On behalf of the PVRI Board, it is my privilege to provide you with an update on the activities of our organisation for 2019, and a glimpse of 2020 and years to come.

Our Board of Directors held productive meetings throughout 2019 to discuss our overall strategy for the four years ahead; review progress of our two main PVRI conferences (the Annual World Congress and the Annual Drug Discovery & Development Symposium) and of our Pulmonary Circulation journal. Review of the PVRI Task Forces and several other PVRI trademark activities, such as the Innovative Drug Development Initiative (IDDI) and international meetings held under the PVRI banner, also came under critical review.

One of our major successes this year was to secure CMIREF funding for a tripartite initiative that was refined in early 2019. This includes:
1. Creation of a PVRI GoDeep Registry which will involve deep phenotyping capturing demographics, haemodynamics, imaging, and serum biomarker parameters susceptible to be used for sophisticated research purposes. This effort based in Giessen, Germany, will be spearheaded by Werner Seeger, the next PVRI President. The plan is to link registries from a number of carefully chosen centres of excellence in various parts of the world.
2. Creation of a PVRI GoGlobal Registry which will bring under one umbrella several registries from centres around the world headed by PVRI delegates. I will head this endeavour along with researchers at the Hopkins Bloomberg School of Public Health. As opposed to the GoDeep Registry, the data entries for GoGlobal will be limited here to demographics and diagnoses, with the aim to provide a global overview of the prevalence and aetiology of PVD and PH throughout the world.
3. PVRI Global Health representation, a project spearheaded by Stuart Rich (one of the PVRI founders) and Paul Corris (PVRI Chief Scientific Medical Officer). This third initiative will strive for effective advocacy for PV to be recognised as a global disease burden.

In summary, we envision the Global PVRI Registry to become a major clinical repository available for centres around the world for the purpose of research and promoting both understanding and treatment of pulmonary vascular diseases. Our ultimate goal remains to create a reliable landscape of PVD prevalence, patient demographics, and types of disease (across the 5 groups of the PH classification) specific to distinct parts of the world. This resource will strengthen our connections with institutions dedicated to Global Health, such as the World Health Organization (WHO), the Non-Communicable Disease Alliance (NCDA), and the World Heart Foundation (WHF) with which we have acquired membership.

The 13th PVRI Annual World Congress in Barcelona proved to be a major achievement and success for the PVRI, reaching record attendance of nearly 500 scientists, and new heights in terms of content and quality of presentations. Aside from innovative basic science topics, there was much emphasis on the power of registries around the world and novel and old (repurposed drugs) therapies for PAH. Much credit goes to the scientific organising committee composed of Sore Sauer-Pullammetz, Sébastien Bannet, Stevee Provencher, Anna Hemmes, Christophe Guignabert and Joan Albert Barberá.

This meeting was immediately followed by the Symposium of the 1st International Consortium for Genetic Studies in PAH organised by Nick Morrell under the PVRI banner, which was also well attended and of excellent scientific quality. At this time, we are looking forward to the 14th PVRI Annual World Congress, in Lima, Peru, with its scientific programme that highlights areas in hypoxia and high altitude. The 2020 Annual World Congress will immediately be followed by the Symposium of the 2nd International Consortium for Genetic Studies in PAH, again organised by Nick Morrell.

The 6th Annual Drug Discovery & Development Symposium, held in Paris in July 2019, also exceeded our expectations, both in terms of number of delegates and quality of science. The programme focused on Phase 2 clinical trials, both failures and successes, general designs and relevant endpoints, and strategies aimed at improvement. A second focus was on novel clinical trials based on pre-clinical models exploring new pathways. The success of this meeting was in great part due to the efforts of the organising committee composed of Gérard Simonneau, Marc Humbert, John Newman and Stuart Rich. The last afternoon session of this two-day meeting was devoted to the IDDI presentations and discussions led by Paul Corris, Syenko Nikhiko and Peter Fernandes. The 7th Annual Drug Discovery & Development Symposium will be held in Boston on 29-30 June 2020. Visiting historic Boston in June will certainly be a treat and the meeting promises to expose novel scientific ideas and attract a large audience.

The PVRI continued to award several grants in support of research and international travel. We have sponsored various PV meetings throughout the world. Examples include several meetings in Cairo, Egypt, organised by Ayman Farghali and Sir Magdi Yacoub. A PH meeting for children was held in Mumbai, India, organised by Prashant Bobhate, a further one in Tabriz, Iran, organised by Farid Rashidi. The EU Task Force leaders Carmine Dario Vizza and Stefano Ghio organised their second successful EU Task Force meeting in Catania, Italy. Feedback from these meetings was very positive and input of PVRI speakers was much appreciated. Our Disease Task Forces have progressed in several areas, such as the Imaging Position Statement, and the creation of the new Infection in Pulmonary Vascular Disease Task Force.

From a PVRI educational standpoint, we are happy to announce that Martin Johnson and Colin Church will be launching the second phase of the PVRI Digital Clinic in Lima, in collaboration with our Marketing Manager Aaron Shefras. The course includes 12 clinical cases provided by PH experts from various academic PH centres. Special thanks go to them for their extraordinary work in providing highly educational and challenging cases. We would also like to acknowledge our sponsors for this project, MSD. Without their trust and support, this important work would not be possible.

Our Pulmonary Circulation journal received its third Impact Factor of 2.075. It remains strong with a slight upward trend in original manuscript submissions and scientific content. Efforts are underway to improve our overall quality and raise our Impact Factor. We are greatly indebted to our Editors-in-Chief Jason Yuan and Nick Morrell, and Deputy Editors Kurt Steinmark and Irene Lang, as well as all journal reviewers for their hard work and enthusiasm.

In closing, we have achieved a great deal but have many challenges and wonderful opportunities ahead. We will continue to build bridges among our colleagues throughout the world. We owe our progress to all of you involved at so many levels.

Best wishes for continuous PVRI growth in the years to come!
Together with my colleagues in Giessen, Friedrich Grimminger and Ardeschir Ghofrani, I first became involved with the PVRI in its foundation period, mainly through Ghazevan Butros, whose passion and enthusiasm for a worldwide society of pulmonary hypertension physicians inspired us not only to join, but even to co-host one of the early PVRI Annual Meetings in Giessen, Germany, in January 2014, in collaboration with the meeting of the Excellence Cluster “Cardio-Pulmonary System” (ECCPS).

Although I have been involved with the PVRI for many years, it really was a great surprise when I was asked to become the President and I was truly taken aback by the enormous progress this small organisation had made since I first joined. To name but a few of its achievements, Pulmonary Circulation journal, our global expanding membership, our e-learning course, our Annual Congresses and Drug Symposia, which attract an increasing number of participants from all over the world, the invaluable work of our Task Forces, both in the regions and through our disease focus groups, our growing Roundtable of industry members and our collaborations with the Food and Drugs Administration (FDA) and the European Medicines Agency (EMA). Our work has led to many breakthroughs and discoveries and the promotion of real progress in treating our patients with this severe and still often devastating disease.

However, the above does not fully encompass the real character of the organisation and what I most admire about it. The PVRI was founded on three guiding principles - inclusiveness, diversity and integrity - and it has never lost touch with these values.

Inclusiveness of everyone! The PVRI thrives on open debate and discussion where everyone has a voice. I believe that it is this lack of hierarchical structure that encourages innovation. Only through open dialogue and blue-sky thinking can we achieve new discoveries and breakthroughs, but also learn from each other and accomplish clinical excellence.

Diversity in the sense of its global reach (we have members in 103 countries), but also diversity of medical disciplines, uniting heart and lung specialists, basic, translational and clinical scientists, leaders in the field and promising young scientists as well as the enormous range of disease specialists reflected in our many ‘Disease & Specialty’ and ‘Regional’ Task Forces.

Integrity we can only achieve what we do by the hard work and engagement of so many unpaid, active members from around the world. It is this engagement and support, as well as the commitment and tireless hard work of our staff, that makes a difference to the disease and the quality of life of our patients. Our active members are the heartbeat of this organisation. I cannot express my gratitude enough for what you do!

During my presidency, I want to build on these values and bring our mission to the furthest corners of the world, where we are currently not represented or only have a few members. I believe that we need to unite everyone in the fight against pulmonary vascular diseases and I will actively encourage collaborations with key medical societies, such as ISHLT, ESC, ATS, AHA and WSPHA on the international level, in addition to the numerous national organisations that address the topic of PH and pulmonary vascular diseases.

Many joint symposia and forums are already planned for 2020 and 2021, such as the joint ESC-PVRI Symposium in Lima, the ISHLT-PVRI joint Symposium in Montreal in April 2020, as well as the PH-Day during the ISHLT Annual Conference in Sydney, Australia in 2021. Social gatherings will continue during the ATS, as well as a new Get-Together in Europe during the ERS or ESC meetings.

Furthermore, I want to encourage the next generation of PH physicians to come into the fold of the PVRI by making the PVRI Annual World Congress, THE PH meeting where our younger members can discuss their work with eminent research leaders in this field. I am hugely encouraged by the large number of abstracts which have been submitted to our Lima meeting.

As part of my presidency, I will focus on developing a global registry in PH and I want to express my heartfelt thanks to the CMREF for their invaluable support in this pursuit. This will enable the PVRI to set up intercontinental registries either focusing on deep phenotyping GoDeep or focusing on a more global survey GoGlobal of PH patients all over the world. An ongoing effort alongside these activities will be the work we already undertake with the World Heart Federation, Non-Communicable Disease Alliance and the World Health Organization. Effective advocacy with global health providers for pulmonary hypertension to be acknowledged as a global burden is a key to getting better healthcare provision for our patients.

Finally, for our patients we need better and new drugs and treatment options. Our Innovative Drug Development Initiative, spearheaded by Paul Corris, Sylvia Nikkho and Peter Fernandes, and the work we do with the FDA and EMA are of vital importance in this pursuit and we shall continue to support effective clinical trials with our colleagues from pharma as best as we can.

However, the workload ahead should not deter us, but fill us with determination and enthusiasm to improve the care of our patients globally.

I am thus hugely grateful for this opportunity and wish to thank the PVRI Board of Directors and all our members for your confidence and trust in me. I will embark on this role with a great sense of responsibility and look forward to the exciting journey that lies ahead.

With my most humble thanks.

Welcome from the incoming President 2020/21
2019 marks the end of my sixth year with the PVRI. I have now worked with three inspiring Presidents, all of whom have brought their unique vision to our charity, which is continuing to expand across the globe. In a world that seems to be growing in conflict, the PVRI continues to succeed in bringing people together and uniting diverse stakeholders in open dialogue and a common aim. The PVRI ethos of collaboration, openness and inclusiveness runs through the entire organisation, but truly comes to life during our scientific meetings. Sharing in the experience of so many people coming together from different parts of the world with a common purpose, is still as inspiring and humbling to me as when I attended my first PVRI meeting in 2014. Our Barcelona Annual World Congress at the beginning of the year, was indeed a very special event with nearly 500 delegates from 39 countries. Congratulations to Soni Savai-Pullamsetti, Sébastien Bonnet and team for putting together an excellent scientific programme.

Our Drug Discovery & Development Symposium has become THE meeting for academia, pharma and regulators in the field. Stuart Rich and John Newman must be recognised and congratulated for their vision in masterminding this annual event. Together with Gérald Simonneau and Marc Humbert; they coordinated this year’s scientific meeting in Paris, which attracted an audience of over 120 participants. Over 80 delegates stayed for the open meeting of the PVRI Innovative Drug Development Initiative that followed. We should take great pride in facilitating these important stakeholder interactions that are vital in advancing our understanding of the disease. I would like to thank our industry partners and Roundtable members for their ongoing support.

