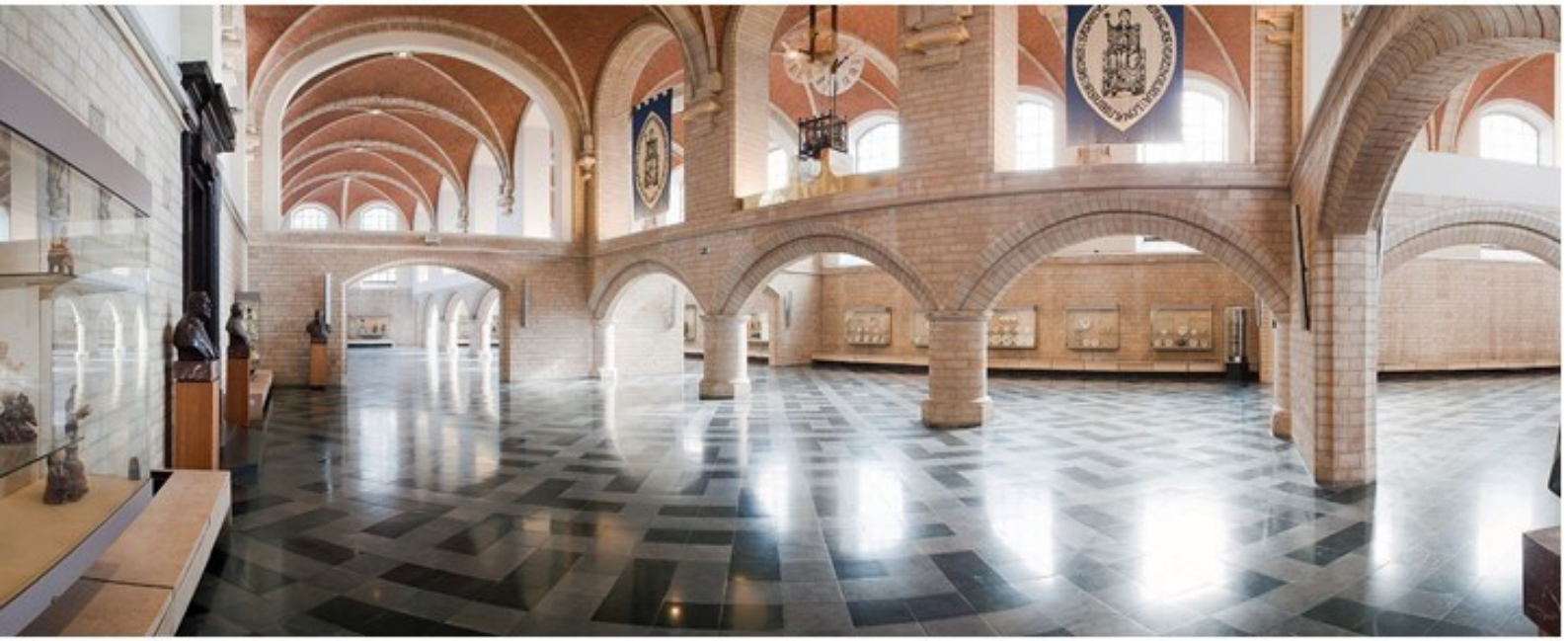
ICA International CTEPH Association

INTERNATIONAL **CTEPH** CONFERENCE **2017**



June 9-10, 2017

Aula Pieter De Somer, Leuven, Belgium

Conference program

www.cteph-association.org

ICC 2017 Committees

The International CTEPH Association would like to thank the members of the Scientific Committee and the members of the Abstract Review Committee for their contribution to the scientific program and abstract review process.

SCIENTIFIC COMMITTEE

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Welcome letter

Dear Colleagues and Friends,

On behalf of the International CTEPH Association, we would like to extend a warm welcome to all participants of the International CTEPH Conference (ICC) 2017. The International CTEPH Association is very pleased that the conference has attracted so much interest, making it the largest gathering of experts active in the field of CTEPH since the inaugural Cambridge conference in 2011.

Progress in surgical and medical treatment continues to improve the outcome for our patients with CTEPH. In addition, novel treatments, such as balloon angioplasty, have proven to be successful and continue to offer new perspectives. The conference program will present the latest and most significant developments in basic and clinical research, imaging and diagnosis, and, of course, medical and surgical treatment.

We trust that the conference will provide a vibrant and interactive opportunity for the presentation of the original research and exchange of the state-of-the-art knowledge regarding CTEPH, to raise awareness of CTEPH and to help scientists and clinicians translate the latest findings into improvements in patient care. It is our aim that the major strength of the meeting will be the opportunity for clinicians, surgeons, intensivists, radiologists and researchers to interact and exchange ideas.

On this occasion we would like to express our thanks to the pharmaceutical companies, who have provided funding for this event. Of note, the ICC sponsors did not have any influence on the selection of the faculty or the content of the scientific program.

Finally, we hope that you will enjoy this conference that takes place in the beautiful setting of one of the oldest universities of the world.

Yours sincerely,

Prof Gérald Simonneau

Prof Marion Delcroix

In compliance with UEMS-EACCME® guidelines, all speakers/chairpersons participating in this program have disclosed potential conflicts of interest that might cause a bias in the presentations.

Scientific programme

DAY ONE – Friday 9th June 2017

08:00 – 08:50 **Registration and welcome coffee**

08:50 – 09:00 **Welcome and introduction**
G. Simonneau and M. Delcroix

09:00 – 10:30 **Plenary session 1**
From acute pulmonary embolism to CTEPH
Chair: M. Madani and E. Klok

- Epidemiology of CTEPH and link with PE
  O. Sanchez
- Can we prevent CTEPH after acute PE?
  S. Konstantinides
- CTEPH pathology
  P. Dorfmüller
- Genetics of CTEPH
  N. Morell
- Plenary discussion

10:30 – 11:00 **Coffee break and poster viewing**

11:00 – 12:30 **Plenary session 2**
Diagnosis and imaging
Chair: M. Delcroix and M. de Perrot

- Diagnostic algorithm and differential diagnosis
  W. Auger
- What diagnostic modalities do we need for treatment decision?
  E. Mayer
- New global CTEPH registry – patient disposition
  J. Pepke-Zaba
- US CTEPH registry
  K. Kerr
- Plenary discussion

DAY ONE – Friday 9th June 2017

12:30 – 14:00 **Lunch break and poster viewing**

14:00 – 15:45 **Plenary session 3**

Pulmonary endarterectomy

E. Mayer and G. Simonneau

- Surgical treatment – state of the art
  D. Jenkins
- Case-based presentations
  - What is the distal limit of operability?
    M. Madani
  - What is the distal limit of operability?
    E. Fadel
  - Chronic thromboembolic disease without PH
    M. de Perrot
  - Case of unilateral occlusion
    E. Fadel
  - Short-term ECMO for post-operative endobronchial haemorrhage
    S. Guth

16:15 – 17:45 **Plenary session 4**

Medical therapy

I. Lang and B. Meyns

- Medical therapy
  X. Jaïs
- Bringing medical therapy to surgery or intervention
  M. Delcroix
- Persistent PH post-surgery
  J. Pepke-Zaba
- Plenary discussion

18:30 – 19:45 **Surprise Event**

20:00 – 22:30 **Dinner**

DAY TWO – Saturday 10th June 2017

07:45 – 08:15 **Registration**

08:15 – 10:15 **Plenary session 5**

Balloon pulmonary angioplasty

E. Fadel and H. Matsubara

- Real-life BPA techniques – images from the cath lab
  T. Ogo
- Challenging post-procedural complications
  I. Lang
- PEA/BPA: hybrid approaches
  C. Wiedenroth
- Rescue BPA
  P. Brenot
- Plenary discussion
- CTEPH management – present and future
  N. Kim

10:15 – 10:45 **Coffee break and poster viewing**

10:45 – 11:45 **Plenary session 6**

Future CTEPH landscape – hot topics

M. Delcroix and D. Jenkins

- The RACE study
  G. Simonneau
- Biomarkers in CTEPH
  M. Toshner
- Exercise MRI in CTEPH
  G. Claessen
- Screening for CTEPH
  E. Klok

DAY TWO – Saturday 10th June 2017

11:45 – 13:15 **Abstract session**
Oral presentation of the best five abstracts
J. Pepke-Zaba and A. Torbicki

13:15 – 13:30 **Closing remarks**
Abstract award ceremony
G. Simonneau

13:30 – 14:45 **Lunch and individual departure**

Accreditation

The UEMS-EACCME ® has granted 9 credit points to the International CTEPH Conference 2017.

Biographies of the Scientific and Abstract Committee Members

Professor Marion Delcroix

Marion Delcroix was born in Belgium and studied medicine at the Free University of Brussels, where she graduated in 1987. She was research assistant and senior research assistant of the Belgian National Fund for Scientific Research (FNRS) for 5 years. She then specialised in pneumology. She is presently working in the Department of Pneumology of the University Hospitals of Leuven and is professor of Medicine and of Respiratory Physiology at the Catholic Universities of Leuven and Kortrijk. As director of the Centre for Pulmonary Vascular Diseases in Leuven, she has been involved in the routine care of over 1000 patients with pulmonary hypertension and has participated to the main pivotal trials for the treatment of pulmonary arterial hypertension (PAH). She was invited expert at the Third, Fourth and Fifth World Symposia on Pulmonary Arterial Hypertension, is vice-president of the Belgian Task Force for Pulmonary Hypertension, and founding member of the International CTEPH Association. She has over 100 publications, with research interest focusing on pulmonary circulation and gas exchange, cardiac imaging, and on the role of inflammation in the pathogenesis of PAH and CTEPH.

