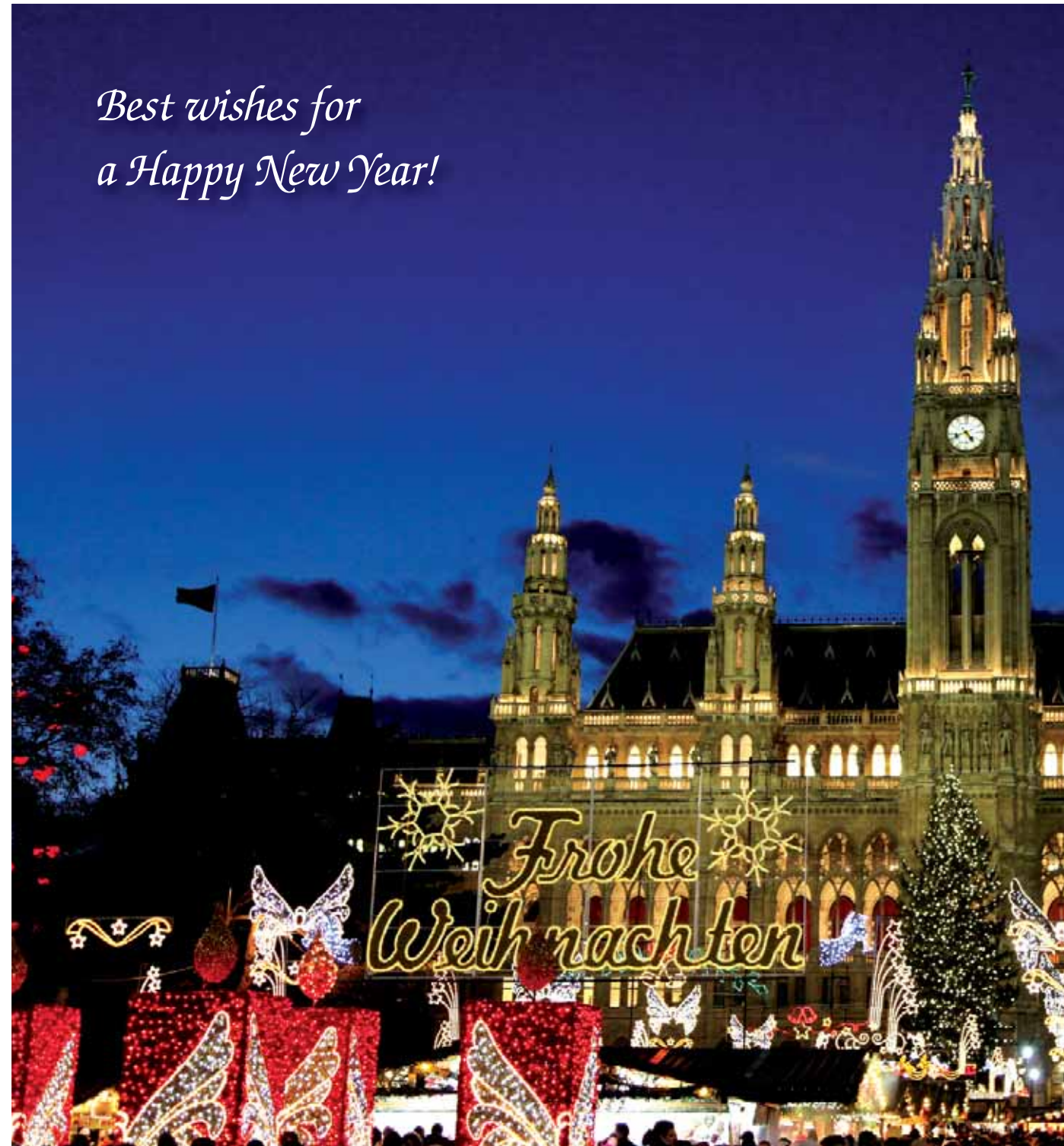




PULMONARY HYPERTENSION Mariposa Journal

WINTER 2013 • PHA EUROPE'S OFFICIAL JOURNAL • N. 11

*Best wishes for
a Happy New Year!*





Austria
Belgium
Bulgaria
Czech Republic
Finland
France
Germany
Greece
Hungary
Ireland
Israel
Italy
Latvia
Netherlands
Norway
Poland
Portugal
Russia
Slovakia
Slovenia
Spain
Sweden
Switzerland
Turkey
Ukraine

Editor’s memo

Winter 2013 edition

Dear friends,

welcome to the Winter 2013 edition of Mariposa, PHA Europe’s official journal. Another busy and exciting year has gone by and I take this opportunity to thank our member associations for their precious collaboration and support. Together we have come a long way and have made big steps forward in the process that we trust will ultimately lead to better conditions for all PH patients across Europe. This year, we have mainly concentrated on awareness raising, advocacy, and empowerment of the European PH community.

AWARENESS RAISING

PHA Europe’s awareness raising efforts were part of the global celebrations for the 2nd World PH Day, May 5. Some 40 PH associations in five continents took part in the campaign with unique events. A dedicated website was set up by the US PH association (PHA) to showcase the various activities conducted around the world.

PHA Europe’s campaign for World PH Day revolved around the theme of sports. People around the globe were encouraged to take part in sporting activities to “Get breathless” for PH. This was a powerful way of highlighting the restrictions on physical activity faced by PH patients and provided an effective way to raise further awareness of PH, bringing the challenges these patients face to the forefront. “Get breathless for PH” events were organized in 15 European countries. In line with the campaign’s focus on sport an additional 6 countries took part in the Ironman European tour. IRONMAN is possibly the toughest one-day endurance event in the world: races start with a 3.8 km swim, followed by a 180.1 km bike ride and ends with a 42.2 km run to the finish line.

“Get Breathless for PH” activities were first launched on the ski slopes of Japan and continued with sports matches, concerts, flash mobs, dances and fun runs in different countries, with excitement peaking on World PH Day itself, when Pope Francis offered a powerful message of encouragement to all PH sufferers during a Sunday Service in St Peter’s Cathedral Rome. The campaign attracted worldwide media attention, boosted by a host of celebrities who advocated the cause. Highlights included Latvia’s sports day which was opened by Olympic walking champion Aigars Fadejevs, whilst in Greece the Deputy Minister of Health, Marios Salmas, launched a city bike race and was cheered on by over 3,000 supporters.

ADVOCACY

PHA Europe’s advocacy activities mainly related to the follow-up of the PH Policy brief and Call to action, presented at the European Parliament in 2012, and the International Patient and Carer Survey, the key findings of which were presented at the 2012 General Annual Meeting. The data resulting from the survey reinforced the arguments put forward in the PH Policy brief, i.e. to the need to advocate for an integrated and multidisciplinary PAH care that includes the emotional, social and practical impact of the disease as well as unmet information needs. The findings of the survey were presented to the PH community with a scientific poster on “The impact of PAH on the lives of patients and carers” exhibited in February at the World Symposium on PH as well as at the August Annual Congress of the European Respiratory Society. Thanks to the active collaboration of the member associations a number of events were organized across Europe to promote the Policy brief and survey materials (both of which were translated into almost all European languages) and advocate for better standards of care for PH patients.

This year we were able to obtain the assistance of an EU-public affairs specialist to support our advocacy activities at EU level. This support consisted of carefully monitoring of EU initiatives to identify opportunities to provide input and possibilities for participation, as well as advice and practical support on how to get engaged. In the course of the year PHA Europe submitted a number of letters and position papers to the EU institutions (European Parliament, Commission, Council) on different topics of interest to PH (the EU Chronic conditions reflection process, health inequalities, Horizon2020). PHA Europe also supported a CTEPH research project submitted to E-Rare.

Thanks to the productive ongoing collaboration with the European Organization for Rare Disease, Eurordis, two PHA Europe representatives are now part of its Drug Information Transparency and Access (DITA) task force. PHA Europe has also recently become a full member of the European Patients’ Forum (EPF). Eurordis and EPF are both very important EU level stakeholders representing patient and it is an honour to be able to participate in their activities.

Relations with the scientific PH community and industry are also very important in terms of advocacy and awareness raising. PHA Europe is active in the work of a task force of the European Respiratory Society (task force on rare lung diseases) and advisory

committees of the European Lung Foundation (patient advisory committee). It was also present at the annual Congresses of the main scientific societies: the European Society of Cardiology and European Respiratory Society. These congresses are key events which are consistently attended by thousands of healthcare professionals, industry and other interested parties. Our presence enables us to present PHA Europe’s activities, distribute materials, to have access to the most up to date information on research and treatments and expand our contacts, an essential ingredient of networking, while also serving to raise awareness of PH among top medical and industry professionals. We were heartened again this year to note the large number of symposia, posters, abstract sessions, tutorials and other sessions specifically dedicated to Pulmonary Hypertension at the congresses of both the cardiology and respiratory societies.

In the course of the year PHA representatives were also invited to attend and/or speak at a number of conferences and industry events, including the 2nd Annual Pharmaceutical Pricing and Reimbursement Forum in Vienna, the Bayer Nurse Workshop in Stockholm, the AOP PH forum in Vienna, and many more. These occasions presented a welcome opportunity to provide the patient perspective and provide powerful insights into unmet patient needs and issues.

COMMUNITY BUILDING

PHA Europe strongly believes that patient associations play a fundamental role in providing invaluable support to patients and their families and that every European country should have a strong association in place. Over the years it has developed programmes to support the development of new associations and to provide ongoing support to established organizations.

The “White Spots” programme’s aim is to establish contacts and encourage patients, caregivers or physicians in the countries where there are no associations to create one. PHA Europe offers assistance in setting up, provides institutional and legal support concerning statutes and registration, basic start up informational tools (website, Facebook) and educational materials and resources. A number of countries have entered this programme in the past year.

Regular communication and exchange between PHA Europe and its members is vital to heighten awareness and engagement. Language barriers sometimes result in delays in the association’s activities. The “Fellowship” programme was set up with the aim of providing the national associations with an English-speaking part time assistant to act as the liaison person with PHA Europe and also assist with the national association’s day to day work. The pilot project launched this year ran successfully in nine countries (Bulgaria, Czech Republic, Greece, Hungary, Latvia, Poland, Portugal, Slovenia & Slovakia); the programme will be extended to further countries in the coming year.

PHA Europe believes that IT and social media can also play a vital role in the empowerment of the PH community. Therefore, it has invested time and resources in the unification of websites and Facebook pages for its members in order to enhance the role of social media and be able to conduct joint online campaigns. PHA Europe is also active with its own Facebook page and reg-

ularly updates its website. This activity will be increased next year with better use of social media to include Twitter.

CONCLUSIONS

It is hugely gratifying to see how our community has gradually built up over the years, the high level of activities the member associations conduct at country level and the enthusiastic support and participation in PHA Europe projects and campaigns. We are proud to have a broad and highly representative membership, with 29 patient associations from 25 countries. The increasing engagement of our member associations was also demonstrated by the very high attendance at the PHA Europe General Annual Meeting which took place in September 2015. Fifty five PH Patient Leaders from 27 countries were present and took an active role in the five day meeting. The atmosphere at the meeting was wonderful and the feedback extremely positive. PHA Europe looks forward to working with the member associations in successfully continuing its projects and activities in 2014 and future years. Thank you again for your collaboration.

I take the opportunity also to thank our industry partners for their trust and generous support of our activities, which has allowed us to build up our association and to come this far. We look forward to a long and mutually beneficial future collaboration.

Best wishes for a very Happy New Year!

Gerald Fischer
PHA Europe President

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FEBRUARY

Vienna, February 15
2ND ANNUAL PHARMACEUTICAL PRICING AND REIMBURSEMENT FORUM

Juan Fuertes, Member of the PHA Europe Board, represented PHA Europe at this important gathering of industry representatives. In his presentation he spoke about the expectations of PH patients to obtain the best possible treatment at an affordable price.



Brussels, February 27
EURORDIS GALA DINNER FOR RARE DISEASE DAY

Hendrik Ramaker, President of PH Belgium (FL) represented PHA Europe at the Annual Gala Dinner held by Eurordis, the European organization for rare diseases, held within the context of the International Rare Disease Day celebrations. He was a guest of Bayer.



MARCH

Nice, February 27-March 1
WORLD PH SYMPOSIUM

PHA Europe was asked to give its patronage this important event and was present with five representatives of national PH associations and a stand. On this occasion PHA Europe presented its scientific poster on “The impact of PAH on the lives of patients and carers”.



Athens, March 14-16
BAYER ANNUAL SYMPOSIUM

Pisana Ferrari represented PHA Europe at this important annual scientific event organized by Bayer. Over 350 medical professionals from 38 countries attended this meeting.



APRIL

Rome, April 19
EUPATI, EUROPEAN PATIENTS' ACADEMY ON THERAPEUTIC INNOVATION

More than 180 delegates from 28 countries participated at this meeting. Juan Fuertes attended on behalf of PHA Europe. The objective of EUPATI is by 2017 to train 100 patient experts from across Europe, to provide online education resources for a further 10,000 patient representatives and create an Internet library with information for anyone with an interest in medicines and R&D.



MAY

Sao Paulo (Brasil), May 4
LATIN WORLD PH DAY

Over one hundred patients and Latin American PH leaders held the regional celebration of WPHD in Brasil. PHA Europe's representative, Juan Fuertes, addressed the audience at the opening ceremony.



May 5
WORLD PH DAY

Over 40 PH patient associations worldwide took part in the celebrations for the 2nd World PH Day. Fifteen PHA Europe national affiliates organized “Get breathless for PH” events and a further six countries were involved in the Ironman European Tour, which formed an integral part of the campaign.



Dubrovnik, May 31-June 1
EURORDIS MEMBERSHIP MEETING

Over 200 participants from over 30 countries took part in this important annual meeting of the European organization for Rare Diseases. PHA Europe, Member of EURORDIS, was represented by Juan Fuertes.



JUNE

Stockholm, June 7-8
BAYER NURSE WORKSHOP

Hall Skaara, President of PHA Norway, and Patrik Hassel, President of PHA Sweden, were invited to speak at this scientific conference on behalf of PHA Europe. The workshop brought together 140 nurses from 18 countries, working in the field of PH.



AUGUST

Amsterdam, August 31-September 4
EUROPEAN SOCIETY OF CARDIOLOGY (ESC) ANNUAL CONGRESS

PHA Europe was present with a booth at the ESC Annual Congress and was represented by PHA Europe's Vice President, Pisana Ferrari, and Marzia Predieri, from the Italian PH Association AIPI. The Congress was attended by almost 30.000 professionals from all over the world.



SEPTEMBER

Barcelona, September 7-11
EUROPEAN RESPIRATORY SOCIETY (ERS) ANNUAL CONGRESS

PHA Europe was present with a booth at the ERS Annual Congress and was represented by Pisana Ferrari, Juan Fuertes, Irene Delgado, President of ANHP, and Regina Prenderville, from PHA Ireland. At the ERS PHA Europe presented its scientific poster on “The impact of PAH on the lives of patients and carers”. The Congress was attended by 21.000 professionals from all over the world. During the ERS Congress Juan Fuertes took part in the annual meeting of the EUROPEAN LUNG FOUNDATION'S PATIENT ADVISORY COMMITTEE.



Barcelona, September 11-15 **PHA EVENT**
PHA EUROPE GENERAL ANNUAL MEETING (GAM)

Fifty five PH Patient Leaders from 27 countries participated in the 2013 GAM. The agenda included medical updates and presentations on advocacy, communication & social media as well as interactive sessions with PHA Europe's industry partners.



NOVEMBER

Budapest, November 9
HUNGARIAN PATIENT MEETING

PHA Europe's President Gerald Fischer was invited to attend the annual meeting of the Hungarian PH patient association and speak about the activities of PHA Europe. The meeting was attended by over 140 patients and caregivers, PH physicians and other stakeholders.



Vienna, November 22-24
6TH ANNUAL CENTRAL AOP EUROPEAN PH MEETING

PHA Europe President Gerald Fischer was invited to attend this very informative meeting which gathered over 100 participants from all around the world. The meeting was chaired by Prof. Irene Lang, from the Vienna University, and had a very prestigious panel of speakers.



Santiago de Chile, November 22-23
LATIN PH DAY AND LATIN LEADERS SUMMIT

Latin American PH leaders meet annually for the Latin PH Day. The national PH organization of Chile was founded on this occasion. Juan Fuertes attended the meeting representing the link between the Latin American associations and Europe and made a presentation on how to manage a non-profit organization.



DECEMBER

Vienna, December 7-8 **PHA EVENT**
PHAEE BOARD MEETING

PHA Europe Board Members met in Vienna to review the projects and activities of 2013, the annual accounts and discuss future action and strategies.



London, December 9
DRUG INFORMATION TRANSPARENCY AND ACCESS (DITA) MEETING

Luc Matthysen and Juan Fuertes represented PHA Europe at this meeting which discussed important issues such as pharmacovigilance, transparency in clinical trials data and access to treatment. Luc and Juan are both members of the DITA task force.



Brussels, December 10
EUROPEAN VOICE PRESS CLUB

Juan Fuertes attended the official launch of the ERS paper “Lung Health in Europe: Facts and Figures” which addresses the challenges of lung diseases, including pulmonary hypertension.



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General Annual Meeting

Barcelona, September 11-15, 2013

The General Annual Meeting (GAM) represents one of PHA Europe's central activities. It provides PH patient association leaders with the opportunity for sharing information and experiences, mutual learning, networking, education, and skills development. The GAM also serves as a platform to showcase European and national initiatives and activities and discuss future strategies and projects. This year's meeting took place from September 11 to 15 in Castelldefels, near Barcelona (Spain). The turnout in Barcelona was the highest to date: 27 countries represented, of which four were newcomers: Austria, Belgium, Bulgaria, Czech Republic, Denmark, Finland, France, Germany, Greece, Hungary, Ireland, Israel, Italy, Latvia, Norway, Netherlands, Poland, Portugal, the Republic of Macedonia, Russia, Serbia, Slovenia, Slovakia, Spain, Sweden, Switzerland, the Ukraine. A total of sixty-five persons - patient representatives, speakers and guests - participated in this year's General Annual Meeting.

TEN YEAR ANNIVERSARY FOR PHA EUROPE

This year's GAM marked PHA Europe's tenth anniversary. During his welcome speech the President briefly told the participants the history of the organization. It all started in September 2003 in Vienna, thanks to the initiative of Bruno Kopp (President of PHeV, the German PH association). Bruno had organized a booth for PH at the European Respiratory Society's annual congress and had invited a small group of PH Leaders from eight European countries to attend. At this meeting the enthusiasm was such at being together for the first time and understanding the great potential of joining forces that it was very quickly decided to create a European association. As the years went by new national associations joined and PHA Europe became increasingly structured and

and organized, with a big step taken in 2010 to have full time staff. Today PHA Europe is a well recognized stakeholder in for PH and has conducted many successful activities in the areas of awareness, advocacy and lobbying, as well as support to patient associations through specific programs ("Fellowship" and "White Spots" programs). It has a very broad and representative membership, currently standing at 29 member associations from 25 countries. The President welcomed the four "newcomers" to the GAM from Denmark, the Former Yugoslav Republic of Macedonia, Serbia and the Ukraine. The President concluded this part of the meeting by thanking the representatives of the national associations for their hard work and enthusiastic collaboration, without which none of PHA Europe's achievements would have been possible.



Front row, from left: Pisana Ferrari, Irene Delgado, Luc Matthysen, Gergely Meszaros, Evgeniy Komarov, Luca Piali, Maria Joao Saraiva, Hendrik Ramaker. Middle row, from left: Yosef Gotlieb, Teresa Carvalho, Malene Thordal Johnsen, Solvi Mones, Ioanna Alisandratou, Melanie Gallant Dewavrin, Yana Yakovleva, Lolita Indriksone, Natalia Maeva, Juan Fuertes, Mihaela Raykova, Florence Szabo, Dominika Tepla, Luisa Sciacca della Scala, Gerald Fischer, Ocsana Alexandrova, Tuulia Naepi, Veronika Jelinkova, Todor Mangarov. Back row: Günther Thimm, Tadeja Ravnik, Klara Klanca, Daniela Naydenova, Antonios Konaratos, Wim Colle, Patrik Hassel, Eszter Csaboda, Regina Prenderville, Joanna Hryc, Sofie Moldenawer Christensen, Marzia Predieri, Hans-Dieter Kulla, Agnieszka Bartosiewicz, Yveta Makovnikova, Gjurgica Kjaeva, Annalena Bonelli, Peter Makovnik, Jolanda Velink, Maiia Vinnik (partially hidden), Lydia Benallouch.



