



**ISHLT ACADEMY
MASTER CLASS IN
PULMONARY HYPERTENSION**

TUESDAY, APRIL 4, 2017

**SEAPORT F
SEAPORT G**

2PM–7PM

SCIENTIFIC PROGRAM CHAIRS

Co-Chair: Myung Park, MD
Houston Methodist, Houston, TX, USA

Co-Chair: Ioana Preston, MD
Tufts Medical Center, Boston, MA, USA

SCIENTIFIC PROGRAM COMMITTEE

Members: Robert Frantz, MD, Mayo Clinic, Rochester, MN, USA

Marco Guazzi, MD, PhD, IRCCS Pol San Donato, Milan, Italy

Manreet Kanwar, MD, Allegheny General Hospital, Pittsburgh, PA, USA

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

COURSE SUMMARY

The Pulmonary Hypertension (PH) Master Class is intended for members with higher levels of expertise (completed the core curriculum course on PH and/or primary practice in PH \geq 5 years) who have managed patients with one or more of the topics intended for discussion. The course setting will generate a highly interactive environment composed of a smaller group of individuals designed to enhance individual expertise and network development. Utilizing the concept of “convergent discussion” and the technique of “audience response system,” faculty moderators will use complex situations and controversial statements during practical case presentations in order to lead the group through active audience participation, towards specific answers based on practice gaps and learning objectives.

The four topics covered include: Advanced Pulmonary Arterial Hypertension (PAH Group 1); PH Due to Left Heart Failure (PH Group 2); PH Due to Advanced Lung Disease (PH Group 3); and Chronic Thromboembolic PH (CTEPH, PH Group 4). The topics chosen will cover the major aspects of various forms of PH and highlight the significant differences in the diagnosis and management of these entities.

One to two references on each topic will be recommended reading to participants in preparation for this class. We anticipate that this method of collaborative and interactive learning will lead to application and integration of new knowledge into participant practice.

PRACTICE GAPS

- 1.** Advanced medical therapies and treatment algorithms for PAH have evolved tremendously in the recent years. The management of patients with advanced PAH and acute (or acute on chronic) right heart failure poses real challenges: their mortality is extremely high and salvage therapies used as bridge to a successful transplantation are complex. The implementation of advanced supportive measures in a failing right heart such as parenteral prostanoid replacement therapies, inotropic support and ECMO are often delayed or never utilized in advanced PAH patients. Recently published literature identified significant gaps in the recognition of signs that require adjustment of therapies in PAH management, including usage of parenteral therapies. Therefore, there is a real practice gap that limits their appropriate use in selected patients.
- 2.** The presence of right ventricular dysfunction in the setting of systolic heart failure (PH due to heart failure with reduced ejection fraction, PH-HFrEF) represents a complex challenge for practitioners. The use of advanced support systems such as LVADs in the setting of right heart failure needs very careful consideration, including detailed evaluation of the RV function with noninvasive and invasive tests; heart failure specialists may lack the intricate details of these complicated patients and will benefit from the expertise of our speakers in addressing these challenging and commonly encountered issues. Lastly, PH from left ventricular diastolic dysfunction (PH-HFpEF) is often refractory to currently available therapies and its management is very challenging. Moreover, therapies that are effective for PAH Group 1 have not been carefully evaluated in PH due to left heart disease (PH Group 2). Therefore, a deep understanding of the pathophysiology of PH Group 2 and how it differs (or not) from PAH Group 1 is necessary before making a decision of whether or not to treat.
- 3.** PH due to advanced lung disease (PH Group 3) occurs in the vast majority of patients being evaluated for lung transplant and is associated with increased mortality. Identification of PH, correct determination of PH type and decision whether or not to treat are complex aspects of care of patients with advanced lung disease. Because many patients in this category are being diagnosed when PH is already advanced, there is a clear gap in early recognition of PH Group 3. Assessment of patients with lung disorders such as pulmonary fibrosis, COPD and sarcoidosis for the presence and severity of PH is therefore an important step in improving their management.
- 4.** Annual incidence rates of deep venous thrombosis (DVT) and pulmonary embolism (PE) are approximately 0.5 to 1.0 per 1000 inhabitants and CTEPH develops in approximately 2-15% of patients who had a PE. Therefore, the estimated prevalence of CTEPH is high. Several reports highlight the significant gaps that exist between guidelines and clinical practice in regards diagnostic approaches and management of CTEPH, such as the lack of utilization of the ventilation/perfusion scan for screening and delay or omission of referral to a specialized surgical center for evaluation of operability. Therapies for CTEPH have been recently diversified and include sophisticated surgical approaches such as pulmonary endarterectomy, balloon angioplasty and medical treatment. Therefore, reviewing and clarifying the diagnostic algorithm, as well as different therapeutic approaches will be of great benefit for physicians and allied health care practitioners.

