Review
ISSN 2413-0516

Pulmonary Hypertension in Saudi Arabia: Prevalence, Patient Characteristics, and Current Management

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(Submitted: 20 August 2025 – Revised version received: 15 September 2025 – Accepted: 16 October 2025 – Published online: 26 December 2025)

Abstract

Objective: This review provides a comprehensive overview of the literature on Pulmonary Hypertension (PH) in Saudi Arabia.

Methods: Data about the appearance of PH in Saudi Arabia were collected, and the epidemiology, treatment options, and types of medications were discussed.

Results: Studies indicate an alarming increase in PH prevalence, driven by a combination of genetic predispositions, lifestyle changes, and improved diagnostic methods. The demographic profile of PH patients in Saudi Arabia also reveals a concerning trend, with women being affected at a rate higher than men which could be attributed to hormonal influences. Increased awareness, early diagnosis, and the development of targeted treatment strategies are essential for improving outcomes for patients with PH. PH symptoms are managed with a combination of medications, lifestyle changes, and sometimes surgery, depending on severity and underlying causes. Endothelin Receptor Antagonists, Phosphodiesterase-5 Inhibitors, Prostacyclin Analogues, and Soluble Guanylate Cyclase Stimulators, along with novel therapeutic agents such as Sotatercept (activin signaling inhibitors), are the primary treatment pathways for PH. Fasudil (a Rho-kinase, ROCK inhibitor), Tacrolimus (calcineurin inhibitor), estrogen inhibitors, and AMP-activated protein kinase (AMPK) activators are also discussed as potential options.

Conclusion: This review offers a concise overview of pulmonary hypertension in Saudi Arabia, highlighting ongoing challenges and advancements in its management.

Keywords: Pulmonary, hypertension, epidemiology, pharmacology

Introduction

Pulmonary arterial hypertension (PH) is a progressive and severe disease with a 1-year mortality rate surpassing 20% in high-risk groups.[1] This condition is characterized by a mean pulmonary arterial pressure (mPAP) greater than 20 mmHg at rest, as measured through right heart catheterization, and by increased pulmonary arterial pressure, which leads to significant illness and death.[2,3] It occurs spontaneously when the blood vessels in the lungs become narrowed, blocked, or damaged, making lung blood flow difficult and causing the heart to weaken and fail.[4] PH is often underdiagnosed due to its subtle onset and nonspecific symptoms such as dyspnea on exertion, fatigue, and exercise intolerance. However, there are different groups of PH, which may overlap, so proper diagnosis, classification, and treatment are crucial. Based on the WHO classification system, these groups are categorized based on their cause.[5] PH is categorized into 5 groups: Group 1: PH, which includes Idiopathic PH, Heritable PH, Drug-Induced PH, and PH associated with conditions such as connective tissue diseases and congenital heart defects. Group 2: PH due to Left Heart Disease, Group 3: PH due to lung diseases and/or Hypoxia, Group 4: chronic thromboembolic PH (CTEPH), and Group 5: PH with unclear multifactorial mechanisms.[2]

Several studies have examined the epidemiology, treatment options, and clinical outcomes of PH; however, in Saudi Arabia, studies were limited until 2020, and their number has increased gradually up to 2025. The prevalence of PH varies with underlying conditions; for instance, it is estimated to affect 15% of patients with systemic sclerosis[6] and predominantly affects women 1.8 times higher than men.[7] The pathophysiological mechanisms underlying PH include vascular remodeling, smooth muscle proliferation, and endothelial dysfunction.[8] These changes lead to increased pulmonary vascular resistance and right ventricular strain.[9] Various factors contribute to these processes, including genetic predisposition, environmental triggers, and circulating inflammatory mediators.[10] Also, PH is likely to be more prevalent in the developing world because of the presence of risk factors like schistosomiasis, HIV, Chronic viral hepatitis, uncorrected congenital heart diseases, hemolytic anemias, and smoking.[11] This review aims to provide an overview of the current understanding of the pathophysiology, clinical classification, clinical manifestations, diagnostic challenges, and pharmacological treatments, as well as their effectiveness, as well as the challenges faced in managing this condition in Saudi Arabia.

