Pre-Congress MEETINGS

Wednesday 25th January 2017

PVRI Board of Directors Meeting
Chair: PVRI President Paul Corris
1000-1700

PVRI Molecular/Animal Task Force & The Women’s Health Task Force
Sex Talks
Chairs: Anna Hemnes, Mandy Maclean
Aromatase inhibitors such as anastrozole are being considered as novel therapies for PAH. We aim to discuss the sometimes paradoxical pre-clinical data and clinical data surrounding the roles of endogenous estrogens and estrogenic pathways in PAH, women’s health and pregnancy.
• How do we reconcile these, what are they telling us about our models and experimental design?
• How should we translate pre-clinical findings to the clinic so that we facilitate new therapies becoming available to our patients?
1000-1700

PVRI Paediatric Task Force Meeting
Chairs: Ian Adatia, Maria del Cerro
1400-1700

PVRI Board of Directors Dinner
1900-2200
# 11th PVRI Annual World Congress on Pulmonary Vascular Disease

## Thursday 26th January 2017

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<thead>
<tr>
<th>Time</th>
<th>Session</th>
<th>Speaker(s)</th>
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<tr>
<td>0800-0810</td>
<td><strong>Plenary 1</strong></td>
<td><strong>Development, stem cells &amp; pulmonary hypertension</strong>&lt;br&gt;Chairs: Mark Ormiston, Kurt Stenmark</td>
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<tr>
<td>0810-0840</td>
<td><strong>Professor Sheila Glennis Haworth Lecture:</strong></td>
<td><strong>Advances in lung development: Relevance to pulmonary hypertension</strong>&lt;br&gt;Ed Morrisey</td>
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<tr>
<td>0840-0855</td>
<td><strong>Introduction to IPSC &amp; CRISPR</strong></td>
<td>Darrell Kotton</td>
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<td>0855-0910</td>
<td><strong>Historical perspective of stem cells in PAH</strong></td>
<td>Mark Ormiston</td>
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<tr>
<td>0910-0925</td>
<td><strong>Inducible pluripotent stem cells &amp; CRISPR for disease modeling</strong></td>
<td>Eric Austin</td>
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<td>0925-0940</td>
<td><strong>Bioengineering the lung</strong></td>
<td>Harold Ott</td>
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<tr>
<td>0940-1000</td>
<td><strong>Discussion</strong></td>
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<tr>
<td>1000-1015</td>
<td><strong>Break</strong></td>
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<tr>
<td>1015-1045</td>
<td><strong>Plenary 2</strong></td>
<td><strong>High altitude, oxygen sensing &amp; metabolism</strong>&lt;br&gt;Chairs: Stephen Archer, Ken Weir</td>
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<tr>
<td>1045-1100</td>
<td><strong>HIF-2α &amp; pulmonary hypertension</strong></td>
<td>Andrew Cowburn</td>
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<tr>
<td>1100-1115</td>
<td><strong>Adapting to high altitude: Epo &amp; iron</strong></td>
<td>Max Gassmann</td>
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<td>1115-1130</td>
<td><strong>Novel HIF-1α independent roles of the VHL protein signaling</strong></td>
<td>Evangelos Michelakis</td>
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<td>1130-1145</td>
<td><strong>Therapeutics for high altitude illness</strong></td>
<td>Erik R Swenson</td>
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<td>1145-1215</td>
<td><strong>Discussion</strong></td>
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<tr>
<td>1215-1330</td>
<td><strong>Lunch</strong></td>
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**Pulmonary Circulation Editorial Board Meeting**
Controversies in PAH: It’s debatable

RV failure in PAH is initiated by RV ischemia
Moderator: Lisa Mielniczuk // Pro: Harm Bogaard // Con: Ryan Tedford
This debate encourages the audience to consider the RV and vasculature in PAH as an integrated unit to consider the role that vascular disease plays in the RV, not just in the pulmonary circulation.

Use of endpoints that combine morbidity and mortality, overstate therapeutic benefit of PAH therapies
Moderator: Ron Oudiz // Pro: Paul Forfia // Con: Thenappan Thenappan
This debate will expose a concerning trend in study design in which morbidity endpoints are combined with mortality, creating a composite endpoint that is called ‘mortality’ which may overstate the efficacy of new therapies.