Professor Elie Fadel

Elie Fadel is the head of the Department of Thoracic and Vascular Surgery and Heart-Lung Transplantation at the Marie Lannelongue Hospital and Professor of Thoracic and Cardio-Vascular Surgery at the Paris-Sud University. He also leads the Laboratory of Surgical Research, INSERM E999 at Marie Lannelongue Hospital and is mainly involved in basic science research on pulmonary vascular disease and right heart failure. Prof. Fadel's surgical expertise focuses on surgery for local extended (T4) lung cancer, mediastinal tumours, tumours involving vessels or spine, aortic surgery and surgery for pulmonary hypertension, such as lung and heart-lung transplantation, as well as pulmonary endarterectomy for CTEPH. He is a member of professional societies, including the French Society of Cardio-Thoracic Surgery, the International Society of Heart-Lung Transplantation, the European Association for Cardio-Thoracic Surgery and the American Association for Thoracic Surgery, and serves as board member of the International CTEPH Association.

Doctor David Jenkins

David Jenkins is a consultant cardiothoracic surgeon and clinical director at Papworth Hospital, Cambridge, UK. He graduated from medical school at the University of London in 1989, with a first class intercalated BSc in physiology, and trained in surgery in London. Following a 2-year formal research period investigating ischaemic preconditioning and myocardial protection at the Hatter Institute at the University College London, Dr. Jenkins was awarded Master of Surgery from the London University in 1997. He undertook his cardiothoracic surgical training in West London at the Harefield, Hammersmith and St. George's hospitals, and was appointed as Consultant at Papworth Hospital in 2001. Dr. Jenkins has experience in all aspects of adult cardiac surgery. His specialist experience includes surgical treatment of pulmonary hypertension, intra-thoracic transplantation, and mechanical circulatory support including extracorporeal membrane oxygenation. In 2004, he became lead surgeon for the UK national pulmonary endarterectomy program. Dr. Jenkins is an executive member and audit lead of the Society for Cardiothoracic Surgery in Great Britain and board member of the International CTEPH Association. His research interests are related to myocardial protection, pulmonary hypertension and pulmonary endarterectomy.

Professor Nick H. Kim

Nick H. Kim is clinical professor of Medicine in the Division of Pulmonary and Critical Care Medicine at the University of California, San Diego (UCSD). He is director of both Pulmonary Vascular Medicine and of the PCCM Fellowship Training Program at UCSD. Prof. Kim graduated from Harvard University, Cambridge, Massachusetts, and received his medical degree in 1994 from the University of Chicago Pritzker School of Medicine, Chicago, Illinois. He completed his internal medicine internship and residency at the University of Chicago and a year of anaesthesiology training at the University of California, San Francisco. After completing his fellowship training in pulmonary and critical care medicine at UCSD in 2001, he joined the faculty specialising in pulmonary vascular medicine. Prof. Kim's clinical and research interests are in pulmonary hypertension, CTEPH and balloon pulmonary angioplasty. He is a board member of the International CTEPH Association and an investigator for multiple clinical trials in pulmonary hypertension. He has published in numerous peer-reviewed journals and has lectured internationally on the subject of pulmonary hypertension.

Professor Irene Lang

Irene Marthe Lang is deputy chair of the Department of Cardiology at the Medical University of Vienna (MUV), Vienna, Austria. Prof. Lang carried out her medical education and residency at the University of Vienna, before taking on a 5-year post-doctoral research fellowship at the University of California, San Diego. She has been a professor of Vascular Biology at the MUV since 2004, where she leads a clinical and experimental group in vascular medicine focusing on pulmonary vascular biology and right ventricular function. Furthermore, Prof. Lang is a founding member of the International CTEPH Association and an active interventional and structural cardiologist.

Professor Michael M. Madani

Michael Madani is professor of Surgery and Chief of Cardiovascular and Thoracic Surgery at the University of California, San Diego (UCSD). Prof. Madani is also director of UCSD Sulpizio Cardiovascular Center – Surgery and director of the UCSD Pulmonary Thromboendarterectomy program. Prof. Madani received his medical degree from the University of Toronto and moved to San Diego in 1999, after completing his surgical residency in Berkshire Medical Center, University of Massachussetts Medical School. Prof. Madani's clinical interests include pulmonary thromboendartectomy (PTE), mitral valve repair, minimally invasive and robotic cardiac surgery, as well as surgical treatment for heart failure, and heart and lung transplantation. He is currently a member of various national and international committees and is actively involved in multiple areas of clinical research. He also serves as secretary-general for the World Society of CardioThoracic Surgeons and as board member to the International CTEPH Association. Prof. Madani has over 100 publications and has contributed to many distinguished textbooks in cardiothoracic surgery. Well-known in the international arena for his expertise in PTE, Prof. Madani is a gifted speaker and has given more than 160 presentations in the national and international arena during his distinguished career.

Professor Hiromi Matsubara

Hiromi Matsubara is currently director of the Department of Clinical Science and Cardiology at the National Hospital Organisation Okayama Medical Center, Okayama, Japan. After receiving his medical degree from Okayama University Medical School and finishing his residency in internal medicine at National Okayama Hospital, he began his fellowship at the National Cardiovascular Center in Osaka, Japan in 1990. He became an assistant professor of the Department of Physiology II at Okayama University Graduate School of Medicine and Dentistry in 1993. He then became an assistant professor at the Department of Cardiovascular Medicine in 1997 and again promoted in 2000 to associate professor of Cardiovascular Medicine. He then became clinical professor of Department of Cardiovascular Medicine and the Director of Department of Cardiology at National Hospital Organization Okayama Medical Center. He also serves as director of the Department of Clinical Science since 2010. His investigative interests have focused on clinical and physiologic aspects of pulmonary hypertension. He started performing balloon pulmonary angioplasties (BPA) for CTEPH in 2004 and has conducted approximately 1800 BPAs in more than 300 patients during the last 12 years. Under his guidance, the Department of Cardiology at National Hospital Organization Okayama Medical Center became the largest BPA centre in Japan.

Professor Eckhard Mayer

Eckhard Mayer is professor of Thoracic Surgery and director of the Department of Thoracic Surgery at Kerckhoff Heart and Lung Center, Bad Nauheim, Germany. Since 1989 he has been involved in the first successful European Pulmonary Endarterectomy Program at Mainz University hospital and has been the director of this program at Mainz and Giessen universities and the Kerckhoff Heart and Lung Center for more than 20 years. He has published widely in the field of CTEPH and has been actively involved in the Venice 2003, Dana Point 2008 and Nice 2013 Pulmonary Hypertension (PH) World Symposia as well as in several European and national PH guideline conferences. Prof. Mayer is a founding member of the International CTEPH Association.

Professor Joanna Pepke-Zaba

Joanna Pepke-Zaba graduated from Warsaw University School of Medicine in Poland before undertaking a fellowship in respiratory physiology at Papworth and Addenbrooke's Hospitals, University of Cambridge, which resulted in a PhD. She is the lead physician and director of the National Pulmonary Vascular Diseases Unit at Papworth Hospital, Cambridge, UK. Her research has concentrated on the translational programmes in the field of pulmonary hypertension (PH), with specific interest into CTEPH and idiopathic pulmonary arterial hypertension. She has published over 120 papers in the field of PH and serves on various educational scientific international committees and is an active member/fellow of the European Respiratory Society, European Society of Cardiology and American Thoracic Society. She is committed to training in respiratory medicine and was reappointed as Recognised Teacher by the Clinical School of the University of Cambridge. She is a founding member of the International CTEPH Association and has been Honorary Senior Visiting Fellow of the University of Cambridge, School Of Clinical Medicine since 2011.

Professor Gérald Simonneau

Gérald Simonneau is head of the Department of Pneumology and Intensive Care Medicine at Hôpital Kremlin Bicêtre, Paris-Sud University, France. In addition, Prof. Simonneau has served as director of the French National Reference Centre for Pulmonary Hypertension since 2004 and director of the Research Unit "New Therapeutic Approaches for Pulmonary Hypertension" INSERM U999, since 2010. He has published widely in the fields of pulmonary hypertension, pulmonary vascular diseases and pneumology in peer-reviewed journals, including *New England Journal of Medicine*, *Lancet*, *Annals of Internal Medicine*, and *Circulation*. He is founding member and chair of the International CTEPH Association and has been president of the working group on pulmonary circulation of the European Society of Cardiology. Prof. Simonneau has received the pulmonary arterial hypertension research award of the European Respiratory Society in 2011. Lastly, he was named among "World's Most Influential Scientific Minds" in the field of Clinical Medicine (Thomson Reuters) in 2014 and 2015.