EDUCATIONAL GOALS

The overarching goal is to provide an advanced learning opportunity for specialists in the field of PH on the treatment of PAH Group 1, as well as PH in the setting of advanced left heart and lung disease and in chronic thromboembolic PH; all these entities have very different approaches in regards to management.

TARGET AUDIENCE

Pulmonologists/Respirologists, cardiologists, thoracic surgeons, nurses, physician assistants and allied health professional with experience in PH. The course is intended for health care professionals with primary practice that is focused in PH for at least 5 years or completed the ISHLT core curriculum course on PH.

LEARNING OBJECTIVES

Upon completion of the Master Class, participants will be able to:

1. Initiate advanced therapies for PAH
2. Apply advanced supportive measures in severe right heart failure
3. Manage complex patients with HFpEF and HFrEF complicated with PH
4. Understand pathophysiology, correctly diagnose and manage PH associated with advanced lung disease of different etiologies, such as emphysema, pulmonary fibrosis or sarcoidosis.
5. Correctly define the type and severity of vascular compromise in CTEPH
6. Determine the best therapeutic option in CTEPH, such as surgical eligibility, medical treatment or invasive nonsurgical approaches (balloon angioplasty).

ACCREDITATION STATEMENT

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

CREDIT DESIGNATION STATEMENT

ISHLT designates this live activity for a maximum of 4.5 *AMA PRA Category 1 Credits*.™ Physicians should claim only the credit commensurate with the extent of their participation in the activity.

ANCC CREDIT

Amedco is accredited as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation.

This course is co-provided by Amedco and the International Society for Heart and Lung Transplantation (ISHLT). Maximum of 4.25 contact hours.

ACPE CREDIT

This activity may be eligible for ACPE credit, see final CPE activity announcement for specific details.

DISCLOSURE

Current guidelines state that participants in CME activities must be made aware of any affiliation or financial interest that may affect the program content or a speaker's presentation. Planners, Faculty and Chairs participating in this meeting are required to disclose to the program audience any real or apparent conflict(s) of interest related to the content of their presentations or service as Chair/Planner. These disclosures will be distributed at the meeting. Additionally, all speakers have been asked to verbally disclose at the start of their presentation if a product they are discussing is not labeled for the use under discussion or is still investigational.

SCIENTIFIC PROGRAM SCHEDULE



1:15 PM – 2:00 PM
REGISTRATION AND COFFEE

2:00 PM – 2:15 PM
WELCOME AND INTRODUCTIONS
Myung Park, MD, Houston Methodist,
Houston, TX, USA
Ioana Preston, MD, Tufts Medical Center,
Boston, MA, USA

2:15 PM – 3:15 PM
SMALL GROUP INTERACTIVE
DISCUSSION A: Advanced PAH:
Challenges In Management
Moderator: **Robert Frantz, MD**

2:15 PM Case Scenario: Idiopathic PAH
with Advanced Right Ventricular Failure
Paul Corris, MB FRCP, Freeman Hospital,
Newcastle Upon Tyne, United Kingdom

Teaching/Discussion Points

- Understanding of optimal management strategies for RV failure (diuretics, inotropes, pressors).
- Discuss the role of atrial septostomy and ECMO.
- Timing of lung transplantation and factors impacting outcome.

2:45 PM Case Scenario: PAH with Suboptimal Control on Double Combination Therapy: What Next?
Stephan Rosenkranz, MD, University Heart Center, Cologne, Germany

Teaching/Discussion Points

- Utilization of optimal therapy lags behind guideline recommendations.
- Recognition of disease progression requires sophisticated integration of clinical, imaging and hemodynamic parameters that is insufficiently employed.
- Discussion of when to initiate parenteral therapy and various options available.
- Recognition of practice gaps suggesting that many PAH patients with advanced disease are not initiated on parenteral therapies and discussion of possible factors influencing this outcome.