PAH can be cured with early combination therapy
Moderator: Ioana Preston // Pro: Paul Hassoun // Con: Duncan Stewart
This debate considers whether adding more expensive PAH drugs is wise or whether we are targeting the wrong pathways and need entirely new classes of therapeutic agents.

The hemodynamic criteria for diagnosing Group 1 PH are wrong: The criteria for wedge and mPAP pressures are too high
Moderator: Joanna Pepke-Zaba // Pro: John Ryan // Con: Hap Farber
This debate encourages the audience to consider the consequences of allowing wedge pressure >15 to be consistent with PAH, particularly as Group 2 PAH is much more common and can be similar in appearance on echo. The risk of indication drift to include Group 2 patients is considered (particularly in light of Group 1 therapeutics being generally ineffective in Group 2 patients). In addition, new data suggests that the mPAP threshold to diagnose PAH may currently be set too high.

Break

Moderated poster session 1: Clinical
Moderated poster session 2: Basic science
Moderators: Elena Goncharova, Brian Graham, Soni Pullamsetti, Usha Raj, Martin Wilkins
Several senior moderators will ensure every poster is briefly presented by the author and discussed.

END OF DAY ONE SCIENTIFIC SESSIONS

PVRI ANNUAL GENERAL MEETING FOR ALL PARTICIPANTS & GUESTS

NETWORKING COCKTAIL RECEPTION FOR ALL PARTICIPANTS & GUESTS

PVRI Pharma Task Force Meeting
PVRI Publications Committee Meeting & Dinner
Plenary 3
What’s hot in the basic science of pulmonary hypertension
Chairs: Jane Leopold, Mandy MacLean

miRs in PAH // Sebastian Bonnet 0800-0820

Reduced BMPR2 promotes endothelial-to-mesenchymal transition via HMGA1 and its target slug // Marlene Rabinovitch 0820-0840

The Wnt pathway in PAH // Vinicio de Jesus Perez 0840-0900

Discussion 0900-0920

Fatty acid metabolic defects & right ventricular liptoxicity in human pulmonary arterial hypertension // Evan Brittain 0920-0940

Chymase: A multifunctional player in pulmonary hypertension associated with lung fibrosis // Ralph Schermuly 0940-1000

Oestradiol metabolism and right ventricular function // Corey Ventetuolo 1000-1020

The role of hedgehog in lung repair and regeneration // Tien Peng 1020-1040

Discussion 1040-1050

Break 1050-1110

Plenary 4
Clinical Case Round Table
Chairs: Bradley Maron, Thenappan Thenappan
Presentation of six clinical cases by young clinicians followed by a round table discussion of case management by senior faculty.

Expert Presenters: Gaurav Choudhary, Brian Graham, Rebecca Kameny, Shelby Kutty, Bradley Maron, Thenappan Thenappan.


Introduction 1110-1120

Case 1: Group 1 PH congenital heart disease
Presenter: Shelby Kutty
Discussants: Jeff Fineman, Dunbar Ivy, Maria Jesus del Cerro 1120-1135

Case 2: Group 1 patient with vasodilator testing - management of responsive patient
Presenter: Bradley Maron
Discussants: Erika Berman-Rosenzweig, Anna Hemnes 1135-1150

Case 3: Group 2 PH diagnosis and treatment
Presenter: Thenappan Thenappan
Discussants: Erika Berman-Rosenzweig, Ronald Oudiz, Stuart Rich 1150-1205
Case 4: Group 3 PH - Sleep apnea
Presenter: Gaurav Choudhary
Discussants: John Granton, Nicholas Hill, Stephen Mathai

Case 5: Paediatric PH: BPD
Presenter: Rebecca Kameny
Discussants: Dunbar Ivy, Robin Steinhorn, Bernard Thébaud

Case 6: Group 4 PH
Presenter: Brian Graham
Discussants: Paul Corris, Steven Kawut, Ron Oudiz

