

## 50 years of DNA structure discovery

In 1953 the structure of DNA, double-stranded helix chains, was described. These molecules, found within the nucleus of each cell, carry the genetic information that directs the synthesis of proteins necessary for the reproduction and function of all cells in our organism. This discovery helped us to understand how this information is stored and how these molecules can be duplicated by separation of the strand and synthesis of a complementary one to be transmitted to progeny. Thus, DNA in genes carries genetic information, and proteins, synthesized during all biological activities, provide the means to execute this information.

This signifies that we inherit our main characteristics, physical ones such as the colour of the eyes, or skin, as well as the capacity to respond to the environment and, to a certain point, our intellectual capacities and abilities, from our progenitors through inheritance of the genetic code of DNA contained in the genes.

In the last 50 years, many studies have been conducted to ascertain how this process works and what happens when mistakes occur that can lead to diseases. Development of biotechnology has been rapid and it is greatly helping the molecular biology studies undertaken in different fields, which are important for a wide range of activities, from identification of a person (paternity studies, identification of small samples for forensic medicine...) to knowledge of inherited diseases and

predisposition to others such as cancer, neurological abnormalities, etc. Some traits are determined by one gene, but many others need several genes to complete one function (probably all the abilities and intellectual capacities).

Recent description of the number of genes in our genomes is leading to the study of their function and participation in so many different activities of our cells. The implications of all these findings for medical care are obvious: first of all, for diagnosis of why something is not working properly and for the use of that information to diagnose the same abnormality in another patient and second, for therapy. It is easy to imagine that if we know which gene is affected and causing a specific disease, it would be good to replace it by a normal one to recover cell function. This therapeutic approach - gene therapy - is developing at high speed and there are already some demonstrations that it works in selective and severe immunodeficiencies.

There is a long way to go before we can understand the full complexity of our body, first in health conditions and later in disease situations. However, huge developments are in progress for the benefit of all.

**Dr T. Espanol**  
Immunology Unit  
Hospital Vall d'Hebron  
Barcelona- Spain

## Jeffery Modell Foundation Establishes World Immunodeficiency Network (WIN)

### Grant Support Offered to 57 Patient Organisations in 50 Countries

The Jeffrey Modell Foundation announced an exciting new initiative on behalf of patient groups around the world... the World Immunodeficiency Network. WIN offers funding support through grant applications as well as "know how" and assistance in preparing publications, disseminating information, web site development, family days, advocating for product safety and availability and much more. WIN connects 57 patient groups in 50 countries, includ-

ing the US, Canada, Mexico, Central and South America, Europe, Eastern Europe, Asia, Africa, Australia, and New Zealand. WIN has established a world renowned Medical Advisory Board who will review and act upon grants within 30 days.

WIN patient grant applications are offered on the JMF website: [www.info4pi.org](http://www.info4pi.org). Simply click to "WIN."



# UPDATE

Spring 2004

## Chairman's Message



### Dear all,

On behalf of the IPOPI board I wish you all the best for 2004. We hope it will be a good year for you all.

2004 will be a busy and exciting year for IPOPI. In October we will have the bi-annual meeting in Versailles together with ESID and INGID. We are working on the programme and it will soon be available on the following website (<http://www.esid2004.org>).

Part of the bi-annual meeting will be the elections of board members. Within a few month the NMO's will receive papers to nominate candidates for the Board. It is advisable to start thinking early about nominations.

Because IPOPI is growing and

there is a lot of work to be done we will start looking for a full time Chief Executive. We hope we can introduce this new CEO in Versailles.

It is also the intention to have our own IPOPI headquarters, away from PiA, who have kindly offered us shelter all these years.