Our society is continuing to grow in members, activities and revenues. Our network has expanded to 6,300 professionals from 103 countries. We are increasing our partnerships and collaborations with other medical societies, patient organisations and industry colleagues, as a fundamental strategy of our mission. This has resulted in many joint scientific symposia and international meetings. To name but a few, these included the 1st Symposium of the International Consortium of Genetic Studies in PAH, led by Nick Morrell; two different Paediatric Symposia, one in India, led by Prashant Bobhate and the other in Mexico led by Humberto Garcia; the 2nd EU Task Force meeting, in Italy under the leadership of Stefano Ghio and Dario Vizza; and the Pulmonary Hypertension AHC Science & Practice series in collaboration with the Aswan Heart Centre in Egypt under the leadership of our most eminent Trustee Sir Magdi Yacoub. Together with the other PVRI co-sponsored meetings held in Spain, Egypt, Bahrain, Germany, China, USA, India, Mexico, Iran, France, Thailand, Panama, Ukraine and Italy, we reached out to an audience of over 3,000 international delegates.

During the year, we have awarded a total of $100,000 in grants to our members in support of international travel, and regional PH meetings. Our scientific journal Pulmonary Circulations is increasing in recognition and international reach with over 840 articles published since its first edition in 2011. A huge thank you to Jason Yuan, Nick Morrell, Kurt Stenmark, Irene Lang and the whole PC editorial board for all their efforts.

I am excited by the expansion of our educational activities and launch of the second phase of the PVRI Digital Clinic, featuring twelve patient cases. We thank MSD for their financial support and all our colleagues from the various PH centres for providing the course materials. Particular thanks, however, go to Martin Johnson and Colin Church; the Scientific Content Managers, as well as Aaron Shefras, our Marketing Manager. Their huge contributions must not be underestimated.

A significant part of our work is achieved by our active Task Forces in the regions around the world and in the specific disease areas. Our Imaging & Pulmonary Hypertension Task Force, led by David Kirby, David Levin, Andrew Peacock and Andy Swift, has developed an important algorithm that will help in the diagnosis and management of the disease, and our Innovative Drug Development Initiative, led by Paul Corris, Sylvia Nikkho and Peter Fernandes, is working on the publication of a series of position statements on clinical trial design, biomarkers and endpoints. We have set up the new Infection in Pulmonary Vascular Disease Task Force, led by our President Emeritus Ghazwan Butrous, which held its first meeting in Egypt in October and addresses HIV, schistosomiasis and other infectious diseases in the context of pulmonary hypertension.

‘Our ambition is to unite pulmonary hypertension professionals globally in our fight against this deadly disease, remaining true to our values and purpose: advancing science through collaboration to improve patient care.’

Stephanie Barwick CHIEF EXECUTIVE OFFICER 2019
The immense level of activity of all our Task Forces is the engine of the PVRI that will lead to new scientific and clinical discoveries. I encourage you to read their detailed reports on pages 45-73. I would like to thank all our Task Force leaders and members for their invaluable contributions.

2019 has been a critical year for the PVRI to achieve financial stability for the future. We have been successful in the award of a substantial grant from the Cardiovascular Medical Research and Education Fund in the USA. The grant is spread over five years and will help us develop a Global Registry to measure the prevalence of pulmonary hypertension worldwide. In the fight against this deadly disease, we must firstly demonstrate that it is not rare, but a global burden that affects so many more people than the current estimates suggest.

This initiative will encompass PVRI GoDeep, a deep phenotyping database, under the leadership of our next President Werner Seeger at the Justus-Liebig-University in Germany, and in parallel PVRI GoGlobal, a survey that records various PH databases and activities from around the world where the data cannot be included in GoDeep. GoGlobal will be led by Paul Hassoun, our current President, at Johns Hopkins School of Medicine in the USA and researchers at the Hopkins Bloomberg School of Public Health.

But data alone cannot effect change in the absence of effective advocacy. Directing our attention to global health and providing a voice to improve current policies in healthcare and drug availability will be a major focus from next year.

These efforts will be led by Stuart Rich and Paul Corris. Our collaborations with the Institute for Health Metrics and Education at the University of Washington in the USA, as well as our membership to the World Heart Federation, Non-Communicable Disease Alliance and World Health Organization will be vital in these endeavours.

These achievements, however, cannot fully encompass the true character of the PVRI. The passion and drive of our members is the greatest gift of our community, which can never be summarised in a report. It took over 700 volunteers to achieve all the activities that are detailed in these pages. Their work is our heartbeat. Through them we contribute to making lives better and we enlighten our souls in the process, through the networks and relationships that emerge.

As a collective community of members, partners, staff and wider professional networks, we are united in our mission to improve patient care.

There will be challenges ahead, but we have much to be proud of.

---

Stephanie Barwick
Chief Executive Officer
Canterbury, UK
Our people

...are our strength. We are united in our mission to improve patient care.

Friendship through science.

700 proactive volunteers worldwide in 2019

In 2019, over 700 volunteers have proactively contributed to our activities, bringing our mission to life.

Our network has expanded to 6,300 PH professionals in 103 different countries.

We achieve our mission through the work of our Task Forces.

Our Regional Task Forces spread awareness and understanding of pulmonary vascular disease throughout the world (see full reports on pages 45-58).

The work of our Disease & Specialty Task Forces leads to new scientific and clinical discoveries (see full reports on pages 59-73).

PVRI Global network by regions:
- North America: 23%
- South America: 9%
- Africa: 5%
- Australia, New Zealand & Pacific Regions: 4%
- Central Asia: 1%
- China & East Asia: 8%
- Europe: 28%
- India: 7%
- Middle East: 4%

Our members are the heartbeat of our society. Through them we contribute to making lives better.
Our network

In 2006, the PVRI was started by five people. Now, our network expands to 6,300 professionals in 103 countries. We achieve our mission through the work of our Task Forces and all our active members throughout the world.
...our purpose

is to enhance global understanding of pulmonary vascular disease through education and research.

During 2019, we supported scientific meetings in Spain, Egypt, Bahrain, Germany, China, USA, India, Mexico, Iran, France, Thailand, Panama, Ukraine and Italy, reaching out to an international audience of over 3,000 professionals.

The first PVRI meeting was held in Malta in 2007 with 25 people. Since then, we have established two annual international scientific meetings - the Annual World Congress on PVD and the Drug Discovery & Development Symposium.

Our 13th Annual World Congress in Barcelona, was attended by 475 people.

We have become the leading voice of authority on pulmonary vascular disease worldwide.

475 delegates attended our Annual World Congress in Barcelona
We speak science...

The PVRI supports PH meetings all over the world, all year round.

<table>
<thead>
<tr>
<th>Date</th>
<th>Event</th>
<th>Location</th>
<th>Highlights</th>
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<tbody>
<tr>
<td>30 Jan-3 Feb</td>
<td>PVRI 13th Annual World Congress on PVD</td>
<td>Barcelona, Spain</td>
<td>• 475 delegates from 39 countries&lt;br&gt;• 56 presentations in 11 plenary sessions&lt;br&gt;• 171 scientific abstracts&lt;br&gt;• 85% of delegates rated event as excellent</td>
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<tr>
<td>2 Apr</td>
<td>PH meeting CAIRO, EGYPT</td>
<td></td>
<td>• 3 invited speakers&lt;br&gt;• 6 scientific sessions</td>
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<td>3-6 Apr</td>
<td>Joint PVRI-ISHLT Symposium during the ISLT 30th Annual Meeting &amp; Scientific Session</td>
<td>Orlando, USA</td>
<td>• 171 scientific abstracts&lt;br&gt;• 213 telecast viewers nationwide</td>
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<tr>
<td>13-14 Apr</td>
<td>Pulmonary Hypertension in Indian Children</td>
<td>Mumbai, India</td>
<td>• 87 delegates&lt;br&gt;• 12 international speakers from 7 countries&lt;br&gt;• 7 scientific sessions</td>
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<tr>
<td>27-28 Jun</td>
<td>3rd Pulmonary Hypertension meeting TABRIZ, IRAN</td>
<td></td>
<td>• 40 delegates from 39 countries&lt;br&gt;• 12 international speakers from 7 countries&lt;br&gt;• 7 scientific sessions</td>
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<tr>
<td>2-Aug</td>
<td>Asia Pacific Accreditation Cooperation PH Forum</td>
<td>Bangkok, Thailand</td>
<td>• 180 delegates</td>
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<tr>
<td>13-14 Apr</td>
<td>Pulmonary Hypertension in Indian Children</td>
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<tr>
<td>24-26 Sep</td>
<td>PVRI Eastern European Task Force meeting</td>
<td>Kiev, Ukraine</td>
<td>• 150 delegates&lt;br&gt;• 10 scientific presentations</td>
</tr>
<tr>
<td>18-19 Oct</td>
<td>PH AHC Science &amp; Practice</td>
<td>Aswan, Egypt</td>
<td>• 22 senior faculty&lt;br&gt;• over 150 delegates</td>
</tr>
<tr>
<td>21-22 Oct</td>
<td>1st International Symposium of Infection and PVD</td>
<td>Aswan, Egypt</td>
<td>• 28 senior faculty&lt;br&gt;• scientific sessions on HIV, schistosomiasis, tuberculosis&lt;br&gt;• over 100 delegates</td>
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JANUARY FEBRUARY MARCH APRIL MAY JUNE JULY AUGUST SEPTEMBER OCTOBER
...our publication is the first of its kind exclusively focused on the pulmonary circulation and diseases of the pulmonary vascular system.

Our research platform is the world.

Since the first volume in 2011, we have published 845 articles including:
- 53 editorials
- 148 reviews
- 443 original research articles
- 103 guidelines & other
- 98 case reports

First impact factor awarded in 2017.
Pulmonary Circulation Report
Jason Yuan & Nick Morrell, Editors in Chief

2019 has been another great year for Pulmonary Circulation. We continue to see increasing numbers of articles submitted and now receive many good quality papers. We have been able to raise the bar for acceptance and we are pleased to say that it is now becoming more difficult to get published in Pulmonary Circulation!

As can be seen from the table of most popular articles below, our most popular articles tend to be clinical rather than basic-science, and comprehensively review articles are also frequently downloaded. A highlight amongst the review articles we have published recently was the report of the PVRI taskforce on lung vascular imaging. While we are pleased to have many high-quality clinical manuscripts submitted to the journal, we would encourage our audience, especially PVRI members and members of the journal’s editorial board, to submit their basic and translational research manuscripts.

As at the time of writing this report (end October 2019), we have published 100 articles, including high-quality reviews, original research articles, and case reports.

Since the first volume in 2011, we have now published 845 articles in Pulmonary Circulation:

- Editorials (53) 6%
- Reviews (148) 17%
- Original Research Articles (444) 52%
- Guidelines & Other (103) 12%
- Case Reports (90) 11%

We receive most submissions from the USA, followed by China, the UK, Japan, and Germany, though we receive articles from all around the world. Please consider us when deciding where to publish your article - we would be delighted to receive it.

Strong Impact Factor
Our impact factor remains strong for a journal of our stature at 2.075. The impact factor is a measure of the frequency that an average article in a journal has been cited in a given year. To have an impact factor consistently over 2.0 is an excellent indicator for a young journal when we have been increasing the number of articles published.

