

SAPH2019

The 12th Annual Conference of
The Saudi Association for Pulmonary Hypertension

14-16 FEBRUARY 2019 | GULF HOTEL, BAHRAIN

FINAL PROGRAM

ORGANIZED BY:



IN COLLABORATION WITH:



PVRI
Pulmonary Vascular
Research Institute

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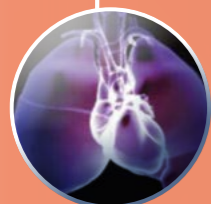
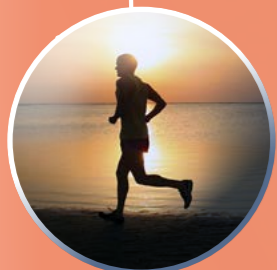
About Us

The Saudi Association for Pulmonary Hypertension (SAPH) is a medical and research body that is considered as a part of the Saudi Thoracic Society (STS) and is devoted to increase the awareness and knowledge of Pulmonary Vascular Diseases (PVD), and to facilitate advances in the treatment of affected people within the Kingdom of Saudi Arabia.



Objectives

- To investigate the local incidence, prevalence and pathobiology of PVD in Saudi Arabia
- To identify local characteristics of the illness including its morbidity and mortality
 To develop national guidelines for the diagnosis and treatment of PVD
- To provide expertise that will address issues related to the education and training of healthcare professionals in the field of PVD
- To focus on improving the treatment of PVD, by promoting basic and clinical research
- To promote public awareness through different means including print and electronic media and public seminars



Dear Colleagues,

On behalf of the Scientific and Organizing Committee, it gives us great pleasure to invite you to SAPH2019, the 12th Annual Conference of the Saudi Association for Pulmonary Hypertension which will be held at Gulf Hotel, Bahrain on 14-16 February 2019.

SAPH2019 aims to create a platform for regional medical professionals to engage in exchange of knowledge and expertise on the latest and best practice methods in pulmonary hypertension.

As a continuation of successful collaboration between the Saudi Association for Pulmonary Hypertension (SAPH) and the Pulmonary Vascular Research Institute (PVRI), it has been proven that it is highly successful and also helped in introducing physicians in the Middle East and North African countries to the world experts in the field of Pulmonary Hypertension and to the most advanced development in understanding and managing Pulmonary Hypertension.

The scientific committee is planning a comprehensive Scientific Program covering all aspects of Pulmonary Vascular Diseases in both Adult & Children, from the basics of Pathobiology and Pathophysiology, Diagnosis, Classification and Management to the most advanced development in Therapies and Interventional Modalities. The program will be covered by the internationally recognized leaders in the field of Pulmonary Hypertension.

The conference will be complemented by an exhibition to showcase and meet the leaders in the industry.

We look forward to welcoming you to SAPH2019 in Bahrain.

With my best regards,



Abdullah M. Aldalaan, MD
 Chairman, the 12th Annual Conference of Saudi Association for Pulmonary Hypertension (SAPH2019)
 President, Saudi Association for Pulmonary Hypertension
 Consultant Pulmonologist & Intensivist
 Department of Medicine
 King Faisal Specialist Hospital and Research Centre
 Riyadh, Saudi Arabia

Bahrain - which means "two seas" - was one of the first states in the Gulf to discover oil and to build a refinery. The Kingdom of Bahrain is an archipelago of 33 islands covering a total area of 741 square kilometers in the Arabian Gulf, located east of the Kingdom of Saudi Arabia and northwest of Qatar. The capital of Bahrain is Manama, and the population of Bahrain is approximately 1.41 million.

Bahrain was one of the first countries in the Middle East to discover crude oil. Though oil revenues continue to provide a sizeable proportion of Bahrain's gross domestic product (GDP), the country is expanding in the financial services, real estate and construction, and manufacturing sectors. Bahrain's GDP grew by 2.5% in 2017 despite a stagnation of the oil sector. Bahrain implemented the first Free Trading Agreement with the United States. Bahrain is also a member of the Cooperation Council for the Arab states of the Gulf (GCC).

Climate

The climate of the small archipelago of Bahrain is desert, mild in winter and very hot in summer. In winter, from December to February, at times there can be even warm days, with peaks around 30 °C (86 °F), when the wind blows from the south, but in these months this wind is rare. Sometimes, especially in January and February, cold air masses from the north can bring some cool and windy days, in which the daytime temperature can drop to around 15 °C (59 °F), and that of the night to around 10 °C (50 °F). Summer in Bahrain is very hot and sunny, with highs around 38/40 °C (100/104 °F) from June to September; lows are around 29/32 °C (84/90 °F) from June to September, and the humidity from the Persian Gulf makes the heat hard to bear. August and September are the months with the worst combination of humidity and temperature. However, hotels, offices, and restaurants are equipped with air conditioning.

Language

Arabic is the official language and the language of daily life. English is understood in many places and Farsi and Urdu also are spoken by the large numbers of Indian and Persian residents.

Clothing

Most men wear a traditional long robe called a thobe. Wealthier people tend to wear thobes tailored in a more Western style, with side and breast pockets and collars and French cuffs. Men also wrap their heads with a scarf called a Gutra. Women cover their clothes with the traditional black cloak, which goes over the head, and wear a veil of thin black gauze over the face. Some

younger women in the cities leave their faces or even their heads, uncovered, but this is rare. Bahrain's cultural name is Bahraini.

Currency

The Bahraini dinar is the world's second most valuable currency and the country has a growing and increasingly diversified economy. Currency Code is BHD. The Bahraini dinar is subdivided into 1000 fills. And is usually written with three decimal points (For example- BD 2.103)

Photography

As for photography, obtaining permission prior to photographing buildings or individuals is a 'must'. In case you are subject to Bahraini court orders arising out of indebtedness, or other legal disputes, you may be prevented from departing Bahrain until your cases are resolved.

Landmarks and architecture

King Fahad Causeway

This giant cultural landmark worth \$1billion links Saudi Arabia with Bahrain and is one of the longest bridges between the two nations. Unveiled in 1986, the causeway helped end the separation of 25,000-year between the two nations.

King Fahad Causeway is actually a combination of several bridges and dams. The five bridges that were completed as part of this project had a combined length of 12,430 meters. The Causeway actually comprises three parts, once extending from Khobar to man-made island of Saudi Arabia close to Bahrain border. Another one begins from this artificial island to Umm-al-Nasan, and the third one joins Umm-al-Nasan to the main island of Bahrain.

Bahrain World Trade Center

The Bahrain World Trade Center (Bahrain WTC) is a 240m tall twin tower complex located in proximity to King Faisal Highway and is ranked the second tallest building in Bahrain, following the twin towers of Bahrain Financial Harbour. The towers were built in 2008 by Atkins, the multinational architectural firm. This is also the first skyscraper in the world to incorporate wind turbines in its design. The two towers are linked by three sky-bridges, each holding a 225kw wind turbine, totaling 675kw wind power production. It is said that the wind turbines contribute to 11 to 15 percent of the total power consumption of the towers'.



Badges:

Name badges must be visible and used at all times, anywhere at the conference venue, and off- site social activities.

Colors:	Description:
Green:	Faculty (all access)
Red:	Delegate (all access)
Eggplant:	Exhibitor (all access)
Yellow:	Organizer (all access)

CME Certification:

This Conference is accredited by The Saudi Commission for Health Specialties (SCFHS) for 18 CMEs. In order to receive the CMEs, it is required to fill in the Evaluation and CME Acquisition Form which will be available at the Registration Desk throughout the conference.

Though Certificate of Attendance hard copies are not accepted by the SCFHS, but if you like to have one, kindly request through our website.

Conference Packets:

Conference Packets will be distributed to registered participants at the Registration Desk.

Faculty Check in:

There is a dedicated faculty lounge & preview room for faculty's registration and badge collection and is operational at the same time as the registration desks.

Food & Beverage:

Coffee breaks and Lunch will be open to registered delegates with conference badge. The hotel also offers a variety of all-day dining restaurants to choose from.

Automated Teller Machines (ATM):

There is an ATM located in the Hotel Lobby

Rules:

Smoking Policy in the Hotel: The entire hotel is non smoking. Mobile Phones- Delegates are kindly requested to keep their mobile phones in the off mode in meeting rooms when scientific sessions are in progress.

Parking:

24 hours valet parking is available at the Conference venue.

Prayer Room:

Prayer rooms are available in the Event Centre.

