



## Advocating for primary immunodeficiency diseases globally Access to Appropriate Treatment 22nd - 29th April 2013

### coming Events

#### APRIL

12<sup>th</sup>

Dunedin Patient Meeting

22<sup>nd</sup> – 29<sup>th</sup>

PID Awareness Week

29<sup>th</sup>

All4Good 2013

School Mufti challenge

World Day of Immunology

#### MAY

20<sup>th</sup>

Circus Quirkus Hastings

21<sup>st</sup>

Circus Quirkus Gisborne

Visit

[www.idfnz.org.nz](http://www.idfnz.org.nz)

for more in-depth

and up-to-date information  
and registration froms



Visit our new online shop

[www.idfnz.org.nz/shop](http://www.idfnz.org.nz/shop)

For a growing range of  
items for sale which directly  
benefit the Foundation.  
Supporting Patient Events  
and Services.

**IDFNZ supports World PI Week - a global campaign which aims to raise awareness and improve diagnosis and treatment of primary immunodeficiencies as an increasingly important group of diseases.**

Current diagnosis levels suggest that around 1 in 8-10,000 people have a genetic primary immunodeficiency that significantly affects their health. \* In a person with a working immune system, the body recognises pathogens and responds with the production of immune cells and antibodies against that organism. The pathogen can then be eliminated, and the immune system 'remembers' the threat and can respond more efficiently to the same pathogen in future, thus preventing repeat infections.

People with PID are missing one or more of the vital weapons the body has against infection. Deficiencies in immunity can vary in severity, depending on whether one or several parts of the immune system are non-functional.

Thanks to medical advances, most PID's can be successfully treated. With proper therapy most patients are usually able to lead more normal lives. Children usually can go to school, play with friends, and take part in sports. Most adults with PI are leading productive lives in their communities.

The key is early diagnosis and appropriate medical treatment – hence the early warning signs of Primary Immune Deficiency need to be communicated to the wider medical profession as well as parents, school teachers, day care centre employees, researchers and nurses to increase the understanding of these diseases and facilitate early diagnosis. IDFNZ offers a wide range of free resources promoting this message - visit [www.idfnz.org.nz](http://www.idfnz.org.nz) to view these. **Free copies of posters and pamphlets for your workplace and community can be ordered by emailing [info@idfnz.org.nz](mailto:info@idfnz.org.nz).** The Investigate Immunity series of information on the immune and blood systems has been specially developed for classroom use – order a free teaching resource pack now for your school; enrol your school in our **All4Good** school programme to help raise awareness of PID and funds to promote the work of IDFNZ and KIDS Foundation in your community.

\*Chapel, H and Arkwright, P.D. What Are Immunodeficiencies? (2008)

### Celebrating WORLD DAY OF IMMUNOLOGY on the 29th April

Learn more about your immune system and how primary immune deficiencies affect people in our communities.

#### 7 Warning Signs When to suspect primary immune deficiency



**IDFNZ**  
Immune Deficiencies Foundation  
of New Zealand

IDFNZ supports people in New Zealand  
living with primary immune deficiencies.

For more information, visit [www.idfnz.org.nz](http://www.idfnz.org.nz)



## All4Good 2013



Yes it is that time of year again, we are encouraging schools to support KIDS Foundation **All4Good** as their social science subject. In addition to the rewards of helping less fortunate, medically fragile youths in their community, the Foundation is also offering some fantastic prizes for schools participating this year. All members are encouraged to promote this programme in their own school. **Entry forms are online at [www.all4good.org.nz](http://www.all4good.org.nz) and free teacher resource packs can be ordered from the National office [info@idfnz.org.nz](mailto:info@idfnz.org.nz) or 0508 300 600.** Pass on the postcard enclosed in this newsletter to your child's school.