Promotional activities
Pulmonary Circulation’s Editors and SAGE Publishing promoted the journal at various international conferences, including the PVRS Annual World Congress in Singapore, the PVRI Discovery and Development Symposium, the American Thoracic Society (ATS) International Conference, and the American Heart Association Scientific Sessions.

Top downloaded articles during 2019
1. Management of prostacyclin side effects in adult patients with pulmonary arterial hypertension
Authors: Martha Kingman, Christine Archer, Chris Chiu, Mary Bartlett, J. Beckmann, Robin Hohnfels, Sandra Lombardi
Published: 11 July 2017
Downloads: 309

2. Natural plant products in treatment of pulmonary arterial hypertension
Authors: Lili Xiang, Ying Li, Xi Dong, Djuro Kosanovic, Ralph Theo Schermuly, Xiaohui Li
Published: 25 June 2018
Downloads: 825

3. Advanced interstitial lung fibrosis with emphysisma and pulmonary hypertension with no evidence for interstitial lung disease on high resolution CT
Authors: Vyse Foris, Luke Brick, Philipp Douchain, Gabor Kovacs, Elvira Stach, Prithvi, Horst Aischwiler
Published: 8 July 2019
Downloads: 692

4. Pathophysiology, incidence, management, and consequences of cardiomyopathies in pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension
Authors: Meghan M. Cindulik, John J. Ryan, Stephen L. Archer
Published: 8 March 2019
Downloads: 578

5. Pulmonary hypertension subjects exhibit right ventricular transient exhalation dilation during supine exercise stress-echocardiography
Authors: Rosa E. Velayo, David Rancourt, Melkon Hacopian, Dennis Altherton, Mylan C. Cohen, Joel A. Wirth
Published: 7 June 2019
Downloads: 450

6. Early histological changes of pulmonary arterial hypertension disclosed by invasive cardiopulmonary exercise testing
Authors: Yusuf J. Bhatti, Alexandra J. Rito, Alexander Kompel, Konstantinos Dimopoulos, Laura C. Price, Harpreet Ranu, Athir Welts, S. John Wort, Caim M. McCabe
Published: 16 April 2019
Downloads: 414

7. Early intervention should we conduct therapeutic trials for mild pulmonary hypertension before onset of symptoms?
Authors: Jessica H. Hurson, Robert P. Franz, Evan L. Brittain
Published: 16 April 2019
Downloads: 409

8. Echocardiographic assessment of right ventricular function in experimental pulmonary hypertension
Authors: Zhongkai Zhu, Durst G. Galadie, Ailing Li, Bianca Rodriguez, Chenxin Ge, Haiyang Tang, Richard D. Minshall, Wei Huang, Ailing Chen
Published: 13 June 2019
Downloads: 377

9. Right heart in pulmonary hypertension: from adaptation to failure
Authors: Xiaofeng Ren, Roger A. John, Wei Dong Gao
Published: 2 August 2019
Downloads: 372

10. Distinct plasma gradients of microRNA-204 in the pulmonary circulation of patients suffering from WHO Groups I and II pulmonary hypertension
Authors: Leonard E. Estephan, Michael G. Geronardi, Chad M. Kosanovich, Michael G. Ribano, Yingze Zhang, Nancy Petro, Annie Watson, Xiaohui Li, John C. Sembrat, Mauricio Rojas, Dmitry A. Goncharov, Marc A. Simon, Elena A. Gonzalda, Anjali Vaidya, Akia Smith, Jeremy Mazurek, Yuchi Han, Stephen Y. Chan
Published: 28 March 2019
Downloads: 372

11. Ambrosetti for treatment of inoperable chronic thromboembolic pulmonary hypertension (CTEPH)
Authors: Pilar Escobar-Suárez, Haim Bondjanova, Paula S. Curtis, Irene Lang, Anton Wolk, Noordgraaf
Published: 20 May 2019
Downloads: 370

12. Drug repositioning in pulmonary arterial hypertension: challenges and opportunities
Authors: Daniel Grinnan, Cory Trankle, Adam Andruska, Bruce Bloom, Edela Spieringskins
Published: 4 March 2019
Downloads: 367

13. Physiotherapy practice in pulmonary hypertension: physiotherapist and patient perspectives
Authors: Carol Koen, Sally Fowler-Davis, Sonam McLean, Jane Manson
Published: 29 June 2019
Downloads: 365

Authors: Benoit Rancoule, Lloyd D. Harvey, Ramon J. Ayon, Alessandra Babichova, Sebastien Bonnet, Stephan Y. Chan, Jason X. J. Yuan, Viridis de Jesus Perez
Published: 18 January 2018
Downloads: 364

15. Right ventricular outflow tract velocity time integral to pulmonary artery systolic pressure ratio—a non-invasive metric of pulmonary arterial compliance differs across the spectrum of pulmonary hypertension
Authors: Priyanka T. Bhattacharya, Gregory S. Troutman, Frances Mio, Aril K., Fox, Maniak S. Tamar, Payman Zaniari, E. Wilson Grandin, Jonathan N. Menachem, Edo Y. Blirat, Julia A. Chirino, Tula Mandje, Kevin A. Smith, Steven M. Kauw, Paul R. Forba, Anjali Vaidya, Jeremy A. Mazurek
Published: 14 May 2019
Downloads: 351
...our patients are at the heart of our mission.

We strive for excellence in clinical care, research and education to improve the lives of our patients.

Providing our members with ways to enhance their understanding of this complex disease is essential to improving patient care.

Over the past ten years, we have created an online learning library of over 900 educational materials. These include lecture recordings, abstract posters and interviews with world renowned physicians.

In 2018, we launched the PVRI Digital Clinic, an online learning course that features a series of patient cases and shadows a real clinical environment.

It is aimed at specialists anywhere in the world, who want to improve their understanding of the diagnosis and management of PVD, and connect to leading specialists in the field.

Our aim is to produce a platform that will become an evolving learning tool that retains its relevance in the ever changing world of PH.

We thank MSD and GSK for their financial support of the PVRI Digital Clinic.

Let our learning lead to action.
Phase 2 of our e-learning initiative, the PVRI Digital Clinic, has undergone development since the first week of June 2019 and will be launched at our Annual World Congress in Lima, Peru on 30 January 2020.

Following the successful prototype development of the course, and thanks to funding received from MSD, phase 2 incorporates nine additional patient cases and houses a publishing platform whereby the PVRI can upload new patient cases quickly and at low cost. This means that the course can be continuously updated and expanded in the future.

We are sincerely grateful to our funding partners, MSD, for their support during this phase of development, which will enable all PVRI members to learn more about pulmonary vascular diseases and interact with other clinics around the world. Our aim is to produce an evolving learning tool that retains its relevance in the ever-changing world of PH.

What’s involved in Phase 2?
We have improved the Digital Clinic as an educational tool in the diagnosis and treatment of pulmonary vascular diseases.

The PVRI Digital Clinic includes the combination of real patient data, bespoke specialist advice and easy-access, highly relevant, peer-reviewed learning materials. We can now efficiently add more patient cases and increase the complexity of the course. This enables the learning experience in the field to remain relevant and truly effective. So far, a total of 12 cases are available.

Submit your patient case
To enable the PVRI to grow the Digital Clinic with additional and varied case studies, in a manner that is consistent and scalable, the PVRI is reaching out to clinics across the world to request that professionals upload their own patient cases for consideration. To submit your case study to the Digital Clinic, please email info@pvrinstitute.org.

What lies ahead for the e-learning platform?
There are a number of improvements in the pipeline for the Digital Clinic, some of the new features are detailed below.

Optional ongoing therapy
After completion of the ‘Treatment’ stage, users will be able to participate in the ongoing therapy of their online patients, should they opt to. They will receive email alerts advising them that their patient has experienced a complication, such as heart problems, pregnancy or adverse drug side effects. Users will then be able to re-enter the PVRI Digital Clinic to manage the complication and complete the case.

Improved learning library
The learning library is unique to our course. It currently features any relevant peer-reviewed reference material, such as articles, journals, books, guidelines. With the improved library, users are able to search and filter, while other relevant materials will appear as ‘related’ content, enabling the user to easily jump from one learning item to another.

Improved course layout
Although the course has been tested by 15 clinicians, further improvements, such as the layout of videos, can be made to further improve the user experience. We propose utilising ‘Google Analytics’ and ‘Hot Jar’, technologies to inform us where users are searching and filter, while other relevant materials will appear as ‘related’ content, enabling the user to easily jump from one learning item to another.

Improved course layout
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We thank United Therapeutics for sponsoring 75 PVRI memberships for doctors in developing countries.

...our partners include medical societies, patient groups, industry sponsors, drug regulators and global health organisations.

Strategic alliances and global partnerships are central to our mission.

We work with...

Medical societies
Industry partners
We thank all our Roundtable members for their support in 2019.

World health organisations

Drug regulators

Patient groups

We thank Bayer for their financial support to the PVRI Innovative Drug Development Initiative (IDDI)
...our PVRI grants enable our members to achieve their potential.

We are a small charity, but with the support from others, we can support our members.

During 2018/19, we have awarded $500,000 in grants to our members.

These included:

$260,000 for BMPR2 research sponsored by The Dinosaur Trust
$190,000 for two Fellowships
$40,000 for a feasibility study of an imaging database in deep phenotyping of PH associated with respiratory disease
$26,000 as co-sponsorship of worldwide Task Force and PH meetings
$17,000 of Travel Grants to our young scientists
$13,000 of Travel Grants supported by GSK for young scientists to participate in the ERS and ESC Conferences
$12,000 to our members to speak at international PH meetings
$1,300 for the Rupert Swift Award to a promising young scientist, supported by the Swift family
$500 for the best young abstract presenter at our Annual Congress in Barcelona, sponsored by the Butrous Foundation Young Investigator Award

$500,000 awarded in grants to our members during 2018/19
Vascular stress promotes fibrotic remodelling of the pulmonary arterioles, and this raises the intriguing possibility that BMPR1A is important in maintaining the health of the lung endothelium, and that its loss leads to pulmonary hypertension in a mouse model. The disease can be improved in these animal models through using molecules that inhibit TGFβ2. This raises the intriguing possibility that BMPR1A is important in human pulmonary arterial hypertension, and that inhibiting TGFβ2 may have a therapeutic effect. We will not aspire to develop our understanding of the clinical relevance of our findings and potential for novel therapeutics.

Together with my mentor, Dr Bradley Martin, we aim to characterise the effect of mild pulmonary hypertension on right ventricle-pulmonary vascular function and to identify novel opportunities to therapeutically target the pathobiology of early-stage PAH in the monocrotaline pah model without pulmonary artery endothelial cells from human pulmonary artery endothelial cells in response to oxidant stress. Currently, we are working to understand the biofunctionality of endothelial Csk as it relates to vascular fibrosis and early PAH pathogenesis.

Using a novel approach to isolate pulmonary artery endothelial cells from the monocrotaline PAH model without cell culture (Wertheim et al., PLoS One 2018), we observed that early-stage PAH is characterised by a unique endothelial transcriptomic profile that is enriched for fibrosis genes. Network medicine analysis of the endothelial transcriptome identified C-terminal Src kinase (Csk) as a predicted regulator of endothelial fibrosis in vivo.

We further demonstrate that endothelial Csk is upregulated in early-stage PAH in vivo, and that Csk accumulates in cultured human pulmonary artery endothelial cells in response to oxidant stress. Currently, we are working to understand the biofunctionality of endothelial Csk as it relates to vascular fibrosis and early PAH pathogenesis.