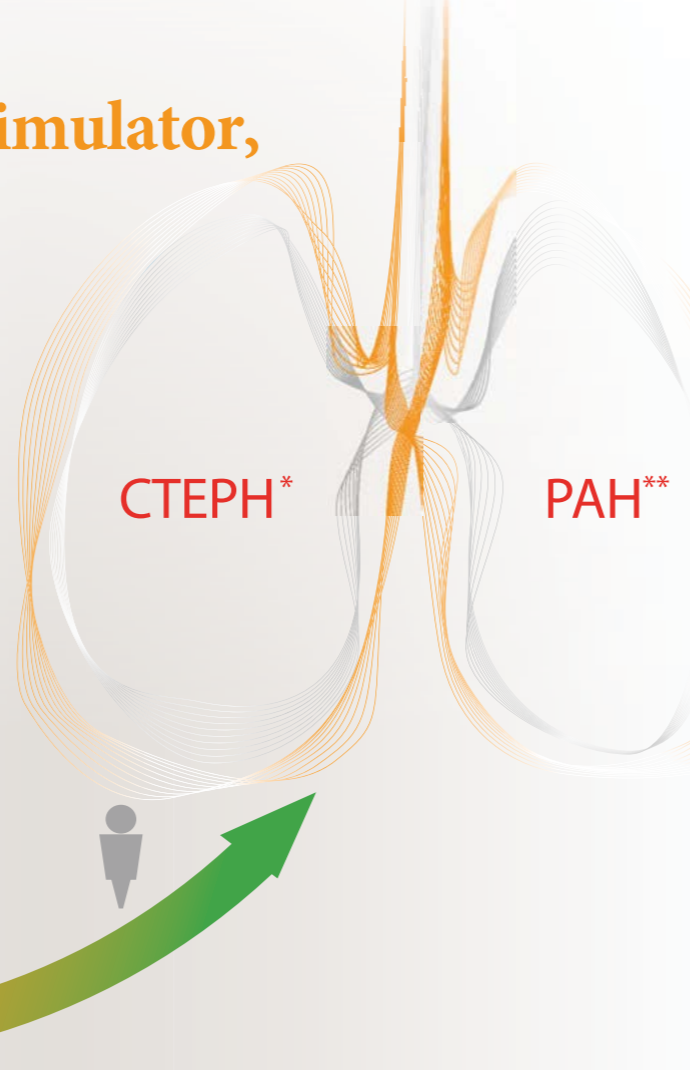
Evacuation Assembly Point:

In case of an emergency evacuation procedure please proceed in an orderly fashion to the open area in front of the Events Centre. Please follow the instructions of the Hotel Staff Wardens at all times.



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*CTEPH: inoperable or persistent/recurrent after surgery
 **PAH: in monotherapy or in combination with ERA
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References:
 1-Ghofrani HA et al. Future Cardiol 66-155: 6; 2010.

Abbreviated Prescribing Information: Adempas 0.5 mg, Adempas 1 mg, Adempas 1.5 mg, Adempas 2 mg, Adempas 2.5 mg Film Coated Tablet. Composition: What Adempas contains: The active substance is riociguat. Each tablet contains 0.5 mg, 1 mg, 1.5 mg, 2 mg or 2.5 mg riociguat. The other ingredients are: Tablet core: cellulose microcrystalline, croscopolidone, hypromellose, lactose monohydrate, magnesium stearate and sodium laurylsulfate (see end of section 2 for further information on lactose). Film-coat*: hydroxypropylcellulose, hypromellose, propylene glycol and titanium dioxide (E 171). *1 mg, 1.5 mg, 2 mg and 2.5 mg tablets also have: ferric oxide yellow (E 172) *2 mg and 2.5 mg tablets also have: ferric oxide red (E 172). **Indications:** Adempas contains the active substance riociguat. Riociguat is a type of medicine called a guanylate cyclase (sGC)-stimulator. It works by widening the pulmonary arteries (the blood vessels that connect the heart to the lungs), making it easier for the heart to pump blood through the lungs. Adempas can be used to treat adults with certain forms of pulmonary hypertension, a condition in which these blood vessels become narrowed, making it harder for the heart to pump blood through them and leading to high blood pressure in the vessels. Because the heart must work harder than normal, people with pulmonary hypertension feel tired, dizzy and short of breath. By widening the narrowed arteries, Adempas leads to an improvement in the ability to carry out physical activity. Adempas is used in either of two types of pulmonary hypertension: **Chronic thromboembolic pulmonary hypertension (CTEPH).** In CTEPH, the blood vessels of the lung are blocked or narrowed with blood clots. Adempas can be used for patients with CTEPH who cannot be operated on, or after surgery for patients in whom increased blood pressure in the lungs remains or returns. **Certain types of pulmonary arterial hypertension (PAH).** In PAH, the wall of the blood vessels of the lungs are thickened and the vessels become narrowed. Adempas is only prescribed for certain forms of PAH, i.e. idiopathic PAH (the cause of PAH is unknown), heritable PAH and PAH caused by connective tissue disease. The doctor will check this. Adempas can be taken alone or together with certain other medicines used to treat PAH. **Contraindications:** Adempas should not be taken: if patient is taking certain medicines called PDE-5 inhibitors (e.g. sildenafil, tadalafil, vardenafil). These are medicines used for the treatment of high blood pressure in the arteries of the lungs (PAH) or erectile dysfunction. If patient has severe liver problems (severe hepatic impairment, Child Pugh C). If patient is allergic to riociguat or any of the other ingredients of this medicine (listed in section 6). If patient is pregnant. If patient is taking nitrates or nitric oxide donors (such as amyl nitrite) in any form, medicines often used to treat high blood pressure, chest pain or heart disease. This also includes recreational drugs called poppers. If patient has low blood pressure (systolic blood pressure less than 95 mmHg) before starting first treatment with this medicine. If patient has increased pressure in the pulmonary circulation associated with scarring of the lungs, of unknown cause (idiopathic pulmonary pneumonia). If any of these applies, patient should talk to the doctor first and should not take Adempas. **Warnings and precautions:** patient should talk to the doctor or pharmacist before taking Adempas if: patient has recently experienced serious bleeding from the lung, or if patient has undergone treatment to stop coughing up blood (bronchial arterial embolisation), patient takes blood-thinning medicines (anticoagulants) since this may cause bleeding from the lungs. Doctor will regularly monitor the patient. Patient feels short of breath during treatment with this medicine, this can be caused by a build-up of fluid in the lungs. Patient should talk to the doctor if this happens. patient has problems with the heart or circulation, patient is older than 65 years. Patient's kidneys do not work properly (creatinine clearance < 30 ml/min) or if patient is on dialysis as the use of this medicine is not recommended. patient has moderate liver problems (hepatic impairment, Child Pugh B). patient starts or stop smoking during treatment with this medicine, because this may influence the level of riociguat in the blood. Patient will receive Adempas only for special types of pulmonary arterial hypertension (PAH), 1. There is no experience in the use of Adempas in other types of PAH. Use of Adempas in other types of PAH is therefore not recommended. Doctor will check if Adempas is suitable. **Children and adolescents:** The use of Adempas in children and adolescents (under 18 years of age) should be avoided. **Other medicines and Adempas Doctor or pharmacist should be informed if patient is taking, have recently taken or might take any other medicines, in particular, medicines used for: high blood pressure or heart disease (such as nitrates and amyl nitrite) in any form, as patient must not take these medicines together with Adempas. high blood pressure in the lung vessels (the pulmonary arteries), as patient must not take certain medicines (sildenafil and tadalafil) together with Adempas. Other medications for high blood pressure in the lung vessels (PAH), such as bosentan and iloprost, can be used with Adempas, but patient should still tell the doctor. erectile dysfunction (such as sildenafil, tadalafil, vardenafil), as patient must not take these medicines together with Adempas. fungal infections (such as ketoconazole, itraconazole), HIV infection (such as ritonavir), epilepsy (e.g. phenytoin, carbamazepine, phenobarbitone), depression (St. John's Wort), preventing rejection of transplanted organs (cyclosporin), joint and muscular pain (niflumic acid), cancer (such as erlotinib, gefitinib), stomach disease or heartburn (antacids such as aluminium hydroxide/magnesium hydroxide). These anticancer medicines should be taken at least two hours before or one hour after taking Adempas. nausea, vomiting (feeling or being sick) (such as granisetron). **Smoking** If patient smokes, it is recommended that he/she stops, as smoking may reduce the effectiveness of these tablets. Patient should tell the doctor if he/she smokes or if he/she stopped smoking during treatment. **Pregnancy and breast-feeding** Pregnancy: Adempas must not be used during pregnancy. If there is a chance the patient could become pregnant, she should use reliable forms of contraception while she is taking these tablets. Patient is also advised to take monthly pregnancy tests. If she is pregnant, she thinks she may be pregnant or is planning to have a baby, she should ask her doctor or pharmacist for advice before taking this medicine. **Breast Feeding:** If patient is breast-feeding or planning to breast-feed, she should ask her doctor or pharmacist for advice before taking this medicine because it might harm the baby. Doctor will decide with her if she should stop breast-feeding or stop treatment with Adempas. **Driving and using machines** Adempas has moderate influence on the ability to drive and use machines. It may cause side effects such as dizziness. patient should be aware of the side effects of this medicine before driving or using machines. Adempas contains lactose: if patient has been told by a doctor that he/she has an intolerance to some sugars, he/she should tell treating doctor before taking these tablets. **Possible side effects:** Like all medicines, this medicine can cause side effects although not everybody gets them. The most serious side effects are: coughing up blood (common side effect), acute bleeding from the lungs may result in coughing up blood, cases with fatal outcomes were observed (uncommon side effect). If this happens, patient should contact doctor immediately as patient may need urgent medical treatment. Overall list of possible side effects: **Very common:** may affect more than 1 in 10 people headache, dizziness, indigestion, swelling of limbs, diarrhoea, feeling or being sick. **Common:** may affect up to 1 in 10 people: inflammation in the digestive system, reduction of red blood cells (anaemia) seen as pale skin, weakness or breathlessness, awareness of an irregular, hard, or rapid heartbeat, feeling dizzy or faint when standing up (caused by low blood pressure), coughing up blood, nose bleed, difficulty breathing through nose, pain in the stomach, intestine or abdomen, heartburn, difficulty in swallowing, constipation, bloating. **Uncommon:** may affect up to 1 in 100 people: acute bleeding from the lungs. Patient should contact the doctor immediately as patient may need urgent medical treatment. **Reporting of side effects:** If patient gets any side effects, patient should talk to the doctor or pharmacist. This includes any possible side effects not listed in this leaflet. By reporting side effects, more information on the safety of this medicine can be provided. **Marketing Authorisation Holder:** Bayer Pharma AG, 13342 Berlin, Germany. **This leaflet was last revised in August, 2016****



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 Tel.: +966 12 657 16 75
 Fax: +966 12 653 49 92
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Saudi Thoracic Society

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Saudi Thoracic Society (STS) is a scientific foundation and a leading resource for improvement of lung health in Saudi Arabia. Its mission is to promote the prevention, diagnosis, and treatment of chest diseases through leadership, education, research, and communication. STS was established in 2002 and it is affiliated with King Saud University in Riyadh.

