5th European Conference on Neonatal and Pediatric Pulmonary Vascular Disease

University Medical Center Groningen, The Netherlands

Thursday
10 November 2022

Friday
11 November 2022

www.pediatricpvd.com

The 5th European Conference on Neonatal and Pediatric Pulmonary Vascular Disease is accredited by:

This program is accredited by the European Board for Accreditation of Continuing Education for Health Professionals (EBAC) for 14 hours of external CE credits. In compliance with EBAC guidelines, all speakers/ chairpersons participating in this program have disclosed or indicated potential conflicts of interest which might cause a bias in the presentations.

Each participant should claim only those hours of credit that have actually been spent in the educational activity.

To claim the credits participants will be asked to complete an evaluation survey. Each participant should claim only those hours of credit that have actually been spent in the educational activity.

Table of content

Welcome

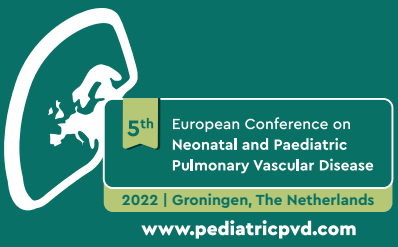
Dear colleagues and friends,

We are excited to welcome you to the city of Groningen for the 5th edition of the European Conference on Neonatal and Pediatric Pulmonary Vascular Disease. In many infant and childhood disorders, pulmonary vascular disease forms an essential component or comorbidity that will greatly impacts the child's wellbeing and prognosis. Due to improving cross borders organization and global cooperation in both basic and clinical research, we progressively succeed to translate advancing insights into the clinical care for our patients. However, there is still a lot to do! This conference will share and discuss current scientific evidence regarding diagnosis, treatment, and outcome of neonates, children, and adolescents with pulmonary vascular disease, as well as innovative concepts, future directions and cutting-edge research.

Due to the COVID-19 pandemic we had to postpone the celebration of the current lustrum-edition for one year, but now we are very happy to meet you all again and to share ideas in person. For this lustrum edition we are back at the University Medical Center Groningen where it all started almost ten years ago with the 1st European Conference on Neonatal and Pediatric Pulmonary Vascular Disease.
We would like to invite everyone to celebrate this joyous event with us at the conference dinner Thursday night at the Prinsenhof. For those of you staying in town a little longer, we will offer some "diversion" on Friday evening...

We wish you all an inspiring and exciting stay in Groningen!

On behalf of the conference committee,
Prof. dr. R.M.F. Berger, Chair



General information

Conference committee

- R.M.F. (Rolf) Berger
- J.M. (Menno) Douwes
- E. (Manolis) Mavrogiannis
- C. (Chantal) Lokhorst
- M. (Mark-Jan) Ploegstra
- M.T.R. (Marc) Roofthooft
- E. (Eva) Gouwy
- R. (Rosaria) Ferreira

Venue

The 5th European Conference on Neonatal and Pediatric Pulmonary Vascular Disease will take place at:

University Medical Center Groningen
Rode Zaal
Hanzeplein 1, 9713 GZ Groningen

The University Medical Center of Groningen is one of the biggest medical centers of the country. It serves as referral center for the north of the country as well as neighbouring regions of Germany. It is also a national referral center for a number of rare diseases among which pediatric pulmonary hypertension. One of the cores of the UMCG is its congenital heart diseases center serving pediatric as well as adult patients. Moreover, the UMCG is a prestigious transplantation center, allowing transplantations of more organs than any other medical center in the Netherlands. >

The southern part of the UMCG, where the conference will also be held, has been built in the 90's. The architectural design of the building has been focusing on the healing process, creating a city in the city. The hospital is close to the city center. The patient rooms are on the periphery of the building allowing every room to have view and light. The buildings in the center of the UMCG have streets and patios. It is thus possible for the visitors to walk in those indoor areas and feel that they are part of the community. Visitors, students and staff create a vibrant atmosphere. In addition to that, exhibitions, conferences and other events take place here. The UMCG is also a university campus for the faculty of medical sciences of the University of Groningen. The northern part of the buildings complex has lecture halls and buildings for research, among which the European Research Institute of Biology of Ageing.