2:15 PM – 3:15 PM
SMALL GROUP INTERACTIVE
DISCUSSION B: Challenges in PH Due to Left Heart Disease: An Increasingly Recognized Complication
Moderator: **Marco Guazzi, MD, PhD**

2:15 PM Case Scenario: PH Due to Heart Failure with Reduced Ejection Fraction
Ray Benza, MD, Allegheny General Hospital, Pittsburgh, PA, USA

Teaching/Discussion Points

- Hemodynamic definition and its limitations
- Epidemiology and phenotypes. Understanding the role of the right ventricle in HFrEF
- Pathophysiology of PH-HFrEF and the importance of mitral regurgitation
- Discussion of transplant and advanced support in the presence of PH
- Targeting the pulmonary microcirculation; a critical appraisal of current therapies for PH-HFrEF

2:45 PM Case Scenario: PH Due to Heart Failure with Preserved Ejection Fraction
Dario Vizza, MD, University of Rome, Rome, Italy

Teaching/Discussion Points

- Hemodynamic and clinical definitions;
- Epidemiology and phenotypic characteristics.
- Challenging LV filling and diastolic properties: exercise vs fluid loading
- Challenges in the management of patients with HFpEF and significant RV dysfunction. Discussion whether targeting the pulmonary vasculature is effective and safe.

3:20 PM – 4:20 PM
SMALL GROUP INTERACTIVE
DISCUSSION ROTATIONS
A AND B REPEATED

4:20 PM – 4:45 PM
COFFEE BREAK

4:45 PM – 5:45 PM
SMALL GROUP INTERACTIVE
DISCUSSION C: PH Due to Advanced
Lung Disease
Moderator: Steven Nathan, MD

4:45 PM Case Scenario: PH Due to
Combined Chronic Obstructive Lung
Disease and Pulmonary Fibrosis
Fernando Torres, MD, University of Texas
Southwestern Medical Center, Dallas, TX, USA

Teaching/Discussion Points

- Understand the implications of PH in diffuse parenchymal lung disease.
- Epidemiology: advances in screening techniques and diagnostic algorithms.
- Interpretation of hemodynamic testing in the setting of advanced lung disease.
- Challenges in the treatment of advanced pulmonary vascular disease in patients with COPD and IPF.
- Clinical trial conundrums.

5:15 PM Case Scenario: PH Associated
with Sarcoidosis
Oksana Shlobin, MD, Inova Fairfax Hospital,
Falls Church, VA, USA

Teaching/Discussion Points

- Is PH complicating sarcoidosis different to the PH of other forms of diffuse parenchymal lung disease?
- What factors are unique to sarcoidosis? If PH in sarcoidosis similar to PAH Group 1?
- Epidemiology: the role of PH screening in sarcoidosis.
- Implications with regards to transplantation.
- Treatment options, limitations of current therapies.

4:45 PM – 5:45 PM
SMALL GROUP INTERACTIVE
DISCUSSION D: CTEPH
Moderator: Manreet Kanwar, MD

4:45 PM Case Scenario: CTEPH with
Mixed Type 3 and Type 4 Disease
Irene Lang, MD, Medical University of
Vienna, Vienna, Austria

Teaching/Discussion Points

- Distal disease: What is the right therapeutic approach?
- Is there a role for combined surgical (PTE) and medical (BPA) approach in type 3/4 combined disease?
- Surgical challenges in patients with distal disease.
- Optimizing patient selection for surgical candidacy in patients with high pulmonary vascular resistance and distal disease.

5:15 PM Case Scenario: CTEPH with
Discordant Diagnostic Results Between
Imaging, Hemodynamics and Clot
Burden
William Auger, MD, University of California
San Diego Medical Center, San Diego, CA, USA

Teaching/Discussion Points

- Patient selection for optimal therapeutic options for CTEPH.
- Predicting surgical outcomes from pre-op data.
- Diseases that mimic CTEPH.
- Identifying patients with likely post PTE persistent PH.

5:50 PM – 6:50 PM
SMALL GROUP INTERACTIVE
DISCUSSION ROTATIONS
C AND D REPEATED

6:50 PM – 7:00 PM
CLOSING REMARKS
Myung Park, MD, Houston Methodist,
Houston, TX, USA
Ioana Preston, MD, Tufts Medical Center,
Boston, MA, USA