



## **ISHLT ACADEMY: CORE COMPETENCIES IN PULMONARY HYPERTENSION PROGRAM**

### **Program Chairs**

Colin Church, BSC(hons), MBChB, FRCP, PhD, Golden Jubilee National Hospital, Glasgow, UK  
Ryan J. Tedford, MD, Medical University of South Carolina, Charleston, SC, USA

### **Faculty**

Colin Church, BSC(hons), MBChB, FRCP, PhD, Golden Jubilee National Hospital, Glasgow, UK  
Paul Corris, MB, FRCP, Freeman Hospital, Newcastle University, Newcastle upon Tyne, UK  
Robert Frantz, MD, Mayo Clinic College of Medicine, Rochester, Minnesota, USA  
Mardi Gomberg-Maitland, MD, MSc, George Washington University Health and Medical Sciences, Washington DC, USA  
John Granton, MD, University of Toronto, Toronto, Canada  
Marco Guazzi, MD, PhD, San Paolo Hospital, Milan, Italy  
Brian A. Houston, MD, Medical University of South Carolina, Charleston, SC, USA  
Steven Hsu, MD, Johns Hopkins Hospital, Baltimore, MD, USA  
Denise J. Lewis, RN, BSN, George Washington University, Washington, DC, USA  
Bradley A. Maron, MD, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA  
Dana P. McGlothlin, MD, Kaiser Permanente, San Francisco, CA, USA  
Steven Nathan, MD, Inova Fairfax Hospital, Falls Church, Virginia, USA  
Joanna Pepke-Zaba, PhD, FRCP, Royal Papworth Hospital, Cambridge, UK  
Ioana Preston, MD, Tufts Medical Center, Boston, MA, USA  
Ryan J. Tedford, MD, Medical University of South Carolina, Charleston, SC, USA  
Jean Luc Vachiery, MD, Erasme University Hospital, Brussels, Belgium

### **Target Audience**

This course is primarily designed to benefit clinicians and allied health professionals who are in the early stages of their careers, or who are in training and/or are part of a new program, or desire an update on the current state of the field.

### **Professional Practice Gap this Activity Addresses**

Pulmonary hypertension (PH) is a life-threatening condition commonly encountered in patients with advanced heart and lung disease. Despite significant advances in the field, patients with PH are complex and their management can be challenging. Recognition of the PH problem and delay in diagnosis remains an issue and this contributes to the poor long term survival of these patients. Misidentification of the causes and misclassification of PH leads to poor survival rates.

### **Educational Need for this Activity**

This course is designed for:

- those who are in the early stages of their careers or
- who are training or
- who are part of a new program

Future clinicians must have this essential training in order to manage patient treatment and improve patient outcomes by learning to recognize pulmonary vascular disease when it is present and then determine the underlying etiology. This course will help to highlight the risks of misclassification of PH and the misuse of approved specific therapies for pulmonary arterial hypertension.

### **This activity is designed to:**

- increase the learner's knowledge in their understanding of the approach to the diagnosis and management of PH.
- improve competence by applying strategies to systematically and effectively assess the etiology of PH in patients.
- improve performance in reducing misclassification of PH and misuse of approved therapies for arterial hypertension leading to better patient outcomes.

### **Learning Objectives**

At the conclusion of this educational activity, participants will have improved competence and professional performance in their ability to:

1. Discuss context and historic background for pulmonary hypertension.
2. Define and classify pulmonary hypertension.
3. Evaluate pulmonary hypertension as an increasingly recognized condition of many causes that is uniformly associated with reduced survival.
4. Review the FDA approved indications for all of the currently available specific therapies for pulmonary arterial hypertension.
5. Identify the risks of misclassification of PH and misuse of approved specific therapies for pulmonary arterial hypertension.
6. Discuss what physicians are using currently to treat PH patients.
7. Apply the management strategies for patients with advanced right heart failure including mechanical support
8. Discuss the essential interaction with allied health professionals including specialist nurse, palliative care

### **Acknowledgement of Financial Commercial Support**

No financial commercial support was received for this educational activity.

### **Acknowledgement of In-Kind Commercial Support**

No in-kind commercial support was received for this educational activity.

### **Satisfactory Completion**

Learners must listen to all seven modules while following along with the visual slides, and complete an online evaluation, to receive a certificate of completion. If you are seeking continuing education credit for a specialty not listed below, it is your responsibility to contact your licensing/certification board to determine course eligibility for your licensing/certification requirement.

### Physicians

International Society for Heart & Lung Transplantation (ISHLT) is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians.

