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Review Article

Acute Management of Right Ventricular Dysfunction in Pulmonary Hypertension Associated with Interstitial Lung Disease

Evan Hindoro[1], Fariz Nurwidya[2]*, Vidya Gilang Rejeki[1], Efriadi Ismail[2]

[1]Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Indonesia
[2]Department of Pulmonology and Respiratory Medicine, Faculty of Medicine, Universitas Indonesia

Abstract

Pulmonary hypertension (PH) associated with interstitial lung disease is classified as Group 3 PH. This condition is characterized by poor survival and reduced quality of life, while scientific evidence supporting specific therapies for this subgroup remains limited. Moreover, right ventricular (RV) dysfunction is often found to be more severe in Group 3 PH, despite pulmonary vascular resistance being generally lower than in Group 1 PH. Various precipitating factors, such as acute hypoxemia, infection, arrhythmia, and exacerbation of underlying lung disease, may worsen pulmonary arterial pressure and trigger acute right ventricular failure. Current therapeutic strategies focus on managing these precipitating factors, controlling arrhythmias, optimizing ventilation and hemodynamics through appropriate preload optimization, inotropic and vasopressor support, and the selective use of pulmonary vasodilators.

Keywords: Acute Right Ventricular Dysfunction, Interstitial Lung Disease, Pulmonary Hypertension, Right Ventricle

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***Corresponding author**: fariz.nurwidya@ui.ac.id

Introduction

Interstitial lung disease (ILD) is a heterogeneous group of lung disorders involving damage to the pulmonary parenchyma with similar clinical, radiographic, and pathological characteristics. This disease is characterized by infiltration of inflammatory cells into the alveolar walls and involvement of the pulmonary interstitial space, leading to increased alveolar permeability and impairment of ventilation and perfusion. These processes are followed by fibroblast proliferation and excessive collagen deposition as a response to injury caused by inflammation and reparative processes of the epithelial and endothelial surfaces. [1–3] Multiorgan involvement in ILD is part of the chronic inflammatory process of the disease; however, involvement of the cardiovascular system is a consequence of ILD-associated pulmonary hypertension. [3]

Pulmonary hypertension associated with chronic lung disease is classified as Group 3 PH according to the classification system of the World Symposium on Pulmonary Hypertension (WSPH). Group 3 PH requires special categorization due to limited evidence supporting PH-specific therapies in this particular group.[4] Recent data indicate that Group 3 PH is the second most common type after Group 2 PH and is associated with poorer survival, reduced quality of life, and substantial medical costs.[4–5] In addition, Prins KW et al. found that right ventricular systolic dysfunction, as measured by fractional area change (FAC), was more severe in patients with Group 3 PH compared with those with Group 1 PH, despite lower mPAP and pulmonary vascular resistance.[5]

Right Ventricular Physiology

Embryological anatomical development results in functional and structural differences between the right ventricle and the left ventricle. The right ventricle has a crescent-shaped structure with extensive trabeculation and a thinner wall compared with the left ventricle, which has a thicker myocardium designed to withstand high systemic pressure but not volume overload. In contrast, due to its crescent-shaped geometry and prominent trabeculation, the right ventricle is well adapted to handle high-volume loads but not high-pressure loads, as pulmonary pressure is approximately less than one-tenth of systemic pressure in healthy individuals. [6–7] (Table 1).

Indeed, changes in left ventricular afterload do not appear to result in a significant reduction in stroke volume, in contrast to the right ventricle, which is highly sensitive to changes in right ventricular afterload (in the form of

pulmonary arterial systolic pressure, mean pulmonary arterial pressure [mPAP], and pulmonary vascular resistance), leading to a marked decrease in stroke volume.[8] Coronary perfusion of the right ventricle occurs during both systolic and diastolic phases, whereas in the left ventricle it occurs only during diastole; therefore, under conditions of increased right ventricular pressure load, elevated intramural pressure may lead to myocardial ischemia by reducing perfusion pressure to the right ventricle. (Figure 2).