David Roberts, United Therapeutics



Alma Rubio, Actelion



Left: Carl Hicks, VP of the PHA



The three winners for best World PH Day campaign: Poland (1st prize, center), Bulgaria (2nd prize, left), Greece (3rd prize, right)



Prof. Gérald Simonneau, France





From left: Pisana Ferrari with Jutta Ulbrich, Silke Gerlach & Yvonne Engler from Bayer Healthcare



Thomas Smogavetz, Ironman



Alma Rubio, Actelion (center)



ANNUAL ASSEMBLY

This more “institutional” part of the meeting was devoted to the renewal of the PHA Europe Board and Auditors, the presentation of the annual report and accounts and the review and approval of new membership applications. The Ukrainian association, along with the Greek association, were formally accepted as new members by the assembly. The new Board and Auditors for the 2014-2016 mandate remains unchanged with respect the the previous one apart from Juan Fuertes who was replaced by Vittorio Vivencio as 2nd Auditor.

MEDICAL UPDATE

It was a very big honour for PHA Europe to have as its guest medical speaker for this year’s GAM, Head of Pulmonology at the Hôpital Kremlin Bicêtre, Paris-Sud University, France. His presence at this year’s GAM also had a very high symbolic value for us as he attended the 2003 meeting in Vienna and strongly encouraged us to set up a European PH patient association, assuring us his support. We will always be very grateful to him for this. Prof. Simmoneau’s very interesting presentation summarised twenty years of progresses in the areas of PAH and CTEPH treatments, surgery, management strategies and looked at future developments eith regard to research on new molecules.

GAM SESSIONS

The GAM as always provides the opportunity to present the year’s projects and activities. There were sessions scheduled on World PH Day and Ironman, the International Patient and Carer survey follow up, the “White Spots” and “Fellowship” programs, advocacy at EU and national level, social media and communications, future projects for PHA Europe. A one-day session was also held during which our industry partners were invited to attend and address the members. The feedback from the participants was very positive. During the welcome dinner on the first day, three associations received awards for the best World PH Day national campaigns (1st prize Poland, 2nd Bulgaria - BSPPH - and 3rd Greece) and special mentions were given to Ireland and to the Italian patient association AMIP (for its flash mob where the Pope sent his greetings to PH patients).

CONCLUSIONS

The atmosphere at the meeting was, as always, warm and friendly, there was laughing and fun as well as some very touching and emotional moments. We have tried to convey this wonderful atmosphere by featuring more photos with respect to past years, which capture not only the meeting itself but the time spent to socialize and chat, laze on the beach, visit the sites in Barcelona and enjoy nice food and wine! Big thanks to Mihaela Raykova and Hall Skaara for the great photos. Most importantly, thank you to all those present for helping us make the annual meeting a truly unique and inspirational event!

Pisana Ferrari





The Fifth World Symposium on Pulmonary Hypertension: Inroads in Understanding, Diagnosing and Treating the Disease

by Yosef Gotlieb, PhD, PHA Europe, PHA Israel

The World Symposia on Pulmonary Hypertension (WSPH) started in Geneva in 1973 and have been held every five years after its convocation in Evian, France in 1998. The symposia have traditionally reviewed progress made in pulmonary hypertension science and paved the way for further advancement in research, diagnosis and treatment. It was a great honour for PHA Europe to be asked, along with PHA and leading medical societies in Europe, to grant it's patronage to the Fifth WSPH, which took place in Nice, France from February 27 to March 1, 2013. The presence of PHA Europe and PHA at the meeting was cordially noted from the podium and both groups were afforded an information booth at a central location of the conference venue. Indicative of the collaborative reception PHA Europe received was the inclusion of a scientific poster submitted by the organization for display at a dedicated poster session. The poster, "The Impact of PAH on the Lives of PAH Patients" was exhibited alongside major medical studies concerning the disease and outlines the results of a study on the psycho-social needs of PH patients and their carers. The study has influenced thinking and policy concerning approaches to PH treatment and integrated support. Five delegates from PHA Europe attended: President Gerald Fischer (Austria), Vice Presidents Pisana Ferrari (Italy) and Mélanie Gallant Dewavrin (France), Yosef Gotlieb (Israel) and Hall Skaara (Norway). They were accorded great courtesy by the organizers including participation at official dinners and other events. The symposium was attended by over 1,000 medical professionals and other specialists from all over the world. It provided an excellent opportunity for exchanges between the conferees and the patient associations delegates and enabled networking with key PH stakeholders including clinicians, researchers, and leading staff of pharmaceutical and biomed concerns. Further, the meeting provided a unique channel to become updated by the most prestigious and renowned specialists around the world about recent developments in PH research, diagnosis, treatments, case management, surgical and other interventional modalities and future strategies for screening and identification of groups at risk for the disease.



As a former patient who has been active in advocacy and support, attending the Fifth World Symposium on Pulmonary Hypertension (PH) held in Nice from February 27 through March 1, 2013, was enlightening, stimulating and affirming. The opportunity to observe over one thousand medical scientists, clinicians and researchers pooling their knowledge and experience in an effort to understand, effectively treat and ultimately vanquish the disease offers hope for all who have been touched by it.

I attended the conference as a delegate of the European PH Association, where I represent the Israel PH organization. Significantly, the European PH Association, which encompasses over twenty national groups, was asked along with the American association to endorse this scientific conference by the Symposium's organizers. This is a most notable milestone, representing growing recognition of the importance of patient representatives as a major partner-stakeholder along with clinicians, researchers and industry in a united community aimed at ameliorating and eventually overcoming this rare and disabling disease.

An Assembly of Leading Clinicians and Scientists

Prior to the start of the Symposium, members of twelve topic-specific task forces assembled to elaborate the results of pre-symposium preparatory meetings that were conducted over the previous 18 to 24 months. These task forces reviewed the major advances of pulmonary vascular science that have taken place since the previous Symposium (held at Dana Point, California in 2008), assessed research that has been undertaken since that time, identified questions of basic and clinical significance that the task force members were asked to consider, defined gaps in the knowledge base, and identified areas of research collaboration that have yet to be undertaken. A ten-member Symposium steering committee consisting of leading figures in PH science and treatment guided the task force groups and organized the conference. The Fifth World

Symposium on Pulmonary Hypertension was conducted under the patronage of the Pulmonary Circulation and Right Ventricular Function Working Group of the European Society of Cardiology, the European Respiratory Society, the Association for European Pediatric and Congenital Cardiology, the International Society for Heart & Lung Transplantation, as well as the European and American PH Associations.

The proceedings were opened and closed by two of the leading lights in PH science, Prof. Gérard Simonneau (Department of Pulmonary and Critical Medicine, University of Paris Sud) and Prof. Nazzareno Galiè (Institute of Cardiology, University of Bologna), who were instrumental in convening the meeting.

In the period since the first World Symposium was held in Geneva in 1973, interest in PH has burgeoned. At that time, a pioneering group of twelve experts were convened by the World Health Organization to characterize a sharp and deadly rise in incidences of high pulmonary vascular pressures associated with aminorex, a "diet" drug that acts to inhibit appetite. While PH had been described in the literature, it had not yet been well-characterized and the twelve experts, some of who remain active in PH treatment and were present at the Fifth Symposium, were tasked to come up with a clinical classification of the disease.

It was not until 1998 that a second World Symposium was convened, in Evian, where one hundred experts were present. In the fifteen years since then, symposia have been conducted every five years: In Venice in 2003 where the number of participants doubled to 200; in Dana Point, California (2008), where 350 attendees were presented, and; the fifth meeting, in Nice where more than a thousand participants were in attendance (February 27-March 1, 2013).

The exponential growth in the number of specialists convening to share and diffuse knowledge and acquaint themselves with the most updated diagnostic tools and therapies attests both to mounting interest in this rare disease and the many



still unanswered questions concerning those who contract PH, how it is acquired, in what ways the lives of these patients can be improved and, hopefully, how the disease might one day be prevented and perhaps cured.

During the three days of meetings twelve plenary sessions corresponding to each of the Tasks Forces were conducted with PH specialists from around the world presenting updated findings, policy recommendations, treatment protocols and diagnostic algorithms to the entire body of Symposium participants.

TASK FORCES AND SESSIONS

Each session stressed changes in understanding and the implications of the developments in the areas studied by the task forces. In an effort to expand the knowledge, pool and share expertise, each session presented questions to the entire assembly and asked for feedback from the doctors in attendance concerning matters of practice in the treatment of PH. The responses were collected by electronic voting, tallied in real time, and presented during each session to further discussions and provide greater insight.

Pathology and Pathobiology

The first session was presented by the **PATHOLOGY AND PATHOBIOLOGY** task force and dealt with the causes and mechanisms of the disease. A prominent finding of this group was that pulmonary veins, in addition to arteries, are involved in and undergo changes (remodeling) as a result of the disease. Further research has been called for to describe the relationship between clinical manifestations and the pathological evidence of this phenomena, including, a call for the improved vascular imaging of veins.

Evidence of involvement by the lymphatic system in the pathogenesis of the disease has also been observed, and the necessity of further research concerning its role was cited. The task force noted that the severity of damage to vascular cells in PH correlates with genetic influences and that genetic markers will likely help in determining a patient's presentation with the disease along two dimensions: a) time of symptom onset (early versus late), and b) disease severity. The genetic impact of the disease can be seen in terms of cell proliferation

and cell death resistance, increased signaling resulting in greater cell production, genetic instability (such as mitochondrial fission), and immune destruction.

The role of cellular inflammation in PH is now thought to be a central element in the disease, though its role is not yet understood. It is believed that inflammation constitutes a "second hit" which when accompanied by a known genetic marker of PH, Bone Morphogenetic Protein Receptor Type II (BMPR2) Deficiency, results in the disease. The presence of a BMPR2 mutation requires an additional factor before PH is manifested.

Genetics and Genomics

The second task force to offer their findings dealt with **GENETICS AND GENOMICS**, which presented findings that further illuminated the results given by the pathology and pathobiology group. An important distinction is made between heritable pulmonary hypertension (hPAH) and idiopathic Pulmonary Hypertension (iPAH). Eighty percent of families suffering from hPAH are positive for the BMPR2 mutation, while 20 percent of these families have no currently identifiable genetic mutation. In families with patients suffering from iPAH only 27 percent have been found with BMPR2 mutation. Not all people with BMPR2 mutations develop PAH, though 27 percent do. Importantly, there is a gender disparity in the development of PH in people carrying the BMPR2 deficiency mutation, with the incidence being much higher in women (42%) than in men (15%).

A major finding presented by the Genetics and Genomics task force involved the discovery of a second marker of the disease which could be used to identify people at risk in the twenty percent of hPAH families that do not carry the BMPR2 mutation. This marker, Potassium Channel Subfamily K Member 3 (KCNK3) is expressed by the proliferation of smooth muscle cells in the pulmonary artery. It appears to be equally manifested in both sexes and the clinical signs include early onset and progressive loss of function. Although people with the disease who have this marker currently have a negative prognosis, the discovery of this marker offers a new focus for medical therapy.

Prof. Marc Humbert of Paris who is both a veteran expert on PH and a medical ethicist discussed the ethical issues in offering genetic counseling to patients and their families,

especially to people with fPAH and their families, as well as to those with the idiopathic form of the disease. The dilemma derives from the difficulties in presenting possible and probable clinical scenarios to people at risk for the disease in the absence of favorable prognoses and treatment options. It is recommended that genetic testing for the disease should be conducted independently of genetic counseling since many people at risk for the disease are disinclined to learn about possible negative outcomes of this propensity. Other ethical quandaries pertain to reproductive issues concerning the counter-indication of pregnancy in women who have the disease.

The consensus of the Genetics and Genomics task force was that genetic influences clearly pivotal in determining the severity and progression of the disease as well as who is at risk at developing it. A fuller understanding of the genetic architecture of the disease, the pathways through which it develops, and its association with other diseases are critical areas for future research.

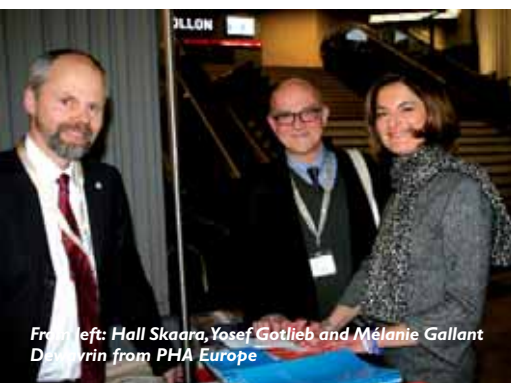
Definitions and Classifications and Particularities of Different PAH Subgroups

This was the subject of the third session. The system of PH typologies was introduced at the 1998 Evian Symposium and differentiated between five classes of PH disease on the basis of pathophysiology, clinical presentation and therapeutic options. The classifications have remained largely constant, although revisions have been made. These classes include:

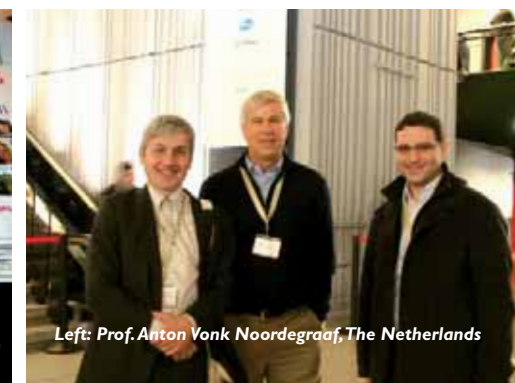
1. Pulmonary Arterial Hypertension (PAH), which involves remodeling of the pulmonary arteries and is often associated with other conditions such as liver or kidney disease, HIV, or connective tissue disease;
2. Pulmonary Venous Hypertension (PVH), which derives from diseases of the heart, the ventricles or the valves of the left-side of the heart or the compression of the central pulmonary veins;
3. Pulmonary Hypertension associated with Disorders of the Respiratory System or Hypoxemia, including interstitial lung disease and chronic obstructive pulmonary disease (COPD);
4. Pulmonary Hypertension caused by Chronic Thrombotic or Embolic disease (CTEPH), and;
5. Pulmonary Hypertension caused by disorders directly affecting the Pulmonary Vasculature, including capillaries.

Since the Dana Point Symposium in 2003 diagnosis of PH is made on the basis of hemodynamic criteria. The presence of PH is defined of in patients who have mean pulmonary arterial pressure (PAP) at rest greater than 25 milliliters of mercury (Hg). Normal levels are considered to be 20 mm Hg and below. The intervening levels, greater than 20 but less than 25 constitute an ambiguous area regarding treatment, especially since it is now largely recognized that PH intervention achieves best results when introduced as early as possible. Accordingly, this task force considered the possibility of introducing Borderline PH as a disease-related classification. This is particularly relevant in people with connective tissue disease, especially scleroderma, where the presence of borderline PAP levels correlates highly with eventual worsening and full-blown PH. The task force's consensus is that the hemodynamic determination of the disease is variable and that people with intervening levels of 20-25 ml Hg must be followed closely and their status considered in clinical context. A second issue dealt with this group was chronic heart disease (CHD) which is a life-long illness that affects one percent of all children around the world: Among these individuals, five to ten percent of those who live into adulthood will develop PAH, particularly those individuals who have Eisenmenger Syndrome. Since the survival rate of CHD has steadily increased, the number of patients living into adulthood has risen and, accordingly, so has the number of those who develop PH. In some cases, these heart defects are operable, although PH can also develop subsequent to surgery. For this class of pulmonary hypertension, pulmonary vascular resistance may be a better indicator of the disease than resting mean pulmonary arterial pressure since it can detect the disease even in the presence of normal levels of pressure.

Drug-induced and toxin-related PH remains a major concern of the PH community and determining levels of risk for development of the disease for individuals who use risk-associated drugs is a subject of ongoing research. Studies at multiple centers prompted this task force to develop a four-tier risk classification of drugs as having Definite, Likely, Possible and Unlikely risk association with PH. Aside from diet pills known to produce PH, there is evidence that selected serotonin reuptake inhibitors (SSRIs) may be associated with the disease. Also, PAH has been observed in people treated with the anti-cancer agent Dasatinib, as well as those who have



From left: Hall Skaara, Josef Gotlieb and Melanie Gallant
D. V. from PHA Europe



Left: Prof. Anton Vonk Noordegraaf, The Netherlands



Rino Aldrighetti, President of the PHA
and Gerry Fischer, President of PHA Europe



been administered some types of interferon (types 1, 2, 3) and amphetamine-like drugs.

A major conclusion of this task force is that a detailed history of drug use should be an integral part of screening for the disease. PH is also found as a co-morbidity of sickle cell anemia: Studies conducted in Brazil, France and the US suggest that ten percent of adults with this condition will develop pulmonary hypertension. Accordingly, patients with these diseases should be routinely screened for pulmonary hypertension.

Pathophysiology

Task Force number four concerned itself with the **PATHOPHYSIOLOGY** (focusing on exercise and the right ventricle (RV) of the heart) of pulmonary hypertension. PH results in insufficient blood flow the right ventricle and the level of pressure in the chamber remains constant both at rest and during exercise. This leads both to lower exercise capacity and reduced cardiac output. An imbalance between pressure and flow damages the right ventricle and constitutes a major pathology of PH. Further research into the molecular and cellular basis of RV pathophysiology is needed given that RV injury contributes significantly to morbidity and mortality in patients with PH.

Epidemiology and Registries

Task Force Five on **EPIDEMIOLOGY AND REGISTRIES** reviewed conclusions deriving from analysis of registries from different countries. Analysis of the French registry (2003-2008), as well as a study of 3,515 PH patients in the US conducted in 55 centers, found an increase in the number of men presenting with PH, a finding supported by the Spanish registry. Despite this increase, the US data indicates that 80 percent of PH patients are women and that the average age of onset has been increasing. The UK and Irish registry has found an increase in the number of cases of PH with comorbidity has been rising. A significant addition to the epidemiology of PH is found in the Chinese registry, which finds average PH incidence at younger ages (36 years plus/minus 13) occurring without comorbidities. Whether the phenotype of PH being seen by Chinese experts is different from those observed elsewhere remains an open question.

Analysis of historical data was shown that since the early 1990s survival with PH has significantly increased as a result of

new treatments. However, there remains a persistent lag in time-to-diagnosis, which continues to pose a major impediment to effective treatment. Patients who begin therapy at advanced stages of the disease have less benefit from treatment. There is a need to undertake prompt assessment, particularly of individuals at risk of being “rapid progressors,” i.e. people whose disease progression is likely to be more rapid and with earlier unfavorable outcomes than usually seen.

The consensus statement calls for a global registry that will minimize national and environmental variability and which would be comprised of rigorous, high-quality, complete and uniformly assembled data. Selecting centers capable of gathering sound and complete data is a major priority.

Caution was advised in generalizing data for long-term patients to newly diagnosed ones, since the former is based on documentable history which is not present in new patients.

Diagnosis and Prognosis

The sixth task force, focused on **DIAGNOSIS AND PROGNOSIS** discussed the possibility that cardiac output may be a preferred measurement for PAH in the early stages of the disease and for high risk populations.

Right heart catheterization remains the best diagnostic tool for PH although its results must be seen in clinical context. This is especially true in determining disease progression.

Therapy and Standards of Care

The seventh task force, on **THERAPY AND STANDARDS OF CARE** considered the issue of monotherapy versus combinational therapy. The recommendation is that those patients presenting with Functional Class Four should be immediately started with combinational therapy.

Combinational therapy is often indicated for patients at other functional classes as well, although an ongoing question relates to whether polytherapy should be initiated at the start of treatment or introduced sequentially. A clear recommendation on this question requires further research.

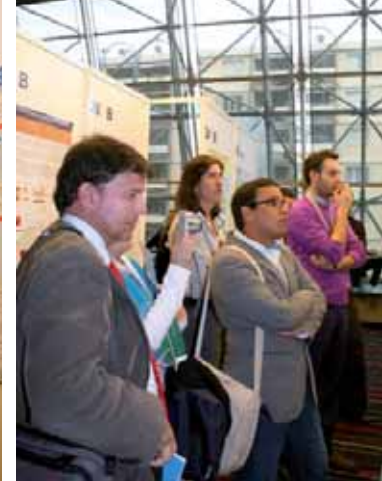
The session also considered whether treatment algorithms should differ by PH type. The recommendation is that there is no reason to alter existing protocols for the treatment of PAH. However, with respect to PH deriving from connective tissue disease, new research indicates that aggressive therapy is warranted given the serious outcomes of people with this form of



Prof. Ekkehard Gruenig, Germany, with Pisana Ferrari, VP of PHA Europe



Andrea Birtley, United Therapeutics, visiting the poster area



From left: Gerry Fischer, President of PHA Europe, with Prof. Nazzareno Galie, Italy

the disease. Those PH patients suffering from congenital heart disease have had good treatment outcomes with existing therapies, although those with right-left shunt disease may have their conditions worsened by intravenous treatments. No change in treatment protocol was recommended for those patients whose PH is associated with HIV. Concerning patients with portopulmonary hypertension, further research was called for to clarify the mixed results of current therapies.