Credit Designation Statement - ISHLT designates this enduring material activity for a maximum of **6.00 AMA PRA Category 1 Credit(s)<sup>TM</sup>**. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

### Accreditation Statement



In support of improving patient care, this activity has been planned and implemented by Amedco LLC and International Society for Heart & Lung Transplantation. Amedco LLC is jointly accredited by the Accreditation Council for Continuing Medical Education (ACCME), the Accreditation Council for Pharmacy Education (ACPE), and the American Nurses Credentialing Center (ANCC), to provide continuing education for the healthcare team.

### Nurses (ANCC) Credit Designation

Amedco LLC designates this activity for a maximum of **6.00 ANCC** contact hours.

### Pharmacists (ACPE) Credit Designation

Amedco LLC designates this activity for a maximum of **6.00** knowledge-based CPE contact hours.

**NOTE to Pharmacists: The only official Statement of Credit is the one you pull from CPE Monitor. You must request your certificate within 30 days of your participation in the activity to meet the deadline for submission to CPE Monitor.**

## SCIENTIFIC PROGRAM

### WELCOME AND OVERVIEW

Length: 2 mins

Ryan J. Tedford, MD, Program Co-Chair  
Medical University of South Carolina, Charleston, SC, USA

### SESSION 1: Let's Start at the Start! General Introduction to RV Physiology and Pulmonary Hypertension

Length: 68 mins

**1. What is this New Pulmonary Hypertension? Definitions and Classifications**

Bradley A. Maron, MD, Brigham and Women's Hospital, Harvard Medical School, Boston, MA, USA

**Teaching/Discussion Points**

- a. Definitions and hemodynamic classification of pulmonary hypertension
- b. WHO clinical classification
- c. Diagnostic evaluation
- d. Update from World Symposium 2018

**2. Understanding RV Physiology and Coupling to the Pulmonary Circulation**

Steven Hsu, MD, Johns Hopkins Hospital, Baltimore, MD, USA

**Teaching/Discussion Points**

- a. Normal RV morphology, physiology and function
- b. RV/PA coupling and ventricular interdependence
- c. RV adaptation to pressure overload
- d. Pathophysiology of RV failure and the right heart failure syndrome in PH

**3. Right Heart Catheterization: Let's Do It Right**

Brian A. Houston, MD, Medical University of South Carolina, Charleston, SC, USA

**Teaching/Discussion Points**

- a. Pulmonary artery catheter and how to perform a diagnostic right heart catheterization
- b. Common mistakes and pitfalls with hemodynamic measures
- c. Indications and performance of vasoreactivity testing
- d. When to consider left heart catheterization for LVEDP assessment

### SESSION 2: The True Pulmonary Vascular Disease: Group 1 PAH

Length: 65 mins

**1. Understanding the History and Pathophysiology of PAH**

Ioana Preston, MD, Tufts Medical Center, Boston, MA, USA

**Teaching/Discussion Points**

- a. Pathology and pathobiology of PAH
- b. Cellular and molecular pathways implicated in PAH
- c. Genetics of PAH

**2. What Treatments Can I Give? Management Strategies and Pharmacotherapy**

Robert Frantz, MD, Mayo Clinic College of Medicine, Rochester, Minnesota, USA

**Teaching/Discussion Points**

- a. Adjunctive therapies and calcium channel blockers
- b. Available PAH specific therapies
- c. Pivotal trials
- d. Updated treatment algorithm

**3. How Do I Know When to Change Treatment? All About Risk**

Mardi Gomberg-Maitland, MD, MSc, George Washington Univ Health and Medical Sciences, Washington DC, USA

**Teaching/Discussion Points**

- a. Importance of risk assessment
- b. Key components of risk assessment and why they are important
- c. Utilization of risk assessment tools in the management of PAH
- d. When to assess risk and what to do about it therapeutically

**SESSION 3: But the LV is Normal So It Can't Be Left Heart Failure: Understanding Group 2 Pulmonary Hypertension**

Length: 63 mins

**1. Pathophysiology and Diagnosis of Group 2 PH**

Jean Luc Vachieri, MD, Erasme University Hospital, Brussels, Belgium

**Teaching/Discussion Points**

- a. Pathophysiology of pulmonary hypertension in left heart disease
- b. Epidemiology and impact of pulmonary hypertension in HFrEF, HFpEF, and valvular heart disease (including patients undergoing surgical and percutaneous, eg TAVR and MitraClip, valve interventions)
- c. Definitions and terminology of WHO group 2 PH
- d. Outcomes and risk predictors

