



# PULMONARY HYPERTENSION MariposaNews

n. 5 • Summer 2011

SUMMER ISSUE OF THE NEWSLETTER OF PHA EUROPE,  
EUROPEAN PULMONARY HYPERTENSION ASSOCIATION

Austria

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Bulgaria

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Greece

Hungary

Israel

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Netherlands

Norway

Poland

Portugal

Slovakia

Slovenia

Spain

Sweden

Switzerland

Turkey



from top left: the founding members of the new Latvian PH association; ten year anniversary celebrations for AIPI, Italy and PH ev, Germany; new cardiac echo equipment being donated by AMIP, Italy; workshop with journalists, Poland

- Activities of PH patient associations in Europe and around the world
- EURORDIS 4th Workshop of the Council of European Federations • PHA Europe at Lisbon, Vienna and Dresden events • Monitoring developments in the European Institutions and NGO's • Update on PH treatments and research on PH drugs • Upcoming events for 2011

PHA EUROPE 



Dear friends,  
welcome to the Summer 2011 edition of Mariposa News. This year's Spring issue was dedicated exclusively to our first pan-European "Breathtaking" awareness campaign, so that we are now reporting on activities and events having taken place in the period January to August 2011.

It has been a very intense last few months and I would like to thank the national patient associations for sharing their achievements with us and providing precious input for Mariposa News. In the course of this year PHA Europe representatives have been asked to present the PH patient perspective at a number of important scientific events (Lisbon, Vienna, Dresden...) and brief reports of these meetings are included in the section which follows on to the national associations' news.

PHA Europe was also invited by EURORDIS, the EUROPEAN RARE DISEASE ORGANIZATION, to speak about the "Breathtaking" campaign at a workshop of the Council of European Federations. This interesting meeting provided a wonderful opportunity to meet with leaders of other European rare disease patient networks such as our own and learn more about the great work EURORDIS is doing in the area of rare diseases and empowerment of patient groups. Pisana Ferrari, who attended on behalf of PHA Europe, has written a summary of the main points. We are very honored that EURORDIS has recently posted our slide presentation on its website.

As we are going to print, preparations are well under way for our General Annual Meeting which is taking place in Barcelona September 8-11. We are very gratified by the exceptional turnout: 45 PH patient leaders from 22 countries: Austria, Belgium, Bulgaria, Czech Republic, France, Finland, Germany, Hungary, Israel, Italy, Latvia, the Netherlands, Norway, Poland, Romania, Russia, Slovenia, Slovakia, Spain, Sweden, Switzerland, Turkey. The meeting has a very wide ranging program with interactive sessions about issues of common interest and presentations by distinguished speakers from the medical and nursing professions, the pharmaceutical industry as well as leaders of PHA UK and PHA USA.

It will be a busy summer, with PHA Europe also being present at both the EUROPEAN SOCIETY OF CARDIOLOGY (ESC) and EUROPEAN RESPIRATORY SOCIETY'S (ERS) Annual Congresses.

The ESC Congress will be held in Paris from the 26th to the 31st of August. As in past years PHA Europe is an exhibitor. We will also have a booth at the ERS Congress which is taking place in Amsterdam from the 24th to the 28th of September. At the ERS we will be taking our "White Paper" project one step further with the organization of an expert roundtable bringing together top medical and nurse PH specialists, patient representatives and industry. More details will follow in our next issue. Another important event coming up shortly is the ESC Educational Course on Pulmonary Hypertension which will be held in Sophia Antipolis, France, October 14-15. PHA Europe has been asked to speak about "What PH patients want" within the context of the Annual "G5". This meeting brings together the five main stakeholders involved in PH patient care: physicians, nurses, industry, regulatory authorities and patients.

The section of Mariposa News carrying information about recent developments at European level is shorter than usual and we had to leave out the links/reading suggestions because of space constraints. We have included the main developments and will catch up in the next edition.

Thank you all again for your support and collaboration. It has been a very successful year so far and I look forward to seeing you all in Barcelona to discuss our future projects.

Gerald Fischer  
President PHA Europe

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## AUSTRIA

### Patient meeting in Linz, 2 April



Over 100 persons attended the PHA Austria patient meeting in Linz (this is a new record for us!).

Speakers included Dr. Regina Mascherbauer, an Austrian PH specialist who spoke about

new PH therapies, Dr. Wille-Wagner, a psychologist, and Dr. Kerle, whose presentation was about oxygen therapy.

### Orlando Hospital, 24th of March



Gerald Fischer with a PH nurse (left) and the President of the PHA patient association Orlando (right side)

Gerald Fischer, PHA Austria President, was in Florida during the month of March 2011 and was invited by the Orlando Hospital to speak about PH. During his visit he met with the President of the PHA Orlando support group, PH nurses and some representatives of the pharmaceutical industry.

### 1st Austrian PAH-CHD Expert Forum in Vienna, 6-7 May

This expert Forum organized by Actelion addressed the issue of Pulmonary Hypertension associated with congenital heart diseases. Gerald Fischer was asked to speak about PHA Europe and the importance of having a strong European PAH association.

### Opening Party of the Ludwig Boltzmann Institut (LBI) in Graz, 30 June

The Ludwig Boltzmann Institut is a research center for PH with 20 researchers. It is financed for the next 4 years (hopefully longer) with a budget of 9 million! This

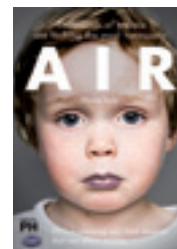
is a tremendous success for the whole PH community.

### Other activities

- At the end of 2010 Gerald Fischer attended a seminar on Health Technology Assessment at the London School of Economics and he received a diploma in "Healthcare decision making".
- Gerald Fischer had the opportunity of meeting the Austrian minister of Health Dr. Stöger to discuss social benefits for PH patients.
- PHA Austria has set up a sms donation hotline for the organisation: 0664 660 8888.
- PHA Austria is part of the team charged with starting up an umbrella organisation for rare diseases in Austria.
- PHA Austria sent out a special "thank you note" and report of activities to all its sponsors and supporters.

### New fundraising campaign, planned for 2011

We have started working on a new awareness campaign for 2011 (see photo).



### Joint meeting PHA Austria and PH e.v. Bavaria in Graz

On the 14th and 15 th of July we held our first meeting together with PH e.v. Bayern (Bavaria) in Graz. On the Friday we organized a visit of the LBI. On the Saturday a number of lectures were scheduled on PH and PH related topics:

- Presentation of the activities of the LBI research institute, Prof. Andrea Olschewski, Graz.
- Threshold of PH, Dr. Kovacs, team of Prof. Horst Olschewski, Graz.
- Right Heart Catheter, Dr. Susanna Desole, Innsbruck.
- Chronic Thrombo-Embolic Pulmonary Hypertension (CTEPH), Prof. Irene Lang.
- State of the art in PH, Prof. Horst Olschewski.

Eva Grassmugg, Selbsthilfegruppe Lungenhochdruck



Photos from Graz event: Prof. Horst Olschewski and Eva Grassmugg at the LBI; visit of the LBI; group photo with (from left) Eva Grassmugg, Gerhard Volmar (President of the German-Bavaria group); view of the meeting room



## BELGIUM - HTAP BELGIQUE

### Recents events

Here are some photos of events that Rosie and I or Rosie on her own have attended since the end of 2010. We both went to the EPPOSI WORKSHOP, November 17th (see photo in the company of Vladimir Tomov, Confederation of Health Protection, Bulgaria).



*Rosie Matthysen with Prof. Jean Luc Vachiéry*

Rosie has taken part in two meetings of the European Medicines' Agency (EMA) Scientific Advice Working Party (December 2nd and 28-30 March 2011). At these meetings she was asked to present the patient's point of view. We also attended a

Workshop at the FOUNDATION ROI BAUDOUIN, on the 18th of December.

Rosie regularly takes part in the master classes organized at Erasme-ULB Hospital (see photo of meeting of November 24th, with Prof. Jean-Luc Vachiéry).

*Luc Matthysen, HTAP Belgique*



*Luc and Rosie Matthysen at the EPPOSI WORKSHOP*



## CZECH REPUBLIC

### Annual Congress of the Czech Society of Cardiology, May 1-4

At the beginning of May we took part in the XIX Annual Congress of the Czech Society of Cardiology. Within the context of our activities at the Congress we also highlighted the "Breathtaking" campaign posters. For doctors present we had prepared some tests, focusing on the thromboembolic form of PH (CTEPH).

### Newsletter

At the end of 2010, the Czech PH Association issued its first newsletter *Bulletin 2010*. This first edition contains a summary of our activities since the starting up of the association, the life story of Dr. Josef Oškera, a PH patient, information about PH and other important related topics, cross-words etc. Feedback has been extremely positive

and we are very happy with the success of this initiative.



*The Czech PH patient association's booth at the XIX Annual Congress of the Czech Society of Cardiology*



### PHA Europe Breathtaking campaign

The Czech Association also participated in "Breathtaking" awareness campaign for Rare Disease Day 2011. Our target was the medical community, in particular general practitioners. That's why we chose to publish the "Breathtaking" campaign poster in "Medical Tribune", the only newspaper about medical science, clinical practice and healthcare in the country, which has very wide distribution.

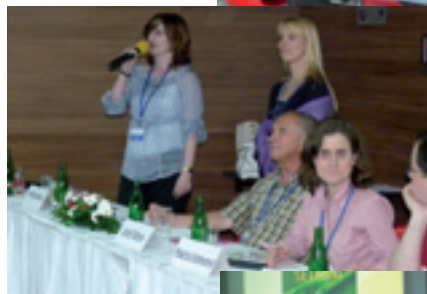
### Patient meeting Seč, June 11-12

In June we held our regular annual meeting with the members of our association. An important guest was Prof. Aschermann, a leading Czech cardiologist. Lectures were on the following topics: Innovations in the treatment of PH with a focus on clinical studies of new drugs (Dr. Pavel Jansa), Food and diet regime in PH (Dr. J. Šatrová) and PH incidence in families (Dr. Regina Votavová). There was also an evening entertainment and relaxation program.

### Future projects

We are currently preparing for the annual rehab/reconditioning stay and putting together materials for the 2011 newsletter.

*Katerina Novaková, Sdružení Pacientiů s Plicní Hypertenzí*



Photos of the patient meeting in Seč.  
Left: Katerina Novaková speaking



[www.plicni-hypertenze.cz](http://www.plicni-hypertenze.cz)

## FINLAND

### Setting up a new patient group

Hello from Finland! I am a PH patient working as a nurse in Helsinki. I first made contact with PHA Europe through a Finnish doctor who had met Pisana at a conference in November 2010. It was very useful to exchange advice and information about what was going on in Europe in the area of PH.

I would like to tell you about the latest developments in setting up PHA Finland.

In May 2011 I met with a group of PH patients at a weekend meeting in Espoo organized by the FINNISH HEART ASSOCIATION. I was glad to meet some very enthusiastic people who were interested and want to be involved in the association. I told them about PHA Europe and promised to keep them posted on activities and information about PHA Europe and to forward them the Mariposa Newsletter.

Some of them expressed the difficulty of understanding English, but I promised to translate articles into Finnish if they wished. I received a good mailing list, and will

be in contact with them and send them the latest newsletters.

I have been lucky to be in contact not only with people from the FINNISH HEART ASSOCIATION (Sydänliitto) but also the FINNISH BREATHING ASSOCIATION (Hengityslitto Heli).

They have promised to help us establish PHA-Finland.