In cooperation with Congress by design: CONGRESS BY DESIGN®

Questions during the conference?
The Congress by design staff will be at the registration desk during the conference, please do not hesitate to come to us with your questions. In case of an emergency, you can call us at +31 (0) 88 089 8101.

Faculty Members

S. Abman	Children's Hospital Colorado Denver, USA
M. Beghetti	University Hospitals of Geneve, Switzerland
R. Berger	University Medical Center Groningen, The Netherlands
H. Bogaard	Amsterdam University Medical Centers, The Netherlands
D. Bonnet	Necker University Hospital Paris, France
M. del Cerro	Hospital Universitario Ramón y Cajal, Madrid, Spain
R. Coleman	Baylor College of Medicine, Houston, USA
D. de Wolf	University Hospitals Brussels and Gent, Belgium
A. Everett	Johns Hopkins University School of Medicine, Baltimore, USA
J. Fineman	University of California, Benioff Children's Hospital, San Francisco, USA
M. Friedberg	University of Toronto, Hospital for Sick Children, Canada
R. Grady	Washington University School of Medicine, St. Louis Children's Hospital, USA
T. Humpl	St. Elisabethen-Krankenhaus Lörrach, Germany
D. Ivy	University of Colorado School of Medicine, Children's Hospital Colorado Denver, USA
D. Kiely	Sheffield teaching hospitals NHS foundation trust, University of Sheffield, UK
C. Lokhorst	University Medical Center Groningen, The Netherlands
E. Mavrogiannis	University Medical Center Groningen, The Netherlands
S. Moledina	Great Ormond Street Hospital, University College, London, UK
M. Pierro	Maurizio Bufalini Hospital, Cesena, Italy
M. Ploegstra	University Medical Center Groningen, The Netherlands
M. Prapa	St George's University Hospitals NHS Foundation Trust, University of London, UK
N. Subhedar	Liverpool Women's NHS Foundation Trust, UK
I. van der Sar	Erasmus Medical Center Rotterdam, The Netherlands
M. Westerterp	University Medical Center Groningen, The Netherlands
N. Wijbenga	Erasmus Medical Center Rotterdam, The Netherlands
B. Willemse	University Medical Center Groningen, The Netherlands

Program

Day 1 - Thursday, 10 November

07.30 - 08.30	Registration and coffee	
08.30 - 10.00	Welcome/Introduction	R. Berger
08.30 - 08.40	**Pediatric Pulmonary Hypertension: A short Introductory update course** **Chairs: R. Berger & D. Ivy**	
08.40 - 09.05	Diagnostic work up in 2023, the essentials	D. Ivy
09.05 -09.30	Treatment strategies in 2023, the essentials	D. Bonnet
09.30 - 09.50	Hemodynamic definition of PH in childhood	J. Fineman
09.50 - 10.00	Panel discussion: How to handle the new PH definition in children?	M. Beghetti
10.00 - 10.30	Break	
10.30 - 12.20	**Molecular Mechanisms in PAH, towards new therapies?** **Chairs: A. Everett & S. Abman**	
10.30 - 10.55	Inflammation in PAH: lessons to be learned from atherosclerosis	M. Westerterp
10.55 - 11.20	The role of inflammasome in the pathobiology of PAH	E. Mavrogiannis
11.20 - 11.45	New PAH therapies and trials to emerge from (pre-)clinical work	H. Bogaard
11.45 - 12.10	Mechanisms of right ventricular adaptation in neonatal and pediatric PH	M. Friedberg
12.10 - 12.20	Discussion	
12.30 -13.30	Lunch	

Program

Day 1 - Thursday, 10 November

13.30 - 15.15	**Advanced intensive care in neonates, children and adolescents with PH** **Chairs: Kieboom & D. Bonnet**	
13.30 - 13.55	Lessons from a pediatric ECLS program	R. Coleman
13.55 - 14.20	Lessons from a pediatric lung transplant program	B. Willemse
14.20 - 14.45	Lessons from a Potts program	M. Grady
14.45 - 15.15	Panel discussion: which strategy to follow...	
15.15 - 15.45	Break	
15.45 - 17.30	**Comorbidities in neonatal and pediatric pulmonary vascular disease** **Chairs: B. Willemse & S. Moledina**	
15.45 - 16.10	Pulmonary Hypertension and Down syndrome	T. Humpl
16.10 - 16.35	Pulmonary Hypertension and TBX4-variants	M. Prapa
16.35 - 17.00	Importance of phenotyping in children with PH	M. del Cerro
17.00 - 17.15	Discussion	
18.30	We will enjoy the Welcome Conference Dinner at the Prinsenhof.	