**2. Management and Treatment of Group 2 PH**

Marco Guazzi, MD, PhD, San Paolo Hospital, Milan, Italy

**Teaching/Discussion Points**

- a. Therapeutic options
- b. Pivotal trial data
- c. Management guidelines

**3. PH in LVAD and Transplant Candidates: Before, During and After**

Robert Frantz, MD, Mayo Clinic College of Medicine, Rochester, Minnesota, USA

**Teaching/Discussion Points**

- a. Risks associated with pulmonary hypertension in heart transplantation
- b. Management of pulmonary hypertension in heart transplant candidates, including the role of PH specific therapies and LVAD implantation
- c. Management of right ventricular failure in LVAD recipients
- d. Bridging strategies

## SESSION 4: Just Treat the Lung Disease: Understanding Group 3 Pulmonary Hypertension

Length: 41 mins

### 1. *Diagnosis and Classification of Group 3 PH*

Paul Corris, MB, FRCP, Freeman Hospital, Newcastle University, Newcastle upon Tyne, UK

#### Teaching/Discussion Points

- a. Epidemiology of Group 3 PH
- b. Prognostic significance of PH in COPD and ILD (including idiopathic pulmonary fibrosis and ILD related to connective tissue disease)
- c. Pathophysiology of PH in COPD and ILD, including pulmonary arterial and venous involvement and vasoactive and profibrotic mediators
- d. Significance of PH in advanced lung disease patients being considered for lung transplantation

### 2. *Treatment of PH in Mild/Moderate/Severe Lung Disease with PH*

Steven Nathan, MD, Inova Fairfax Hospital, Falls Church, Virginia, USA

#### Teaching/Discussion Points

- a. Review the therapeutic options based on clinical trial data in Group 3 PH
- b. Discuss the challenges of mixed PH classification patients

## SESSION 5: Clots and Oddities? A Brief Exploration of CTEPH and Group 5 Pulmonary Hypertension

Length: 42 mins

### 1. *A Whistlestop Tour of CTEPH: Current Managements and New Advances*

Joanna Pepke-Zaba, PhD, FRCP, Royal Papworth Hospital, Cambridge, UK

#### Teaching/Discussion Points

- a. Epidemiology with a focus on incidence and risk factors
- b. Review what is known about the evolution from acute to chronic thromboembolic disease
- c. Describe the contribution small vessel arteriopathy plays in the development of CTEPH
- d. Describe the issues surrounding operability and PEA surgery
- e. Review the appropriate role for specific medical therapy and BPA in CTEPH

### 2. *All You Need to Know About Group 5 PH*

Dana P. McGlothlin, MD, Kaiser Permanente, San Francisco, CA, USA

#### Teaching/Discussion Points

- a. Review the causes of PH with unclear/multifactorial mechanisms
- b. Focus on epidemiology, mechanisms of PH and role of PH specific therapies in sarcoidosis and hematologic disorders

## SESSION 6: What To Do When the Drugs Stop Working: Advanced PH Management, RV Failure and Transplant

Length: 46 mins

### 1. *What To Do When the RV Stops Performing: Medical Management (How to Mend a Broken Right Heart)*

John Granton, MD, University of Toronto, Toronto, Canada

#### Teaching/Discussion Points

- a. Review the precipitating factors
- b. Discuss management principles, including the role of pulmonary vasodilator therapies, oxygenation, volume management and inopressor use

### 2. *What To Do When the RV Stops Performing: Mechanical and Surgical Management*

Paul Corris, MB, FRCP, Freeman Hospital, Newcastle University, Newcastle upon Tyne, UK

#### Teaching/Discussion Points

- a. Indications for lung transplantation in PAH
- b. Timing and utilization of atrial septostomy
- c. Mechanical circulatory support indications and bridging strategies

## SESSION 7: Miscellaneous Cases: Pregnancy, Perioperative and Nursing

Length: 45 mins

### 1. *How to Nurse the Patient with End Stage PH*

Denise J. Lewis, RN, BSN, George Washington University, Washington, DC, USA

#### Teaching/Discussion Points

- a. Approach to nursing care
- b. Palliative/hospice care
- c. Anticipatory care
- d. Quality of life assessments

### 2. *Surgery, Pregnancy and PAH: The Unholy Alliance?*

John Granton, MD, University of Toronto, Toronto, Canada

#### Teaching/Discussion Points

- a. Reported perioperative morbidity/mortality with cardiac and non-cardiac surgeries in PH patients
- b. Risks of anaesthesia and mechanical ventilation in PH patients
- c. Risks of certain operations in patients with PH (eg laparoscopy, lobectomy, orthopedic surgery)
- d. Preoperative considerations and planning (Principles of intra- and post-operative management)
- e. Risk and outcomes of PAH and pregnancy

## SUMMARY / EVALUATION

Length: 2 mins

Colin Church, BSC(hons), MBChB, FRCP, PhD, Program Chair

Golden Jubilee National Hospital, Glasgow, UK