We are planning a meeting for August of this year. In October Bayer is planning to organize a PAH-nurses' training course. I will also be attending this training. Very excited about it!

Warm regards and see you all in Barcelona.



*Jaana Rajahalme*

## FRANCE

### Family Weekend

Last May we held the 4th edition of our *Weekend familles-enfants* (WEEF). Céline, who is one of our Vice-Presidents (her son Paul, 10, died last year from PH), and is in charge of relations with pediatric patients' parents, invited 50 people to spend the weekend together in a hotel in the countryside in Jura (Eastern part of France). There were seven patients' families, among which two grand mothers and a godmother. Some came a very long way, even from the south-west part of France! Those who had already been to a WEEF were very excited to get together, especially five teenagers who asked for a room where they could all sleep together (they did not sleep much during the weekend...)! Three cardio-pediatricians including Prof. Maurice Beghetti, who came from Geneva with his family, took part in the WEEF, as well as a psychologist and two baby sitters. It was our best family weekend ever! The baby sitters were a key factor for the success of the weekend, because the children were busy and having great fun all the time while the parents were chatting or at the sessions. Another reason for the success is the doctors' support in enrolling families (this had not been the case previously). We think that the participation of Prof. Bonnet from the Necker Hospital in Paris (French reference center for Children with congenital heart defects) in the last two years has made a turn because he is very enthusiastic about what we do. Clearly the WEEFs are not only beneficial for sick children, but also for the whole family. It helps to understand how to live better with PH and enables parents to share experiences and feelings. There is always a very emotional Sunday morning when parents talk in a group with the doctor and Céline is there to moderate. I was also there, as an "ex pediatric patient now transplanted". I think realizing that one can live such a long time with PH and enjoy life was quite encouraging for them. We were all very satisfied with our weekend, exhausted but certain that we had brought some happiness and not only... We had to make the decision, however, that in future we will not accept more than 50 people for one weekend otherwise it would lose its conviviality. Another aspect is that we have to find dedicated funds for this

project (the participants only paid for the travel and a contribution of 15€ per adult) and the budget issue needs to be further addressed.

### Patients Regional Meetings "RR"

This year we have already held two regional meetings, one in Saint Lô in Normandy, with the doctors of the competence center in Caen, which gathered only 50 people despite the fact that Caen is a center with 180 patients. The other meeting was in Chedigny, with a doctor from Tours. There was not many people here either (Tours is a much smaller center anyway). We have come to the conclusion that for a first patients meeting in a region, it is much better to locate it next to the center (these meetings were 50 km away from the hospital). Planning it in the countryside when people already know the association is no problem but new patients may be reluctant to take the risk of losing time!



top left: poster session on Sunday morning at the WEEF  
top right: teenager girls meeting at the WEEF

### Sports events "A pleins poumons"

A great part of our week-ends was busy with sports or entertaining events. The most important were those in Toulouse, Lyon and Sète.

For the 7th time we held an ultra-marathon between Toulouse and Port la Nouvelle (a port on the Middle Sea). People run or cycle along the "Canal du Midi and Canal de la Robine" for two days. Runners all had a 'godchild' patient. This patronage brought much emotion, and helped patients exchange with runners and also encouraged patients and members to support the event and the runners. About 180 people took part in the race and many others were supporting along the way. There was a report on one of the most important national TV channels, "TF1", in the famous "Journal de Claire Chazal". This contact with journalists was difficult to obtain and took a long time so we consider ourselves very lucky. In Lyon, Sylvain, our President, and his friends organized a sports day with several running races for individuals, adults, children or fam-



Photos from the regional meeting in Tours



ilies. Over 200 participants came, not many of them concerned by the disease (the people concerned were mostly Sylvain's family and friends), who just came to run. It was a good occasion to raise awareness (see blog on our website). During the weekend in Sète (near Montpellier, on the Middle sea), we organized a race in the city, a visit of the city, a dinner for the 'official people' where Mrs Anh Dao Traxel (adopted daughter of former President Jacques Chirac) was invited, and a concert of Patrick Fiori, a French singer in front of 1800 people. Just before the concert, Dr Amedro from Montpellier, a pediatric cardiologist, made a presentation on PH.

### Patient Study

Since year 2009, we have been working on a patient study about the PH care they experience in the centers, and how they live with PH in everyday life. The questionnaires of the quantitative phase have just been gathered (about 275) and are currently being analysed. We expect to have the results in October 2011 and are thinking of how we will publish them.

These are a few of the activities we wanted to share with you!

*Mélanie Gallant Dewavrin, HTAP France*

[www.htapfrance.com](http://www.htapfrance.com)

## GERMANY

### Happy anniversary! 15 years for PH e.v. Germany and 10 years for the René-Baumgart Foundation

This year the General Assembly of PH e.v. Germany on April 16th, 2011 was a special one, a moment to celebrate and look back at our achievements and progress made in the treatment of PH. 15 years ago the association was founded, in 1996, and 10 years ago, in 2001, the René Baumgart-Foundation. In these years a variety of drugs and treatment options for patients with PH have been developed. But it was also a moment to look ahead, because despite the treatment options there is still no cure for patients suffering from Pulmonary Hypertension. There is a lot to do for the association and the foundation.

Prof. Ekkehard Grünig, Heidelberg, gave a short review of the history of PH diagnosis and treatment and mentioned the researchers who have received a René Baumgart Foundation award in the past. He presented the work of Mrs Karen Olsson, who received the award in 2011 for her work on an artificial lung which will be able to help to bridge the time for patients waiting for transplantation.

In his greetings to the assembly the patron of PH e.v., Dr.

Erwin Vetter, pointed out that thanks to PH patient associations awareness for the disease has improved not only in Germany but also in Europe, where the cooperation of PH associations helps to raise awareness at all levels of health politics. He underlined the importance of the foundation in the public health system, being a helpful part of research funding.

Before the anniversary ceremony started with speeches, greetings and music, elections were held, and the Board of PH e.v. Germany was reelected for another two years.

### Charity Concert for PH Germany's René-Baumgart Foundation

The aim of PH Germany's René-Baumgart Foundation is to help people suffering from Pulmonary Hypertension by raising awareness and supporting research in the field of PH. The regional police choir and orchestra of Karlsruhe and Bruchsal with 170 musicians and singers gave a charity concert at the Community Centre in Bruchsal on Saturday, May 7th 2011. Over 600 people attended the concert, the proceeds of which went to the René-Baumgart Foundation. The audience was delighted by a variety of songs and pieces of operas, pop, gospels and musicals. For the final song "Conquest of Paradise" all 170 musicians gathered on the stage to round off a marvelous evening.

*Günther Thimm, Pulmonale Hypertonie e.V.*

*Charity Concert for the René-Baumgart Foundation in Bruchsal*



[www.phev.de](http://www.phev.de)

### HUNGARY

#### Networking with Hungarian pulmonologists

A good relationship has been built between Tüdőér Egylet and the Hungarian Association of Pulmonologists. We attended a scientific session of the Allergy and Breathing Pathology Section of the this Association in Eger on 24-26th March, 2011. We had the opportunity of speaking to the physicians and meeting some charming ladies who had undergone lung transplant. We received momentum and encouragement from them.

#### Annual patient meeting



*Eszter Csabuda (first from right), President of the association, at the patient meeting with some of the guests*

financial plans of 2011. The success of the “Breathtaking” campaign in Hungary in terms of improved awareness of PH and the association were also discussed and assessed.

#### Joining RIROSZ, Hungarian association for rare diseases

Based on the decision of the Hungarian PH association’s board, on January 28th, 2011 we applied for membership (which was granted) to RIROSZ, the Hungarian association for rare diseases. This relationship has enabled us to

organize common events. We won a tender, the aim of which is to elaborate the strategic objectives of the association.

The strategy will address medical healthcare, the quality of life and the social needs of patients with rare diseases. The long term aim is to lay down a nationwide plan. The Strategic Conference of RIROSZ was held on July 6-8, 2011 in Balatonfüred.

#### Access to PH drugs

No major changes have occurred in access to PH treatments in our country. Revatio, Tracleer, Volibris and Ventavis are the most used ones. The withdrawal of Sitaxentan did not impact us, because there were no patients on this drug. It is good news that in spite of the difficult economic situation and the fact that other non PH drugs become more expensive and their subsidy has been abolished, no restrictions have been introduced in the treatment of PH. Prostacyclins can be used on a case by case permission, but obtaining it is really difficult. A great restructuring may occur in the national medical healthcare system, and the position of rare diseases in this process is not predictable.

*Gergely Mészáros, Tüdőér Egylet*



*First from the left Kristof Karlocai MD, VP of the association, the second from left Eszter Csabuda, President of the association, at the patient meeting*

[www.phev.de](http://www.phev.de)

### IRELAND

#### A busy year for PH!



*from left to right Irene Bergin, Sonia Morrison, Prof S. Gaine and Pisana Ferrari*

2011 has been a very busy year for Pulmonary Hypertension in Dublin. We completed our involvement with the “Blue lips” campaign and received our certificate from the Guinness Book of

Records which now has pride of place in the PH Unit. In April we had our first PHA Ireland meeting for a number of years and it was a great success with over a hundred people turning out for the occasion. We were delighted to have the Vice-President of PHA Europe, Pisana Ferrari, over to speak and her message was greatly appreciated by all in attendance. We hope to facilitate a patient support group later this year as a result of the meeting and garner more involvement from Irish patients in their organisation. Wishing all our friends in Europe a pleasant summer!

*Caitriona Minnock, PHA Ireland*

[www.centreforlunghealth.com](http://www.centreforlunghealth.com)



## ISRAEL

The Israel Pulmonary Hypertension Association has had an exceptionally busy few months of fruitful activity.

### Conference for Primary Care Providers

On June 16th at Tel Aviv's seafront Dan Hotel, nearly one hundred primary care clinicians (physicians and nurses)



convened for a conference sponsored by the Israel PHA on the subject of "Pulmonary Hypertension: Profile of the Disease". Israel's most senior PH specialists presented lectures in sessions on disease identifi-

cation and symptomatology, diagnosis, treatment options, and research.

Additionally, a panel consisting of the director of a pediatric pulmonary rehabilitation department, a family physician, a mental health specialist and patients' advocate, a medical logistics specialist, and a patient provided their experiences concerning the complexity of caring for a PH patient.

The organizers believe that the event served to significantly expand the Israel PH community, enhance awareness of the disease, and cultivate a commitment to screening and support from among the clinicians in attendance. We have also agreed to continue professional education concerning the disease in the future. As a result, we have already begun planning future activities of this kind.

There was post-conference coverage of the event in one of Israel's premiere, online medical forums. Included were synopses of the main lectures.

The Israel PH Association is grateful for the Tom Lantos Innovation in Community Service Award that we received, which provided essential funding for the conference. Our appreciation is extended to the US Pulmonary Hypertension Association for facilitating and administering the grant. Such support and collaboration goes far in making the PH community a truly international one.

### Yarid Shira

Two weeks after the conference, the Israel PH community experienced a bitter-sweet celebration that brought people from throughout the country to Park Raanana, a beautiful setting in the heart of Israel. The event was characterized by an incredible outpouring of support during a day filled with excitement, color, booths and health-related activities like Tai Chi and yoga. Vendors selling everything from fresh

produce to garments donated twenty-five percent of their proceeds to the Israel PH Association for the purchase of mobile oxygen systems.