Professor Adam Torbicki

Adam Torbicki is head of the Department of Pulmonary Circulation and Thromboembolic Diseases of the Centre of Postgraduate Medical Education based at the Cardioangiology Department of the European Health Centre in Otwock, Poland. Prof. Torbicki graduated from Warsaw University School of Medicine in 1978, and held fellowships at the Universities of Freiburg (Germany) and Pavia (Italy). He served as head of the Department of Chest Medicine at the Institute of Tuberculosis and Lung Diseases in Warsaw, Poland from 1998 to 2011, a reference centre for pulmonary hypertension and embolisms. He specialised in internal medicine, cardiology and angiology. Since 1982, Prof. Torbicki has been involved in research related to pulmonary circulation with special interest in non-invasive evaluation, prognostic staging and follow-up of pulmonary hypertension and right ventricular function. He contributed as principal investigator and/or member of steering committee to many trials assessing pharmacologic treatments in pulmonary hypertension. He is the author of more than 250 scientific publications (HI = 50) and co-authored the European Society of Cardiology Guidelines on Pulmonary Hypertension published in 2006, 2009 and 2015. Furthermore, Prof. Torbicki is chairman of the task force that published the European Society of Cardiology Guidelines on Diagnosis and Treatment of Pulmonary Embolism in 2000, 2008 and 2014. He was chairman of the Working Group on Pulmonary Circulation and Right Ventricular Function of the European Society of Cardiology (2004 – 2006), president of the Polish Cardiac Society (2004 – 2007) and vice-president of the European Society of Cardiology (2010 – 2012).

Abstracts selected for oral presentation

EFFICACY AND SAFETY OF MACITENTAN FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM THE RANDOMIZED CONTROLLED MERIT STUDY

N.H. Kim, G. Simonneau, A.M. D'Armini, P. Fedullo, L. Howard, X. Jaïs, D.P. Jenkins, Z.C. Jing, M. Madani, N. Martin, E. Mayer, K. Papadakis, D. Richard, H.A. Ghofrani

Objective

The randomized, double-blind MERIT study (NCT02021292) evaluated the endothelin receptor antagonist macitentan in primary inoperable CTEPH.

Methods

Patients in WHO functional class (FC) II–IV with a pulmonary vascular resistance (PVR) ≥400 dyn·sec/cm^5 and 6-minute walk distance (6MWD) of 150-450 m were randomized 1:1 to placebo or macitentan 10 mg once daily and treated for 24 weeks. Inoperability/eligibility was assessed by an independent adjudication committee. Phosphodiesterase type-5 inhibitors (PDE-5i) or oral/inhaled prostanoids were allowed for patients in WHO FC III–IV. The primary endpoint was PVR at Week 16, expressed as percent of baseline. The main secondary endpoint was mean change from baseline to Week 24 in 6MWD. Exploratory endpoints included mean change from baseline to Week 16 in cardiac index (CI) and mean right atrial pressure (mRAP), and percent of baseline N-terminal pro B-type natriuretic peptide (NT-proBNP) at Week 24.

Results

80 patients were randomized to macitentan (n=40) or placebo (n=40). At baseline, 61% were receiving PAH therapy; 96% PDE-5i. Compared with placebo, PVR and 6MWD significantly improved in the macitentan-treated group (Table). The treatment effect on PVR and 6MWD was similar regardless of baseline PAH treatment. Exploratory endpoints also improved in the macitentan-treated group (Table). Most common adverse events (macitentan versus placebo) were peripheral edema (22.5% vs. 10.0%) and decreased hemoglobin/anemia (17.5% vs. 2.5%). Five patients (placebo) prematurely discontinued treatment. Two patients (placebo) died.

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EFFICACY AND SAFETY OF MACITENTAN FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM THE RANDOMIZED CONTROLLED MERIT STUDY

Conclusion

In the MERIT study, macitentan led to significant improvements in cardiopulmonary hemodynamics and exercise capacity in inoperable CTEPH patients and was well tolerated.

Table

Variable	Baseline (N=40) Mean (SD)		Post-baseline* (N=40) Mean (SD)		% of baseline Geometric mean (95% CL)		Treatment effect (Ratio)** (95% CL) p value
	Placebo	Macitentan	Placebo	Macitentan	Placebo	Macitentan	
PVR, dyn·sec/cm^5 (primary endpoint)	984 (487)	929 (380)	899 (476)	723 (454)	87.2 (78.5, 96.7)	73.0 (63.6, 83.8)	0.84 (0.70, 0.99) p=0.04
NT-proBNP, pg/mL	1793 (2075)	2204 (2943)	1433 (1450)	1553 (2252)	90.9 (76.5, 107.9)	72.6 (61.2, 86.2)	0.79 (0.63, 0.99) p=0.04
	Baseline (N=40) Mean (SD)		**Post-baseline* (N=40) Mean (SD)**		**Change from baseline Mean (SD)**		**Treatment effect (Difference)** (95% CL) p value**
	Placebo	**Macitentan**	**Placebo**	**Macitentan**	**Placebo**	**Macitentan**	
6MWD, m (secondary endpoint)	351 (74)	353 (88)	352 (121)	388 (83)	1.0 (83.2)	35.0 (52.5)	34.0 (2.9, 65.2) p=0.03
mRAP, mmHg	8.7 (5.2)	10.4 (8.1)	8.6 (5.3)	7.8 (5.9)	−0.1 (3.5)	−2.6 (8.4)	−1.47 (−3.77, 0.84) p=0.21
CI, L/min/m^2	2.2 (0.6)	2.2 (0.5)	2.2 (0.5)	2.6 (0.8)	0.0 (0.4)	0.4 (0.7)	0.43 (0.18, 0.67) p<0.001

*Post-baseline is Week 16 for PVR, mRAP and CI, and Week 24 for 6MWD and NT-proBNP
**From ANCOVA model adjusted by variable at baseline as covariate

RIOCIGUAT FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): 2-YEAR RESULTS FROM THE CHEST-2 LONG-TERM EXTENSION

G. Simonneau, A.M. D'Armini, H.A. Ghofrani, F. Grimminger, M.M. Hoeper, P. Jansa, N.H. Kim, M. Wilkins, A. Fritsch, N. Davie, G. Xing, E. Mayer

Objective

Riociguat significantly improved 6-minute walking distance (6MWD) and other efficacy endpoints in patients with inoperable or persistent/recurrent CTEPH in the 16-week CHEST-1 trial. We present 2-year data from the CHEST-2 long-term extension.

Methods

The primary endpoints were safety and tolerability; secondary endpoints included 6MWD, WHO functional class (FC), *N*-terminal prohormone of brain natriuretic peptide (NT-proBNP), survival, and clinical worsening-free survival. Kaplan–Meier analyses and a Cox proportional-hazards regression model were used to assess correlation between efficacy parameters and long-term outcomes.

Results

Of 243 patients who completed CHEST-1, 237 (98%) entered CHEST-2. At this cut-off (March 2014) most patients had been treated for ≥2 years. Riociguat was well tolerated and no new safety signals were identified. Serious adverse events occurred in 129 (54%) patients (8% considered drug-related) and 14 (6%) patients discontinued therapy because of adverse events. Mean±SD 6MWD improved from CHEST-1 baseline by 50±68 m (n=162) and WHO FC improved/stabilized/worsened in 39/58/3% of patients (n=170). 2-year survival and clinical worsening-free survival rates were 93% and 82%, respectively. 6MWD and NT-proBNP at CHEST-1 baseline and Week 16 correlated significantly with long-term survival and clinical worsening-free survival, and WHO FC correlated with clinical worsening-free survival.

Conclusion

Riociguat has a good long-term safety profile and is the first pharmacologic therapy for CTEPH to show sustained clinical effect. The correlation of 6MWD and NT-proBNP with long-term survival emphasizes the prognostic value of these endpoints in patients with CTEPH.

MULTICENTER, OBSERVATIONAL SCREENING SURVEY FOR THE DETECTION OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION FOLLOWING PULMONARY EMBOLISM

N. Coquoz, D. Weilenmann, D. Stolz, V. Popov, A. Azzola, J-M. Fellrath, H. Stricker, A. Pagnamenta, S. Ott, S. Ulrich, S. Györik, J.-D. Aubert