The specific aims of the Society are:

Promoting and coordinating activities in the field of respiratory medicine.

Fostering research activities in the field of respiratory medicine.

Organizing and coordinating regular national and regional meetings.

Publishing a newsletter and a journal of international repute.

Publishing and updating clinical practice guidelines in the field of respiratory medicine.

You are invited to become a member of the Joint STS-ERS Membership. You will be a valuable member of our mailing list and it is important to keep you informed on relevant updates, articles, news and announcements.

To Register, kindly visit our website (www.saudithoracic.com)

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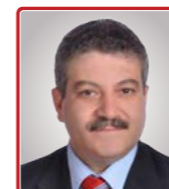
Abdullah M. Aldalaan, MD
Chairman, the 12th Annual Conference of Saudi Association for Pulmonary Hypertension (SAPH2019)
President, Saudi Association for Pulmonary Hypertension
Consultant Pulmonologist & Intensivist
Department of Medicine
King Faisal Specialist Hospital and Research Centre
Riyadh, Saudi Arabia



Nawal Al Gubaisi, MD, SSC-Med, SF-PD
Chairman, Scientific Committee
Consultant Pulmonologist
Pulmonary Hypertension Specialist
The Head of Pulmonary Medicine Unit
King Fahd Medical Military Complex
Al Khobar, Saudi Arabia



Hassan S. Alorainy, BsRC, RRT, FAARC
Executive Director, The 12th Annual Conference of Saudi Association for Pulmonary Hypertension (SAPH2019)
Senior Clinical Respiratory Specialist
Riyadh, Saudi Arabia



Hussam Sakkijha, MD
Consultant, Pulmonary, Critical Care and Sleep Medicine
King Fahad Medical City, Riyadh
Riyadh, Saudi Arabia



Majdy Idrees, MD, FRCPC, FPVRI
Adjunct Professor of Respiratory Medicine
University of British Columbia, Vancouver, Canada
Head, Pulmonary Vascular Unit
Prince Sultan Military Medical City
Riyadh, Saudi Arabia

MEMBERS



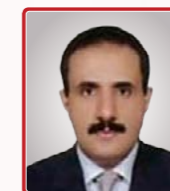
M. Ali Al Azem, MD, MHA, FCCP
Chairman, Department of Critical Care
Consultant, Pulmonary, Critical Care & Sleep Medicine
King Fahad Specialist Hospital, Dammam
Dammam, Saudi Arabia



Khalid Al Najashi, MD, MBBS
Consultant, Interventional ACHD and Pediatric Cardiology
Prince Sultan Cardiac Center
Prince Sultan Military Medical City- Riyadh
Riyadh, Saudi Arabia



Maha Al Dabbagh, MD
Pediatric Pulmonologist
King Fahd Armed Forces Hospital, Jeddah
Jeddah, Saudi Arabia



Saleh Aldammas, MD
Consultant, Pulmonary and Sleep Medicine
Prince Sultan Military Medical City
Riyadh, Saudi Arabia



Badr R. Al-Ghamdi, MD
Associate Professor and Consultant Pulmonologist
Chairman Department of Medicine
College of Medicine, King Khaled University
Director of Chest unit, Aseer Central Hospital
Abha, Saudi Arabia



Hanan Al Rayes, MD
Consultant Rheumatology,
President of Saudi Society for Rheumatology
Deputy Director of Medicine Department
Prince Sultan Military Medical City
Riyadh, KSA

EXECUTIVE COMMITTEE



Manal S. Al Hazmi, MD
Consultant, Pulmonary and Critical Care
Medicine
King Fahad Specialist Hospital, Dammam
Dammam, Saudi Arabia



Omar Tamimi, MD
Department of Pediatrics
Princess Nora bint Abdul Rahman University
Riyadh, Saudi Arabia



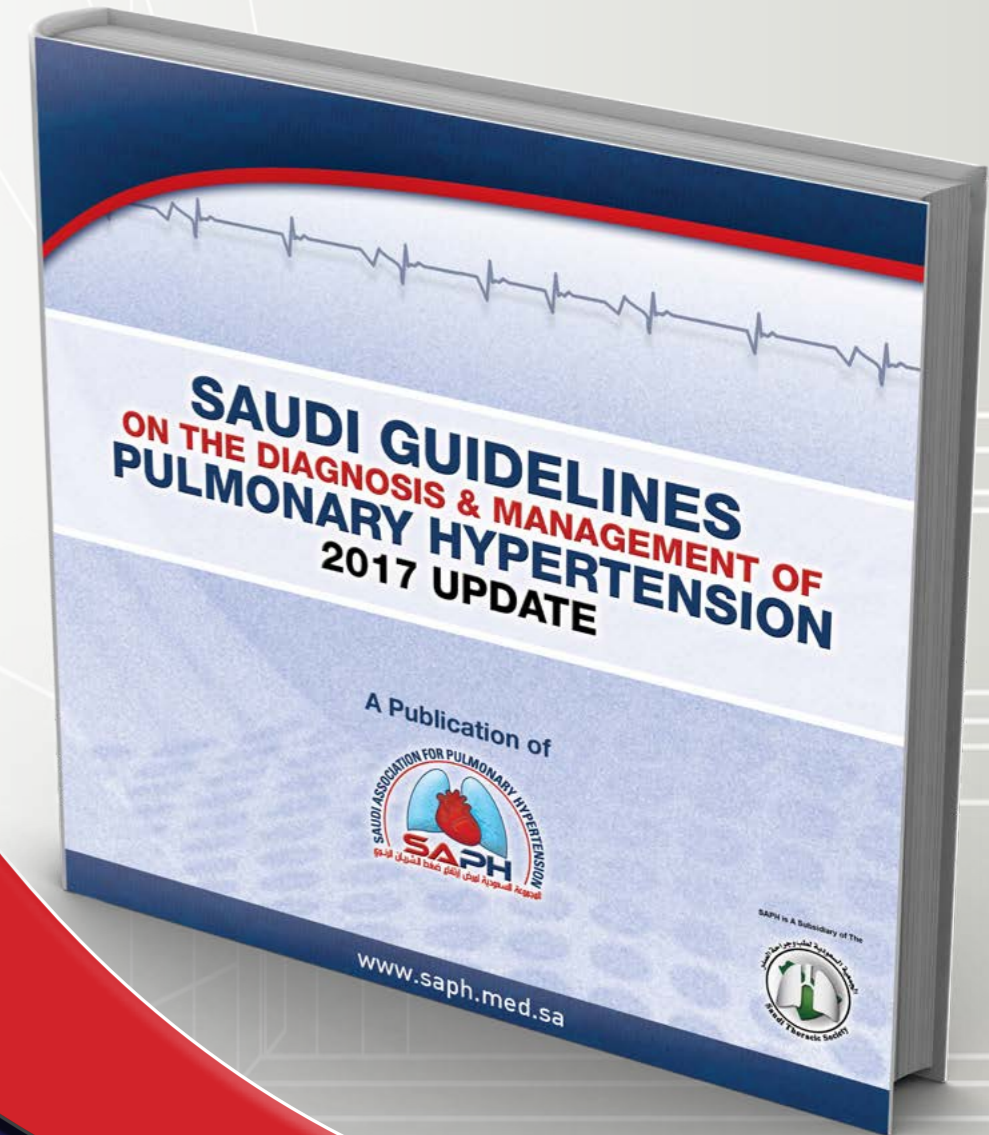
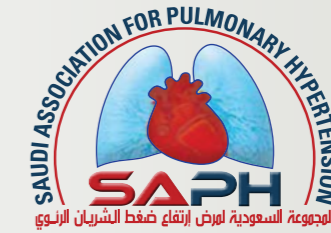
Abdulmajeed Al-Otay, MD
Consultant Pediatric Cardiologist,
Head of Cardiac Imaging Unit,
Prince Sultan Cardiac Center
Riyadh, KSA



Bader Alghamdi, MD
Consultant, Pulmonary Medicine and
Pulmonary Hypertension
Director, Internal Medicine Residency
Program, KAMC-Jeddah
Assistant Professor, Internal Medicine and
Pulmonary, KSAU-HS
Department of Medicine, Division of
Respirology
King Abdulaziz Medical City KAMC- Jeddah
Jeddah, Saudi Arabia



Hanaa Banjar, MD, FRCPC
Associate Professor of Pediatrics and
Pediatric Pulmonology, Alfaisal University
Consultant Pediatric Pulmonology,
Department of Pediatrics
King Faisal Specialist Hospital and
Research Centre
Riyadh, Saudi Arabia



SAUDI GUIDELINES ON THE DIAGNOSIS AND MANAGEMENT OF PULMONARY HYPERTENSION 2017 UPDATE

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SAPH2019

The 12th Annual Conference of
The Saudi Association for Pulmonary Hypertension

14-16 FEBRUARY 2019 | GULF HOTEL, BAHRAIN

CMEs Important Advisory

TOPICS:

- Updated Strategy on Diagnosis and Management of Asthma and COPD
- Current understanding of Interstitial Lung Disease
- Lung Infections
- Some other selected topics in Pulmonary Medicine

TARGET AUDIENCE:

- Adult/ Pediatric Pulmonologists
- Allergists & Immunologists
- Family Medicine & Primary Care Physicians
- Internists
- Fellows in Training
- Intensivists
- Allied Health and Nursing

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If Practicing in Saudi Arabia:

- **SCFHS** no longer accepts hard copy of Certificate of Attendance.
- **SCFHS** accepts only Electronic Submissions of **CMEs** done by the Saudi Thoracic Society.
- To acquire your **CMEs**, you must complete the Conference Evaluation Form and fill and sign the **CMEs** Acquisition Form at the Registration Desk.
- **SCFHS** has recently required all practitioners in Saudi Arabia to register on their website as "Mumaris". If you have not yet, you must register to enable us to add your **CMEs**.

