

# Epidermolysis Bullosa in Australia and New Zealand

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## KEYWORDS

- Epidermolysis bullosa • Dystrophic epidermolysis bullosa
- Epidermolysis bullosa simplex

Australia has a population of about 24 million people, and New Zealand, separated from Australia by a 2-hour flight, and consists of the North Island and South Island, has about 5 million people. Australia is a larger land mass than the continental United States, and 90% of its population are concentrated in eight major cities, all of which, except the capital, Canberra, are on the coast. These are the capital cities of the former territories/colonies of the United Kingdom, which only federated into one nation in 1901. Hence, managing patients with an orphan disease presents particular difficulties of distance and tradition. The largest populations are Sydney, New South Wales (4 million); Melbourne, Victoria (3 million); Perth, Western Australia; Brisbane, Queensland; Adelaide, South Australia; Canberra, Australian Capital Territory; Hobart, Tasmania; and Darwin, Northern Territory (**Fig. 1**).

The first epidermolysis bullosa (EB) clinic to be established was in Sydney, by Mark Eisenberg, a general practitioner with a personal interest in EB. He had the support of the late Brien Walder,

head of dermatology at Sydney Children's Hospital (SCH), and Kieran Moran, a pediatrician was brought in as well. In 1996, I was invited by Walder to assist with this clinic as the dermatologist, since I had been taking care of the EB registry patients at Rockefeller University in New York from early 1994. At that stage there, was ad hoc electron microscopy (EM) in different units and no immunofluorescence mapping (IFM) in Australia. I set up a national diagnostic laboratory for IFM and EM for EB based at St George Hospital, Sydney, within the anatomic pathology department. From 1996, the laboratory has received specimens from around Australia and New Zealand and some surrounding Asian countries shipped in Michel's media.<sup>1-4</sup> EM is provided by C.W. Chow of Royal Children's Hospital, Melbourne. In addition, I began seeing adults with EB at St George Hospital and in my part-time private practice, assisted by my practice nurse, Lesley Rhodes.

Since this first EB clinic was established in Sydney, two other EB clinics started, one at Royal Children's Hospital, Melbourne, run by

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Funding has been provided by the St George Hospital for salary support for clinical time at St George Hospital and Sydney Children's Hospital with epidermolysis bullosa (EB) patients. Premier Dermatology Clinical Trials, Kogarah, for salary support for EB nurse and research staff. University of New South Wales for medical students and scholarships for Masters of Science and PhD students.

Much of this work was supported by feeder grants from DebRA Australia, DebRA New Zealand and DebRA in New South Wales, South Australia, Victoria and Queensland. Premier Dermatology Clinical Trials have supported salaries and conference expenses for fellows and researchers Eleni Yiasemides, Linda Martin, Niken Trisnowati, Ningning Dang, Matt Kemp, Adam Rubin, Becca Cummins, Anna Liza Agero, Wenfei Yan, Supriya Venugopal, and Lizbeth Intong, and medical students John Frew, Julien Lahmar, Mary Alice Nading, Yong Kho, and EB nurse Lesley Rhodes. Support from SEALS, Anatomic Pathology, St George Hospital, Sydney, for the national EB diagnostic laboratory and the molecular diagnostic laboratory at Royal Brisbane Hospital, Brisbane, for storage and assistance with genetic testing and prenatal diagnosis.

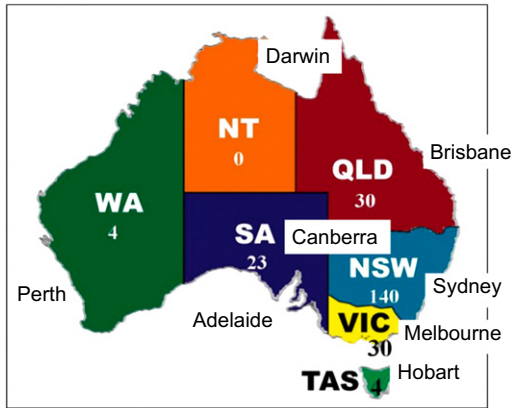
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Dermatol Clin 28 (2010) 433–438

doi:10.1016/j.det.2010.02.009

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**Fig. 1.** Geographic distribution of the major cities and states/territories of Australia showing the numbers of EB patients in the Australasian EB registry from each area.

dermatologists George Varigos, John Su, and David Orchard, and in Adelaide at the Women's and Children's Hospital, originally by Julie Wesley and now by Lachlan Warren. The EB clinic at SCH was expanded with the addition of a medical geneticist, Anne Turner, in 2000, and an additional dermatologist, Orli Wargon, in 2003, after the death of Walder, and a part time EB nurse in 2007, Louise Stevens. In addition, physical, occupational, and pain therapists attend the clinic, which is held monthly as well as a social worker. Newborn infants with EB usually are transferred to SCH from other hospitals in the state of New South Wales.

When EB patients are 16 years of age, they transition to St George Hospital, Sydney, another teaching hospital of the University of New South Wales Medical School. Here the more severe patients with recessive dystrophic EB (RDEB) are reviewed every 3 months in the ambulatory care

unit, with a full skin check after a bath, possible biopsies, and infusions as needed, while they are reviewed by designated experts in hematology, renal, endocrinology, pain service, gastroenterology, as needed (Fig. 2).

In New Zealand, Nick Burchall established the service for EB, based in Auckland, now assisted by Deanna Purvis, and Dystrophic Epidermolysis Bullosa Research Association (DebRA) New Zealand raised funds for several regional EB nurses who assist with the management of EB patients around the country with him and regional dermatologists.

### AUSTRALASIAN EB REGISTRY

In 2005, the author established a national EB registry based at St George Hospital. There are currently 242 patients enrolled, 140 of whom are in New South Wales (see Fig. 1). According to figures by the DebRA charities in New South Wales, Victoria, and SA and the newly formed DebRA Australia, there are likely to be about 400 patients with more severe forms of EB and probably 1000 or more if milder cases are included. DebRA New Zealand has about 40 members (A Kemble-Welch, personal communication, May 2009), and 11 are currently enrolled in the registry. To be enrolled, patients have to be examined by one of the EB clinic dermatologists and have confirmatory biopsies or genetic testing to confirm the subtype of EB. Currently the prevalence of EB is 10 per million population, but in reality it will be higher than this.<sup>3</sup>

### PROVISION OF CARE

In Australia, the public hospital clinics are all free to patients with citizenship or permanent resident status. What is not covered is the cost of the



**Fig. 2.** Transition to adult care at St George Hospital with accommodation for patients at Bezzina House and review at the Ambulatory Care Center with infusions and patient education.

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