Objectives

Primary objective

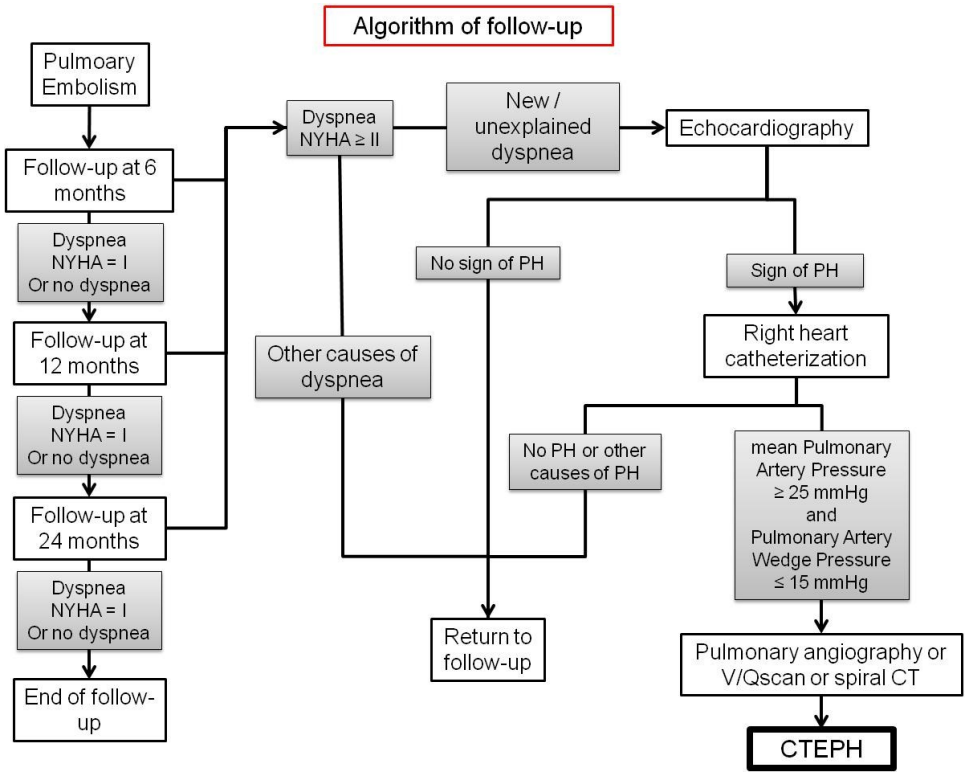
Evaluate the incidence rate of chronic thromboembolic pulmonary hypertension (CTEPH) following pulmonary embolism (PE)

Secondary objective

Test the usefulness of a dyspnea based screening algorithm for diagnosing CTEPH after PE

Methods

In a prospective, multicenter, observational phase V study, conducted between 2008 and 2015 we screened 1699 patients with PE in 11 Swiss pulmonary hypertension (PH) centers. 602 confirmed acute PE patients were included and followed over two years.



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MULTICENTER, OBSERVATIONAL SCREENING SURVEY FOR THE DETECTION OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION FOLLOWING PULMONARY EMBOLISM

Results

Our main finding is a cumulative CTEPH incidence of 0.79% over two years. We identified 4 cases on 508 patients who completed the follow-up. The CTEPH incidence rate was 0.37% persons/year after PE in Switzerland. A match between the Swiss PH registry and our study database of all the 1699 screened patients strengthened our results. According to the registry, we missed no CTEPH case during our follow-up. Four additional cases occurred in the 1193 patients excluded or incompletely screened. Therefore, our protocol didn't segregate a higher risk population. The dyspnea based screening algorithm yielded a sensitivity of 100% and a specificity of 90.5%. Echocardiography in newly dyspneic patients showed a negative predictive value of 100%.

Conclusion

To our knowledge, this multicenter prospective study on CTEPH is one of the largest cohorts conducted in PE patients. The screening algorithm was able to detect all CTEPH cases occurring in the 2 years following PE. The low 0.79% incidence, when compared to the ~25 new diagnosed cases/year in Switzerland, raises the importance of silent embolic events in the CTEPH pathogenesis.

PREOPERATIVE MONOTHERAPY WITH PHOSPHODIESTERASE-5 INHIBITOR, ENDOTHELIN RECEPTOR ANTAGONIST, OR RIOCIGUAT DOES NOT AFFECT OUTCOMES AFTER PULMONARY THROMBOENDARTERECTOMY

T.M. Fernandes, B. Hsu, D.G. Papamatheakis, D.S. Poch, P.F. Fedullo, N.H. Kim, K.M. Kerr, V.G. Pretorius, M.M. Madani, W.R. Auger

Objective

Use of pulmonary arterial hypertension (PAH)-targeted oral medical therapy prior to pulmonary thromboendarterectomy (PTE) is common among patients referred to our center. Whether the use of various classes of these medications affects outcomes from this procedure has not been adequately investigated.

Methods

In order to assess class effects on PTE outcomes, we reviewed consecutive patients referred to UCSD for PTE and compared patients on no PAH-targeted medical therapy to those treated with monotherapy with either phosphodiesterase type 5-inhibitors (PDE5i), endothelin receptor antagonists (ERA), or soluble guanylate cyclase stimulator (sGCS). Patients on combination therapy were excluded. A multivariate logistic regression was completed to examine the effects of monotherapy with one of the three classes on outcomes from PTE surgery (including mortality, residual pulmonary hypertension with PVT >400 dyn.sec.cm-5, and reperfusion lung injury).

Results

Of the 521 patients operated on between 2013 and 2015, 48.0% were on pre-operative PAH-targeted medical therapy. After excluding those on combination therapy, there were 80 patients on PDE5i, 68 patients on sGCS, and 16 on ERA. In a multivariate logistic regression model which adjusted for age, sex, BMI, right atrial pressure, PVR, cardiac index, history of VTE, and disease level, there was no significant increase in the odds a patient pre-treated with any of the three classes of PAH-targeted oral medications on post-operative mortality, residual pulmonary residual pulmonary hypertension, reperfusion lung injury or pulmonary hemorrhage (Table 1).

Conclusions

Preoperative monotherapy with PDE5i, sGCS, or ERA does not significantly increase the incidence of any important morbidity or mortality after PTE surgery.

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PREOPERATIVE MONOTHERAPY WITH PHOSPHODIESTERASE-5 INHIBITOR, ENDOTHELIN RECEPTOR ANTAGONIST, OR RIOCIGUAT DOES NOT AFFECT OUTCOMES AFTER PULMONARY THROMBOENDARTERECTOMY

Table 1. Multivariate Logistic Regression for Single Treatment Effect on Various Post PTE Outcomes

	Number of Patients	Mortality OR (95% CI)	Residual PH OR (95% CI)	Reperfusion Lung Injury OR (95% CI)	Pulmonary Hemorrhage OR (95% CI)
No Drug	271	Reference	Reference	Reference	Reference
PDE5i	80	1.552 (0.202-8.187)	2.332 (0.862-6.221)	1.869 (0.954-3.601)	1.773 (0.651-4.550)
sGCs	68	NA	1.845 (0.619-5.147)	1.598 (0.763-3.223)	1.029 (0.277-3.089)
ERA	16	5.414 (0.233-52.172)	2.377 (0.320-11.593)	1.845 (0.464-6.108)	NA

PTE = pulmonary thomboendarterectomy; PH = pulmonary hypertension; RPE = reperfusion edema; PDE5i = phosphodiesterase type 5 inhibitor; sGCs = soluble guanylate cyclase stimulator; ERA = endothelin receptor antagonist (Of note, there were no deaths among patients on sGCs alone and no episodes of pulmonary hemorrhage among patietns on ERA alone so OR cannot be calculated for either of these groups.)

BALLOON PULMONARY ANGIOPLASTY FOR PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: A JAPANESE MULTICENTER REGISTRY

A. Ogawa, H. Ito, H. Matsubara on behalf of the Japanese Circulation Society Joint Working Group for the "Statement for balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (JCS 2014)"

Objective

Balloon pulmonary angioplasty (BPA) is recently reported to be effective for selected patients with chronic thromboembolic pulmonary hypertension (CTEPH). However, all reports are results from a relatively small cohort treated at a single center. We conducted a retrospective multicenter registry to evaluate safety and efficacy of BPA.

Methods

A total of 308 patients who underwent BPA between November 2004 and March 2013 were registered at 7 institutions in Japan. Data were retrospectively analyzed to evaluate clinical outcome and complications. Survival from the initial BPA session was evaluated.

Results

Hemodynamic parameters were significantly improved in 249 patients in whom all sessions were completed. Mean pulmonary arterial pressure decreased by 19 mmHg after final BPA. In 196 patients who underwent right heart catheterization at follow-up, improvement of hemodynamic parameters was maintained. Complications occurred in 36.6% of a total of 1408 sessions. Most of them were related to procedure: reperfusion pulmonary injury (17.8%), hemoptysis (14.0%), and pulmonary artery perforation (2.9%). Death occurred in 12 patients during follow-up. The leading causes of death were right heart failure, multi-organ failure, and sepsis. Overall survival was 99.6% at 1 year and 98.4% at 3 years after the initial BPA session.

Conclusions

The first multicenter registry in Japan revealed that BPA performed at experienced pulmonary hypertension centers is a safe and effective therapeutic option in patients with CTEPH. Further investigation is needed to confirm long-term safety and efficacy of BPA.

POOR SUBPLEURAL PERFUSION AS A PREDICTOR OF FAILURE AFTER BALLOON PULMONARY ANGIOPLASTY FOR NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Y. Taniguchi, P. Brenot, X. Jaïs, C. Garcia, O. Planche, E. Fadel, M. Humbert, G. Simonneau

Background

It has been shown that 'poor subpleural perfusion' (PSP) in the capillary phase of pulmonary angiography (PA), which is related to microvasculopathy in chronic thromboembolic pulmonary hypertension (CTEPH), predicts poor outcome after pulmonary endarterectomy (Tanabe, Chest 2012). We assessed the association between PSP and failure of Balloon Pulmonary Angioplasty (BPA) in 101 non-operable CTEPH.

Method

Subpleural perfusion in the capillary phase of PA was classified as normal or poorly perfused as previously described. Patients were divided according to hemodynamic results after the last BPA: a failure group (defined as mean PAP >30mmHg and PVR decrease <20% after BPA [n=11]) or a success group [n=90].