The gathering, which took place under brilliant skies along a lake (a lake, in Israeli terms, is a pond elsewhere), was mixed with sorrow. The fair is named in honor of Shira Dinur, a 26 year-old PH patient much beloved by members of the Association. No less adoring of Shira were her friends, who as volunteers put together the festival in her honor with great commitment and pizzazz. The organization was exceptional and the coverage was extensive throughout the Israeli media. It was a wonderful tribute to Shira and is slated to continue each year.

Yarid Shira was sponsored by the Israel PHA Association,

the Ra'anana Municipality, corporate sponsors, small businesses and others.



### Second Support Group

A support group, Israel's second in the north of the country has spun-off from a highly successful one in the center of the country. A number of meetings have been held and there is optimism that this group will become rooted and offer PH patients mutual support and a venue for the exchange of information in that region.

### Israeli and American Firm Team on New PH Treatment

The announcement in June that an Israeli firm, Pluristem Therapeutics, and an American company, United Technologies, have partnered to advance a possible new treatment for PH based on cellular technologies was met with great anticipation by the PH community. Initial findings offer hope that after several more years of applied research and clinical trials, the new treatment based on stem cells will lead to a breakthrough for PH patients the world-over. Dr. Frida Grynspan Gotlieb, the chair of the research session at the PH: Profile of the Disease conference is the vice president of research and development of Pluristem, which is located in Haifa, Israel.

The Israel PH Association looks forward to building on the achievements of the past few months and further expanding and deepening the range of our programs.

*Yosef Gotlieb, PH Israel*

## ITALY - AIPI

### Ten year anniversary celebrations!



This year AIPI celebrated its 10th anniversary on the 3rd of April, at its annual patient meeting. Over 140 patients and family members from all over Italy attended the one-day event which was held in Bologna. All but one

of the seven founding members were present and we started the meeting by going back to how it all started, with the monthly meetings of our first tiny support group, then looking to how the association has gradually expanded over the years: our first website and newsletter, our first annual assembly... A lot has been achieved in these years: we now have almost one thousand members (700 of which are patients) and offer many services to support patients and their families including telephone help lines, message board, publications, audiovisual materials, professional psychological assistance, help with logistics, visits to patients in hospital, economic aid.

During the morning session we had two interesting presentations, the first of which on recent provisions for the disabled, and then an update on PH treatments by Prof. Nazzareno Galie from the PH Unit at the University of Bologna.

After lunch we enjoyed *Prosecco* and the wonderful "birthday" cake with the AIPI logo which Marzia Predieri, our Treasurer, had kindly organized. In the afternoon we were treated to live music, with two singers. Riccardo Rossini, a PH patient and member of the association sang some wonderful American songs from the 50s and 60's. Selene Lungarella is a charming young professional singer whose father suffers from PH. She sang some very nice pieces from her latest CD and other well known Italian hits. Everybody was singing along with her and we all enjoyed ourselves a lot.



### New graphic layout for AIPI newsletter

We have recently done some restyling on our AIPI quarterly newsletter. The new graphic layout is more colourful and bright and we have new sections with different topics.



### New publications

AIPI has recently printed an updated patient survival guide and translated into Italian and published the Spanish association's "Guide to the emotional aspects of PH". We would like to take this opportunity to renew our warmest thanks to Irene Delgado, President of

ANHP, who very kindly allowed us

to translate it and use. This is a wonderful example of how patient associations can work together across borders. We are sure that this excellent publication will be very useful to our Italian patients.



### Excursions

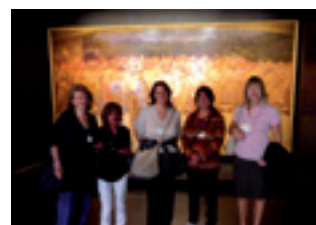
This is something very new for AIPI. The first excursion was a "joint" visit (AMIP and AIPI, the two Italian PH patient associations) to the beautiful gardens of the Quirinale, official residence of the Italian President of the Republic, in Rome. Marzia Predieri from AIPI and Luisa Sciacca della Scala, President of AMIP, with a few members and patients from both groups, enjoyed touring the gardens and the nice lunch in a nearby trattoria. The second excursion was in Milan, with a visit to the Museo del '900. Three PH patients and two AIPI members enjoyed a guided tour through the museum which features art from the 20th century.

*Pisana Ferrari, AIPI Italy*



left: Photos of the annual patient meeting: distributing materials, the meeting room, Riccardo and Selene singing for us

right top: Lunch after the visit to the Quirinale gardens in Rome  
right: Excursion to Milan's Museo del '900



## ITALY - AMIP

**Patient meeting**

Our annual 2011 patient meeting took place in Rome on June 17-19. We met at the Hotel Domus Carmelitana, the same venue as in past years. The highlight of the event was the Saturday doctor-patient meeting. This year we had doctors coming from both the Rome and Pavia specialized centres. They updated us on developments in research and available treatments for PH. The patients had the opportunity to ask questions, talk about their problems and, most importantly, they had all time to be listened to.

After a meeting of this kind the patients are really confident that they are in good hands. I think this is a fantastic result! This year Rome "awarded" us some very beautiful sunny days, we had all the time we needed to learn (a lot), but also to enjoy ourselves, make friends and share experiences.

**New projects and fundraising**

During the last few months we have put a lot of effort into fundraising. For this activity we must thank our members, some of whom have been really fantastic: Emma was able to involve the whole town where she lives, Chieti, to take part in a beautiful Folk Concert and Claudio organized in his town, Gualdo Tadino, a match between the "Italian Show-men Team" and the local players. Not to mention the many initiatives our tireless friend Vittorio organizes for us. Lately we were among those who benefited from the "Heart Football Match" that the "Italian Actors' Football Team" played in Rome. The match was broadcast on Rai2 TV - national channel.

The projects we finance are always rather ambitious: after the Cardiac Echo we bought last year for the Rome Specialized Centre, we decided to donate the same equipment to Pavia's Center. Both Centers are following hundreds of PH patients and it is very important that patients benefit from the very best possible facilities to improve quality of their follow-up. The association tries to "fill in" where the Public Health system fall short on...



AMIP patient meeting on the roof garden of the hotel. In this photo 9 Italian regions are represented. Third from left: Luisa Sciacca della Scala, AMIP President, and 4th from left Maria Pia Proia, Past President

We are also still paying a Research Grant for a young doctor in the Ospedale Pediatrico Bambin Gesù in Rome. She works with Dr. Parisi who has been treating children with PH for over 10 years in this Hospital. We think that it is important to encourage this facility as it is one of very few available in Italy for pediatric patients.

**A documentary about PH**

Last autumn some of our patient members were interviewed by Federico Bonelli and Marzia Papagna (a film maker and a journalist who happen to be my son and a young friend of mine). Up to that time Federico and Marzia knew practically nothing about PH. The patients to be interviewed were chosen at random, depending on their availability during the few days we had at our disposal to go and meet them in different regions of Italy. The filming was done with simple technical means, but with our great satisfaction the long interviews we were able to collect provided a wealth of emotions and courage. From these interviews we put together a documentary, the first one ever made in Italy, that tells of a journey inside the lives and houses of PH patients. One discovers that every patient must face more or less the same problems: the difficulty in arriving at a correct diagnosis and, even more, the difficulty in living with a disease that is unknown not only to one's friends and environment, but often also to the doctors and institutions that should take care of the patients. The documentary also tells about the hope and new energy the patients draw from the progress of scientific research about

from left: Drs. Mezzapesa and Badagliacca (Policlinico Umberto I, Rome), Alfieri (Bambin Gesù, Rome), Camporotondo and Rainieri (San Matteo, Pavia), Luisa Sciacca della Scala, Drs. Poscia, Nocioni and Papa (Policlinico Umberto I, Roma) with the plaque of the Cardiac Echo equipment it is donating to the Pavia hospital



### ITALY - AMIP

their illness and from being able to rely on the competent Specialized Centers that we are lucky to have in Italy. The documentary "Living with PH" is especially dedicated to all those persons who are suddenly faced with this serious and unknown disease; it will help them to see how others were able to overcome their same problems.



On the 5th of March we launched the documentary with an event led by well known journalist Paolo di Giannantonio (RAI TV national channel). The

patients that had helped us were present and also the doctors that had lent us their scientific supervision. A press release was sent to the media. At the moment it is possible to see the documentary both from our web site or directly from:

<http://vimeo.com/20636471>

We had very positive feedback from the media: an interview on Rai3. The video can also be seen on You Tube:

<http://www.youtube.com/watch?v=J0cjABeZQ90>

We have recently received an invitation to take part in a program on TV2000 (private channel). Our documentary will be broadcast next September on SKY TV and we have been invited to present it at the next Viareggio Health Festival that is an important event here in Italy. We are sure that all this visibility is going to bring good results.

*Luisa Sciacca della Scala, AMIP Italy*

[www.assoamip.net](http://www.assoamip.net)

### LATVIA

#### **A PAH patient organisation was founded in Latvia!**

Greetings from Latvia! Latvia is located at the Baltic sea in north-east of Europe. The population is about 2 million and the capital is Riga. PHA Latvia was founded in July 2011 by Ieva Plume, Zane Lazdina, Lolita Indriksone, Dr. Andris Skride and Dr. Ainars Rudzitis. Dr. Andris Skride and Dr. Ainars Rudzitis are coordinators and specialists for PH in Latvia. There are 30 patients with a diagnosis of PAH

in the Registry of Pulmonary Arterial Hypertension in Latvia. The first meeting of PHA Latvia will be held in August 2011 in Riga.

*Ieva Plume*



*The founding members of the Latvian PH association. Ieva Plume is second from left*

### NETHERLANDS

#### **A great man passed away**

To our grief, Wim Patist, the founder of PHA Netherlands died on Friday the 7th of January 2011. Wim and four other persons were the initiators of our foundation, which was created in 2003. With the support of Dr. Boonstra, Dr.



Vonk-Noordergraag and Dr. Berger, it was the start of a long-time drive to represent PH patients in the Netherlands and abroad. Wim Patist had always time and advice for his fellow-sufferers. He was the first chairman of the foun-

dation "Stichting Diagnose PH" in 2003. Unfortunately his health went downhill and he had to undergo a lung transplant in that year. For that reason he had to step down as chairman of the foundation. In 2004 the name of the foundation was changed to "Stichting PHA Nederland". Soon after an improvement of his health, Wim Patist became again Chairman of the foundation in 2006. In the following years many goals were achieved, including the Awareness Campaign in 2007, a website, events to meet other PH-patients and much more. As his health troubled him continually, he had to step down again as Chairman of the foundation. This he did with pain in his heart. On January 2009, Ferdinand Bolsius was elected as his

successor. Wim Patist remained available as advisor and as a never ending source of information. Besides his activities for our foundation, he also was an active Board member of an organization called "Harten Twee" (Two hearts).

In 2009 Wim Patist started the foundation "Organ Transplant Mediation", which is active in trying to improve the chances of a lung transplant for PH patients. Regrettably this foundation lost a very active member of the Board. Although we knew that Wim's health was deteriorating, his death came still as a shock to us. We all shall miss him, it will be strange not being able anymore to contact him for advice or for some pep-talk.

We thank Wim Patist for all he has done for PH patients, he has and will be an example for all of us. Wim Patist will always be remembered by us all.

### Yearly Patient meeting

On the 2nd of April we held our yearly meeting for patients in Amsterdam. Several speakers did a presentation about many aspects of PH. Professor Vonk Noordegraaf (see photo in the middle below) from the Vumc told us about the newest developments in research for new medicines and about better methods of diagnosis. In the VU Medical Centre they cooperate closely with other universities in the world. There was also a very interesting presentation from Dr.