For Hard Copy Certificate:

If practicing outside Saudi Arabia, or need a hard copy, please communicate with us after the conference via this email: (saphsts@gmail.com), we will send you the signed certificate electronically for you to print at your convenience.

DAY 1 - THURSDAY, 14 FEBRUARY 2019

EVENING SESSION

17:00 - 18:45 Registration

18:45 - 19:00

Opening Ceremony and Welcome Notes

Abdullah Aldalaan - KSA

19:00 - 20:30 Session 1: Keynote

Chairs: Khalid Bin Thani - Bahrain | Saleh Aldammas - KSA

19:00 - 19:30 | The Keynote Lecture: No Discovery and the Path to the Nobel Prize

Marc Humbert - France

19:30 - 20:00 | PAH; Year in Review

Hussam Sakkijha - KSA

20:00 - 20:30 | Q & A Panel Discussion

**20:30 DINNER LOCATION: GCC TERRACE SWIMMING POOL AREA
GULF HOTEL, BAHRAIN**

END OF 1ST DAY

DAY 2 - FRIDAY, 15 FEBRUARY 2019

MORNING SESSIONS

07:30 - 08:30 Registration

08:30 - 10:00 Session 2: PAH Basic Science

Chairs: Abdullah Aldalaan - KSA | Nazzareno Galiè - Italy

08:30 - 08:50 | The Complex Pathology of PAH

H. Ardeschir Ghofrani - Germany

08:50 - 09:10 | PAH Pathobiology; New Classes of Drugs in PAH Treatment

Marc Humbert - France

09:10 - 09:30 | PAH Genetics; Update and Future Role for new PAH Therapy Targets

Allan Lawrie - UK

09:30 - 09:50 | Phenotyping of PH, the Future Goal!

Majdy Idrees - KSA

09:50 - 10:00 | Q & A Panel Discussion

10:00 - 10:30 COFFEE BREAK

10:30 - 12:00 Session 3: 6th World PH Symposium

Chairs: Gérald Simonneau - France | Badr Al Ghamdi - KSA

10:30 - 10:50 | The New Threshold of PH Diagnosis; Is it Rationalized?

Gérald Simonneau - France

10:50 - 11:10 | Diagnosis Algorithm/Classifications; Key New Changes and Evidence

Luke Howard - UK

11:10 - 11:30 | Comparing Risk Stratification Strategies; Simplicity and/or Accuracy?

Oliver Sitbon - France

11:30 - 11:50 | The Final look at PAH Treatment Algorithm

Nazzareno Galiè - Italy

11:50 - 12:00 | Q & A Panel Discussion

12:00 - 13:30 LUNCH BREAK

DAY 2 - FRIDAY, 15 FEBRUARY 2019

AFTERNOON SESSIONS

13:30 - 15:00 Session 4: PAH/PH Treatment; Current Guidelines

Chairs: Jean Luc Vachiéry - Belgium | Nasser Albusaidi - Oman

13:30 - 13:50 | Treating PH Due to Left heart Diseases, Sum of all Evidence
Jean Luc Vachiéry - Belgium

13:50 - 14:10 | PH Due to Chronic Lung Diseases, Where do we Stand?
Luke Howard - UK

14:10 - 14:30 | PVOD; An Update; Genetics, Pathology and Beyond
Oliver Sitbon - France

14:30 - 14:50 | Insights from the SAUDI PH Registry (PAH Data)
Abdullah Aldalaan - KSA

14:50 - 15:00 | Q & A Panel Discussion

15:00 - 15:30 COFFEE BREAK

15:30 - 17:00 Session 5: Challenging Cases

Chairs: Discussants: Nazzareno Galiè - Italy | Olivier Sitbon - France | Majdy Idrees - KSA

15:30 - 16:00 | PAH in Pregnant Women
Fayez Alahmadi - KFSH&RC, Riyadh, KSA

16:00 - 16:30 | A Familial Pulmonary Arterial Hypertension Case
Hanan Fan - KFAFH, Jeddah, KSA

16:30 - 17:00 | A Case of Portopulmonary Hypertension
Mohammed Qasem - PSCC, Al Qassim, KSA

19:30 - 22:00 GALA DINNER AT RITZ CARLTON BAHRAIN (MASAYA PAVILION)

Buses will Leave from Gulf Hotel Main Entrance (in front of Lobby Area) at 19:30 Sharp

END OF 2ND DAY

DAY 3 - SATURDAY, 16 FEBRUARY 2019

MORNING SESSIONS

08:00 - 10:00 Session 6: PH in Pediatrics/ CHD-PAH/ CTD-PAH

Chairs: Mustafa Al Refae - KSA | Hana Banjar - KSA

08:30 - 08:50 | PAH in Pediatric Patients; A State of Art
Alessandra Manes - Italy

08:50 - 09:10 | Congenital Heart Defects; When to Close and when not to Close?
Hani Sabour - UAE

09:10 - 09:30 | CHD-PAH Burden and Treatment; A State of Art
Jean Luc Vachiéry - Belgium

09:30 - 09:50 | CTD-PAH; A State of Art
Nawal Al Gubaisi - KSA

09:50 - 10:00 | Q & A Panel Discussion

10:00 - 10:30 COFFEE BREAK

10:30 - 12:00 Session 7: CTEPH

Chairs: Eid Alqurashi - KSA | Maryam Alqaseer - KSA

10:30 - 10:50 | Pulmonary Vascular Injury, Role in CTEPH Development
H. Ardeschir Ghofrani - Germany

10:50 - 11:10 | BPA Technique, Patient Selection and Outcome
Nazzareno Galiè - Italy

11:10 - 11:30 | Medical Therapy of CTEPH; Update!
Bader Alghamdi - KSA

11:30 - 11:50 | Insights from the SAUDI PH Registry (CTEPH Data)
Sarfraz Saleemi - KSA

11:50 - 12:00 | Q & A Panel Discussion

12:00 CLOSING REMARKS | Abdullah Aldalaan - KSA

12:10 - Please be advised: Lunch will be served.

END OF 3RD DAY



Abdullah M. Aldalaan, MD

Chairman, The 12th Annual Conference of
Saudi Association for Pulmonary Hypertension (SAPH2019)
President, Saudi Association for Pulmonary Hypertension (SAPH)
Consultant Pulmonologist & Intensivist
King Faisal Specialist Hospital and Research Centre
Riyadh, Saudi Arabia

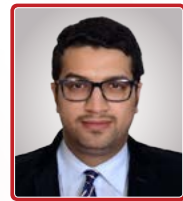
Dr. Abdullah M. Aldalaan graduated from King Saud University in Riyadh. He received his residency training at Duke University Medical Center in North Carolina, USA; and completed his fellowship training in Pulmonary and Critical Care at University of Virginia, USA. He obtained American Board of Internal Medicine, Pulmonary Medicine and Critical Care Medicine. Since then, he has been practicing as a Pulmonologist and Intensivist at King Faisal Specialist Hospital and Research Center (KFSH & RC) in Riyadh, Saudi Arabia.

He established the following at King Faisal Specialist Hospital & Research Centre:

- Lung Transplant Program, in 2003 (the first and only one among the Arab countries)
- Pulmonary Hypertension Treatment Program, the only program in the area which provides comprehensive diagnostic and therapeutic protocols for patients with pulmonary hypertension, which includes all available internationally recognized medical interventions in this field, and which has become a reference center in the whole area for both education and treatment.

His areas of interests are Lung Transplantation and Pulmonary Hypertension. However, he runs inpatient and outpatient Pulmonary Services at KFSH & RC which covers a wide range of pulmonary diseases. For 4 years from 2010 to 2014, he served as Section Head of the Pulmonary Medicine, Department of Medicine.

In addition to his clinical responsibilities, he is also the Director of Ambulatory Care Services. He is a Board Member of the Saudi Thoracic Society. He held several Scientific Committee positions and has participated in several research publications. He is also currently the President of Saudi Association for Pulmonary Hypertension (SAPH).



Fayeze Khalid S. AlAhmadi, MD

Pulmonary Fellow,
King Faisal Specialist Hospital and
Research Centre
Riyadh, Saudi Arabia

Dr. Fayeze AlAhmadi is currently a second-year fellow at King Faisal Specialist Hospital and Research Centre (KFSH&RC)-Riyadh. He completed the Residency Training Program and the Fellowship Training in Pulmonary Medicine at the same Hospital.