Through scientific training and career development opportunities, the PVRI Basic Science Fellowship has provided a valuable platform to launch my career as a physician-scientist with expertise in pulmonary vascular medicine. I look forward to ongoing collaboration with the PVRI in the years ahead.

Bradley Wertheim  
FELLOWSHIP GRANT  
In pulmonary arterial hypertension (PAH), endothelial cell dysfunction and oxidant stress promote fibrotic remodelling of the pulmonary arterioles, leading to right heart failure and premature death. Mortality and hospitalisation increase incrementally beginning at a mean pulmonary artery pressure below values that were previously thought to be pathogenic. Therefore, treatment of early-stage PAH may have clinical benefit, but specific therapies do not exist for such patients because the mechanisms of endothelial cell dysfunction and vascular remodelling in early PAH are not known.

Katherine Clapham  
FELLOWSHIP-HP GRANT  
I’m thankful for the PVRI’s support of my proposal: Novel Mechanisms of EndoMT in Pulmonary Arterial Hypertension. The research fellowship, generously provided by the PVRI, gave support for my position in the laboratory of my mentor Hyung Chon.

We aimed to investigate the function of the bone morphogenetic signalling component BMPR1A (ALK-3) in the cells that line blood vessels (endothelial cells). We created a mouse model in which we deleted BMPR1α from endothelial cells specifically and found that these mice developed pulmonary hypertension. Further experiments suggested that deletion of BMPR1α from the endothelium of these mice might lead to pulmonary hypertension, in part through increased activity of another molecule named TGFβ2.

We treated the mice with endothelial deletion of BMPR1α through administering a molecule that specifically inhibits TGFβ2 in the lung endothelium and found that the pulmonary hypertension in these mice improved.

We have also begun to study BMPR1A mutations, which were identified in individuals with pulmonary arterial hypertension by our collaborators Drs Nick Morrell and Stefan Gatz. We conducted experiments to examine the impact of the identified mutations on the function of the BMPR1A protein. This work is still ongoing. Overall, our work demonstrates that BMPR1A is important in maintaining the health of the lung endothelium, and that its loss leads to pulmonary hypertension in a mouse model. The disease can be improved in these animal models through using molecules that inhibit TGFβ2. This raises the intriguing possibility that BMPR1A is important in human pulmonary arterial hypertension, and that inhibiting TGFβ2 may have a therapeutic effect. We will not aspire to develop our understanding of the clinical relevance of our findings and potential for novel therapeutics.

Lucilla Piccari  
RESEARCH GRANT  
Research  
physician, Pulmonary  
Hypertension Unit, Pulmonology  
Department, Hospital del Mar-IMIM, Barcelona, Spain

As a Fellow of the PVRI, I was given the opportunity to develop my project: Feasibility study on the creation of an imaging database in deep phenotyping of Pulmonary Hypertension associated with Respiratory Disease (REHAR - Spain).

This is the first step in an international multicentre study on pulmonary hypertension (PH) associated with chronic lung disease, where data from PH registries in three European countries will be collected and analysed to facilitate phenotypes in Group 3. PH. The project focuses on deep imaging/phenotyping, and is particularly directed at the study of severe PH within this group.

Three national networks of PH centres with longstanding experience in CLD-PH have been considered for initial inclusion:

• the Spanish Registry for PH associated with Respiratory Disease (REHAR - Spain)
• the National Audit on Pulmonary Hypertension UK
• the Italian Pulmonary Hypertension Network (IHPHENet - Italy)

The coordinators, Dr John Wort for the UK, Dr Patrizio Vitulo for Italy and Lucilla Piccari for Spain, are currently screening, with the help of a survey, candidate centres for technical requirements relating to the imaging platform and data management.

At the same time, we are establishing the framework for the deep phenotyping analyses that will be performed, in collaboration with Professor Horst Olschewski and Dr Gabor Kovacs from the Ludwig Boltzmann Institute for Lung Vascular Research, Graz, Austria, and Dr Andy Swift from the Department of Infection, Immunity and Cardiovascular Disease of the University of Sheffield, UK. Both groups have long and established expertise in the analysis of imaging data in pulmonary hypertension and have previously collaborated on a PVRI project in this very field (see the PVRI Imaging Task Force Annual Reports on the PVRI website at www.pvri.org.uk).

Imaging from all patients will be analysed by a panel of radiologists using a predefined visual score while Artificial Intelligence will be used to assess the images with machine-learning techniques. With this two-pronged approach we believe we will be able to identify key characteristics in the lung parenchyma and vasculature and to link it to clinical, functional and haemodynamic data in order to build phenotypes within Group 3.

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Sébastien Bonnet  
**RESEARCH GRANT**

Thanks to the support of the PVRI/The Dinosaur Trust, we have initiated an international (North America, Europe) collaborative effort aimed to better understand the role of BMPR2 signalling network in PAH. Specifically, in France, Dr Perros and his colleagues have established the first rats’ model with disrupted BMPR2 signalling and demonstrated that these rats exhibit a phenotype similar to the one seen in humans with BMPR2 mutations (25% or so spontaneously developed PAH).

In Canada, after establishing the rat colony, Dr Ranchoux, a post-doctoral fellow, was able to duplicate Dr Perros findings. Based on this, Drs Nadeau, researcher associate, and Victoria Toro, PhD student, have explored the consequences of aging in this rat population in order to better understand the clinical complications that may arise in PAH patients with BMPR2 mutation, as well as healthy carrier.

During this, they have demonstrated that 20% (95%CI:12-28) of the 82 Bmpr2+/β71 rats versus 4% (95%CI:0-9) of the 84 age-matched wild-type littermates spontaneously developed mammary masses and ulcerations requiring euthanasia. Mammary masses were histologically diagnosed by an anatomical veterinary pathologist as fibroadenomas (hormone-dependent benign breast tumours predisposing to cancer) and adenocarcinoma in 83% and 17% of cases, respectively. Tumours predominantly occurred in females (93% of cases), were generally observed after postnatal day 322. We also demonstrated that female with tumours had greater PA pressures, suggesting that tumours development may accelerate/promote PAH development in BMPR2 mutant rats.

We are now investigating this further.

In conclusion, we demonstrated for the first time that BMPR2 mutated animals are predisposed to breast cancer. Development of breast cancer promotes the development of PAH.

Frédéric Perros  
**RESEARCH GRANT**

Through the funding support of the PVRI/The Dinosaur Trust grant, we were able to characterise at the haemodynamic, histological, electrophysiological, and molecular levels, the first rat lines ever created with monoallelic mutations in the gene encoding bone morphogenetic protein receptor 2 (Bmpr2), the main genetic risk factor for heritable pulmonary arterial hypertension (PAH). The results of this study were published in a high-ranked cardiovascular journal: Hautefort A et al. Circulation. 2019.

Noticeably, we were able to show a not-yet-studied effect of Bmpr2 mutation specifically on the right ventricle (RV), which compromised intrinsic function, even in non-overloaded states, suggesting that BMPR2 mutations may have a negative effect on RV function in patients with PAH. Totale, differences in survival and disease severity in BMPR2 mutation carriers have been explained mainly by more severe pulmonary vascular involvement, leading to a more severe and faster disease trajectory. However, because RV function is the main determinant of prognosis and disease severity, RV function may play a significant role in the different clinical phenotypes of BMPR2 mutation carriers and noncarriers. This is a shift in paradigm from an exclusive role of BMPR2 dysfunction in vascular remodeling to its highly probable involvement in RV failure/maladaptation.

Our studies now focus on a more detailed evaluation of the cardiovascular function, which will determine to what extent the RV (and the left ventricle of the Bmpr2 mutant rats) is compromised and the adaptability of those hearts to increased load.

- **17** Travel Grants awarded to young scientists for best abstracts at Barcelona Annual World Congress 2019
- **20** additional Travel Grants awarded to our members to participate in international PH meetings during 2019
We will do whatever it takes.

...our passion is to establish a global map on the prevalence of the disease.

Uniting our community in a collective voice of advocacy to improve healthcare and drug availability for all patients worldwide.

Many findings indicate substantially higher prevalence of the disease worldwide.

We are working with global health partners to map the worldwide burden of the disease to improve healthcare for all.

During 2019, we obtained a 5-year grant from the Cardiovascular Medical Research and Education Fund in the USA to establish a Global Registry to measure the burden of the disease.

- PVRI GoDeep
  Deep-phenotyping database to facilitate improved research.

- PVRI GoGlobal
  Collecting global data on the prevalence of the disease.

- Effective advocacy to improve healthcare for PVD patients worldwide.

Our Global Health Mission

The global presence of pulmonary hypertension (PH) in certain regions of the world is unknown, as available data is mostly limited to the developed countries. Global estimates of PH have been reported to range from 20 million to 70 million. However, many findings indicate substantially higher prevalence of the disease than what is currently assumed.

The PVRI will embark on a mission to collect reliable data and establish a global map on the prevalence of PH and its variability among various regions around the world. This initiative will be entitled the PVRI Global Registry Initiative.

During 2019, we were awarded a substantial grant from the Cardiovascular Medical Research and Education Fund (CMREF) to establish this initiative. The PVRI Global Registry Initiative is a tripartite approach that includes the following:

• GoDeep - a deep phenotyping database
  The GoDeep project is a deep phenotyping PH registry and will be led by Werner Seeger at the Justus-Liebig University Giessen, Germany.

• GoGlobal - a survey of PH registries where data cannot be included in GoDeep
  The GoGlobal project will be led by Paul Harbour at the Johns Hopkins School of Medicine, USA in partnership with researchers at the Hopkins Bloomberg School of Public Health.

• Global Health - advocacy to global health providers
  Global Health will be under the scientific leadership of Stuart Rich, Northwestern University, USA and Paul Corris, PVRI Chief Scientific Medical Officer, UK.

PVRI Global Health activities

Global Burden Disease (GBD)/the Institute for Health Metrics and Education (IHME)

The IHME database has been commissioned by the World Health Organization and is undertaken by Dr. Greggory Roth Adjunct Associate Professor of Global Health and Health Metrics Science at the IHME, and Associate Professor of Medicine/Cardiology at the University of Washington School of Medicine.

The IHME database is currently based on access to death certification records globally, where the cause of death has been right heart failure.

The first IHME report is expected in 2021. We aim to be fully prepared by developing position statements, guidelines and recommendations for governments, healthcare departments and other stakeholders, who seek information on PH/PVD and care provision for patients.

The World Heart Federation (WHF)

In 2018, the WHF gave their full membership status to the NCDA, which will allow free registration to various international meetings and symposia. Paul Corris and Majdy Idrees will be participating in the ‘Global NCDA Alliance Forum 2020’, which will take place from 9-11 February 2020, in Sharjah, United Arab Emirates.

As a member of the NCDA, the PVRI has access to the ‘Knowledge Action Portal’, which is an online communication tool supported by the WHO and visited by a wide range of stakeholders, including governments globally.

We aim to update the portal regularly, including features, such as work undertaken by our Regional and Disease & Specialty Task Forces.

PVRI Global Health Working Group

We have established a Global Health Working Group which will have its first meeting on 2 February 2020 after our scientific sessions in Lima.

The scope of the group will include:

• Development and implementation of a strategy surrounding the publication of relevant position statements, guidelines, country-specific documents for patient care in readiness for the WHF’s first release of data in 2020.

• Communication and advocacy to global health providers. This will include representation at various global health forums and symposiums and activity both on and offline.