# Recent Events

## IPOPI XIIth Biennial Meeting

Vicki Tattley, our vice chair, represented New Zealand Primary immunodeficient patients and IDFNZ at the recent International Patient Organisation for Primary Immunodeficiencies (IPOPI) XIIth Biennial Meeting – held in Florence, 3-6 October 2012. Vicki was able to meet and network with patient delegates from all over the world. Catching up with the many overseas doctors and patient representatives that we have been fortunate to have had as speakers here in New Zealand was another benefit.

The talks gave an insight into the challenges of living with PID for patients in other countries, many facing difficulties accessing the high quality of treatment we enjoy here in New Zealand. The conference included updates on latest testing and treatment options developing for PID. Vicki was also able to update IPOPI members on progress being made here in New Zealand. New Zealand has been involved with IPOPI since its inception and like all organisations there has been growth and change along the way. IPOPI continues to strive for more National Member Organisations – six new ones were admitted at this Conference.

IDFNZ representation at IPOPI was made possible by a Lotteries grant.



Vicki at the IPOPI XIIth Biennial Meeting in Italy

# Lecture Series

## Promoting Awareness of Primary Immune Deficiency Disorders



Professor Cant with Vice Chair Vicki Tattley

IDFNZ was thrilled to be able to welcome Professor Andrew Cant BSc MD FRCP FRCPCH - Consultant in Paediatric Immunology and Infectious Diseases, Newcastle General Hospital, Director Children's Bone Marrow Transplant Unit, Great North Children's Hospital Newcastle, Hon Clinical Professor of Paediatric Immunology, Institute of Cellular Medicine, Newcastle University, UK, when he visited New Zealand last year.

Professor Cant was able to speak on behalf of IDFNZ at a range of venues in Auckland, Wellington, Christchurch and Dunedin – communicating latest information on diagnosis and treatment of primary immunodeficiency disorders to medical professionals across New Zealand.

Professor Cant's talk on PID Warning Signs, SCID and Newborn Screening was presented to doctors at Starship Hospital and was video conferenced live to doctors across New Zealand and the Pacific Islands. If patient members wish to view a recording of this presentation or communicate it to their General Practitioners it can be accessed at: <https://www.starship.org.nz/for-health-professionals/paediatric-update/2012-archive/pid-warning-signs-scid-and-newborn-screening/>.

This lecture series was made possible with a CSL Education grant.

## Australasian Society of Clinical Immunology & Allergy (ASCI) Conference.

IDFNZ was proud to sponsor the 2012 ASCIA conference held in Wellington. As always the event was well supported by New Zealand and Australian medical professionals keeping up to date on clinical immunology. International keynote speakers included: Professor Andrew Cant (UK), Professor Carlos Camargo (USA), Dr Jörg Kleine-Tebbe (Germany), Professor Ulrich Wahn (Germany), plus approximately 60 other speakers and chairs contributing to the meeting. The New Zealand organising committee included Dr Miriam Hurst as chair, Dr Shannon Brothers, Dr Marianne Empson and Ms Pauline Brown. The content of the conference was excellent providing delegates with latest information from around the world.



IDFNZ Board members Ali Coombes, Laura Hannah, Judith Dickson, Nick Metson and Adriaan Bosch kindly represented IDFNZ at the conference, attending talks and staffing our display stand.

# Medical Update

## Evogam – a Subcutaneous Immunoglobulin Produced from New Zealand Plasma.

Medsafe finally approved the registration of Evogam shortly before Christmas 2012. This new immunoglobulin product is manufactured by CSL Behring in Melbourne using the same processes as for Intragam P but is then concentrated into a product suitable for subcutaneous use. This allows people with immunodeficiency to treat themselves at home. Subcutaneous treatment will work for some, but not all, people. If you are interested in accessing this new form of treatment then you should talk to your local immunologist in the first instance. They are all aware of the new product and are being kept informed of developments as we move to make the product more widely available.

New Zealand Blood Service (NZBS) supported the clinical trials that CSL used to support the registration of the new medicine. This support was conditional on the people involved having on-going access to the subcutaneous product following completion of the clinical trials. This worked well and these people have now transferred to the registered version of the medicine.

