Definition, classification and diagnosis of pulmonary hypertension (Including imaging techniques)

The prostacyclin (prostaglandin I2 (PGI2)) pathway is an important therapeutic target in pulmonary arterial hypertension (PAH). Reduced levels of PGI2 are associated with pathogenic changes within the lung vasculature and therapies that aim to restore PGI2 signalling have been shown to play a key role in PAH management . Evidence to support the use of therapies targeting the PGI2 pathway was first generated in the 1990s when intravenous epoprostenol, was demonstrated to provide symptomatic and haemodynamic benefits and to improve survival. As a result, epoprostenol was the first drug to be approved for PAH. The European Society of Cardiology/European Respiratory Society (ESC/ERS) guidelines cite different levels of recommendation and evidence for each therapy, with epoprostenol having the highest level (IA) in patients with WHO FC III or IV disease. Intravenous prostanoids require continuous infusion via a central venous catheter due to their short half-lives . Serious side-effects may occur on interruption or withdrawal of i.v. prostanoids , which can result from pump malfunctions or if the line is erroneously compromised. A compromised line can lead to cardiovascular collapse

Subcutaneous treprostinil was first approved for use as a PAH treatment in 2002 and in the current ESC/ERS guidelines it is recommended for use in patients in WHO FC III or IV. Subcutaneous treprostinil is administered as a continual infusion via a s.c. catheter . It has a longer half-life than epoprostenol and as a result, the risk of haemodynamic compromise within a short period of time due to rebound pulmonary constriction appears to be lower. It is available in pre-mixed vials Although the s.c. mode of administration offers advantages, the delivery of a continuous parenteral infusion is not without practical challenges. Infusion site pain and reaction were the most common adverse reactions reported in clinical trials, occurring in 85% of patients. Failure to address infusion site pain may lead to discontinuation of treatment Although site pain is not dose-dependent and is manageable in the majority of patients, local, topical or systemic analgesics may be required Patients with chronic pain may need referral to a pain specialist. Practical advice and support delivered in a timely manner helps patients to overcome their concerns about treatment. With an effective management plan and support from an experienced multidisciplinary team, dealing with the complexities of therapies that target the PGI2 pathway becomes more manageable.

PAH treatment algorithm, risk stratification and treatment goals

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.

Clinical trial designs and emerging therapies (including Paediatric PH)

Pulmonary hypertension is commonly found in daily practice. But some specific etiology like pulmonary artery hypertension is a rare but progressively lethal disease. Therefore the new 2022 ESC/ERS pulmonary hypertension guideline adjust the diagnosis criteria of pulmonary hypertension, emphasize early diagnosis and early efficient combination treatment for these patients. For this goal many image modalities were suggested for the diagnosis and the disease severity evaluation for them. Some high risk specific groups, like those with connective tissue disorder or gene disease, are suggested to have a screening process for them.

What's new in post WSPH

Section: PH and left heart disease

Speaker: Liu Wei-Shin 劉維新

Abstract:

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.

PAH management in special conditions

(ICU, pregnancy, peri-OP, palliative /end of life care)

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.