Also new in 2004 will be the changing of the focus on plasma derived products from safety to affordability. Up till 2003 the safety of plasma derived products was the hottest item during meetings from PPTA, EPFA and our own biannual meetings. I noticed some changes in this, starting with the meeting from EPFA in Vienna in October last year. Safety is still an issue but no longer the main issue. All parties, including the manufacturers, are aware that there can never be a 100% safety guarantee. But also all parties involved are convinced that nowadays plasma derived products are as safe as possible. This does not mean that nothing need be done about safety anymore. Beside safety, economics / affordability will become more and more important. IPOPI will maintain the contacts with industry, regulators and other users of plasma derived products, such as the World Federation of Haemophilia about this new challenge.

**Kees Waas**  
IPOPI Chair

## WHO AND IVIG

We were surprised during the autumn to learn that WHO had excluded immunoglobulin from its list of essential medicines. In view of the benefits to people with primary immunodeficiencies IPOPI felt that it could not allow this situation to go unnoticed and contact was made with WHO. Our overtures were well received by the Blood Transfusion Safety Team.

Through the IPOPI Blood Committee, steps were taken to advise others of the situation and information was distributed to ensure that submissions were made to the WHO Expert Committee on the Use of Essential Drugs. Those early submissions were made by IPOPI and ESID in view of the extremely short time frame. However since then, because of work pressures and demands on staff time, the

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## International Patient Organisation for Primary Immunodeficiencies

Alliance House, 12 Caxton St,  
London SW1H 0QS

<b>Chairperson</b> Kees Waas Netherlands	Anneli Larsen Sweden
<b>Secretary</b> Keith Gray United Kingdom	Katri Molarius Finland
<b>Treasurer</b> Stephen Baxter New Zealand	Maria Nolan Ireland
<b>Secretariat</b> David Watters United Kingdom	Joy Rosario South Africa
<b>Committee Members</b> Eva Brox Norway	<b>Alternate Member</b> Vicki Modell USA
	Richard Barr USA

## Contacting Your Executive Committee Members

### SECRETARIAT

David Watters  
Alliance House, 12 Caxton Street  
LONDON, SW1H 0QS, UK  
david@pia.org.uk  
Tel and Fax: 00 44 207 222 3545

### CHAIRMAN

Kees Waas  
Burg. Krijgsmangeerde 274942 AV  
Raamsdonksveer, NETHERLANDS  
Keeswass@worldonline.nl  
Tel: 00 31 162 517 210

### SECRETARY

Keith Gray  
6 Great Meadow Way, Fairford Leys  
Aylesbury, HP17 8FT, UK  
keith.gray@btopenworld.com  
Tel: 00 44 1494 654 604

### COMMITTEE MEMBERS

Eva Brox  
Borgundvegen 478  
Aalesund N-6015, Norway  
evabrox@online.no  
Tel: 00 47 70 14 65 65

Stephen Baxter  
The Vicarage  
43 Queen Street, Pukekohe, New Zealand  
sjbaxter@clear.net.nz  
Tel: 00 64 9238 4985

Teresa Espanol  
Hospital General Vall d'Hebron  
Pg. Vall d'Hebron, 119-129  
Barcelona 08035, Spain  
tespanol@vhebron.net  
Tel: 00 34 93 274 6832

Katri Molarius  
Kuutamokatu 8G 73,  
Espoo 02210, Finland  
katri.molarius@helsinki.fi  
Tel: 00 358 50 367 7730

Maria Nolan  
"Croneview" Cronelea  
Shillelagh Co. Wicklow, Eire  
crone@eircom.net  
00 353 552 9133

Joy Rosario  
PiNSA, Suite 236, Private Bag X10  
Musgrave, 4062, South Africa  
jgrosario@mweb.co.za  
00 72 31 34566

Anneli Larsson  
Vaktelvägen 45, Örebro 703 48, Sweden  
larsson.272818@telia.com  
Tel: 00 46 19 272 818

Bianca Pizzera  
Via Vittorio Veneto, 8  
Arona (No) 28041, Italy  
bianca.pizzera@libero.it  
Tel: 00 39 322 45554

Richard Barr  
225 Crossroads Boulevard  
381, Carmel, California 93923, USA  
grbarr@mindspring.com  
Tel: 00 1 831 624 3795