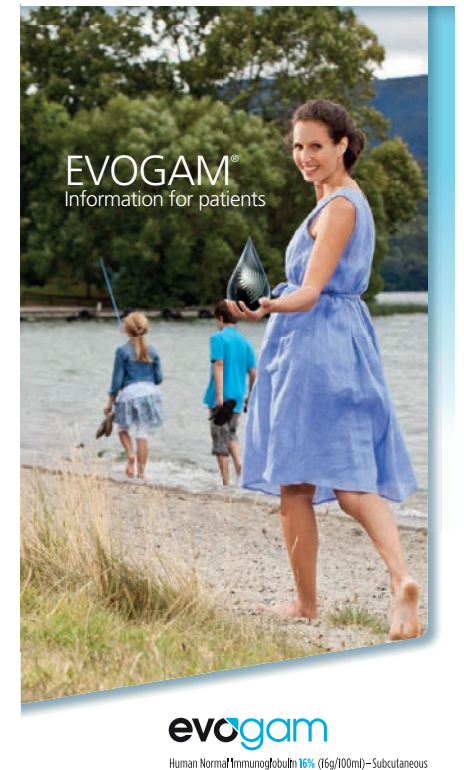
NZBS is now working with CSL to develop timelines for a formal launch of the product sometime in the middle of the year. A number of issues need to be sorted to achieve this.

Firstly we need build up supplies of Evogam and also ensure that we have more than one batch of the product available to ensure certainty of supply before too many people switch to the new product.

Secondly we need to ensure that nursing staff receive education and training in the administration of the product. This will be particularly important for the smaller centres which did not participate in the clinical trials.

CSL have developed a patient information brochure on the product. This is available on the NZBS website (<http://www.nzblood.co.nz/Clinical-information/Transfusion-medicine/Information-for-recipients/EVOGAM-Patient-Information>).

Article written by Peter Flanagan, NZBS.



## Our People

### Living with SCID – A mother's reflections 21 years on

Sheridan was born June 1992. He was a healthy 7lbs 6oz, there were no complications during labour or during my pregnancy. Everything was fine until he was 6 weeks old.

I remember the first time I took him to the doctor he was so sick with a chest infection and he was given antibiotics and paracetamol; we visited the doctors two or three times over the week, once at the GP and twice to the emergency doctors until finally we took him to the emergency department at the hospital.

This was our very first hospital stay, little did I know it would be one of many, one of many hospitals and one of many stays, eventually the hospital would become our home for the next 12 months.

Sheridan was diagnosed with several chest infections, gastroenteritis, pneumonia, ear infections, and asthma and repeatedly treated with antibiotics orally and intravenously. He was continuously sick we would spend two or three days at home and then be back in hospital there would be the same routine including constant blood test, intravenous fluids because he was so dehydrated from a combination of vomiting and diarrhoea and painful thrush in his mouth which made it too painful for Sheridan to drink orally.

The severity of what was happening to him at the time was both upsetting and confusing. "What is wrong with him" I would ask in a panicked tone "is he going to be okay" I watched my baby boy go from being a chubby jolly little baby to being literally skin and bone, he had lost so much weight he was almost down to his birth weight. The reality of what was happening at the time was that Sheridan's blood test did not give any answers or reasons for Sheridan's symptoms or why he was constantly unwell.

Sheridan had finally been discharged from hospital after spending another week or two in the hospital, he still wasn't very well but as the test results did not give us any answers there was little they could do in the hospital, we were sent home with a course of antibiotics and paracetamol and advised to come back if we needed too.

At the time Sheridan still wasn't drinking orally because the thrush in his mouth was very thick and painful so it was hard to keep his fluids up. Later that night he was having trouble breathing.

Continued on page 6







Thank you to all of our friends at Smith & Smith across the country for continuing to support KIDS Foundation.