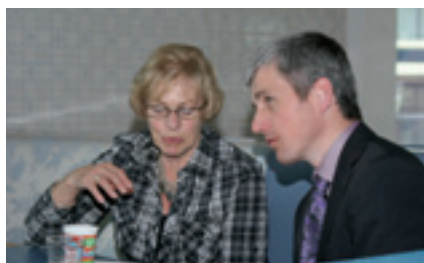
W. van der Bij, a lung specialist at the Medical Centre of Groningen, about all the difficult aspects of lung transplantation. He also mentioned the problem of the availability of lungs for transplantation in the Netherlands. At this moment we have a waiting period of more than three years. Especially for PH patients this is a big problem!

Of course we always take care to offer a good lunch and we like to close the meeting with a cheerful end of the day activity. This time we had a comedian who pretended to be a professor and the presentation ended in very funny chaos!

### Quality criteria for optimal care

In cooperation with two other lung patient organisations we are working on a plan to formulate quality criteria for the optimal care of patients. We try to know what kind of care the patients prefer to get, so they can live in an optimal condition of life. This project is not as simple as it seems to be. At this moment we have had two meetings with ten patients with the help of professional discussion leaders. Now they are making a survey for all the patients. At the end of this survey we hope to be able to produce a document which we can distribute to all the health care professionals who take care of the PH patients.

*Ferdinand Bolsius and Maaïke Atres, PHA Nederland*



*From left: lunch at the patient meeting, Prof. Vonk Noordegraaf meeting his patients and Maaïke Atres' mother selling postcards she has made herself to raise funds for the association*

[www.pha-no.com](http://www.pha-no.com)

## NORWAY

PHA Norway will have its first patient meeting in September on the 24th and 25th in a spa resort outside of Oslo. An invitation to the meeting has been sent to Norwegian PH-doctors who have been encouraged to forward the invitation to their patients. The main purpose of the meeting is to create an opportunity for patients to get acquainted, exchange information, and give and receive mutual support. Also on the agenda:

- A leading PH-specialist will lead a session about PH treatments in Norway.

- A representative from Germany will present the organization and activities of the German PH association as well as the development of future PH-drugs.
- The leader of Feiringklinikken, a cardiac rehabilitation clinic in Norway, and a psychologist on the staff, will present their program for PH patients.

The meeting has been made possible through grants from PHA USA and Actelion.

*Hall Skaara, PHA Norway*

[www.pha-no.com](http://www.pha-no.com)

## POLAND

### New website

The Polish PH Association (PPHA) has found a new sponsor who will contribute towards improving the association's website. The new website will have a better graphic layout, a platform which will help patients contact doctors, and an easier way to access the association. PPHA is in the process of finalizing the agreement with the sponsor.

### Meeting of National Forum of Rare Diseases' Treatments

On the 16th of June 2011 PPHA took part in a meeting of the National Forum of Rare Diseases' Treatments. The Polish association has become a member of this forum. The forum will be able to support associations with treatment issues, through advocacy and lobbying with officials and organizing meetings. A member of the PPHA had a conversation with journalists and gave away PH materials. The National Forum is planning a Congress of Rare Diseases on the 20th of September 2011.

### Workshops for journalists



PPHA took part in special workshops for journalists in four Polish cities: Szczecin, Lublin, Poznan and Wrocław. The main aim of these workshops was to educate the regional media.

Representatives of the regional public administrations were also invited to the meetings. The agenda of each meeting was the same - a doctor described the symptoms, diagnosis, and treatment of PH and a patient spoke about life with PH - about difficulties and fears. All the meetings were held in a public administration buildings and the workshops were held under the patronage of the cities' Mayors.

Participants received PH materials and a USB pen drive with the PPHA logo, and a special Polish cake - a blueberry cake with a little note saying: "This is not a lipstick or a blueberry cake!" Blue lips are a characteristic of PH.

After the workshop in Lublin, regional Polish TV showed a special program and film about PH. Two doctors spoke about PH in medical terms and two patients described their life with PH and their problems. The young reporter of this film, when he came back from a camerawork, said that he didn't realize that PH was such a serious disease.

The film is on the following website:

<http://www.tvp.pl/lublin/reportaze/zdarzenia-magazyn-reporterow/wideo/9-czerwca-2011/4660470>



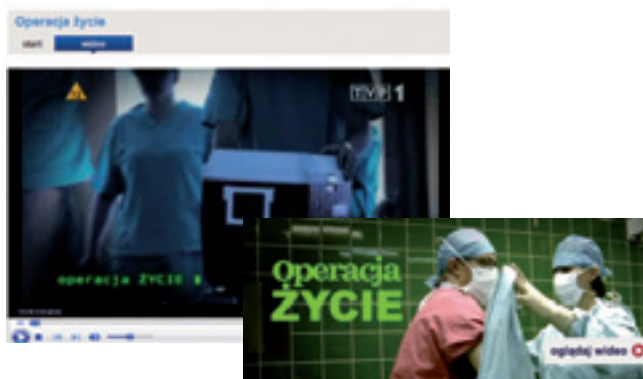
### General Assembly and patient meeting

PPHA is planning a General Assembly on the 3rd and 4th of September 2011. Members have to vote for the new board and other official positions. Members also will discuss future plans.

### "Life Surgery"

Polish TV showed a special documentary about transplantation called "Life Surgery". Thirteen episodes of "Life Surgery" showed how people live before and after transplantation. The authors wanted to show all the problems and difficulties related to organ transplantations. One part of this film is about PH. This part showed a young woman who was waiting for a transplant. Viewers could see how PH destroyed the women's life (the young woman has now had a transplant and her health is improving from day to day). The film is on the following website:

<http://www.tvp.pl/vod/dokumenty/spoleczenstwo/operacja-zycie>



Agnieszka Bartowieszc, PHA Polska



## SLOVENIA

**Patient Association set up**

I am very happy to inform our PHA Europe friends that in April 2011 the Slovenian PH patient association was formally established.

We have a Board of which I am President, the Vice President is Stanko Podbregar and our Secretary Maks Soršak.

We do not have a webpage yet but we have opened a Facebook page: *Pljučna Arterijska Hipertenzija*.

I really look forward to meeting you all in Barcelona in September!

*Klara Klančar, Društvo Za Pljučno Hipertenzijo Slovenije*

## SPAIN

It has been a long and fruitful year for the ANHP with plenty of activities and promises of new important projects that hopefully will extend to other PHAs improving the quality of life of PH patients. We have grouped these activities by subject, because a chronological account would be too heavy for the readers.

**Presence at Scientific Congresses**

*SEPAR winter edition in Córdoba*

SEPAR winter edition was in Córdoba where the ANHP President, Irene Delgado, and the Vice-President, Rosa García, held a press conference with the media. Dr. Balóira and Sara Toledano were there to present the medical and psychological aspects of PH. Irene made her presentation on how patients live with the disease.



*Irene Delgado and Rosa García at SEPAR*

The ANHP was present at three congresses during the first six months of 2011. SEPAR winter and summer are the biggest events for pulmonologists in Spain. The

At the SEPAR summer edition, held in Oviedo, we had our own stand, as in past editions. ANHP was the only patient organization present giving away materials and

information to specialists, promoting awareness of PH and motivating the clinicians to hand out information to their patients about ANHP.

During the congress, we also participated in the preparatory meeting for the RARE RESPIRATORY DISEASES YEAR OF SEPAR 2012. As usual, ANHP is the only association that has the

capacity to attend these meetings and make the voice of the patients heard. We also had the chance to finally meet Dr Jaime Morales, from Mexico, who, together

with Migdalia Denis, is the biggest defender of PH patients in Latin America. ANHP renewed its vows to support the Spanish speaking PHAs in that continent offering its guides and materials for their use.

The third Scientific Society, event that we were invited to, was NEUMOMADRID, a regional pulmonology congress. As ANHP coordinator, I presented the difficulties that rare diseases patients face in different regions of Spain. We have to highlight that it was the patients' intervention that received the most response because of its contents that raised awareness on the damage made to patients by the diverging policies of the decentralized health authorities.

And last but not least, we had our booth at the WEEK OF THE HEART organized by the SOCIETY OF CARDIOLOGY. This event is held in Plaza de España in Madrid and 20,000 visitors pass by the booths and get information that ranges from healthy food for the heart to the common arterial hypertension or rare diseases such as PH.



*ANHPs booth at SEPAR*

## SPAIN

### Follow up to the “Breathtaking” campaign

As you know through the special edition of the Mariposa News, ANHP organized events in the two major cities of Spain, Madrid and Barcelona. The placard posting in Barcelona Metro Stations with long staircases were displayed one more month than agreed and it hit the “three million views” by metro passengers!

### ANHP General Assembly

This year we organized the General Assembly in Aranjuez. Dr Pilar Escribano, one of the most renowned cardiologists treating PH at the 12 de Octubre Hospital, shared her knowledge with the patients and was highly appreciated.

### Genetic Study of PH

ANHP together with the 12 de Octubre Hospital will fund a genetic study on idiopathic PH. The project foresees the study of possible genetic mutations in 400 test subjects. The study will be conducted by the INGEMM, INSTITUTO DE GENÉTICA MÉDICA Y MOLECULAR. It is a two year project that will provide additional scientific evidence on the genetic origin of familial PH using state of the art equipment and the most innovative techniques.

### Training for Healthcare Professionals

Irene Delgado was involved in the training of nurses and physicians specialized in PH with her presentation “The Other View”, providing the insight of the patients and sensitizing healthcare professionals specialized in PH of four hospitals.

### Defense of patients regarding access to medication

ANHP has requested the pharmacovigilance of two generics of sildenafil after receiving allegations of clinical worsening since the beginning of its use. We have also made an intervention before the health authorities regarding the lack of use of prostacyclins in one hospital treating over eighty PH patients.

### Tom Lantos Innovation in Community Service Award: First Seminar on Psychological care for PH Patients

The Tom Lantos Award for Innovation in Community Service has been a turning point in the psychological care of PH patients in Spain. The Spanish PH association ANHP has always promoted the principle of considering the PH patient as a whole, not as just a “faulty” organ. After publishing the guide “Emotional aspects of PH”, there was need to step up and involve the Health Authorities in this holistic view of the disease.

The Gilead sponsorship of the Tom Lantos grant provided us with the opportunity of organizing the first Seminar on Psychological Care for PH Patients addressed to psychologists and psychiatrists in the public hospitals of Madrid.

As Irene Delgado said, “Medicines to treat the body are not the only need we have. PH patients must get emotional support to face the fear of our prognosis and we must have tools to reduce the challenge of this disease that often impedes access to a job or adjusting to the society. Psychological support is paramount for many patients in their struggle to adapt to their condition and make the most of their lives”. Psychological support is paramount for many patients in their struggle to adapt to their condition and make the most of their lives.

Getting the involvement of authorities was hard. It was painful to hear and actually see that because of the small number of patients PH is at the bottom of the priorities within the health system. However, unexpected strong support came from the Coordinator of the Regional Mental Health Office of Madrid and he made this seminar possible. On June 7, we gathered twelve psychologists and psychiatrists from the public hospitals and Dr Petersen was with them throughout the day.