These activities aim to achieve recognition of PH as an important, high prevalence, global disease that is currently largely neglected.
From a scientific standpoint, the kick-off presentations for the meeting were appropriate with a historical perspective on hypoxic pulmonary vascular contraction (Jason Yuan) and the role of redox signaling (Jeremy Ward). With pulmonary hypertension complicating many common diseases (HFpEF for group 2 and COPD for group 3) and not being considered rare any longer, a focus on the effects of PAH and other co-morbid conditions was is order, with various animated pro-con debates.

Delving on the complexity of the pulmonary hypertension syndrome, an update on ongoing large research activities, such as the Pulmonary Vascular Disease-omics (PVDOMICS) initiative, provided by Jane Leopold, was timely. In addition to important updates on clinical manifestations of the syndrome, basic haemodynamic measurements (with very informative presentations by Gabor Kovacs, Ronil Telford, Horst Oechslin, Euan Brittain, Jean-Luc Verheye, and Rebecca Vanderspork), and exciting new technologies. This included sound wave therapies for circulatory disorders in the Professor Stuart Rich Lecture, given by Hitoshi Shimakawa, or the breath volatolome for PAH-diagnosis, presented by Sylvia Cohen-Kaminsky.

There was a plethora of information on cutting edge basic research regarding cell heterogeneity and epigenetic regulation (Professor Sheila Claire’s Haworth Lecture’s masterly presentation, by Thomas Bis辜负), the importance of miRNA profiling (Stephen Chan and François Potus), methylation (Son Savai-Pullamsetti), and genetics (David Mottin).

The importance of registries for PAH patient care, research, world collaboration (an important focus of interest for the PVRI in the next five years) was emphasised in a section entirely devoted on the power of registries around the world with loads of information provided by Marc Humbert, Pieter Escobedo-Subías, Geoff Strange, Abdallah Al-Ossian, and Maria Jesus del Cerro-Marín.

Another focus was current and novel therapies and repurposing drugs for PAH from the industry perspective (Michael Colahan), seasoned clinicians (Ap Vonh and Diederik Slot, physician-scientist: Edith Franke Speikerkotter) and basic scientists (Francos de Marín).

A record 170 abstracts were submitted to the congress, which were presented over a number of sessions, including our increasingly popular rapid-fire format.

PVRI Award Winners
2018 Lifetime Achievement Award
Paul Corris.

2018 Achievement Award
Bradley Maron.

2018 Achievement Award
 Aaron Shefras.

2018 Certificate of Excellence
Silvia Nikolov and Peter Fernandes (AOV leaders).

2018 Rupert Swift Award
François Potus.

2018 Buitroux Foundation Young Investigator Award
Kurt Prins.

Fourteen medications targeting the endothelin, nitric oxide and prostanoids in pathways have been approved for PAH in the last 25 years. However, in the last five years, the most important recent advance in PAH medical management have not been related to the discovery of novel drugs or pathways, but to the development of better strategies with an emphasis on early combination therapy and escalation of treatments based on systematic assessment of clinical response using a multiparametric risk stratification approach. In that context, the current treatment algorithm provides the most appropriate initial strategy, including monotherapy, or more often initial combination therapy.

Further treatment escalation is required in cases low-risk status is not achieved in planned follow-up assessments. In the modern management era, many PAH patients are doing well while on therapy. However, a number of patients still do not reach the treatment goals with double or triple combination therapy and there is a clear unmet need to develop novel strategies and target novel pathways.

The meeting was a great success, with excellent conferences and discussions between presenters, moderators, and attendees. There were many highlights, including a keynote lecture on drug discovery and development in PAH since the early days of pure vasodilator therapies to the recent age of multiple approved agents targeting the three pathways. There was also a good analysis of a series of recent negative phase 2 clinical trials and a discussion on the best use of animal models for drug development. There were interesting talks on better use of surrogate endpoints and biomarkers and lessons from heart failure colleagues on how to develop creative strategies to improve success in Phase 2 trials. Further discussions focused on what to do when the primary endpoint of a study fails and whether responder and non-responder analysis can help us in negative clinical trials, with the examples of tocolzumab and mabsin studies in PAH. Current clinical trials targeting novel pathways were then presented with a lot of enthusiasm by different investigators (PAMP-1 inhibitor, frotoban, anti-endoglycine, dopamine ß monooxygenase inhibitor…).

The conference closed with a superb presentation on pulmonary artery denervation in PAH with a device-based approach.

After such a nice event, we look forward to the next PVRI Drug Discovery & Development Symposium, which will be held in Boston in July 2020.

See you there!
In 2019, the PVRI China Task Force has continued to encourage collaboration at international level. This effort has resulted in several international joint publications and promoted both clinical and translational science in PVD research in China.

The main activities include:

- Education courses on pulmonary vascular disease (PVD)
- National and international conferences
- Implementing national strategies in PE
- PVD management

Education courses of pulmonary vascular diseases (PVDs):

6-11 May 2019: The Pulmonary Hypertension Academy, a multicentre and multidisciplinary education platform for the diagnosis and management of pulmonary vascular disease, arranged a week education programme in Beijing China, with the participation of Peking Union Medical College, China-Japan Friendship Hospital, Peking Union Hospital, Beijing Friendship Hospital, Beijing Anzhen Hospital, Beijing Chaoyang Hospital and Beijing Hospital. More than 20 young fellows from different parts of China participated in the education programme. The Academy provides an excellent learning and communication opportunities for young physicians, who have a primary interest in pulmonary hypertension.

July 2019: Two education programmes, devoted to standardising the diagnosis and treatment of pulmonary vascular disease, were organised by the Chinese Medical Journal. These courses provided education on several important aspects, including diagnosis and treatment of pulmonary hypertension, as well as standardised operational procedures for imaging pulmonary hypertension. More than 10 multidisciplinary physicians jointly explored and contributed on the clinical practice of evaluation and management of pulmonary arterial hypertension.

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To provide support for individualised diagnosis and treatment of pulmonary embolism and pulmonary hypertension, a multidisciplinary consultation platform for diagnosis and management of pulmonary hypertension and pulmonary vascular diseases was established by the Assembly of PE-PVD group within the Chinese Thoracic Society. By the end of October 2019, more than 100 experts from 30 centres have participated in the programme. The platform provided an excellent learning and communication opportunities for physicians.

12-14 October 2019: To increase the awareness of thrombosis, further activities were conducted around World Thrombosis Day (12 October), including an educational event for patients and the public, MDT consultation for PE-DVT patients, social media activities for increasing awareness of WTD, and an education programme for physicians.

National and international conferences

14-16 April 2019: The 11th National Congress on Pulmonary Embolism and Pulmonary Vascular Diseases and the 9th International Conference of Pulmonary Circulation Disorders were held in Hohhot, China, with the collaboration of the Pulmonary Vascular Research Institute (PVRI) and the Chinese Thoracic Society (CTS). The symposium covered updated guidelines for pulmonary hypertension, CTEPH, and venous thromboembolism (VTE), with new information on the use of antithrombotics and thrombosis prevention. The latest knowledge to improve diagnosis, treatment and prevention of pulmonary embolism and pulmonary hypertension was shared. The PVRI also enhanced its educational programme during the meeting.

October 2019: The China Heart Congress, the one and a half day pulmonary vascular disease session was attended mostly by cardiologist physicians. Professors Xianhong Chen and Lan Zhao gave lectures in the meeting, covering topics on advances in PAH, as well as congenital heart disease and medical and surgical treatment of CTEPH.

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Implementing National Strategies on PE- and PVD programme

The formulation of national policies is an important prerequisite and guarantee for the prevention and treatment of PE-DVT. The Ministry of Health in China has clearly included PE-DVT prevention and treatment into hospital assessment indicators to ensure it is well implemented. During the year, more than 500 hospitals participated in the VTE prevention programme.

13-15 April 2019: The China National Standardised Diagnosis and Treatment System project was launched at the 11th National Congress on Pulmonary Embolism and Pulmonary Vascular Diseases and the 9th International Conference of Pulmonary Circulation Disorders.

13-17 October 2019: CTEPH Master Class, a multicentre and multidisciplinary education platform for diagnosis and management of pulmonary vascular diseases, arranged a week education programme in Peking Union Medical College, China-Japan Friendship Hospital. More than 30 young fellows from 30 hospitals all over China participated in this education programme, which provided an excellent learning and communication opportunities for young physicians with an interest in CTEPH.

The Pulmonary Vascular Research Institute (PVRI) China Task Force continues its active participation in organizing national and international conferences and education courses on pulmonary vascular disease (PVD) in China. The Task Force also aims to extend update understanding of pulmonary embolism and pulmonary hypertension among Chinese physicians, exchange clinical experiences and to create future opportunities for international collaboration.
PVRI Eastern Mediterranean & Saudi Arabia Report 2019
Paul Hassoun & Abdullah Al-Dalaan

Educational events
• Pulmonary Hypertension Master Class Course: 14-15 Oct 2019, King Faisal Specialist Hospital & Research Centre, Riyadh, KSA (in collaboration with University of California San Diego).

Regional activities
• 31st PVRI Annual World Congress on Pulmonary Vascular Disease, 31 January to 3 February 2019, Barcelona, Spain
• SAPH 12th Annual Conference, 14-16 February 2019, Manama, Bahrain (see following report)
• Middle East & North Africa PH forum, 25-26 October 2019, Dubai, UAE

Research (current initiatives)
• EMEA CTEPH Registry: An international prospective registry investigating the epidemiology, diagnosis and treatment of CTEPH patients in EMEA countries (RAC # 2161033)
• Expert Exposure Registry: Riociguat in Patients with Pulmonary Hypertension (RAC # 2161038)
• Systematic Prospective Follow Up for Better Understanding of Clinical Characteristics of Patients with PH Disease (SAUDIPH) Registry (RAC # 2171148)
• Prevalence of Genetic Factors in Saudi Patients with Idiopathic Pulmonary Hypertension (RAC # 2161062)
• Transition of Bosentan to Macitentan in the treatment of Pulmonary Hypertension – comparison of clinical outcome (RAC # 2171190).

12th Annual Conference of the Saudi Association for Pulmonary Hypertension (SAPH), 14-16 February 2019
SAPH2019 held in Bahrain on 14-16 February 2019, brought together physicians from the Middle East and North African countries and world experts in the field of pulmonary hypertension to engage and collaborate on the most advanced developments in the understanding and management of the disease.

The on-going collaboration between the SAPH and the Pulmonary Vascular Research Institute (PVRI) continues to be highly successful, as illustrated by the increased numbers of participants at SAPH 019, which included 30 international and regional faculty members, and 190 delegates (up from 120 in 2018).

The three-day event consisted of eight whole sessions on pulmonary hypertension, with 20-minute lectures followed by a 10-minute discussion. A Gala Dinner was held at the Ritz Carlton at the end of the second day.

An exceptional inaugural session, starting with an opening ceremony and welcome notes by SAPH 2019 representatives, was followed by lively and thought-provoking discussion, highlighting new technological innovations and medical findings, which were reviewed and debated by all speakers, including PVRI invited speakers Professors Allan Lawrie, University of Sheffield, and Luke Howard, Imperial College London, as well as international speakers.

Stimulating topics included:
• PH in paediatrics
• CTD-PH
• 6th World PH Symposium
• Lectures from PVRI invited speakers

Case study presentations covered the entire spectrum of PH, including PH in pregnancy and a case of post-pulmonary hypertension. Three discussants, leading challenging case presentations, analysed each presented case.

The conference concluded with a session on chronic thromboembolic pulmonary hypertension (CTEPH), led by the most prominent speakers. The quality-driven programme promoted active participation from the delegates.

We would like to thank the two PVRI sponsored speakers, Allan Lawrie and Luke Howard, for their input and contributions.

