General Measurement

Pulmonary hypertension (PH) is a complex and severe medical condition characterized by elevated blood pressure in the pulmonary arteries, leading to significant morbidity and mortality if untreated. The condition involves multiple pathophysiological mechanisms that can result in right heart failure due to increased resistance in the pulmonary circulation. The World Symposium on Pulmonary Hypertension (WSPH) has developed a comprehensive classification system to facilitate the understanding, diagnosis, and treatment of PH.

The classification of PH is divided into five major groups: Pulmonary Arterial Hypertension (PAH), PH associated with Left Heart Disease, PH associated with Lung Diseases and/or Hypoxia, PH associated with pulmonary artery obstruction, and PH with Unclear and/or Multifactorial Mechanisms Diagnosing PH requires a multifaceted approach. The primary diagnostic tool is right heart catheterization, which measures hemodynamic parameters such as mean pulmonary arterial pressure (mPAP), pulmonary capillary wedge pressure (PCWP), and pulmonary vascular resistance (PVR). PH is defined hemodynamically as a mPAP greater than 20 mmHg at rest. Additional diagnostic methods include echocardiography, pulmonary function tests, and imaging studies such as CT and MRI to assess the pulmonary vasculature and cardiac function. In 2024, significant advancements have been made in the field of multimodality imaging techniques for the pulmonary circulation. AI and machine learning algorithms are being integrated into imaging modalities to enhance image interpretation, detect subtle abnormalities, and predict disease progression. Combining different imaging modalities, such as PET/CT or PET/MRI, provides comprehensive information by merging anatomical and functional data, improving diagnostic accuracy and patient management

Special Circumstances in Pulmonary Hypertension

Chen, Zheng-Wei (陳政瑋)
Division of Cardiology, Department of Internal Medicine, National Taiwan University Hospital Yunlin Branch

Abstract

Pulmonary Hypertension (PH) presents unique challenges across various life circumstances due to its chronic nature and associated risk factors. This discussion highlights the special considerations in managing PH during birth control, pregnancy, surgical procedures, and travel, emphasizing the need for a multidisciplinary approach to care.

Birth Control: Effective contraceptive counseling is essential for women with PH of childbearing age, considering the significant risk of contraceptive failure. While most hormonal contraceptives are generally safe, certain PH treatments may reduce their effectiveness, underscoring the need for individualized care.

Pregnancy: Pregnancy in severe PH cases entails substantial maternal and neonatal mortality risks. Although recent advances have slightly improved maternal outcomes, pregnancy remains generally contraindicated for women with PH due to its unpredictable progression. Clear counseling and options like early termination, adoption, or surrogacy are vital for informed decision-making.

Surgical Procedures: Surgery poses high risks for PH patients, including potential right heart failure and increased mortality rates, particularly when emergency surgery is required. A tailored assessment involving a multidisciplinary team is essential to evaluate surgical necessity, urgency, and the individual's health status.

Travel and Altitude Exposure: Patients with PH face risks from hypoxia at high altitudes, which can exacerbate symptoms. Oxygen supplementation is recommended during air travel for certain patients, and short-term hypoxia is generally tolerable in stable patients. However, travel beyond 2,500 meters should be avoided without oxygen support.

This speech provides critical insights for managing PH patients across complex situations, ensuring optimized, patient-centered care.

Modern Pharmacological Treatments and Drug Interaction

Wu Chun Hsien

In recent years, advancements in the pharmacological treatment of pulmonary arterial hypertension (PAH) have shown promising developments, as highlighted in the 2024 TSOC pulmonary hypertension guidelines. This presentation, titled Modern Pharmacological Treatments and Drug Interaction of PAH, focuses on the latest therapeutic pathways and drug options available for managing PAH, with special attention to novel therapies such as sotatercept.

Sotatercept, a first-in-class fusion protein targeting the transforming growth factor-beta (TGF-β) superfamily, represents a groundbreaking addition to PAH therapy. It works by restoring the imbalance between pro-proliferative and anti-proliferative signaling in pulmonary vascular cells, addressing a key pathogenic pathway in PAH. Clinical trials have shown sotatercept's potential to improve pulmonary vascular resistance and enhance patient outcomes by targeting vascular remodeling, a critical factor in PAH progression.

In addition to traditional therapies targeting the endothelin, nitric oxide, and prostacyclin pathways, these new treatment modalities provide a more comprehensive approach to managing PAH. However, with these advancements comes the increased complexity of drug interactions. The combination of multiple agents raises the potential for pharmacodynamic and pharmacokinetic interactions, which necessitates careful consideration in clinical practice. Physicians must evaluate patient-specific factors and continuously monitor for adverse effects, especially when combining sotatercept with other PAH therapies.

Intervention Therapies and Management of Advanced Right Heart Failure

台中榮總 梁凱偉 醫師

The management of patients with acute decompensation of pulmonary arterial hypertension (PAH) is based on the treatment of the precipitating factors, the implementation of a careful fluid management plan, and the use of strategies to reduce right ventricular (RV) afterload and improve cardiac function and tissue perfusion.

Fluid volume optimization by intravenous diuretics is the cornerstone of treatment for right-sided heart failure in PAH patients. Loop diuretics are commonly used due to their rapid onset of action and efficacy. Inotropes, such as low doses of dobutamine (2.5 $\mu g \cdot kg^{-1} \cdot min^{-1}$), can be initiated with progressive up-titration in cases of persistent signs of low cardiac output. Vasopressors may be required to rapidly restore blood pressure and vital organ perfusion. Among drugs with a prominent peripheral arterial vasoconstrictor action, norepinephrine and vasopressin should be the drugs of choice. Use of target PAH medication including a prostacyclin analogue can effectively and dramatically decrease the RV afterload.