Program

Day 2 - Friday, 11 November

07.30 - 08.30	Registration and coffee	
08.00 - 10.00	**How to monitor your pediatric PH-patient** **Chairs: C. Lokhorst & R. Grady**	
08.00 - 08.25	Do we use risk stratification and risk scores in pediatric PAH?	S. Moledina
08.25 - 08.50	New ways to asses pediatric patients with PAH	A. Everett
08.50 -09.15	Clinical value of new imaging techniques in PAH	D. Kiely
09.15 - 09.40	Genetics in neonatal and pediatric PAH: ready for clinical risk assessment?	M. Prapa
09.40 - 10.00	Discussion	
10.00 - 10.30	Break	
10.30 - 12.25	**Clinical issues in neonatal pulmonary vascular disease** **Chairs: S. Abman & Kooi**	
10.30 - 10.55	Knowledge gaps in neonatal PH	N. Subhedar
10.55 - 11.20	Diagnosing PH in neonates and preterms: how and when?	D. de Wolf
11.20 - 11.45	Fetal growth restriction, pre-ecclampsia and neonatal PVD: common vascular pathophysiology?	M. Pierro
11.45 - 12.10	"Developmental lung disease" in WSPH classification: how to move forward...?	S. Abman
12.10 - 12.25	Discussion	
12.30 -13.30	Lunch	

Program

Day 2 - Friday, 11 November

13.30 - 14.00	**Best Poster presentations, oral** **Chairs: J. Douwes & M. Friedberg**	
13.30 - 13.40	Speckle tracking Echocardiography imaging to predict outcomes in pediatric PAH patients	G. Shatabdi
13.40 - 13.50	The additional prognostic value of pulmonary arterial compliance in children with IPAH/HPAH	E Gouwy
13.50 - 14.00	A multidisciplinary approach to treat PH in severe BPD: effect on sildenafil usage	E. Olson Jackson
14.00 - 15.15	**New technologies in de pediatric PAH arena** **Chairs: D. Kiely & T. Humpl**	
14.00 - 14.20	The E-nose in diagnosing PAH	I. van der Sar N. Wijbenga
14.20 - 14.40	Wearables and implantables in pediatric PH	M. Ploegstra
14.40 - 15.00	Artificial intelligence entering the PH	C. Lokhorst
15.00 - 15.15	Discussion	
15.15 - 15.45	Break	
15.45 - 16.45	**Challenging cases in pediatric PAH** **Chairs: M. Roofthooft & R. Berger**	
16.45 - 17.00	Closure and farewell	
18.00	This evening we will have a goodbye dinner at Bar1672	

Abstracts

Speckle tracking Echocardiography imaging to predict outcomes in pediatric PAH patients

Shatabdi Giri; Prashant Bobhate

Background: Pulmonary arterial hypertension(PAH) is a progressive disease with varying ages of presentation and is usually irreversible. The primary cause of death relates to deterioration of right ventricular (RV) function. Non-invasive assessment of RV longitudinal systolic strain predicts future right heart failure, clinical deterioration and mortality in patients with PAH. However, its prognostic value for pediatric PAH population is poorly defined. We aimed to use Speckle tracking echocardiography(STE) imaging to assess outcomes in pediatric PAH patients.

Methods: Ours was a prospective observational study done in single centre tertiary care hospital. Patients <18 years with PAH were included in study and those with significant intra/extra cardiac shunt or transient PH were excluded. Study duration:-June 2009-June 2022. STE was used to assess RV function at first visit. Primary end point included all-cause mortality, patients who required Potts shunt or those who were referred for heart and lung transplant. Statistical analyses was done using SPSS 20 software and a p value of <0.05 was considered significant. ROC curve analysis with equal importance to sensitivity and specificity was done and Youden's index was calculated as maximum distance from line of equality.

