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Rhythm Disorders in Pulmonary Arterial Hypertension (PAH) - What Therapeutic Options are Available ?

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Abstract

Rhythm disorders in PAH are rare. In practice, they pose a therapeutic management challenge. Right ventricular dysfunction associated with hypoxia influences the prescription of antiarrhythmics; some of them are even contraindicated. We report three cases of idiopathic PAH complicated by atrial flutter (2 cases) and atrial fibrillation (1 case). The patients experienced right heart failure after a stabilization phase with specific vasodilators (bosentan, sildenafil, iloprost). They were treated with oral amiodarone, alongside treatment for right heart failure. Potassium levels were normal. Beta-blockers were not prescribed due to severe right ventricular dysfunction with arterial hypotension. Unfortunately, two patients developed hyperthyroidism secondary to amiodarone. Through these observations, we will discuss the different therapeutic options in these situations, referencing the literature.

Keywords: Rhythm disorders, PAH, right ventricular dysfunction, arterial hypotension, hyperthyroidism, therapeutic options.

Introduction

Pulmonary arterial hypertension is a rare and severe condition. It has significant impacts on the right-sided chambers of the heart, leading to medium and long-term dilation of the right atrium and right ventricle, with severe right ventricular dysfunction. The increase in pulmonary vascular resistance results in chronic right ventricular overload(1), leading to increased right atrial pressure, progressive atrial dilation, and electrophysiological remodeling that creates a pro-arrhythmogenic substrate, which can lead to supraventricular arrhythmias (SVTs) (2,3). The prevalence of SVTs in patients with PAH is between 10 and 25% (4, 1.5–7). The loss of atrial systole, combined with the altered hemodynamics related to PAH, promotes their occurrence and is associated with clinical worsening, characterized by a decline in New York Heart Association (NYHA) functional class, reduced performance in the 6-minute walk test, and poor prognosis (4,8). Survival is particularly reduced in these patients (4,9).

The mechanisms behind rhythm disorders are diverse, including increased susceptibility to afterdepolarizations, enhanced heterogeneity of repolarization, and upregulation of fibrosis. These mechanisms lead to the initiation and maintenance of reentrant circuits in the right heart, resulting in atrial rhythm disorders. Other anomalies can occur, such as sinus bradycardia, sinus tachycardia, first-degree atrioventricular block, right bundle branch block, and ventricular arrhythmias. Rajdev et al. (10) analyzed arrhythmias in PAH and concluded that ventricular tachycardia is less frequent than bradycardia, which is a concerning sign. This suggests that PAH leads to cardiac conduction system disease. Other studies have

shown that atrial fibrillation (AF) and atrial flutter induced by PAH are the most common arrhythmias. They are a cause of disease decompensation and increase morbidity and mortality (11). The loss of atrial systole, atrioventricular desynchronization, and changes in heart rate are even more harmful in the presence of right ventricular dysfunction.

Restoring sinus rhythm can improve hemodynamic status and the six-minute walk test. In practice, therapeutic options are limited by the negative inotropic effect of antiarrhythmics and the proarrhythmogenic effects on right ventricular dysfunction.

Observation

We report the case of three patients who have been under our care and have been followed for PAH for over five years, treated with bosentan, sildenafil, and inhaled iloprost. We had two cases of atrial flutter and one case of atrial fibrillation (Figures 1 and 2). They experienced right heart failure after a stabilization phase with specific vasodilators. They were treated with oral amiodarone, along with treatment for right heart failure. Potassium levels were normal. Beta-blockers were not prescribed due to severe right ventricular dysfunction with arterial hypotension (Figures 3 and 4). Unfortunately, two patients developed hyperthyroidism secondary to amiodarone.

Discussion

According to data from the literature, SVTs secondary to PAH typically manifest on average 3.5 years after the initial PAH diagnosis (8,18). The incidence depends on associated comorbidities and the progression of the disease; these are often symptomatic arrhythmias that are more easily detected because they cause a deterioration in functional and hemodynamic status (4). Regarding treatment, our patients poorly tolerated medical therapy (digoxin, amiodarone, bisoprolol). They were treated with external electrical cardioversion (EEC). The therapeutic management of supraventricular arrhythmias in the context of PAH has not been the subject of specific recommendations, and there has been no specific research on the effectiveness of rate control or rhythm control strategies. Small series have suggested that restoring sinus rhythm was associated with improved functional class and survival (4, 9,12). Other therapeutic options provide good results for these arrhythmias: electrical cardioversion (EEC) is effective but short-term, and the risk of recurrence is significant, especially when the right atrium is dilated (13). When comparing the outcomes of ablation against EEC and antiarrhythmic therapy, ablation shows better results in terms of recurrence, improvement in quality of life, increased walking distance, and reduced right heart failure (14).