ANHP has recently received a request from one of the public hospitals to fund a research on the impact of psychological therapy in reducing morbidity and mortality of PH. There will be one group of patients that will attend psychological group therapy, and those patients who will need it will be followed up by a psychiatrist on individual basis. Their progress will be compared with a control group, and we expect to see a measurable difference in their clinical evolution in favor of the patients with psychological support.

This first seminar on psychological care for PH patients made possible by the Tom Lantos Award is a milestone in the evolution of PH care.

“We have opened a door for new challenges, we are small and in dire needs of funds but we have hope and a strong will”, says Irene Delgado.

### Development of institutional capacities

Since the beginning of my activity within the ANHP, I have received training in Orphan Drugs and Rare Diseases (V INTERNATIONAL CONGRESS ON ORPHAN DRUGS AND RARE DISEASES in Seville), patients representation at European level (EURORDIS Summer School), and have been included in the pool of experts of the European Medicines Agency.

*Juan Fuertes, Asociación Nacional Hipertensión Pulmonar*

## SWITZERLAND - HTAP REVIVRE

### 1st French-speaking Congress for PAH Patients, Lyon

This Congress was organized by HTAP France in Lyon on the 9 and 10th of October 2010. We had the great pleasure to be invited by HTAP France to this meeting and the even greater pleasure of participating! The venue was wonderful and perfectly organized, with the rooms, meals, conferences and workshops all grouped together in a very nice residence in the middle of a large park. Oxygen had been provided for in the rooms and conference areas, there were meals without salt and nurses at hand at all times for patients on intravenous therapy... the organizers had thought of absolutely everything! The level of the conferences was extremely high and included presentations on:

- PAH in general: pathophysiology of the disease, diagnostic process, treatment strategies, clinical research, future therapies;
- specific issues: pediatric PAH, PH and congenital heart disease, PH and connective tissue disease, PH in respiratory diseases;
- surgery: double lung transplant and thromboendoarterectomy;
- genetic issues.

There were very interesting workshops on:

- every day life with specific therapies: intravenous, inhaled, oxygen;
- PH patient management and education, social aspects (specific to France);
- physical rehabilitation.

There were also workshops on the more personal aspects of PH, which were quite emotional and intense, and during which we had the opportunity of sharing and exchanging experiences on a deeper level:

- the worries of caregivers and people living with patients;
- PAH and men issues (dedicated to men only);
- PAH and women (for women only);
- psychological support;
- interpersonal relations;
- problems of persons who had undergone transplant.



*Prof. Marc Humbert, from the PH center at Clamart, France*

A very big thank you to HTAP France on our behalf! They invested a lot of time and energy convincing all the parties interested of the necessity of such an initiative and carried it out incredibly well. Thank you Sylvain, Mélanie and the members of the Board, thank you to all the volunteers who helped with the organization. Thank you to the doctors who believed in this project and who took part. There were 16 of us from Suisse romande taking part and we were all absolutely delighted!

### New contact details

**President:** Lydia Benallouch

**Vice President:** Edmond Dupertuis

**Secretary:** Marie-Régine Laub

**Coordinator:** Monika Sorge Maître

*Monika Sorge Maître, HTAP Revivre*



*The HTAP Revivre delegation at the 1st Francophone Congress for PAH patient, Lyon*



## SWITZERLAND - SCHWEIZER SPHV

### Annual General Meeting

After a successful year of activity our first ANNUAL GENERAL MEETING was held at the Congress Hotel in Olten on the 19th of March. At the centre of the well organised meeting was a speech by Professor Dr Rudolf Speich of the University Hospital of Zurich. Its subject was "Therapies for PH yesterday, today and tomorrow". A number of sponsors of the association also participated with stands: whoever was interested could acquire information through brochures, leaflets or by engaging in conversation with the representatives of the sponsor either after the conference or during the break.



Therese Oesch, President of SPHV, at the patient meeting in Olten

The event started with a welcome coffee during which participants could make contact, exchange experiences and get to know each other. The meeting was kicked off at 10 by an introduction by Therese Oesch, President of SPHV. She reported on the activity undertaken so far by the young SPHV.

She also gave a short overview of the planned events. The association has 117 members.

Thereafter began the conference by Professor Speich which was illustrated by a power point presentation. The speaker illustrated the different forms of PH and diagnosis. He explained that the latter is often marred by difficulties, since doctors were and are still far too unfamiliar with this condition and do not suspect PH (shortness of breath, normal X ray, misinterpretation of ECG, although typical alterations are present). Although in most cases the need for treatment is urgent, through incorrect diagnosis the adequate treatment can be delayed for up to two years or more,



Participants at the SPHV patient meeting enjoying lunch together

which worsens the prognosis for the patient. A heart scan allows to calculate the pressure difference between the ventricle and the atrium and the sum of vascular pressure allows to evaluate the possibility of PH, which however by itself does not make a definitive diagnosis possible. The examination by means of a right heart catheterization alone is acknowledged by the experts as the universal standard method to detect PH. The right heart catheterization is the most reliable method to diagnose PH, while other methods deliver additional valuable elements although they do not allow a reliable clarification eg echo as a screening method. The right heart catheterization together with a exercise stress test allows an early diagnosis. Specialists agree that PH starts with average pressure in the pulmonary arteries greater than 25 mmHg, more recently equal or greater than 20 mmHg.



Photos of the patient meeting in Olten. Franz Fischer, SPHV delegate at PHA Europe annual assembly, is first on the right in bottom photo

### Regional Meetings

In the course of 2011 we will organize at least three regional meetings in the Basel, Chur and Zurich areas. Through these meetings we intend to involve also those patients who cannot any longer leave home for too long.

### PH-Flyer

We now have a new leaflet on PH with our contact details. We have already distributed this leaflet to all patients and doctors in Switzerland. The leaflet is also available on our web page in PDF format. Please click on the following link:

[http://lungenhochdruck.ch/pdf/PPH-Flyer\\_1.pdf](http://lungenhochdruck.ch/pdf/PPH-Flyer_1.pdf)



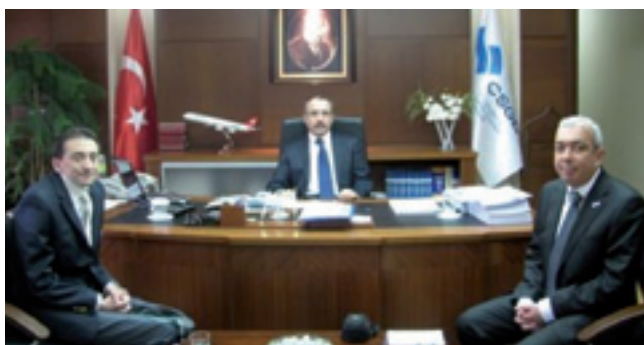
Bruno Bosshard, SPHV

## TURKEY

**Meeting with the Health and Social Affairs Minister**

In recent months we have had a meeting with the Minister of Health during which we conveyed the problems of our association and the PH patients. Our Minister has given instructions to some of his officials in order to try solving our problems. We had previously submitted a report explaining these difficulties. And now it has been decided to create a rare and orphan diseases department within the the Ministry. Our association and the head of this department will cooperate and we will lay the foundations with a pilot scheme.

Following this, we also met the Minister of Labor and Social Security during which we had the opportunity of explaining the problem of lack of payment for some medicines. Remodulin, with whose authorities we had met during the PHA USA congress in 2010 and which was then not available in our country, is now a repaid and approved medicine. We are now starting to iron out problems with Revatio. We started more facilitative works either by buying medicines or our treatment abroad, and promised to develop this activity.



The Minister for Labour and Social Affairs Ömer Dinçer with PHA Turkey President Hasan Saruhan (left) and Kamil Hamidullah

Our close relationship with both ministries continues. We will hold a coordination meeting once every 6 months.

**PH at the theatre or in films**

We have started to explore the possibility of including a play named "11 Eylül (1967/2007)" which is about the life of Ümit Atli, in the repertoire of state theatres. We are also meeting with potential sponsors in order to shoot a TV series or a worldwide film, if we can get support. The main objective of these initiatives are to raise awareness on the problems of patients with rare diseases and the importance of organ donation and organ transplantation.

**International Congress**

We have started pre-studies for our biggest project so far and we are intending to set to work this September. Our 1st International Congress will be held in September 2012 with participation of our fellow doctors from America, Europe and Israel; patients from America and Europe; our friends from the society (Associations), our doctors in Turkey; and in the seat of honor our patients and their relatives. We will be together with our worldwide family for 4 days in the south of our country. If it is approved, we also would like to host the Europe General Assembly in 2012.

**New Board**

**President:** Hasan Saruhan  
**Vice President:** Zeynep Tıraş  
**Treasurer:** Nuriye Yanıkoglu  
**Secretary:** Emine Çayıroğlu  
**Member:** Aslı Leman Atli

Ümit Atli, Pulmoner Hipertansiyon Derneği

[www.pha-turkey.com](http://www.pha-turkey.com)

## UNITED KINGDOM

**Recent activities of PHA UK**

In February PHA UK officially entered the GUINNESS BOOK OF RECORDS with its Blue Lips campaign and this is a wonderful result for the PH community worldwide. Many European patient associations took part and helped in collecting and we can all be proud. PHA UK has been very busy on other projects, such as the two

recent pieces of research it commissioned on the impact of PAH on the lives of people affected by the condition: the ImPAHct survey and "The lived experience of pulmonary hypertension". Other recent activities include a publication on the emotional sides of PH, "Listening to you, listening to me", the annual Family weekend and a charity concert and many more... Read about these activities on the latest edition of EmPAHsis, the official journal of PHA UK at <http://www.phassociation.uk.com>

Pisana Ferrari

[www.phassociation.uk.com](http://www.phassociation.uk.com)

### RUSSIA



The Charity Foundation “Natasha” is the only organization in Russia which helps patients with pulmonary hypertension.

#### PH treatments

To date, Russia has licensed only two drugs for the

treatment of pulmonary hypertension: iloprost/Ventavis and bosentan/Tracleer. However, the first drug is not widely used due to low awareness among physicians and specialists. Both medications are not included in the list of ‘soft’ drugs, which means that the citizens of Russia with PH have to buy them with their own money. It is very difficult, given that the wage level for a family is much less than the cost of one package of drugs.

Since early this year some patients with PH were able to obtain financing for the acquisition of bosentan/Tracleer from the government.

#### Lung Transplant

Perhaps the main news of the past six months is that a second successful double lung transplantation was conducted in our country. The first lung transplant was held

in August 2006, but due to unresolved legal issues, the continuation of such operations was delayed by more than 4 years. Carrying out lung transplant gives great hope for patients with pulmonary hypertension who are at the stage of acute decompensation. To date, several dozen patients with pulmonary hypertension are on the waiting list.

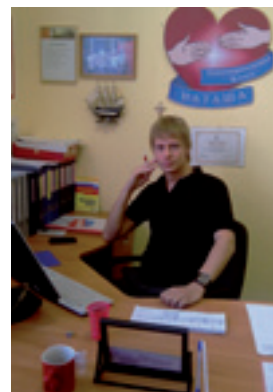
#### Registries

This spring, the Natasha Foundation has begun to create a single registry of patients with PH in Russia. Today 45 people are included. This is only a small portion of people with PH in our country, but we are actively working to expand the roster.

#### Web

Work has begun to set up a large internet resource on pulmonary hypertension. On this website, patients with PH will be able to find answers to their questions, learn about medications, medical centers, treatment options, understand the symptoms and diagnostic methods of examination. Also here they will be able to share experiences and support each other.