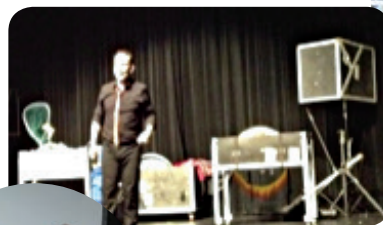


# All4Good Heroes

As an **All4Good workplace**, Smith and Smith has assisted our work in the community. The second half of 2012 was full on with an amazing variety of fundraising events. Some notable highlights were:

**Marty Putz star of Razzmatazz**

**Our Razzmatazz Shows** - helped out by Smith & Smith staff from Invercargill, Dunedin, New Plymouth and Manukau.



Dunedin staff at Razzmatazz Aug 2012



**The Hamilton Smith & Smith Car Day**  
Brendan Judd, a Hamilton member, enjoying a fun day



Thank you, Smith & Smith, for making our children smile with joy ...

**Madison Merrick**

**St Joseph's Takapuna School raised \$1000** via a mufti day

**Irene and Angela raised \$2340** by running the Rotorua Marathon - read their story at [www.idfnz.org.nz](http://www.idfnz.org.nz)

**Proving distance is no barrier, Michael Merrick raised an amazing \$36,720**, running the Dubai Marathon for Madie. Helped by pupils and staff at Qatar Intl School.

**Mandy Bennett raised over \$12,000.** Mandy worked tirelessly coordinating a special event at Spencer on Byron for Madison, as well as overseeing a host of other fundraising initiatives.

**The Dragon Trust** kindly donated a special pushchair for Madison to use overseas.



**Highbrook branch** held an amazing quiz night,  
**Christchurch branch** held a monster garage sale,  
**Rotorua branch** a sausage sizzle.  
There were many more events across NZ.



*We can't mention everyone here.... but be assured whichever*



*cause you are supporting, your efforts really do make a difference!*



**A sweet start to 2013** – Smith & Smith have collaborated with Cadbury to offer a range of tasty Cadbury snacks for sale at Smith & Smith customer service points around the country.

These smart point of sale displays give customers the opportunity to relax with a sweet treat whilst waiting for Smith & Smith experts to repair faulty windscreens. Smith & Smith clearly strive to deliver excellent customer service, and of course each sale benefits KIDS Foundation.

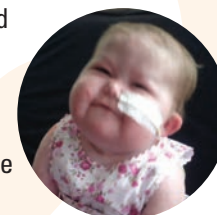
**Members can support this fundraiser by popping into their local Smith & Smith for their chocolate bars.**

**Zoe Edwards**

**Dance for Zoe team who raised \$1565** for gorgeous Zoe

**Lisa Mullins** who is running a marathon in Whakatane for Zoe supported by her Fonterra colleagues.

**The HB Williams Turanga Trust and WH Williams Trust** for supporting Zoe



**Well done, Shirley, for raising over \$2700 for our cause ... thank you!**

Shirley Davy of Auckland ran the Auckland half marathon to raise funds for IDFNZ KIDS Foundation (view the Howick and Pakaranga Times online article at <http://www.times.co.nz/news/pounding-pavements-paying-off.html>) Shirley's gorgeous son Sam has MMIHS, an extremely rare medical condition. When Sam was born, Shirley and her husband Francis were told he was unlikely to survive a year. Sam has spent more than half his life in Starship Children's Hospital. Shirley's aim in running the half marathon on October 28 was to put the spotlight on the Kids Foundation and how it supports Sam and children like him.



What I need to mention here is that Sheridan would have been 9 and a half months at this stage so this gives an indication of how long his battle had been going on for, keeping in mind his first hospital stay was at 6 weeks old.

Sheridan was crying, not a normal cry but in pain, his face was red and he was shaking. My dad drove us to the hospital and we rushed into the emergency department, they sent for a paediatrician straight away and we were seen immediately. I remember being really scared at this point and we were sent to the children’s ward.

I remember they did a lumbar puncture that night and I couldn’t cope with seeing my baby go through that again so my dad stayed with him.