Table 1. Physiological and anatomical differences between the right and left ventricles[7]

Parameter	Right Ventricle	Left Ventricle
Shape	Crescent-shaped	Elliptical
Structure	Two myocardial fiber layers	Three myocardial fiber layers
Wall Thickness (mm)	1–5	8–10
Circulation	Low pressure, low resistance	High pressure, high resistance
Stroke Volume (mL)	70–90	70–90
Ejection Fraction (%)	65	70–80
Ventricular Pressure (diastolic; mmHg)	0–8	4–12
Ventricular Pressure (systolic; mmHg)	15–30	90–140
***Afterload* (dyne·s/cm^5)**	Pulmonary vascular resistance < 250	Systemic vascular resistance 800–1,200
Adaptation to disease	Tolerant to increased preload	Tolerant to increased afterload

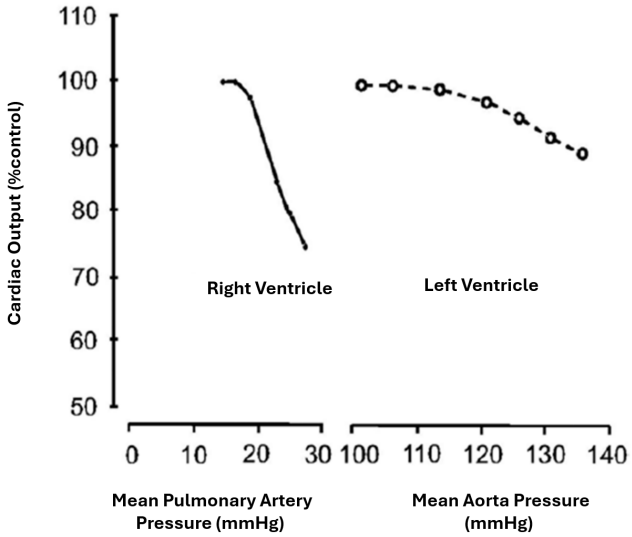

Figure 1. Differential stroke volume responses of the right and left ventricles to afterload elevation[7]

Right Ventricular Dysfunction in Group 3 Pulmonary Hypertension

Right ventricular dysfunction is a strong predictor of clinical outcomes in almost all pulmonary vascular diseases. Parameters of right ventricular systolic function are obtained through echocardiographic assessment, including tricuspid annular plane systolic excursion (TAPSE) and fractional area change (FAC) measured by two-dimensional (2D) transthoracic echocardiography, as well as right ventricular ejection fraction (RVEF) assessed by three-dimensional (3D) transthoracic echocardiography.[9] Right ventricular systolic function is highly sensitive to changes in afterload due to structural differences between the left and right ventricles, as previously described.[7]

In pulmonary disorders, ventilation–perfusion mismatch resulting from alveolar damage in chronic obstructive pulmonary disease (COPD), chronic hypoventilation in obesity hypoventilation syndrome, pulmonary arterial vasoconstriction due to chronic hypoxia at high altitude, and pulmonary capillary damage and fibrotic processes are

vasoconstriction due to chronic hypoxia at high altitude, and pulmonary capillary damage and fibrotic processes are responsible for increased right ventricular afterload.[10–11] interdependence occurs. With changes in right ventricular afterload, right ventricular dilatation leads to redistribution of wall stress throughout the right ventricular wall, causing displacement of the interventricular septum toward the left ventricle. This results in impaired left ventricular filling and ultimately a reduction in cardiac output.

It has been reported that interventricular septal contraction contributes approximately 30–40% of total right ventricular cardiac output; this contribution is lost in conditions of right ventricular dilatation that displace the interventricular septum toward the left ventricle.[7] In Group 3 pulmonary hypertension, chronic right ventricular dysfunction may undergo acute clinical deterioration, characterized by failure of oxygenation and perfusion. This deterioration may be triggered by precipitating factors such as infection, pregnancy, anemia, fever, non-adherence to treatment, fluid retention, arrhythmias, venous thromboembolism, procedural stress, and acute exacerbation of the underlying disease. In several reports, this condition is also referred to as acute cor pulmonale. [7,8,10-12]

Because the right and left ventricles are enclosed within the same pericardial space, a phenomenon known as ventricular

Mechanisms of Right Ventricular Functional Deterioration in Group 3 Pulmonary Hypertension

In acute cor pulmonale, identification of precipitating factors is the primary focus to enable prompt management; however, in many cases, the triggers of this decompensated phase cannot be identified, allowing disease progression to continue and leading to multiorgan failure. Precipitating factors increase myocardial oxygen demand in right ventricular myocytes and worsen hypoxic conditions in ILD, resulting in increased pulmonary vascular resistance. Increased right ventricular afterload leads to right ventricular ischemia due to reduced right coronary perfusion, ultimately culminating in right ventricular failure.

