



# DEBRA NZ Newsletter

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## DEBRA NZ Conference 2007 will be in Auckland, Friday 17<sup>th</sup> to Sunday 19<sup>th</sup> August.

Our guest speakers include Prof Marcel Jonkman and EB Nurse Jose Duipmans from the Netherlands, and Prof Dedee Murrell from Sydney.

We are combining with the NZ Dermatologists and Dermatology Nurses Conferences for a half day seminar on EB on Saturday afternoon, 18th August. Our international speakers will be part of an outstanding panel of experts including Prof John McGrath from the UK, who will give presentations on EB research, care and treatment.

Other medical professionals and friends, family and supporters of DEBRA NZ are also invited to this session. It will be a great opportunity for the medical profession and the public to learn more about EB.

The DEBRA NZ Conference will be based at the Copthorne Hotel, 196–200 Quay St, Auckland, with the Saturday afternoon session across the road at the Hilton Hotel on the waterfront.

There will be a fun children's programme including art and drumming on the Saturday and an outing on the Sunday, and a crèche for the younger ones.

### Accommodation and Travel Arrangements

Please confirm if you will be attending asap by phone, email, or mail, letting us know your travel plans, number needing accommodation at the Copthorne on which nights, special dietary needs and ages of children who will be in the crèche or part of the children's programme.

We are very pleased to be able to subsidise your travel costs with the help of a grant from the NZ Lotteries Grants Board. Please let Anna know asap if you need help with booking airfares to Auckland

(ph. 04 389 7316 or email: trustees@debra.org.nz).

We hope accommodation costs will be subsidized with further grants that have been applied for. Our aim is to make sure cost doesn't stop people with EB and their families from being able to spend this time together and learn about the latest research and developments in EB care.

**Sad news:** We are very sorry to inform those of you who had not already heard, that Neville Stevenson died on 8th January this year. He was 59 years old, and was an active member of DEBRA NZ from it's beginning. Neville was a wonderful and courageous man and will be very sadly missed by all of us. His parents Noel and Ella wrote: *"To our special DEBRA family. A special thank you for standing by us and Neville when things got bad. It helped us immensely to have so many of you at his funeral. Our sincerest love and blessings to you all."*

## Gene therapy in Italy for Junctional EB

An Italian research team has treated a 36-year old man with Junctional EB with skin grafts that contained corrected copies of the laminin-5 gene. The team at University of Modena, led by Professor Michele de Luca, has recently reported the results in the medical journal Nature Medicine.

The experimental treatment removed pieces of the patient's skin from his palm. Epidermal skin stem cells were then grown in the laboratory from these biopsies and corrected copies of the laminin-5 gene were inserted into the cells.

These corrected cells were then grown into sheets of skin in the laboratory. The new skin was grafted to the front of the patient's thighs, where the skin had many non-healing wounds. In total, nine sites were grafted on his thighs. These grafted areas had healed completely after a week and a year later the areas were strong and blister free, while the non-grafted areas continued to blister. The patient will receive additional grafts over time and the results in all areas will be documented.

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## Gene therapy in Italy for Junctional EB

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"This effort by Professor De Luca's group provides a key 'proof of principle' that confirms the assumption that gene therapy may work for JEB", DeBRA Scientific Advisory Board Member Dr. Jo-David Fine said. "Longer term study will be done to see if the skin cell corrections remain effective over time."

DeBRA Europe is a partner in a European Union-funded program, Skintherapy, which will help to take the work further.

DeBRA Scientific Advisory Board Member Dr. Jouni Uitto added that 'in research we have to proceed cautiously because this approach is experimental and promising results in a single patient must be reproducible in other patients. In addition, this approach is expensive and not approved in the United States or any country except Italy. Finally, this approach is suitable only for a selected subset of patients with EB. Nevertheless, it is a promising start for future gene therapy for EB.'

DEBRA will provide updates about this work as they become available. Professor De Luca is to be one of the guest speakers at the Australasian EB Professionals Meeting in Sydney and the DEBRA Australia Conference this June.

### Introducing Logan and family

Hello everybody, we are Nichola Edwards, Graham Andersen and our beautiful wee boy is Logan Andersen.



As some of you already know we live in Ngongotaha, Rotorua. Graham was born and bred here. I am originally from Carterton in the Wairarapa and have now been living here for nine years. Graham and I have been together for just over five years now and are planning to tie the knot in March next year which is really exciting.

On 29<sup>th</sup> September 2006 we had wee Logan. Our pregnancy wasn't the greatest anybody could ever have and if there was a complication to go with pregnancy, we got it!! Logan was subsequently born via caesarean 5 weeks premature weighing in at 4lb 3oz, but he was safe and well.