He attended several conferences and was very much active in presenting posters, he also presented "KFSH&RC CTEPH Experience" in the 10th Annual Central European PH Meeting, Vienna 2018. He is also a member and coordinator of Taiba Doctors Charitable Society.



Nazzareno Galiè, MD

Full Professor of Cardiology Alma Mater Studiorum,
University of Bologna
Director Post-graduate School of
Cardiovascular Diseases
Bologna, Italy

Nazzareno Galiè, MD, heads the Pulmonary Hypertension Centre at the Institute of Cardiology and is Associate Professor of Cardiology at the Medical Faculty of the University of Bologna, Italy. He also teaches at the Postgraduate Medical Schools of Cardiology, Pulmonary Diseases, and Rheumatology at the University of Bologna. He is Director of the International Master Degree in Pulmonary Vascular Diseases of the University of Bologna. He has authored 107 scientific publications indexed in Pub - Med on heart failure, heart transplantation, and Pulmonary Hypertension. Professor Galiè is a Scholar of the Italian Society of Cardiology,

Fellow of the European Society of Cardiology (FESC), and Honorary Fellow of the Royal College of Physicians (FRCP), UK. He is a Past-Chairman of the working group on Pulmonary Circulation of the European Society of Cardiology and of the joint task force of the European Society of Cardiology and the European Respiratory Society for the guidelines on Pulmonary Hypertension.



Nawal Al Gubaisi, MD, SSC-Med, SF-PD

Chairman, Scientific Committee,
Consultant Pulmonologist
Pulmonary Hypertension Specialist
The Head of Pulmonary Medicine Unit
King Fahd Medical Military Complex
Khobar, Saudi Arabia

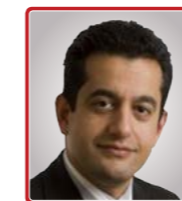
Dr. Nawal Al Gubaisi received her Medical Degree from King Faisal University. Following this, she did her postgraduate training both in Internal Medicine Program and Pulmonary Medicine from Saudi Board. Also, she got a HERMES European Diploma in Respiratory Medicine. She did her fellowship from UBC Canada along with training in Right Heart Catheterization. Currently, Dr. Nawal Al Gubaisi is the Head of Pulmonary Medicine Unit at King Fahd Medical Military Complex, Dhahran, Saudi Arabia. She is working as a Consultant Pulmonologist and is specialized in Pulmonary Hypertension.



Bader Alghamdi, MD

Assistant Professor, Consultant in Pulmonary Medicine,
Pulmonary Hypertension and Clinical Exercise Testing
King Saud bin Abdulaziz University for
Health Sciences (KAUH-HS)- Jeddah
Jeddah, Saudi Arabia

Dr. Alghamdi attended medical school at King Abdulaziz University in Jeddah. Following this, he completed his residency training at King Abdulaziz Medical City (KAMC)-Jeddah in Internal Medicine Program. Then, he completed his fellowship at Queen's University-Kingston-Canada in Pulmonary Medicine, Pulmonary Hypertension and Clinical Cardiopulmonary Exercise testing. Currently, Dr. Alghamdi is a Consultant in Pulmonary Medicine and Pulmonary Hypertension at KAMC-Jeddah, Director of the Internal Medicine residency program at KAMC-Jeddah and Assistant Professor in Internal Medicine and Pulmonary in King Saud bin Abdulaziz for Health since (KSAU-HS)- Jeddah.



Hossein Ardeschir Ghofrani, MD

Professor, Pulmonary Vascular Research,
Justus Liebig University, Giessen, Germany
Head, Pulmonary Hypertension Division,
Pulmonary Vascular Research Institute
University Hospital in Giessen

Giessen, Germany

Hossein A. Ghofrani received his medical degree from the Medical School at Giessen University in Germany. He is Professor for Internal Medicine at University Hospital Giessen and Marburg GmbH. He currently is Head of the Pulmonary Hypertension Division, Department of Internal Medicine, at Giessen. He also leads a collaborative group on Cardiopulmonary Vascular System research. In addition, he is Director of Pneumology at the Kerckhoff Heart and Lung Center in Bad Nauheim, Germany.

Prof. Ghofrani has participated in the therapeutic development of surfactant for the treatment of acute respiratory distress syndrome (ARDS); prostanoids, PDE inhibitors, combination therapies, and soluble guanylate cyclase activators and stimulators for pulmonary hypertension; endothelin antagonists for chronic lung disease and pulmonary hypertension; and tyrosine kinase inhibitors for pulmonary vascular diseases. He has received four awards for investigations in pulmonary vascular science and is a reviewer for several medical scientific journals including the American Journal of Respiratory and Critical Care Medicine, European Respiratory Journal, Circulation, and Lancet.



Luke Howard, MA, MB, BChir, DPhil, FRCP
Consultant Respiratory Physician
Imperial College Healthcare,
NHS Trust, London
London, United Kingdom

Dr. Luke Howard, DPhil, FRCP is a consultant respiratory physician who specializes in cardiopulmonary medicine and exercises physiology. He specializes in diseases of the pulmonary circulation, in particular, Pulmonary Hypertension and pulmonary embolism, and through his interest in exercise physiology has specific expertise in unexplained breathlessness. He works in close collaboration with his colleagues in cardiology, rheumatology, and hematology to provide a comprehensive assessment of conditions leading to exercise limitation. He consults for patients and athletes alike and has close links with the GB Rowing Team. He undertook his undergraduate training at the University of Oxford where he also completed his doctorate in altitude physiology and then his clinical training at the University of Cambridge, qualifying in 1996.

He trained in London and Cambridge, being appointed to the National Pulmonary Hypertension Service at Hammersmith Hospital in 2006 as a consultant with an honorary senior lecturer position at the National Heart and Lung Institute, Imperial College London. His research interests include iron physiology, cardiopulmonary hemodynamics, exercise physiology, Pulmonary embolism, and remote patient monitoring.



Marc Humbert, MD, PhD
University Paris-Sud - University Paris-Saclay
Head of the Department of Pulmonology and
Intensive Care Respiratory
Pulmonary Hypertension Reference Center
Assistance Publique Hôpitaux de Paris
LabEx LERMIT - DHU Thorax Innovation (TORINO)
Section Editor, European Respiratory Journal
Kremlin Bicetre, France

Marc Humbert, MD, PhD, is Professor of Respiratory Medicine at the South Paris University (Université Paris-Saclay) in Le Kremlin-Bicêtre, France since 1999. In addition to his academic responsibilities, he is the Director of the Severe Asthma Clinic and National Reference Centre for Pulmonary Hypertension, Department of Respiratory and Intensive Care Medicine, Hospital Bicêtre, Assistance Publique Hôpitaux de Paris, France. Along with membership to several scientific councils and institutes, Marc Humbert is the current Director of the INSERM Unit "Pulmonary Hypertension: Pathophysiology and Innovative Therapies" and of the "Thorax Innovation" University Hospital Department. He has been the Chief Editor of the European Respiratory Journal from 2013 to 2017. He has published more than 700 peer-reviewed articles, mostly in the field of pulmonary hypertension.

He is a Fellow of the European Respiratory Society (FERS Foundation Fellow) and has received several distinctions including the 1997 François Brenot Award and the 2006 Courmand Lecture Award from the ERS, the 2009 Descartes-Huygens Award from the Royal Netherlands Academy of Arts and Sciences, the 2016 Rare Disease Award of the Fondation de France (Fondation Eliane and Gérard Pauthier), and the 2018 ERS Award for Lifetime Achievement in Pulmonary Arterial Hypertension. Since 2017, Marc Humbert is the vice-coordinator of the European Reference Network for rare and low prevalence respiratory diseases (ERN-LUNG). Clarivate Analytics listed Marc Humbert as one of the world's highly cited researchers in the field of Clinical Medicine (2018).



Majdy Idrees, MD, FRCPC, FPVRI
Adjunct Professor of Respiratory Medicine
University of British Columbia, Vancouver, Canada
Head, Pulmonary Vascular Unit
Prince Sultan Military Medical City
Riyadh, Saudi Arabia

Dr. Idrees is an Adjunct Professor of Pulmonary Medicine at the University of British Columbia, Vancouver, Canada and the former Head of Pulmonary Division and the Director of the Pulmonary Vascular Disease Unit at Prince Sultan Military Medical City (Armed

Forces Hospital), Riyadh, Saudi Arabia. He was the first to establish the nucleus of pulmonary vascular diseases/pulmonary hypertension medicine in the Arab Gulf countries and is the founder, and the former head, of the Saudi Association for Pulmonary Hypertension. He received his MBBS. degree from King Saud University in Riyadh, Saudi Arabia, and did his postgraduate training in both Internal Medicine and Pulmonary Medicine at the University of British Columbia, Canada from 1992 - 1997. He had his American Board degree in Pulmonary Medicine in 1996, and the Canadian Board in Pulmonary Medicine in 1997.

His major area of research is related to pulmonary hypertension and pulmonary vascular diseases, and also to airways diseases. He has more than 50 publications in peer reviewed journals that include book chapters, original papers, and review articles. He is the primary author of the Saudi Guidelines for the management of Pulmonary Hypertension and a coauthor of the Saudi Guidelines in both Asthma and COPD. He is a manuscript reviewer and co-editor of many medical journals.