Results: Out of total 198 patients in pediatric PAH registry, 155 belonged to Group 1 PAH. 72 patients had significant intra-cardiac and extra-cardiac shunt lesion. Out of the remaining 83 patients,16 were lost to follow-up or had incomplete data, so a total of 67 patients were included in study cohort. The mean age at diagnosis was 7.56 ± 5.19 years with male:female ratio of 1.09:1. Majority presented with easy fatigue and right heart failure. Median follow-up:-1.16years(2 months-7 years). Study cohort was broadly divided into 2 groups:- Group 1-Alive(43) and Group 2-Expired(7)/Potts shunt(17) and functional ECHO parameters were compared between the two groups. Mean event free survival of study cohort was 4.2+0.3 years with 1, 3 and 5 year survival of 68%, 64% and 64% respectively. RV Global strain(RVGLS)< -12.95%,RV Free wall strain(RVFWS)< -16.65%,right atrial strain(RA strain)>14.25% ,RV fractional area change(RVFAC)>18.7% and RV ejection fraction(RVEF)>21.85% were associated with better outcomes. Patients showing better outcome as well as better 1, 3 and 5year survival had significantly lower RVGLS(p value-0.001) RVFWS(p value-0.023) and significantly higher RA strain(p value-0.002), RVFAC(p value-<0.001) and RVEF(p value-0.02). RVFWS and RVFAC were found to be most sensitive(82%) and RVEF was found to be most specific(74%) marker for predicting survival based on ROC curve analysis.

Conclusion: Predicting all-cause mortality and classifying PAH patients aides clinical decision-making. The ability for repeated assessment of RV function enables evaluation of disease progress which is feasible with functional Echo parameters. STE can be a useful marker to predict survival and outcomes in pediatric patients with PAH.

A multidisciplinary approach to treat PH in severe BPD: effect on sildenafil usage

Delphine Yung; Emma O. Jackson; Alyssa Blumenfeld; Greg Redding; Sara Berkelhamer; Laurie Eldredge

Background: Bronchopulmonary dysplasia (BPD) associated pulmonary hypertension (PH) is more common in severe BPD and leads to worse outcomes. The management strategy of BPD-PH is complex and has evolved toward common use of sildenafil.

Methods: We describe our center's multidisciplinary BPD consult team and systematic approach to BPD-PH, which emphasized frequent screening echocardiograms, optimization of respiratory support, and aggressive treatment of co-morbidities including PDA and ASD closure. Sildenafil was generally started only after lack of improvement in PH by echocardiogram and after confirmation of PH by cardiac catheterization.

Results: The BPD team followed 91 patients with severe BPD from 2018-2020, with minimum follow up of 1 year and 60 of those patients had an echocardiogram diagnosis of PH without critical congenital heart disease. All had PH resolution, including 3 who later died, 27 patients with PDA device closure and 5 with ASD device closure. Sildenafil was given to only 5 patients, with 2 stopping early due to adverse events. The other 3 (2 started by referring hospital) had PH resolve by echo after optimization and were allowed to outgrow the dose.

Three patients underwent cardiac catheterization for pulmonary hypertension with mean pulmonary artery pressures (mmHg): 21, 22, 30. Two deaths were due to complications of IVH, and one with tracheostomy emergency after discharge home.

Conclusion: Our systematic multidisciplinary approach led to much lower mortality and lower use of sildenafil in BPD-PH patients than has been reported in the literature, with complete resolution of PH. Unique features of our approach included aggressive PDA and ASD device closure and starting sildenafil only after ongoing optimization of respiratory support demonstrated lack of improvement of PH by echocardiogram.

STE imaging to predict outcomes after reverse Potts shunt in patients with Idiopathic PAH.

Shatabdi Giri; Prashant Bobhate

Background: Potts shunt has been suggested as effective palliative therapy and a bridge to heart-lung transplant for patients with Idiopathic pulmonary artery hypertension (PAH) not associated with congenital heart disease. However, not all patients may benefit from this high-risk procedure.
Objectives: 1. Correlation of functional parameters of right atrium and ventricle with invasive hemodynamics and lab parameters. 2. Utility of functional parameters to assess outcomes after reverse Potts shunt.

