PiNSA
PRIMARY IMMUNODEFICIENCY NETWORK OF SOUTH AFRICA

February 2011

Dear Readers

2011 has started of with a bang and we would like to wish you and your families a safe, healthy, prosperous and above all happy year. Here is hoping that the floods that we have been experiencing have not affected you and your loved ones.

To say that 2011 is going to see PiNSA moving from strength to strength must be the understatement of the year so far.
As you know, the PiNSA Committee met in November 2010 and we had a wonderful day thrashing out the future work that needs to be done for Primary Immunodeficiency. You will find included in this newsletter, a report back from our Chairman - Joy Rosario.

In this edition of the newsletter, you will also find a story written by Karen Heese, which will explain PID from a patient and not a parent's view point.

- We would like to keep both parents and patients informed as much as possible and to enable us with this; we are asking that you send us your questions and stories.

Dr Monika Esser will be answering our new FAQ (frequently asked questions) section, so please email us with your queries.

- You will also find tips on how to make your child's visits to hospital more bearable.
- How to cope with your child's school and the barrage of questions asked by their educators, peers and their parents.

Enjoy the read and remember to email us should you need any assistance, advice and we will put you in touch with one of our Executive committee members who will be able to help you. We would just like to remind you, that all information, queries etc are treated as confidential.

Karen's story

I had just turned 29 when I was diagnosed with a primary immune-deficiency (PID). It was rather bewildering; all I had been told was to go to an oncology centre for treatment with what sounded like an undesirable "blood product" - intravenous immunoglobulin (Polygam). In my excellent GP's defence, she herself, after impressive detective work with Lancet laboratories in an attempt to understand why I was not immune to Rubella despite a repeated MMR, knew nothing of PIDs and thought that I would be filled in by the haematologist to whom I had been referred. I learnt very little during my consultation other than the need to return for Polygam a few days later and its considerable expense, as well as, rather importantly, the need to pack lunch. A friendly, but ill-informed nurse also told me that I stood a chance of HIV infection from the Polygam.

After my first treatment, I returned home to an empty house, with my husband out of town studying, with chills and aches worse than any I had experienced in the previous decade of admittedly already patchy health and decided that I had to find another way to deal with my condition. After a little research, I found out that there was little alternative to Polygam, but that the side-effects I had experienced were not unusual and could be managed with Panado. I dropped the rather extreme antihistamine which had had a disconcerting sedative effect on me (to the point of fainting) and made sure that I rested before and after the treatment. I resolved not to be afraid of needles and had a hot bath before treatment to swell my veins up. But more than anything, I found that information and information sharing was the best way of coping with my new life.

Within a few months, I established that my condition was not X-linked agamma-globulinanemia, as I had initially been told, but Common Variable Immundeficiency (CVID), and that the child that I was by then already pregnant with, was unlikely to carry my condition (a great relief as I had been expecting a 50% chance of passing on my condition if I had had a boy and the prospect of being responsible for what I found onerous treatment weighed upon me heavily). I also discovered that the side-effects seemed to wane and that with adequate rest, my "new" life was entirely positive. By the time that I turned 30, I was eight months pregnant, and feeling healthier and better than I could remember.

The surprising part of this post-Polygam revelation was that I had never considered myself sickly – admittedly the hard facts suggest otherwise; repeated ear, nose and throat infections as a child, along with bladder infections and then skin and stomach infections as a teenager (resulting in a cumulative absence of one third of my matric year). My twenties, though, showed an increasing frailty to which I had become gradually accustomed; including a painfully infected knee (which I was unwilling to have operated on) and pneumonia (which prompted a soul-searching hospital stay during which I decided to slow down my work commitments and start a family).

I had attributed much of this to a less healthy student lifestyle than my early home life, and then later to the pressures of work commitments. A routine ante-natal screen (which showed up that MMRs had failed to create an immune response) suddenly put all the pieces of the puzzle together, and answered the questions that many friends had asked me directly or muttered under their breath – "why are you always sick?" I was relieved too as a callous doctor whom I had consulted, suspecting that allergies were behind my string of chest infections, had told me that my sickness was "probably" psychosomatic. I was neither crazy nor desperate for attention!

I have been receiving treatment for CVID for eight years, and have two beautiful, healthy little girls. I am also fortunate enough to work from home, as an economist, and am spared commuting and large meetings, as well as an old foe – air-conditioning, that most people have to endure in their places of work. My greatest challenge on a day-to-day basis is the onslaught of snuffles and infections from my children and their friends, but it is a small price to pay for the love and affection which makes sense of my life in my more introspective moments.

The most difficult part of the treatment is going to an oncology centre, where despite the compassionate and cheerful staff, and courageous patients (I have befriended desperately ill people, some of whom have since died), which I find a huge emotional strain. But I try to draw inspiration and gratitude for the good health that I have (even if I almost always have a tissue in hand, nebulise most nights and often need to resort to antibiotics), as well as my extensive privilege (the financial means to insure against illness and treat my condition and the love of my family), as well as the opportunity to engage with others. But most of all, I have been taught the humility of a body that requires what I describe as a monthly "oil change", between which episodes I try to enjoy each day and live positively with the perspective that PIDs have taught me.