To continue to enhance the quality of the conference, SAPH2020, to be held on 13-15 February 2020 in Riyadh, Saudi Arabia, will compile a programme consisting of the most influential topic and shared practices around the globe.

It will also comprise an expert panel of prominent speakers, who will present their expertise in the field of pulmonary hypertension at national level.
The presentations of the meeting can be found in the PVRI website, accompanied by a short but focused comment prepared by Valentina Mercurio.

**Eastern Europe**
During the National Ukrainian Congress of Cardiology, held in Kiev, Ukraine, from 24-26 September 2019, the PVRI Eastern Europe Task Force, led by Yuriy Sireno, held a session dedicated to pulmonary hypertension.

Held in the Olympic Stadium Congress, Dr Abdullah Aldalaan, King Faisal Specialist Hospital & Research Center, Saudi Arabia, presented the following talks: SAUDI Pulmonary Hypertension Registry: Compared to west registries.

PVRI European Task Force 2019 Meeting
The second meeting of the Pulmonary Vascular Research Institute European Task Force was held on 11 October 2019 at Acireale, Sicily.

As in 2017, surrounded by the Sicilian beauty, numerous PVRI international scientists with deep expertise in pulmonary vascular diseases, presented the latest news on pulmonary hypertension (PH), discussing hot topics in this field. The audience, composed of many Italian physicians with interest in pulmonary vascular disease, was impressed by the high scientific profile of the meeting. Presentations, together with a commentary by Alessandra Cuomo and Valentina Mercurio, can be found on the PVRI website.

The main objective of the PVRI European Task Force is to create a network of European and International Colleagues and to share clinical projects in the field of PH. In particular, the main topic of this meeting was the usefulness of registries aimed at better characterising different phenotypes these patients.

As in previous years, many efforts have thus been spent in 2019 in this direction.

In **Group 1 PH**
The PVRI is sponsoring an international project (“SUPPORT”) led by Lucilla Piccarlari, Patrizio Vitulo and John Worth, aimed at clarifying the phenotype of such patients. The feasibility of the study was extensively discussed with the representatives of a dozen Italian centres of the PHNET network; experts in the management of the 3-PH group, in order to plan the kick-off of the Italian partner. Lucilla Piccarlari presented this project during the 2nd meeting of the European PVRI Task Force.

In **Group 2 PH**
The international multicentre collaboration aimed at gaining insights on reversibility or irreversibility of pulmonary hypertension is moving forward; results are expected at the beginning of next year.

In **Group 1 PH**
The international multicentre collaboration on pathophysiological objectives which will never be the aims of randomised clinical trials is also moving forward; results are expected at the beginning of next year.

The 2nd meeting of the European Task Force was held at Acireale in October. Again, as in 2017, the wonderful Sicily has hosted some of the best known PVRI scientists discussing hot topics in the field of pulmonary hypertension.
Annual Report 2019

This year we had two successful meetings to raise awareness about pulmonary hypertension.

12-13 April 2019 Paediatric PAH Symposium Mumbai, India

This one-and-a-half day symposium, co-sponsored by the PVRI and pharma, was held at Kokilaben Dhirubhai Ambani Hospital Mumbai.

Invited speakers sponsored by the PVRI were Professor Sheila Glennis Haworth CBE and Professor Shahin Moledina together with a host of Indian faculty. The symposium was attended by 87 delegates and was covered by nationwide live television, viewed by 122 delegates on day 1 and 110 delegates on day 2.

The event provided a broad insight into the diagnosis and management of paediatric pulmonary hypertension. The conference had a unique design. Each session had two to three didactic lectures followed by three case studies or research papers presented by young paediatric cardiology fellows.

Day 1

The symposium was opened with a welcome note from Dr Snehal Kulkarni and Dr Prashant Bobhate. This was followed by a keynote lecture given by Professor Sheila Glennis Haworth CBE, University College London, UK, discussing advances in diagnosis and management of paediatric PAH over the last five decades. This was followed by a presentation about expert panel on PAH registry from the UK by Dr Shahin Moledina, Great Ormond Street Hospital, UK, and Dr Sanjay from Kerala. Experience of the first paediatric PH registry from India was presented by Dr Anuj Sharma. The day progressed with presentations, including the diagnostic of PH, le echocardiogram, cardiac catheterisation, CT angiogram and exercise testing. Towards the end of the day, talks on the assessment and management of PAH in single ventricular physiology were delivered.

Day 2

Day 2 started with a session on management of paediatric PAH, which was chaired by Prof Dr. IBS Saxty and Dr Snehal Kulkarni. Dr Shahin Moledina presented his experience on upfront combination therapy vs sequential add-on therapy. This was followed by talks regarding assessment of PAH patients on medical therapy, management of PH in the intensive care unit, and caring of PH patients beyond medical management.

The second session raised awareness on the interventional management for patients with PH and the faculty shared their experience regarding the use of atrial flow regulator, Portashunt and lung transplant in India.

The last session of the day consisted of two very useful talks, given by Dr Shahin Moledina and Professor Sheila Haworth, on epigenetics and the future of managing paediatric PH. The conference ended by vote of thanks by Dr Prashant Bobhate.

13 October 2019 Paediatric Cardiac Society

Rajpur, India

The second event was held in association with the Paediatric Cardiac Society of India during their annual conference at Rajpur, India on 13 October 2019. This event had a special focus on humanitarian models of healthcare with an emphasis on developing and sustaining paediatric cardiac programmes in lower and middle-income countries.

The faculty included Dr Mark Cawling, Dr S. Radhakrishnan, Dr Prashant Bobhate and Dr Shriram Kumar. The dedicated PAH session had three lectures followed by three interesting case presentations by young fellows.

2020 Objectives

We have planned a two-day symposium, which is going to be co-sponsored by the PVRI and the international faculty for the same are Professor Pad Hanssen, Johns Hopkins University Medical School, USA, and Professor Michele D’Alto, Ospedale Monaldi, Italy.

The conference, which will be held in Mumbai, is aimed at physiicians, rheumatologists, pulmonologists and cardiologists interested in managing pulmonary hypertension.

More information will be published on the PVRI website.

Annual Report 2019

The Iranian Pulmonary Hypertension Association started in 2014 with the aim of bringing pulmonologists, cardiologists and rheumatologists involved in the field of PVD under one roof, as well as enhancing the quality of service provision and reducing the mortality rate of patients with pulmonary hypertension (PH).

To achieve this, the following was agreed:

• Registration of patients.
• Increasing the number of PH centres.
• Organising educational congresses and programmes.

2019 activities

• Enhancing the understanding of the disease by holding PVD educational programmes every four months in an academic centre.
• Two CME programmes, each lasting for 16 hours, were held in Zanjan and Urmia.
• Two clinical workshops for education of the right heart catheterisation were hosted for pulmonologists in Isfahan and Tehran centres.
• Specific registration systems were operated in six centres: Tabriz, Mashhad, Tehran, Ahvaz, Isfahan and Shiraz, for the purpose of data collection and participation in international research projects in collaboration with European centres.

The 2nd Pulmonary Vascular Diseases Summit Tabriz, Iran, was held on 27-28 June 2019 with sponsored speakers from the PVRI (see following report).

• The 9th Iranian International Congress of Pulmonary disease, critical care and tuberculosis, was held on 12-15 October 2019 in Tehran, Iran. The event was endorsed by the American Thoracic Society (ATS), the European Respiratory Society (ERS) and the Pulmonary Vascular Research Institute.

The pulmonary hypertension session was moderated by Dr Majid MalekMohammadi (Pulmonologist) and Dr Babak Shaer Khachani (cardiologist), Professor Meinhard Kneussl (ERS expert) was a keynote speaker and touched on the topics of pathogenesis, diagnosis and treatment of pulmonary hypertension.

We would like to thank three PVRI sponsored speakers for their input and contributions:

• Professor Marion Dalicqox
• Professor Irene Lang
• Professor Gerald Simonneau.

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The organisers would like to thank all invited speakers, delegates and moreover, all colleagues, who endorsed the congress.

3rd Pulmonary Vascular Diseases Summit Tabriz, Iran, 27-28 June 2019

The international PVD Congress is held biannually by IPHA and the Iranian Society of Pulmonology in Tabriz, Iran. The last Congress was held on 27-28 June 2019 in Tabriz, Iran.

It was well attended with several delegates, including pulmonologists, cardiologists, rheumatologists, internists and GPs. The Congress was officially opened by Professor Gérard Simonneau. Emeritus Professor at the Paris-Sud University, France, and esteemed PVRI Board member.

The CTEPH session, Evaluation of CTEPH patients for anastomosis and management of complications, provided by video conference by Dr Michael Matani (USA), attracted a strong audience.

A second video conference showed Dr Stefano Barco (Germany) presenting two topics:

• Prevention of early complications and late consequences after acute pulmonary embolism
• Use of old and new anticoagulants in special population short bowel syndrome, anti-phospholipid syndrome, elderly pregnancy.

The faculty included 12 international speakers from Belgium, Czech Republic, France: Germany, Japan, Turkey, United States.

PVRI India Report 2019

Prashant Bobhate

PVRI Iran Report 2019

Farid Rashid
In 2019, the North American Task Force has been extremely busy in promoting pulmonary vasculature and right ventricular based research. Indeed, several of our members, Anna Hennes; Steeve Provencher and Sébastien Bonnet, with the help of international members, Soni Savai-Pullamsetti, Joan Albert and Christophe Guignabert, have been in charge of organising the PVRI 13th Annual World Congress in Barcelona, which has been the most attended event in PVRI history.

The meeting offered a new format to our annual meeting with new abstract-based sessions, including oral and rapid-fire sessions (see full report on page 42).

In May 2019, the PVRI was well represented in Dallas at the International American Thoracic Society (ATS), which was organised by Professors Vinicio De Jesus Perez and Sébastien Bonnet. Numerous PVRI members, especially young investigators, had the chance to present their work, and an outstanding and passionate debate on the use of animal models in pulmonary hypertension research took place.

Finally, in November 2019, several members of the PVRI attended the International American Heart Association (AHA) meeting, where two prestigious awards were given to outstanding members of our Task Force. Professor Evangelos Michelakis received the Kenneth D. Bloch Memorial Lecture Award, while Professor Paul Yu received the Dickinson Lecture Award.

Finally, we wish to congratulate several members of our Task Force, who have been nominated to prestigious and important positions within the community, such as Dr Elena Goncherova elected Chair of the Programme Committee for pulmonary circulation of the ATS and who will help Professor Sébastien Bonnet in his duties as Chair; Dr Paul Yu, nominated chair of the 3CPH SCILL Committee at the AHA; Dr Vinicio De Jesus Perez who will assist Dr Yu as the SCILL Vice-Chair; and Professor Sébastien Bonnet, nominated Vice-Chair and upcoming Chair of the 3CPH council of the AHA, and will assist Professor Fumito Ichinose.

In 2019, the South American Task Force has been extremely busy in promoting pulmonary vasculature and right ventricular based research.

Indeed, several of our members, Anna Hennes; Steeve Provencher and Sébastien Bonnet, with the help of international members, Soni Savai-Pullamsetti, Joan Albert and Christophe Guignabert, have been in charge of organising the PVRI 13th Annual World Congress in Barcelona, which has been the most attended event in PVRI history.

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On 20 August 2019, in conjunction with Dr Sonia Muñoz and sponsored by Janssen, the Task Force organised a workshop on pulmonary hypertension in Children at Altitude in Pasto, South of Colombia, which is located at 2,550 metres above sea level. This workshop was attended by approximately 30 participants.

