

## Busy times for PiNSA

### A note from the Chair, Cally Coldbeck

It has been an interesting couple of months for us. Joy Rosario and I had many lengthy discussions over the succession of the Chair of PiNSA and Joy came to the difficult decision due to her work load to take up post of Vice Chair and she put my name forward as Chair to the committee, who seconded this motion.

Both Joy and I had hoped for a smooth, seamless, succession and we can truly say that we have managed incredibly well over the past few months, and with our wonderful committee behind us, we see no reason for the status quo to change.

During this quarter, we have been incredibly busy.

- We have sent our financials off for audit and we are waiting for the report so that we can supply the NPO (non-profit organisation) reports.
  - We have seen our first World Immunodeficiency Day come and go. As a fledgling organisation, we have had to take small steps with regards to awareness this year, and will expand year on year. As they say in the classics "Every day is a school".
  - During the run up to the World Immunodeficiency Day, PiNSA sent out a press release to Reuters, the SABC, ETV and a range of other South African media. We unfortunately did not get the coverage that we had hoped for, but we will definitely be working constantly on these avenues during the course of 2011/2012.
  - We have made contact with John Robbie of 702 and Damon Beard of East Coast Radio inquiring about assistance with air time and/or an interview
- Fundraising this quarter has taken



*Member Meg Coldbeck and her grade 7 class from St John's DSG in Pietermaritzburg along with the rest of the school raised funds for PiNSA with a 'civvies' day.*

in a substantial amount and we are constantly reviewing our options available to ensure fluidity with regards to donations.

Should anyone have any fundraising ideas that they would like to share with us, please do not hesitate to contact Christelle at our PiNSA offices (033-3429941) or pinsahelp@mweb.co.za

So far this year, we have held a talk and civvies day at a school in Pietermaritzburg, have written to other schools about similar fundraising ventures and have received wonderful donations from both. We are holding a Rock & Roll fundraising venture in Pretoria at the end of June, which promises to be a great success.

- We hosted a medical fraternity information breakfast, where

paediatrician Dr J.F. Roos gave a most interesting talk on PID with a question and answer session. A wonderful time was had by all.

- As members of PiNSA, you should have received and completed the IPOPI PID Patient and Caregiver Survey – South Africa; this should assist IPOPI with integrating the National Member Organisations from all over the world. The deadline has been extended, so if you have not filled yours in as yet, please do so. As this is invaluable information needed to improve Patient Support.

## A note from the chair



*Cally Coldbeck*

### A little about me

I am a wife and mother of two beautiful daughters. One of which, Meg, suffers from PID. I was trained at Greys Hospital as a nurse. I stopped nursing in 1987. I served as the Public Relations Officer at Mediclinic, Pietermaritzburg until I fell pregnant with our eldest daughter Lara-Kate. Megan arrived 18 months later much to our shock, horror and delight. With having to constantly visit our previous GP, I eventually became practice manager until he left to work abroad.

I then trained as a landscaper and ran my own company for two years until Megan became incredibly ill and was diagnosed with PID.

In the past year, our family printing business has grown in leaps and bounds and my

husband Mike requested my assistance as Office Manager.

This gives me the opportunity to work from the office or home. I also run the PiNSA office from our factory. We have all the facilities and the most incredible staff, including the PiNSA secretary Christelle, who have come to know and understand Meg's illness.

I have managed to work around all the different needs that a busy mother and wife has with the help of my family, fellow caregivers, Joy Rosario, Christelle and the PiNSA committee.

I am positive that the transition from Vice Chair to Chair will not affect PiNSA in anyway. I look forward to working with each and every one of our members and to seeing that our PiNSA objectives are realised.

## Building a community for patients and families

### Joy Rosario's story – the founder and vice chair of PiNSA

**M**y story really started when my daughter Gabrielle was born in 1977 and now 33 years and 28 operations later (soon to be 29), the story continues. To date she must have had about 500 infusions of Polygam which equates to many hundreds, if not thousands of blood donations!

In 1998 I read an article saying that people with Hyper-IgM were at risk and I went into mommy panic mode, Gaby was 21 at the time. Thanks to IPOPI (International Patient Organisation for Primary

Immunodeficiency) and David Watters of the PiA (Primary Immunodeficiency Association) my fears were allayed and an enduring relationship with both David and IPOPI started.