Occurrence of PH in Saudi Arabia

Historically, the prevalence of PH in Saudi Arabia has been under-recognized, largely due to a lack of comprehensive nationwide screening programs. Recent studies indicate that the prevalence of PH in Saudi Arabia is on the rise, influenced by genetic factors, lifestyle changes, and increased awareness of the disease.[12] PH, notably linked with congenital heart defects, comprises a significant portion of these cases.[13–15] Furthermore, there is growing evidence suggesting that the prevalence of idiopathic pulmonary arterial hypertension (IPAH) in Saudi Arabia is similar to, or potentially higher than, that in Western populations.[16] The detected studies of PH in Saudi Arabia were summarized in Table 1.

The SAUDIPH registry, initiated recently, reported that Group 1 PAH accounts for 57.7% of cases among enrolled patients[7]. The mean age at diagnosis was reported to be 32 years, indicating a relatively young patient population affected by this condition, which is younger than the reported ages in other international registries. The diagnostic process typically

Table 1. **The detected studies of PH in Saudi Arabia**

Hospital (References)	Number of patients	PH patients	Year	Mean age (years)	Female (%)	Patient groups
Tertiary care center, Riyadh[23]	59 with Down syndrome	44 (75%)	1998–2008	9 ± 5.9	39 (66%)	> 50% of systolic systemic pressure
Tertiary care hospital, Riyadh[24]	264	112 pulmonary artery pressure >25 mm Hg (RHC)	2009–2012	55.8 ± 15.8	72.3%	- 10.7% group 1 pf PH - 6.2% in group 2of PH - 65.2% in group 3 of PH - 3.6% in group 4 of PH - 14.3% in group 5 of PH
Prince Sultan Medical Military City/Cardiac Center, Riyadh[25]	128	107	Dec 2009 and Nov 2012	36 ± 9	62.6%	- 72.8% of the patients were in Group 3 and Group 4 of PH - IPAH was the most common subtype - CHD was the second most common PH subgroup
Single tertiary care hospital[26]	A total of 925 patients underwent RHC	368	Jan. 2013 and June 2014	37.8 ± 12.8	62.4%	- 165 (44.8%) Idiopathic PH, - 93 (25.2%) Congenital heart disease + PH - 89 (24.1%) Connective tissue disease + PH - 12 (3%) Heritable PH - 9 (2.4%) Portal hypertension + PH.
Specialized tertiary care center[17]		222	2004–2018	32		- Group 1 of PH = 57%
King Fahad medical City, Riyadh[27]	95	81 (85%)				- 79% had post-capillary PH. - 86% had pulmonary artery systolic pressure (PAPs) - 93% had mean pulmonary artery pressure (PAPm) - 89% had pulmonary artery acceleration velocity
Application of Questioner[28]	794 of SCA, CVD, and DM patients.	79 (10%)	29/7/2020 till 15/11/2020.		52.2%	- PHTN in SCA patients was 31.8%
PH clinics in King Abdulaziz Medical City and King Faisal Cardiac Center, Jeddah[7]	118		2018–2021.	59.73	- PH (*n* = 100, 85%) - Obese (*n* = 57, 48%)	- Pulmonary artery pressure (PAP) was significantly higher in patients with mild right ventricular impairment (*P* = 0.001).
Secondary care hospital, Hofuf, Al-Ahsa[29]	264 chronic cardiovascular conditions	48 (19.5%)	Jan. 2024 to June 2024	20–80	47.2%	- 19.5% of the patients, with the majority having mild or moderate - PH was 55.2% in patients with atrial fibrillation
King Abdulaziz Medical City-Riyadh[30]	454	382	Jan. 2008 and Dec. 2023	44	80 %	- Group 1 (*n* = 223), - Other groups (*n* = 159)

RHC, Right heart catheterization; SCA, Sickle cell anemia; CVD, Cardiovascular disease; DM, Diabetes Mellitus.

involves echocardiography, which is widely available in urban centers but may be limited in rural settings, while right heart catheterization was the gold standard for the diagnosis. PH epidemiology varies worldwide, influenced by genetic, environmental, and socio-economic factors.[17] In Saudi Arabia, the unique demographic characteristics and the prevalence of obesity, diabetes mellitus, hypertension, and CHD explain its epidemiology, and ongoing research and education are necessary to improve patient outcomes.[18] The prevalence of PH in Saudi Arabia ranges from 0.5% to 4.6% in the general population, with higher rates seen in specific subgroups, especially among patients with underlying conditions like congenital heart disease (CHD) and chronic obstructive pulmonary disease (COPD).[19] A study at King Saud University found that among patients undergoing echocardiography, 3.9% were diagnosed with PH, emphasizing the importance of routine screening in high-risk groups.[20]