Methods: This is a prospective single-center study from April 2015- October 2020. All patients with PAH on maximal medical therapy and functional class IV or clinical deterioration were included and Patients with significant intra/extra-cardiac shunt were excluded. The primary end point was :- all cause mortality. Correlation was done between RV functional parameters and hemodynamic and biochemical parameters. RV functional parameters were also compared between patients who benefited from physiological Potts shunt and expired patients. Statistical analyses was done using SPSS 20 software and a p value of <0.05 was considered significant. ROC curve analysis with equal importance to sensitivity and specificity was done and Youden's index was calculated as maximum distance from line of equality.

Results: Our study included 19 patients (14 females and 5 males) with PAH and no intra- or extra-cardiac shunt undergoing Potts shunt (16 surgical Potts shunt and 3 PDA Stent) at a single tertiary care center. The Median follow-up was 38 (range 5-53 months). Right atrial (RA) strain correlated negatively with mean RA pressure (r2= -0.78, p value= 0.001) and NT pro-BNP (r2=-0.62, p value = 0.01) while right ventricular global longitudinal strain (RVGLS) correlated with cardiac index (r2= -0.52, p value= 0.03), Pulmonary vascular resistance index (PVRi) (r2= 0.6, p value= 0.01), and NT Pro-BNP (r2=0.65, p value= 0.003). RV free wall strain (RVFWS) correlated with cardiac index (r2= - 0.53, p value= 0.03), PVRi (r2=0.6, p value= 0.01) and NT pro-BNP (r2=0.57, p value= 0.01). Eight patients died and eleven patients showed sustained clinical and echocardiographic improvement. The patients showing benefit after Potts shunt had significantly lower preoperative RV GLS (-11.35% versus -2.85%, p value:0.002) and pooled RV FWS (-14.8% versus -2.2, p value:0.004) and significantly higher right ventricular fractional area change (FAC) (30.45% versus 7.39%, p value:0.001) than the patients who did not benefit from the procedure.

Conclusion: Functional ECHO parameters and speckle tracking imaging correlates well with invasive hemodynamic and laboratory parameters and can be helpful in predicting short and midterm outcomes after Potts shunt in patients with Idiopathic pulmonary hypertension.

Application nitric oxide generator «Tianox» in the treatment acute neonatal pulmonary hypertension

Artem A. Burov; Victor D. Selemir; Victor V. Zubkov

Background: At present nitric oxide for inhalation is produced by chemical synthesis at stationary stations. The Russian development by a group of authors from the Russian Federal Nuclear Center - All-Russian Research Institute of Experimental Physics of the nitric oxide generator. The technology is based on the process of atmospheric nitrogen oxidation in a nonequilibrium plasma of a gas discharge and is characterized by high operating accuracy and stable maintenance of the concentration of nitric oxide in the respiratory mixture. On the basis of the technology, a device for inhalation therapy with nitrogen oxide "Tianox" was developed and created. The device provides the synthesis of nitric oxide from the air directly during therapy, the supply of NO to the patient's respiratory circuit, adjustment and continuous monitoring of the concentration of NO in the respiratory mixture. Purpose: Evaluation of the effectiveness of inhaled NO-therapy in newborns with acute neonatal pulmonary hypertension in NICU. Materials and methods: In patients of the main group, NO was synthesized from atmospheric air using the Tianox device, in the retrospective control group, NO was delivered to patients from balloons using the SLE 3600 device.

Methods: A total of 109 newborns with acute neonatal pulmonary hypertension: 75 congenital diaphragmatic hernia, 3 cystic adenomatous lung malformation, 1 recurrent pneumothorax, 11 congenital pneumonia, 5 neonatal sepsis, 2 non-immune drops of the fetus and newborn, 2 critical CHD, 10 with multiple malformations.

Results & conclusion: Our study did not reveal significant differences in the effect on the small circulation, clinical course and risk of side effects between the methods of synthesis of NO from atmospheric air and dosing from cylinders. At the same time, the use of cylinders inevitably increases the economic costs of their purchase, turnover and maintenance. In addition, the presence of cylinders with toxic gas (the concentration of NO in the cylinder is 1000 ppm) under high pressure (150 atm.) is less safe for patients and ICU staff than the synthesis of NO from atmospheric air. The connection time of nitric oxide has been reduced to 45 seconds, the decision-making time for connecting nitric oxide by the doctors on duty has been reduced, there is no time for switching cylinders with nitric oxide, there is no occurrence of pulmonary hypertensive crises associated with switching cylinders, no procedure for calibration of monitoring sensors is required, the extension of indications for connecting inhalation of nitric oxide.