Dr. Idrees was awarded and recognized for his achievements in the field of pulmonary vascular diseases by the Pulmonary Vascular Research Institute, and his name was added to the "Pulmonary Hypertension Committee of Honor" chosen by the Spanish Ministry of Health. He has been invited as a guest speaker in many national and International meetings and gave more than 450 lectures in different fields of pulmonary medicine.



Allan Lawrie, PhD
BHF Senior Basic Science Research Fellow
Professor of Translational Cardiopulmonary Science
Head, Pulmonary Vascular Research Group
Co-Director, Donald Health Research Programme
in Pulmonary Hypertension
Department of Infection, Immunity and Cardiovascular Disease
University of Sheffield Medical School, Beech Hill Road
Sheffield, UK

Dr. Allan Lawrie completed his Ph.D. "The effects of ultrasound on vascular gene delivery" in the Department of Cardiovascular Science at the University of Sheffield at the end of 2001. He followed this with a post-doctoral position with Prof. Marlene Rabinovitch at Stanford University, California where he developed his interest in Pulmonary Hypertension. He gained his first independent Fellowship to return to Sheffield at the end of 2004 (Russell Fellowship) and started to build a basic science research group focused on understanding the molecular mechanisms underlying Pulmonary Hypertension.

Dr. Allan also obtained a Medical Research Council Career Development Award in 2008 and now holds a British Heart Foundation Senior Basic Science Research Fellowship (since 2012). Within the newly formed Donald Heath Programme, he leads a talented group of scientists and maintains strong translational links through the Sheffield Pulmonary Vascular Disease Unit at the Royal Hallamshire Hospital working towards his core research interests of drug target identification, novel therapies, and early disease diagnosis.



Alessandra Manes, MD
Coordinator Center of Diagnosis and Therapy of Pulmonary
Arterial Hypertension
Department of Experimental, Diagnostic
and Specialty Medicine - DIMES
Alma Mater Studiorum University of Bologna
Bologna, Italy

Alessandra Manes received her medical degree with the academic honor cum laude at the University of Bologna (UoB) in 1996. She specialized in Cardiology, also graduating cum laude, in 2000 and she earned her Ph.D. in 2003 (Medical Faculty of the UoB). She is a staff member of the Institute of Cardiology of the UoB and serves as the coordinator of the Pulmonary Hypertension Center. Dr. Manes teaches at the International Master's Degree Program on pulmonary Vascular diseases at the UoB.

She has authored more than 200 scientific publications on pulmonary hypertension and heart failure and received the "François Brenot Award as "Promising Young Investigator in Pulmonary Vascular Sciences" of the European Respiratory Society in 1999. She has been included in the faculty of the World Symposium of Pulmonary Hypertension in 2003, 2008 and 2013.



Hani Sabbour, MD, FACC, FHRS, FASE
Consultant Cardiologist
Cleveland Clinic Abu Dhabi
Abu Dhabi, UAE

Dr. Hani Sabbour, MD, FACC, FHRS, FASE graduated with a Bachelor's Degree in basic Medical Sciences and Bachelor's Degree in Medicine and Surgery from Kuwait University Faculty of Medicine in 1994 with first class honors and was twice awarded His Highness the Emirs Gold Medal for academic excellence in both degrees. He subsequently went on to train in Internal Medicine and Cardiovascular Disease at SUNY Stony Brook in New York and was elected resident and fellow of the year several times.

He then completed his training in Clinical Cardiac Electrophysiology at Massachusetts General Hospital and was appointed Clinical Instructor in Cardiology at Harvard Medical School in 2001 as well as SUNY Stony Brook. He is currently American Board of Internal Medicine Certified in Internal Medicine, Cardiology, Electrophysiology, and Board Certified in Echocardiography and Nuclear Cardiology. He has always been heavily involved in teaching and been on the teaching faculty at Brown University since 2006.

He was recently promoted to Clinical Assistant Professor of Medicine and Cardiology at Brown University Warren Alpert School of Medicine. Dr Sabbour has been active in research and is currently an active PI in two international multicenter trials in the field of pacing and ICDs and has had several publications in the field. He was also appointed the Internal medicine residency program director at Al Ain Hospital as well as Arab Board Cardiology fellowship site director. He has been in practice in Rhode Island for 11 years and recently moved to the UAE to be on staff as Consultant Cardiologist at SKMC, Al Ain Hospital and Tawam Hospitals. His main clinical interests are the management of Arrhythmias and Advanced CHF and Pulmonary HTN as well as cardiac imaging.



Hussam Sakkijha, MD
Consultant, Pulmonary,
Critical Care and Sleep Medicine
King Fahad Medical City, Riyadh
Riyadh, Saudi Arabia

Dr. Sakkijha is a Consultant Pulmonologist, Intensivist and a sleep physician at King Fahad Medical City in Riyadh, Saudi Arabia. He is also in charge of Pulmonary Hypertension Program at the same center. He is the co-chair of CTEPH taskforce in the SAPH and contributed extensively to the PH Services in the region. He has many publications in the field and a Co-author of the Saudi Guidelines of the management of PH.



Sarfraz Saleemi, MD
Consultant, Pulmonary Medicine
King Faisal Specialist Hospital and Research Centre
Riyadh, Saudi Arabia

Dr. Saleemi is a Consultant Pulmonologist at King Faisal Specialist Hospital & Research Center in Riyadh, Saudi Arabia. He is the deputy head of the Pulmonary Hypertension program in the hospital and a special SAPH member. He contributed significantly to the promotion of Pulmonary Vascular Disease in Saudi Arabia. His area of interest is related to CTEPH and Porto-pulmonary Hypertension and published extensively in this area.



Gérald Simonneau, MD
Professor Emeritus, Respiratory Medicine
at Paris Sud University
Senior Consultant, National French référence
Center for Pulmonary Hypertension
Marie Lannelongue and Bicetre University Hospitals
Paris, France

Professor Gerald Simonneau is today Emeritus Professor at the Paris-Sud University, France, and Senior Consultant at the National Reference Centre for Pulmonary Vascular Disease in Bicêtre and Marie Lannelongue University Hospitals, France. He has published widely in the fields of Pulmonary Hypertension, Pulmonary Vascular Diseases, and Pneumology, in peer-reviewed Journals including the New England Journal of Medicine, The Lancet, Annals of Internal Medicine, and Circulation.

He has been President of the working group on pulmonary circulation of the European Society of Cardiology and has received the PAH research award of the European Respiratory Society in 2011. Lastly, he was named among the World's Most Influential Scientific Minds in the field of Clinical Medicine (Thomson Reuters) in 2014, 2015 and 2016.



Hanan Fan, MD
Consultant Pulmonologist and Head of Pulmonary Division
Director of Pulmonary Fellowship Program and
Medicine Residency Program
AHA ACLS Instructor
King Fahd Armed Forces Hospital
Jeddah, Saudi Arabia

Dr. Hanan Aladdin Fan is a Consultant Internist & Pulmonologist in Head of Pulmonary Division at King Fahd Armed Forces Hospital, Jeddah. She is also currently working as a Director & Supervisor of Pulmonary Fellowship Program in KFAFH, Jeddah and is leading a role of Chairman of Educational Committee of Internal Medicine Saudi Board Program.

Dr. Hanan Fan have attended several conferences including European Respiratory Society Conference (2018), Pulmonary Hypertension Master class(2017), Italy. She was an active organizer and speaker in World Psoriasis Day, Challenges in Psoriasis management (2018), she have also been a Member of Morbidity & Mortality Committee in King Fahd Armed Forces Hospital in Jeddah during residency.



Olivier Sitbon, MD, PhD
Professor of Respiratory Medicine, South Paris University
French Referral Centre for Pulmonary Hypertension,
Department of Respiratory and Intensive Care Medicine,
Bicêtre University Hospital, Le Kremlin-Bicêtre, France
Le Kremlin-Bicêtre, France

Olivier Sitbon, MD, Ph.D, is a Professor of Respiratory Medicine at the South Paris University and a Consultant at the French Referral Centre for Pulmonary Hypertension, Department of Respiratory and Intensive Care Medicine, Bicêtre University Hospital in Le Kremlin-Bicêtre, France. He also leads the team "Medical and surgical therapeutic innovation in PAH" of the INSERM Research Unit "Pulmonary Hypertension: athophysiology and Innovative Therapies". Prof. Sitbon has conducted extensive research in PAH and he is the Scientific Head of the French Registry of patients with Pulmonary Hypertension.

His investigational activities include clinical studies on factors associated with PAH, prognostic factors, risk stratification and treatment goals in PAH, and the development of new strategies for the treatment of PAH. He co-chairs the "Trials Design and New Therapies for PAH" task force at the 6th World PH Symposium in 2018. He has authored more than 270 peer-reviewed articles on PH and related topics in New England Journal of Medicine, Circulation, Journal of the American College of Cardiology, American Journal of Respiratory and Critical Care Medicine, European Respiratory Journal and Chest, amongst other Scientific Journals.



Jean-Luc Vachiery, MD
Clinical Professor of Cardiology,
Director, Pulmonary Vascular Diseases and
Heart Failure Clinic,
Hôpital Erasme – Cliniques
Universitaires de Bruxelles, Belgium
Brussels, Belgium

Jean-Luc Vachiéry is Clinical Professor of Cardiology and Director of the Pulmonary Vascular Diseases and Heart Failure Clinic at the Hôpital Erasme – Cliniques Universitaires de Bruxelles, Belgium. He received his board certification in Internal Medicine in 1992 and in Cardiology in 1995. Jean-Luc Vachiery has authored several articles, book chapter and participated in many collaborative research projects. This led to the creation of a clinical unit dedicated to Patient Management and clinical research in Pulmonary Hypertension and heart failure in 2008.