Additionally, the increasing rates of obesity and sedentary lifestyles further exacerbate these risk factors.[21] The prevalence of autoimmune diseases, particularly systemic lupus erythematosus and scleroderma, also appears to be a significant contributor to PH in this region.[22] The lack of public awareness and healthcare access in rural areas may further complicate early detection.[15]

Epidemiology, Mortality, and Prognosis of PH

The prognosis for PH remains poor if left untreated, with a median survival of less than three years after diagnosis.[2]

Several studies conducted in Saudi Arabia indicate that early intervention can significantly improve patient outcomes, but management of PH remains inadequate.[24] A multicenter study in Saudi Arabia analyzed patient data over several years and reported a five-year survival rate of 50% among patients diagnosed with PH. This finding is consistent with results from other regions.[31] Key prognostic factors associated with PH in this population include the functional class at the time of diagnosis, the underlying cause of the disease, right ventricular function, and comorbidities such as diabetes and renal dysfunction and it was reported that PH patients in class IV face significantly higher mortality rates than those in class I.[32]

Symptoms and Diagnostic Criteria of Pulmonary Hypertension

Pulmonary hypertension primarily causes exercise intolerance and may present with symptoms such as shortness of breath (dyspnea), significant fatigue, palpitations, and fainting, as well as chest discomfort and palpitations due to heart strain. Awareness of these symptoms is vital, as delays in recognition can delay diagnosis. Diagnosis combines clinical evaluation, imaging studies like echocardiography, and invasive hemodynamic measurements through right heart catheterization, which is the gold standard and is essential for diagnosis, helping to differentiate types of PH and providing key hemodynamic information. Echocardiography helps estimate pulmonary artery pressures and assess heart function. Other evaluations, like pulmonary function tests and imaging studies, are also valuable. Increased awareness and adherence to diagnostic criteria are necessary to improve the early detection and management of PH.[33] Patient education and awareness of PH symptoms are essential for early diagnosis and better prognosis.[34] Despite progress, gaps remain in recognizing and managing the disease, which highlighting the need for local registries to understand its epidemiology better.[35]

Survival Rates and Outcomes

PH is categorized based on the WHO classification system into 5 groups as described before.[2] The research findings from the past two decades outline the state of PH in Saudi Arabia, focusing on pharmacological management, clinical practices utilized, and study the effectiveness and accessibility of these treatments.[36] Patients with PH have shown encouraging survival rates, with 1, 3, and 5 year survival rates of 95.6%, 89.2%, and 74.6%, respectively and those with congenital heart disease-associated pulmonary arterial hypertension (CHD-PAH) had the highest rates, achieving 100% survival at 1 and 3 years.[17]

Treatment and Management

The first comprehensive guidelines for the diagnosis and treatment of PH in Saudi Arabia were published in 2008[37] and covered the most aspects of the disease. Efforts were made to present the guidelines in an easy-to-read format, making them accessible and useful for clinicians treating PH patients, enabling them to select the best management strategies for individuals with specific conditions.[38] Over the past two decades, management strategies for PH have significantly evolved, shifting the focus towards targeted therapies. According to the SAUDIPH registry, 83.7% of patients were receiving specific therapies, including endothelin receptor antagonists (ERAs), phosphodiesterase-5 inhibitors (PDE5Is), and prostacyclin analogs. These therapies have shown positive outcomes; for example, 30% of patients transitioned from functional classes III/IV to I/II, indicating an improvement in functional status.[17] Table 2 shows the currently approved therapeutic medications for PH include endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, prostacyclin receptor agonists (PRAs), prostacyclin analogs (PAs), and soluble guanylate cyclase (sGC) stimulators, in addition to new drugs like the activin signaling inhibitor.[39]

Endothelin Receptor Antagonists

Landmark studies[40,41] have demonstrated the efficacy and safety of endothelin receptor antagonists (ERAs) in treating PH. These drugs play a vital role in managing PH by improving hemodynamics, exercise tolerance, and quality of life, while also slowing disease progression.[42] They work by blocking endothelin-1, a potent vasoconstrictor that elevates pulmonary artery pressures. By relaxing the pulmonary vasculature and decreasing vascular resistance, they enhance blood flow and exercise capacity. Continued research is essential to optimize treatment protocols, particularly in diverse populations like those in Saudi Arabia.