Results

Baseline characteristics and hemodynamics were similar in the two groups (table). In contrast, PSP was observed in 45.5% of patients in the failure group versus 15.6% in the success group (p=0.016). Multivariate analysis revealed that PSP was the only predictor of BPA failure (Odds Ratio 4.524, 95% confidence interval 1.213-16.877, p=0.025).

Conclusion

Poor subpleural perfusion in the capillary phase of pulmonary angiography, suggesting the presence of diffuse microvasculopathy, is associated with significant residual pulmonary hypertension after BPA. In our experience, it affected approximately 10% of non-operable CTEPH who underwent BPA.

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POOR SUBPLEURAL PERFUSION AS A PREDICTOR OF FAILURE AFTER BALLOON PULMONARY ANGIOPLASTY FOR NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Variable	Success group n=90	Failure group n=11	p value
Baseline			
Age (years)	63.5 ± 13.8	62.7 ± 14.5	0.867
Male (n, %)	47 (52.2)	7 (63.6)	0.474
NYHA Fc (I,II / III,IV) (%)	26.5 / 73.5	27.3 / 72.7	0.369
6 minutes walk distance (m)	401 ± 118	380 ± 92	0.627
mean PAP (mmHg)	45.6 ± 10.2	45.6 ± 11.4	0.988
Cardiac Index (L/min/m^2)	2.51 ± 0.48	2.66 ± 0.80	0.393
PVR (wood unit)	8.39 ± 3.25	7.58 ± 2.28	0.422
Poor subpleural perfusion	14 (15.6%)	5 (45.5%)	0.016
After last BPA session			
NYHA Fc (I,II / III,IV) (%)	91.7 / 8.3	44.4 / 55.6	0.002
6 minutes walk distance (m)	442 ± 121	363 ± 106	0.066
mean PAP (mmHg)	29.6 ± 7.5	42.8 ± 8.7	<0.0001
Cardiac Index (L/min/m^2)	3.04 ± 0.70	2.59 ± 0.62	0.047
PVR (wood unit)	3.64 ± 1.61	6.77 ± 2.39	<0.0001
% decrease of PVR (%)	-53.3 ± 19.1	-10.8 ± 9.6	<0.0001
% decrease of mean PAP (%)	-33.2 ± 17.4	-4.5 ± 9.4	<0.0001
Number of patients (mean PAP ≤30mmHg)	53	0	

POSTOPERATIVE LEFT VENTRICULAR FUNCTION IN DIFFERENT TYPES OF PULMONARY HYPERTENSION: A COMPARATIVE STUDY

T. Verbelen, A. Van De Bruaene, B. Cools, D. Van Raemdonck, M. Delcroix, F. Rega, B. Meyns

Objectives

Temporary left ventricular (LV) dysfunction after pulmonary endarterectomy (PEA) for chronic thromboembolic pulmonary hypertension (CTPEH) is well described. True LV-failure has only been described after bilateral lung transplantation (bLTx) in pulmonary arterial hypertension (PAH) patients. We sought to identify factors that contribute to this LV-failure and hypothesized that an atrial septostomy before bLTx could prevent this LV-failure.

Methods

From our database, all PAH patients that underwent bLTx (n=24) and all CTPEH patients that underwent PEA, with a minimal reduction of 800 dynes.s.cm^{-5} (n=27), were selected. Perioperative demographic and echocardiographic data were analyzed.

Results

Results are depicted in tables 1-3. Pulmonary hypertension was diagnosed at significant younger age and time between diagnosis and surgery was significantly longer in PAH patients. PAH patients had significant larger right ventricular (RV) dimensions, but a similar preoperative LV diastolic dysfunction. Surgery caused significant decreases in RV dimensions, to a lesser extent in CTEPH patients, and significant increases in LV dimensions. Atrial septostomy for PAH showed a tendency to increase LV dimensions and cardiac index. Two PAH patients developed postoperative LV-failure. Their mean age at diagnosis, time between diagnosis and surgery, E/A ratio and LV-mass before bLTx were 10.9 years, 4776 days, 2.3, and 68 grams, respectively.

Conclusions

In PAH, diastolic dysfunction before surgery is comparable to CTEPH. However, age at diagnosis is younger and LV preload deprivation lasts longer. This might explain the occasional development of systolic LV-failure after bLTx. Preoperative atrial septostomy might train the LV, by increasing its preload, and avoid postoperative LV-failure.

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POSTOPERATIVE LEFT VENTRICULAR FUNCTION IN DIFFERENT TYPES OF PULMONARY HYPERTENSION: A COMPARATIVE STUDY

Table 1 Demographic data.

	CTEPH - PEA (27)	PAH - bLTx (24)	p
Female/Male	9/18	11/13	0.4018
Age at diagnosis (years)	56.8 ± 14.3	37.4 ± 14.7	$< 1.10^{-4}$
Age at surgery (years)	57.9 ± 14.3	43.0 ± 13.4	$< 1.10^{-3}$
Time between diagnosis and surgery (days)	387 ± 440	2068 ± 1662	$< 1.10^{-5}$

CTEPH = chronic thromboembolic pulmonary hypertension, PEA = pulmonary endarterectomy, PAH = pulmonary arterial hypertension, bLTx = bilateral lung transplantation

Table 2 Pre- and postsurgical echocardiographic data.

	CTEPH - PEA (27)		PAH - bLTx (24)	
	pre	post	pre	post
LVEDD (mm)	37 ± 6	43 ± 6*	37 ± 7	43 ± 6*
LVESD (mm)	23 ± 5	29 ± 6*	22 ± 6	30 ± 6*
LV diastolic area (cm²)	20 ± 7	24 ± 7*	19 ± 5	24 ± 5*
IVS thickness (mm)	11 ± 2	11 ± 2	10 ± 2	11 ± 2
LVPW thickness (mm)	10 ± 2	11 ± 2*	9 ± 2	10 ± 3*
LV mass (g)	120 ± 42	168 ± 56*	104 ± 39	159 ± 53*
LVEF (%)	64 ± 11	65 ± 10	71 ± 11	65 ± 11*
E velocity (m/s)	0.45 ± 0.14	0.64 ± 0.19*	0.67 ± 0.21	0.75 ± 0.23
A velocity (m/s)	0.63 ± 0.18	0.66 ± 0.19	0.72 ± 0.16	0.58 ± 0.18*
E/A	0.80 ± 0.41	1.07 ± 0.53*	0.92 ± 0.31	1.44 ± 0.87
SV (ml)	50 ± 15	65 ± 19*	61 ± 15	65 ± 25
CI (L/min/m²)	2.37 ± 0.79	3.00 ± 0.87*	3.08 ± 0.84	2.75 ± 0.93
RV diastolic area (cm²)	31 ± 5	25 ± 5*	37 ± 10	18 ± 5*

LVEDD = left ventricular end diastolic diameter; LVESD = left ventricular end systolic diameter; LV = left ventricle; IVS = interventricular septum; LVPW = left ventricular posterior wall; LVEF = left ventricular ejection fraction; E = mitral peak early diastolic velocity; A = mitral peak late diastolic velocity; SV = stroke volume; CI = cardiac index; RV = right ventricle; * = significant difference with preoperative value; _ = significant difference with corresponding echo in PEA group.

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POSTOPERATIVE LEFT VENTRICULAR FUNCTION IN DIFFERENT TYPES OF PULMONARY HYPERTENSION: A COMPARATIVE STUDY

Table 3 Echocardiographic data comparing PAH without and with AS.

	PAH-bLTx without AS (16)		PAH-bLTx with AS (6)		
	pre-bLTx	post-bLTx	pre-AS	pre-bLTx	post-bLTx
LVEDD (mm)	36 ± 5	43 ± 5[1]	32 ± 8	39 ± 5[3]	43 ± 5[3]
LVESD (mm)	21 ± 4	28 ± 5[1]	19 ± 5	24 ± 8	28 ± 4[3]
LV diastolic area (cm²)	21 ± 4	24 ± 6	14 ± 2[2]	19 ± 6	24 ± 6[3]
IVS thickness (mm)	10 ± 2	10 ± 2	12 ± 3	10 ± 2	12 ± 2[1]
LVPW thickness (mm)	9 ± 2	11 ± 3	9 ± 1	9 ± 2	10 ± 1
LV mass (g)	101 ± 30	156 ± 51[1]	144 ± 84	127 ± 52	181 ± 54
LVEF (%)	71 ± 12	67 ± 8	66 ± 4	71 ± 10	66 ± 5
E velocity (m/s)	0.64 ± 0.22	0.69 ± 0.19	0.54 ± 0.31	0.74 ± 0.23	0.84 ± 0.23
A velocity (m/s)	0.68 ± 0.17	0.58 ± 0.13	0.74 ± 0.21	0.85 ± 0.22	0.63 ± 0.36
E/A	0.95 ± 0.35	1.15 ± 0.35	0.73 ± 0.12	0.87 ± 0.31	1.34 ± 0.49[3]
SV (ml)	65 ± 18	66 ± 31	43 ± 8[2]	57 ± 9[3]	63 ± 17[3]
CI (L/min/m²)	3.25 ± 0.99	2.99 ± 1.10	1.98 ± 0.30[2]	3.07 ± 0.48[3]	2.51 ± 0.43[3]
RV diastolic area (cm²)	37 ± 10	18 ± 5[1]	41 ± 11	37 ± 8	18 ± 4[1,3]

PAH = pulmonary arterial hypertension; bLTx = bilateral lung transplantation; AS = atrial septostomy; LVEDD = left ventricular end diastolic diameter; LVESD = left ventricular end systolic diameter; LV = left ventricle; IVS = interventricular septum; LVPW = left ventricular posterior wall; LVEF = left ventricular ejection fraction; E = mitral peak early diastolic velocity; A = mitral peak late diastolic velocity; SV = stroke volume; CI = cardiac index; RV = right ventricle; [1] = significant difference with preoperative value; [2] = significant difference with pre-bLTx without AS condition; [3] = significant difference with pre-AS condition; no significant differences were found between PAH-bLTx without AS and PAH-bLTx with AS at the same time-points.