Later we were sent to x-ray things just went from bad to worse. Sheridan started to have convulsions, he was on oxygen and had lines going in left right and centre. This is where we would meet the paediatrician who would give us the answers we were looking for.

Dr Paul Heaton was working that night and thank god! My baby was dying and until this point there was nothing they could do. Dr Heaton did various test and looked at various research over that week always keeping me updated he knew time was running out for Sheridan so a diagnosis was urgent.

He had placed us in isolation and had placed strict rules around access to our room. During the afternoon he and several other paediatricians came into our hospital room where he told me that they had finally found a diagnosis. He said Sheridan had Severe Combined Immune Deficiency. S.C.I.D he explained briefly the condition and told me that I would need to call my family as the air ambulance would be flying us to Auckland’s Starship Children’s Hospital first thing in the morning.

We left for Auckland the next morning, I still had no idea what the heck was happening it was all moving so fast. I am grateful that my family is as supportive as they are; my family saw us off and then drove to Auckland.

We were placed in oncology and we met Dr Ameratanga for the first time, he came back to see us that afternoon and told me that for Sheridan to have any chance of survival he would need a bone marrow transplant which would need to be done in Sydney Australia.

He made all the arrangements and we had 48 hours to get passports and get packing. We were sent to Prince of Wales Children’s Hospital Sydney; it would be another 2 weeks before Sheridan would get to have his transplant. There were complications which set back the timeframe for the transplant, I was to undergo surgery, a bone marrow harvest, and finally Sheridan received his bone marrow transplant on the 14th of April 1993.

We were to spend another two weeks in Sydney before returning back to New Zealand. Sheridan’s recovery was long and rigorous, just because he had received the bone marrow transplant didn’t guarantee Sheridan’s survival. If anything this was our most worrying and frightening time, we knew that there was nothing more that could be done the transplant had to work to save his life. I couldn’t bear to lose him.

Sheridan ended up in intensive care while in Starship Hospital and things weren’t looking good, I was even advised to call family, something they had never advised me to do before.

I was so scared. Sheridan was unconscious and I sat with him and held his little hand and asked him not to leave me. This is the first time I have talked about that moment.

My family drove from New Plymouth to Auckland and arrived that night. The following morning I was still awake and my baby looked at me and said “mama” I couldn’t pick him up because of the tubes everywhere so I just laid next to him and cuddled. The following days he would go from strength to strength.

We would spend then next 10 or so months in Starship in isolation until finally we got the ok to go home to New Plymouth. I couldn’t wait; I had been home a few times over those months when Sheridan’s grandparents or dad would look after him for me. We had also lost my grandmother and grandfather over those 10 months so we were dealing with a lot.

Sheridan turns 21 on the 18th of June this year and I am very proud of him. Although he has IVIG every month and visits to Auckland, surgeries, and various medications and appointments, he is still positive.

He is the quiet achiever and has great friendships. Sheridan celebrates life through living it, he doesn’t see having SCID as sentence, and in fact he has a really good sense of humour about it at times and has learnt to appreciate what he has to live with. He has been described as inspiring and was invited to attend the Young Aspiring Leaders Conference in Wellington in 2011 which he attended.

I want to thank the Kids Foundation for the support they continue to give to our family. Over twenty years the IDFNZ has supported our family and have walked beside us through it all. We are eternally grateful.  
Lisa Tamati.

*Severe combined immunodeficiency (SCID) is a life threatening medical condition, a paediatric emergency. Children with SCID lack virtually all immune protection against bacteria, viruses, and fungi and are prone to repeated and persistent infections that would not normally cause illness in a person with a normal immune system. In a SCID child, these infections can be life-threatening. SCID is estimated to occur in approximately 1 out of every 50,000 to 100,000 births. New born testing for SCID has become routine in recent years across many USA states; IDFNZ is supportive of introducing this initiative into New Zealand to facilitate early diagnosis and treatment.*

## Recent Events

As usual November and December included IDFNZ KIDS Foundation families joining together for Christmas celebrations. We would like to thank our sponsors, staff and volunteers for making these events possible.