Right ventricular dilatation causes leftward displacement of the interventricular septum, resulting in reduced left ventricular cardiac output due to impaired left ventricular diastolic filling. Furthermore, a mismatch between cardiac output and cellular oxygen demand induces a shift in cellular metabolism from aerobic to anaerobic pathways. This process leads to lactate production and lactic acidosis, which further increases pulmonary vascular resistance, thereby creating a self-perpetuating vicious cycle.[10,11] (Figure 3)

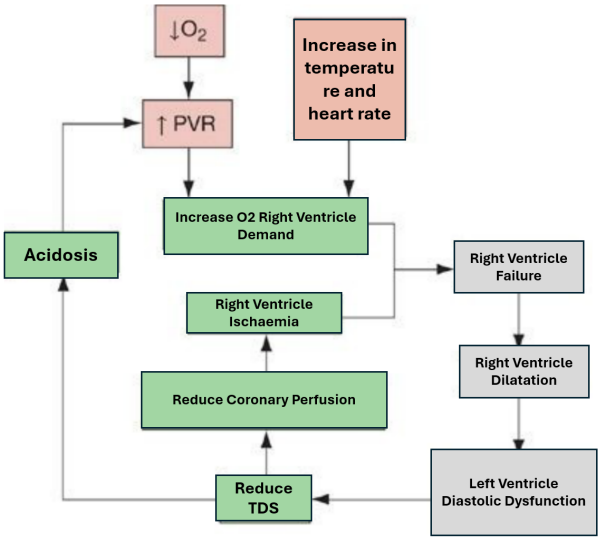

Figure 2. Mechanisms of acute right ventricular dysfunction in pulmonary hypertension due to interstitial lung disease.[11]

Management of Precipitating Factors

The primary management goals in unstable ILD-associated pulmonary hypertension complicated by acute cor pulmonale are to reduce oxygen consumption (VO_2) and to improve oxygen delivery (DO_2) to tissues and organs. Supportive therapy aims to restore adequate tissue perfusion and cellular oxygenation through rational optimization of preload with intravenous fluid administration, reduction of right ventricular afterload, right ventricular support, and optimization of oxygenation, including the use of mechanical ventilation when necessary. However, identification and treatment of precipitating factors remain the top priority.[7,10-11]

In patients with ILD due to idiopathic pulmonary fibrosis (IPF), acute exacerbations may be triggered by factors such as microaspiration and infection, leading to an extension of acute lung injury, clinical deterioration, increased oxygen requirements, and worsening hypoxemia. When infection is suspected as the precipitating factor, management includes broad-spectrum antimicrobial

therapy in combination with macrolides. Point-of-care ultrasound (POCUS)-guided echocardiographic assessment is recommended to exclude acute pulmonary embolism and acute myocardial infarction (AMI).[10-11]

Arrhythmia Management

In patients with ILD-associated pulmonary hypertension as well as those with chronic obstructive pulmonary disease (COPD), the most common tachyarrhythmias are atrial fibrillation and atrial flutter. Both arrhythmias worsen hemodynamic status due to the loss of atrial contraction, which contributes approximately 30% to stroke volume. Significant tachyarrhythmias and bradyarrhythmias reduce cardiac output, further impair organ perfusion, and exacerbate acidosis resulting from hypoxemia and increased oxygen demand.

Management according to advanced cardiac life support (ACLS) guidelines is recommended to maintain or restore sinus rhythm, using various strategies including electrical cardioversion.[7,12] Sinus rhythm preserves atrioventricular synchrony, which is essential for maintaining right ventricular diastolic filling and cardiac output in patients with pulmonary hypertension. In cases of significant bradyarrhythmia, temporary or permanent dual-chamber pacemaker implantation is recommended to preserve atrioventricular synchrony.[7,12] (**Figure 3**).