On 5–8 November 2019, the 17th Congress of the Latin Society of Pediatric Cardiology and Pediatric Cardiovascular Surgery was held in Buenos Aires, Argentina. Esteemed PVRI member Dr Gabriel Diaz presented a talk on pulmonary hypertension. Dr Diaz hopes to share insights of this event in Lima.
The Symposium Scientific Committee would like to extend their thanks and gratitude to the esteemed faculty for making this event a memorable one.

The Symposium on PH in Children will take place in Panama City in 2021.

In this regard, we have taken the following steps:

- The collaborative PH registry continues to grow with the intent that this will provide authorities and funders with a clearer perspective on the PH landscape in South Africa. The total enrolment is now over 300 patients (mainly incident), almost split equally between the Groote Schuur Hospital—University of Cape Town PH Clinic, representing largely state sector patients, and the Milpark Hospital Centre of Chest Disease PH Clinic representing mainly healthcare insured patients. This REDCap-based registry, modelled on the REVEAL registry, will provide not only epidemiological data, but outcome data to this registry. We remain convinced that the only way to overcome the inbuilt inertia we seem to experience is by providing high quality data of the existence of an unmet need.

- The collaborative efforts are also well underway in the planning and organisation of the upcoming collaborative meeting, co-hosted the PVRI Working Group for PH in South Africa and the Jenna Lowe Trust, to be held in March 2020. The meeting will consist of a 2-day medical conference in Johannesburg with an International Faculty per kind sponsorship of the PVRI addressing broad topics of relevance to a general pulmonology and cardiology forum. This will be followed by an additional 2-day patient-centred forum in Cape Town addressing the needs and interests of an increasingly well informed patient advocacy group. This aspect of the meeting is being arranged by the Jenna Lowe Trust, named after an inspirational patient who, along with her equally inspiring parents and sister, went through the tumultuous journey from PH patient to transplant recipient.

This story is tough yet inspirational and well told by the patient’s mother, Gabi, in her recent book “Get me to 21”. We hope to increase both public and medical awareness in this field with this collaborative approach, as well as demonstrate to our patients that their voices are being heard.
Improving the lives of PH patients in Asia Pacific - the right decisions at the right time.

The APAC PH Forum was held in the Marriott Marquis Queen’s Park Hotel, Bangkok, Thailand, on 2–4 August 2019. The forum was attended by a total of 181 delegates from many countries around the world and was sponsored by Actelion.

Scientific Committee members consisted of:
- Kritvikrom Durongpisitkul, Thailand
- Eli Gabbay, Australia
- Gao Hong, Canada
- Hsao-Hsun Hsu, Taiwan
- Decho Jakrapanichakul, Thailand
- Eli Gabbay, Australia
- Geetha Kandavello, Malaysia
- Andrea Low, Singapore
- Annette Pidoux, Australia
- Tan Ju-Le, Singapore

The delegates and speakers attended from the following countries:
- Australia
- Hong Kong
- Japan
- Malaysia
- New Zealand
- Singapore
- Taiwan
- Thailand
- The Philippines
- Vietnam
- United Kingdom

Delegates from the following specialties were represented:
- Cardiology
- Cardiothoracic
- Imaging
- Nursing
- Pharmacy
- Respiratory medicine
- Rheumatology

The forum aimed to share information and best practice. It also encouraged collaboration and friendship amongst professionals in Asia to further improve the lives of our PH patients.

Forum objectives were to:
- Identify barriers to a timely diagnosis, reasons for missed diagnoses and misdiagnoses to enable early detection and management
- Share updates from international guidelines and WSPH 2018, and discuss pragmatic application in the APAC region setting, particularly around:
  - routine risk stratification and follow-up risk assessments - early access to optimal treatment strategies
- Share current best practice and new innovative research in the region so that less well-developed services have a usable model to implement in their own setting
- Increase regional collaborations and foster research relationships, with a focus on establishing PH registries using contemporary tools.
The earliest manifestations of PH are extensively affected by exercise. The exercise task force membership published an annual report in 2019.

We have:
- demonstrated a metabolic plasma signature in ePH
- demonstrated dynamic RV/pulmonary vascular uncoupling by ICPE in ePH2
- demonstrated dynamic RV/pulmonary vascular uncoupling by ICPE in HFpEF3
- described abnormal pulmonary vascular distensibility and blood flow gradient as early findings in PAH4-5
- established an international registry for ePH
- https://clinicaltrials.gov/ct2/results
- https://ichgcp.net/clinical-trials-registry/NCT03954574

Goals/Aims for 2020
- Deep phenotyping of ePH, including imaging and transpulmonary flux of biomarkers (metabolomics, proteomics, transcriptomics and coagulomics)
- Further research into dynamic RV/pulmonary vascular uncoupling to exercise performance and long-term outcome
- Publication of a high impact review of ePH

References
This year, the PVRI High-Altitude Task Force has been very active in scientific interactions. It participated in the 12th Annual World Congress of the International Society of Mountain Medicine, in Nepal.

It is important to emphasise that most of the participants were young women researchers and scientists. Their exposure was part of their educational training and skill development. In addition to their active participation in the scientific presentations, they interacted with several eminent researchers in the field. Another significant development was the participation from Defence organisations, especially the Defence Institute of Physiology & Allied Sciences (DIPAS), Delhi, India. It was an opportunity for my DIPAS colleagues, and several of our PVRI friends, to interact and share scientific interests.

Dr Aastha Mishra, a PVRI member who delivered a talk in one of the sessions, was considered one of the best young speakers by the international jury and was awarded for her excellent work. Among other young researchers, Drs Manjari Rain, Kavita Sharma, Tsering Palmo and Manjula Miglani provided oral or poster presentations.

Dr Qadar Pasha was assigned responsibility to organise and chair a special session on high-altitude research activities by the ISMM scientific committee, as well as delivering in another session; Dr Max Gassmann also delivered a very dynamic and well-received presentation.

Other meetings & conferences
Dr Qadar Pasha was invited to participate and deliver talks in the following two international conferences:
• The PVRI 13th Annual World Congress on PVD, Barcelona, Spain, in January 2019.

It is with mixed feelings that we announce Dr Ghulam Mohammad’s retirement after 30 years of dedicated service to his patients. He was a Senior Consultant and Head of Department of Medicine at the SNM Hospital, Leh, Ladakh; one of our most active PVRI family members from India, and a great friend and collaborator in our high-altitude research. Colleagues at the SNM Hospital gave him a befitting farewell. We salute our colleague for his great contribution to the medical field and scientific initiatives. His contribution has also been enormous in arranging the volunteer subjects at the hospital, whether controls or patients of HAPE (high-altitude pulmonary oedema). He was highly instrumental in developing the High-altitude research Lab at the SNM hospital. He continues his private clinic work and, in addition, has also joined other private hospitals in Leh, as a Consultant. We wish him a healthy and beautiful life ahead and every success in his professional activities.

Future Conference
The 6th International Leh Symposium is to be organised in September 2020, at the most scenic and equally most peaceful land of Buddha, Leh, Ladakh, India.

Further details will be available on the PVRI website in due course, but we invite our all PVRI family members and colleagues from other fields to attend this event.
The PVRI Imaging Task Force continues to grow with membership from different communities, including clinical, imaging, physics and modelling backgrounds.

Achievements in 2019

- The second dedicated PVRI Imaging Symposium was held in Barcelona in 2019. There were over 30 attendees, including new members to the group. Presenters and attendees came from Europe, Africa, the USA and the Asia-Pacific region. The programme offered an overview of state-of-the-art imaging in pulmonary hypertension, imaging approaches in pulmonary embolism, real world challenges and future perspectives such as imaging of vessels, the clinical use of complex measures and the use of artificial intelligence approaches.
- A comprehensive consensus statement by the PVRI Imaging Task Force was published in Pulmonary Circulation in 2019. This included 27 authors from Europe, America, Africa and Asia-Pacific, spanning 10 countries and representation from physicians, radiologists, imaging scientists and computational modellers. This is the first expert consensus publication specifically dealing with imaging and pulmonary hypertension and includes a state of the art overview of the diagnostic pathway for patients with suspected pulmonary hypertension, including 55 summary statements, based on a Delphi approach.
- We encourage members of the PVRI to review the guideline and consider its adoption in their clinical practice.

Goals for 2020-2022

- Aim to contribute regularly to web-based interactive cases on PVRI site
- Explore imaging practices worldwide by conducting and auditing of current approaches within the PVRI community
- Identify priority workstreams from the PVRI meeting in Peru
The Innovative Drug Development Initiative, formerly known as the PVRI Pharma Task Force, was established in January 2017 under the leadership of Paul Corris (PVRI), Sylvia Nikkho (Bayer) and Peter Fernandes (Bellerophon).

Its mission is to provide a platform for academia, the pharmaceutical industry and drug regulators to openly discuss questions surrounding the future of trials comprising novel drugs. It encourages early and continuous dialogue with all stakeholders, including the patient’s perspective, on innovative clinical and regulatory development strategies, with the ultimate aim of finding new ways to fight and treat pulmonary vascular disease and pulmonary hypertension. The IDDI is an integral part of the PVRI’s strategic mission and four backbone initiatives. The current aim of the IDDI is to establish a series of position papers, developed by each IDDI Workstream. These will be released as abstracts, posters, oral presentations and peer-reviewed articles, which will be published in the PVRI’s journal Pulmonary Circulation. The purpose of these statements is to enhance global understanding of the disease and provide a valuable insight into the complexities surrounding new drug discovery in this disease field.

IDDI Workstreams

During 2019, an additional workstream on Repurposing Drugs was formed, in addition to the already established three workstreams of Endpoints, Clinical Trial Design and Biomarkers. All four workstreams are interlinked and mutually supportive, and are led by an academic and a pharma representative:

- **Clinical Trial Endpoints led by**
  - Olivier Sitbon (University Paris-Sud, France)
  - Sylvia Nikkho (Bayer, Germany)
- **Clinical Trial Design led by**
  - Jim White (University of Rochester, USA)
  - Peter Fernandes (Bellerophon, USA)
- **Biomarkers led by**
  - Anna Hommes (Vanderbilt University, USA)
  - Lawrence Zisman (Gossamer Bio, USA)
- **Repurposing Drugs led by**
  - Karl Peters (University of Minnesota, USA)
  - Mark Sudman (Bellerophon, USA)

**Activities during 2019**

Two IDDI Open Forum meetings were held during the year, attracting audiences of over 100 participants, including representatives from academia, patient organisations, regulatory agencies and the pharmaceutical industry. Stimulating presentations and interactive discussions were held at the:

- PVRI Annual World Congress, Barcelona, Spain, January 2019
- PVRI Drug Discovery & Development Symposium, Paris, France, July 2019

In addition, regular telephone conference calls were held throughout the year by each of the respective workstreams, and a meeting of the IDDI workstream leaders was held during the ERS Conference in Madrid in September 2019 to monitor progress on the publication of position statements.

**Clinical Trial Endpoints Workstream**

The workstream on novel endpoints, chaired by Olivier Sitbon and Sylvia Nikkho, includes 27 members and investigates the potential role of utilising composite improvement endpoints, as well as risk scores, as novel clinical endpoints in PAH clinical trials.