***We only have 170 people diagnosed and should have at least 4000: the work is ahead.***



In 2000 David enquired whether I would be prepared to give a go at setting up a South African organisation, the seed money being provided by IPOPI. At the time I was a teacher librarian at Rustenburg Girls High in Cape Town with three months holiday a year! I took on the challenge and PiNSA was constituted at the Red Cross Children's hospital in Cape Town in 2001. I was co-opted onto the IPOPI Board in 2004 and re-elected in 2010 for a term concluding in 2014. This



*Joy Rosario*

international involvement has been important because it has allowed us an international profile but more importantly we have built relationships which have benefited South Africa.

Two moves and two promotion posts later I am in Pretoria and running the Information Resource centre for the National Department of Basic Education. Gaby is lead strategist for Native, a social media and marketing company in Johannesburg.

PiNSA is alive and well with a committee of ten people. What a pleasure to have the new energy and input, for many years I was a one man band offering little more than telephonic support. Dr Monika Esser came on board in 2006 and

we managed to garner funding for a secretary. Cally Coldbeck has now taken up the reins as chair and she is exactly what PiNSA needs, she has the enthusiasm and commitment to carry it through and I am always there as her support and backstop – the organisation is too important for any of us to fall by the wayside. We only have 170 people diagnosed and should have at least 4000: the work is ahead.

## Report back from Lisbon

Joy Rosario attended the IPPC (International Plasma and Protein Congress) on 14 and 15 March 2011 followed by an International Patient Organisation for Primary Immunodeficiencies (IPOPI) Board meeting on 16 March in Lisbon, Portugal. This is her report:

The IPPC meeting was most interesting (presentations available on <http://www.ippc2011.com/#/Home-01-00/> - please e-mail me for login and password) as it gave me an opportunity to speak face to face with Duncan Armstrong of NBI. He is most sympathetic to our situation and indicated that the sub-cut licensing issue of Intragam is on the agenda. We also spoke at length about the possibility of a Sub-Saharan group of stakeholders as awareness of PID is important for countries that surround South Africa. Jean Emmanuel from Zimbabwe presented a paper called "Supply of Plasma derivatives in Low Development Index Countries". He was part of the discussion with Duncan but both warned of raising unrealistic expectations and also

supply of products. My view is that at least we can provide patient support and awareness and that we need to broaden the footprint as at the moment PID is a first world disease. Hopefully the Paediatric congress at Sun City in October will build more contacts in surrounding countries.

The IPOPI Board meeting discussed our financial position and the fundraising proposal, also World Primary Immunology Week. IPOPI had given us a grant for the publishing of materials. Several new national member organisations (NMOs) have come on board, South America being the most active continent in this regard. There will be a second global leaders meeting in London in November to which Dr Monika

Esser has been invited. I go automatically as a Board member. We discussed the agenda for the IPOPI/ESID/INGID meeting for Florence next year. PiNSA needs to raise the funds for our new Chair to attend, it's an important meeting.

Other matters included a MAP report from Dr Espanol (Chair of MAP) and also a website update.

The biggest news is that David Watters, executive director of IPOPI announced his retirement and we, PiNSA, followed up with a congratulatory e-mail. His invaluable support and knowledge will be really missed. Johan Prevot will be taking over from him and I am sure he will be wonderful, I have known him for many years and he has been in our employ for some time now.

## How chronic diseases impact on siblings

As parents and caregivers to a child with a chronic condition, we know that parenting can become a constant balancing act, fraught with anxiety and hyper vigilance.

From the outset, most of the children who have a chronic condition have been ill for years. We have been back and forth to doctors and hospitals to no avail. Just when you thought you were losing your mind and concocting illnesses in your head, you get the final diagnosis and your world falls apart. As parents, it is during this stage that we become all consumed with the care of this child. We investigate every option available to us and even then we try harder to find a cure. This is where the road to find answers becomes fraught with pot holes that you never anticipated.

The sibling can start to play up. They could become tearful, clingy, possibly starts having disruptions in sleep patterns or nightmares. Some may even not want to go to school and sibling squabbling is almost always inevitable.

