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ORIGINAL RESEARCH

The distribution of epidermolysis bullosa in Australia with a focus on rural and remote areas

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ABSTRACT

Background/Objectives: Patients with epidermolysis bullosa (EB) require specialised medical care. In Australia this expertise is located in the major cities, with patients living in rural and remote areas having reduced access to these services. We aim to analyse the geographical distribution of patients with EB in Australia to determine the relevance of this potential geographical disadvantage for this population.

Methods: Using postal codes obtained from the Australian National Diagnostic Laboratory Database for EB and the Australasian EB Registry, living patients with EB in Australia were categorised using the Australian standard geographical classification, remoteness areas. An analysis of EB subtype, including severity was also performed.

Results: A total of 318 patients were categorised, of whom 221 lived in major cities, 65 in inner regional areas, 26 in outer regional areas, four in remote and two in very remote areas. Half the patients living in remote and very remote areas had severe forms of EB.

Conclusions: A significant proportion of patients with EB live outside the major cities in Australia. Half of the patients living in remote and very remote areas had severe forms of EB. Targeted strategies to improve access to EB-specific medical care may be needed for patients living in rural and remote areas.

Key words: epidemiology, epidermolysis bullosa, healthcare delivery, rural.

INTRODUCTION

One-third of the Australian population lives in regional and remote areas. [1] This population tends to experience poorer health outcomes than the two-thirds living in the major cities. [1,2,] There are several reasons for this disparity. A higher proportion of indigenous people live in regional and remote areas and overall have poorer health than the non-indigenous population of Australia. [1,2,] Additionally, limited access to specialist medical care in the rural setting poses a geographical disadvantage. [3] Patients with complex chronic diseases requiring frequent specialist review, as is the case in epidermolysis bullosa (EB), are therefore at a particular disadvantage.

EB is a heterogeneous group of rare genodermatoses characterised by skin and mucous membrane fragility. Four subtypes of EB exist including EB simplex (EBS), dystrophic EB (DEB) (including dominant and recessive forms), junctional EB (JEB) and Kindler syndrome (KS) which all differ in their severity and clinical presentation.[4] EB is a complex and often multi-system disease requiring a multidisciplinary team (MDT) approach to care with specialised clinician input. In Australia EB-specific MDTs are present in specialised EB centres located in Sydney, Melbourne and Adelaide.[4] Dermatologists specialising in EB are additionally located in Brisbane and Perth. Given the rarity of the disease, there are currently no specialised outreach clinics. Patients with EB living in rural and remote areas therefore have to travel long distances to cities to access disease-specific services.

Abbreviations:

ASGC-RA	Australian standard geographical classification, remoteness areas
DEB	dystrophic EB
EB	epidermolysis bullosa
EBS	EB simplex
JEB	junctional EB
KS	Kindler syndrome
MDT	multidisciplinary team

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The aim of this cross-sectional study was to analyse the distribution of patients with EB in Australia to determine whether there is a population of patients living in rural and remote areas exposed to a potential geographical disadvantage.

MATERIALS AND METHODS

The Australian National Diagnostic Laboratory Database for EB was developed at St George Hospital in Sydney in 1996.[5] The Australasian EB Registry was developed in 2006 and approved by the local institutional review board.[6] A cross-sectional analysis was performed using patient data from both the registry and database, which was accessed on the 30 November 2014. Inclusion criteria were living patients with confirmed EB located in Australia with a listed postal code. Data gathered included EB subtype and postal codes which was used to determine their Australian standard geographical classification, remoteness areas (ASGC-RA). The ASGC-RA classification is a measure of rurality or remoteness and is based on road distance to the nearest urban centre[7]. The five categories of the ASGC-RA classification include major cities, inner regional areas, outer regional areas, remote and very remote areas. The percentages of patients in each ASGC-RA category were calculated and rounded to one decimal place. Data were additionally analysed in terms of EB subtype and if patients had a severe form of EB, which included Dowling–Meara and recessive forms of EBS, Herlitz JEB and Hallopeau–Siemens recessive DEB.[4]

RESULTS

The Australian National Diagnostic Laboratory Database and the Australasian EB Registry contained 388 patients with confirmed EB. In all, 23 patients were excluded from the study as they lived outside Australia and 37 were excluded because they were deceased. A further 10 patients were excluded as they did not have a postal code listed. This left a total of 318 patients for inclusion in the study.