*Evgeniy Komarov, Natasha Foundation*



[www.nurm.ru](http://www.nurm.ru) - [www.community.livejournal.com/fond\\_natasha](http://www.community.livejournal.com/fond_natasha)

### LATIN AMERICA

#### Quarterly newsletter

The latest edition of the Latin America Society’s *Hap en Noticias* has been posted on the SLHP’s website. The newsletter contains reports from various national Latin American PH associations about their activities, a number



of very inspiring patient stories as well interesting scientific articles by leading PH medical specialists. Highlights of the last edition include the Mexican Association’s “Walk for hope” in January 2011 and first PH conference in Feb-

ruary 2011, news of two PH patient associations being set up in Guatemala and Salvador and of a new website established in Argentina. There is also a two-page special on the “Breathtaking” campaign in Europe, in particular in Spain, and a report of the PuckerUp4Ph campaign organized by PHA UK.

We have recently received news from Migdalia Denis, Pres-





ident of the Latin America PH Society, about recent legislation on rare diseases in Peru. This law, passed in June 2011, declares of national interest and gives special attention to the treatment of persons with rare and orphan diseases. Rare diseases are defined as those with a risk of death or chronic disability and which are difficult to diagnose and be followed up. These are in most cases not

known as there is little epidemiological data and have profound social implications. The law provides for the creation of a National Plan for the prevention, diagnosis, care, rehabilitation and monitoring of rare or orphan diseases. It establishes a national patient register.

*Pisana Ferrari*

[www.sociedadlatinahp.org](http://www.sociedadlatinahp.org)

## USA

### International Quarterly E-Newsletter



PHA created a quarterly International e-Newsletter for PH association leaders around the world. The first issue was sent out in June 2011 and featured stories on the "Breathtaking" awareness campaign, Israel's Conference on Pulmonary Hypertension and a news story on the role of social media in the PH community in the aftermath of the Japanese earthquake.

If you have not received this newsletter and are interested in receiving future issues, please email:

[International@PHAAssociation.org](mailto:International@PHAAssociation.org)

### PHA Online University and Advances in Pulmonary Hypertension

PHA Online University is an online resource for medical profes-



sionals seeking education about pulmonary hypertension. With new courses and live webinars every month, information about research,

treatment and diagnosis is available at your fingertips at the PHA Online University: [www.PHAonlineuniv.org](http://www.PHAonlineuniv.org) Advances in PH, a medical journal put together by leaders in the PH field, can also be found on the PHA Online University. The latest two issues of Advances in PH focused on Ethical Issues in PH and Group II PH: Striving in Understanding. This journal is distributed to cardiolo-

gists, pulmonologists, rheumatologists and other healthcare professionals worldwide. All of the issues are available online at: <http://www.phaonlineuniv.org/Journal> or you can subscribe for free by contacting: [Medical@PHAAssociation.org](mailto:Medical@PHAAssociation.org)

### PHA Classroom

PHA Classroom is an e-learning tool for patients and caregivers that allows visitors to participate in live events or watch recordings of educational sessions. Topics range from information about PH to tips on living with the disease. Visit [www.PHAAssociation.org/eLearningEvents](http://www.PHAAssociation.org/eLearningEvents) to learn more about this resource.

### PH Clinicians and Researchers (PHCR)

PHCR is a professional medical membership network of over 600 PH-treating physicians and researchers. PHCR currently allows doctors to engage in case-based learning over a listserv and list their practices in a patient Find a Doctor Directory, among its other benefits.

### Save the Date!

The 10th International Pulmonary Hypertension Conference and Scientific Sessions will take place June 22-24, 2012 in Orlando, Florida. The Second International Leaders' Summit for PH association leaders will occur on June 21, 2011. More information about PHA's Conference can be found here: [www.PHAAssociation.org/Conference](http://www.PHAAssociation.org/Conference)



*Meghan Tammaro, PHA*

[www.phassociation.org](http://www.phassociation.org)



The COUNCIL OF EUROPEAN RARE DISEASE FEDERATIONS (CEF) is a platform set up by EURORDIS\* which brings together European rare disease federations or networks. The aim is to foster the exchange of experiences and information. It was a great honor for PHA Europe to be invited to attend and speak at the 4th CEF Workshop in Paris on the 30th of June and 1st of July. PHA Europe had been asked to present its “Breathtaking” campaign as an example of a successful awareness raising initiative around Rare Disease Day (28 February 2011). The “Breathtaking” campaign was held with the support and patronage of EURORDIS.

The CEF workshops aim to enhance and build the capacities of the European federations. The agenda of the 4th workshop, held in Paris on the 30th of June and 1st of July 2011, included an overview of Rare Disease Day 2011, a report on the relevance of the recent EU directive on cross border healthcare and as presentations on some of the tools designed by EURORDIS to empower the federations: e-learning and online patient communities.

It was extremely interesting for me to meet and talk to the delegates from other rare disease federations such as our own, to see how they are organized and work, and share some of our experiences, successes and difficulties. Sixteen rare diseases were represented at the workshop including osteogenesis imperfecta, dysmelia, cystic fibrosis, epidermolysis bullosa, ichthyosis, retina disorders, renal genetic diseases and a number of neurological conditions such as ataxia, hereditary spastic paraplegia, Niemann Pick.

## Rare Disease Day 2011

The meeting opened with Paloma Tejada from EURORDIS who presented the main achievements of Rare Disease Day 2011. This year's edition saw the active involvement of 56 countries, 22 of which from Europe and the others from the rest of the world, thus making it a truly international event. Newcomers included Mexico, Thailand and Panama. An incredible success considering it started out only 4 years ago in 2008 with 18 countries participating. On the 28th of February there were almost 50,000 visitors from 150 countries on the EURORDIS website with 10,000 on Rare Disease Day only. There was a lot of activity also on the social networks with 14,000 ‘friends’ for the official Facebook page. Next year Rare Disease Day will be celebrating its 5th anniversary and EURORDIS its 15th so big celebrations are being planned. Themes for the event change every year and next year's is “Solidarity”: working together for common



Anja Helm from EURORDIS with Balthasar Schaap of the European Federation of Hereditary Ataxias



Pisana Ferrari with Christoph Poincilit from the International Niemann-Pick Disease Alliance



Presentation on advocacy for



goals. It has now been decided to start the groundwork necessary to get Rare Disease Day recognized as an official day in the UN Calendar.

### PHA Europe's "Breathtaking" campaign

In the PHA Europe presentation the campaign's organizational aspects were described in detail from the very first steps with the national associations to the Brussels and Vienna launch events with key medical opinion leaders and press and the awareness raising activities carried out in another ten countries across Europe. Particular emphasis was laid on the achievements of the campaign in terms of:

- extensive dissemination of information and media coverage across Europe;
- increased visibility for the federation (institutions, medical, industry, etc.);
- enthusiastic involvement of national associations;
- empowerment of the national associations (even the "small" associations can do a lot!);
- building up of community spirit.

The PHA Europe slides have recently been posted on the EURORDIS website.

<http://www.eurordis.org/content/council-european-rare-disease-federations>

### EU Directive on cross border health care

Yann Le Cam, EURORDIS' C.E.O., gave a very interesting overview of the recently approved EU Directive on cross border health care. This directive, published in the Official Journal in April 2011, is binding for the Member States who now have 36 months to transpose it into the national law. The new legislation will have very significant implications for all patients across Europe as it will allow patients to seek medical treatment outside their main country of residence (within the EU) and be reimbursed. We wrote a brief report on this in the winter 2010 edition of Mariposa.

Mr Le Cam's presentation focused on the relevance of the Directive to rare diseases. First of all rare diseases are now mentioned in a European binding legislative text and this is an important step forward (Council Communications or Recommendations are not binding eg. the 2009 Council Recommendation on Rare Diseases). The Directive promotes EUROPEAN REFERENCE NETWORKS and specifies that these are especially important for rare diseases. Another point is the special provision for RD patients to facilitate diagnosis in other countries. Mr Le Cam warned that the national transposition of the directive will be difficult and there is a risk that each country will do something different. Coordination is very important. At the first level EUCERD\*\*, a EU rare disease expert committee where patient representatives are present, is working on guidelines on how to transpose the legislation nationally. Quality criteria for health care are also very important and EUCERD is starting to build on this too. Mr Le Cam also said European federations should monitor closely the work of European Reference Networks and follow-up on patient mobility by reporting examples of patients not being reimbursed for treatment received in another country as they should be according to the terms of the Directive.

<http://register.consilium.europa.eu/pdf/en/11/pe00/pe000006.en11.pdf>

### Clinical trials register

The EUROPEAN CLINICAL TRIALS REGISTER is an existing register (it was set up in 2011) which has become public since March 2011. Francois Houyez, EURORDIS Health Policy Director, reported on this new and very useful tool for European rare disease associations. The clinical trials website contains information on the design of each clinical trial, the sponsor, the investigational medicinal products and therapeutic areas involved and the status of the clinical trial (it is compulsory for the sponsors to enter their data in the register).

<https://www.clinicaltrialsregister.eu/>



clinical trials in rare diseases



Dennis Costello presenting the Eurordis Rare Diseases Communities project



from left: Rob Camp, one of the speakers, with Peter Finney of the European Myasthenia gravis association



## The Rare Disease Communities project

Dennis Costello, a web communications manager, presented this very interesting project that provides wonderful networking and information sharing opportunities for rare disease patient organizations worldwide. The RARE DISEASE COMMUNITIES is an online social network which started out in 2009 as a joint initiative of EURORDIS and NORD, the US organization for Rare Diseases. The project is financed by European Commission, the French Telethon (AFM) and the French Federation of Pharmaceuticals (LEEM) and other corporate partners.

The main objectives are:

- collaboration amongst patient groups;
- access to quality information;
- networking/connecting people with similar interests;
- raising awareness;
- sharing patient data (gain insight on from aggregate data).

Ten rare disease communities have already been created and are online and ten more are in the pipeline. Mr Costello took us through a few of these communities (Duchenne disease, Marshal Smith Syndrome, hereditary angioedema) and showed us how the network works. The website is split into three sections: Understand, Meet and Learn. The part I found most interesting is the Meet section, a forum moderated by volunteers, offering a translation service. This is a great opportunity and privilege as the language issue is a major one for all federations or networks such as our own. For practical reasons PHA Europe has adopted English as its working language but this obviously limits the number of people who can take part in our activities, read our publications and use the information on our website. At our annual leaders' meetings we have to ensure that if the President of the national group does not understand English he brings someone along to translate for him/her. Language would be a major obstacle if we ever decided to start up our own forum. The RARE DISEASE



COMMUNITIES forum offers translation services in 5 languages: English, French, Spanish, Italian and German. If a user sees a message on the Forum that is not in one of these languages he can ask for a translation to be made. Within a few hours, the message is translated and uploaded automatically to the website. The user having made the request also receives the translated content via e mail.

The other two sections of the RARE DISEASE COMMUNITIES website are also very useful and interesting. The Understand section features patient stories and photos and the Learn section contains information in the form of FAQ's, documents, recently published scientific articles, updates on upcoming.

The RARE DISEASE COMMUNITIES platform is definitely something that PHA Europe should think of joining in the very near future!

Mr Costello ended his presentation by telling us a bit about the new online patterns in disease management. Patients and families are increasingly:

- searching online;
- using Facebook and Twitter;
- making decisions based on what they find online;
- setting up their own communities.

The web itself is evolving and it is estimated that 34% of all referral traffic is about sharing (blogs, bookmarking, social networks, etc.). The risk of misinformation is high and patient associations have the opportunity of playing a quality assurance role. At the same time the visibility of rare diseases is pretty good but could be much better. Enabling a patient community provides value.