The issue of adapting treatment and care protocols according to existing socioeconomic and geopolitical realities was felt to be beyond a single, overarching recommendation. Applying common sense on a case-to-case basis was advised.

Measures of clinical progress and functional class (FC) were considered by the specialists of this task force. The standard Six Minute Walk Test (SMWT), though widely used, was found to be at best a gross and variable measure. Time to Clinical Worsening (TTCW) is now a preferred endpoint for diagnosis and progress assessment, although the SMWT if modified with before and after measures of oxygen saturation, heart rate and systemic blood pressure has been shown to be relevant as well. The experts believe that in diagnosis and assessing patients clinicians should use a panel of indicators including results of hemodynamic studies, patient self-reporting, SMWT and TTCW.

Responding to dissatisfaction concerning the strength of recommendations and the level of robustness accompanying recommendations made by the tasks forces, the experts sought to clarify areas of obscurity.

They also emphasized that continuing research should encompass both randomized control studies conducted at multiple research centers and meta-analysis of data mined from registries and other statistical sources.

An important recommendation issued by this task force is that when there is an inadequate clinical response to monotherapy followed by combinational therapy, early referral for transplantation should be considered in order to maximize prospects of transplant success. Too often referral for transplantation is delayed, and the patient undergoes the procedure in a degraded state when it is difficult to survive the stresses of surgery and post-op rigors. Additionally, early transplantation should be considered promptly for early non-responders to other therapies.

Goals of Therapy

Task Force Eight, **GOALS OF THERAPY**, examined the prognostic indicators and the role they have in informing therapy. Among these indicators are functional class, hemodynamic results, echocardiogram and MRI readings, the BMPR2 biomarker, TSMW and cardiopulmonary exercise testing (CPET). The validity of Functional Class (FC) III as a separate clinical category was reconsidered and the validity of functional class as a useful indicator of disease progress was also reviewed.

The current classification rubric corresponds to these parameters: FC I involve patients with no limitations on functioning; patients with FC II have no limitations at rest but do face limitations on exertion; those classified with FCIII PH have marked limitations at rest and function with difficulty upon exertion, and; patients diagnosed with FC IV have severe dysfunction.

While functional class correlates with the quality of life, TSMW and survivability indicators, it correlates poorly with hemodynamic results. Importantly, it was noted that improvement in functional class as a result of treatment is a better indicator of positive outcome than the functional class the patient is assigned with at diagnosis or at the start of treatment.



Malcolm Allison, Bayer, and Alma Rubio, Actelion



From left: Yosef Gotlieb, Pisana Ferrari, Mélanie Gallant Dewavrin, Gerry Fischer



Mélanie Gallant Dewavrin with some of the French doctors including Prof. Olivier Sitbon (right)



Gerry Fischer with Dr. C. Gregory Elliott, USA



Prof. Ardeshir Ghofrani, Germany



Young investigator awards



The role of biomarkers in determining diagnosis and prognosis was discussed, though existing markers do not correlate well with hemodynamic results. Future research should pursue a multi-biomarker that would demonstrate both clinical and biologic validity and would serve as a better prognostic indicator.

The Goals of Therapy task force stressed the importance of using a panel of prognostic indicators rather than a single measure as there are problems with all of the existing criteria: Hemodynamic studies describe a situation specific in time; the SMWT is a gross indicator that does not correlate well with survivability and provides a skewed view of overall functional capacity (it is also unspecific concerning the specific effects of therapy); echocardiograms remain a useful, though inexact measure, as is the case with cardiac topographic imaging. Measures of right ventricular functioning are also pertinent and should be considered as a diagnostic and prognostic parameter.

The task force notes that further research is needed to determine whether therapy goals should be different according to the phenotype of the disease, that is, whether the underlying disease state is a significant determinant of which treatment will be most effective for a given patient.

The summary recommendation of this task force was that treatment goals should include the following parameters: Evidence of improved functional class (symptom relief), improved hemodynamic results and normalization of RV function, normal or near normal RV size and function as demonstrated by echocardiography or MRI studies, adjusted BMRP2 levels so they are normal for age and gender, SMWT with performance between 380-440 meters, and oxygen uptake levels (VO₂) of 15 L/min or greater as measured by a cardio-pulmonary exercise test (CPET).

New Trials and Therapies

Task Force Nine, NEW TRIALS DESIGN AND NEW THERAPIES dealt with clinical endpoints in the context of assessing new therapies in clinical trials. Optimal endpoints would be well-defined, reliable, consistent in their results and provide sensitive readings.

The changing “landscape” of pulmonary hypertension shows the improved functioning of patients due to more effective treatments and a larger arsenal of therapies. In order to assess new treatments, greater access to reliable pools of patient data is needed to evaluate both clinical efficacy and the biologic basis of the medication, the latter in order to determine whether the intervention acts on the underlying causes of the disease or only offers symptom relief.

Endpoints differ across the three stages of clinical trials. Phase One entails evaluation of the therapy for safety and in order to determine the limits of dose escalation. Evidence of activity, the presence of any adverse effects, and the range of dosing are also investigated at this stage of the trial.

Phase Two trials focus on efficacy as gauged by changes in hemodynamics and improvement in biomarkers, and clinically by performance on the Six Minute Walk Test (SMWT)

and Time to Clinical Worsening (TTCW). More nuanced effects such as the pharmacology of dose response, differential results across PH types, and impact on biomarkers are also assessed during this stage.

During Stage Three trials the primary endpoints are largely clinical and defined by morbidity patterns and mortality rates, TTCW, changes in functional class, SMWT and other indications such as the number of work days or school days missed. The Pulmonary Vascular Resistance Index (PVRI) measure was presented as an additional measure of clinical status.

The promise of specific classes of drugs were considered by the task force. Among these are vasodilators, which relieve the disease despite that pulmonary hypertension is not solely a disease of impaired vasodilatation. Other classes of promising medications include anti-proliferatives and pro-apoptotics and treatments involving gene therapy and stem cell and progenitor cell therapy. Tyrosine kinase inhibitors were discussed given that they target the vascular endothelial growth and also regulate metabolism and cell proliferation. Among other drug classes that are being investigated for potential benefits in treating PH include beta blockers (which have been shown to improve RV function), medications targeting the sympathetic nervous system (which have been shown to reduce pulmonary vascular resistance), motor inhibitors, medicines acting on nitric oxide and nitrate pathways and vascular mitochondria, monoclonal antibodies, the drug adrenomedullin (a vasodilator), and ranaolazine, an anti-anginal agent.

Non-drug therapies that have been used as bridging therapies to lung transplantation include cardiac resynchronization therapy and ablation, which has been shown to reduce sympathetic overdrive. Also, lung assistance devices have also been employed as a bridging therapy to transplantation. Discussions of regenerative and cell therapies aimed at generating vascularity in an impaired lungs include early attempts with angiogenic cell therapies which showed promise in the lab but which have not been translated into clinical practice. Experimentation with stem and progenitor cells, specifically epithelial and mesenchymal cells, have also been encouraging. Ultimately, a combination of cell and gene therapies could emerge leading to the regeneration of functional lung vascularity, though further work is necessary to translate these approaches for clinical use.

Given the extreme nature of the disease, a bold stand concerning novel therapies, including greater risk tolerance, is counseled. Criteria for prospective PH therapies must balance risk with the potential for significant benefit, including treatments that have toxic effects, in the hope that they can reduce overall mortality from the disease.

CTEPH

Chronic Thromboembolic Pulmonary Hypertension (CTEPH) was the topic dealt with by Task Force 10. This form of PH involves the presence of thrombotic obstruction



Gerald Fischer, President of PHA Europe, speaking at the Gala Dinner



Rino Aldighetti, President of the PHA, speaking at the Gala Dinner



and stenosis in the pulmonary arteries leading to high levels of pulmonary vascular resistance and right heart failure. Surgery, including embolectomy and thrombectomy with the aim of removing the arterial obstructions are regularly used to treat this class of PH.

Pulmonary endarterectomy (PEA) which results in the removal of clots is considered the treatment of choice for CEP_{TH}. Pulmonary angioplasty, which has been less well studied and requires more trials, is also a treatment option. The task force stressed that optimal therapy for a given patient should be explored on a case by case basis using a team approach involving PH physicians, expert radiologists and surgeons.

Section on the Task Forces Dealing with Left Heart and Lung Disease and Pediatric PH

[Note: I was unable to attend the two remaining sessions of the Symposium. Task Force 11 dealt with PH Due to Left Heart Disease and Chronic Lung Diseases, and Task Force, the final session, was dedicated to Pediatric PH. A section reporting on these sessions should be included here].

In addition to the formal sessions conducted by the task forces, poster sessions were offered on an extensive number of topics including Basic Science, Clinical Science, as well as the twelve subjects presented by the task forces in the plenary sessions.

Peer review papers based on the presentations delivered and task force recommendations are being prepared for publication as a special supplement to a major medical journal.

SUMMARY

As I look back on the Fifth World Symposium on Pulmonary Hypertension I recall how deeply impressed I was by this

assembly of experts who converged to pool their knowledge, analytical skill and clinical experience in an effort to aid PH patients and their families. That some of the pioneering investigators and physicians who attended the first meeting sponsored by the WHI in 1972 are still contributing to this field is a remarkable testimony to their persistence. Their success was evidenced by this large and highly scientific conference. The legacy of the pioneers is a flourishing medical concentration, PH science, that crosses specializations and involves clinicians and researchers who endeavor to improve and prolong the lives of patients who have been diagnosed with the disease, identify groups of people at risk of developing it, define the causes and mechanism by which the disease and its sub-types emerge and progress, improve diagnostic tools and genetic screening, and formulate new therapies to manage and, one hopes, eventually prevent and cure the disease.

That over a thousand medical scientists from all over the globe took the better part of a week away from their patients, families and research to travel to the south of France in order to share their knowledge and coordinate action attests to a high sense of purpose and commitment that these specialists share across borders. That the conference organizers and participants included, indeed embraced, representatives of patients associations and included industry professionals in the proceedings demonstrates progressive vision and the recognition that in order to defeat PH, the involvement of all stakeholders is necessary.

As the visual theme of the Fifth World Symposium on Pulmonary Hypertension the conference organizers chose Henry Matisse's 1909 painting *La Danse*. The iconic image of a circle of pastoral dancers suggests unity, sharing and caring. The motif represents the underlying spirit of the PH community and can serve as a model for endeavors, medical and other, where cooperation offers the best prospect for overcoming a common challenge.

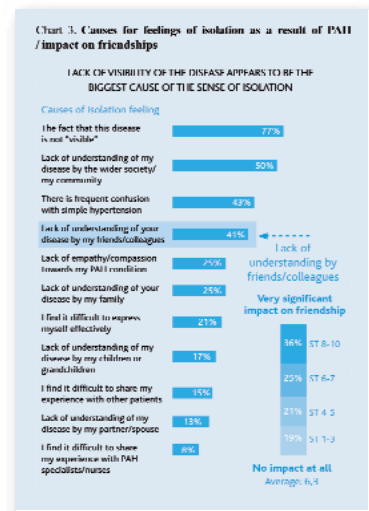


International PAH Patient and Carer Survey

An update of recent activities

INTRODUCTION

The International PAH Patient and Carer Survey (IPCS) was conducted to provide new insights into the wider impact of PAH on patients and carers beyond the clinical definition of the physical burden of the disease. Between 2010 and 2011, a total of 326 PAH patients and 129 carers across five European countries (France, Germany, Italy, Spain and the UK), replied to questionnaires asking about four main areas: the physical and practical impact of PAH, the emotional impact, the social impact, and information needs and provision.



Pisana Ferrari, Vice President of PHA Europe, was a member of the international Steering Committee (consisting of PAH specialists, nurses and representatives from PAH patient organizations), formed specifically to ensure the survey would be relevant at both a medical and practical level. The Steering Committee was also consulted on the analysis of the survey

results, to gain a thorough understanding of the personal perspectives of PAH patients, as well as their carers, and to help shape and endorse information resources to meet the gaps identified by the survey. Made possible through a grant from Actelion Pharmaceuticals, this is the first survey to explore the perspective of the carer as well as the patient on the physical, social, emotional and practical impact of the condition on their lives. It also looked at the information needs of PAH patients and carers and how these might change as the disease progresses.

MAIN IPCS FINDINGS

Top-line survey results revealed:

- The ability to work and the impact on household income is of particular concern to patients and their carers.
- The world narrows for PAH patients and carers as they are increasingly confined to their home with less ability to travel and socialize.
- PAH affects a patient's ability to be intimate with their partner mainly because of low self-esteem and being unable

or afraid to exert themselves physically.

- PAH patient and carers experienced feelings of isolation, mainly as a result of little understanding of the disease among family and friends.
- PAH patients and carers found there were gaps in information about the emotional and financial aspects of living with PAH.

The survey findings underline the need to recognize carers as individuals alongside the patients and provides a compelling argument for both to receive a more comprehensive standard of care, including psycho-social support. Optimum PAH management would ideally require a multidisciplinary approach including physicians, nurses, social workers and psychologists.

COMMUNICATING THE IPCS RESULTS AND CALL-TO-ACTION TO THE PH AND MEDICAL COMMUNITY

Presenting the IPCS findings and draft materials at the PHA Europe GAM 2012

PHA Europe officially presented the full survey results at its Annual General Assembly (GAM) on September 12-16 2012, in Castelldefels, near Barcelona, Spain. On this occasion, draft information resources (Patient and Carer Dialogue Tools) were submitted to the members for discussion and review. The Dialogue Tools provide a summary of the results from the international survey and contain a questionnaire, intended to help patients to document their thoughts, feelings and concerns in a structured way, which can help focus discussions with doctors, family, friends and others, and get the emotional support and best care possible needed to cope with their or their cared one's illness.



The Dialogue Tools were revised by the members and finalized in the weeks following the GAM and the final English edition was printed and distributed in early 2013. Members were encouraged and provided with support from PHA Europe in order to arrange for translations into their own national languages and in organizing local events and activities to promote the findings and Call to Action.



IPCS Scientific Poster at WSPH

The World Symposium on Pulmonary Hypertension (WSPH) started in Geneva in 1973 and has been held every five years after the Evian edition in 1998. The symposia mark the main progresses in pulmonary hypertension science and pave the way for

further advancements. PHA Europe was present at the 5th WSPH, which took place in Nice (France) from February 27 to March 1, 2013, with a booth and five delegates: Gerald Fischer, Pisana Ferrari, Hall Skaara (PHA Norway), Mélanie Gallant Dewavrin (HTAP France) and Yosef Gotlieb (PHA Israel). The symposium was attended by over 1,000 medical professionals from all over the world and provided an excellent opportunity for showcasing our activities and networking with all the key stakeholders in PH.

We were delighted that the Scientific Committee of the WSPH accepted our abstract on the findings of the survey and proud to present the scientific poster on "The impact of PAH on the lives of patients and carers" at a dedicated poster session. Many visitors came to the poster area, including Professors Nazzareno Galiè (see photo) and Gérard Simonneau, Co-Chairmen of the WSPH and Prof. Ekkehard Gruenig from the Heidelberg University (in the photo). Copies of the IPCS findings, executive summary, poster and patient and carer booklets were distributed (in English).

On 28 February, 2013, PHA Europe distributed a media release to healthcare journalists to coincide with the presentation. The aim of the media release was to generate coverage highlighting the results of the International Patient and Carer Survey, on the impact of pulmonary hypertension on a patient's overall quality of life. The release included comments from Prof. Galiè, Prof. Loic Guillevin and Dr Luke Howard. News coverage was secured in key online publications across Europe, resulting in widespread awareness of the challenges faced by PH patients and carers, generating over 80 pieces of online coverage and reaching a potential audience of about 2 Million people on the day of release.

IPCS Scientific poster at ERS congress

Vice President Pisana Ferrari (Italy), Juan Fuertes and Irene Delgado (ANHP Spain) and Regina Prenderville (PHA Ireland), represented PHA Europe at the Annual Congress of the European Respiratory Society (ERS), where the association also had a complimentary booth within the World Village area. More than 20,000 delegates attended the 2013 Annual Congress, which is Europe's largest annual scientific gathering in the field of respiratory medicine. We are very honored that the ERS accepted the scientific poster we had previously presented at the WSPH, summarizing the key findings of the survey, in the form of a poster session dedicated to PH. The ERS Congress provided a wonderful opportunity to further promote the results from the survey. We had many distinguished visitors, including Professors Sean Gaine and Andrew Peacock from the UK (see photo below) and Dr. Joan A. Barbera from Spain.



European Respiratory Review

Shortly after the summer, the IPCS survey Steering Committee submitted the survey results as a paper to the European Respiratory Review. The paper, entitled "Understanding the impact of pulmonary arterial hypertension on patients' and carers' lives" was accepted by the Editorial Board and has now been published.

To view the full article please visit:

<http://err.ersjournals.com/content/current>





<http://err.ersjournals.com/content/22/130/535.full.pdf+html>

Authors are: Loic Guillevin, Iain Armstrong, Rino Aldrighetti, Luke S. Howard, Henrik Ryftenius, Aryeh Fischer, Sandra Lombardi, Sean Studer and Pisana Ferrari, all members of the IPCS Steering Committee.

Scientific poster at ECRD

The survey Steering Committee has also submitted a scientific poster for approval to the European Conference on Rare Diseases and Orphan Products (ECRD), which will be held in May 8-10, 2014, in Berlin. The ECRD provides a unique platform for all rare diseases, across all European nations. With its high participation rate and excellent speakers, the congress covers the latest research, developments in new treatments and information regarding innovations in health care, social care and support at both the European and national levels.

We very much hope that we will be able to share the findings of the IPCS survey at this event and will keep you updated as we hear more.

WIDE-SPREAD NATIONAL ACTIVITIES, COMMUNICATING THE IPCS RESULTS

Multilingual dialogue tools

PHA Europe's 2013 Annual General Assembly was held in Barcelona on September 11-15, in Castelldefels, near Barcelona, Spain. During the GAM an interactive session was scheduled to present the activities that the national associations had held over the past year to promote the findings of the survey and its Call to Action. The GAM also provided the opportunity for the national associations to show and distribute printed versions of the Patient and Carer Dialogue Tools in 12 different languages (Bulgarian, Czech, French, German, Greek, Hungarian, Italian, Latvian, Polish, Slovakian Slovenian and Spanish). Further languages are planned for print shortly (Dutch, Hebrew, Macedonian, Portuguese, Russian and Swedish).

Sharing the IPCS findings across Europe

It has been amazing to see all the activities the different national associations have organized throughout 2013 to raise awareness of the survey findings. Activities have included presentations at patient and scientific meetings with healthcare professionals and interaction with the media. Further activities include organizing roundtable discussions or even collecting further feedback from patients and carers locally:

- **Bulgaria.** On September 10th BSPPH presented the

results of the survey during a round table with members of the Health Committee in the National Assembly, doctors working in the field of PH, nurses, social workers, psychologists, media and other stakeholders. On the 27th of November it organized a press conference to present feedback from the members on the Dialogue tools.

- **Czech Republic.** Distributed the Dialogue Tools to all patients and carers, and collected further feedback. They are now evaluating the findings.
- **France.** Distributed the survey report and Dialogue Tool booklets to newly diagnosed patients, at their patient conference and to each doctor in the expert centers in France. In addition they have also distributed a press release about the materials and the survey.
- **Germany.** Translated the full report and Dialogue Tools and included endorsement of the German, Austrian and Swiss associations. The Dialogue Tools have been sent to all members of the Association and presented at the PHev Annual Meeting.
- **Greece.** Translated the Dialogue Tools and distributed them to hospitals with heart-lung units throughout Greece. They have also held demonstrations with physicians around using the booklets to ensure they are as useful as possible. A press conference and information desks at PESPA also took place this year.
- **Hungary.** Presented the International Survey and Dialogue Tools at their national patient meeting.
- **Italy.** Presented the International Survey at their annual patient meeting. The survey was also quoted in the preface to a new AIPI publication called "Tips for everyday life with PH" and was talked about at press conference in Bologna on the occasion of an important PH scientific symposium in June, which had a big impact on the media.
- **Norway.** Organized a patient and carer workshop and discussed the findings of the International Survey.
- **Poland.** Presented the International Survey results at their patient meeting.
- **Portugal.** AHPH translated the materials developed from the international survey and intends to divulge the findings to the PH community. This activity is currently being integrated into the National Project of Characterization of Portuguese patients with PH.
- **Slovakia.** Presented the survey results at their patient meeting, as well as a presentation by Iveta Makovnicková,

the President of the Slovak PH Association and Dr. Milan Luknár on Slovak Television (STV 2 channel).