In Barcelona, the endpoint break-out session was led by Sylvia Nikkho and Olivier Sitbon. The work was supported by Paul Corris, Christian Meier and Werner Steeger. It focused on composite improvement endpoints, which were already used in clinical trials and allowed individual responders to be identified in a much shorter period of time in comparison to time to clinical worsening endpoints. This changed the placebo response and thereby required a smaller number of patients. However, while these could be clinically meaningful and discriminative of treatment effects based on available post-hoc clinical trial analyses, as for composite improvement endpoints, it was agreed that in future PAH trials treatment effects needed to be compared in addition to dual or even triple therapy.

The aim of the planned publication is to discuss the potential of composite improvement endpoints, as well as risk scores, as novel clinical endpoints in trials, while considering established clinical endpoints as taken care of by Marion Delcroix, Joanna Pepke-Zaba and Gérald Simonneau in the important context of the regulatory and patient’s perspective, and close interaction with the members of the other workstreams.

**Endpoints: Achievement & Plans**

**2020**

- Explore requirements for a potential novel clinical endpoint including patient’s perspective
- Complementary analysis of components of risk scores, evaluation of their weights using available databases
- Evaluation of risk scores used in registries/post-hoc clinical trial analyses

**2019**

- Position Paper
- Evaluation of composite endpoints used in clinical trials
- Evaluation of established endpoints
Clinical Trial Design Workstream

The workstream, chaired by Peter Fernandes and Jim White, discussed the following topics during 2019 with focus on the use of actigraphy to monitor changes in physical activity (PA) and its evolving role in pulmonary hypertension clinical drug development as a primary regulatory endpoint gaining acceptance by regulators for Phase 3 clinical studies.

Patient-focused drug development incorporates the VOICE OF THE PATIENT (VOP) and ultimately places the patient’s first when considering the risk and benefit of treatment. Discussions with patient focus groups have reported that daily physical activity is most meaningful to those living with this disease (FDA PFDD Meeting 26 September 2014) and aligns with every-day management of their illness.

The objective measurement of PA monitoring devices offers a reliable measure of a patient’s daily activity that can be utilised to measure this clinically meaningful outcome. The Food & Drug Administration (FDA) in the USA has recently accepted physical activity as measured by actigraphy as an acceptable regulatory endpoint for a Phase 3 study in IPF. In order to apply this endpoint to other cardiopulmonary disease clinical study designs, the development team will need to deliberate on key questions outlined below:

- What is the minimal % change in PA that is considered meaningful MCID?
- How will MCID be pre-determined/pre-specified?
- What other qualitative data is needed to support this MCID - ie, PROs?

Further working group discussions are planned to outline and deliberate on the role of innovative trial design, including the use of better stratification of patients at enrollment, enrichment strategies, adaptive approaches to trial design and withdrawal studies. The IDDI workstream will collaborate on harmonising their approach towards developing a Master Protocol with actigraphy as a primary endpoint to facilitate its implementation into future cardiopulmonary drug development programmes.

Biomarkers Workstream

The working group on Novel Biomarkers, chaired by Lawrence Zisman and Anna Hemnes, explores this rapidly developing scientific area. The challenges of both clinical trial design and novel surrogate endpoints in Phase 2 studies of anti-proliferative approaches are being considered.

In Paris, the Biomarker subteam provided an overview of biomarkers for PAH. An emphasis was placed on the potential use of biomarkers to predict response to a therapeutic intervention. New approaches to circulating biomarkers were discussed, as well as imaging of the right ventricle and pulmonary vasculature. The European Medicines Agency’s (EMA) view of surrogate endpoints was reviewed and it was recognised that there are currently no validated biomarker surrogate endpoints that meet regulatory criteria for use as approvable endpoints in PAH. Indeed, even NTproBNP does not meet the criteria for use as a surrogate endpoint based on EMA guidelines. Nevertheless, NTproBNP and potentially other biomarkers, have utility to examine the effects of therapy, as well as use as prognostic indicators. For example, HOXDIP and CDH4-15, in particular, are candidate biomarkers. Omic approaches to circulating biomarkers were reviewed, including genomics, transcriptomics, proteomics and metabolomics. An example from the field of proteomics was described wherein a panel of 9 proteins showed prognostic power independent of the REVEAL score. New approaches, such as the use of circulating exosomes, single-cell RNA sequencing, and cell free DNA were reviewed.

Imaging approaches to the right ventricle were discussed, including the use of RV EF from CMRI, and the use of RV strain from echocardiography. 3D reconstruction of the pulmonary vasculature from standard CT images has been reported. Use of 18FDCG and 18FFLT PET imaging have also recently been reported in PAH in relatively small studies. Hyperpolarized Xenon MRI of the lungs is a relatively new technique that allows examination of precapillary blood flow in a quantitative manner and may have utility in evaluating the effect of disease modifying agents in PAH.

Finally, the growing interest in the use of artificial intelligence and machine learning with potential applications to imaging and analyses of large omic databases were reviewed.

Repurposing Drugs Workstream

Launched in February 2019, the workstream Repurposing Drug for PH is chaired by Mark Toshner and Kurt Prins and focuses on developing a roadmap for future drug repurposing, as outlined below.

Work stream members:
- Mark Toshner
- Kurt Prins
- Edda Spiekerkotter
- Harm Bogaard

Introduction

- Introduction to the concept of drug repurposing.
- Place PAH/PH in the wider context of the drug repurposing landscape.
- Brief summary/history of repurposing in PAH.
- Outline of challenges – present and future.

Preclinical pipelines for drug repurposing

- Offering screen approaches.
- Success of screen vs targeted approaches.
- Challenges of animal and cell modelling.
- Use of genetic data.
- Mendelian randomisation.

Repurposing in the modern era

- Summary of clinical trials.gov data.
- Investigator-led studies are more likely to test novel therapeutic strategies, but these are single-centre and have difficulty recruiting and finishing.
- Industry-led studies are expensive, unlikely to test novel therapeutic strategies, but multi-centre and recruit and finish more often.

Challenges in repurposing and trial design

- Industry-wide problems, not exclusive to rare diseases.
- The role of stratiﬁed vs non-stratiﬁed approaches.
- Evolving role of small biotech.

Objectives for 2020

The goal of the IDDI for the next year is to organise a meeting for all workstream leaders during the 3rd PVRI Annual World Congress in Lima, Peru, on 31 January 2020, as well as an ‘IDDI Open Meeting’ immediately following the 7th PVRI Drug Discovery & Development Symposium, which will be held in Boston in June 2020. Emphasis during the year will be to finalise the individual position statements of each workstream ready for publication in Pulmonary Circulation. In addition, the IDDI should crystallise a unified vision for its future that will best serve all its members and diverse stakeholders in the long-term.

A note of thanks

We would like to express our sincere thanks to the dedicated leaders and members of all the four IDDI workstreams.

We greatly appreciate the support from all PVRI Roundtable members, who are part of the IDDI.
The first international meeting of the iPVD consortium took place in Aswan, Egypt from 21-22 October 2019.

The first seminar was chaired by Professor Stephen Chan, USA, and Dr Ahmed ElGuindy, UK. Discussants were Dr Rajkumar Savai, Germany, and Dr Matthias Clauss, USA, with a special remote presentation given by Dr Soni Savai-Pullamsetti, Germany. They discussed the role of inflammation and immunology on pulmonary arterial hypertension and with infection in particular. This was followed by a seminar on the epidemiology of schistosomiasis, HIV, tuberculosis in Africa with Professor Ana Olga Mocumbi, Mozambique, and Dr Harriet Maquita Rizza, Uganda, with a special remote presentation from Professor Russell Stothard, UK, and Dr Amaya Bustinduy, UK.

The topic of HIV and pulmonary vascular diseases was covered in Seminar 2, which was chaired by Dr Navneet Dhillon, USA, with active participation from Professor Stephen Chan, Dr Vinicio de Jesus Perez, USA, and Dr Matthias Clauss, USA. During this session Dr Nicola Petrosillo, Italy, presented the clinical aspect of HIV, pulmonary hypertension via an online presentation.

The final seminar on the first day, chaired by Professor Butrous, and Dr Clauss, was devoted to the discussion of new unpublished findings with a research project on the role HIV and schistosomiasis co-infection in pulmonary vascular disease. New emerging data were presented by Professor Cogolludo, Edgar Gonzalez Fernandez Malave, Spain, and their research fellow Sandra Medrano Garcia. Various topics were discussed, including the role of rheumatic heart diseases, pregnancy and regional and ethnic variation of the response to the current medication of pulmonary hypertension, in addition to the current epidemiological survey and tools in Africa with pulmonary hypertension. A business meeting of the iPVD was devoted to the discussion of new ideas and strategies to develop the work in this field. These included the education and publication strategy, research projects and collaboration, and potential grant applications. In addition, the structure and function of the iPVD consortium was discussed.

The attendees and faculty enjoyed the welcome reception and the Gala Dinner. The iPVD consortium would like to thank the CMFREF for their generous grant to support this symposium, the Magdi Yacoub Foundation, Aswan Heart Centre, and the PVRI Infection & Pulmonary Hypertension Task Force for providing all the logistics.

The subject of schistosomiasis and pulmonary vascular diseases was extensively discussed and chaired by Professor Angel Luis Cogolludo, Spain, and Dr Ahmed ElGuindy, Egypt. Professor Ghazwan Butrous, UK, and Professor Ana Olga Mocumbi, Mozambique, debated the pathobiology the clinical problem in Africa. The discussion was enriched by Dr ‘Azza Farrag’ special presentation of the past and present of schistosomiasis in Egypt.

1 Full recording of the meeting is available at PVRI website.

2 Notes

To improve knowledge, encourage research and optimise the delivery of clinical care of neonates, children and adolescents with pulmonary vascular disease.

Members: 57

We welcome the following new members to the Task Force:
• Dr Xue Qin Liu
• Dr Mariana Cazalas
• Dr Susan Pumacayo-Cárdenas

Missions
To improve knowledge, encourage research and optimise the delivery of clinical care of neonates, children and adolescents with pulmonary vascular disease through:
1 Facilitating the growth of networks for research and clinical care between professionals from different medical specialties, institutions and countries especially in less privileged communities.
2 Disseminating knowledge about the paediatric pulmonary vascular phenotype in clinics caring for adults with pulmonary vascular disease and facilitating smooth transition of childhood survivors of pulmonary vascular disease to adult clinics.
3 Promoting concepts about the foetal and developmental origins of pulmonary vascular disease, and the impact of early events on risk and severity of childhood and adult disease.

2019 activities
• Paediatric & Congenital Heart Disease Task Force Meeting during the PVRI Annual World Congress on PVD in Barcelona January 2019.
• 12th International Conference Neonatal & Childhood Pulmonary Vascular Disease, March 2019, San Francisco, CA, USA, organised by Jeff Fineman (Co-chairs; Shahin Moledina, Maria Jesus del Cerro).
• PPHNET Annual meeting, held in San Francisco, March 2019, organised by Steven Abman.
• 4th European Conference on Paediatric Pulmonary Vascular Disease, Groningen, (Netherlands) 10-12 October 2019, organised by Rolf Berger.

2019 Achievements
We published the results of the survey Acute vasoreactivity test and use of Calcium Channel Blockers in idiopathic Paediatric Pulmonary Hypertension, conducted among the members of the PVRI Paediatric & Congenital Heart Disease Task Force, PPHNET and REHIPED members.

2020 Goals
• To hold the 13th International Conference Neonatal & Childhood Pulmonary Vascular Disease, March 2020, San Francisco, CA, USA, organised by Jeff Fineman.
• Celebrate the 10th Annual Meeting of the Spanish Registry on Paediatric Pulmonary Hypertension in Madrid, Spain, on 21 February 2020.
“Today’s work, tomorrow’s possibility...”