Jean-Luc has co-chaired the Pulmonary Hypertension Council at the International Heart and Lung Society (2002 – 2005), the Working Group on Pulmonary Circulation and Right Ventricular Function at the European Society of Cardiology (2006 – 2008) and the Working Group on Heart Failure at the Belgian Society of Cardiology (2008 – 2008). He was a Task Force member and section editor of the European Guidelines on Pulmonary Hypertension (2009 and 2015). He is the current chair of the PH Council at the ISHLT (2018 – 2020).

Jean-Luc has also been involved in the World Symposium on Pulmonary Hypertension as task force member in 2003, (co-chair of the PH due to heart and lung diseases task force) 2013 and was the co-chair of the Task Force on Pulmonary Hypertension due to left heart diseases for the upcoming World Symposium on Pulmonary Hypertension in 2018. Jean-Luc acts as an expert for several regulatory agencies and Scientific Organizations for matters related to medicines for Pulmonary Hypertension.



Mohammed Qasem, MD
Consultant Cardiologist,
Prince Sultan Cardiac Center- Al Qassim
Head of Noninvasive Cardiology Department
Head of Education and Research Department
Director of Pulmonary Hypertension Clinic
Al Qassim, Saudi Arabia

Dr. Mohammed S. Qasem is currently a Consultant Cardiologist at Prince Sultan Cardiac Center, Al Qassim, Saudi Arabia. He is the Head of Noninvasive Cardiology Department and the Head of Education and Research Department. Also, serving as a Director of Pulmonary Hypertension Clinic at Al Qassim. He completed his education in Medical Bachelor & Bachelor of Surgery at Faculty of Medicine, Jordan University of Science & Technology (JUST), Irbid, Jordan.

His 3-year-fellowship in cardiology at the Jordan University Hospital was an extensive experience in cardiac ward care, CCU, consultation, non-invasive and invasive tests. He actively participated in several conferences and presented a number of abstracts including Role of AFI in early detection of ischemic heart disease in the patient presented with typical chest pain and LM Diseases at ASE2016.



BOSENTOR

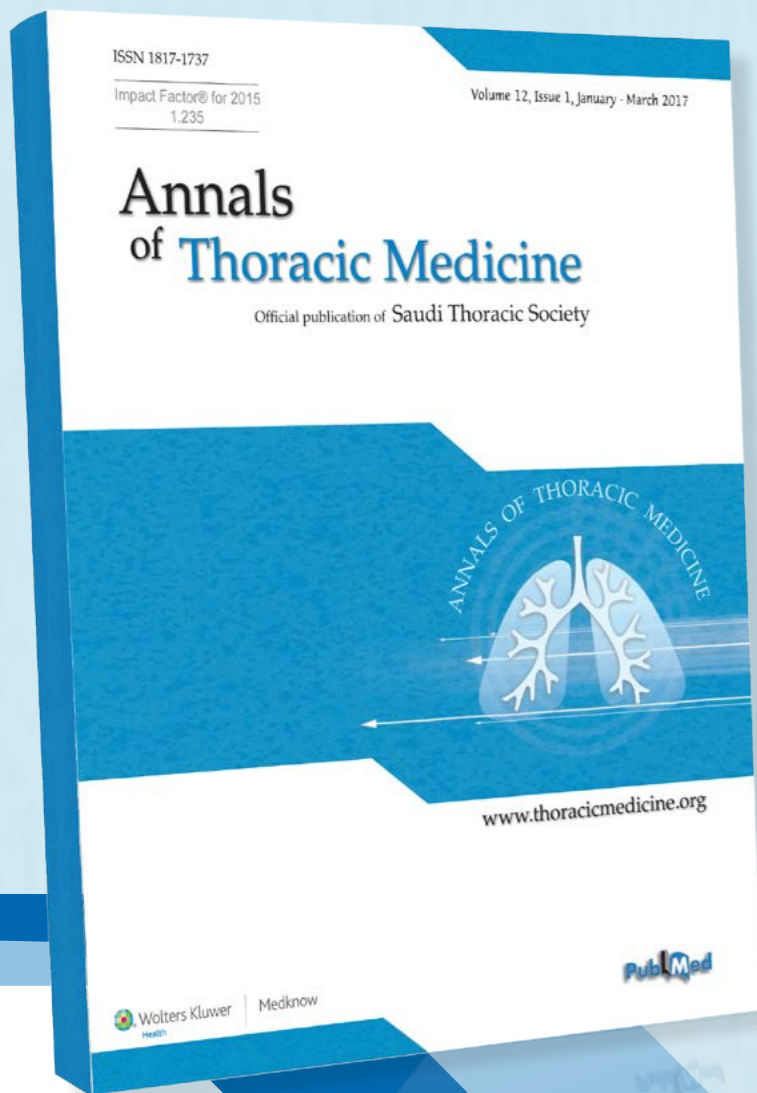
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CHALLENGING CASES

A Case of Portopulmonary Hypertension with Liver Cirrhosis: A Case Presentation and Review Literatures, What to do and what to give?



Mohammed Qasem, MD
Consultant Cardiologist,
Prince Sultan Cardiac Center- Al Qassim
Head of Noninvasive Cardiology Department
Head of Education and Research Department
Director of Pulmonary Hypertension Clinic
Al Qassim, Saudi Arabia

Introduction:

Liver disease and portal hypertension can be associated with pulmonary vascular complications including Hepatopulmonary Syndrome and Portopulmonary Hypertension. Portopulmonary Hypertension occurs in 1-5% of port hypertension cases and carries a poor prognosis; there is no clear management plan in the pulmonary hypertension guideline because such cases are excluded from major randomized clinical trials.

Case Report:

A 53-year old Egyptian male university teacher. He has no cardiovascular risk factors. No evidence of any chronic illness apart from benign prostatic hypertrophy. He was presented to our General Cardiology Clinic with a history of exertional dyspnea and palpitation. Clinic examination revealed raised stable vital signs, body weight 80 kg, height 170 cm and BMI 28. Cardiac examination: raised JVP, loud S2 with palpable and audible P2 component, no cyanosis and no clubbing. CXR revealed prominent pulmonary trunk and proximal pulmonary artery. Echocardiography revealed normal systolic function, grade 1 diastolic dysfunction with normal LV filling pressure, LV systolic D shape indicating pressure overload, moderate TR with PSAP 105 mmHg, dilated RV and RA with preserved RV function TAPSE 24 mm. Patient referred to our clinic for further work-up. Pulmonary hypertension work-up revealed: Normal HRCT, CT of pulmonary artery showed no evidence of acute pulmonary embolism or chronic thromboembolic disease. Pulmonary function test was normal except for reduced DLCO 45%. VQ scan revealed a normal study.

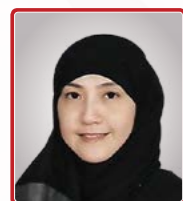
Pulmonary consultation cleared the patient from pulmonology point of view including sleep apnea syndrome as a patient gave a history of snoring. Laboratory result was all normal: CBC, liver enzyme, INR, kidney function test, all hepatitis markers, HIV negative. Normal thyroid function tests, negative ANA, Anti-dsDNA and lupus anticoagulant. Two weeks later, the patient sought medical advice to our ER complaining of presyncope and palpitation, so he was admitted to our intermediate care for observation and for right heart catheterization study (RHC). The hemodynamic study revealed: mean pulmonary pressure (MPA) 60 mmHg, pulmonary vascular resistance (PVR) 13.8 wood, systemic vascular resistance (SVR) 18 wood, RA pressure 9 mmHg and pulmonary vasoreactivity test was positive. 6 minutes walk test 680 m and N-T proBNP 110. Taking into consideration the clinical status, hemodynamics, echo data, and laboratory data, the patient has a low risk. Sildenafil 20 TID and Bosentan 62.5 BID were started. On the same night of the start of medical therapy, the patient had attacked twice, supraventricular tachycardia (SVT), which was terminated by Adenosine. Next day, he got another attack of SVT which terminated with Adenosine and Verapamil 40 mg BID initiated. Electrophysiologist's opinion is to continue medical therapy, no need to intervene. As the patient gave a history of gastrointestinal bilharziasis at the age of 15 years, we take into consideration portal hypertension and chronic liver disease. Abdominal Ultrasound Revealed: Liver cirrhosis with multiple nodular surfaces. The liver average in size displays coarse echotexture with the nodular surface, nodular hyperplasia, no focal lesion, no intra or extrahepatic biliary dilatation and portal vein is 14. Upper gastrointestinal endoscopy revealed portal hypertension gastropathy. Bilharziasis antibodies titer negative. Combined meeting with gastroenterologist revealed patient with liver cirrhosis with child-terracotta Class A with 1-year survival 100%. So the strategy of management is to discontinue Bosentan. However, taking into consideration the pulmonary portal hypertension stage severity, the patient is considered at higher risk and poor prognosis, MPAP > 45mmHg, PVR > 800 dyn/s/cm-5A and RAp 10 mmHg, consequently Macitentan 10 mg planned due to lower liver toxicity. Patient markedly improved after 5 weeks of treatment.