Vicki Modell  
747 Third Avenue  
New York, NY 10036, USA  
vicki@jmfworld.org  
Tel: 00 1 212 819 0200

**Honorary Life President Bob LeBien**

## Note from the Editor...

This edition of Update reflects the variety of some of the initiatives and concerns of primary immunodeficiencies globally. What it doesn't reflect is the cosmopolitan English that comes through in the various articles and reports that arrive. By the time you read Update, it has been through various spell check/ proof reading processes thus sadly losing its truly international flavour. However, remember that the relevance of Update is wholly dependent on people forwarding their articles and news to the editorial board no matter how organic the English and everything sent to us is welcome!

Amongst the various articles in this Update is hidden the fact that Vicki and Fred Modell of the Jeffery Modell Foundation are the recipients of an International Union of Immunological Societies (IUIS) award, an award that was given only once before, to Bill and Melinda Gates of the Gates Foundation. Congratulations to you both, from all of us in the worldwide primary immunodeficiency community, your work is invaluable.

Another article talks about the risks inherent for PID people in keeping domestic animals. Please don't overreact to the article and start looking for homes for your pets! We are running the article because we should all be aware that some diseases can be transferred and we should just be conscious of this fact.

Finally, please support the publication and send news, views or articles to Joy Rosario (Editor) at [jgrosario@mweb.co.za](mailto:jgrosario@mweb.co.za)

## Twelfth International Congress of Immunology to Convene in Montreal

### Presentation of IUIS Award Announced

10,000 clinical and basic immunologists, researchers, scientists and investigators from all over the world will gather this July in Montreal, Canada for the International Congress of Immunology. This meeting is held every three years; the last was held in Stockholm in 2001. This year, the meeting is being hosted by the Canadian Society for Immunology.

Participating in this event of unprecedented scope for the immunology field is the International Union of Immunological Societies (IUIS), which is comprised of 58 organisations worldwide. The IUIS will present a special award to Vicki and Fred Modell at the opening ceremony and welcome reception. This award, created and designed by world renowned artist Niki de Saint-Phalle, was given only once before, to Bill and Melinda Gates of the Gates Foundation. News of the award was announced at the WHO/IUIS meeting in Portugal, by Dr. Fritz Melchers, past president of IUIS.

## Plasma Protein Therapeutics Association



The Plasma Protein Therapeutics Association (PPTA) is the primary advocate for the world's leading producers of plasma-based and recombinant biological therapeutics. The medicines produced by PPTA members are used in treating life-threatening diseases and serious medical conditions including immune system deficiencies, bleeding disorders, burns and shock.

PPTA's member companies are among some of the best-known pharmaceutical manufacturers worldwide and include Aventis Behring, Baxter Bioscience, Bayer Corporation, Biotest, Grupo Grifols, Kedrion, Octapharma and ZLB Bioplasma. Additionally, PPTA represents more than 450 plasma collection facilities through PPTA Source.

As the global representative for the plasma therapeutics industry, PPTA works cooperatively with patient groups, legislators, regulatory agencies and others to address critical issues that impact the industry and those people who depend on plasma protein therapeutics. Since 1978 the Association's global committees and regional working groups have brought together industry experts to focus on important issues relating to the quality, safety and efficacy of plasma therapeutics.

In addition to its issue-driven work, the association provides a number of services. Its International Quality Plasma Program (IQPP) certifies plasma collection centres. A component of the IQPP is the National Donor Referral Registry (NDDR). The NDDR is a US national database of plasma donors permanently deferred for reasons which could impact on the safety of the donated plasma. The system has been in use in the US for more than 10 years and is now accessible on the internet. PPTA is currently exploring expansion of NDDR to Germany.

PPTA provides fractionator certification through its Quality Standards of Excellence Assurance and Leadership Program (QSEAL). With this program companies demonstrate their adherence to the Association's voluntary standards, a set of measures which exceed regulatory requirements.