Abstracts selected for poster presentation

Full text of the abstracts will be available on the ICA website: www.cteph-association.org

Poster session on Friday, 9th of June 08:30 – 15:45

Pathophysiology, epidemiology, diagnosis and prognosis

1. PULMONARY ARTERY SARCOMA AS A MIMIC CTEPH

D. Ambroz[1], P. Jansa[1], J. Kuchar[1], M. Niznansky[2], M. Aschermann[1], J. Linder[2]

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2. EPIDEMIOLOGY AND OUTCOMES OF PATIENTS WITH CHRONIC THROMBO-EMBOLIC PULMONARY HYPERTENSION REFERRED FOR ENDARTERECTOMY BEFORE AND AFTER INITIATION OF ANGIOPLASTY IN THE FRENCH NATIONAL REFERENCE CENTER

M. Amsallem[1,2], J. Guihaire[2,3], S. Guimaron[2], L. Lamrani[2], J. Arthur[2], O. Mercier[2,3], E. Fadel[2,3], P. Brenot[1]

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3. LONGITUDINAL CARDIAC DEFORMATION IS REDUCED BOTH AT REST AND DURING EXERCISE IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AND REMAINS IMPAIRED AFTER PULMONARY ENDARTERECTOMY

M. Claeys[1,2], G. Claessen[1,2], R. Willems[1,2], J.-U. Voigt[1,2], T. Petit[1,2], C. Belge[2,3], P. Claus[1], M Delcroix[2,3], A. La Gerche[1,4]

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4. MULTICENTER, OBSERVATIONAL SCREENING SURVEY FOR THE DETECTION OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION FOLLOWING PULMONARY EMBOLISM

N. Coquoz[1], D. Weilenmann[2], D. Stolz[3], V. Popov[4], A. Azzola[5], J.-M. Fellrath[6], H. Stricker[7], A. Pagnamenta[8], S. Ott[9], S. Ulrich[10], S. Györik[11], J.-D. Aubert[1]

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6. LOW-GRADE SARCOMA ORIGINATING WITHIN THE THROMBOTIC MATERIAL IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

D. Di Pernaa, A. Sciortinoa, C. Giordanob, M. Concardib, M. Agozzinob, V. Graziolia, M. Pina, V.N. Merlia, B. Vaninia, A. Celentanoa, I. Parisia, R. Dorec, E. Arbustinib, A.M. D'Arminia

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7. FAMILIAL CTEPH: A FAMILY WITH TWO AFFECTED MEMBERS TREATED WITH PULMONARY ENDARTERECTOMY (PEA)

D. Di Pernaa, V.N. Merlia, C. Giordanob, M. Concardib, T. Fiantaneseb, A. Sciortinoa, V. Graziolia, M. Pina, B. Vaninia, A. Celentanoa, I. Parisia, E. Arbustinib, A.M. D'Arminia

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8. INCIDENCE OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER ACUTE PULMONARY EMBOLISM – A SINGLE CENTER EXPERIENCE

O. Dzikowska-Diduch, M. Kostrubiec, M. Roik, B. Lichodziejewska, A. Wyzgał, A. Łabyk, Sz. Pacho, K. Kurnicka, P. Pruszczyk

Department of Internal Medicine and Cardiology, Medical University of Warsaw, Poland

9. HIGH SENSITIVITY OF A NON-INVASIVE SCREENING STRATEGY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER ACUTE PULMONARY EMBOLISM

Y.M. Ende-Verhaar[1], D. Ruigrok[2], H.J. Bogaard[2], M.V. Huisman[1], L. Meijboom[3], A. Vonk Noordegraaf[2], F.A. Klok[1]

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10. VENTILATION/PERFUSION SCINTIGRAPHY FOR THE DIAGNOSIS OF CTEPH AFTER SPLENECTOMY

M. Gerges, C. Gerges, G. Karanikas, W. Birkfellner, I. Schwarzinger, M. Publig, N. Skoro-Sajer, R. Sadushi-Kolici, M. Frey, M. Schaffarich, J. Jakowitsch, M. Hacker, W. Klepetko, I.M. Lang

Department of Internal Medicine II, Division of Cardiology, Vienna General Hospital, Medical University of Vienna; Department of Biomedical Imaging and Image-guided Therapy, Division of Nuclear Medicine, Vienna General Hospital, Medical University of Vienna; Center for Medical Physics and Biomedical Engineering, Vienna General Hospital, Medical University of Vienna; Department of Laboratory Medicine, Division of Medical-Chemical Laboratory Diagnostics, Vienna General Hospital, Medical University of Vienna; Department of Surgery, Division of Thoracic Surgery, Vienna General Hospital, Medical University of Vienna

11. CAN WE SELECT THE PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION CANDIDATES FOR PULMONARY ENDARTERECTOMY ON THE BASIS OF MULTIDETECTOR COMPUTED TOMOGRAPHY ANGIOGRAPHY ONLY?

H. Gonzalez[1], M.J. Lopez Gude[2], MT. Velazquez Martin[1], Y. Revilla Ostolaza[3], S. Alonso Charterina[3], M. Perez Nunez[3], R. Morales Ruiz[3], J.L. Perez Vela[4], A. Albarran Gonzalez-Trevilla[1], C.A. Quezada Loaiza[5], C. Ortiz Bautista[1], N. Ochoa Parra[6], I. Ponz De Antonio[1], J.M. Cortina Romero[2], P. Escribano Subias[1]

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12. UNUSUAL OBSTRUCTION OF THE PULMONARY ARTERIES

V. Grazioli, M. Pin, A. Sciortino, V.N. Merli, B. Vanini, A. Celentano, I. Parisi, G. Silvaggio, A.M. D'Armini

Department of Cardio-Thoracic and Vascular Surgery, Foundation "I.R.C.C.S. Policlinico San Matteo", University of Pavia School of Medicine, Pavia, Italy

13. ROLE OF S100A4 IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

P. Hacker[1], M. Kasiri[1], T. Raunegger[1], S. Janik[1], W. Klepetko[1], J. Ankersmit[1], T. Szerafin[2], I. Lang[3], N. Skoro-Sajer[3], R. Sadushi-Kolici[3], B. Moser[1]

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14. RISK FACTORS FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) – IMPORTANCE OF THYROID DISEASE

L. Hobohm[1,2], V. Krieg [1], C. Liebetrau[3], S. Guth [4], S. Koelmel[1], K. Pohl [1], C. Troidl [5], L. Essmann[5], H. Rossmann[6], S. Konstantinides[1], E. Mayer[4], C. Wiedenroth[4]*, M. Lankeit[1,7]*

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15. PROANGIOGENIC AND WOUND HEALING MOLECULAR AND HISTOLOGICAL FINGERPRINT OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

D. Iloska[1], J. Wilhelm[2], S. Guth[3], R. Savai[1,2], R. Schermuly[2], L. Fink[2,4], W. Seeger[1,2], E. Mayer[3], S.S. Pullamsetti[1,2]

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16. QUALITY OF LIFE AMONG CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION PATIENTS IN RUSSIAN FEDERATION

A. Klimenko, N. Shostak, V. Shemenkova

Pirogov Russian National Research Medical University, Department of Faculty Therapy named after A.I. Nesterov, Russian Federation, Moscow

17. MIXED VENOUS OXYGEN SATURATION CAN BE A PROGNOSTIC FACTOR OF HEMODYNAMIC STABILITY IN FOLLOW-UP PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

S. Minatsuki[1], M. Hatano[2], A. Saito[1], H. Maki[1], A. Kiyosue[1], J. Ando[1], I. Komuro[1]

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18. SCREENING OF AUTO-ANTIBODIES IN PATIENT WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) WITH PROTOARRAY® ANALYSIS

A. Naito[1], N. Tanabe[1,2], T. Hiwasa[3], T. Jujo[1,2], T. Sugiura[1], S. Sakao[1], K. Tatsumi[1]

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19. THE EFFECT OF MULTIDISCIPLINARY TEAM APPROACH ON THE IMPLEMENTATION RATE OF MAJOR DIAGNOSTIC AND THERAPEUTIC PROCEDURES OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

D.K. Oh[1], J.M. Song[2], D.W. Park[3], S.Y. Oh[4], J.S. Ryu[5], S.D. Lee[6], J.S. Lee[7], J. Lee[8]