- **Slovenia.** Currently working with leading physician Dr. Barbara Salobir to ensure that every Slovenian PH specialist receives copies of the booklets to distribute among their patients.

- **Spain.** Presented the Inter-

national Survey at SEPAR, the main annual pulmonology congress and during the Week of the Heart in September.

- **Sweden.** Presented the International Survey findings at a Nurse Meeting.
- **Latvia and Russia.** Have translated the materials and have further activities planned.

A big thanks to all our members on sharing information about their activities, their experience and their ideas at the GAM. The discussions were a huge inspiration for all of us.

"TIME TO TALK"



PHA Europe has developed a special section of its website to host the IPCS findings and Dialogue Tools titled "Time to Talk". Simple and easy to use materials are currently being

developed for patients and carers to support them in confidently engaging with healthcare professionals, which will also be available under the "Time to Talk" section of the website. Resources are also being developed to support the member associations in best practice sharing and maximizing the survey findings.

FUTURE DEVELOPMENTS

Findings from the International Patient and Carer Survey, the US Patient and Carer Survey and the IMPACT Study undertaken by PHA UK, uncovered a need for patients and carers to have support and resources to help them build their confidence in engaging in dialogue with their clinicians, in order to truly empower them to manage their care.

PHA Europe is currently discussing together with PHA UK and PHA US a possibility to develop a program that would address those needs. We will certainly keep you updated around the programs progress in coming issues.

On behalf of PAH Europe, I would like to take this opportunity to thank all of our member organizations, patients and carers in their continued support and dedication. We very much look forward to working with you in 2014 and hearing about the activities and work you are all undertaking.

Copies of the full ICPS report are available for download from our PHA EU website www.phaeurope.org

Pisana Ferrari



This year's the ERS Lifetime Achievement Award in Pulmonary Arterial Hypertension was given to the Dr. Robyn Barst's Pediatric PH Research and Mentoring Fund. Carl Hicks, Vice President of the PHA, came to receive the prize on her memory on behalf of Dr. Barst's family. The photo (above) was taken during his presentation at the ERS and shows Maleen Fischer with Dr. Barst.



ERS Lifetime Achievement Award in Pulmonary Arterial Hypertension

Many of you know that my daughter is still with us only because of Dr. Robyn Barst, who more than 25 years ago started to treat children with PH.

This year Maleen was in intensive care in February, fighting for her life and Robyn called the hospital every day! Robyn passed away only 2 month later.

We loved her very much and we are sure that she still takes care of our Maleen.

Please donate to her fund:

<https://www.phassociation.org/Donate/Barst>

Thank you Robyn for everything

Gerald Fischer





AUSTRIA

**PATIENTS MEETING IN VIENNA**

On June the 22nd we held our annual patient meeting, in Vienna. There were about 90 patients from all over Austria. This was our first patient meeting for adults and children. The meeting started at 10:00 am and we had a lot of presentations about studies, new standards and pulmonary function. Upstairs in the second meeting room there were presentations from PH pediatricians for children with PH and their families. In the afternoon we heard a presentation from a representative of the Ministry of Social affairs. The last presentation was from Dr. Martina Schmidt from Actelion, she talked about the procedure for an active molecule to obtain recognition as an "orphan drug". The annual patient meeting was held with the sponsorship of Lego, the toy company.

MEETING IN ST. PÖLTEN

On September, 4th we organized a smaller patient meeting for about 30 patients in St. Pölten. We started with lunch in a comfortable restaurant in the city center. After that Dr. Mörtel, from the main hospital, informed our patients about the latest research studies and the rights of patients who take part in a research study. This topic was extremely important for all patients.

FIRST MEETING IN KLAGENFURT

For the first time we organized a patient meeting in Carinthia, in its capital Klagenfurt. It was a success that there were plenty of patients from all over Carinthia. We also welcomed a representative from the Ministry of Social affairs, to help our patients with all kind of questions.

**CHRISTMAS PRESENT FOR OUR PATIENTS**

As Christmas presents this year we are sending to all our patients a CURE PH scarf with a message to *please avoid cold air*.

ZOORUN

We very successfully organized again the Zoorun in Vienna with over 2000 starters. We were extremely happy about the net income of €22.000,00.

POLITICAL ACTIVITIES

We are proud that our political work is leading to the result that handicapped status and parking permits are combined in one Ministry instead of two as it is currently (Health and Social Affairs). Next year it should be the Ministry of Social Affairs only.

**RESEARCH SUPPORT**

We supported research for non-invasive measurement of pressure and cardiac output with €10.000. which were donated to the University clinics in Vienna (pediatric) and Innsbruck.

PSYCHOLOGICAL SUPPORT

We now offer to patients who are interested psychotherapy sessions in our office in Vienna. These sessions are conducted by a professional therapist, Dr. Maria Resch.

ENERGY FOR LIFE CONCERT

"Energy for Life" organized a classical music concert in Vienna for children with PH in May. "Energy for Life" is a social foundation whose main concern is the integration of socially disadvantaged young people including those with special needs.

**FACEBOOK CAMPAIGN**

With the new campaigns for PHA EUROPE rolling out this year, Austria was the first country to complete one. The incentive for people to get aware of Pulmonary Hypertension was to win an iPad. With a net amount spent of only 200\$ an astonishing 167,000 people saw the page during the 2 weeks during which the campaign was running.

PATIENT MEETING IN INNSBRUCK

We are busy organizing our 5th Patient meeting in Innsbruck which will take place on December, 5th.

Gerald Fischer, Selbsthilfegruppe Lungenhochdruck
www.lungenhochdruck.at
<http://on.fb.me/RzdEcb>



BELGIUM-HTAP

7TH EUROPEAN PATIENTS' RIGHTS DAY.

The EU dedicated the year 2013 to "European citizens and their rights". The EU Commission's goal is to raise citizens' awareness of their new rights resulting from the EU policies. The 7th European Patient rights' Day was held in Brussels on the 16th of May. This event focused on the EU Directive 2011/24/EU on the application of patients' rights in cross-border healthcare and patients' involvement. Mrs Testori-Coggi (European Commission - Director General of DG Health and Consumers) presented the "10 benefits the EU brings to patients".

http://europa.eu/rapid/press-release_MEMO-13-432_en.htm?locale=en

4th ANNUAL CONFERENCE OF THE EUROPEAN PUBLIC HEALTH ALLIANCE

This two-day conference entitled "Brave New World: Inclusive Growth and Well-Being or Vested Interests and Lost Generations?" was held in Brussels on September 4-5 and brought together more than 30 speakers from all across Europe's health professions, experts and civil society along with several institutional speakers, with an audience of more than 200 people. The Conference addressed the current failure amongst European

decision-makers to ensure that ordinary people, their health and well-being are sufficiently protected and promoted.

EPPOSI-STAKEHOLDER DAY 2013

This event was held in Brussels on the 16th of September. All the participants received an update of the core work programmes and how each of them is directly tied to shape future EU policies.

- AIP-INNO: work around innovation to support the policy on creating an innovation ecosystem in line with DG Connects vision.
- AIP-HTA: centers around an initiative to complete the societal elements of the EUNetHTA's HTA Core Model.
- AIP-RD: Rare Disease.
- AIP-CCM: programme on chronic conditions management.

PATIENTS' ASSOCIATIONS DAYS IN HOSPITALS

Last year Luss (the Belgium Patients Platform) started to organize patients' associations days in the hospitals. This year, this initiative was also organized in several hospitals but those hospitals became active partners. The hospitals organized lectures and a conference where the different associations could present their activities. The health care professionals were much more involved and the management recognized the value of the associations. This is a very positive achievement and Luss announce that it will organize these meetings next year as well.

Luc Matthysen, HTAP Belgique



Prof. Marion Delcroix, Belgium

them and they were soon ready to support us. The amount was divided so that the association also contributed its share. This intense financial cooperation is new to our association, and opens great prospects for the future!

After six months of (intensive) preparation we decided on a date for our 4th Annual patient week: October 12 & 13. No less than 80 people participated in the conference. About 10 speakers (professors, doctors and experts) brought the latest information about PH. Topics such as the World Congress in Lyon, the ABC of PH, insurance etc. were discussed.

The program allowed time for relaxation, music and socializing. We had a band performing, in the evening there was a quiz and karaoke and we also organized some workshops (making jewelry, e-bikes, crafts, etc.).

One particular point in the program: medical students were invited to write a paper. They did this exceptionally well. The two winners each received a cash prize of 250 euros each!

This was the "Jossie Habets" award, a prize in memory of Jossie, one of the founders of the organisation in 2003: it was given to to Esther Nossent (VU Amsterdam) and Jasper Vanhoof (UZ



Wim Colle



Hendrik Ramaker

Leuven) to encourage them in their research. Everyone went home tired but happy!

Wim Colle, Vice-President Patiëntenvereniging Pulmonale Hypertensie vzw

www.ph-vzw.be

<https://www.facebook.com/PHBelgium?fref=ts>



Luc Matthysen



BELGIUM-VZW

THE PATIENT WEEKEND, A BIG MOMENT FOR THE ASSOCIATION PH BELGIUM!

Together with Professor M. Delcroix our association put together a program that would provide a nice balance between information sharing and relaxation moments where patients could meet each other. The VZW Board also chose to make this event special for the patients and their families by

selecting a very nice venue, the Crowne Plaza Hotel in Brussels. We absolutely wanted to organize this conference so that it could be free of charge for the patients. Simple mathematics told us that the weekend would cost more than 16.000 euros, an amount that our association could not afford on its own. The solution was found in the dialogue with the pharmaceutical industry. Hendrik and Wim made contact with





BULGARIA-BSPPH

ROUND TABLE ON THE CHALLENGES FOR PATIENTS WITH PAH

The Bulgarian Society of Patients with Pulmonary Hypertension (BSPPH) is one of the 17 patient organisations in Europe which, in the course of 2013, presented the results of the international patient and carer survey recently conducted under the auspices of PHA Europe. This survey highlighted the urgent need for integrated care for patients living with this disease, to include psychosocial support. Bulgaria gathered patients, doctors, journalists, representatives of the institutions and other public figures in a Round Table which was held on 10th of September at the University Hospital for Active Treatment of cardiovascular diseases "St. Catherine" in Sofia. The goal of the meeting was to discuss the findings of the survey and present the new knowledge about the disease and its impact beyond the purely clinical aspects and to stress the need for a more comprehensive standard of care.

Participants at the Round Table included Dr. Ivan Ibrishimov - member of the Health Commission in the NATIONAL ASSEMBLY, Dr. Boyko Penkov - Deputy Minister of Health, Dr. Mariyana Simeonova - Director of the EXECUTIVE AGENCY FOR TRANSPLANTATION, Associate Professor Dr. Nikolay Dimitrov - National Cardiology Consultant, Dr. Stanimir Hasardzhiev - President of the NATIONAL PATIENTS' ORGANISATION and Acc.Prof. Dr. Borislav Velikov - Chairman of the Board of Directors of Aguachim, representative of Actelion for Bulgaria.

Cardiologists, pulmonologists, healthcare specialists, journalists and patients discussed the unsolved problems of PH patients. These discussions went way beyond the formal statements and marked constructive steps towards finding concrete solutions for PH patients.

Currently we have feedback from 12 patients (9 with PAH, 2 with CTEPH, 1 with systemic sclerosis and PAH) and 17 carers, who are willing to participate in the questionnaires developed as a result of the survey. BSPPH planned to present the results at a press conference at 27th November.



THE "BEZDIHANIE" CAMPAIGN WAS AWARDED A PRIZE IN BARCELONA

The "Bezdihanie" (Breathlessness) campaign organized in Bulgaria for WORLD PULMONARY HYPERTENSION DAY was awarded with the second prize at the General Annual Meeting of PHA EUROPE held in Barcelona in September 2013. The Bulgarian initiative was ranked second after the Polish association's campaign, and the campaign of the Greek association was third. BSPPH had organised a half-day dancing event under the motto "Take a Breath for the Pulmonary Hypertension". The event took place on 22 May 2013 in Sofia, in front of the NATIONAL PALACE OF CULTURE and was carried out under the patronage of the Mayor of Sofia Mrs Yordanka Fandakova.

The event gathered many people, revealing to them in an unobtrusive manner what does it mean to constantly remain "breathless" - one of the disease's symptoms. The initiative was covered in the media, in the national television, radio, and printed peer-reviewed publications, as well as in online portals. "The award we received from PHA EUROPE is a springboard for the events we are going to organize in the future. I believe it is a matter of huge importance to speak about pulmonary hypertension and to provide the patients in Bulgaria, as well as their families, with the psychological and healthcare service and expert support which they need. That is because in addition to the health aspect, there is also the social aspect to this disease. Therefore, such initiatives, where people by way of participation demonstrate their compassion, are very important - they provide the emotional support, which for the patients is as important as the proper treatment" said Mrs Natalia Maeva, President of BSPPH. The news about the award was covered in the media, in the Bulgarian National Radio and as well as in the online portal

<http://bit.ly/1c6AEag>

ONE "YES" CAN SAVE UP TO SEVEN HUMAN LIVES

BSPPH was involved in the Donor Awareness Week held from 7 through 13 October 2013. This campaign also took place in the cities of Ruse, Sliven and Pleven. The Donor Awareness Week started with an exhibition " #hashtag", opened by the Mayor of Sofia, Mrs. Yordanka Fandakova. The 8th of October was dedicated to the pulmonary hypertension in Sofia - capital of Bulgaria. At the information booth situated in the public park in front of the National Theatre of Sofia "Ivan Vazov", volunteers and representatives of BSPPH distributed information materials on organ donation and transplantation, as well as about pulmonary hypertension. Communicating with the visitors who stopped by the booth was rather interesting. We gathered quite disparate opinions on the issue of organ donation and transplantation. Many young Bulgarians insisted that their wish to be organ donors be indicated in personal documents. For patients with idiopathic pulmonary hypertension, it is particularly important to gain access to the European clinics performing lung and combined lung and heart transplantations since this could give them back their normal life. The good news is that Bulgaria is already a member of EUROTRANSPLANT, but even so a lot of work lies ahead. One of the major priorities for BSPPH's is to work and continue working for the causes of the National Donor Awareness Campaign.

"FACULTY OF PULMONARY HYPERTENSION": KNOWLEDGE IS HEALTH

The first ever specialized PH training program in Bulgaria, the "University of patients - Faculty of Pulmonary Hypertension" was launched on November 13, 2013 in the hotel "Down Town" in Sofia. This program is organized by BSPPH and is a joint initiative with the National Patients' Organization. The "Faculty of Pulmonary Hypertension" is aimed at providing up-to-date and useful information, training and support to patients with pulmonary hypertension, their families and loved ones, in order to acquire knowledge and skills for self-care and control of this serious disease. It is extremely important for patients to be real partners in the overall diagnosis and treatment process. The Program will run long-term and will rely on joint partnerships between PH medical experts, volunteers and members of BSPPH. One of the main objectives of this activity is to support the creation of a single medical register of patients with different subtypes of PH.

The project plans for training teams to be formed from the three centers in the country that monitor patients with PH. The teams will include a specialist in Cardiology, a coordinator

specialist, a nurse and a patient with PH. Training seminars and discussions also will be organized with approximately 30 patients in Plovdiv, Varna and Burgas. The first training module of the Faculty of PH took place in the hotel Down Town in Sofia and was opened with an exposé by Dr. Stanimir Hasardzhiev, President of National Patients' Organization. Dr. Lyubomir Dimitrov, a cardiologist from the National Cardiology Hospital, commented on the different methods of treatment of the disease in the context of current international practice and modern medical equipment for therapy. The psychologist and psychotherapist Ivo Velitchkov presented the professional perspective of applied psychology with the purpose of adaptation of these patients and their coping with negative mental states caused by pulmonary hypertension. Two patients also took part: Natalia Maeva, Chairman of BSPPH, and Milena Naneva, the longest surviving patient with PAH in the country. In their presentations they addressed the pressing problems of the disease.

PRESS CONFERENCE

On the 27th of November BSPPH organized a press conference at the Bulgarian News Agency on "Challenges for the patients with pulmonary hypertension in Bulgaria: problems and solutions". Nineteen journalists from different media attended the press conference as well as:

- Ass. Prof. Dr. Vasil Velchev - President of the Bulgarian Cardiology society, St. Anna Hospital, Sofia.
- Prof. Dr. Daniela Petkova - Pulmonologist, University hospital "Alexandrovska".
- Dr. Mariana Simeonova - Director of Executive Agency on Transplantation, Ministry of Health.

Discussions focused on the problems of patients with pulmonary hypertension in the country. Natalia Maeva from BSPPH presented the international patient and carer survey and the dialogue tools developed for patients and carers. These had been sent out by BSPPH to the patients with feedback from 31 respondents (9 patients with PAH, 2 with CTEPH, 8 with secondary pulmonary hypertension and 11 carers). All the respondents agreed that for them it is a serious disease; 25% admit that they are constantly depressed; 12% said that they lost a lot of friends; 15% are hiding that they are diagnosed with PH.

Natalia Maeva, Bulgarian Society of Patients with Pulmonary Hypertension - BSPPH

www.bspph.net

www.facebook.com/BSPPH.Bulgaria?ref=hl



First from left: Natalia Maeva, at press conference



BULGARIA-PHA

SECOND NATIONAL CONFERENCE ON PH

The Second National Conference for Pulmonary Hypertension took place on 23rd and 24th of November at the Hotel Augusta SPA in Hisar. The event was organised for patients suffering from PH and medical students, studying in this field. The motto of the conference was "Early diagnosis - Best treatment - Appropriate monitoring - For all PH patients". The best specialists in the field of PH together with 48 patients, 20 guests and 79 medical students from the 6 main medical universities in Bulgaria attended the event. Detailed presentations about the illness, the situation in Bulgaria and the problem with transplantation in the country were made by leading specialists in the field: Prof. Nina Gotcheva, national cardiological consultant, associate Prof. Margarita Tsonzarova - national paediatric cardiology consultant, associate Prof. Yoto Yotov - interventional cardiologist in the Reference Center for PH, Zeregyul Mualibova M.D., cardiologist in Reference Center for PH, Mariana Simeonova M.D., Executive Director of Agency of Transplantation. The event was open for all patients with pulmonary hypertension. The attendance of each participant was fully funded by PHA Bulgaria. We can consider 2013 a successful year for PH patients in Bulgaria. Since the beginning of the year 4 patients started treatment with drugs. So far, 70 patients have the necessary drugs provided by the National Health System in Bulgaria. PHA Bulgaria, together with the National Health System is in the process of preparing an application for thromboen-



derectomy for a CTEPH patient. The operation is scheduled for early 2014 at the hospital of Prof. Klepetko.

Todor Mangarov, PHA Bulgaria
www.apph-bg.org
www.facebook.com/aph.bulgaria



not meet the criteria for getting these cards. The specialist from the Council gave us advice how to appeal against this negative feedback.

RECONDITIONING STAY IN PODEBRADY 2013

As usual in August, our patients, their families, our PH doctor and physiotherapists gathered in spa town of Poděbrady for the annual reconditioning stay. For all of us, it has become a nice tradition to spend one week every summer in this beautiful town, west of Prague, doing light workouts (both in a gym and in a pool) under the supervision of our therapists, short walks along the Labe river or fun quiz tests.

Our patients also had the opportunity to discuss their problems with a PH specialist.

PATIENTS AND CARER SURVEY

This year, thanks to PHA Europe, we had the opportunity to take part in a survey which highlights the issues faced not only by patients with PH but also their family members and friends. Currently we are collecting the booklets and evaluating the findings. We would like to present the results at our meetings and also to make a poster we could present at the Annual Congress of Cardiology in Brno next year.

6 MINUTES FOR YOUR HEALTH

In October 8th our association and PH Dr. Pavel Jansa were invited to Usti nad Labem (a town north of Prague), where a unique project called "6 minutes for your health" was launched. The idea comes from the Czech cardiologist Dr. Iveta Petrova, the main goal is to test your physical condition during the 6 minutes of walking. In a public park in the center of this town you can find a path which is around 150 meters long, where you can measure the distance you walked in 6 minutes and check your results at the information boards situated along the path. No matter what your age or gender is you can try to walk the path at any time of the year and use this test as a potential indicator for evaluating your health condition. As you know PH doctors use the 6 minute walking test to evaluate the physical condition of their patients and of other people who could be diagnosed with this terrible disease.