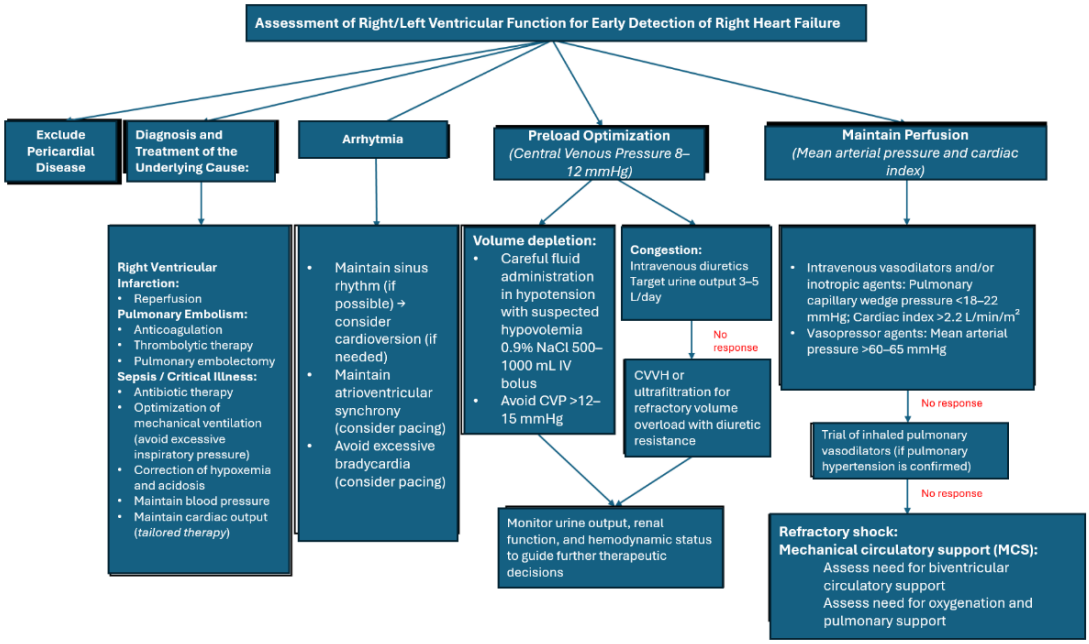

Figure 3. Management of acute right ventricular failure[7]

Ventilatory Management

Oxygenation and ventilation in acute cor pulmonale associated with Group 3 pulmonary hypertension present a particular clinical challenge. Three major factors exacerbate right ventricular dysfunction: hypercapnia, hypoxia, and acidosis. These factors can worsen pulmonary arterial vasoconstriction, even though permanent vascular remodeling and pulmonary arterial narrowing have already occurred in ILD.[10] The use of positive-pressure ventilation, including noninvasive ventilation and even invasive mechanical ventilation, is not uncommon. Application of positive end-expiratory pressure (PEEP) increases intrathoracic pressure and pulmonary vascular resistance. This may reduce right ventricular preload in hypovolemic conditions; however, PEEP also reduces left ventricular afterload and myocardial oxygen demand.[13] **(Figure 4)**

In conditions of right heart failure or preload-dependent states such as cardiac tamponade, the use of PEEP at 5–15 cmH_2O may worsen right ventricular cardiac output, leading to a low cardiac output state (LCOS). Therefore, a low PEEP strategy of 3–5 cmH_2O, or the lowest possible level to achieve a target oxygen saturation >92%, is recommended.

Conversely, in afterload-dependent conditions such as left heart failure without concomitant right ventricular dysfunction, PEEP is recommended to be initiated at 5–10 cmH_2O when pulmonary capillary wedge pressure is not elevated (12–15 mmHg) after adequate optimization of fluid status. When pulmonary capillary wedge pressure is elevated (>15 mmHg), as in acute pulmonary edema, PEEP may be initiated at 8–10 cmH_2O to reduce left ventricular afterload, promote alveolar recruitment, and improve oxygenation.[13]