Through the Patient Notification System (PNS) all interested persons in North America and Canada can obtain up to date information on withdrawals and recalls of plasma protein therapeutics manufactured by member companies.

PPTA also holds an annual congress both in Europe and in the USA, to update all those involved or interested in the sector about policy and scientific developments which have taken place over the year. The International Plasma Protein Congress (IPPC) 2004 will take place in the Hilton Brussels (Belgium) on 9-10 March 2004. The PPTA Plasma Forum will be on the 9-11 June 2004 at the Grand Hyatt, in Washington DC, USA.

PPTA is in regular contact with organisations which represent those with primary immune disease. The Association brings together the PID community with other customer groups on a regular basis in stakeholder forums and in patient workshops. Consultation on upcoming legislation, regulation and market access issues takes place at local, national and international levels.

The Association is happy to provide input to activities of Primary Immune patient groups when invited. We participate in IPOPI's safety and supply group and regularly provide our perspective to annual meetings of different PID groups and have arranged information sessions on subjects of interest to national groups.

PPTA is always happy to hear from Primary Immune organisations, and we can be contacted at the following locations:

**Europe:** +32 2 705 5811  
[ppta@pptaglobal.be](mailto:ppta@pptaglobal.be)

**USA:** +1 410 263 8296  
[ppta@pptaglobal.org](mailto:ppta@pptaglobal.org)

**Japan:** +81 3 5789 5928  
[myoshida@pptaglobal.org](mailto:myoshida@pptaglobal.org)

## CAN YOU CATCH A DISEASE FROM YOUR CAT?

### AND HERE'S THE LATEST FROM THE DOCTORS.....

**M**any years ago, twenty three in fact, my little girl Gaby, my husband and I were on holiday at the beach for a few days. At the time we were living in Pietermaritzburg in a Victorian redbrick house with a lovely garden, two dogs and a cat. Gabrielle had been diagnosed with Hyper IgM and we were used to regular hospital visits and courses of antibiotics, this of course predated infusion therapy.

Anyway, at the beach she presented with swollen glands in the neck which told us she was running yet another ear infection. This time she also had a swollen gland in her groin but the doctor thought it was related to the ear infection so just put her onto a course of antibiotics. She didn't respond and started to spike a fever so we cut the holiday short and took her back to Pietermaritzburg to our normal ear nose and throat specialist. He immediately referred her to a surgeon who instructed me to take her straight to hospital. I asked if I could go home to fetch her pyjamas and he refused – it was then that I knew we were sailing into unknown waters.

They operated for a burst appendix but didn't get that far; the gland in the groin was reacting to some unknown infection and was removed. Gabrielle came out of theatre with all sorts of tubes and drains, a very little creature on a very large theatre trolley. For three days she just got sicker and sicker and the doctors were considering diagnoses such as leukaemia and tuberculosis. On the fourth day the surgeon came in and asked me if I kept cats. I said yes, at the time I was breeding Siamese using Kitty Boy our chocolate point as the tom and had a two month litter of kittens at home. It turned out that one of the kittens had scratched Gaby on her knee and she had developed cat-scratch disease as a result. It was only with the advent of the Internet that I did some searching and found that there can be indeed a relationship with cat-scratch disease, also tick bite fever (a related pathogen) and immunodeficient people.

I am not suggesting that you find homes for your animals, after all I still have a cat. Just be aware that transfer of infection is possible, even in the cleanest of homes and that you should be on the alert for this type of disease. Had I known we would have been spared the trauma of that time and Gabrielle would have had one less scar. She is veteran of 26 operations and this one was the most frightening of them all and entirely unnecessary – she could have gone straight onto the correct antibiotic in the first place.