We are already looking forward to the next year's activities!

Kateřina Nováková
 Sdružení Pacientů s Plicní Hypertenzí
www.plicni-hypertenze.cz



Dr. Pavel Jansa (left) and Kateřina Nováková (right)



CZECH REPUBLIC

ANNUAL ASSEMBLY IN SEČ

Like every year in June leaders of our association and its members met in the beautiful surroundings of the Iron Mountains for their Annual Assembly. The main topic of our meeting was presented by a specialist from the Czech NATIONAL DISABILITY COUNCIL. Due to some recent changes in the law, for the most of our patients with pulmonary hypertension it is almost impossible to obtain or renew health disability cards. When they apply for a new one their application is, in most cases, rejected this goes also for their parking card, with all its benefits. The main reason for rejection is the fact that, from 2012, patients with pulmonary hypertension (but not only) do



FRANCE

INTRODUCTION

At HTAPFrance, we concentrate on information directed to patients and their families, because we know that it is the first action they expect from us, together with supporting research. All our activities give priority to information and getting people to meet and share experiences. That is why all our regional meetings, annual assembly, magazine, internet site, hotline and leaflets focus on this aspect. But we also appreciate it when local volunteers organize activities to raise awareness on PH.

AWARENESS RAISING

Even if PH World Day has been extensively described in Mariposa summer issue, I would like to mention the activities that took place in France on this theme. But not on May 5th! In particular, three events were supporting the idea of “Get Breathless for PH”.

First of all, Agnès who lives near Marseille organized with Rotary International and Curves, both in Gemenos, a big Zumba dance, on the main square of this town. Some members of our association were there to raise awareness and respond to people's questions and tell about their own stories.

A week later we held our traditional Ultra Marathon, 205 kilometers between Toulouse and Port La Nouvelle, along the well known Canal du Midi and Canal de la Robine, to the Mediterranean sea. It took them two days, some were running the all way (we call them “Ultras”), some rode a bicycle the whole

way, and some did a part of it, from 10 kilometers to 190! We used the “Get breathless” t-shirts on the podium, when the journalists and the public were there, but not on the race because we needed more than 400! There were more than 200 runners or cyclists, and about a hundred volunteers to secure the path, organize the catering etc.. There was, as always, a great atmosphere! It was my first time at the Ultra-marathon, although it was the 9th edition... and I could not believe in what I saw and heard. It was so emotional, and people discovering this were so astonished and then seduced! There were many articles in the local press and on the internet.

Last of all our WPHD events, on the 15th of September, we organized a vintage car rally in the South Eastern part of France, just beyond « La Côte d'Azur », in the Verdon Gorges. The rally was organized at a “Countryside day”, during a village festival with local products. The owners of the cars took some patients and participants for a drive on the Cretes Route. The organiser of the car rally is a family member of Philippe (at Ironman Nice) who had PH since 2004 and underwent double lung transplant last winter. Philippe and his wife are very engaged in HTAPFrance as they say they got really precious support during those difficult times. For the first time in this village 80 prestigious vehicles were gathered.

Later on this year, we had two unusual projects I want to tell you about.

1st HTAP FRANCE WORKSHOPS,
4th OF OCTOBER

For many years we have been observing differences in the follow-up protocols between “competence centers” and it has been concerning us a lot, as a source of anxiety for patients not being sure they are in the right place to take care for their PH. After discussions with the experts, we understood that, in order to ensure the best care everywhere in France we should have the different teams to discuss all together. So for the first time this year, we took advantage of having already a venue booked for our Patients' Congress, to plan on the day before a workshop on PH care in France. All the competence centers were invited, and of the 23 centers, 17 were present. Ideally, they would have sent a physician, a member of the team, and a patient. In most cases we had a doctor and a patient. 40 people were present. We had decided to do a short presentation in which we would explain the objectives, and then address 6 different topics. The subjects we had selected, and had been agreed by Prof. Gérard Simonneau, were:

- PAH and pregnancy.
- When to prescribe prostacyclin?
- How to prevent complications on a permanent catheter?
- PAH patients in the emergency room: how to “signalize” them?
- CTEPH: screening and treating them.
- The use of right catheterization.

It was a very positive experience, which has helped us understand the differences and have a real discussion on the topics. We managed to get agreements on some points but agreed that most topics would give us work for a few years before harmonization of the practices! The most satisfying aspect is that everyone present at the meeting showed a real will to cooperate.

2nd PH PATIENTS' CONGRESS IN LYON,
5-6th OCTOBER

Right after the HTAPFrance workshops, we welcomed 210 people from France, including from the overseas island “La Réunion” and from Switzerland (Lydia and Monika came with 12 people). Patients, families, friends, doctors, nurses, psychologists... In the course of the two days, in five different meeting rooms, there were 30 sessions on various subjects. From the basics of PH to intimacy issues, from PAH and scleroderma to transition from pediatrics to adults departments Very practical questions of everyday life were put forward and answered by top experts in PH and by professionals such as social workers, psychologists, nurses... The participants did really learn a lot, and met other patients or carers, made friends, and took home many tips and positive energy.

Mélanie Gallant-Dewavrin, HTAP France
www.htapfrance.com
<https://www.facebook.com/pages/HTAP-FRANCE/288261994529082>





GERMANY

FAMILIES WITH CHILDREN SUFFERING FROM PH MEET AT THE AMMERSEE

Families with children suffering from a rare disease often have the problem of not finding other families to exchange information and experiences. The families may feel isolated and alone and need emotional support. The phev association organized the first family reunion for families with children suffering from pulmonary hypertension in Diessen on April 5-7. The families had the opportunity to meet other families affected by the disease, to learn and exchange ideas with each other. Dr. Christian Apitz from Giessen and Dr. Alfred Hager from Munich presented recent developments in research and treatment. A lecture on resilience was made by the psychologist Kathrin Veller and there was a workshop with the psychologist Christina Sokol, Hildesheim and Dr. Hoppenworth, Osnabrück. A very enjoyable boat trip on Lake Ammersee strengthened the sense of community and at the end everyone agreed that such meetings should be held again!

**16TH ANNUAL MEETING OF PHEV**

The 16TH ANNUAL PH PATIENT MEETING was held in Frankfurt am Main on October 25-26-27, and was attended by a total of approximately 250 patients, their families and other interested parties. The event was opened by the Deputy Chairman of the association, Günther Thimm, who represented the Chairman, Hans-Dieter Kulla, absent for family reasons. The meeting was held under the patronage of the former Social Minister, Dr. Erwin Vetter. The focus of the presentations at the Annual PH patients meeting were results of the 5th WORLD SYMPOSIUM ON PULMONARY HYPERTENSION held in Nice on March 27-1st of March 2013. Several speakers addressed this issue and reported on the main outcomes of the World Symposium. Apart from the lectures, numerous workshops were also on the program. Topics ranged from malnutrition in PH to breathing and inhalation techniques, Tai Chi, progressive muscle relaxation and autogenic training, medical technology in PH, rehabilitation, patients' rights, the impact of chronic illness on the family and partnership. The program allowed ample opportunities for personal interactions and exchange of experiences.

New treatments for PAH

Professor Ardeshir Ghofrani, from the LUNG CENTER OF THE UNIVERSITY HOSPITAL in Giessen and Marburg, reported on the new treatment opportunities for targeted PH therapy presented in Nice. All drugs developed up to now are vasodilators, which aim at relaxing the constricted vessels. Drugs against the vessel wall thickening and the uncontrolled

growth of cells in the blood vessels are not yet available, said Dr. Ghofrani. Imatinib, an antiproliferative agent that is used in the treatment of tumors, has not been approved by the regulatory authorities due to high risks for PH therapy, the principle has proved to be effective and further studies will be undertaken on this pathway. New drugs discussed in Nice also included Macitentan, a vasodilating agent an "endothelin receptor antagonists", and Riociguat, a stimulator of the soluble guanylate cyclase (sGC). Both drugs are now approved in some countries and approval in Europe is expected to take place in the near future. Professor Ghofrani presented the results of recent studies on these drugs. In this context Prof. Ghofrani also strongly urged patients to participate in clinical trials. As meta-analyses clearly demonstrate, participation in studies improves the prognosis since all those who take part

are supervised closely by specialists at PH centers. In addition, clinical studies are vital for any future progress on PH therapies.

Paradigm shift in the exercise training therapy

Prof. Ekkehard Grünig, Head of the Pulmonary Hypertension Center of the CHEST CLINIC AT THE UNIVERSITY HOSPITAL of Heidelberg, reported on a new study on exercise training therapy in PH. This study shows that exercise training can increase the contractile capacity of the heart, improve walking distance, oxygen uptake and the quality of life of patients. This is also demonstrated in a new training study with 90 patients in Heidelberg. The German association phev warmly thanked Professor Grünig for supporting multiple training projects. Reporting on further results from the Nice symposium, Prof. Grünig spoke of the need for more careful observation of patients with borderline elevated pulmonary arterial pressure - the so-called borderline PH - and a better standardization of the right heart catheter examination. Professor Grünig also recommended as supportive measures for PAH patients, amongst others, the coordinated use of diuretics, anticoagulants, possibly - long-term oxygen therapy, the control the level of iron and ferritin, regular flu shots, supervised rehabilitation and psychosocial support.

Surgery for chronic thromboembolic pulmonary hypertension (CTEPH)

News on CTEPH from the Nice symposium were represented by Prof. Heinrike Wilkens of the Department of Internal Medicine V - Pneumology, Allergology, Respiratory and Environmental Medicine at the University Hospital of the Saarland. If there is a diagnosis of CTEPH pulmonary endarterectomy (PEA) is the treatment of choice. However, it should be carried out at a surgical center which has extensive experience with this operation. Determining the operability of the patient depends on the experience of the surgeon and is critical. If the patient is inoperable, or pulmonary hypertension recurs after surgery, a therapy for PAH drugs is indicated. This, however, requires further studies.

Looking across the border

Dr. Sven Günther, who has worked at the National Ref-

erence Center for Pulmonary Hypertension in the center for PH at the Hôpital Bicêtre in Paris, reported that a significantly higher number of patients are being treated with prostacyclins (in the form of a long term intravenous infusion) in France with respect to Germany. In addition, Günther stressed the similarities of the PH management on both sides of the Rhine and advocated to pool expertise and experience.

Journalism Prize for FACT

The € 3000 Journalist Award this year went to Andreas Rummel for his contribution on the "Dramatic consequences of the organ donation scandal" for the German magazine "FACT". Rummel spoke of the problem of lack of donors and shortage of organs. In a short video Rummel spoke of a 33-year old patient with PH who is on the waiting list at the University Hospital of Jena for heart and a lung transplant. If organs are not available soon she will die. At the award ceremony in Frankfurt he brought some good news, the PH patient has now received the much needed transplant and is regaining her energies slowly but steadily. Special thanks go to the main sponsors of the event: Actelion Pharmaceuticals Germany GmbH, Bayer Vital GmbH and Pfizer Pharma GmbH, and to the other sponsors GlaxoSmithKline GmbH & Co. KG und Techniker Krankenkasse Hamburg. Furthermore, the supply of oxygen at the meeting was made possible by the Vereins der GTI medicare GmbH and VIVISOL Germany GmbH, a very big thank goes to them as well.

IRONMAN

Phev volunteers were present and helped man the booth, distribute materials and talk with visitors at the Ironman race in Berlin, held as part of the World PH Day celebrations (see photo below on the right hand side with cheque for their important contribution to the campaign). The PHA Europe collaboration with Ironman was held in five other countries: Austria, France, Norway and Switzerland.

Hans -Dieter Kulla , pulmonary hypertension e.V.

www.phev.de

<https://www.facebook.com/pages/Lungenhochdruck-Deutschland/386143468153378>



From left: Günther Thimm, Prof. Ekkehard Gruenig and the winner of the Journalist award, Andreas Rummel



Günther Thimm with Prof. Ardeshir Ghofrani



Berlin Ironman race



GREECE

A NEW PATIENT ASSOCIATION, PH GREECE!

The “Hellenic Pulmonary Hypertension (H.P.H.)” was founded in 2013 in order to achieve the dream of the current President, Mrs. Ioanna Alisandrato, which is to help patients with PH in the Greek territory, to inform them about the disease and to represent them in the international PH Community.

**WORLD PH DAY**

Our first event, “Get Breathless for PH”, took place in May 2013 (World PH day), and consisted in a big cycling event in Athens. This activity was such a great success, with very high participation, wide media coverage and the presence of the Ministry of Health, that we were awarded a prize by PHA Europe for our effort. During World PH Day

we also organized a press conference, during which the President of H.P.H. and two distinguished medical experts not only answered questions about the disease, but also pointed out, among other things, to the need for early diagnosis and the need to inform the general public about PH.

PATIENT AND CARER DIALOGUE TOOLS

The Greek association also participated in the PHA Europe project related to the follow up of the Patient and Carer Survey. It developed the two dialogue tools, “Living with PAH”, the two letters for health care professionals and the

executive summary with the title “The Impact of PAH”, all of them for the patients and the caregivers. These materials were translated into Greek, since our main goal was to raise awareness. More precisely, we wanted to hand these valuable tools out to every patient and caregiver, to provide better and more accurate information concerning the effects of this rare disease, to show people that they are not alone and to assure them that they are surrounded by a team which can support them. In order to accomplish these objectives, we first visited Greek Hospitals in Athens and in Thessaloniki, where we met doctors and asked them to help us in promoting these leaflets to patients and to caregivers. Afterwards, with the collaboration of numerous mayors, we set up kiosks in different municipalities, where, for a month, we gave out information and offered advice and help to anyone who came along to our booths.

CONFERENCE OF THE GREEK ALLIANCE OF RARE DISEASES

In October we participated at the Conference organized by the Greek Alliance of Rare Diseases, in Thessaloniki, where we had the opportunity to present to the rest of the Rare Dis-

eases Community the work already accomplished by our newly founded Association.

A “PATIENTS IN POWER” CONFERENCE

But our actions didn’t stop there. In November, our Organization took part in the 2nd Greek Patients’ Conference “Patients in Power”, in Athens, where we presented our work - for which we even received an award.

FACEBOOK, TV AND OTHER

For a whole month, every single day, we uploaded on the social media a photo with a rare purple flower and a message for PH, which was shared more than 700 times. We also took part in a weekly TV program about health, with the purpose to inform the public about our rare disease. And, following all these activities, our Organization succeeded in being registered in the Greek data base for rare disease patient’s organizations.

MEDICAL WORKSHOP

Finally, we participated in a medical seminar-workshop for PH, organized by specialized doctors from the Onasio Cardiac Surgery Centre (O.C.S.C.), during which Cardiologists, Pulmonologists and other doctors were informed about the disease, in order to be able to identify immediately its symptoms, and therefore give an early diagnosis.

Through all these initiatives, which have been very constructive and were backed by every single person who participated in them, not only did we have the opportunity to raise awareness among doctors about the severe consequences of pulmonary hypertension at every level of our lives, to have them at our side, to give the chance to patients and to caregivers to have a better quality of life thanks to the leaflets we



distributed and to assure a continuous collaboration with municipal authorities, but we also had the opportunity to find new patients who sought help from us and to open up the path to spread more awareness. We cannot wait to see what our next steps will be!

Ioanna Alisandrato, PHA Greece

www.hellenicpulmonaryhypertension.gr

www.facebook.com/HellenicPulmonaryHypertension



Ioanna Alisandrato receives 2nd prize for best World PH Day campaign





HUNGARY

NATIONAL PAH PATIENT MEETING

The 3rd national patient meeting was organized by Tüdőér Egylet (PHA Hungary) on November 9, 2013. More than 140 patients, physicians and other stakeholders participated.

The Hungarian PH association is a young organization founded in 2008 by Kristof Karlocai (MD, expert of PH) and 40 patients, including Eszter Csabuda, President of Tüdőér Egylet. Eszter was properly diagnosed and put on therapy only after 8 years of misdiagnosis. The founding of the association was strongly supported by PHA Europe.

The association celebrated its 5th anniversary this year and Eszter emphasized that the goals of the association, which remain the same, are complex: to give hope, to show alternatives, to demonstrate power to tackle with the disease day by day and to fight for access to drugs which are already on the market for the Hungarian patients. It is also important to help with psychosocial support and to raise awareness of the disease, because patients not being treated may pass away within 3-5 years. 170 patients are diagnosed in Hungary and there could probably be other 250 undiagnosed patients across the country.

The first topic was the general surgical risk and specific pre- and postoperative considerations in PAH. Kristof Karlocai, MD, Vice President of Tüdőér Egylet, physician at the Semmelweis University, Department of Pulmonology, informed the audience about the risks of surgery and how such risks can be minimized in PAH. Mr. Karlocai emphasized that proper planning is critical and it is highly advisable to involve PAH experts in such planning. Tamás Forster, professor of the II. Medical Clinic and Centre of Cardiology gave insight to the proper, post-surgery treatments of PAH patients.

The second topic was the experience of PAH coordinators. Gyöngyi Máté, pediatric PAH coordinator of Gottsegen György Hungarian Institute of Cardiology, demonstrated with lot of photos how the Centre of Pediatrics tried to make the disease easier for children. Edit Takács, adult PAH coordinator of the same institution shared the experience with adult congenital (GUCH) PAH patients.

The third topic was the financial aspects of PAH therapy. Márta Szegedi, MD, representative of National Health Insurance Fund, held a lecture about the financial impact of PAH drugs



and treatments. She presented detailed data on how much money is reimbursed regarding the treatments. She also gave information about different centres and type of applications.

In the last part of the meeting two guests speakers presented their experience from different European countries. Alma Rubio, representative of Actelion, highlighted the most important ongoing projects, including World PH Day, the Ironman European Tour and the International Patient and Carer Survey. She stressed that the attitude of the pharmaceutical industry had dramatically changed in past years and that it fully realizes the importance of patient groups.

Gerry Fischer, President of PHA Europe, also attended the meeting and spoke about the current European situation, tasks, purposes and cooperation between national organizations. He also introduced the main projects and outlined PHA Europe's main initiatives. He closed his presentation with a short emotional video about his young daughter, who suffers from PAH.

Eszter Csabuda, Tüdőér Egylet

www.tudoer.hu

[https://www.facebook.com/pages/Tüdőér-](https://www.facebook.com/pages/Tüdőér-Egylet/151123348280359)

[Egylet/151123348280359](https://www.facebook.com/pages/Tüdőér-Egylet/151123348280359)



Prof. Tamás Forster, MD



Eszter Csabuda



Prof. Kristof Karlocai



From left: Gerald Fischer, Eszter Csabuda, Alma Rubio, Márta Szegedi and Gergely Meszaros



IRELAND

ANNUAL MEETING FOR PATIENTS AND THEIR FAMILIES IN DUBLIN

The annual Pulmonary Hypertension meeting for patients and their families was held on 27 April 2013 in Dublin. Experts in their field addressed a packed audience on how patient groups can use their collective voice to inform health policies and improve the lot of the PH patient overall.

In his opening presentation Prof. Sean Gaine from the Mater University Hospital stated that PH is different in everyone – “Sometimes it can be the only disorder a person is suffering with, while in other cases, there may be other comorbidities involved”. Prof. Gaine provided a clinical overview of the nature of PH, the symptoms and the delays in diagnosis. Mr Iain Armstrong from PH UK opened his presentation with a call to PH patients in Ireland to use their experiences to make their collective voice heard – “I passionately believe that patients are an under-used resource” he said. He went on to discuss that PH can mean an uncertain future for patients. Mr Armstrong presented data from an ImPAHct study into pul-

monary arterial hypertension (PAH) and its impact on aspects of the sufferers’ quality of life rather than the clinical disease. A diagnosis of PH means an uncertain future, he said. There is also a financial impact, as 75% of people in the study said the illness had a significant impact on their ability to work. If you have money worries at the same time as having this illness, this can have a major impact on quality of life. Every patient has their own unique story and their voice is crucial. Mr Armstrong invited the attendees to consider and reflect on how they felt when they were diagnosed - many employers and indeed insurance companies do not understand what it means when a patient has been diagnosed with PH. This presentation was followed by a lively and information Q&A session.

Dr Noel McCaffrey from Dublin City University began his talk by stating that there is not a single illness where exercise cannot play a therapeutic role. “One of the benefits of engaging in an exercise programme is that it limits isolation because people go to their exercise classes and meet

others there”. He stressed that exercise can greatly improve quality of life - “if you have reduced strength - you will have trouble doing certain tasks”. Dr McCaffrey presented slides showing that no matter what age the patient is, the rate of physical and cognitive decline can be slowed by the patient becoming involved in a structured exercise programme. One of the things that stops PH patients exercising is that their heart and lungs are often incapable of supporting 5 minutes of exercise, this can become frustrating for them so we break that down into five one minute sessions to make it less stressful for the patient. Sometimes patients say “Doctor I am not getting any better” but it is important for patients to note that halting the decline in any disease is a very positive outcome.