At 3-4 days old we noticed some blisters on two toes of both feet and some under his chin and that is when we were told he had suspected EB. This was a real shock to us as neither of us had ever heard of this. Never-the-less we are very lucky as specialists seem to think he has the simplex version of EB. We are currently awaiting the biopsy results which went to Sydney last week thanks to Waikato Hospital, Hamilton.

We all wish to say a **HUGE thank you to DEBRA NZ** for the superb support you have given us, especially Deanna who has given us a fair bit of her valuable time. Another **BIG** thank you to the other families who deal with EB in their daily lives. Your support, visits to us, words of wisdom and advice has been such an enormous help and without you, I am sure both Graham and I would be near the point of insanity by now.

We are looking forward to putting a lot of faces to names at this years EB conference in Auckland in August. Until then, take care and if there is ever anything we can do to return all the favours we have been given please do not hesitate to contact us :o)

Email: [granic@kol.co.nz](mailto:granic@kol.co.nz)

## A Catch up With the Riordan Family: Hannah turns 2 !

Hannah turned 2 this January. She adores the Wiggles so had a wiggly party. It was a small celebration this year as she had the chicken pox, the spots appearing the day before her birthday. We were very apprehensive because of her EB but she coped very well with little itching and her skin handled the extra blistering well. The worst was her tummy which is very sensitive and many of the pox left large raw blistered areas.

Her highlight so far this year would have to be a visit to Wiggles World in Australia where she met 3 of the wiggly characters. We were also lucky enough to meet the Woodward family from the Sunshine Coast who have 15 mth old Lochlan who also has dystrophic EB. Thanks Deanna for putting us in touch with them!

Hannah is an active 2 year old who loves to discover new things. She plays hard, sings loud, laughs, is stubborn, has a gentle nurturing side and a feisty destructive side. Her allergies have improved heaps which helps to keep her skin more intact. She visited the local beach and local swimming pool for the first time this summer. Hannah is doing well and developing as a 2 year old should.

Jon and Louise wish we had even half the energy Hannah has. We are busy doing renovations on our home. Jon has installed a new kitchen and built a deck. Louise tries to fit in a bit of decorating when time and space permit.

We look forward to seeing you all at the DEBRA NZ Conference in Auckland.

Hope your days are going well,

Louise & Jon

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### DEBRA Conferences & events coming up:

- Australasian EB Professionals Conference, 14th June 2007, Sydney
- DEBRA Australia Conference, 15th to 17th June 2007, Penrith, NSW
- DEBRA NZ Conference, 17-19 August 2007, Auckland
- EB Adventure Camp, 20-26 August 2007, National Park & Ruapehu
- Chile EB Symposium, 27-28 September 2007, Santiago
- DEBRA Europe/International Conference, 2-4 November 2007, Avignon, France
- DEBRA NZ Families Camp, 25-30 April 2008, Rotorua

*The 3 NZ nurses, Jacqui, Deanna and Sharon, and Anna & Martin will attend the EB Professionals meeting on medical & research issues in Sydney. They will also participate in the 2nd DEBRA Australia Conference, along with a few more DEBRA NZ members.*

**\* The winter Adventure Camp will be held again this year at National Park and Mt Ruapehu from Monday 20<sup>th</sup> to Sunday 26<sup>th</sup> August. All young adults with EB are welcome. Please contact Anna KW or Deanna Forsyth if you are interested in joining us.**

## Directors report

Anna Kemble Welch

The last year has been very busy for DEBRA NZ. The annual conference in July 2006 was the biggest yet with about 85 adults and children from around New Zealand enjoying a weekend with other DEBRA families in Rotorua; we held our first winter adventure camp for teens and young adults with EB near Mt Ruapehu; and a third nurse has joined the DEBRA nursing team part time to help with support and advice for people with EB in the South Island. It has been great having Michal McCracken's help with DEBRA admin stuff for a few hours a week, particularly with the fundraising applications.

Membership continues to grow as existing EB families hear of us for the first time and new babies are born with blistering skin and diagnosed with EB. Recent TV coverage and magazine articles have increased the awareness of EB amongst the public and medical professionals and helped improve understanding.