Risk stratification in adult and pediatric pulmonary arterial hypertension: a systematic review

Chantal Lokhorst; Sjoukje van der Werf; Rolf M.F. Berger; Johannes M. Douwes

Background: Currently, risk stratification is the cornerstone of determining treatment strategy for patients with pulmonary arterial hypertension (PAH). Since the 2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension recommended risk assessment, the number of studies reporting risk stratification has considerably increased. This systematic review aims to report and compare the variables and prognostic value of the various risk stratification models for outcome prediction in adult and pediatric PAH.

Methods: A systematic search with terms related to PAH, pediatric pulmonary hypertension, and risk stratification was performed through databases PubMed, EMBASE, and Web of Science up to June 8, 2022. Observational studies and clinical trials on risk stratification in adult and pediatric PAH were included, excluding case reports/series, guidelines, and reviews. Risk of bias was assessed using the Prediction model Risk Of Bias ASsessment Tool. Data on the variables used in the models and the predictive strength of the models given by c-statistic were extracted from eligible studies.

Results: 84 studies were eligible for inclusion, with this review focusing on model development (n = 20), validation (n = 13), and enhancement (n = 12). The variables used most often in current risk stratification models were the noninvasive WHO functional class, 6-minute walk distance and BNP/NT-proBNP, and the invasive mean right atrial pressure, cardiac index and mixed venous oxygen saturation. C-statistics of current risk stratification models range from 0.56 to 0.87 in adults and from 0.69 to 0.78 in children (only two studies available). Risk stratification models focusing solely on echocardiographic parameters or biomarkers have also been reported.

Conclusion: Studies reporting risk stratification in pediatric PAH are scarce. This systematic review provides an overview of current data on risk stratification models and its value for guiding treatment strategies in PAH.

The additional prognostic value of pulmonary arterial compliance in children with IPAH/HPAH

Eva Gouwy; Johannes M. Douwes; Douwe Postmus; Mark-Jan Ploegstra; Meindina G.Haarman; Rolf M.F. Berger

Background: Pediatric pulmonary hypertension (PAH) is a rare, progressive pulmonary vascular disease with pulmonary vascular remodeling leading to an increase in pulmonary vascular resistance (PVRi) and a decrease of pulmonary arterial compliance (PACi). PVRi and PACi form respectively the steady and pulsatile component of right ventricular afterload which is an important determinant of right heart failure and therefore prognosis in PAH. It is debated whether the relationship between PVRi and PACi, expressed as their product RC-time, is constant. The aim of this study is to determine the relationship between PVRi and PACi and to determine the prognostic value of PACi in addition to PVRi.

Methods: In this longitudinal retrospective study all children with idiopathic and hereditary PAH (IPAH/HPAH) treated in the UMCG that had a diagnostic heart catheterization between 1993 and 2021 were included. The relationship between PACi and PVRi and the relationships between PACi, PVRi, and RC-time and disease severity and 1-, 3-, 5-, 7- and 10-year outcome were assessed.

Results: In the 62 included patients (33 females) a relationship between PACi and PVRi was shown, however RC-time was found not to be constant. PVRi and PACi were associated with markers of disease severity (WHO functional class, NT-pro-BNP, Uric acid, Pulmonary artery acceleration time). PACi is associated with 1-,3- and 5-year transplant-free survival. PACi is associated with 1-year transplant-free survival independent of mPAP and PVRi. PVRi is associated with 3-,5-,7- and 10-year transplant-free survival. RC-time is associated with 10-year transplant free survival.

Conclusion: The relationship between PACi and PVRi is not constant. PACi is associated with disease severity and short(er) term outcome in children with IPAH/HPAH, especially 1-year transplant-free survival which is predicted by PACi independently of mPAP and PVRi. Therefore, PACi is of additional prognostic value next to PVRi especially on short term prediction.