Ms Karen Redmond, Cardiothoracic Surgeon in the Mater University Hospital, Dublin expressed her special interest in PH and outlines new techniques in transplantation. She said “Organs are a bit like engines; sometimes they just need to be tweaked and sometimes they just have to be replaced”.

Ms Redmond discussed the transplantation process beginning with “the gift of a donation - which is a gift of life”. Often grieving families can take great comfort from knowing that they have done something to give life to another person and this is reinforced when they receive thank you letters from the recipients. She stressed that “transplants are only necessary in a small amount of patients and there can be contraindications in patients who may have cancer or cardiovascular disease”. She also spoke about improvement in the quality of life when organ transplantation is successful. However concerns of rejection and/or infection can never be ruled out.

Following the meeting, a report of the event appeared in one of the medical journals, the journalist stated that... “one interesting aspect of the Patient Meeting involved four empty chairs. Rather than taking their seats on the stage behind a table, all of the speakers opted to sit with the PH patients and their families as they listened to each other’s presentation and fielded questions. This epitomised the spirit of the event - patients, their families and the medical professionals together in the drive to improve the lives of those with Pulmonary Hypertension”.

This was a very apt description of the day and feedback was very positive. The 2014 meeting for patients and families is scheduled for Saturday, 10 May.

ALL IRELAND SCIENTIFIC PH MEETING

On the 28th of October an important scientific meeting on PH was organized in Dublin, at the Royal College of Physicians, with leading Irish PH physicians as speakers. The meeting was well attended and very successful.

Regina Prenderville, PHA Ireland

<http://www.pulmonaryhypertension.ie/pha-ireland/>



Prof. Sean Gaine, Ireland



From left: Regina Prenderville and Iain Armstrong





ISRAEL

**12TH ANNUAL PATIENT MEETING**

PHA Israel held its twelfth annual conference in early October at Kibbutz Nir Etzion, located on the slopes of Mt. Carmel, south of the city of Haifa. Israel PHA chair, Atty. Joni Berg and executive director Aryeh Cooperman began the conference at 17:00 on Oct. 9th by welcoming those in attendance and surveying the activities that had taken place over the past year.

A Community of Hope, Expert Panel

Board member Yosef Gotlieb then made a presentation on "Building a Community of Hope," describing the international PH community, including relations with other national PH associations, the World Symposium on Pulmonary Hypertension (Nice, France, February 2013) and the 2013 Conference of the European Pulmonary Hypertension Association (Casteldefells, Spain, September 11-15, 2013). A panel discussion by several of Israel's leading PH specialists followed. Chaired by Dr. Ben Fox of the Rabin Beilinson Medical Center in Petach Tikva, the other panelists included: Prof. Neville Berkman of the Hadassah University Medical Center in Jerusalem, who spoke on new research and treatment protocols; Dr. Yochai Adir of the Carmel Medical Center in Haifa, who addressed combinational therapy in PH treatment, and; Dr. Mike Siegel of the Sheba-Tel HaShomer Medical Center, whose talk dealt with the genetic basis of PH. The experts then addressed questions and individual concerns asked by conference participants.

Patients Rights

Following dinner, two evening sessions were conducted. Aryeh Cooperman spoke on "Patient Rights and the National Insurance Institute (NII)," which was followed by a talk on legal aspects of the relationship between patients and insurers given by Atty. Sharon Peres Ben-Haim, an expert in the field.

Workshops

During the second day of the conference (Oct. 10), four workshops over two consecutive sessions were offered. During

the first session, workshops on listening and communication for couples and a second one on glassmaking were conducted. During the second session, demonstrations of Qigong and vegetable sculpting were presented.

On the basis of feedback received, the conference admirably succeeded in updating the participants on treatment and patients rights, in networking patients and their families with other people who have PH, and in offering patients and families a pleasant few days away from their daily routines spent in the company of other members of the Israel PH community.

TREATMENT INFORMATION DAYS, YAD SHIRA (SHIRA FAIR)

PHA Israel has planned a number of information days regarding specific treatments. Individuals who are treated with iloprost (Ventavis) and their carers have been invited to a special meeting that will take place in January 2014 in the city of Netanya. Future events of this type are planned for the upcoming period with respect to other therapies.

The Board of the Israel PH Association has already begun preparation for the third annual Yad Shira, a "happening" dedicated to the memory of Shira Dinur, a young woman who passed away several years ago from PH-related complications. Since then, friends of Shira and the Israeli PH Association have sponsored special events, the first being an outdoor fair and the second, last year, consisting of a cocktail reception. The forthcoming event in March 2014 will encompass performances by prominent musicians and a standup comedian, a book launch of the Hebrew translation of a guide to PH prepared by the US Pulmonary Hypertension Association and published with the support funds raised by the previous Yad Shira events, and a fundraising lottery.

Yosef Gotlieb, PHA Israel

www.phisrael.org.il - <http://on.fb.me/1bPDL5v>



ITALY-AIPI

ANNUAL PATIENT MEETING

On the 1st of April 2013 AIPI held its 10th annual assembly, which was attended by over 150 patients and family members. The agenda included a medical update by Prof. Nazzareno Galiè, a presentation on recent developments in disability legislation in Italy and the annual report of activities. Elections for the new Board were held and Pisana Ferrari was confirmed as President (third mandate). Lunch was followed by live music and entertainment (with famous TV artist Claudio Serra), time for socializing and the traditional raffle.

NEW PUBLICATIONS

Since the beginning of the year four issues of our quarterly magazine *AIPInews* have been issued. We have also translated and printed the patient and carer booklets developed in the aftermath of the International Patient and Carer Survey on the broader impact of PAH on patients.

Finally, we have produced an illustrated booklet for PH patient and carers called "Tips for every day life". All these resources can be downloaded directly from the AIPI website.

**AIPI CELEBRITY CALENDER FOR 2014**

In the course of 2013 AIPI was very fortunate to be included in an artistic project by one of its members and supporters, celebrity photographer Gianluigi di Napoli. Gianluigi has a passion also for circuses and circus life, has been the official photographer of the "Cirque du Soleil", has published books about the circus and is very good friends with the most famous clown in the world, David Larible. He created a project called "MASKS", with the idea of engaging celebrities to wear and be photographed with clown make up and decided to dedicate his project to raising awareness and funds for PH and for AIPI. Twelve very famous Italian female icons took part MASKS: Olympic swimmer Federica Pellegrini, actresses Maria Grazia Cucinotta, Valeria Solarino and Carolina Crescentini, the étoile dancer of La Scala Petra Conti and many



more. The clown make up was designed and executed by David Larible. The celebrities also posed for Gianluigi without clown make up and some beautiful “diptychs” were thus realised. The best photos were exhibited twice, once in Milano at the MIA, Milano International Art Fair, one of the most important photographic exhibition in the world, and in Bologna, on the occasion of an international PH conference. Twenty-four of these diptychs were selected to be part of the 2014 calender for AIPI. Promotion activities and press conferences held in Milano and Bologna resulted in great media coverage and interest of the general public. The calendars for

2014 are available for free on request, please write to marzia.predieri.1@gmail.com.

EXCURSIONS

This year's AIPI annual excursion was to the beautiful city of Cagliari, in the island of Sardinia. They participants spent three days visiting the island's historical sites and nature as well as enjoying its great food!

Pisana Ferrari, AIPI Italy

www.apiitalia.it

<http://www.facebook.com/AIPIItalia?ref=ts&fref=ts>



Tor des Géants is famous to be one of the hardest of the world, never less than 3-4000 meters of altitude above sea level.

“Scocciapilaccia”

This year the “Butteri of Vejano” wanted to dedicate their annual gathering to PH patients. They competed on horseback in the “scocciapilaccia” (breaking the pot) traditional game and offered us a very pleasant day in their company in a beautiful country spot. We were offered a picnic and the proceeds from a raffle.

Karate international competition

Our young testimonial Carolina Amato, the Italian Karate champion, is still wearing our colours in national and international competitions...and winning her gold medals.

IPHNET NETWORK LAUNCH!

On November the 29th all the members of our Scientific Committee met in Rome during the workshop “Pulmonary hypertension: the network as a tool to correct diagnosis and treatment”. The iPHNET network is today a reality. Many PH Centres of Excellence will be able to work together and cooperate with other Italian hospitals. Only a few years ago this achievement was only a dream, now we are already beginning to see the hoped-for good results. We are sure to see much more in the near future. 25 hospitals from all over Italy were represented. In each of them PH patients are being treated, it is absolutely important that the doctors cooperate with the PH Expert Centers.



ITALY-AMIP



SPORTS EVENTS

As usual our association activities were many and many of them had the double purpose of spreading awareness about PH and fundraising to sustain our projects. We were lucky to have many friends who offered their help. A special mention in this respect goes to the runners we made friends with during last year's Rome Marathon: many of them are still running for us! Maurizio and Francesco were the first ones, today there are dozens and all feel very involved and aware of doing something important for PH!



Tagliacozzo run, Sept 8

Before the start the athletes blew soap bubbles in the air. The entire proceeds from the registrations was donated to AMIP.

Tor des Géants, the Endurance Trail in the Alps

Stefano Corrado ran for us to the finish. It was a real challenge, done to witness how hard it is to breathe for PH patients. The



Prof. Dario Vizza, Italy

PARTICIPATION AT SCIENTIFIC MEETINGS

A very important part of AMIP's job is to bring the voice of PH patients to the institutions and to the medical world. We did this the whole year round, by taking part in meetings and workshops where rare diseases were being discussed. Our President, Vittorio, was invited and able to bring the PH patients' point of view in presentations during several events in Rome, Capri (2nd Focus on PH, May 3/4, a meeting where major international experts were present), Bari, Florence etc.



POPE FRANCIS

We would like to finish this article with nice news: to thank Pope Francis for the words of encouragement he spoke during last World PH Day, Vittorio found the way to send him a copy of the “Mariposa Journal”, one of AMIP's newsletter “Insieme” and a very special membership card of our association. He must have welcomed the gifts because he sent back a Special Blessing. Vittorio wanted to share this with all of you!

Luisa Sciacca della Scala, AMIP Italy

www.assoamip.net

<http://www.facebook.com/AssociazioneMalatiDiIpertensionePolmonare?ref=ts&fref=ts>



LATVIA

GRANT OF €20.000 FOR THE OXYGEN PROJECT

PHA Latvia won a grant of €20,000 euros with a project called "Heartbreathe" in the social projects' contest organized by the Charity fund "Ziedot" and the State Stock Company "Latvia's State Forests". Now our organization has 15 stationary oxygen concentrators and 3 portable concentrators for PH patients in Latvia. Home care oxygen therapy doesn't receive reimbursement from the health care system in Latvia and this work lies entirely on patients' shoulders.

EVENT RARE AND FRIENDLY

PHA Latvia, in association with Rare Disease Society "Caladrius" and The Centre for Disease Prevention and Control" (CDPC) of Latvia, organized a Rare Disease Day 2013 event called "Rare and Friendly", on the 27th February, in Riga. We spent the day in a warm and cheerful atmosphere. To support rare disease patients, especially kids, popular personalities of Latvia - singers Intars Busulis, Jenny May and hockey club "DINAMO RĪGA" player Arvids Rekis joined a photo session

with us and our friends from other rare disease patient organizations of Latvia. Opening the event, World ashihara-karate champion Vitaly Bereznev said that he had to win a great combat in the World championship to achieve the champion title, but for patients with rare diseases every day is like a struggle, because it is a struggle for life. After the photo session "We are friendly" a press conference took place ("We talk"). The event caught media attention - TV3 news, daily newspapers "Diena" and "Neatkarīgā Rīta Avīze", radio SWHPlus, and specialized internet portals and others - who all talked about "Rare and Friendly".

CHARITY SPORTS GAMES AT WPHD 2013

The Charity Sports Games on 5th May in Salaspils near Riga attracted great interest and there were 224 participants ready to "Get breathless for PH" on the sunny spring day - 150 children and 74 adults. The National TV channel LTV1 featured a story about our Charity Sports Games on Daily News. There was an interview with Ieva Plume, Dr. Ainars Rudzitis and Dr. Andris Skride, PH experts in Latvia, and with Zane Lazdina, one of the PH patient who needs lung transplantation. The main target of the Charity Sport Games was fundraising for three PH patients in Latvia - Zane (31), Jurijs (27), and Alexander (30).

HEART HEALTH CAMP

The Heart Health Camp 2013 of PHA Latvia was held from the 3rd to the 6th of June in Saulkrasti, on the Baltic Sea. There were 39 participants from 3 rare disease associations of Latvia: PHA Latvia, the Cystic Fibrosis Association of Latvia, the Rare disease association "Caladrius". Participants of the camp took part in a visual art workshop and a flower decoration workshop. We also had a relaxation program with a yoga specialist and we did exercises in the mornings. On the last camp day we visited the Museum of Munchausen in Dunte. At the camp there was a program for children with leisure time activities, sport games, and lessons based on the Montessori learning method. We met new friends and



have learned new hobbies. More information is available on the PHA Latvia FB page.

LOBBY WORK

PHA Latvia is continuing to work on improving the health care system for PH patients and other rare disease patients in Latvia. We are part of the Pharmacy consultation Committee and the National Plan for Rare Diseases work group of the Ministry of Health. We have repeatedly turned to the Prime Minister of Latvia and to the Social Committee of the Parliament of Latvia with a requests to change the discriminative and limited system of drug reimbursement for rare disease patients in Latvia and we have made a request for transplantation possibilities for PH patients abroad, as well as other important issues. The National Plan for Rare Diseases 2013-2016 has been adopted by the Ministry of Health but the plan does not solve the main problems of rare disease patients, including those of PH patients in Latvia.

Ieva Plume will participate in the international conference "National activities related to Rare Diseases" on the 14th of November, 2013 in Vilnius, Lithuania.

OTHER PROJECTS

- PHA Latvia has been part of the international patient and carer survey's project, for which it has translated and printed the survey materials.
- PHA Latvia is a participant in the PHA Europe "Fellowship" program
- This year PHA Latvia supported the first pulmonary endarterectomy in Latvia.
- PHA Latvia is the main partner of the project "Development of Health NGO's Network in Latvia" for a Patients' Ombud Office of Latvia to be achieved with European Union funds.

Ieva Plume, PHA Latvia

www.phalatvia.lv/en/ - <http://on.fb.me/Vx0udp>



NETHERLANDS

CARE FOR LUNGS

In June 2013 Riet van der Ekart organised a big event in the south of The Netherlands, in Schijndel. Riet is a PH patient. The event was called: "24 Hours Care for Lungs". With the help of many volunteers she organised a wonderful day and a wonderful night. Many teams walked 24 hours for PH and there were many other activities, such as a football competition. The result: 60.000 euro! The donation was given to the medical team of the Vumc in Amsterdam. The donation will be used to finance scientific research on hereditary PH. Prof. Dr. Anton Vonk Noordegraaf was very pleased with this donation and this support for research.



From left: Prof. Dr. Anton Vonk Noordegraaf (Vumc Amsterdam), Riet van der Ekart (PH patient) and Ferdinand Bolsius (President PHA-NL)

In november 2013 Riet van der Ekart received a nomination from the Vumc for this event: "24 Hours Care for Lungs".

Leny van der Steen, PHA Nederland

www.pha-nl.nl - <http://on.fb.me/TFBsJT>
www.hebhartvoorlongen.nl

NORWAY

PH PATIENT MEETING

PHA Norway held a patient meeting in March. It was a small group of 20 people, and we had a very nice two and a half day conference at the Feiring rehab clinic north of Oslo. The last half day was dedicated to discussions about the patient and carer survey. I presented the survey and it's main findings. Afterwards everyone filled out the appropriate dialogue tool. We used this as a basis for group work. I divided the attendees into two groups: patients and carers. Since we had two small groups of 10 people, everyone was able to contribute to the discussions. It was especially beneficial for the carers to also discuss how the disease has affected them. We then had one person from each group present the results of the discussion in plenum.

The discussions confirmed that most findings in the survey coincide with conditions here in Norway. However, it appears that families in Norway are not as affected economically as many other places in Europe, as we have a very good welfare system in Norway. Furthermore, when it comes to the loss of libido issue, it was hard to conclude. It seems that this is an issue even people in liberated Norway find hard to discuss with others in a group discussion setting. However, couples will probably raise this issue when they are on their own.

The main conclusions from the discussions were that families often had a very hard time before a diagnosis is given. Especially when the patient was undiagnosed and some doctors found it hard to believe that they were sick. When a diagnosis was given, it often came as a relief even if the



diagnosis was serious. It actually seemed to strengthen the couples' relationships. Families also noted that priorities are different after the diagnosis. For instance, family and friends become more important and housework less important. People also expressed that they have learned to live in the moment.

BAYER NURSE WORKSHOP, STOCKHOLM

The annual Bayer Nurse workshop took place in Stockholm on June 7 and 8, 2013, attended by 140 nurses from 18 countries, working in the field of PH. Patrik Hassel, from PHA Sweden and myself, were invited to address the participants. I used this opportunity to present PHA Europe and its activities. I spoke about the aims of the umbrella organization and how PHA Europe supports the national associations and strives to achieve the best possible standards of care for all European PH patients. Patrik Hassel, who is father to a young child with PH, spoke of his family's personal experiences with the disease.

Hall Skåra, PH Norway
www.pha-no.com
<http://on.fb.me/TDzyKI>

Hall Skåra speaking in Stockholm at the nurse workshop



A TELEPHONE HELP LINE FOR PH PATIENTS AND THEIR CARERS LAUNCHED IN POLAND

Both telephone number and E-mail addresses are on the PHA Polska website and on the association's leaflets and brochures. All questions and inquiries are being answered by experienced patients with a long PH history.

PHA POLAND AT "EFFECTIVE COOPERATION WITH THE MEDIA" TRAINING

In July 2013, PHA Polska participated in the media training meeting for patients' associations organized by Bayer. The training included, among others, the following topics: communication, media relations, PR tools, legal aspects of external communication in pharmaceutical companies.

PHA POLAND AT "WHITE SATURDAY" IN EUROPEAN HEALTH CENTRE

On July 6, PHA Poland participated in the "White Saturday" event organized in the European Health Centre in Otwock. PHA Poland had its own booth with educational and promotional materials on PH.

EXPERIENCE THE "BETTER LIFE" - BAYER 150 ANNIVERSARY EXHIBITION

At the end of July 2013, PHA Poland representatives took part in the fascinating exhibition organized by Bayer to celebrate the 150th anniversary of the company. An interactive exhibition showed how the company improved the quality of life for people around the world.

EUROPLAN CONFERENCE

On September 27-28 PHA Polska participated in the Europlan Conference (European Project for Rare Diseases National Plans Development). On the first day the Polish situation with respect to Europlan was presented. The EU, together with EURORDIS, have rated Poland's engagement in the treatment and care of rare disease patients and execution of European Committee's recommendations as regards the preparation of a Rare Diseases National Plan. During the second day the conference participants discussed the importance of rare diseases treatment organization within the health care system. PHA Polska was represented by: Alicja Morze, Barbara Bieniasz, Mirosław Zieliński the President of the National Forum for the Therapy of Rare Diseases - ORPHAN

REGIONAL PATIENTS' MEETING

On October 19, PHA Polska, together with the Cardiology Clinic of the Medical University of Lodz, organized a one day patient meeting. Doctors, nurses and patients met on that day to learn more about the PH and PHA Polska's activities. PHA Polska's activities were presented by Alicja Morze.

Prof. Marcin Kurzyna, from the European Health Centre, presented some of the results from the international patient and carer survey and talked about the impact of PAH on the lives of patients and carers. At the end he also explained how the treatment and care system of PH patients is organized in Poland. Dr. Łukasz Chrzanowski from Cardiology Clinic of the Medical University of Lodz spoke about the diagnostics and treatment of PH.

Both patients and their carers were very satisfied with the meeting's programme. They were glad that they could speak with the medical experts about their disease and meet other patients who experience similar issues and problems.



AGNIESZKA BARTOSIEWICZ - PHA POLSKA IN NEWSWEEK POLSKA

Agnieszka Bartosiewicz was interviewed by Newsweek (national weekly magazine, October 28, 2013) and talked about her experience with PH. Prof. Adam Torbicki and Prof. Marcin Kurzyna from European Health Centre described the medical aspects of the disease.