The primary ERAs include Bosentan, Ambrisentan, and Macitentan. Bosentan, the first approved for PAH, improves exercise capacity[43] while Ambrisentan, a selective ETA receptor antagonist, is known for its favorable side effect profile and effectiveness in reducing clinical worsening.[42] Similarly, Macitentan has shown improvements in morbidity and mortality.[44] While ERAs can be used alongside other treatments, they may have side effects such as liver function abnormalities and peripheral edema. Regular monitoring is crucial, especially with Bosentan, due to potential hepatotoxicity.[45]

Phosphodiesterase Type 5 Inhibitors

Phosphodiesterase type 5 inhibitors (PDE5Is) are an important treatment option for pulmonary hypertension. They inhibit the enzyme phosphodiesterase type 5, increasing levels of cyclic guanosine monophosphate (cGMP) in the pulmonary blood vessels. This leads to vasodilation and improved blood flow, alleviating symptoms of PAH. Sildenafil (Revatio) is the commonly used PDE5I, and clinical studies have demonstrated that it improves exercise capacity, functional class, and hemodynamic parameters in PH patients.[46] The SUPER-1 study found significant enhancements in the 6-minute walk distance (6MWD) for those treated with sildenafil compared to a placebo.

Tadalafil is another PDE5i approved for PH, with a longer half-life and a favorable safety profile that improves 6MWD and hemodynamic measures.[47] PDE5 inhibitors are often combined with other PH treatments, such as endothelin receptor antagonists, to enhance effectiveness.[48,49]

Prostacyclin Analogs

Prostacyclin analogs (PCAs) are medications used to treat PH by mimicking the effects of prostacyclin, a potent vasodilator. These drugs dilate both pulmonary and systemic arteries, reducing pulmonary vascular resistance and lowering pressure in the pulmonary artery.[50] They also inhibit the proliferation of vascular smooth muscle cells, promoting long-term vascular remodeling.[50,51]

Table 2. **The most used and new drugs to treat PH, their mode of action and side effects**

Treatment	Medication group	Drug	Mode of action	Refs.	Side effect
Used drugs	Endothelin Receptor Antagonists	- Bosentan - Ambrisentan, - Macitentan	- Blocking endothelin-1, a potent vasoconstrictor	40–45	Causing liver enzyme elevation, peripheral edema, anemia, vasodilatory effects like headache, flushing, and nasal congestion.
	Phosphodiesterase Type 5 Inhibitors	- Sildenafil (Revatio) - Tadalafil	- Inhibit the enzyme phosphodiesterase type 5 - Increasing levels of cGMP in the pulmonary blood vessels	46–49	Causing non-arteritic optic ischemic neuropathy, chorioretinopathy, glaucoma, and optic atrophy.
	Prostacyclin Analogs (Pas),	- Epoprostenol - Treprostinil - Iloprost	- Mimicking the effects of prostacyclin. - Inhibit the proliferation of vascular smooth muscle cells	50–53	Causing jaw pain, diarrhea, flushing, headaches, nausea, vomiting
	Soluble Guanylate Cyclase (Sgc) Stimulators	- Riociguat	- Crucial in the nitric oxide signaling pathway, - Converting GTP to cGMP), which regulates vasodilation and smooth muscle relaxation	54	Cause headache, dizziness, nausea, diarrhea, and hypotension
New drugs	Rho-kinase (ROCK) inhibitors	- Fasudil	- Inhibit myosin light chain phosphorylation and relax constricted vascular smooth muscles	56	Causing drops in blood pressure, dizziness, fatigue, and potential cardiovascular collapse
	Estrogen inhibitors	- Aromatase inhibitors (anastrozole) - estrogen antagonists (fulvestrant)	- Exhibit contradictory effects on pulmonary vasculature, with certain forms having antiproliferative and proapoptotic actions		Causing joint/muscle pain, fatigue, mood changes (anxiety, depression), headaches, high cholesterol, and bone thinning (osteoporosis)
	AMP-activated protein kinase (AMPK) activators	- Metformin	- Lower the risk of cardiovascular diseases - Activates AMP-activated protein kinase - Reducing vascular remodeling and cell proliferation - Enhancing endothelial NO synthase activity and relaxant effects in blood vessels.	63	The role of AMPK activation in PH is complex and still under study
	Bone morphogenetic protein 2	Tacrolimus Sotatercept (activin signaling inhibitor)	- Potent BMP2 activators	65–69	Causing postoperative inflammation, ectopic bone formation, osteoclast-mediated bone resorption, and inappropriate adipogenesis
Non-pharmacological treatments	- Lifestyle modifications - Sleep breathing disorder management - Healthy diet, low in sodium and rich in vitamins - Supplemental oxygen - Continuous Positive Airway Pressure (CPAP) therapy - Exercise training as yoga, meditation, or deep breathing - Psychosocial support - Vaccinations, for influenza and pneumonia - Lung transplantation			73–77	No detected side effect