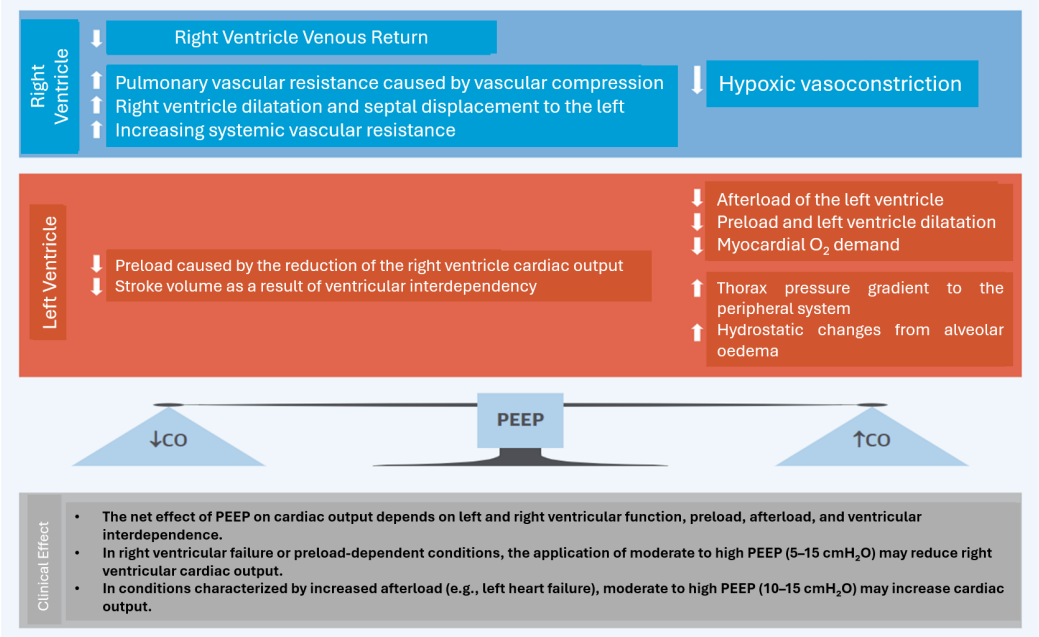

Figure 4. Physiological effects of PEEP on cardiac output and left and right ventricular function[13]

Hemodynamic Management

Preload optimization

In hypervolemic states, fluid administration worsens hemodynamics in right heart failure due to excessive right ventricular distension, which displaces the left ventricle and impairs left ventricular diastolic filling, resulting in reduced left ventricular cardiac output. This condition also exacerbates tricuspid regurgitation. Intravenous diuretic therapy is therefore the preferred approach in right ventricular dysfunction with signs of venous congestion to achieve euvolemia.

The target urine output is 3–5 liters per day, in accordance with the intravenous diuretic protocol using furosemide in the Cardiorenal Rescue Study in Acute Decompensated Heart Failure (CARRESS-HF). If urine output does not reach the target within the first 24 hours, furosemide dose titration and combination therapy should be performed based on the urine response to the initial dose. At 48 hours, if urine output remains below target, low-dose dopamine at renodilatory doses (2 µg/kg/min) may be administered.

After 72–96 hours, if urine output still fails to reach the target of 3–5 liters per day in the presence of persistent congestion and right heart failure, renal replacement therapy is recommended.[7] (Table 2 and Table 3)

Conversely, in hypovolemic patients who are fluid-responsive, intravenous fluid administration is recommended with a target central venous pressure (CVP) of 8–12 mmHg. Fluid administration should be guided by hemodynamic echocardiography and only performed in fluid-responsive patients. Fluid resuscitation is not recommended when CVP exceeds 12–15 mmHg or when signs of right ventricular volume overload are present, such as interventricular septal flattening during diastole; in such cases, a decongestive strategy is preferred.[7,14]

Inotropes and Vasopressors

The administration of inotropes and vasopressors in acute right ventricular dysfunction aims to restore organ perfusion, improve tissue oxygenation, and correct acidosis by enhancing right ventricular contractility and by converting unstressed volume within the splanchnic circulation—resulting from loss of peripheral vascular tone—into effective circulating volume (stressed volume).[14]

Acute right ventricular dysfunction is frequently accompanied by signs of impaired tissue perfusion or cardiogenic shock with a “right ventricle–dominant shock” or “biventricular shock” profile. Therefore, therapeutic targets include maintaining a mean arterial pressure (MAP) > 60–65 mmHg and a cardiac index (CI) > 2.2 L/min/m². These targets are not only intended to preserve organ perfusion but also to improve right ventricular perfusion and restore interventricular septal function to generate adequate right ventricular cardiac output by increasing left ventricular afterload.[7,14–16] (Figure 5)