#### Joy Rosario

My introduction to zoonoses, diseases that can spread from animals to man, came in the form of an annoying upper respiratory infection that would not clear up. Many years ago I used to buy imported parrots from a quarantine station, bring them home, and tame them for sale as pets. I knew that this practice was slightly injurious to my health, as I sported bruises and scars from parrot bites so severe that a total stranger was prompted to give me the phone number of a battered women's shelter! But I didn't realize that these assaults to my flesh were not the only injuries I could sustain from my daily contact with pet birds.

My doctor suspected that I had contracted psittacosis, a chlamydeous infection of birds that can cause pneumonia in humans. Although easily cured in healthy young adults as myself with a course of antibiotics, this disease can be life threatening in immunocompromised persons, such as the very young and the elderly. After hearing this frightening news, I started to treat every incoming parrot for this disease as well as test

them prior to sale to make sure I would not spread this disease to an unsuspecting pet owner.

Since that time I have learned that it is important for my doctor to know my hobbies and my occupation so that he can be aware of diseases that other patients may not be at risk for. And I became aware of the potential for legal liability if one of my customers should become sick following the sale of one of my animals.

Most zoonoses are very difficult to "catch". The organism that causes psittacosis is the same one that causes the very common chlamydial eye and respiratory infections in cats. However, the spread of this organism from cat to human is much rarer than from bird to human. However, in this day and age it is much more likely that one day we will place a pet into a home with an immunocompromised person. Lowering of the immune system by chemotherapy for cancer, prolonged use of steroids, or HIV infection increases the

risk that a pet kitten may spread one of a variety of diseases to an unsuspecting new owner.

Ringworm is probably the most common zoonoses of cats. It was quite common when I was working in the veterinary hospital to see a kitten come in covered in ringworm lesions, and to find that the owner was also vigorously scratching at suspicious-looking red scaly patches of skin! The ringworm fungus was cultured from 4-35% of asymptomatic cats at four different cat shows and studies of catteries suggest that as many as 40% of all cats may be asymptomatic carriers of ringworm. If you have EVER had ringworm in your cattery, chances are it will pop up again. I advise all cattery owners to inspect their kitten sales contracts carefully and consider putting in a warning about the possibility of ringworm infection. Otherwise, you may see yourself in court someday trying to defend yourself against a demand for reimbursement for the costs of medical treatment for a six year old and all the children in his first grade class. I have not yet heard of a situation where a cattery owner was held liable for severe fungal infection in an AIDS patient, but that is probably only a matter of time.

Bacterial Infections can be spread from cats to humans. Bacteria are usually secondary infectious agents that follow viral or mycobacterial infections. Animals with diarrhea can be a source of infection to humans, as diarrhea more effectively contaminates the environment with pathogens than does a formed stool buried neatly in kitty litter. Most bacterial zoonoses (campylobacteriosis, streptococci, staphylococci) clear up readily with antibiotics. The most common bacterial zoonosis is the gram-negative organism Pasteurella. Approximately 60-75% of normal cats carry these bacteria in their mouths. Cat bite wounds should be cleaned carefully with antibiotic cleansers such as Nolvasan. An antibiotic ointment should be applied. Inflammation of the wound site or fever following a cat bite indicate that medical attention is required and systemic antibiotics may be indicated.

Salmonella bacteria can be shed in cat stools and are more common in cats fed raw meat or those that catch wild birds. Hand washing following litter box cleaning or handling of stools is effective in limiting spread to humans, as infection follows a fecal to oral route. Another gram-negative bacteria causes tularemia infections. These are found in cats that catch wild rabbits or rodents and these bacteria may also be spread to humans. This, of course, is not a pathogen we need to worry about in our closed catteries.

Cat Scratch Disease is one everyone has heard of, but only very recently has the causative agent been identi-

fied. The bacteria Rochalimaea henselae was cultured from over 40% of cats surveyed in a population of shelter cats. But more importantly to us as cattery owners, the organism was also found to be carried by cat FLEAS. The disease causes systemic illness and lymph node lesions and can be very serious in immunocompromised individuals. Antibiotic therapy usually cures the disease without complications in healthy young adults. Clearing the bacteria from infected cats requires long-term antibiotic treatment, however, and cats may be continuously or intermittently infected indefinitely. Again, caution is urged following cat bites and scratches and the potential for the transmission of these bacteria should be another impetus to keeping the cattery free of fleas. Cat Scratch Disease is a fairly common disease with an incidence of about 0.8 cases per 100,000 population. Again, the risk of causing severe infections in immunocompromised pet owners needs to be considered.