Epoprostenol is a well-known prostacyclin analog and was the first agent approved for severe PH. Due to its short half-life, it requires continuous intravenous administration. Also, Treprostinil has longer half-lives and can be given subcutaneously or intravenously which improves exercise capacity and quality of life.[52] Similarly, Iloprost can be inhaled or administered intravenously, also enhancing exercise tolerance.[53] A meta-analysis indicated that prostacyclin analogs lead to better survival and exercise capacity than other therapies. However, common side effects like headache, gastrointestinal disturbances, and jaw pain may limit their use.

Soluble Guanylate Cyclase

Soluble guanylate cyclase (sGC) stimulators like Riociguat are crucial in the nitric oxide (NO) signaling pathway, converting guanosine triphosphate (GTP) to cyclic guanosine monophosphate (cGMP), which regulates vasodilation and smooth muscle relaxation. Clinical trials have shown Riociguat significantly improves exercise capacity, functional class, and hemodynamics in patients with idiopathic PH and chronic non-operable/residual thromboembolic PH, especially in those unresponsive to other treatments.[54]

Other Novel Therapeutic Agents

Although there is significant progress in PH therapy, the prognosis for many patients, especially those with advanced disease, is still poor. The importance of the novel therapeutic targets of ROCK inhibition using fasudil/AT-877ER, BMP2 inhibition using tacrolimus and sotatercept, estrogen inhibitors using anastrozole and fulvestrant, and AMPK activation using metformin, on clinical and laboratory parameters as well as on the pulmonary vascular hemodynamics of PAH patients was documented below.[55]

Rho-kinase Activity

Rho-kinase (ROCK) inhibitors are strong vasodilators that inhibit myosin light chain phosphorylation and relax constricted vascular smooth muscles via novel mechanisms. Fasudil, a ROCK inhibitor, has been approved in Japan and China to prevent cerebral vasospasm in aneurysmal subarachnoid hemorrhage. Many investigations have shown that ROCK activity is increased in PAHs, and ROCK inhibition with intravenous Fasudil and oral Fasudil hydrochloride (AT-877ER) has been studied for its vasodilator impact, which may restrict PH progression.[56]

Estrogen Inhibitors

A significant increase in the female/male ratio among PH patients suggests a potential role of estrogen in its pathophysiology,[57] although female PH patients tend to have a milder disease course than males. Animal studies indicate estrogen may protect against PH development, contributing to the unclear role of estrogen in this disease, often referred to as the estrogen paradox.[58] Estrogen metabolites exhibit contradictory effects on pulmonary vasculature, with certain forms having antiproliferative and proapoptotic actions, while others promote inflammation and proliferation.[59] Estrogen derivative, E2 can mitigate vascular proliferation through protective metabolites and influence VEGF gene expression while also stimulating endothelial cell proliferation. Aromatase inhibitors like anastrozole and estrogen antagonists such as fulvestrant are critical in PH research.[60–62]