Table 2. Diuretic dosing protocol used in the CARESS-HF study[7]

Assessment	Recommendation
Urine output targets ***(assessed daily from baseline up to 96 hours)***	□ Urine output > 5 L/day → reduce the current diuretic regimen if desired □ Urine output 3–5 L/day → continue the current diuretic regimen □ Urine output < 3 L/day → refer to the diuretic grid table
24-hour assessment	□ Urine output recommendations as above □ Proceed to the next step in the table if urine output < 3 L/day
48-hour assessment	□ Urine output recommendations as above □ Proceed to the next step in the table if urine output < 3 L/day □ Consider dopamine or dobutamine at 2 μg·kg⁻¹·min⁻¹ if systolic blood pressure (SBP) < 110 mmHg and ejection fraction < 40% or if right ventricular systolic dysfunction is present □ Consider nitroglycerin or nesiritide if SBP > 120 mmHg (regardless of ejection fraction) and severe symptoms are present
72–96-hour assessment	□ Urine output recommendations as above □ Proceed to the next step in the grid table if urine output < 3 L/day □ Consider dopamine or dobutamine at 2 μg·kg⁻¹·min⁻¹ if SBP < 110 mmHg and ejection fraction < 40% or if right ventricular systolic dysfunction is present □ Consider nitroglycerin or nesiritide if SBP > 120 mmHg (regardless of ejection fraction) and severe symptoms are present □ Consider hemodynamically guided intravenous therapy, LVAD support, dialysis, or ultrafiltration if indicated

Table 3. Diuretic Dosing Table (Diuretic Grid) (*Diuretic grid)*

Current Dose	Daily Loop Diuretic Dose	Recommended Thiazide
<80 mg	40 mg IV bolus + 5 mg/hour	No
81–160 mg	80 mg IV bolus + 10 mg/hour	Metolazone 5 mg once daily
161–240 mg	80 mg IV bolus + 20 mg/hour	Metolazone 5 mg twice daily
>240 mg	80 mg IV bolus + 30 mg/hour	Metolazone 5 mg twice daily

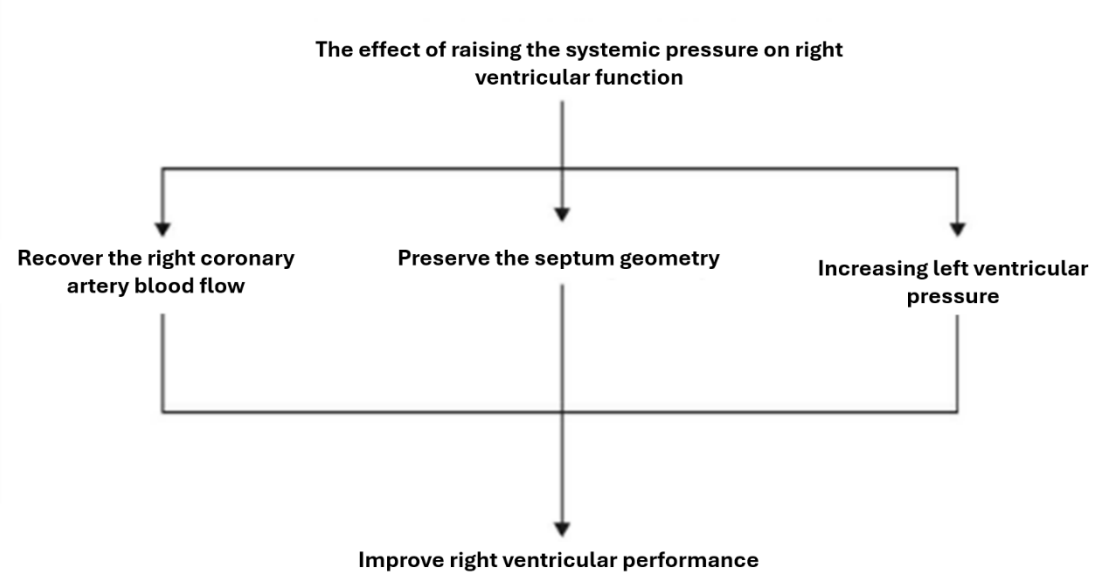

Figure 5. Effect of systemic pressure elevation on right ventricular systolic function