Extra-Corporeal Membrane Oxygenation (ECMO) may stabilize oxygenation and RV function and reverse end organ dysfunction. ECMO support has become an established bridging tool to transplantation in patients with irreversible right heart failure. It also provides a therapeutic window for medically stabilizing critically decompensated PAH patients.

Balloon atrial septostomy (BAS) creates an iatrogenic right-to-left interatrial shunt, decreases the filling pressure of the right heart, increases the filling pressure of the left heart, increases systemic cardiac output and oxygen delivery at the expense of systemic arterial desaturation for subjects with PAH. It is recommended for PAH with advanced right heart failure as a palliative therapy or a bridge to lung transplantation. BAS is not recommended for PAH patients with a mean right atrium pressure greater than 20 mmHg. Furthermore, BAS should be performed by experienced structural heart interventionists at the PAH referral center. Image guidance during BAS is recommended due to distorted anatomy in PAH with a very dilated right atrium and a small left atrium.

Diagnostic Algorithm for Pulmonary Hypertension

Chun-Yuan Chu MD. (朱俊源)
Division of Cardiology, Kaohsiung Medical University Hospital

Abstract:
The ultimate diagnosis of pulmonary hypertension (PH) is established through right heart catheterization, which should be performed at a PH center by an experienced team. The main aim of the diagnostic algorithm is to discern those patients who need to be referred to a PH center and who should undergo invasive hemodynamic assessment. The time between diagnostic steps should be minimized. At each step, patients with a low likelihood of PH and a more plausible alternative cause (e.g. significant left heart disease) for their symptoms may be identified and managed with an alternative diagnostic approach.

Nevertheless, patients with interstitial lung disease and suspected PH, and patients with pulmonary conditions or left heart disease and suspected severe PH should be referred to a PH expert center. In case of a high probability of PAH or CTEPH, especially if there are signs of right heart failure, a fast-track referral to a PH center is recommended at any point during the clinical workup.

Most patients who undergo diagnostic evaluation for PH present with symptoms of dyspnoea, exercise intolerance and/or clinical signs of right heart failure. A stepwise diagnostic approach for these patients, starting with simple, noninvasive tools and followed by more complex diagnostic methods, including the assessment for common cardiac and pulmonary conditions

Risk Assessment and Treatment Goals for Pulmonary Hypertension

Pulmonary hypertension (PH) is a complex and progressive condition characterized by elevated blood pressure in the pulmonary arteries, which can lead to right heart failure if untreated. Effective management of PH requires a comprehensive risk assessment, a structured treatment algorithm, and clear treatment goals.

Risk Assessment: The risk assessment for PH involves evaluating the patient's clinical status, hemodynamic parameters, exercise capacity, and biomarkers. Key indicators include the World Health Organization (WHO) functional class, six-minute walk distance (6MWD), levels of brain natriuretic peptide (BNP) or N-terminal pro b-type natriuretic peptide (NT-proBNP), and echocardiographic findings. This multifaceted approach helps stratify patients into low, intermediate, or high-risk categories, guiding treatment decisions and prognostication.

Treatment Algorithm: The treatment algorithm for PH is tiered based on the patient's risk category. For patients with pulmonary arterial hypertension, advanced therapies may include endothelin receptor antagonists (ERAs), phosphodiesterase-5 inhibitors (PDE-5i), prostacyclin analogs, or sotatercept. Combination therapy is strongly recommended, and parenteral prostacyclin should be considered for patients with high-risk status. Regular follow-up and reassessment are crucial for optimizing therapy.

Treatment Goals: The primary goals of PH treatment are to relieve symptoms, restore right ventricle function, improve hemodynamic parameters, enhance quality of life, and prolong survival.

Effective management of pulmonary hypertension thus hinges on a thorough risk assessment, adherence to a structured treatment algorithm, and the pursuit of clearly defined treatment goals.

Therapeutic Strategies for Pulmonary Hypertension

吳書豪醫師
台北馬偕紀念醫院心臟內科

In this short talk, we review management of some special conditions in patients with pulmonary hypertension. Besides general principles, we focus on update from the 7th World Symposium of Pulmonary Hypertension.

We will go through several topics. The first is how to manage these patients with right heart failure in the setting of intensive care unit. The key is to keep adequate output and perfusion, with medication and mechanical circulatory support if needed. Second, the experts do not recommend pregnancy although they do not prohibit it. It is emphasized to manage delivery in an experienced center. Cesarean section is mostly favored. For peri-operative management, we need careful evaluation, watching certain risk factors and acknowledging that deterioration may happen even in relative stable patients. Anesthesia is a critical part. In palliative/end-of-life care, the experts suggest regular assessment. It is not necessarily “end-of-life” care, it is more for how to live well.

For these special conditions, there are few trials. We need more specialists to work on how to better manage these situations.

Comparison of WSPH and TSOC guideline

In this section, we will discuss about new proposals of PH with left heart disease in WSPH 2024, includes the new classification of group 2 PH, PH with borderline PAWP (between 12 to 15 mmHg). Also, the results of clinical trials of PH-left heart disease in the past 6 years will be reviewed as well.