

OUTCOMES OF PULMONARY REHABILITATION IN SEVERE PULMONARY HYPERTENSION – CASE REPORT

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OUTCOMES OF PULMONARY REHABILITATION IN SEVERE PULMONARY HYPERTENSION - CASE PRESENTATION (Abstract): Chronic pulmonary artery hypertension (CPAH) is characterized by progressive increase in vascular resistance and pulmonary artery pressure, leading to right heart failure and death. Diagnosis algorithm is complex, and the underlying cause often remains undefined. We present the case of a 74-year-old male patient, whose diagnosis was disputed for several months between cardiologists and pulmonologists, being eventually diagnosed with severe pulmonary hypertension and hypoxic respiratory failure of chronic thromboembolic etiology intricate with diffuse interstitial lung disease. The new therapeutic approach with riociguat failed and the treatment was stopped, then the patient addressed Iasi Pulmonary Rehabilitation Clinic for an inpatient pulmonary rehabilitation program, comprising functional assessment, nutritional and psychological counseling, mild inspiratory muscle training sessions, low workload endurance exercise, followed by relaxation and appropriate breathing techniques. After two weeks, oxygen saturation, shortness of breath, fatigue, sleep, anxiety and depression symptoms had improved while 6-minute walk test distance did not change significantly. **Keywords:** PULMONARY HYPERTENSION, RESPIRATORY FAILURE, PULMONARY REHABILITATION.

Chronic pulmonary artery hypertension (CPAH) is a debilitating disease of the pulmonary vasculature, characterized by progressive increase in vascular resistance and pulmonary artery pressure, leading eventually to right heart failure and death. Diagnosis algorithm is complex, and the underlying cause often remains undefined.

The current treatment strategy for

CPAH patients includes three main steps, the initial approach including physical activity, supervised rehabilitation, and psychosocial support while pharmacological therapy is indicated only as a second step, according to the vaso-reactive or non-reactive type of CPAH and determinants of prognosis (1, 2, 3, 4). Pulmonary rehabilitation and supervised exercise, added to

pharmacological treatment options increase exercise capacity and quality of life of these patients, although there are small cohort trials proven otherwise (5).

CASE REPORT

We present the case of a 74-year-old male patient, with a complex medical history, who was admitted in November 2016 to the Pulmonary Rehabilitation Clinic of the Iasi Rehabilitation Hospital for an inpatient pulmonary rehabilitation program, his major complaints being severe dyspnea, fatigue, sleep disturbances, and an overall diminished quality of life and physical function while under long-term oxygen therapy at home. The patient was a former mild smoker (5PY) and not exposed to respiratory hazards.

Clinical examination revealed oral cyanosis, telangiectasis, splitted second heart sound left parasternal 2nd interspace, severe

regurgitation murmur in tricuspid area, BP = 100/60 mmHg, arrhythmic heart sounds (HR = 95/min), oxygen saturation SpO₂ = 88% in ambient air, with no sign of pulmonary or systemic congestion, no finger clubbing.

His past medical history has recorded a diagnosis of prostatic adenocarcinoma Gleason 5, eight years ago, under hormone therapy for seven years, discontinued by the urologist due to occurrence of progressive shortness of breath, fatigue, and weight loss (10 kg in 9 months), leading to repeated hospitalizations in pulmonology and cardiology services.

The case was disputed for several months (May-September 2016) between cardiologists and pulmonologists, being eventually diagnosed with severe pulmonary hypertension (precapillary type) and hypoxemic respiratory failure, following the recommended algorithm (1) (tab. I).

TABLE I
Diagnostic approach and results

Investigation	Results
Echocardiography	Dilated right chambers, PAPs estimated = 86 mmHg; mild systolic dysfunction RV, moderate functional and pulmonary tricuspid regurgitation, normal LV, mild to moderate mitral regurgitation, without hemodynamic significance
ECG, Holter	Atrial flutter with variable conduction
Bodyplethysmography, Spirometry & DLCO	FVC = 120%, FEV1=123%, FEV1/FVC = 78,47%, MEF50 = 130,5%, TLC = 121,8%, RV = 134,5% (normal values), DLCO = 42,7% from predicted values (fig.1)
Chest X-ray	Bilateral diffuse interstitial opacities
CT of the chest	Bilateral ground-glass opacities, fine subpleural reticular opacities, several mediastinal adenopathies, and three nodular peribronchial opacities
Lung perfusion scan	Multiple segmental perfusion deficits (moderate-severe) (fig. 2)
Right heart catheterization	PAP = 70/27/42 mmHg, PCP = 9/5/3 mmHg, PVR 6,34uW, CI = 3.7l/min/m ²
Immunology	c-ANCA, p-ANCA, ANA, anti-La/SS-B, anti-Ro/SS-A, anti-SM, anti SCL70, anti-RNP = negatives
Tumor markers	CEA, CEA 19-9, NSE, Cyfra 21-1, PSA (under hormone therapy) within normal range
Others	BNP = 207pg/ml; 400 pg/ml

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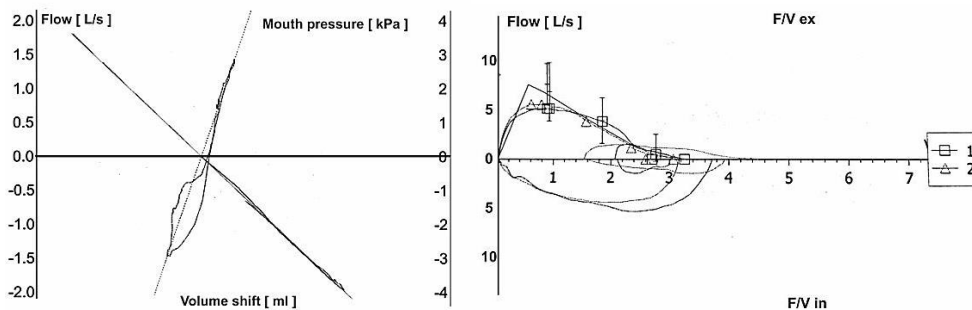