Tips for making IVIG hospital stays easier

- Surround your child with happy, caring, capable and child-friendly staff
- Try to get a semi-private ward to avoid cross-infections
- If you have a lap-top computer, take this along with videos (don't forget your earphones!)
- A special 'hospital' box, which holds all their little treasures, pictures, crayons, colouring book and a diary. The diary was suggested by a child psychologist so that our children can express their emotions and feelings in their very own 'for their eyes only' diary. Let your child decorate this box themselves (they love it). A special teddy, toy, blanket or pillow – all make for a more comfortable stay and happier child.
- Take along some healthy snacks and cool drinks to last you both through the day. You will find this a lot cheaper than the kiosk at the hospital
- Always remember the CHILDREN'S BILL OF RIGHTS and make staff aware of these should you need to

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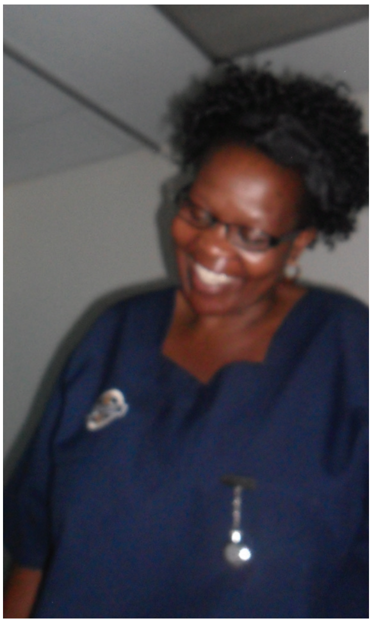
Children's Bill of Rights

- We will tell you who we are and what our role in your healthcare is.
- We will provide a place for one family member to spend the night or stay as close to you as possible at all times.
- Your family can stay together as much as possible. If this is not possible, the people caring for you will explain why.
- We will explain all the things to you and speak in language you can understand.
- You can ask at any time what is happening to you and why.
- You can make choices whenever possible. Please ask questions about anything that is unclear to you or your family.
- Let us know what play and learning activities you like to do to help you grow and learn while in the hospital.



Tips for school

Sending your child to school for the first time and or a new school, even a new grade is a daunting event for parents and children alike. Throw in the fact that your child has PID and it becomes quite a nightmare. There is little to worry about if you follow these helpful tips (all tried and tested).

- Educate the head of the school and teachers as to what PID and your child's condition is.
- Download our 'letter to educators' which you will find on our website.
- Advise other parents to your child's condition.
- Children are to use a hand sanitizer after having used the bathroom or blowing their noses – which should be placed at the door of the classroom.
- You are to be alerted if there are any communicable diseases in the school.
- Advise that your child will have more absenteeism than the other children and will need to have a catch up facility available to him/her.
- Implore that your child is not treated differently and is allowed to participate in all activities – this is most important in maintaining your child's healthy self esteem.

Useful websites

The IPOPI ***www.ipopi.org*** and IDF ***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.

Change of office location

The PiNSA office base has relocated to Pietermaritzburg. The reason is purely logistical as Cally Coldbeck, the Vice Chair of the association, and our secretary Christelle Henning. reside in Pietermaritzburg. Rina has been a marvellous asset for the last two years and it is because of her (and the funding for a secretary from the Binding Site) we have had a regular newsletter. She will continue working with Dr Esser on the national registry so to those who have not returned your signed consent forms, please let Rina have them as soon as possible – as we need to keep a signed consent form for the registry.
Send your consent form to Rina at rina@sun.ac.za.

Note from the chair

Cally Coldbeck (Vice-Chair) and I decided to have a face-to-face meeting to do a catch-up on a number of issues and also a hand-over of documents, equipment etc as the secretary will be in Pietermaritzburg. Cally has the space and a trained person under her wing so we believe this will be the solution. The new secretary is called Christelle Henning, so welcome Christelle to the PiNSA fold!
When Cally and I met we went over the following;

- Statements and claims for the audit: we need to have an auditor's report for the NPO (non-profit organisation) annual reports
- The constitution, I have asked Marcia Boyle of the IDF in the US, Stephen Baxter of IDF NZ and Jose Drabwell of the PiA UK for their constitutions so that we can remodel ours. Ours was based on the Irish PiA constitution originally and needs to be updated (all overseas NMO's (National Member Organisation).
- Governance procedures like file name conventions, e-mail conventions etc. Also what is expected in terms of reporting. e.g. our strategic plan with objectives, monthly financial (petty cash) and work plan reports, a monthly membership update etc.
- Membership: Cally has kindly taken on the job of updating the membership list. We have a new membership form which we are asking people to complete. Remember at this stage there is no cost to being a member of PiNSA, but it will come at some point – for the first time we have had to pay a subscription to IPOPI (International Patient Organisation for Primary Immunodeficiency) to be recognised as an NMO with voting rights.
- Support and information: we worked out a modus operandi for this objective and there will be a structure to patient support. Medical issues go to the MAP (Medical Advisory Panel headed by Dr Esser) and others we will manage. No patient or family will feel left on their own.
- Advocacy: Responsibility for the quarterly newsletter (will be secretary,) and website updating (also secretary but at the moment me).
- Fundraising was discussed and there are a number of strategies including online donation websites.
- Lobbying: issues such as the treatment being on essential medicines and PID on list of chronic diseases are this remit – we need to know who is on what medical aid and any hassles experienced. This information will be kept confidential and we will only use it with your permission.
- Treatment and training: this is the remit of our MAP doctor and her panel, also our immunology nurse

Other than that IPOPI will be conducting a patient survey, more to come on that and we are also preparing for World Immunology Day on 29th April 2011. All ideas will be more than welcome but please bear in mind that implementation is key and capacity is strained. This is a volunteer organisation, only the secretary is funded.

Joy Rosario
PiNSA chairperson