AMP-activated Protein Kinase (AMPK) Activators

Metformin is a well-known antidiabetic medication that improves insulin sensitivity, enhances fatty acid oxidation, and reduces oxidative stress. This makes it a potentially effective metabolic therapy for PH. Previous studies have suggested that metformin can lower the risk of cardiovascular diseases in patients with PH. These benefits are not solely due to the drug's blood sugar-lowering effects; they may also involve its ability to improve lipid metabolism, modulate inflammatory responses, and enhance the functions of endothelial and vascular smooth muscle cells. Additionally, metformin activates AMP-activated protein kinase (AMPK), which can promote the vasodilatory and antiproliferative effects of nitric oxide.[63]

Bone Morphogenetic Protein 2

The transforming growth factor-β (TGFβ) superfamily, focusing on BMP2 (bone morphogenetic protein 2), is discussed in many studies. BMP2 is recognized for its diverse functions, which include organ development both in the skeletal and extra-skeletal systems, bone regeneration, and angiogenesis.[64] In the context of PH, mutations in BMP2 are primarily found in heritable pulmonary arterial hypertension. Conversely, a reduction in BMP2 expression without mutations has been observed in idiopathic pulmonary arterial hypertension as well as in various other forms of PH. This mutation or deficiency in BMP2 leads to dysfunction and apoptosis (cell death) of pulmonary endothelial cells, increased cellular proliferation, vascular remodeling, and the loss of small peripheral pulmonary vessels. As a result, there is a progressive rise in pulmonary arterial resistance, which initiates and sustains PAH. Researchers have explored the potential of Tacrolimus and Sotatercept, both potent BMP2 activators, to alleviate symptoms of PAH and reverse its underlying pathology.[65–68]

Tacrolimus

Among drugs approved by the U.S. Food and Drug Administration, tacrolimus have been identified as potent activators of BMPR2, which reversed experimental pulmonary arterial hypertension.[67] This drug restored the normal function of pulmonary artery endothelial cells and, despite its side effect profile, it can be considered as an ideal candidate for use in PH patients. Based on these findings, a randomized, double-blind, placebo-controlled phase II trial was initiated to evaluate the safety and tolerability of low-dose tacrolimus in stable PH patients over a 12-month period.[69]

Sotatercept

Sotatercept (Winrevair) is an innovative inhibitor of the activin signaling pathway that significantly improves outcomes for PH patients with Group 1. It reduces the risk of death, the need for lung transplantation, and hospitalization while enhancing exercise capacity, improving functional class, and decreasing clinical worsening events. Sotatercept works by binding to free activins and growth differentiation factors, thereby restoring the balance between growth-promoting and growth-inhibiting signals in the pulmonary arteries.[70]

Traditional therapies primarily focus on expanding blood vessels through vasodilation, while Sotatercept targets the cellular mechanisms that contribute to the pathological remodeling of blood vessels in the lungs. It specifically addresses the disrupted balance of growth and inhibitory signals within the TGF-β signaling pathway, a crucial mechanism in the pathobiology of PH. Sotatercept is effective for a wide range of PH patients, even in patients previously considered untreatable. In the ZENITH study, 172 patients with advanced PH who were at high risk of death within one year were treated either with Sotatercept or a placebo.[70] Only 17.4% of patients in the Sotatercept group were at risk, compared to 54.7% in the placebo group, representing a 76% risk reduction (Hazard Ratio: 0.24; $P < 0.001$). In another study, the effectiveness of Sotatercept was tested on 320 patients with PH. In the Sotatercept group, 10.6% experienced at least one primary endpoint event, compared to 36.9% in the placebo group.[71]

Combination Therapy

Combination therapy is the standard approach for managing Group1PH. This strategy involves using two or more medications from different classes to enhance therapeutic effects, better control symptoms, and improve patient outcomes.[72] The choice of medications typically depends on the specific type of PH, the severity of the disease, the patient's tolerance, and their response to previous treatments. Additionally, lifestyle

changes and supportive therapies can complement pharmacological treatments.