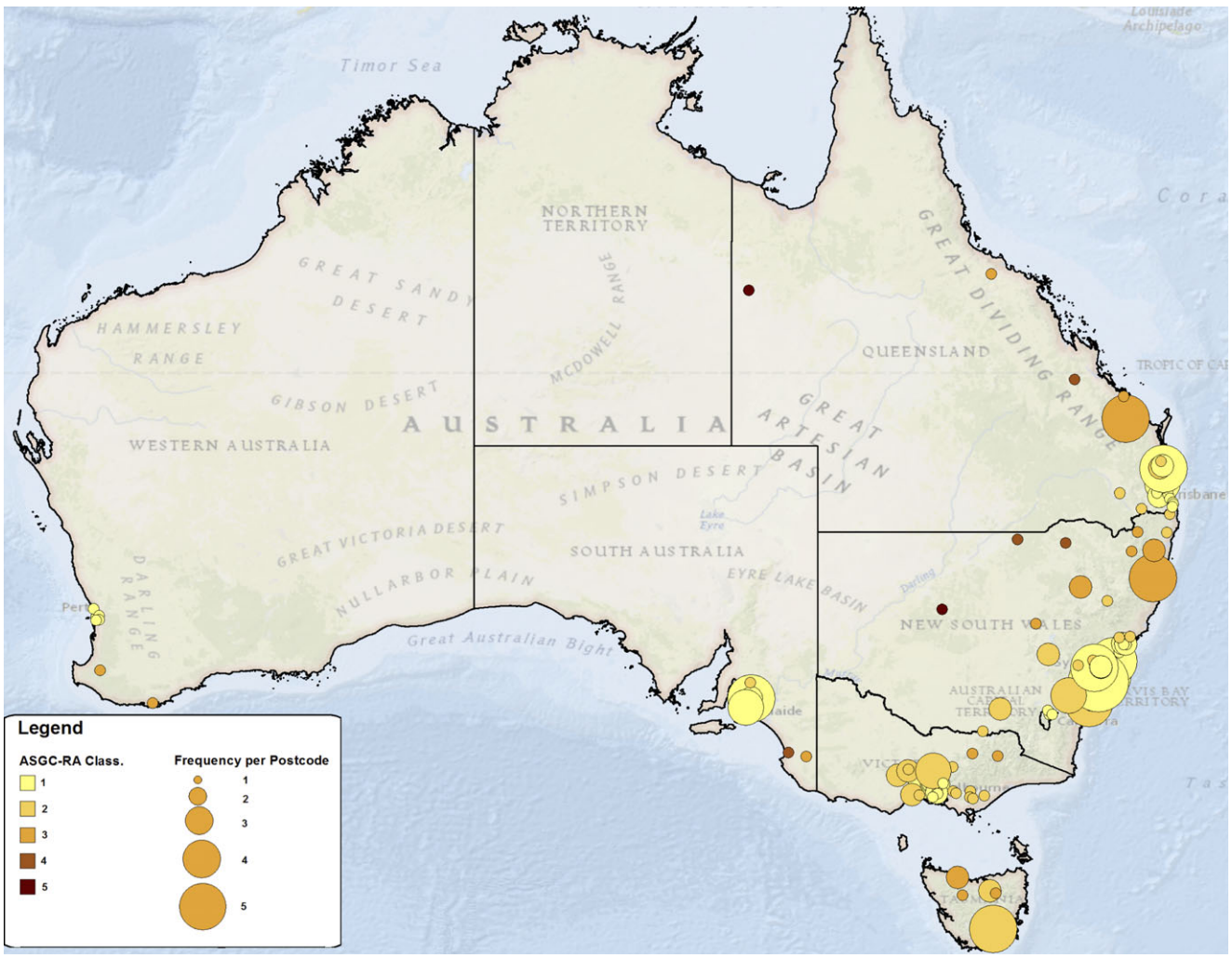

Figure 1 Map of Australia showing the geographical distribution of patients with epidermolysis bullosa and the corresponding Australian standard geographical classification, remoteness areas for each patient. 1, major cities, 2, inner regional areas, 3, outer regional areas, 4, remote areas, 5, very remote areas

As defined by the ASGC-RA classification system, 221 patients lived in major cities, 65 in inner regional areas, 26 in outer regional areas, four in remote and two in very remote areas, as can be seen in Figures 1 and 2. This gave percentages of 70, 2, 8, 1% and 0.6%, respectively. Figure 3 shows a comparison between the proportion of patients with EB and the population distribution of Australia across each ASGC-RA category, using data from the 2006 census. A total of 161 patients had EBS, 108 had DEB (68 with dominant DEB, 28 with recessive DEB and 12 with a form of DEB not yet classified), 17 had JEB, one patient had KS and 31 did not have an EB subtype yet classified. Of the 40 patients who had severe subtypes of EB, 29 lived in major cities, seven in inner regional areas, one in an outer regional area, two in remote areas and one in a very remote area.