<http://www.rarediseasecommunities.org/en>

## E-learning

EURORDIS strongly believes in the empowerment of patient representatives. Its training programs and resources are designed to strengthen the capacity of rare disease patient representatives to take part in clinical trials and become involved in the drug development and regulatory process in Europe. This was possibly the most interesting topic of the workshop.



The training program is something all patient organizations should encourage their leaders and members to explore and use. Francois Faurisson, EURORDIS Clinical Research Advisor, presented the program to all the delegates at the workshop, who were connected to internet and could follow online from their own pc. The first topic we looked at was “methodology” and this covered:

- study design
- controlled trials
- randomization
- endpoints
- criteria of inclusion
- analysis of results.

For each of these topics there is a “lesson”, then a section with definitions of all the key terms, another with a few examples of real clinical studies carried out and, finally, a quiz where one can test oneself. At all times during the training session one can consult the very comprehensive glossary. We all had the opportunity of trying it out personally.

Another very interesting section is the one on the ethics of clinical trials, where issues such as the principles involved, the autonomy and respect of the person, informed consent and ethics committees are discussed in detail. At the end of this session there is a quiz as well.

<http://www.eurordis.org/training-resources>

## Conclusions

I found this meeting to be extremely interesting and useful. EURORDIS is doing some really great work in the field of rare diseases in many different ways through its awareness campaigns, the representation of patient interests, advocacy, dissemination of information, training and empowerment of patient organizations. I was impressed by the commitment and dedication as well as the professional skills of all the persons working for the organization and presenting at the workshop. PHA Europe and indeed all rare disease patient organizations have much to gain from a close collaboration with EURORDIS



and, through EURORDIS, with other organizations across Europe. For the official report and all the slide presentations see:

<http://www.eurordis.org/content/council-european-rare-disease-federations>

## Notes

\* EURORDIS is a non-governmental patient-driven alliance of patient organizations representing 479 rare diseases patient organizations in over 45 countries.

More info at [www.eurordis.org](http://www.eurordis.org)

\*\* EUCERD is the EUROPEAN UNION COMMITTEE OF EXPERTS ON RARE DISEASES. It was established in 2009 and is charged with aiding the European Commission with the preparation and implementation of Community activities in the field of rare diseases, in cooperation and consultation with the specialized bodies in Member States, the relevant European authorities in the fields of research and public health action and other stakeholders acting in the field. More information at <http://www.eucerd.eu>

Pisana Ferrari

## Next PHAE newsletter

The Winter 2011 issue of Mariposa News is due out in mid-December. The first section will contain reports of activities of the national associations from September to December 2011. **We would be very grateful if all the member associations could send their contributions by the end of October 2011.** We can provide for translation into English if necessary.

Any photos sent should be in the highest resolution possible (minimum 1 MB). Please remember to send them as a separate attachment and not as part of a Word document. Many thanks in advance!

Mariposa News will be circulated in PDF format to all our member associations who will also be receiving twenty copies of the printed version by ordinary post. Anyone interested in being included in our mailing list for the PDF or in receiving printed copies can contact Pisana Ferrari at [p.ferrari@phaeurope.org](mailto:p.ferrari@phaeurope.org)



### **Pulmonary Hypertension: Leading the way to the future Lisbon, 12-13 March 2011**

The 6th BAYER SYMPOSIUM ON PH was held in Lisbon on 12-13 March and I was kindly invited to speak about "The relevance of patient organizations for patients". About 360 persons from 37 countries attended the event which featured a faculty of world-renowned PH medical specialists. Many of these I had already had the pleasure of meeting at the ESC, ERS and other international congresses: Professors Robert Naeije and Marion Delcroix (Belgium), Nazzareno Galiè (Italy), Marius Hoepfer and Ekkehard Grünig (Germany), Horst Olschewski and Irene Lang (Austria), Pavel Jansa (Czech Republic), Adam Torbicki (Poland), Miguel Angel Sanchez (Spain). The symposium was divided into three parts, the first of which about non PAH conditions such as chronic thromboembolic pulmonary hypertension (CTEPH), chronic obstructive pulmonary diseases (COPD) and interstitial pulmonary diseases (ILD), for which at present no satisfactory treatments exist. Some cases of CTEPH are potentially curable with pulmonary endoarterectomy (PEA) but the management of non operable CTEPH still remains a challenge.

The longer term perspectives for PAH patients are also a still challenge and this was the topic of the second part of the symposium. Despite progress in diagnosis and treatment in the last 15-20 years PAH is still devastating and has a very poor prognosis. One of the presentations, by Dr. Ioana R. Preston (USA) was about combination therapies. These are now widely used but many questions still remain, including whether this approach is more effective than mono-therapy, which combinations should be used and how, and finally if they are cost effective. In theory combination therapy has at least two advantages over mono-therapy: the drugs used target different pathogenic pathways and the use of more than one drug can overcome dose-limiting toxicities of individual agents. Most combination studies up to now have involved add-on therapies and it is not yet clear whether starting upfront is more beneficial. Dr. Preston pointed out that starting with two drugs upfront has certain disadvantages:

- patients might have done well on mono-therapy alone;
- unwillingness to de-scale if patients do better;
- costs.

My own presentation had been included in a session about personal aspects of PH and in particular PH patient needs. During this session two very interesting presentations touched upon issues that are not often addressed: the psychosomatic aspects of PH and the relevance of patient training.

The first presentation, by Prof. Kai-Uwe Kuhn (Germany), focused on the incidence of anxiety and depression among PH patients and the fact that these are underdiagnosed and under-treated. It is generally assumed that when seriously ill persons show symptoms of depression these are to be considered "normal" reactions to a very serious and life-threatening physical condition. As a result of this the quality of life and even the prognosis of patients can be significantly affected. Prof. Kuhn believes that antidepressant treatments can in fact lead to a better ability to take part in physical exercise and lead to an increased will to live better. He concluded by saying that these treatments can be very effective and well tolerated if administered correctly.

In his talk about exercise training Prof. Ekkehard Grünig spoke about a study he has conducted on 30 patients in WHO class II-IV on stabilized on optimal treatment over a 6 week period. This study demonstrated significant improvements in the 6 minute walking test, in quality of life, and functional class. In a subsequent study on 58 patients covering a 2 year period it was possible to see the longer term effects of exercise training. Here again the results were very good, with better survival and longer time to clinical worsening in the group exercising with respect to the control group. Prof. Grünig stressed that exercise is well tolerated when patients are closely supervised but further investigation is necessary for out-patient settings in order to establish whether it is safe.

The aim of my presentation was to explain the vital role that patient associations can play in providing information, support and networking opportunities to patients and family members and in furthering the cause for patients on a more general level. I gave examples of the different types of educational material and services that most patient associations offer their members through telephone help lines, websites, message boards, newsletters, publications, audiovisual materials, patient meetings, social activities, professional psychological support, legal advice for insurance and pensions, financial aid, etc. On the more general level I spoke about how patient associations can represent patients' interests and contribute to raising awareness, both nationally and at the European level. I took the opportunity to show some slides of our "Breathtaking" campaign for Rare Disease Day 2011. One of the points I raised is that optimal patient management can be gained only through collaboration between all the stakeholders involved and in particular with the medical professionals. A very positive development I mentioned was the fact that the current ESC/ERS GUIDELINES on PH specifically encourage medical professionals to work with the patient associations.

The third and last session of the Symposium looked to the



future of PAH management with presentations on future therapies currently being studied and on the importance of registries. On the first topic Thomas Pulido (Mexico) and Ardeschir Ghofrani (Germany) reported on ongoing research on PAH, which is looking very promising: cell and gene therapies, tyrosine kinase inhibitors (imatinib), soluble guanylate cyclase (riociguat). Dr. Tomas Pulido (Mexico) ended his talk on a positive note by saying that novel treatments are progressing as the knowledge of the disease increases and whilst maybe not all the potential therapies will be effective on humans, some will definitely go on to reduce mortality from this disease.

The importance of registries was highlighted by Dr. Abilio Reis (Portugal) who said that the huge costs of PAH in terms of treatments and surgical options demand an active engagement of providers, payers and health authorities, providing them with data for the necessary regulatory processes. New information technologies can now help physicians to build up data bases with epidemiological and clinical information which in turn can be used to create registers.

I would like to end by thanking Bayer for giving this great opportunity to address important patient issues at such a prestigious event.

*Pisana Ferrari*

*Photo at beginning of article, from left: Malcom Allison from Bayer, Prof. Robert Naeije, Pisana Ferrari and Prof. Marius Hoeper.*



**First Austrian Expert Forum on PAH-CHD  
Vienna, May 6-7 2011**

PHA Europe President, Gerald Fischer was invited to speak at the Actelion FORUM FOR CONGENITAL HEART DISEASE AND PULMONARY ARTERIAL HYPERTENSION which was held in Vienna on 6-7 May 2011. He was asked to speak about "The importance of a strong European PH patient association" and he was delighted to have the opportunity to address this prestigious and very interesting meeting.

Mr Fisher began his presentation by reminding the audience that despite considerable progress in diagnosis and treatment

PH is still an incurable disease with a very poor prognosis. Patients are even now being diagnosed with up to two years delay and start being treated late. Earlier access to treatment could improve prognosis significantly. Not all patients in Europe have access to approved drugs and surgery facilities also vary from country to country. Much remains to be done in order to achieve optimal PH patient management for all patients in Europe.

Patient associations can provide invaluable information, support and networking opportunities for patients and, on a more general level, promote the cause of the PH community as a whole through advocacy, lobbying and awareness campaigns. More awareness about PH is important not only for the patients themselves ("finding" the patients, earlier treatment and better diagnosis) but can also impact significantly on medical professionals, national governments and EU institutions. Many countries in Europe still have no patient associations at all or small groups that need to be further consolidated. Mr. Fischer stressed the fact that all patients should be able to benefit from the support provided by patient associations and that joining forces across borders is vital in order to reach critical mass, PH being a rare disease. Patient associations can also greatly benefit from working together at European level as this involves sharing problems/achievements and learning from each other. Different cultural backgrounds across Europe provide a wide and enriching range of views and unique opportunities for debate about new ideas and future strategies.

The key message Mr. Fischer wished to put across in his talk is that patient associations play a crucial role in providing correct information about the disease, treatments options, surgery, day to day life with the disease. This is all the more necessary in the era of internet where massive amounts of information are readily available and patients are increasingly searching the web to find out about their disease (in this respect they are more "mature" than in the past, more proactive). Doctors often do not have the time for lengthy and detailed explanations. The information patients find on the web can be difficult to understand, confusing or even incorrect. A website set up by a patient association with updated, correct, clear and concise information in easy-to-understand language is a vital tool for patients and can also become a "neutral" platform for industry, institutions, doctors and nurses. Patients should be informed of all treatment options. Quality of life is very subjective and treatments should be adjusted according to individual needs and preferences. Ultimately it should be the patients deciding which drugs they prefer! Mr. Fischer concluded his talk by saying that the goal of PHA Europe is that PH become the example of a well organized disease community.

*Pisana Ferrari*

*Photo at beginning of article: Prof Irene Lang (Austria) at the Expert Forum (1st from left on the podium).*



**International PH Nurse Workshop  
Dresden, June 17-18**

It was a great honour for me to be asked to speak on behalf of PHA Europe at Bayer's 2ND INTERNATIONAL PH NURSE WORKSHOP held in Dresden 17-18 June 2011. About 130 nurses and health care professionals from 14 countries were present at this two-day event. The agenda covered three main topics: the science of PH, the way patient care is organized in different countries and how this care can be improved in future.