Non-Pharmacological Treatments

Non-pharmacological approaches can complement medical treatments and improve the overall management of PH, contributing to a better quality of life. Patients with hypoxia can benefit from various non-pharmacological treatments, including lifestyle modifications, long-term oxygen therapy, and sleep breathing disorder management, which significantly improve the quality of life.[73] A heart-healthy diet, low in sodium and rich in vitamins, is essential for managing blood pressure and reducing fluid retention. Supplemental oxygen can improve oxygen delivery to tissues, relieve symptoms, and enhance exercise tolerance. Addressing sleep disorders, particularly sleep apnea, is also critical, as they can worsen PH symptoms. Continuous Positive Airway Pressure therapy can be beneficial for individuals with obstructive sleep apnea. Participating in support groups can provide emotional support and help manage anxiety and improve mental health. Additionally, a structured program that combines education, exercise training as yoga, meditation, or deep breathing, and psychosocial support can foster both the physical and emotional health of PH patients. Also, vaccinations, particularly for influenza and pneumonia, are vital to prevent respiratory infections and enhance overall health. Finally, for patients with advanced PH who do not respond to medical therapy, lung transplantation may be considered.[74]

Lung Transplantation in Saudi Arabia

Unfortunately, many patients with end-stage PH do not respond adequately to medical therapies because the management is complex and requires careful assessment. Based on multi-parameter assessments, patients can be classified as at high risk if their estimated 1-year mortality is >20% or with signs of severe right ventricular dysfunction, or with right ventricular failure and secondary organ dysfunction.[2] Lung transplantation is a critical treatment option for these patients because it can offer a chance for significant improvement in quality of life and survival. While the most common indication for lung transplantation in Saudi Arabia remains pulmonary fibrosis, any patient with severe PH may be considered for transplantation.[75] The significance of lung transplantation for end-stage lung diseases as a critical treatment option was detected by King Faisal Specialist Hospital and Research Center (KFSHRC), located in Riyadh.[76] Comprehensive evaluations are conducted to ascertain the patients' fitness for surgery and Lung Transplantation. Organ donation in Saudi Arabia operates under a regulated system managed by the Saudi Center for Organ Transplantation. Across all indications, 3-month survival post-lung transplant has increased from 82% to 91%.[77] However, recent advances in pre- and post-operative management have resulted in considerable improvements in early post-transplant survival rates. A total of 80 lung transplantation surgeries were performed between 2010 and 2015.[78] The 30 and 90-day mortality rates were 12.5% and 17.5%, respectively, and survival rates were 87.5% at 30 days, 82.5% at 90 days, 81.2% at 1 year, 67.9% at 2 years, and 62.1% at 5 years. Despite the advances in lung transplantation in Saudi Arabia, several challenges persist.[79] The primary hurdles include the shortage of organ donors and the need for increased public awareness regarding the importance of organ donation.

Current Practices, Challenges, and Future Directions

In Saudi Arabia, the treatment of pulmonary hypertension follows the 2020 guidelines established by the Saudi Pulmonary Hypertension Society. These guidelines emphasize customizing treatment for different PH classifications, focusing on group 1 pulmonary arterial hypertension and inoperable cases in group 4 chronic thromboembolic pulmonary hypertension. Combination therapy is the standard for group 1 PAH and is sometimes used for group 4. Despite advancements, managing PH remains difficult due to the need for greater awareness and education among healthcare providers and patients. The primary treatments include endothelin receptor antagonists and phosphodiesterase type 5 inhibitors, while access to advanced therapies like prostacyclin analogues remains limited. Training healthcare personnel for complex treatments is also a challenge. Future research should prioritize multi-center studies to improve understanding and personalize treatment based on genetic and phenotypic factors. Overall, enhancing detection, management, and research is essential for better patient outcomes in Saudi Arabia.

Conclusion

The management of pulmonary hypertension in Saudi Arabia involves various pharmacological treatments that can enhance patient outcomes. Ongoing research is essential for optimizing treatment protocols and understanding the long-term effects of these medications within the Saudi population. Future studies should adopt multidisciplinary approaches to investigate the efficacy and safety profiles of these treatments. Adhering to clinical guidelines is crucial, and recent advancements in pharmacotherapy offer promising potential for improved outcomes. This review emphasizes the complexity of pulmonary hypertension and the necessity for tailored treatment approaches, along with continuous research in this field.

Conflict of Interest

No potential conflicts of interest are found that might be relevant to the contents of this manuscript.

Funding

No funding was provided.

Author Contribution

The author designed the study, collected the data, wrote the manuscript, and approved the final version.

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