Computational modeling for pulmonary arterial hypertension patients

Sander J. B. Schomaker; Rolf M.F. Berger; Tineke Willems;
Johannes M. Douwes; Cristóbal A. Bertoglio

Background: Mathematical models based on biophysical principles formalize the relation between properties of the cardiovascular system and its function. Therefore, by using least squares approaches, important parameters related to pulmonary arterial hypertension (PAH) can be extracted from clinical data. The goal of this research is to develop a 0D lumped parameter model to model the cardiovascular system, which is computationally efficient but still representative of the cardiovascular and in particular right circulation physiology. With such a model physiological correct pressures and flow rates within the blood vessels, and pressures and volumes within the heart can be modelled. Next, a fitting algorithm is created to get a personalized 0D lumped parameter model of the PAH patients. In the further, the model may help doctors diagnose different groups of PAH patients and determine at what stage of illness the PAH patient is. The personalized model can also be used to simulate exercise conditions.

Methods: The proposed 0D lumped parameter model within this research is a closed loop system – written in terms of differential equations – consisting of the right heart, pulmonary circulation, left heart, and systemic circulation. The equations are solved for pressures and flow rates computationally using a tailor-made numerical method. To create a personalized model the pressure data obtained by cardiac catheterisation, MRI flow-rate data, and the stroke volume of the left and right ventricle are used. The sum of the squared error is minimized by using an appropriate fitting algorithm. Important parameters related to PAH which are estimated with the model are resistances and compliance in the pulmonary artery, and stiffness and contractility of the right ventricle. With these parameters the stage of PAH but possible also different types of PAH can be determined. So far, personalized models are created for two idiopathic PAH/heritable PAH (IPAH/HPAH) patients who differ in symptoms during physical activity. Finally, the modeled resistances in the pulmonary and systemic circulation, and compliance in the pulmonary circulation are compared to the clinically measured values for these two patients. Also, the stiffness and contractility of the right ventricle are estimated with the model.

Results & Conclusion: For the resistances and compliance within the pulmonary circulation a relation is observed with the values determined within the clinic. The highest resistance and compliance in the pulmonary circulation are found for the IPAH/HPAH patient within WHO FC III and WHO FC II, respectively. The highest contractility and stiffness of the right ventricle are found for the IPAH/HPAH patient within WHO FC III. By fitting the model to the data of the two patients it is shown that the computational model of the cardiovascular system can be used to extract information about the patients.

Case presentations

Constrictive pericarditis presenting as right heart failure in case of Eissenmenger: A Case Report

Shatabdi Giri (1); Prashant Bobhate (2); Anuj Sharma (3)

Authors affiliation:
1. Kokilaben Dhirubhai Ambani Hospital, Mumbai, India
2. Kokilaben Dhirubhai Ambani Hospital, Mumbai, India
3. Sri Sathya Sai Sanjeevani Hospital, Palval, India

A Difficult Case of Systemic Sclerosis and Pulmonary Hypertension

Claire Parker (1); Jeff Fineman (2)

Authors affiliation:
1. UCSF Benioff Children's Hospital, San Francisco, USA
2. University of California, Benioff Children's Hospital, San Francisco, USA

A child with pulmonary hypertension and bronchopulmonary dysplasia - treatment strategy: how long should we continue PAH therapy?

Joanna Kwiatowska (1); Rafal Surmacz (2)

Authors affiliation:
1. Department of Pediatric Cardiology and Congenital Heart Defect, Medical University of Gdansk, Poland
2. Department of Pediatric Cardiology, Poznan University of Medical Sciences, Poland

Social Program

Welcome dinner
The Welcome dinner will take place at Prinsenhof, Grand Café. The restaurant is located in one of the oldest and most beautiful monumental buildings in the city.

Starting time: 18.30

Martinikerkhof 23
9712 JH, Groningen

Good-bye dinner
The Good-bye dinner dinner will take place at Bar 1672.

The restaurant is located in the heart of the city center where the ships used to trade, the rich artistic history is the main ingredient of Bar 1672.

Starting time: 18.00

Gedempte Zuiderdiep 15
9711 HA, Groningen

Notes

Notes

Sponsors

Platinum

MSD

BAYER

Gold

Occlutech

Salveo Medical

Silver

HeartMedical EuropeBv

ferrer for good

Supporter

gossamerbio


5th
European Conference on
Neonatal and Paediatric
Pulmonary Vascular Disease
2022 | Groningen, The Netherlands
www.pediatricpvd.com