Because portopulmonary hypertension is associated with liver disease, this makes challenges to determine the regimen of treatment, monotherapy, combination therapy, sequential therapy or upfront therapy especially if the patient has stage 4 severe portopulmonary hypertension like our patient. Patients with PoPH have been excluded from almost all RCTs in the PAH field (except for the PATENT study, which included 13 patients with PoPH).

Conclusion:

Although Portopulmonary Hypertension is a form of Pulmonary Arterial Hypertension, it is considered a specific form due to concomitants, Chronic Liver Disease and Liver cirrhosis that may need Liver transplantation. These patients should be referred to a Liver transplantation center in cooperation with Pulmonary Hypertension centers.

A Familial Pulmonary Arterial Hypertension Case



Hanan Fan, MD
Consultant Pulmonologist and Head of Pulmonary Division
Director of Pulmonary Fellowship Program and
Medicine Residency Program
AHA ACLS Instructor
King Fahd Armed Forces Hospital
Jeddah, Saudi Arabia

Introduction:

Idiopathic PAH (iPAH) and heritable PAH (HPAH) are rare in the general population and estimated to be 5 to 15 cases per one million adults. The term heritable PAH (HPAH) includes familial PAH (PAH that occurs in two or more family members) and simplex PAH (a single occurrence in a family) when a pathogenic variant has been identified. Most heritable PAH (75%) is caused by a pathogenic variant in BMPR2. Transthoracic echocardiographic screening of at-risk family members every few years is recommended by WHO guidelines to enable earlier detection and treatment. However, the role of molecular genetic testing for early diagnosis of at-risk family members has yet to be established.

Case Report:

A 38 years old lady presented late with progressive shortness of breath to the cardiology service. Patient was fully worked up and found to have severe pulmonary arterial hypertension of unknown cause that eventually admitted to ICU and referred to Riyadh for transplantation. Unfortunately, patient died before actual transplantation. She was not known to have any chronic illnesses. No history suggestive of connective tissue disease and was not taking any medication. She was a mother of 7 daughters and 2 sons. The eldest daughter 3 days post-delivery of her first pregnancy in Riyadh, complained of sudden shortness of breath and died. It was reported as sudden cardiac arrest of unknown cause. The second daughter presented to Taif hospital with shortness of breath while pregnant in 7-months gestational age. She underwent an emergent cesarean section and unfortunately died due to an unknown sudden cardiac arrest.

My patient Eman (current 27 years old), the forth daughter, presented to our hospital in 2009. She was pregnant in first trimester. She was screened by echo because of the family history mentioned above and was normal. Patient delivered via normal spontaneous vaginal delivery without complications. At the same time, all other siblings were screened by echo and were normal. My patient presented again in 2011 with progressive shortness of breath which started gradually since post-delivery. Clinical examination revealed stable vital signs, cardiac examination raised JVP, loud P2. CXR revealed prominent pulmonary trunk. Echo showed RVSP 95 mmHg, moderately severe tricuspid regurgitation, markedly dilated right atrium, dilated right ventricle, moderately decreased systolic function. TR velocity 4.6m/sec. IVC dilated and collapsing. No pericardial effusion. Right heart catheterization mPAP 90 mmHg. HRCT no parenchymal lung disease. Ventilation perfusion scan showed low probability for pulmonary embolism. Rheumatological serologies were negative. Sleep study was normal. Pulmonary function test FVC=74, FEV1=73, ratio=80, TLC=68, DLCO=73. 6MWT patient walked 298 m (531m predicted) 56%. Patient underwent transesophageal echo to assess for small VSD. Patient was started on sildenafil then bosentan was added. After wards bosentan was changed to macitentan. Serial echos assured decreased RVSP and patient symptoms improved to WHO FC I. In June 2017, patient complained of newly exertional shortness of breath WHO FC II-III. Right heart catheterization revealed PA 88/44, mean 64 mmHg, PCWP 12 mmHg, CI 2.7. Patient lately developed iron deficiency anemia and gained significant weight which could contribute to her current symptoms.

In 2013, during the course of managing Eman, her aunt 58 years old has presented to our hospital with severe shortness of breath. Echo revealed severe pulmonary hypertension, RVSP >120 mmHg. Patient died next day. For that, we considered genetic testing of all the siblings who already had normal echos. Taking in consideration that Eman developed pulmonary arterial hypertension after initial screening of normal echo result. Genetic results revealed positive BMPR2 for Eman and another sister Intissar. The other sister had early right heart catheterization and early targeted therapy was initiated. She was managed carefully during her pregnancy and delivered through cesarean section uneventfully. Patient is asymptomatic, good physical activity with better clinical course.

Conclusion:

Echocardiogram is the screening tool of choice for patients with familial pulmonary hypertension in their families as it is noninvasive and readily available. However, genetic testing can better define the actual risk for another family member to develop FPAH, especially when a BMPR2 mutation has been identified in a PAH patient in the same family. Genetic testing enable earlier treatment and better outcome than screened by echo alone especially when other add-on risk factors as pregnancy is cautiously considered. As to the future, when sufficient understanding of the aberrant actions of the mutated BMPR2 points to either preventive treatments or specific therapies to reverse pulmonary arterial hypertension and restore health.

PAH in Pregnant Women



Fayez Khalid S. AlAhmadi, MD
Pulmonary Fellow,
King Faisal Specialist Hospital and
Research Centre
Riyadh, Saudi Arabia

Introduction:

Idiopathic Pulmonary Arterial Hypertension (iPAH) is characterized by progressive increase in pulmonary arterial pressure and pulmonary vascular resistance which lead to right ventricular failure and death. Idiopathic PAH is aggravated by the physiologic changes during pregnancy. Pregnancy in patients with iPAH is contraindicated because of high maternal and fetal mortality, most physicians recommend early termination of pregnancy in patients with iPAH.

Case I:

A 27 year-old lady, housewife, who was referred to our center with a diagnosis of idiopathic PAH based on right heart catheterization which was done in 2011 and showed MPAP 40 mmHg. She was pregnant, G3 P1+1, 36 weeks of gestation. She had Dyspnea FC II. She was on Sildenafil, Macitentan and Ilioprost. Macitentan stopped. Pro-BNP 66, ECHO showed, EF > 55%, Moderately dilated RV & RA, PASP showed severe PHTN, TRV 2.9 m/sec. After a Multidisciplinary Team Meeting, the consensus was to proceed with Elective Cesarean Section. In OR, during epidural anesthesia, the patient arrested twice, CPR initiated and ROSC was achieved. Delivery of the baby was done successfully. Peripheral arterio-venous ECMO was inserted. We started her on Sildenafil, Macitentan and Treprostinil. ECHO showed EF 20 %, with global hypokinesia. Day 3 post op., ECHO showed improvement of the LV function, EF > 60 %. Day 4 post op., she had hypotension and her hgb dropped. US abdomen showed hematoma at the surgical site and she underwent emergent exploratory laparotomy and 3L of blood was evacuated. Day 5 post op., she was decannulated from ECMO and later on extubated. Days later, she improved clinically and hemodynamically and she was discharged home. As a follow up, she was maintained on Sildenafil, Macitentan and Treprostinil. She is currently FC I, Pro BNP improved from 10,253 >> 135. Three months later we did for her right heart cath. and the results showed: MPAP 53, PCWP 15, PVR 10 WU, CI 2.3, CO 3.7. (Severe pre-capillary PHTN with positive vasoreactivity test!).

Case II:

A 24 year-old lady known with Sickle Cell trait, Primigravida, she was referred to us with a presumptive diagnosis of iPAH at 31 weeks of gestation. She had dyspnea FC IV. ECHO showed, EF > 55%, impaired diastolic function, severely dilated RV with impaired function, severely dilated RA, PASP 95 mmHg, TRV 4.8 m/sec, with +ve PFO. Pro-BNP was 634. We started her on Sildenafil and IV Prostacyclin. The decision of the multidisciplinary team was to proceed with elective cesarean section at 33 weeks of gestation. Few days later, she started to have lower abdominal pain, contractions and fetal distress. So, she underwent emergency caesarean section. Post op., she became hypotensive despite being on inotropic support. So, veno-arterial ECMO was inserted. Macitentan was resumed in addition to Sildenafil and Treprostinil. As a follow up, she had abdominal distension and hypotension, bedside U/S abdomen showed large abdominal collection, Emergency exploration was done for her and hematoma was evacuated. Days later, underwent Decannulation of ECMO and Left groin exploration and Left femoral artery thrombo-embolctomy was done. Later on, she developed Septic shock and she had cardiac arrest, she was intubated and while the ECMO team was cannulating at bedside as a life saving measure. She was bleeding from the left groin (from the old cannula site). She had asystole for 45 minutes and unfortunately, she passed away.

In conclusion:

The aim of presenting these cases is to demonstrate that management and treatment of iPAH in a pregnant woman is a complex task and carries a high risk for morbidity and mortality, a multidisciplinary team should be involved (obstetrician, pulmonologist or cardiologist and anesthesiologist).

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Agnes Kohl, MPharmSc
Head of Business Development

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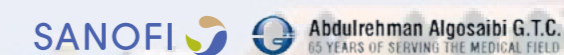
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Telephone: (+966) 11 462 2515 Ext. 12

Mobile: (+966) 53 390 0411

Email: saphsts@gmail.com



 **CONTACT:**

Contact No.: (+966) 53 390 0411

Email: saphsts@gmail.com

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