CONFERENCE OF PULMONARY CIRCULATION SECTION OF THE POLISH CARDIAC SOCIETY

The Conference of the Pulmonary Circulation Section of the Polish Cardiac Society took place on November 8-9 in



From left: Dr. Grzegorz Kopeć, Agnieszka Bartosiewicz PHA Polska, Alicja Morze PHA Polska, Prof. Adam Torbicki

Białystok. There were hundreds of medical experts and guests who discussed on the latest achievements in pulmonary circulation. PHA Polska was represented by Agnieszka Bartosiewicz and Alicja Morze. The association had its stand there. The conference provided a great opportunity to meet and talk with the medical experts.

The conference participants were able to learn more about PHA Polska activities and read the abstract of some the results from the international patient and carer survey.

There were also many copies of the booklets for PH patients and carers with the international survey results available. Link below:

<http://www.tvp.pl/bialystok/edukacja/recepta-na-zdrowie/wideo/14112013/13010878>

Agnieszka Bartosiewicz, PHA Polska
www.tetniczenadcsnienieplucne.pl
<http://on.fb.me/Rzeo0U>



PORTUGAL

This year the Pulmonary Hypertension Association of Portugal (PHAP) embarked on a “revitalization” of its activities, starting with the arrangement of a space that will function as an office and meeting room. It has also engaged volunteers in the legal field and social workers in order to improve a helpline service for its members and to promote better quality of life for patients and caregivers.

PHAP's main activity in 2013 was main activity was to defend the health care service for patients with pulmonary hypertension. Since this disease is not considered a chronic disease by the Portuguese law, PHAP alone and in association with other platforms has been fighting for the creation of a Chronic Patient Statute, having joined a committee that incorporated a petition put to the Portuguese Parliament.

The present economic and political situation of our country is difficult and the need to reduce costs and decrease the deficit has impacted severely on health services, motivating the refusal of certain hospitals to accept patients with PH who are not in their residence area. In this context we developed

a campaign together with social media and political parties which have led to a TV interview and a request for clarification in the Parliament, raising public awareness for this serious situation that may impair quality of life and life expectancy. PHAP has also made an investment in renewing and updating its Facebook page, in producing information brochures and a PH Patient kit that will be available soon.

In 2013 we had the support of Teresa Carvalho, thanks to the PHA Europe Fellowship program. Teresa is an important element in the dynamics of PHAP.

PHAP intends to develop a set of projects for 2014 including psychosocial support to the patients and the launch of a survey about the quality of life of patients. We hope to achieve our goals in 2014 and we hope to continue with the support of Teresa.

Maria Joao Saraiva
 Associação Portuguesa de Hipertensão Pulmonar
<http://on.fb.me/13sawnu>



REPUBLIC OF MACEDONIA



My name is Gjurgica Kjaeva. I come from a little town called Gevgelija, located in a small country in the center of the Balkan Peninsula, the Republic of Macedonia.

I was diagnosed with PH in 2012. My life has changed completely ever since my diagnosis. They say there is light at the end of the tunnel. Every day, I enter that long purple tunnel. I can see the light at the end of it. It seems close, it's getting blue, I try to reach it, but I can't. I try every day but I never seem to reach the light at the end of the tunnel. Big clouds of change, grief and difficulty overshadowed my life and changed it to the core.

Luckily I am married to a wonderful man - Ratko - who supports me. We have a nine-year-old son Boris, and my family is my biggest support in this life-threatening battle. It is very difficult to live with PH every day. I can't go out with friends, so they started abandoning me. I was suddenly reduced to living a life in bed, gasping for each breath, being unable to do any physically challenging task, or play with my child in the park, like everybody else. But in all darkness there is light, every yin has a yang so balance can be restored!

So when I was invited to the annual meeting of the European patient association, and I attended this event, my life started going into a new direction. I met all these wonderful people who were coping with this illness every day, just like me. We are tied in one inherent knot, connected with an eternal bond, in our struggle to get better, to find a cure, to live. I discovered my new PH family. I made so many new friends; I met so many extraordinary people who are fighting every day for their right to live, as a unity, as a whole. Being part of the PH family is the greatest gift, the biggest treasure that I gained from this hardship. I love my family PH and only together we can fight this.

I started thinking “what can I do for my PH family to make our lives better, what can I do to change the world”? During the event I had the opportunity to tell my story, “Mermaid in sky”, a story about my new, changed, life, how I felt and how I hoped for a chance for a better life with a happy end.

The GAM became a very important piece of the puzzle during my struggle with PH. It was a place where I regained hope and strength to continue on fighting. All these new friends gave

me the idea and showed me the path, so when I got back home I was ready to start a movement and make a difference.

After returning home, I immediately initiated the establishment of an association for people with PH under the name of “APH Moment plus”. On October 10, 2013, the association was officially established.

The first step was to start informing the people about Pulmonary Hypertension. There are many people still that have no idea that this disease exists. So I started where it was closest to me, at city I was born in, Gevgelija. I managed to get public support at a humanitarian concert which was held in the name of a young boy who lost his life in the fight against Leukemia.

I made a presentation, with the support of my cardiologist from Skopje, in order to inform citizens and the medical personnel of Gevgelija and introduce them to the seriousness of this illness, the symptoms, what the are medications and everything else which is connected to PH. Our performance was recorded by local TV stations and it was aired several times on television. When the people in the audience understood what this disease meant, how it was treated here in Macedonia, they weren't feeling indifferent anymore.

After this I was invited to talk with the mayor of Gevgelija, who promised his full support in helping us to spread the word about PH to health institutions in Macedonia, especially the Health Insurance fund of Macedonia and the Ministry of Health. And what especially touched me was that even one child made a presentation at his school for Pulmonary Hypertension, and I realized my message was beginning to get heard. I am a graphic designer so I started working on the information materials. I made the logo, flyers and with the help of many people, my PH family, I began the struggle for promotion, and hope that our voice will be heard and my work won't be for nothing. I met two other patients with PH, a 22-year-old girl with PH Together with the support of both of our families and our cardiologist, coordinator of Urgent Diagnostic Center “Philip II” Ass. Dr Ivan Milev, we succeeded to put the word out, and raise the issue on national television, our Macedonian Radio Television station.





Along with the organization for Rare Diseases “Life with challenges” and the organization for patients with HAE, we submitted a request for a public debate in the Parliament and requested the Public Health Committee to submit an amendment to ensure better medical care for rare diseases such as ours. Our battle is still ongoing at the moment, but we are not giving up. I would like to inform all competent medical doctors that they are the ones that can change the life of patients with PH, only if they know to recognize the symptoms and provide treatment for their patients. We have also scheduled meetings with the Health Insurance Representatives, the Minister of Health as well



as professor M.D. Kedev, who is chief of staff at the biggest clinic for cardiovascular diseases. We also have the support of the Special Hospital for Surgical Diseases “Philip II” as well as from the Academic and cardiac surgeon M.D. Zán Mitrev.

We have created a Facebook fan page which is growing in number every day, and currently has 650 friends and supporters all over the world, with 8.544 weekly total reach. The development of our new web page is in progress.

My biggest wish and hope for people suffering from this disease is to at least obtain basic rights to medical insurance, and, most importantly, get their medications. Access to treatment and medications is vital! We need to make people, especially doctors, familiar with the symptoms of this disease. I hope we can spread the word, inform the world and help those in need, because knowledge is power and health is the most important thing in life.

Gjurgica Kjaeva, President APH Moment
<http://on.fb.me/1kUSn5x>



SLOVAKIA

In the 2nd half of the year 2013, PHA Slovakia organized various interesting activities. During the summer months we were not so active, but we have more than compensated for this during the autumn.

REHABILITATIVE STAY, VYŠNÉ RUŽBACHY

From September 4 to 8 we organized a five-day rehab stay for 30 members of PHA Slovakia. One of the main objectives of the meeting was to teach patients a series of simple exercises to improve their health and wellbeing. We focused



on proper body posture, breathing exercises and stretching. The program was enhanced with balneotherapy and psychotherapeutic treatments. We also enjoyed doing some funny



interactive psychological games aimed at improving non-verbal communication. The whole meeting was accompanied by the exchange of experience between patients and discussions in smaller or larger groups, individual consultations and expert advice. There was very pleasant atmosphere among the patients during the stay.

XXXIV. CONFERENCE SSVPL – HIGH TATRAS

17.-19. October 2013

This year we had the opportunity to attend the first Conference of the Slovak Society of General Practitioners in the High Tatras on October 17-18 with our own stand. At the end of the conference Iveta Makovníková gave a speech for general practitioners to increase attention to early diagnosis of patients with PH. The leaders of the Slovak Society of General Practitioners expressed a wish to give a more space to the topic of PH on next year's congress.

PHOTO COMPETITION "THE BEAUTY OF THE MOMENT"

In early June of this year we announced a photo competition entitled "The beauty of the moment" for patients before and after organ transplantation. All associations with transplanted patients participated in this project. At the end of The deadline for sending photos was the end of October. The evaluation of the competition will be on 31st January of next year. The winning photographs will be shown in photo exhibitions all around the country in the ten biggest cities until mid-2015.

MOVIE: "WHEN LUNGS DON'T HELP"

At the beginning of the year we worked with the staff of public television STV on a 20-minute movie called "When the lungs do not help", which describes the problems of Daniel. Daniel is a patient with pulmonary hypertension. The first part of the film was premiered at the celebration of the World PH Day. In October, the film won the first prize in in the category of documentary films contest CIMES 2013 in Baden-Baden. The first part of the movie is about Daniel and his life before lung transplantation surgery and the second part (still to be developed) will document changes in his life after the surgery. The movie should be finished by the end of the year 2013. Its premiere is planned on January 2014 at the opening of the exhibition "The beauty of the moment" in Bratislava. In this film we want to point out the qualitative difference in the lives of patients before and after transplantation and thus support the idea of organ donation. The film, with English subtitles, will be posted on the RareConnect website.

OTHER ACTIVITIES

- Creating a professional advisory team, containing people from medical and healthcare professionals.
- Change of statute - extension of our activities, change of official name of the association in English language.

- Taking part in the PHA Europe General Assembly in Barcelona.
- Slovak Philharmonic Concert for disabled.
- Five workshops - with medical experts and representatives of industry in Slovak Alliance for Rare Diseases and two meetings regarding upcoming photo exhibition
- Publishing activities - 2 types of leaflets published, 2 photographic books about our activities and one poster for patients in PAH centres.
- Contributions to the Journal of the Slovak Alliance of Rare Diseases, called "Minorit".
- Article in the Dutch PH association magazine and also an article in the Public Health Insurance journal.
- Various posts on internet websites
<http://plnielanu.zoznam.sk/c/2270/zlodej-dychu-utoci-nestoji-za-vasimi-zdravotnymi-problemami-plucna-hypertenzia> and www.nusch.sk
- Contribution on public television STV 1 and commercial television Markíza about the needs of PH patients for oxygen breathing apparatus.
- On 29th of July MD. Milan Luknár and president of PHA Slovakia, Iveta Makovníková attended a two-hour talk show on Radio Slovensko.



- On 29th of September MD. Eva Goncalvesová attended a Health news oriented TV show in television TA3.
<http://www.ta3.com/clanok/1027634/plucna-hypertenzia.html>

Iveta Makovníková
Združenie pacientov s pľúcnou hypertenziou
www.phaslovakia.org
<http://on.fb.me/Ud1DUf>





SLOVENIA

PICNIC ON THE RIVER

On the 13th of October Društvo za pljučno hipertenzijo Slovenije organized a picnic on a tourist boat through the capital of Slovenia on the river Ljubljanica. We were able to organize the event thanks to the help of Medis d.o.o. They help us a lot when it comes to spreading awareness and we are looking forward to working closely with them in the near future. We would also like to use this opportunity to thank them for all their support.

Tadeja and I told the group about our trip to Barcelona for the PHA Europe annual meeting. We explained what kind of presentations we listened to and how big PHA Europe's plans are. A lot of attention was focused on providing psychological support for PH patients. Dr. Barbara Salobir, who was present at the picnic, supported the idea with open arms. We will try to get this message across to every lung unit in Slovenia. We made a promise to do as much as possible in the coming year to come to spread awareness of the disease. We will also focus on increasing the number of members of our association.

We will make sure to spread the word of PH. We want the general public to have a better understanding of the symptoms that point to PH and that the time required to diagnose and treat the disease is shortened.

MARATHON IN LJUBLJANA

Awareness about PH was spread during a marathon that took place in Ljubljana on the 27th October. The marathon was a huge success. A record breaking 19.335 runners participated, among them 5 girls that represented our association. They really trained a lot and all of them reached the finish line without much effort. The oldest participant of the Marathon was the 79-year-old Slovenian Kazimira Lužnik, who successfully made it through the whole course. A big thank you once again to our girls: Maruša Kolačko, Monika Koselj, Kristina Klemenčič, Tajsi Stanojevič in Janja Klančar

Klara Klancar

Društvo Za Pljučno Hipertenzijo Slovenije

<http://www.facebook.com/PljucnaHipertenzija>



From left: Tadeja Ravnik and Klara Klancar



Klara Klancar (5th from right, next to Gerald Fischer), was awarded first prize for the Best Rare Disease Day campaign 2012 Castelldefels, September 12

In memory of Klara

As we are going to print the entire European PH community is still under the shock of the news of Klara Klancar's tragic and premature death. Klara had been to our annual meetings and made many friends in our community. She had a warm and charming personality and all remember her with great fondness.

We wish to take this opportunity to thank Klara for the enthusiasm, time and dedication she put into setting up the association in Slovenia and conducting many very successful activities.

In particular, in 2012, with her association she won the first prize for the best awareness raising campaign to celebrate Rare Disease Day. For this and other events she managed to engage many famous Slovenian celebrities and considerably contribute to raising the profile of PH in her country.

All our thoughts are with Klara's two daughters, her husband and family.

Dear Klara, you will be greatly missed.

Pisana Ferrari

SPAIN

AWARENESS RAISING

The beginning of 2013 was marked by the launch of the “Blue Words” campaign. ANHP made an awareness raising video featuring hope and joy of life within the limitations imposed by PH. The video features PH patients who dance, walk and talk about the activities that can be done in spite of the disease. This video has been posted in social networks and blogs reaching an audience that had never heard of PH. As a result, ANHP was interviewed for several radio broadcasts and there were articles in the written media. The objective of the video was not only to raise awareness but to lower the negative impact of the news on the newly diagnosed. This way, when they surf the net in search of information, they get not only the fatal 2,8 years of life expectancy but a coping mechanism and a reason to make the best of their condition to improve their quality of life.

SUPPORT GROUPS AND MEMBERS' EVENTS

The association has facilitated two gatherings of patients throughout the year, one in Madrid and one in Barcelona. These gatherings give the chance to mingle with other patients in a relaxed atmosphere, enjoy the company and bring forward new plans and projects to help the continuation of the association through fund raising, which is quite needed as ANHP does not receive any governmental support and has suppressed the membership fee.

RESEARCH

Meetings at the Universidad Autónoma de Madrid (UAM) have produced results and the association has a working agreement with the College of Psychology to adapt the CAMPHOR questionnaire (the only existing tool to measure quality of life in PH patients) to the Spanish speaking community. ANHP strongly believes that attitude and emotional wellbeing increase quality of life and life expectancy. When the work with the UAM College of Psychology is consolidated, ANHP intends to promote a study that will bring evidence on impact of psychological factors in physical condition and progress of the disease.

Two genetic studies on gene mutations influencing the development of PH and its progression received the support of the association. One of them is the continuation of the study funded in 2012 and led by Dr. Pilar Escribano from 12 de Octubre Hospital, and the second study is conducted by Dr. Adolfo Balboa at Vigo University, supported by the association with more patients who sent their blood samples so that the number of findings could consolidate the validity of the preliminary results.

ATTENDANCE AT SCIENTIFIC EVENTS

ANHP has been present to a number of scientific congresses and events to increase the profile of the disease amongst health professionals and reach out to patients through their doctors and nurses. This is the list of events in 2013:

- XIV Pulmonology winter meeting, Seville (7-9 February).
- VI Orphan Drug and Rare Diseases Congress, Seville (February 14-16).
- Launch of the SEPAR “Year of transplant” (Pulmonology and Thoracic Surgery Scientific Society), Barcelona (February 20).
- V CIBERER working day (Rare Disease Spanish Research Consortium), Madrid (February 27).
- FEDER (Spanish Federation of Rare Diseases) Expert Meeting, Madrid (March 2).
- Supported by ANHP, the research team of the 12 de Octubre Hospital presented a poster on their findings at the XXVII Human Genetics Congress in Madrid (April 10-12).
- Neumomadrid Congress, Madrid (April 11).
- AHELMU breakfast (scientific meeting organized by the Rare and Ultra-rare Disease Pharmaceutical Companies Consortium) in Barcelona (April 23).
- SEPAR Annual Congress, Barcelona (June 14-17).
- Meeting on pharmacovigilance organized by the Spanish Ministry of Health, Madrid (September 30).
- Scientific Watch, Pompeu Fabra University, Barcelona (October 10).
- Nursing training at the Basque Country (October 18).
- II International PH Day organized by 12 Octubre Hospital (November 8).
- II working day on Adherence to therapies in Madrid (November 14).

WORK WITH SPANISH AUTHORITIES AND OTHER ORGANIZATIONS

- Celebration of Rare Disease Day at the Senate (March 8).
- FEDER Madrid Assembly Meeting (March 21).
- Spanish Patient Forum Assembly, Barcelona (March 21).
- Presentation of the “Call to Action” to the Spanish Ministry of Health (April 15).
- FEDER General Extraordinary Assembly, Madrid (May 31).
- Reinforcement of the Pharmacovigilance Programme with new leaflets for doctors and patients involvement.
- PHA Europe GAM, Castelldefels (September 11-15).
- Week of the Heart, organized by the Spanish Society of Cardiology, Madrid (September 24-28).
- Mostra d'associacions de la Mercè, Barcelona (September 24).
- Meeting with the Catalan Ministry of Health, Barcelona (September 27).
- Meeting with the CEO of La Paz Hospital to make an intervention on misuse of off-label erectile dysfunction

sildenafil for HAP (October 30).

- Board of Directors meeting of the Latin PH Society in Santiago de Chile (November 22).

FUND RAISING AND AWARENESS RAISING EVENTS

- First Padel tennis Tournament in Fuenlabrada, Madrid.
- World PH Day spinning frenzy organized by Bayer at a sports center, Barcelona, with an ANHP information spot (May 5).
- ANHP charity concert, Madrid (October 4).
- IRONMAN Lanzarote, IRONMAN Zurich and IRONMAN Hawaii with the participation of Spanish athletes wearing the ANHP logo.
- Algete and Valencia races with participation of athlete Patricio Doucet wearing the ANHP logo.

ACTIVITIES WITH THE PHARMACEUTICAL INDUSTRY

- Working meeting at Actelion, Barcelona (October 21).
- Bayer press conference on CTEPH (October 29).
- Patient open doors day at Lilly (November 22).

JOINT ACTIVITIES WITH PHA EUROPE

- EUPATI Congress in Rome (April 19).



SWEDEN



PH PATIENT REGISTRY

PH Sweden has been invited to attend a working group together with PH professionals to discuss how the annual report of the national registry of PAH patients can be more useful for patients and carers.

TV INTERVIEW

In November the Swedish national radio broadcast a long interview with one of PHA Sweden's members, Peter Bäärnhielm. In the interview Peter described his life journey which changed direction when he was diagnosed with PH.



- World PH Day regional celebration for Latin America in Sao Paulo, Brasil (May 4).
- EURORDIS General Assembly and DITA face to face meeting in Dubrovnik, Croatia (May 30 to June 1).
- Latin leaders summit, Santiago de Chile (November 22).
- Latin PH Day, Santiago de Chile (November 23).
- Several DITA (Drug Information Transparency and Access) task force meetings hosted by EURORDIS.

Note: All listed activities counted on the active attendance of ANHP either as participants or speakers. Due to the limitation of space it is impossible to elaborate on the contents, for further information on specific activities ANHP will publish the Activity Report 2013.

Irene Delgado, Juan Fuertes
Asociación Nacional de Hipertensión Pulmonar

www.hipertensionpulmonar.es

<http://www.facebook.com/hipertensionpulmonar>



NURSE WORKSHOP

Patrik Hassel shared his story being a father to a child with PH at the Bayer nurse workshop held in Stockholm in June 7-8. In the presentation Patrik referred to the patient and carer survey and how hard it sometimes is being a carer. He shared his despair, frustration, joy and hopes and afterwards he got a lot of positive feedback about the presentation.