# All4Good Payroll Giving

## Want an easy way of making regular, no fuss donations to the Foundation?

Payroll Giving allows you to give a chosen amount to a charity directly from your regular pay – you just let your employer know how much, how often and ask if they can set it up for you.

## What's the advantage for you?

- Your tax rebate is already accounted for so you don't have to save receipts and submit a tax claim. So, if you decide to give \$30 each pay cycle, your tax rebate of 33.3% will be automatically calculated so you will only be giving \$20 from your pay.
- It is a simple, direct way to support IDFNZ KIDS Foundation. 100% of your donation reaches the Foundation.
- Once it's set up with your employer you don't have to do anything further.
- You might also like to encourage others in your organisation to join you in supporting the IDFNZ KIDS Foundation- be our 'All4good Hero' within your workplace

To find out how to start Payroll Giving, please contact us at [info@idfnz.org.nz](mailto:info@idfnz.org.nz)

## Hand-made Cards

Quality Hand-made cards for all occasions. At last a range of simple original cards that speak for you.

Photography has always been a hobby and a passion for patient member Michele Kay Sharman. A welcome release from the stress of living with a chronic health condition. Michele has turned her hobby into an opportunity for IDFNZ by offering members and supporters a new range of quality greetings cards perfect for any occasion.

A range of 12 cards is available singly and / or in packs of 6 – available from our online shop – each purchase directly benefits IDFNZ.



## Our Supporters

Smith & Smith  
Lion Foundation  
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## Sam's All4Good Team

When Sam Davy was born, his parents Shirley and Francis were told he was unlikely to survive a year. Sam has a rare medical condition resulting in severe gastrointestinal failure. Sam has beaten the odds, but to do so has spent more than half his life in hospital.

Through this journey to wellness Sam and his family have been supported by KIDS Foundation and last year Shirley ran her first half marathon to help put the spotlight on IDFNZ Kids Foundation and how it supports Sam and children like him.

This year she has raised the bar and is leading a whole team to run the full Auckland marathon in November. The team is named in honour of Sam, the inspiration for Shirley running. The funds raised will go to IDFNZ KIDS Foundation All4Good fund to support their work with other families facing similar medical challenges.

Shirley has not only volunteered herself, she has also enlisted family members from all over New Zealand and Australia to help the Foundation.

In her own words: "Last year I set myself the personal challenge of running my first half marathon for IDFNZ - no mean feat for this couch potato! The online fundraising was easy to set up, and then it was just down to me to get out and pound the pavement. I had to make time for training, but eventually the enjoyment overcame the pain..... and the satisfaction of running over the Auckland Harbour Bridge then over the finish line was worth it! Nearly \$3000 later, I finished - thanks to the financial sacrifice of many individuals who sponsored me to run for IDFNZ! I am amazed at how generous and supportive people were..... so this year I threw the gauntlet to my extended family and they have taken up the challenge to run either the half or full marathon! My brother and his wife are flying in from Perth, as are my sisters from Gore, Dunedin & Havelock North, a sister-in-law from Invercargill and a niece from Christchurch! This year I am going for my first FULL Marathon, which I'm nervous about, but happy to be running and raising awareness for IDFNZ. If you want to join us on November 3, we would love to have you as part of Sam's All4Good Team! - Shirley Davy.

Members who would like to join Shirley and take up her challenge should first register for the marathon at <http://www.aucklandmarathon.co.nz>

And then visit <http://www.givealittle.co.nz/cause/SamsAll4Good> to set up a free champion's page of their own to join as a member of Sam's All4Good team

Not up to running? How about logging onto <http://www.givealittle.co.nz/cause/SamsAll4Good> to leave a donation and a message of encouragement to the team.

This edition of **intouch** has been kindly sponsored by

**Four Winds**  
FOUNDATION LIMITED  
supporting people with mental health issues