Parasitic Diseases are also potential zoonoses. Roundworm eggs can infect humans, particularly children, through a fecal to oral route. Tapeworm eggs, on the other hand, are not directly infectious to people. Feline strains of coccidia and giardia do not appear to be infectious to humans. Much media attention has been brought to the zoonotic potential of toxoplasmosis, a type of coccidia that can cause birth defects in unborn babies. However, poorly cooked meat is a much more significant source of infection than pet cats. Oocysts (eggs) must incubate for three days before becoming infectious to people, so regular litter box sanitation and hand washing following this procedure is adequate to prevent exposure. Pregnant women may be advised to wear gloves when handling cat feces or, better yet, assign another family member to this job.

Viruses are extremely species specific. Feline Immunodeficiency Virus, Feline Infectious Peritonitis, and Feline Leukemia Virus cannot cause illness in people. A cat cannot catch your "cold", although there is indication that canine corona virus infection may potentiate FIP in cats.

The purpose of this article is not to scare you into thinking that owning cats is hazardous to your health. But every hobby has associated risks, and your physician should be aware of the potential that the illness you have may be related to your hobby. I also want to caution breeders that when they are selling kittens, perhaps questions as to the presence of immunocompromised persons in the potential new home may be appropriate. In any case, a good sales contract should deal with the potential of zoonoses and offer you protection against being sued for large medical bills. I

recently declined to sell a kitten into a home with an AIDS patient. It hurt me deeply, as I know the comfort that a pet can give, but I was not willing to risk that one of my kittens could cause severe illness in an immunocompromised person.

Keeping a cattery clean, free of fleas, and feeding fresh cooked or processed foods can not only keep your cats healthier, but may also keep you healthier as well. Do

not ignore cat scratches or bites and make sure your kitten customers are kept well informed so we can make sure that everyone's contact with these wonderful animals is as pleasant as possible!

#### Lorraine Shelton

This article is from PetStation  
<http://www.petstation.com/cat-zoonoses.html>

### \* Pet-transmitted infections : diagnosis by microbiologic and immunologic methods. CM.Litwin Pediatr.Infect.Dis. J. Sept.2003

Most infections in humans are spread in the community; however, some transmitted from pets are on the increase. The review article (\*) stated: "Infections detected after dog bites could be around 10%, but those produced by cats could be as high as in 80% of cases and the risk of wound infection is greater in older people and those immunocompromised" Other infection routes such as contact with feces,urine, saliva and even aerosolization are also possible.

The common use of household pets, such as dogs and cats, or exotic animals, which are increasingly popular, can give rise to infections which, at times, are difficult to suspect and diagnose and, very important, to treat. Some infections such as cryptosporidium can run a chronic clinical course and are difficult to eradicate when the immune system is not perfect. Congenital defects in primary immunodeficient patients rende them much more susceptible to infections with chonic sequelae.

Advice on contact with pets must be individualized to each patient since each defect carries a different susceptibility to different infections, which also varies according to their personal circumstances; however, in any event it is better to avoid this possible source of infections. A well cared for and veterinary controlled dog or cat might be harmless, but several measures must be taken when they are hadled by an immunodeficient patient: hand-washing after touching the animal, not cleaning the habitat ( contact with feces is one of the mechanisms of infections ), etc.

Patient-physician communication is essential for decision making in these situations.

#### Dr T. Espanol

Immunology Unit  
Hospital Vall d'Hebron  
Barcelona

### WHO AND IVIG...continued

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deadline has been extended considerably and the meeting which had been due in April 2004 has been deferred until the autumn of 2005.