LUNCH

END OF DAY TWO SCIENTIFIC SESSIONS
**Saturday 28th January 2017**

### Plenary 5
**Inflammation & immunity in PAH**
**Chairs:** Raed Dweik, Tim Lahm

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<tbody>
<tr>
<td>0800-0830</td>
<td><strong>KEYNOTE LECTURE:</strong> Macrophages and the regulation of inflammation // Luke O’Neill</td>
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<td>0830-0845</td>
<td><strong>Fibroblast - macrophage crosstalk in PH</strong> // Kurt Stenmark</td>
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<td>0845-0900</td>
<td><strong>Treating the patient with a BMPR2 mutation: Towards personalized medicine</strong> // Michaela Aldred</td>
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<td>0900-0915</td>
<td><strong>Low-Dose FK506 (tacrolimus) in end-stage pulmonary arterial hypertension</strong> // Edda Spiekerkoetter</td>
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<td>0915-0930</td>
<td>Discussion</td>
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### Plenary 6
**What’s new, what’s hot clinical?**
**Chairs:** John Granton, Roham Zamanian

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<tr>
<td>0930-0945</td>
<td><strong>Update on management of group 5 PH in sickle cell disease</strong> // Mark Gladwin</td>
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<td>0945-1000</td>
<td><strong>What’s new in CTEPH</strong> // Joanna Peke-Zaba</td>
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<tr>
<td>1000-1015</td>
<td><strong>New master protocol for clinical trials in pulmonary hypertension</strong> // Stuart Rich</td>
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<td>1015-1030</td>
<td><strong>Why do we get short of breath with exercise</strong> // David Systrom</td>
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<tr>
<td>1030-1045</td>
<td><strong>Pulmonary Circulation: How is our journal doing?</strong> // Jason Yuan</td>
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<td>1045-1100</td>
<td>Discussion</td>
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<tr>
<td>1100-1130</td>
<td>Break</td>
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| 1130-1230 | **Moderated poster session 3: Clinical**  
**Moderated poster session 4: Basic science**  
**Moderators:** Elena Goncharova, Brian Graham, Soni Pullamsetti, Usha Raj, Martin Wilkins  
Several senior moderators will ensure every poster is briefly presented by the author and discussed. |
| 1230-1300 | Discussion |
| 1300-1400 | Lunch |
Plenary 7
Functional and molecular imaging in PAH
Chairs: Ian Adatia, Kurt Prins

An introduction to imaging in PH // Robert Frantz 1400-1415
Exercise imaging of the RV in PH // Aaron Waxman 1415-1430
Imaging RV coupling and RV strain // Berend Westerhof 1430-1445
Imaging the human pulmonary circulation in PH // David Kiely 1445-1500
Is PET scanning feasible in human PAH? // Lan Zhao 1500-1515
Diagnosing & quantifying PH in populations // Steven Kawut 1515-1530
Discussion 1530-1600
Break 1600-1615

Plenary 8
Talks from 1 slide
Chairs: Antonio Lopes, Julio Sandoval

A unifying mechanism for the mitochondrial metabolic phenotype of PAH-dysregulation of the mitochondrial calcium uniporter // Stephen Archer 1615-1630
Are the endpoints used in PAH RCTs appropriate? // David Badesch 1630-1645
Exosomes in PAH // James Klinger 1645-1700
The role of iron and HIF in PAH? // Rubin Tuder 1700-1715
Discussion 1715-1745

END OF DAY THREE SCIENTIFIC SESSIONS

PVRI GALA DINNER & MEET THE MENTORS 1900 onwards
Sunday 29th January 2017

Plenary 9
Chair: Stuart Rich

PROFESSOR GHAZWAN BUTROUS LECTURE: 0800-0830
Putting big data in perspective // Charlie Hindmarch

Lessons from GWAS vs whole exome - whole genome studies in PAH 0830-0850 // Anna Hennes
National biological sample & data repository for PAH: Genotyping & biomarkers in the here and now // William Nichols 0850-0910
Discussion 0910-0940
Break 0940-1000

Plenary 10
New directions in the clinical management of PAH 0900-1020
Chairs: Todd Bull, Hap Faber

What’s in the PAH therapeutic pipelines? 1000-1020 // Ardeschir Ghofrani
Combination therapy for Group 1 PH: Respect for ambition? 1020-1040 // Jim White
Use of Selexipag in Group 1 PH 1040-1100 // Seane Gaine
Emerging metabolomics targets 1100-1120 // Greg Lewis
New devices for PH patients-RVADs, noninvasive measures of CO 1120-1140 // Jonathan Rich
New biomarkers 1140-1200 // Horst Olschewski
Discussion 1200-1230

END OF DAY FOUR SCIENTIFIC SESSIONS

Afternoon Travel

We LOOK FORWARD to meeting you all in Miami...