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20. THE FIRST CASE OF AN EXTRASKELETAL MYXOID CHONDROSARKOMA IN THE PULMONARY ARTERY

I. Opitz[1], H. Klein[1], B. Bode[2], L. Freitag[3], D. Schneiter[1], S. Ulrich[3], F. Maisano[4], W. Weder[1]

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21. VALUE OF DUAL-ENERGY CT DERIVED IODINE MAPS FOR THE ASSESSMENT OF REGIONAL LUNG PERFUSION IN CTEPH COMPARED TO V/Q SCAN USING CT PULMONARY ANGIOGRAPHY AS A STANDARD OF REFERENCE

I. Opitz[1], M. Eberhard[2], M. Zaatar[1], W. Weder[1], S. Ulrich[3], T. Frauenfelder[2]

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22. SINGLE CENTRE PROSPECTIVE STUDY OF PATIENTS WITH CHRONIC THROMBOEMBOLIC DISEASE (CTED)

E. Swietlik, D. Taboada, A. Ruggiero, N. Screaton, E. Bales, L. Harlow, A. Fletcher, J. Cannon, K. Sheares, D. Jenkins, C. Ng, S. Tsui, J. Dunning, J. Pepke-Zaba, M. Toshner

Papworth Hospital NHS Foundation Trust - Cambridge/GB

24. HYPOCAPNIA AS A PROGNOSTIC MARKER OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

J.S. Ota-Arakaki, R.P. Ramos, E.V.M. Ferreira, C.M.S. Messina, A. Cepeda, C. Costa, M.T. Rodrigues, C.G.Y. Verrastro, L.E. Nery

Division of Respiratory Diseases, Department of Medicine, Universidade Federal de São Paulo, São Paulo, Brazil

25. INSPIRATORY MUSCLE WEAKNESS CONTRIBUTES TO EXERTIONAL DYSPNEA IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

R.P. Ramos, E.V.M. Ferreira, J.V. Rolim, I. Ivanaga, G. Figliolino, E. Brito, L.E. Nery, J.A. Neder, J.S. Ota-Arakaki

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Medical treatment and interventional treatment

5. TRANSITIONING FROM SILDENAFIL TO RIOCIGUAT IN PATIENTS WITH PERSISTENT OR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION IMPROVES EXERCISE CAPACITY AND HEMODYNAMICS

S. Darocha[1], M. Banaszkiewicz[1], A. Pietrasik[2], A. Dobosiewicz[1], M. Florczyk[1], R. Mańczak[1], M. Piłka[1], J. Norwa[1], J. Kępski[1], M. Wieteska[1], S. Szmit[1], A. Torbicki[1], M. Kurzyna[1]

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[2-] Medical University of Warsaw, Department of Cardiology, Warsaw, Poland

23. EFFICACY OF PULMONARY HYPERTENSION-TARGETED DRUGS FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER PERCUTANEOUS TRANSLUMINAL PULMONARY ANGIOPLASTY

R. Yanagisawa[1], T. Inami[1], M. Kataoka[2], H. Ishiguro[1], K. Takeuchi[1], H. Kikuchi[1], A. Goda[1], H. Yoshino[1], T. Satoh[1]

[1-] Second Department of Internal Medicine, Kyorin University School of Medicine; [2-] Department of Cardiology, Keio University School of Medicine

26. RESIDUAL RIGHT VENTRICULAR DYSFUNCTION AND REMODELING AFTER BALLOON PULMONARY ANGIOPLASTY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: PREVALENCE, CLINICAL CHARACTERISTICS AND PREDICTORS

D. Ambroz[1], P. Jansa[1], J. Kuchar[1], M. Niznansky[2], M. Aschermann[1], J. Linder[2]

[1-] 2nd Department of Medicine - Department of Cardiovascular Medicine, First Faculty of Medicine, Charles University and General University Hospital in Prague; [2-] 2nd Department of Surgery - Department of Cardiovascular Surgery, First Faculty of Medicine, Charles University and General University Hospital in Prague

27. BALLOON PULMONARY ANGIOPLASTY FOR CTEPH: 3-YEARS EXPERIENCE FOCUSED ON SAFETY AND EFFICACY IN 128 PATIENTS

P. Brenot, L. Moisson, C. Garcia, B. Gerardin, E. Fadel

Marie Lannelongue Hospital, Le Plessis Robinson, France

28. WARFARIN PHARMACOGENETICS IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

D. Di Pernaa, C. Giordanob, V. Favallib, A. Smirnovab, M. Urtisb, A. Sciortinoa, V. Graziolia, M. Pina, V.N. Merlia, B. Vaninia, A. Celentanoa, I. Parisia, E. Arbustinib, A.M. D'Arminia

Department of Cardio-Thoracic and Vascular Surgery; Centre for Inherited Cardiovascular Disease, Transplant Research Area; Foundation "I.R.C.C.S. Policlinico San Matteo", University of Pavia School of Medicine, Pavia, Italy

29. PREOPERATIVE MONOTHERAPY WITH PHOSPHODIESTERASE-5 INHIBITOR, ENDOTHELIN RECEPTOR ANTAGONIST, OR RIOCIGUAT DOES NOT AFFECT OUTCOMES AFTER PULMONARY THROMBOENDARTERECTOMY

T.M. Fernandes[1], B. Hsu[1], D.G. Papamatheakis[1], D.S. Poch[1], P.F. Fedullo[1], N.H. Kim[1], K.M. Kerr[1], V.G. Pretorius[2], M.M. Madani[2], W.R. Auger[1]

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30. RIOCIGUAT FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM A PHASE II LONG-TERM EXTENSION STUDY

M. Halank[1], M.M. Hoeper[2], H.A. Ghofrani[3,4,] F.J. Meyer[5], G. Stähler[6], J. Behr[7], M. Fletcher[8], P. Colorado[9], S. Nikkho[10], S. Hoernig[11], F. Grimminger[3]

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31. FOCUSED BALLOON PULMONARY ANGIOPLASTY FOR TRULY NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION – THE UK EXPERIENCE

S.P. Hoole[1], G. Coghlan[2], J. Cannon[3], K. Sheares[3], M. Toshner[3], D. Taboda[3], C. Ng[4], D. Jenkins[4], J. Pepke-Zaba[3]

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32. EFFICACY OF PTPA FOR LONG-TERM OUTCOME IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

T. Inami[1], M. Kataoka[2], H. Ishiguro[1], R. Yanagisawa[1], K. Takeuchi[1], H. Kikuchi[1], A. Goda[1], H. Yoshino[1], T. Satoh[1]

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33. EFFICACY AND SAFETY OF MACITENTAN FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM THE RANDOMIZED CONTROLLED MERIT STUDY

N.H. Kim[1], G. Simonneau[2], A.M. D'Armini[3], P. Fedullo[1], L. Howard[4], X. Jaïs[5], D.P. Jenkins[6], Z.C. Jing[7], M. Madani[1], N. Martin[8], E. Mayer[9], K. Papadakis[8], D. Richard[8], H.A. Ghofrani[10]

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34. COMPLICATIONS WITH THE USE OF THE NEW ORAL ANTICOAGULANT DRUGS IN THE SETTING FO CTEPH TREATMENT – REPORT OF TWO CASES

I.A. Andrade Melo[1], S.T. Lima[2], R. Machado[3], G. Fortunato[3], M. Kalil[3], R. Lima[3], R. Oliveira[3]

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35. SAFTEY AND EFFICACY OF OUR CURRENT STRATEGY OF BALLOON PULMONARY ANGIOPLASTY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

N. Mitsutaka, M. Hiromi

Okayama Medical Center, Okayama, Japan

36. BALLOON PULMONARY ANGIOPLASTY FOR PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: A JAPANESE MULTICENTER REGISTRY

A. Ogawa[1], H. Ito[2], H. Matsubara[1] on behalf of the Japanese Circulation Society Joint Working Group for the "Statement for balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (JCS 2014)"

[1-] Department of Clinical Science, National Hospital Organization Okayama Medical Center, Okayama, Japan;
[2-] Department of Cardiovascular Medicine, Okayama University, Okayama, Japan

37. RIOCIGUAT FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): 2-YEAR RESULTS FROM THE CHEST-2 LONG-TERM EXTENSION

G. Simonneau[1], A.M. D'Armini[2], H.A. Ghofrani[3,4], F. Grimminger[3], M.M. Hoeper[5], P. Jansa[6], N.H. Kim[7], M. Wilkins[8], A. Fritsch[9], N. Davie[9], G. Xing[10], E. Mayer[11]

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38. POOR SUBPLEURAL PERFUSION AS A PREDICTOR OF FAILURE AFTER BALLOON PULMONARY ANGIOPLASTY FOR NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Y. Taniguchi[1], P. Brenot[2], X. Jais[1], C. Garcia[2], O. Planche[1], E. Fadel[2], M. Humbert[1,2], G. Simonneau[1,2]

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39. IS PRESSURE-WIRE USEFUL TO PREDICT REPERFUSION PULMONARY EDEMA AFTER BALLOON PULMONARY ANGIOPLASTY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION?