Since the extension of the deadline IPOPI has been at pains to make sure that a wide representation of support is achieved for those initial statements. The paperwork can accessed electronically at [www.who.int/medicines/organization/par/edll/procedures.shtml](http://www.who.int/medicines/organization/par/edll/procedures.shtml) or hard copy can be obtained through the Secretariat in London, along with copies of the IPOPI submission. If you want further information please talk to David Watters at the Secretariat or Bianca Pizzera who chairs the Committee responsible for this area of IPOPI's work.

### New Physician Algorithm Now from the Jeffery Modell Foundation

#### FOUR STAGES OF IMMUNOLOGIC TESTING PROVIDED BY JMF MEDICAL ADVISORY BOARD

In an effort to advance diagnosis, the JMF has now added a Physician Algorithm to the 10 Warning Signs Poster. The algorithm identifies four specific stages of immunologic testing. The earlier stages are generally appropriate for primary care physicians before the necessity of a specialist referral. The algorithm, reflecting a consensus of the JMF Medical Advisory Board, is presented as a public service by JMF and is supported by grant funds from the CDC, the generosity of the NIH, the PPTA and several global plasma producers. The algorithm is available on the JMF website, [www.info4pi.org](http://www.info4pi.org).

## Meet Your Trustee



**Name:** Anneli Larsson

**Age:** 37

**Family:** Husband and two sons Carl 11 and Oscar 9

**Occupation:** Part time administrator for PIO, Sweden, combined with voluntary work for IPOPI.

When our second child was born with a primary immunodeficiency disease (PI) and his condition required parental care, I decided to put my working life on hold. However, soon thereafter, I found myself involved in the Swedish Patient organisation, PIO. It is a challenge and great responsibility to work for the first established patient organisation in the world for people with primary immunodeficiencies. PIO was established in 1978 by Maj-Lis Hellström. All the work Maj-Lis, Kerstin Torstenson and many others have done over the years, and still do, is a valuable heritage. We on the PIO Board do our best to continue and build on their valuable contribution.

I am of course very grateful to be involved, having had the opportunity to work for a cause I truly care about. It is my conviction that a transparent and effective patient organisation is needed both on national and global levels. We need to be one voice throughout the organisation and protect patient's interests wherever and whenever they appear. The

span is very wide and requires continuous attention in order to provide pragmatic support and knowledge regarding medicines, treatments, political decisions etc.

Patient organisations also have a very important role in bringing people together to offer an opportunity for the exchange of knowledge and experiences. One of my responsibilities in PIO is to arrange summer camps for children with PI, which includes their families. Our ambition with these camps is to support the families in the precarious and often very difficult situation associated with living with a PI. I might add, that last year I promised myself to never put myself through the hard work of arranging a camp again! But when I saw how the kids and parents connected on so many different levels I knew that all the work was worthwhile... families became friends, little boys and girls got themselves pen pals and everyone really appreciated the chance to meet with people in a similar situation. So in August I arranged the third summer camp. This year we had the great privilege to have Professor Anders Fasth as our guest speaker. The session for the parents was about genetics and gene therapies. Anders Fasth also had two special sessions about the immune system, one for children with PI and one for siblings. These sessions were much appreciated.

When I'm not working with PIO or IPOPI I love to spend time with my family and just be there for the boys when they return from school. Children grow so fast so it is only for a few years that I can drive them, watch them play football and listen when they practise the electric guitar and synthesiser. Music and football are both big common interests in our family. I used to play football when I was young and I can still challenge my boys in penalty kick competitions and I sometimes win! I also appreciate meeting with good friends and taking hikes in the woods. Public debates on problems of modern society have also always been a great interest of mine and I have been involved in different movements and projects.

It is a great experience to meet and cooperate with people in National Member Organisations (NMO's) across the world. So many people have dedicated their lives to improve the situation for people with PI. It is a privilege to learn and gain influenced from you all.