If we look at chronic disease through the eyes of the sibling, we see it as a disease that takes their parents attention away from them. We see it as a disease that favours one child and therefore it gives rise to sibling rivalry. Often they will see the disease as one that has robbed them of a happy home life because the parents are tired, bickering or unable to play with them due to time constraints. Often siblings fear that their brother or sister is going to die and before you know it, your entire family is heading down a

slippery slope of despair.

This is where the family as a whole has to put the brakes on and reassess what is important to them as a whole. Life has to get back to normal, even though normality seems light years away.

- Start with reinstating family meal times and round the table (not the coffee table).

This gives the family a wonderful platform to discuss all the aspects

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of the illness. Each family member can give their version of the fear, hurt, frustration and anxiety they are feeling. It is also good for children to witness their parents being emotionally 'normal'.

- Take time out to sit and talk to siblings on a one on one basis.
- Question how they are feeling about life, the disease, their school and the family.
- Ask if they are afraid of anything and then deal with the answers accordingly.
- Let the children paint what they are feeling.
- Ask them what colour hat they are wearing – red for angry etc.
- Cuddle, hold and caress your children. Make them feel that the world is still the way they thought it was before the

diagnosis.

- Play games with your children, not only will this benefit the whole family dynamic, but it will bring laughter back into a tense situation.
- If Mum has to do the hospital run, let Dad order take outs and have picnics on the lawn, or have a video evening.
- Organise more play dates for the sibling. Removing them from the constant vigilance that comes with the condition and adding a sense of normality to life.

The situation becomes more serious when the sibling shows signs of severe depression. This is where you immediately call in professional help. For younger child you can explain this as going to see a 'talking doctor'. There are varying methodologies which can be utilised to treat the sibling. The correct 'talking doctor' will be able to ascertain which avenue is the best suited to your child.

Do not despair, some of the most important findings in siblings of children with a chronic disease are very positive. Despite sometimes suffering from depression and anxiety, the sibling also has the ability to develop a stronger ability to show compassion than a child who has not lived in a home with chronic disease.

Above all, life as you knew it will not be the same again. It can actually be better! Once the panic has passed, the reality of your new life will ease into your bones. Keep your family and friends close, as they are the very people from whom you can gain the most strength.

## Frequently asked questions

Dr Monika Esser of Pinsa's Medical Advisory Panel answers your questions about Primary Immunodeficiencies

**Q:** *If you or your child were diagnosed with PID what would be the first questions you would ask?*

**ME:** Can it be cured/treated? What treatment is available? What is the expected quality of life? What is the life expectancy? Could it have been prevented?

**Q:** *Can Primary Immune Deficiency be cured?*

**ME:** Certain forms of Severe Combined Immune Deficiency and an increasing number of PID's, even Chronic Granulomatous Disease can be cured by a successful bone marrow or cord blood transplant. Therefore early diagnosis is so important before irreversible organ damage has occurred. Gene therapy holds great promise for curing PID's, but later onset of cancers is still a problem. The more common antibody deficiencies are usually treatable with Immunoglobulins assisted by antibiotics and vaccinations.

**Q:** *How safe is the local immunoglobulin therapy in terms of transfer of infections e.g. Hepatitis and AIDS?*

**ME:** Locally available Immunoglobulin (Ig) is as safe as any overseas product because the safety and quality controls, including nucleic acid amplification (NAT) testing for Hepatitis C and HIV are as strict and sometimes stricter than that for overseas products. Blood donations are from healthy unpaid volunteers. However it is a blood derived product and very rarely virus contamination (not HIV) has occurred overseas. Therefore the product, like any other transfusion should only be used for the correct medical indications and judiciously to be

available for PID patients who depend on a regular supply.

**Q:** *How safe is the long term use of immunoglobulin therapy?*

**ME:** The safety profile of long term Immunoglobulin therapy has been well established. All blood derived products are registered with the Medicines Control Council of South Africa in terms of their safety, efficacy and quality. There

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**[pinsahelp@mweb.co.za](mailto:pinsahelp@mweb.co.za)**



has been no documentation of residual effects with long-term use of intravenous immunoglobulins. Follow up of patients who have been on regular IVIG for decades now shows improved quality of life and equivalent ability to work compared to a person without PID. But treatment should be supervised by a doctor who regularly sees the patient holistically. Too little Ig treatment and excessively long intervals between treatment (generally more than three weeks in the case of intravenous Ig) can also result in recurrent infections and the outcome can be almost as bad as without treatment. This is not a complication of the treatment but of incorrect treatment and it is avoided by regular serum trough levels of IgG and medical check ups. As Ig transfusions do not prevent all infections, the PID patient may also need to be on antibiotics in addition sometimes, or require additional nutrition.