Milrinone is considered the inotrope of choice in right ventricular dysfunction associated with pulmonary hypertension because it possesses properties that reduce pulmonary vascular resistance while enhancing right ventricular contractility. In addition, in patients with a history of beta-blocker use, milrinone is often preferred. A limitation of milrinone is its hypotensive effect; therefore, vasopressor agents are frequently co-administered to maintain mean arterial pressure (MAP). Dobutamine is commonly used in daily clinical practice due to its minimal hypotensive effect and shorter onset of action compared with milrinone. However, the DOREMI trial found no difference in clinical outcomes between these two agents, even in patients with right ventricular dysfunction. The SOAP II trial demonstrated increased mortality with dopamine compared with norepinephrine; however, this study predominantly included patients with cardiogenic shock due to left ventricular dysfunction. [7,14–16]

Pulmonary Arterial Vasodilators

Increased right ventricular afterload can be addressed by administering pulmonary arterial vasodilators to reduce pulmonary vascular resistance. There are three major pathways targeted by pulmonary arterial vasodilator therapies: prostacyclin analogues, phosphodiesterase type 5 (PDE-5) inhibitors, and endothelin receptor antagonists. The use of pulmonary arterial vasodilators has been shown to improve clinical outcomes and increase six-minute walk distance; however, these benefits have been demonstrated only in specific populations, namely Group 1 pulmonary hypertension (idiopathic pulmonary arterial hypertension, IPAH) and Group 4 pulmonary hypertension (chronic thromboembolic pulmonary hypertension, CTEPH).[7]

In pulmonary hypertension associated with ILD, study findings have been conflicting. The INCREASE trial demonstrated that patients with ILD-associated pulmonary hypertension confirmed by chest computed tomography (CT) and right heart catheterization who received inhaled treprostinil experienced an improvement in six-minute walk distance of approximately +31.1 meters (95% CI 16.9–45.4, $p < 0.001$) and a 15% reduction in NT-proBNP from baseline.[17] Nevertheless, the ARTEMIS-IPF and ARTEMIS-PH trials showed that ambrisentan was discontinued early due to increased rates of acute IPF exacerbations, hospitalizations, and mortality, leading to a Class III recommendation in the 2022 European Society of Cardiology (ESC) pulmonary hypertension guidelines.[4]

Worsening ventilation–perfusion mismatch may exacerbate hypoxemia as a result of nonselective vasodilation in fibrotic lungs and may potentially cause pulmonary edema due to increased pulmonary blood flow in fibrotic lung tissue with high permeability.

In acute right ventricular dysfunction due to pulmonary hypertension, the use of pulmonary vasodilators such as inhaled nitric oxide (NO) can rapidly reduce right ventricular afterload while preserving systemic perfusion, as NO is rapidly inactivated by hemoglobin within pulmonary capillaries, resulting in selective pulmonary vasodilation. Its effects are limited to ventilated lung regions, thereby reducing pulmonary vascular resistance and improving oxygenation without increasing intrapulmonary shunt. This approach has been shown to improve cardiac output and oxygenation in intensive care unit patients with acute right ventricular failure.[7,14–16]

Conclusion

Pulmonary hypertension associated with interstitial lung disease is classified as Group 3 pulmonary hypertension, for which current therapeutic options remain limited and are associated with high mortality and comorbidity rates. Right ventricular dysfunction is a strong predictor of poor clinical outcomes in pulmonary hypertension. Acute right ventricular dysfunction may occur in response to various precipitating factors, particularly infection, arrhythmias, acidosis, acute pulmonary embolism, high-output states, and acute exacerbations of the underlying disease; therefore, identification and management of precipitating factors are essential. Management of acute right ventricular dysfunction in ILD-associated pulmonary hypertension includes arrhythmia control, ventilatory management, maintenance of hemodynamic stability through preload optimization and rational use of inotropes and vasopressors, and the administration of pulmonary arterial vasodilators.

Acknowledgment

None

Conflict of Interest

The authors affirmed no conflict of interest

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