The nursing profession plays a vital role in the management of PH as I have experienced personally both as a PH patient and as a patient representative. Persons suffering from a chronic and progressive disease such as PH require regular monitoring and frequent visits to the PH clinics. Over time relationships tend to build up with health care professionals. Nurses provide invaluable support in terms of patient education and training and this also has positive effects in terms of compliance to treatments and on outcome.

It was extremely interesting to hear from nurses in Spain, France, Sweden, Poland and the United States, how patient care is organized in these countries. It emerged from the presentations that the level of patient care in all of these countries is very high, with great attention being placed on individual situations and needs.

Most nurses worked in multidisciplinary contexts with other healthcare professionals. All showed concern and sensitivity for patients facing critical situations such as the moment of diagnosis, adjusting to changes in therapy, learning to use new treatments, as well as for quality of life issues. It was touching to see their dedication and commitment, and to hear some speak of the great emotional burden involved.

It was also interesting to hear about the varying levels of responsibility of nurses in the different countries, the United Kingdom being the most "advanced" in this respect. Wendy Gin Sing, specialist nurse from Hammersmith Hospital Imperial College explained that in the UK nurses can define/change treatment strategies for patients and prescribe drugs.

Deborah McCollister, from the University of Colorado Denver PH Clinic, touched upon another key issue for PH patients, the incidence of depression. A recent US study she quoted shows that 55% of PH patients suffer from

moderate to severe depression and that half of these patients were not receiving any treatments to help them in coping. In her view this is an issue that needs to be further addressed.

As a patient representative I was delighted to hear that nurses generally work well with patient associations and in some instances collaborate on the production of educational material. Iain Armstrong is Nurse Consultant with the Royal Hallamshire Hospital, Sheffield, and is also Chairman of the UK patient association PHA UK. He was not present but Wendy Gin Sing delivered his talk about the development of educational material for patients. Iain believes health literacy is a key issue which can improve outcome (compliance, side effects, what to look out for!). Unfortunately, for a number of reasons which include age, background and educational opportunities, the health literacy of most patients is quite low. Materials should take this into account and be written in simple, clearly understandable language, with attention to colours and graphic layout. Generally, they should encourage correct modes of behaviour and build on life experience.

My brief for the workshop was to speak about "Living with new lungs". In my presentation I analyzed the different steps leading up to the transplant:

- *Before:* having end stage disease, the screening process and being listed.
- *After:* the post operative stage, going home, starting life again.
- *Now:* 9 years later, leading a normal, active life.

I tried to bring out all the different aspects, which include many organizational difficulties as well as the more emotional sides. As anyone who has gone through it knows the entire process of going through transplantation is very traumatic and requires practical and psychological support as well as good personal coping skills. It is a very valid therapeutic option which can really offer a new life to PH patients, but currently the shortage of donors and subsequent long waiting lists do not make it accessible to all those who are eligible. Much remains to be done in this respect to raise awareness and promote organ donation. More could be done also in terms of the education of patients who are often left to their own devices when 'moving' from the PH clinic they are familiar with, to a 'new' transplant centre. They would greatly benefit from being 'bridged' from one to the other and from having a better understanding of what they will be going through.

I would like to end by thanking Bayer HealthCare Pharmaceuticals for their kind invitation and for promoting and supporting this event.

*Pisana Ferrari*

*Photo at beginning of article: Deborah McCollister (USA) speaking at the Nurse Workshop.*

The most important news in recent months in the area of public health to report is the final approval of EU legislation on cross-border healthcare\*. The EU PARLIAMENT AND COUNCIL DIRECTIVE ON THE APPLICATION OF PATIENTS' RIGHTS IN CROSS-BORDER HEALTHCARE is binding for the EU Member States who must transpose it into national law by 25 October 2013. Pending final approval, we had reported on this issue in the MARIPOSA NEWS winter 2010 edition.

This EU Directive provides rules for facilitating the access to safe and high-quality cross-border healthcare and promotes cooperation on healthcare between Member States, in respect of national competencies in organizing and delivering healthcare. Despite its limitations there is no doubt that the approval of this directive represents a major step forward for all European patients. The new directive contains the following provisions (Source: *EU Council press release*):

- as a general rule, patients will be allowed to receive healthcare in another member state and be reimbursed up to the level of costs that would have been assumed by the member state of affiliation, if this healthcare had been provided on its territory;
- instead of reimbursing the patient, member states of affiliation\*\* may also decide to pay the healthcare provider directly;
- for overriding reasons of general interest a member state of affiliation may limit the application of the rules on reimbursement for cross-border healthcare;
- member states may introduce a system of prior authorization to manage the possible outflow of patients; a member state of affiliation may refuse to grant prior authorization if the patient seeking cross-border healthcare will be exposed to an unacceptable safety risk, if the general public will be exposed to a substantial safety hazard, if the healthcare is to be provided by a healthcare provider that raises serious concerns relating to compliance with standards and guidelines on quality and safety, or if the healthcare can be provided on its territory within a medically justifiable time-limit.

- in order to manage incoming flows of patients and to ensure sufficient and permanent access to healthcare within its territory, a member state of treatment may adopt measures concerning access to treatment where this is justified by overriding reasons of general interest;
- member states will have to establish national contact points that must provide patients with information about their rights and entitlements and practical aspects of receiving cross border healthcare;
- cooperation between member states in the field of healthcare has been strengthened, for example, in the field of e-health and through the development of an European network which will bring together, on a voluntary basis, the national authorities responsible for e-health; another example is rare diseases, where the Commission will have to support member states in cooperating in the field of diagnosis and treatment capacity;
- the recognition of prescriptions issued in another member state has been improved; as a general rule, if a product is authorized to be marketed on its territory, a member state must ensure that prescriptions issued for such a product in another member state can be dispensed in its territory in compliance with its national legislation;
- sales of medicinal products and medical devices via internet, long-term care services provided in residential homes and the access and allocation of organs for the purpose of transplantation fall outside the scope of the directive.

\* Directive 2011/24/EU of the European Parliament and Council of 9 March 2011 on the application of patients' rights in cross-border healthcare - Official Journal of the EU 4.4.2011: <http://eur-lex.europa.eu/LexUriServ/LexUriServ.do?uri=OJ:L:2011:088:0045:0065:EN:PDF>

\*\* Country of affiliation: art.3. b (i) and (ii) and (c) of the directive define the persons to whom the directive applies and the meaning of country of affiliation.

*Pisana Ferrari*

## Update on PH treatments and research

### Clinical trials register now public

The European Clinical Trials Register, established in 2011, has become public since March 2011. The clinical trials website contains information on the design of each clinical trial, the sponsor, the investigational medicinal products and therapeutic areas involved and the status of the clinical trial. This is an extremely useful tool for patient associations in monitoring ongoing research and informing their members about recruiting opportunities. A search on "pulmonary arterial hypertension" shows 85 documents (date of search 8.8.2011).

The website is hosted by the European Medicines Agency.

[www.clinicaltrialsregister.eu](http://www.clinicaltrialsregister.eu)

### New indications for approved PAH treatments

On 17 March 2011 the Committee for Medicinal Products for Human Use (CHMP) of the European Medicines Agency adopted a positive opinion recommending a variation to the terms of the marketing authorization for the medicinal product Revatio (EMA/CHMP/207927/2011). The indication for Revatio now includes paediatric patients aged 1 year to 17 years old with pulmonary arterial hypertension. Revatio had previously received authorization for adult patients with pulmonary arterial hypertension classified as WHO functional class II and III.

<http://bit.ly/oV7S3H> (shortened link).



## Upcoming events for 2011

**27-31 August, Paris, France**

### **ESC ANNUAL CONGRESS**

PHA Europe will be present as an exhibitor at the EUROPEAN SOCIETY OF CARDIOLOGY'S ANNUAL CONGRESS 2011. This Congress is currently the largest cardiovascular medicine meeting in the world. It covers all disciplines, from basic research to clinical practice. Around 30.000 persons took part in the 2010 ESC Congress in Stockholm, Sweden.



**8-11 September, Barcelona, Spain**

### **PH EUROPE ANNUAL GENERAL ASSEMBLY**

PHA Europe is holding its ANNUAL GENERAL ASSEMBLY in Barcelona September 8-11. At the time of going to print, 45 PH patient leaders from 22 countries, have confirmed their presence at the meeting: Austria, Belgium, Bulgaria, Czech Republic, France, Finland, Germany, Hungary, Israel, Italy, Latvia, the Netherlands, Norway, Poland, Romania, Russia, Slovenia, Slovakia, Spain, Sweden, Switzerland, Turkey. The meeting has a very wide ranging program with interactive sessions about issues of common interest and presentations by distinguished speakers from the medical and nursing professions, the pharmaceutical industry as well as leaders of the UK and US PH patient associations (PHA UK and PHA USA). During the meeting PHA Europe will also be holding elections as the current Board's mandate ends December 2011. This photo is from the 2010 Annual General Assembly.



**24-28 September, Amsterdam, The Netherlands**

### **ERS ANNUAL CONGRESS**

PHA Europe will also be present with a booth at the EUROPEAN SOCIETY FOR RESPIRATORY MEDICINE'S ANNUAL CONGRESS in Amsterdam. This Congress is the world's broadest respiratory gathering. It involves not only leading experts in respiratory medicine, but also in other areas, covering the entire spectrum of research and practice in respiratory medicine. Over 18.000 persons attended the 2010 edition in Barcelona. During the ERS PHA Europe will be holding an EXPERT ROUNDTABLE MEETING with key medical opinion leaders, members of the nursing profession, representatives of patient organizations and the industry. The aim of this meeting is to work towards a common policy statement on PH (White Paper project). PHA Europe will also be attending meetings of the ERS Educational Task Force "GPs meet Rare Lung Disorders" and of the European Lung Foundation Patient Advisory Committee.



**14-15 October, Sophia Antipolis, France**

### **ESC EDUCATIONAL COURSE ON PULMONARY HYPERTENSION**



This course is organized by the ESC WORKING GROUP ON PULMONARY CIRCULATION with its target audience being largely cardiologists, pulmonologists and other specialists involved in PH care. This year the course will address gaps of knowledge, of evidence and of organization. It will be followed by the Annual G5 Meeting of the main stakeholders involved in PH management: physicians, nurses, patients, regulatory authorities and pharmaceutical industry ("G5"). PHA Europe has been asked to attend and present the PH Patient perspective.

## **NEXT YEAR**

### **2012 AT A GLANCE**

#### **Summary of main events:**

- EUROPEAN CONFERENCE ON RARE DISEASES, Brussels, Belgium, May 23-25
- PHA USA INTERNATIONAL CONFERENCE, Orlando, Florida, June 22-24
- EUROPEAN SOCIETY OF CARDIOLOGY, Munich, Germany, August 25-29
- EUROPEAN SOCIETY OF RESPIRATORY MEDICINE, Vienna, Austria, September 1-5
- PHA EUROPE GENERAL ANNUAL ASSEMBLY (date and venue to be defined)

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## AIMS OF PHA EUROPE

Pulmonary 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.



- a. Cross section of normal pulmonary artery
- b. Reduced lumen of pulmonary vessels due to cell proliferation and remodelling in advanced Pulmonary Arterial Hypertension
- c. 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.