Conclusion

Rhythm disorders induced by PAH are proportional to the severity of the disease and its impact on the right-sided heart chambers. The use of antiarrhythmic medications is limited due to their negative inotropic effects on a severely dysfunctional right ventricle. Reducing atrial fibrillation or atrial flutter via external electrical cardioversion (EEC) is possible, but ablation is a better alternative to prevent recurrences and improve patient quality of life.

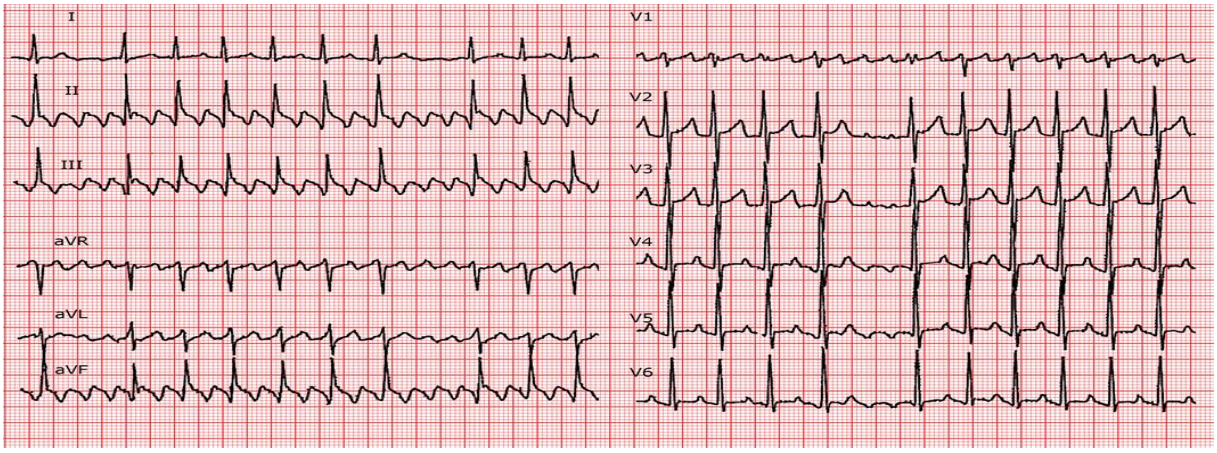

Figure 1 : Atrial Flutter

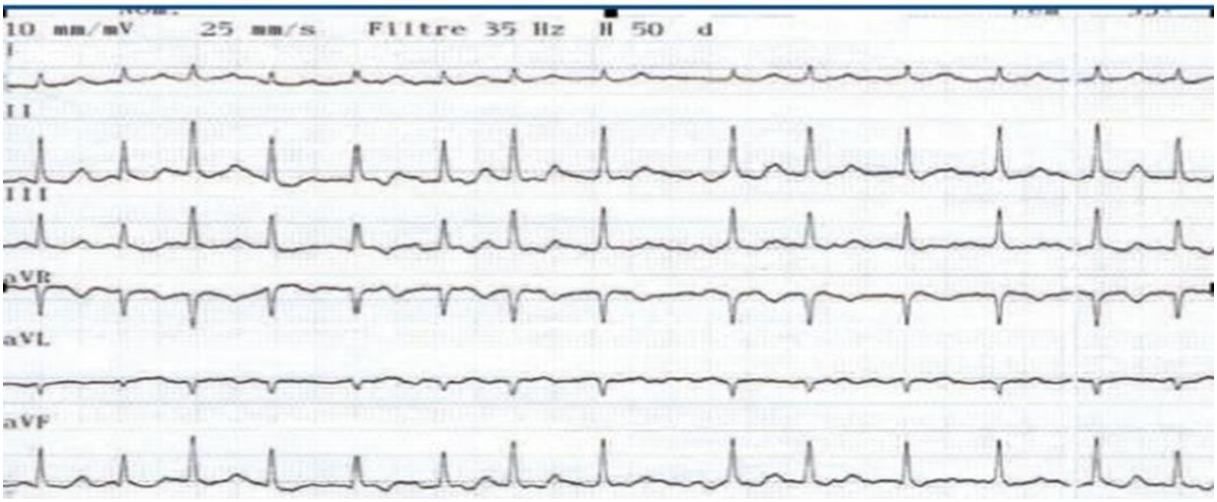

Figure 2 : Atrial Fibrillation

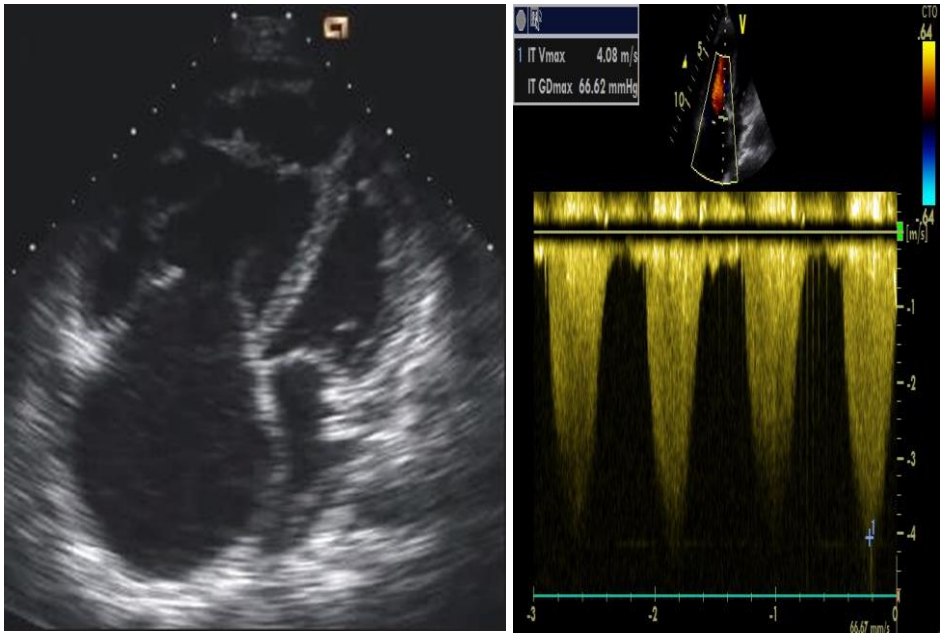

Figure 3: Dilation of Right-Sided Chambers; PAPs Calculated on Tricuspid Regurgitation Flow

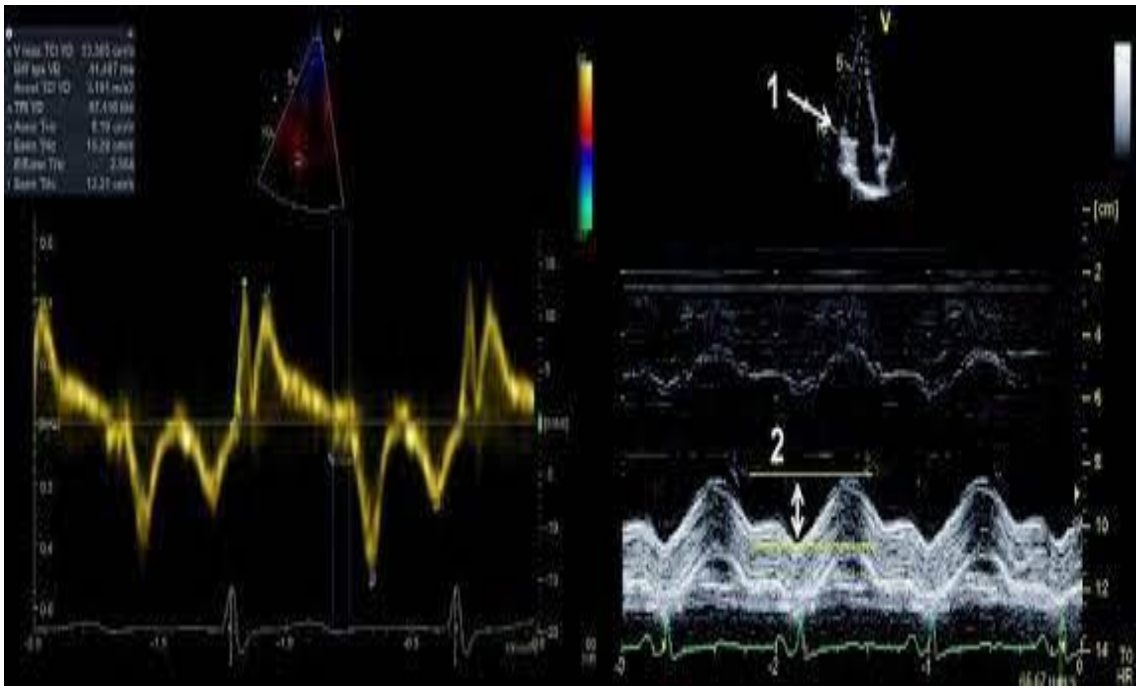

Figure 4: Assessment of Right Ventricular Function: Tissue Doppler (Calculation of S (RV) and TM Calculation of TAPSE)

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