raise awareness for this patient group, whose diagnosis is delayed, and the specialist referral centers should be determined and the referral chain should be operated.

Table 2. Pharmacological treatments given to patients

Treatment	Patient number	Side effect	Combination therapy	Drug monotherapy
Endothelin receptor antagonists				
Bosentan	8	2	7	1
Macitentan	5	-	4	1
Ambrisentan	3	-	3	-
Phosphodiesterase type 5 inhibitors				
Sildenafil	9	-	9	-
Tadalafil	5	-	5	-
Guanylate cyclase stimulators				
Riociguat	7	-	4	3
Prostacyclin analogues				
Iliprost (inhaled)	3	1	3	-
Subkutan PG	1	-	1	-
IP receptor agonists				
Selexipag	1	-	1	-

Conflict of Interests

Authors declare that there are none.

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