M.T. Velazquez Martin[1], A. Albarran[1], I. Hernandez[2], S. Mayordomo[1], Y. Revilla[3], A. Roldan[4], M.J. Lopez Gude[5], J.M. Cortina[5], S. Alonso[3], A. Quezada[2], P. Pilkington[2], J.L. Perez Vela[6], C. Jimenez[4], F. Sliwinski[1], P. Escribano[2]

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40. RECURRENCE OF PLUMONARY HYPERTENSION AFTER BALLOON PULMONARY ANGIOPLASTY

J. Yamashita[1], N. Tanaka[2], M. Goto[1], R. Ito[1], K. Hoshino[1], N. Murata[1], T. Chikamori[1], H. Ogino[3], A. Yamashina[1]

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41. EVALUATION OF THE ACUTE EFFECT OF BPA BASED ON LESION TYPE USING PRESSURE WIRE IN CTEPH

K. Yanaka, K. Nakayama, T. Shinke, H. Otake, Y. Shinkura, N. Tamada, H. Onishi, N. Emoto, K.-I. Hirata

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Poster session on Friday, 9th of June 16:15 – 18:30 and Saturday 10th of June 07:45 – 13:30

Medical treatment and interventional treatment (continued)

42. REGRESSION OF LEFT MAIN COMPRESSION IN PATIENT WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER BALLOON PULMONARY ANGIOPLASTY

N.M. Danilov, Y.G. Matchin, K.V. Mershin, K.S. Chernichka, M.B. Karabasheva, O.V. Sagaydak, A.Y. Demchenkova, M.G. Mitroshkin, I.E. Chazova

Russian Cardiology Research Complex, Moscow, Russian Federation

43. EFFECT OF BALLOON PULMONARY ANGIOPLASTY ON RIGHT VENTRICULAR AFTERLOAD IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

C. Gerges[1], M. Gerges[1], N. Skoro-Sajer[1], R. Sadushi-Kolici[1], B. Moser[2], S. Taghavi[2], W. Klepetko[2], H. Matsubara[3], I.M. Lang[1]

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44. EFFICACY OF PERCUTANEOUS TRANSLUMINAL PULMONARY ANGIOPLASTY FOR CHRONIC PULMONARY THROMBOEMBOLISM WITHOUT PULMONARY HYPERTENSION

T. Inami[1], M. Kataoka[2], H. Ishiguro[1], R. Yanagisawa[1], K. Takeuchi[1], H. Kikuchi[1], A. Goda[1], H. Yoshino[1], T. Satoh[1]

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45. THE ROLE OF ECHOCARDIOGRAPHY TO DEMONSTRATE THE EFFECTS OF THE BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC HYPERTENSION

B. Lichodziejewska, O. Dzikowska-Diduch, M. Roik, S. Goliszek, K. Kurnicka, D. Wretowski, A. Łabyk, K. Irzyk, P. Pruszczyk

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46. RESULTS OF BALLOON PULMONARY ANGIOPLASTY

W. Magoń, J. Stępniewski, M. Waligóra, K. Jonas, P. Podolec, G. Kopeć

Department of Cardiac and Vascular Diseases, Jagiellonian University Medical College at John Paul II Hospital in Krakow, Poland

47. CHANGES IN SURFACE ELECTROCARDIOGRAM AFTER HEMODYNAMICALLY EFFECTIVE BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

M. Piłka[1], M. Kurzyna[1], S. Darocha[1], A. Pietrasik[3], R. Pietura[2], R. Mańczak[1], M. Wieteska[1], M. Florczyk[1], A. Dobosiewicz[1], A. Torbicki[1]

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48. REFINED BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION - 100 BPA SESSIONS WITHOUT MAJOR COMPLICATIONS IN A SINGLE CENTER

M. Roik, D. Wretowski, A. Łabyk, K. Irzyk, B. Lichodziejewska, M. Kostrubiec, O. Dzikowska-Diduch, P. Pruszczyk

Department of Internal Medicine and Cardiology, Centre for Management of Venous Thromboembolism, Medical University of Warsaw, Infant Jesus Teaching Hospital, Poland

49. BALLOON PULMONARY ANGIOPLASTY - A CURATIVE METHOD IN VERY ELDERLY PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

M. Roik, D. Wretowski, A. Łabyk, K. Irzyk, O.l. Dzikowska-Diduch, B. Lichodziejewska, P. Pruszczyk

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50. FIRST RESULTS OF BALLOON PULMONARY ANGIOPLASTY FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) PATIENTS IN BRAZIL

F.S. Souza[1], I. Melo[2], M. Leite[3], R. Abreu[4], M.G. Ferreira[5], P. Ribeiro[6]

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51. REFINED BALLOON PULMONARY ANGIOPLASTY FOR THE TREATMENT OF NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): INITIAL EXPERIENCE IN A CTEPH CENTRE

Y. Taniguchi*, O. Planche*, S. Bourkhis, A.S. Rangheard, F. Parent, O. Sitbon, M. Humbert, D. Montani, L. Savale, G. Simonneau, X. Jaïs

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52. HEMODYNAMIC AND CLINICAL IMPROVEMENT AT FOLLOW-UP AFTER SAFE REFINED BALLOON PULMONARY ANGIOPLASTY IN NON SURGICAL PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

M. Velazquez[1], A. Albarran[1], I. Hernandez[2], S. Alonso[3], M. Perez[3], S. Mayordomo[1], C. Jimenez[4], A. Quezada[2], M.J. Lopez Gude[5], J.M. Cortina[5], A. Roldan[4], B. Coto[4], N. Tovar[4], J.L. Perez Vela[6], P. Escribano[2]

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53. IMPACT OF THE EFFECT OF ADDITIONAL BALLOON PULMONARY ANGIOPLASTY FOR RESIDUAL PULMONARY HYPERTENSION AFTER PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

J. Yamashita[1], N. Tanaka[2], M. Goto[1], R. Ito[1], K. Hoshino[1], N. Murata[1], T. Chikamori[1], H. Ogino[3], A. Yamashina[1]

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Surgical treatment

54. PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: INITIAL SINGLE INSTITUTION EXPERIENCE

A. Acosta, M. Camporrotondo, M.L. Talavera, A. Meretta, M. Lopez, C. Lanfranco, A. Romera, V. Cortes, R Henquin, M. Diez

Hypertension Pulmonary team, Cardiovascular Institute of Buenos Aires (ICBA), Argentina

55. CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION - 10 YEARS SINGLE CENTER EXPERIENCE

M. Bohacekova, M. Kaldararova, T. Valkovicova, I. Simkova

Department of Cardiology and Angiology, Medical Faculty, Slovak Medical University and National Institute of Cardiovascular Diseases, Bratislava, Slovak Republic

56. TEN YEARS EXPERIENCE OF EXTRA-CORPOREAL LIFE SUPPORT AFTER PULMONARY ENDARTERECTOMY

D. Boulate, J. Bardet, S. Mussot, D. Fabre, F. Stephan, P. Brenot, P. Dartevelle, O. Mercier, E. Fadel

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57. PULMONARY ARTERY DENERVATION IN PATIENTS WITH RESIDUAL PULMONARY HYPERTENSION AFTER PULMONARY THROMBOENDARTERECTOMY: ONE-YEAR OUTCOMES

A. Chernyavskiy, N. Novikova, E. Pokushalov, A. Romanov, A. Edemskiy, E.N. Meshalkin

Siberian Federal Biomedical Research Center, Novosibirsk, Russia

58. PERSISTENT EXERCISE LIMITATION (EL) AFTER SUCCESSFUL PULMONARY ENDOARTERECTOMY (PEA): PREVALENCE AND CAUSE

A.G. Corsico[1], A.M. D'Armini[2], R. Di Domenica[1], V. Conio[1], V. Grazioli[2], A. Sciortino[2], M. Pin[2], V.N. Merli[2], F. Meloni[1], C. Klersy[3], I. Cerveri[1]

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59. ATRIAL ARRHYTHMIA IS COMMON AND ASSOCIATED WITH INCREASED LENGTH OF STAY AFTER PULMONARY THROMBOENDARTERECTOMY

T.M. Fernandes[1], B. Hsu[1], D.G. Papamatheakis[1], D.S. Poch[1], P.F. Fedullo[1], N.H. Kim[1], K.M. Kerr[1], V.G. Pretorius[2], M.M. Madani[2], W.R. Auger[1]

[1-] University of California, San Diego; Division of Pulmonary and Critical Care Medicine; [2-] University of California, San Diego; Department of Surgery; Division of Cardiothoracic Surgery

60. SURVIVAL BENEFIT IMPROVEMENT OF PULMONARY ENDARTERECTOMY WITH EXPERIENCE GAINED IN A NATIONAL EXPERT CENTER

I. Hernandez Gonzalez[1], M.J. Lopez Gude[2], M.T. Velazquez Martin[1], Y. Revilla Ostolaza[3], S. Alonso Charterina[3], M. Perez Nunez[3], R. Morales Ruiz[3], J.L. Perez Vela[4], A. Albarran Gonzalez-Trevilla[1], C.A. Quezada Loaiza[5], C. Ortiz Bautista[1], N. Ochoa Parray[6], I. Ponz De Antonio[1], P. Escribano Subias[1], J.M. Cortina Romero[2]

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61. PULMONARY ENDARTERECTOMY IN OCTOGENARIAN POPULATION: SAFETY AND OUTCOMES

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