DEBRA NZ Conference, Rotorua, July 2006



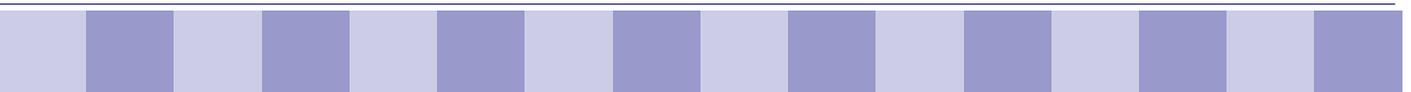
### Winter Adventure Camp, August 2006

The EB adventure camp last winter was a great success. EB may mean painful blisters and wounds and hours of daily bandaging for some, but the brave and good humoured group were undaunted by rain and snow to make the most of their experiences. They climbed or were hoisted up a 20 meter wall at the Outdoor Pursuits Centre and abseiled down, skied, tobogganed and snow boarded on Mt Ruapehu and canoed down the Whanganui River.

The group of young (and not so young!) New Zealanders and Australians with EB and a couple of their friends spent a wonderful week around National Park with DEBRA nurses Deanna Forsyth and Jacqui Finnigan; Sandor as breakfast cook, van driver and general entertainer, and me as camp mum. We were lucky to have the help again of instructor Jim Gilligan with his experience of our previous EB Camp and expertise in adaptive recreation. Give him a roll of duck tape and he can do anything!

### International EB Research and DEBRA Conferences

Participating in the International EB Research Conference in Ireland and the DEBRA Europe/International Conference in Austria in October last year was a great privilege for me, making and reinforcing important links to international expertise and connecting with other DEBRA groups from around the world. Having a NZ presence at these important events has made it possible to bring together the fantastic line-up of international speakers for our conference this year.





## DEBRA NZ Nurses update

Hello everyone,

We would like to welcome a new addition to the team: Sharon Cassidy. Sharon has been a nurse for 25 years and specialises in paediatrics, burns and wound care. She has a wealth of skills & knowledge and we look forward to her input into the DEBRA nursing team.

### The DEBRA NZ nursing cover is now as follows:

**Jacqui** – Upper North Island (Hamilton and North)

**Deanna** – Mid and lower North Island (Sth of Hamilton)

**Sharon** – South Island

It was great to see everyone at the conference last year in Rotorua. Jacqui and Deanna enjoyed their time at the winter camp at Mt Ruapehu and we are all looking forward to this year's conference in Auckland and the adventure camp following. We feel privileged to be a part of this and be welcomed into the DEBRA family.



Jacqui, Deanna & Sharon with Anna KW at Butterfly Creek

### Updates:

**Jacqui:** I continue to work 2 days a week with the community nursing team based at Starship. I have been working for DEBRA NZ on other days and have spent time, this year and last, liaising with various specialists within Starship, Auckland City and Middlemore Hospitals in an attempt to ensure the total health needs of clients and their families are met. I thoroughly enjoy working with DEBRA NZ and am excited about what we may achieve in 2007 and beyond. I will be giving a presentation on nursing management of clients with EB at the Dermatology Nurses' Conference in August, increasing awareness of EB amongst health care teams.

**Deanna:** My year started with two months skiing near Lake Tahoe in California. It was a great time and now I am happy to be home and getting stuck back into work with DEBRA NZ. During winter last year I completed a postgraduate paper in wound management at Massey University. It was very worthwhile and provided me with current and up to date wound care practices that I can use in my work with EB skin care. I also participated in a Dermatology Conference in Taupo in September 06. DEBRA NZ included an EB brochure in every pack for each Dermatologist and Nurse which was a great way to educate more people about EB.

**Sharon:** I currently work 3 days a week as a Registered Nurse and the Wound Resource Nurse for the Paediatric Department at Christchurch Hospital, and have recently started a community nursing service.

I am a member of the NZ Wound Care Society, Australia, New Zealand Burns Association, NZ Paediatric Society, and Nurses for Young Persons in Aotearoa. Last year I completed my Postgraduate Diploma through Otago.

This year I will present a paper at the NZ Wound Care Society Conference on EB and attend both the Australia and NZ DEBRA Conferences and Camp. I am very excited to be working within the DEBRA family and look forward to advocating for your needs.

### Standardised Bandage List:

We are in the process of working with Dr Nick Birchall to renew the current bandage list. This should be completed with renewed Special Authority letters being sent out before the end of June.

If you have any products that are working well (or not so well) and would like to share your experience with others, if you have heard of a new product you may like to try, or if you would like more information about any of the products on the list, please feel free to contact any of the nurses and we will help where needed.

### EB Nurse contact times:

It is always great to hear from you with good news or with things you may like help with. Please help us organise our DEBRA working day by contacting us Monday to Friday between 9am – 5pm if possible. Outside of these hours please email or text us and we will then contact you as soon as we are able. Of course, if there is anything urgent feel free to contact us at anytime. Similarly if you would like to be contacted in a certain way or at a certain time please let your EB Nurse know so she can work in with your schedule.

Kindest regards,

**DEBRA NZ Nursing Team**

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