Patrik Hassel, PHA Sweden

<http://pah-sverige.se/> - <http://on.fb.me/WcaOWZ>



SWITZERLAND-SPHV

INTERNATIONAL RARE DISEASE DAY

On 23 February 2013 the Swiss PH Association was present at a meeting of the International Rare Disease Day in the Irchel Campus of the University of Zurich. The target audience of this event was the group of patients with rare diseases, their relatives and patient organizations, health professionals, scientific researchers and politicians. There were about 300 participants and more than 30 Patient associations. Among the prominent participants were present: the director of the Swiss Federal Office of Public health, several directors of cantonal public health offices, and the Chairman of the association of the Swiss health insurances SantéSuisse, the President of the Swiss Academy of Medical Sciences etc. The main subject-matter involved questions concerning patients' access to treatment, insurance, promotion of research and social matters. Several members of the SPHV board presented our association with posters and had useful exchanges and discussions with other participants and representatives of other patient organizations.

THIRD GENERAL ASSEMBLY

The third general assembly of the Swiss PH Association took place on 23 March 2013 in the Congress Hotel in the railway junction Olten, between Zurich, Basel and Bern. Twenty three members were present. The main item on the agenda was the presentation of the annual financial statement and the election of the Board members. The board members remained unchanged with the exception of the Vice President. It was stated that the financial situation of the association, which has more than 100 members, can be considered as healthy. In addition new rules concerning donation and sponsoring were decided. The President informed the participants about the planned

Therese Oesch



activities in the current year. The first was the charity concert of two amateur choirs and of the group "Oesch die Dritten", internationally known from the TV program "Musikantenstadel" and the second was the presence of the SPHV at the IRONMAN race in Zurich in the summer.

WORKSHOP AT MEDISERVICE

On the 22nd of April the members of the association were invited to a workshop which was sponsored by the mailing pharmacy MediService and a pharmaceutical company. An experienced motivation trainer was invited for a speech under the title "Self-Inspiration and to Motivate other People". Twenty-four members of SPHV attended this event. The presentation was much appreciated by the attendees.

NATIONAL PATIENTS MEETING

On 1 June 2013 the traditional patient meeting was held in Olten. Fifty-seven persons were present. The topics of the meeting were "Social Insurance Consulting", a speech of Ms. Regula Palladino of an important mailing pharmacy and "Travelling with Oxygen" a speech Mr. Daniel Gutbrod, of a leading gas provider.

REGIONAL MEETINGS

In the current year several regional meetings took place in Chur, canton of Grisons, in Zurich, in Brunnen, central Switzerland and in Bern. At these meetings the patients have the opportunity to contact each other and to discuss about their concerns. The President and the Vice President of SPHV were attending all of these meetings. The most attended meeting was that in Zurich with 18 attendees.

MEETING FOR FAMILIES WITH PH CHILDREN

As usual, a meeting for families with PH children was organized from 31 August to 1 September 2013. Five families had an appointment at the entrance of Zurich Zoo. They hiked about half an hour in a forest where they had a nice barbecue at a camp fire. Subsequently they had a night-time visit in the zoo with accommodation in a yurt tent.

IRONMAN ZÜRICH

PHA Europe organized the presence of the association at the Ironman Zurich from the 26th to the 28th July. The aim was to raise awareness of the disease in the general public and to improve its awareness. Several members of the SPHV as well as representatives of PHA Europe were present at a booth. Some of the athletes raced with the slogan "Breathless for PH". After an initial uncertainty about the process of the event at the beginning, SPHV members and volunteers found many interested and engaged athletes and supporters and they distributed brochures, pens, flyers and "Get breathless" t-shirts. Therefore the presence of the PH associations at this event was considered as a success.

PHA EUROPE GENERAL ASSEMBLY IN BARCELONA

PHA Europe sent a representative to the GAM, the father of a PH child who is scientist in the field of PH drugs.

PH E.V. MEETING IN FRANKFURT

This meeting was held from 24-27 October in Frankfurt. As in past years a large delegation of SPHV members attended this meeting which provided a great opportunity to meet other people with PH, as well as doctors and scientists who are working in this area. The bus trip to Frankfurt was sponsored by a pharmaceutical company.

Therese Oesch, SPHV

www.lungenhochdruck.ch - <http://on.fb.me/1dfrZ2B>





RIOCIGUAT FDA APPROVAL

On October 8, 2013, the U.S. Food and Drug Administration approved Adempas (riociguat) to treat adults with two forms of pulmonary hypertension. Adempas belongs to a class of drugs called soluble guanylate cyclase stimulators that help arteries relax to increase blood flow and decrease blood pressure. It is intended for patients with chronic thromboembolic pulmonary hypertension (CTEPH) after surgery or patients who cannot undergo surgery, to improve their ability to exercise. Adempas is also indicated for patients with pulmonary arterial hypertension (PAH) of unknown causes, inherited or associated with connective tissue diseases, to improve their ability to exercise and to delay clinical worsening of their condition.

The clinical trial evaluating the safety and effectiveness of Adempas to treat PAH included 443 participants randomly assigned to take Adempas 1.5 mg or 2.5 mg, or placebo, three times daily. After 12 weeks of treatment, the 6-minute walk distance in patients treated with Adempas improved by an average of 36 meters (about 118 feet) more than in patients treated with placebo. Adempas is marketed by Bayer HealthCare Pharmaceuticals Inc., based in Wayne, N.J.

Source: <http://www.fda.gov/newsevents/newsroom/pressannouncements/ucm370866.htm>

MACINTENTAN FDA AND EMA APPROVALS

On October 18, 2013, the U.S. Food and Drug Administration approved Opsumit (macitentan), a new drug to treat adults with pulmonary arterial hypertension (PAH), a chronic, progressive and debilitating disease that can lead to death or the need

for lung transplantation. Opsumit belongs to a class of drugs called endothelin receptor blockers, which act to relax the pulmonary arteries, decreasing blood pressure in the lungs.

Opsumit's safety and effectiveness were established in a long-term clinical trial where 742 participants were randomly assigned to take Opsumit or placebo. The average treatment duration was about two years. In the study, Opsumit was effective in delaying disease progression, a finding that included a decline in exercise ability, worsening symptoms of PAH or need for additional PAH medication. Opsumit is marketed by San Francisco-based Actelion Pharmaceuticals US, Inc.

Source: <http://www.fda.gov/newsevents/newsroom/pressannouncements/ucm371362.htm>

European Medicines Agency

On Dec 20 the European Commission approved Opsumit for the long term treatment of pulmonary arterial hypertension (PAH) in adult patients of WHO Functional Class (FC) II to III.

Source: <http://ec.europa.eu/health/documents/community-register/html/newproc.htm>

TREPROSTINIL FDA APPROVAL

On December 20, 2013, the U.S. Food and Drug Administration approved Orenitram (treprostinil) extended-release tablets for the treatment of pulmonary arterial hypertension (PAH) in WHO Group I patients to improve exercise capacity. The primary study that established efficacy (FREEDOM-M) included predominately patients in WHO functional class II-III and etiologies of idiopathic or heritable PAH (75%) or PAH associated with connective tissue disease (19%). Orenitram is a trademark of United Therapeutics corporation, Silver Spring, Md, USA.

Source: <http://www.accessdata.fda.gov/scripts/cder/drugsatfda/index.cfm?fuseaction=Search DrugDetails>

Next PHAE newsletter

The next Mariposa Journal will be issued in June 2014 (Spring edition). It will also feature reports from the national associations about activities in their countries, updates on research and treatments and any other interesting and relevant developments at European level. We would be very grateful if the members could send their contributions by **the 15th of May at latest**. These may be sent in the national language and we will provide for translation. Any photos should have a minimum resolution of 1 MB. The newsletter will be available in PDF format and in a printed version. The PDF will be posted on the PHA Europe website. Printed copies will be sent by normal post to the national associations. *Many thanks in advance!*

2014

February 25 - Brussels, Belgium

EURORDIS GALA DINNER FOR RARE DISEASE DAY

The Black Pearl Gala Dinner is a fund raising event organised by EURORDIS within the context of celebrations for INTERNATIONAL RARE DISEASE DAY. President Gerald Fischer, Pisana Ferrari, Hendrik Ramaker and Luc Matthysen have been invited to attend the gala dinner on behalf of PHA Europe.

May 5 - Worldwide

3rd WORLD PH DAY

PH associations around the world are expected to take part in the annual PH World Day (40 took part in 2013). PHA Europe and its affiliates will be running the "Get breathless for PH" awareness campaign developed last year, centred around sports and solidarity for those who are not able to physical activity, ie PH patients.

May 8-10 - Berlin, Germany,

7th EUROPEAN CONFERENCE ON RARE DISEASES & ORPHAN PRODUCTS

The ECRD is a platform for all rare diseases across Europe, bringing together all stakeholders: patients' representatives, academics, health care professionals, industry, payers, regulators and policy makers. It is a biennial event, providing the state-of-the-art of the rare disease environment, monitoring and benchmarking initiatives. PHA Europe will be attending.

June 2-3 - Paris, France

INTERNATIONAL CONFERENCE ON CTEPH

This two-day conference is organised by the International CTEPH Association and features a very prestigious panel of international speakers. PHA Europe has been invited to attend.

June 20-22 - Indianapolis, Indiana

PHA USA INTERNATIONAL CONFERENCE AND LEADERS' SUMMIT

PHA's 11th International Conference will bring together PH patients, caregivers and medical professionals for three days of education, support and networking. PHA's Europe President and other representatives will be attending this meeting.

August 30-September 3 - Barcelona, Spain

EUROPEAN SOCIETY OF CARDIOLOGY'S ANNUAL CONGRESS

PHA Europe will be present as an exhibitor at the EUROPEAN SOCIETY OF CARDIOLOGY'S ANNUAL CONGRESS 2012. This Congress is currently the largest cardiovascular medicine meeting in the world. It covers all disciplines from basic research to clinical practice.

September 6-10 - Munich, Germany

EUROPEAN RESPIRATORY SOCIETY'S ANNUAL CONGRESS

PHA Europe will also be present with a booth at the EUROPEAN SOCIETY FOR RESPIRATORY MEDICINE'S ANNUAL CONGRESS 2012. This Congress involves not only leading experts in respiratory medicine, but also in the entire spectrum of research and practice.

September 17-21 - Barcelona, Spain

PHA EUROPE GENERAL ANNUAL MEETING

PHA Europe will be holding its 2014 GENERAL ANNUAL MEETING in Castelldefels (Barcelona) September. PH Patient Leaders from 27 European countries attended the 2013 meeting.

October - Sophia Antipolis, France (date to be defined)

PH EDUCATIONAL COURSE

This yearly international course has as its target audience mainly cardiologists, pulmonologists and other specialists involved in PH care. PHA Europe is regularly invited to attend and to present the patient perspective in the context of the G6, a meeting which brings together the major stakeholders in PH.

Members of PHA Europe and contact details

AUSTRIA

Selbsthilfegruppe Lungenhochdruck
Wilhelmstraße, 19 - 1120 Wien
www.lungenhochdruck.at
info@lungenhochdruck.at

BELGIUM

HTAP Belgique, Asbl
Première Avenue, 83 - 1330 Rixensart
htapbelgique@hotmail.com

Patiëntenvereniging Pulmonale

Hypertensie vzw
Rode Dries 10 - 2288 Bouwel
www.ph-vzw.be - hendrik.ramaker@skynet.be

BULGARIA

Pulmonary Hypertension Association
37 Hadji Dimitar str, ent. B, fl. 2, ap. 3
5800 Pleven
www.apph-bg.org - todormangarov@abv.org

The Bulgarian Society of Patients with
Pulmonary Hypertension - BSPPH
1 Geneva str. - 1142 Sofia
www.bspph.net - bspph@gmail.com

CZECH REPUBLIC

Sdružení Pacientů s Plicní Hypertenzí
P.O. Box 47, Praha 4, 14021
www.plicni-hypertenze.cz
info@plicni-hypertenze.cz

FINLAND

PHA Finland
c/o Tuulia Näppi
Keskijuoksu 19 J 38 - 02920 Espoo, Finland
pha.fin@gmail.com

FRANCE

HTAP France
c/o Ludvine Chantraine
Allée des Charmes, 8
21380 Asnières les Dijon
www.htapfrance.com
secretariat@htapfrance.com

GERMANY

pulmonale hypertension e.v.
Wormser Straße, 20
76287 Rheinstetten
www.phev.de - info@phev.de

GREECE

Hellenic PH
Mail Box 52700, 145 72 Drosia, Athens
www.hellenicpulmonaryhypertension.gr
www.facebook.com/HellenicPulmonaryHypertension

HUNGARY

Tüdőér Egylet
19, Cházár András Utca - 1146 Budapest
www.tudoer.hu
csabuda.eszter@t-online-hu

IRELAND

PHA Ireland
c/o Regina Prenderville
56 Eccles Street, Dublin 7, Ireland
<http://www.pulmonaryhypertension.ie/pha-ireland/>
rprenderville@mater.ie

ISRAEL

PHA Israel
13 HaShoshan Street - Beit Shemesh 99590 Israel
www.phisrael.org.il - aryeh35@gmail.com

ITALY

Associazione Ipertensione Polmonare
Italiana Onlus
Via Vigoni, 13 - 20122 Milano
www.aipiitalia.it - pisana.ferrari@alice.it

Associazione Malati di Ipertensione
Polmonare Onlus
Via Bagnoregio, 51 - 00189 Roma
www.assoamip.net - info@assoamip.net

LATVIA

PHA Latvia
Str. Celtnieku 6a - 35, Salaspils
Latvia LV - 2121
www.phlatvia.lv/en/ - info@phlatvia.lv

NETHERLANDS

Stichting PHA Nederland
P.O. Box 627
3800 AP Amersfoort
www.pha-nl.nl - info@pha-nl.nl

NORWAY

PHA Norway
c/o Hall Skåra
Kantarellgrenda 6 - 1352 Kolsås
www.pha-no.com - hall@pha-no.com

POLAND

Polskie Stowarzyszenie Osób z
Nadciśnieniem Płucnym i Ich Przyjaciół
ul. Wieluńska 3, 81-084 Gdynia
www.phapolska.org
bartosiewicz.agnieszka@phapolska.org
admin@phapolska.org

PORTUGAL

Associação Portuguesa de Hipertensão
Pulmonar
Avenida Dr. Luis Navega, 38-42 - 3050 Mealhada
www.aphp-hp.org - mariajsaraiva@gmail.com

REPUBLIC OF MACEDONIA

APH Moment
st.Tanche Kamberov, 29 - 1000 Gevgelia
www.facebook.com/pages/APH-moment-Macedonia/1404172929805917
gurgica.wix.com/mermaidinsky#!home-eng/c8t9
pmmomentplus@gmail.com

RUSSIA

Natasha Charity Fund
c/o Evgeniy Komarov - Kondratevsky prospect
House 57, apartment 6 - St.-Petersburg
www.community.livejournal.com/fond_natasha_otzovitesnam@mail.ru - www.nurm.ru

SLOVAKIA

Združenie pacientov s pľúcnou hypertenziou, o.z.
Lučenská, 31 - 990 01 Veľký Krtíš
<http://phaslovakia.org/>
hypertenzia.pluc@gmail.com

SLOVENIA

Društvo Za Pljučno Hipertenzijo Slovenije
Slovenska 29, 1000 Ljubljana
dbph.slovenije@gmail.com
<http://on.fb.me/JWvIjE>

SPAIN

Asociación Nacional de Hipertensión Pulmonar
Avenida de las Artes, 7
28300 Aranjuez - Madrid
www.hipertensionpulmonar.es
informacion@hipertensionpulmonar.es

SWEDEN

Nätverket för PAH
c/o Patrik Hassel
Kärrtorpsvägen, 37
121 55 Johanneshov
www.pah-sverige.se - patrik@pah-sverige.se

SWITZERLAND

Schweizer PH-Verein (SPHV) für
Menschen mit pulmonaler Hypertonie
c/o Bruno Bossard
Im Rossweidli 1 - 8045 Zürich
www.lungenhochdruck.ch - www.sphv.ch
President: praesi@lungenhochdruck.ch
Webmaster: bosshard@lungenhochdruck.ch

Association HTAP Revivre

c/o Mme Lydia Benallouch-Meier
Chemin des Sports 16 - CH - 1203 Genève
www.saph.ch/SuisseRomande
President: lydia_mb@bluewin.ch
Coordinator: monika.sorgemaitre@hcuge.ch

TURKEY

Pulmoner Hipertansiyon Dernegi
Meriç Sokak Milas 2000 Is Merkezi 5A
Blok/26 - Bestepe Ankara
www.pha-turkey.com - info@pha-turkey.com

UKRAINE

Ukraine Association of Patients with Pulmonary
Hypertension
Panasa Myrnoho str.,14 (litera A)
01011, Kiev
www.pha.org.ua - info@pha.org.ua

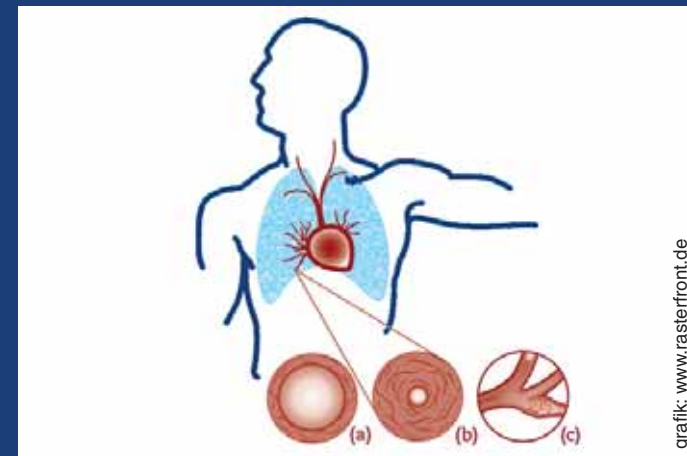


AIMS OF PHA EUROPE

Pulmonary Arterial Hypertension causes breathlessness and is a rare, progressive and currently incurable lung disorder. PHA EUROPE, European Pulmonary Hypertension Association, was founded in 2003 and is registered in Vienna, Austria, as an international non profit organisation. PHA EUROPE is an umbrella organization bringing together Pulmonary Hypertension patient associations across Europe. The primary objective of PHA EUROPE is to establish a narrow cooperation between the members and the European institutions, international organizations and public institutions worldwide and work towards achieving the best possible standards of care for all European Pulmonary Hypertension patients.

WHAT IS PULMONARY ARTERIAL HYPERTENSION?

In patients with Pulmonary Arterial Hypertension characteristic changes occur within the pulmonary circulation, which include thickening of the linings and obstruction of the small pulmonary blood vessels. They are both structurally and functionally abnormal. In severe cases up to 80% of these very small blood vessels are rendered non-functional. As a result the pressure in the pulmonary circulation rises well above normal and this places strain on the right side of the heart. This strain can cause the heart to enlarge and the patient may develop heart failure. This is a disease that can affect all ages and is more commonly seen in females. Pulmonary Arterial Hypertension has an estimated prevalence of about 50 per million population.



- Cross section of normal pulmonary artery
- Reduced lumen of pulmonary vessels due to cell proliferation and remodelling in advanced Pulmonary Arterial Hypertension
- Longitudinal section with pathological changes within the vessels (proliferation, deposition of blood clots, thickening) causing difficulties for the heart to pump blood through the lungs

TREATMENT OF PULMONARY ARTERIAL HYPERTENSION

Over the past decade a number of evolving therapies that either use complex delivery systems such as 24-hour intravenous or subcutaneous drug infusion, drug inhalation and, more recently, oral medications, have transformed the outlook for PAH patients. PAH is a condition that can be rapidly progressive and needs careful, ongoing expert care and management. The disease can be insidious in its onset, with unexplained breathlessness and tiredness as its two main symptoms. If undiagnosed and/or inappropriately treated the average life expectancy for these patients is estimated to be around 2-3 years. For those patients who fail to respond to medical therapies double lung, or in some cases, heart and lung transplantation may be appropriate.

THE FUTURE FOR PULMONARY ARTERIAL HYPERTENSION

While a cure for this aggressive and life threatening disease is still some way off, there is much to be optimistic about. There are an ever increasing number of therapies available for the effective treatment for Pulmonary Arterial Hypertension, which improve the quality and length of life. The aim for the present should be to ensure that all patients with Pulmonary Arterial Hypertension have access to centres of excellence in the diagnosis, management and ongoing treatment of this disease.

OFFICE AUSTRIA:

Gerald Fischer, President - Wilhelmstrasse, 19 - 1120 Vienna - tel. +43 1 4023725 - fax +43 1 4093528 - info@phaeurope.org

OFFICE ITALY:

Pisana Ferrari, Vice President - Via G. Vigoni, 13 - 20122 Milano - mob. +39 348 4023432 - p.ferrari@phaeurope.org

www.phaeurope.org