Fig. 1. Body plethysmography and flow-volume loop showing normal values for pulmonary volumes and flows

Etiology of CPAH was supposed to be chronic thromboembolism possibly intricate with diffuse interstitial lung disease (idiopathic pulmonary). Due to the diagnosis of post embolic pulmonary hypertension distal type, associated with inoperability, in June 2016 the patient has started treatment with pulmonary vasodilator riociguat 1 mg

tid for 2 weeks, then 1.5 mg tid but after one week he has developed paroxysmal nocturnal dyspnea, leading to reassessment of treatment and etiology of PAH, the pulmonary interstitial lung disease being now considered the leading culprit and only diuretic and anticoagulant treatment was continued (August 2016).

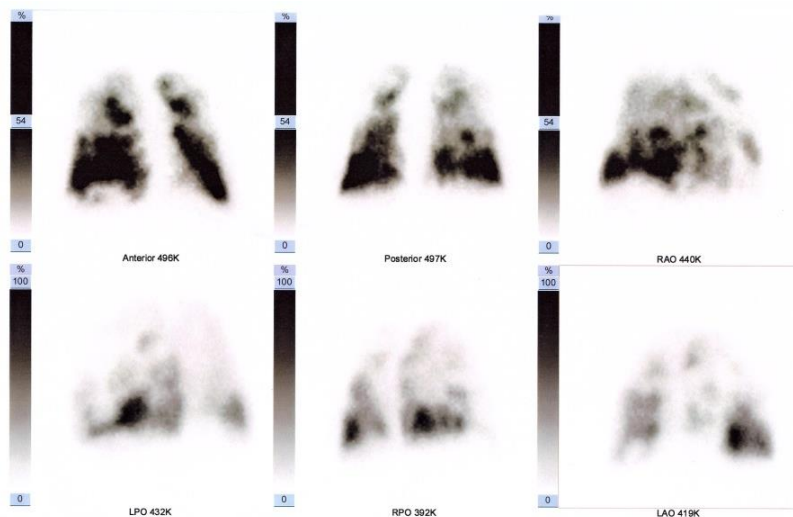


Fig. 2. Multiple segmental perfusion deficits (moderate-severe) in lung perfusion scan (chronic thromboembolism intricate with diffuse interstitial lung disease)

During admission in Iasi Respiratory Rehabilitation Clinic, the patient has followed a comprehensive respiratory rehabil-

itation program, comprising functional assessment, nutritional and psychological counseling, mild inspiratory muscle train-

ing sessions, low workload endurance exercise, followed by relaxation and appropriate breathing techniques (6, 7, 8, 9). After two weeks, oxygen saturation, shortness of breath (assessed through Borg and mMRC scales), fatigue, sleep, anxiety and depression symptoms had improved while 6-minute walk test (6MWT) distance had no significant improvement.

DISCUSSION

This case is particular through diagnosis difficulties of CPAH and as a consequence etiologic treatment had not the anticipated beneficial results. Prognosis indices are represented by clinical evidence of right ventricle failure, the progression of symptoms, WHO class and 6MWT distance (10, 11). Unsolved thromboembolic obstruction in the pulmonary arteries leads to group 4 pulmonary hypertension, according to World Health Organization. Pulmonary endarterectomy is the treatment of choice for chronic thromboembolic pulmonary hypertension as potentially curative therapy, but about one-third of patients, including our case, are not suitable candidates for the surgery, either due to distal and inaccessible nature of the lesions or comorbid conditions, requiring pharmacologic treatment options (12).

Riociguat is a member of the therapeutic class of soluble guanylate cyclase stimulators, associated with significant improvements in 6-minute walk distance, pulmonary vascular resistance, and time to clinical worsening (13, 14), being the only agent approved for treating both chronic thromboembolic hypertension and pulmonary arterial hypertension. The novel mechanism of riociguat lies in its ability to restore the homeostatic and therapeutic effects of nitric oxide that are diminished because of phenotypic alterations associat-

ed with pulmonary hypertension (14) but in the presented case it seemed to have no positive results, questioning the vascular etiology of CPAH.

On the other hand, several small size randomized controlled trials reported that trained CPAH patients have decreased fatigue and dyspnea, higher level of physical exercise, improved distance to 6MWT, quality of life, and cardiorespiratory function (15, 16) but the optimal method of exercise, intensity, and duration of the training are still under debate. Because of the positive outcomes, pulmonary rehabilitation was introduced in the recent guidelines of treatment and management of PAH. A low workload endurance exercise and weightlifting training under close medical supervision, thorough patient education regarding his disease, nutrition, and the safe level of exercise were recommended (1, 17).

In the clinical case discussed above, the initiation of pulmonary rehabilitation was relatively late, and outcomes were negatively affected by the unclear cause of the PAH, rapid evolution of the symptoms, and the negative prognosis of the main underlying disease.

Still, features demonstrating an improved quality of life as the diminished severity of dyspnea, improved sleep pattern, and psychiatric symptoms of depression and anxiety were displayed after two weeks of a comprehensive rehabilitation program.

CONCLUSIONS

Our study highlights that the core issue of the multidisciplinary approach to complex clinical cases of pulmonary hypertension remains the close cooperation between cardiologists and experienced pulmonary rehabilitation experts, to improve outcomes and ultimately, survival of these severely impaired patients.

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