**Q:** *Primary Immune Deficiency is*

*a chronic condition. Has the state established the infrastructure for home therapy e.g. home nurses, like in other countries.*

**ME:** There is currently no assistance or plan for home therapy for PID treatment. This would have to be motivated for by the concerted efforts of organizations such as PiNSA supported by the medical profession.

**Q:** *Could the medical fraternity advise a standard procedure in terms of clinical management of anyone with PID, obviously tailored to the need of the individual? Could this be made available on the website or in the brochure going to the doctors?*

**ME:** Standard guidelines for diagnosis and investigations are available eg JMF guidelines – and the IDF (see useful websites). Treatment is often very individual, other than the standard guidelines for Immunoglobulin replacement. Specific medical protocols for individual diagnoses are available.

**Q:** *Is it safe for patients with Primary Immune-deficiency to receive the seasonal flu vaccine?*

**ME:** The flu vaccine, now in stock in many pharmacies (although some stocks are already said to be running low) and to be made available in government clinics from April, is not "live" and therefore safe for PID patients who do not have severe T-cell deficiencies. Although it could be useful for PID patients' immunity to the flu virus, it is advisable to consult the patient's physician before having the vaccine. Before receiving the vaccine, the PID patient should also be in good health. If younger than two and a half, two vaccines

will be required, four weeks apart. Preventative measures like hand-washing should nonetheless continue throughout the flu season.

**Q:** *Is it necessary that the polygam goes at 100ml per hour or can one speed up by perhaps 150ml per hour and then 200ml per hour after a patient received polygam for more than say 2 years?*

**ME:** My experience is that the overall Infusion speed after the first 15 to 30 minutes (30 to 60 ml/hour in adults and 0.01-0.02ml/kg/minute in children) after the first three infusions can be advanced carefully, but if any side effects especially flushing or rash occur – step back immediately to previously tolerated dose. Depending on the individual patient and the diagnosis I have

witnessed some very ‘non standard’ infusion speeds! If the patient has any IgG antibodies – like most Common Variable or Hypogammas do, I would not push the limits past any slight discomfort and not go past 150ml /hour infusion (120-150ml/hour) speed, (and children 0.08ml/kg/min) especially with home infusions. Use a 20 drop/ml infusion set. Also some patients are premedicated with Phenergan (antiallergy) and or steroids (solucortef) and in this context I would also be careful – slower infusions may then not need these medications. Special caution for patients with kidney problems is advised.

### National registry

**Q:** *Bearing in mind patient confidentiality, is there a national*

*registry of patients with PID (as they have in other countries) and if not, would the medical profession like one developed?*

**ME:** A national registry with support from the industry has been established in 2006 and is stored on a secure server at Tygerberg Hospital, University of Stellenbosch. Only patient data with full patient/parent consent are stored (registered study with ethics number) and all data is strictly confidential. Information exchange of registry results with other countries and continents enables us to see different genetic patterns of diseases and incidences. It may be possible in future to assess the outcome on specific PIDs with good record keeping. This will result in better patient care, while serving to increase awareness for these rarer conditions.

## Are you on the PID registry?

All South African patients with PID are encouraged to provide their details for inclusion on PID Registry. The PID Registry is part of an ethically approved confidential research project at UCT and Stellenbosch universities. Dr Monika Esser, one of the doctors responsible for the register highlights the important role that this information

plays for research and lobbying. ‘The registry provides objective evidence needed to change policies, inform medical aids and improve patient outcomes through data sharing’. *For information about the registry, or to submit data contact Rina at [rina@sun.ac.za](mailto:rina@sun.ac.za) or fax number 021-9389138/4005 or 0722100206 (mobile).*

## Useful websites

The IPOPI [www.ipopi.org](http://www.ipopi.org) and IDF [www.primaryimmune.org](http://www.primaryimmune.org) websites have useful information and Patient and Family handbooks available on different kinds of Primary Immunodeficiencies, should you like to read more on your or your child’s diagnosis.