DISCUSSION

The results of this cross-sectional study demonstrate that almost one-third of the living patients known to have EB in Australia live outside the major cities. The number of patients with EB living in regional and remote areas is equal to just under what would be expected if the prevalence of EB were uniform throughout the Australian population.[1] Interestingly, of the patients with severe subtypes of EB, three lived in remote and very remote areas, which was half of all of the patients living in these locations. This was surprising as our initial thought was that patients with EB, particularly those with severe disease, would tend to gravitate towards large tertiary hospitals. It is worth noting, however, that although it is well established that patients in rural and remote areas tend to have poorer

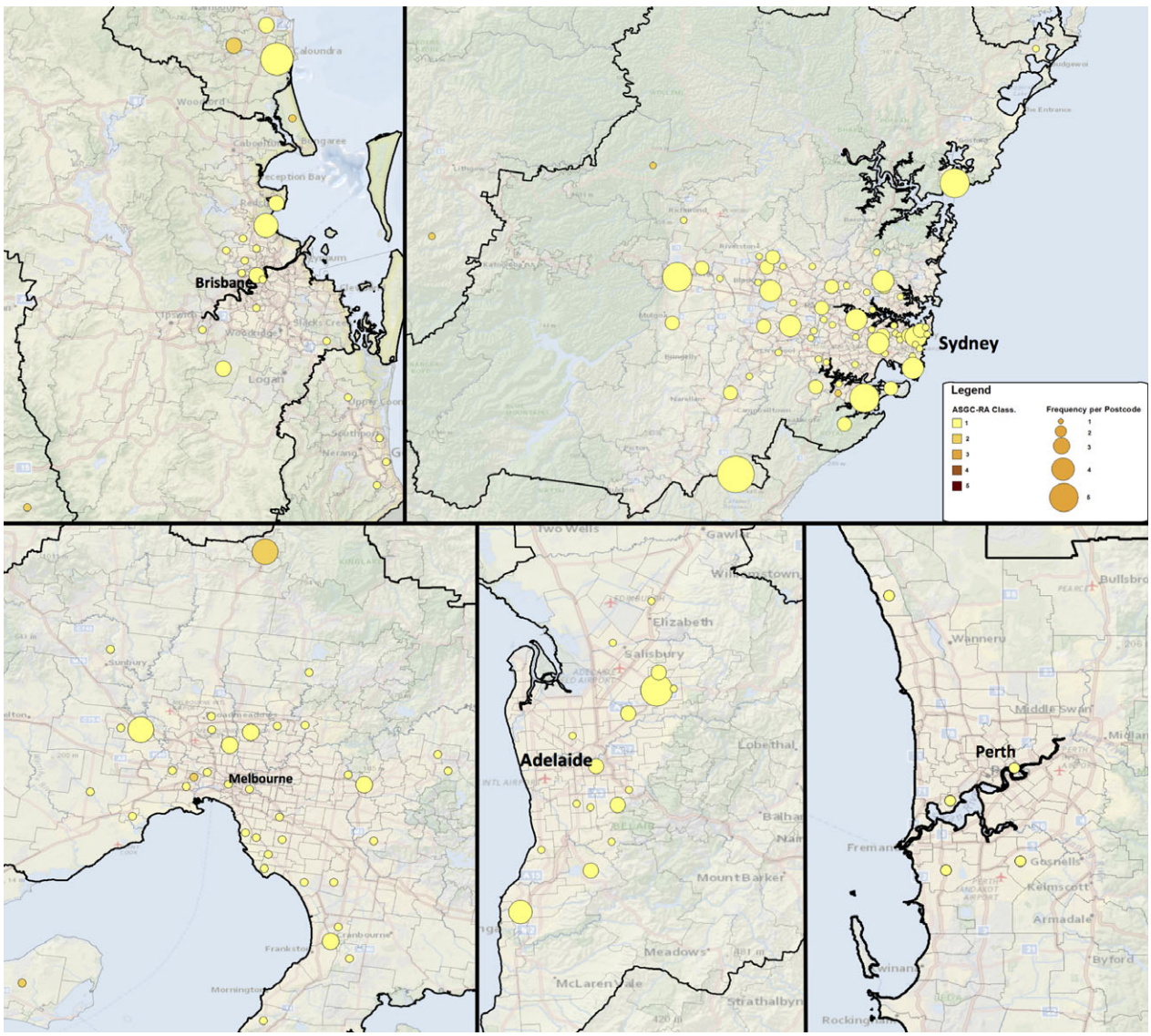

Figure 2 Map of the major cities of Australia showing the geographical distribution of patients with epidermolysis bullosa and the corresponding Australian standard geographical classification, remoteness areas for each patient. 1, major cities, 2, inner regional areas, 3, outer regional areas, 4, remote areas, 5, very remote areas

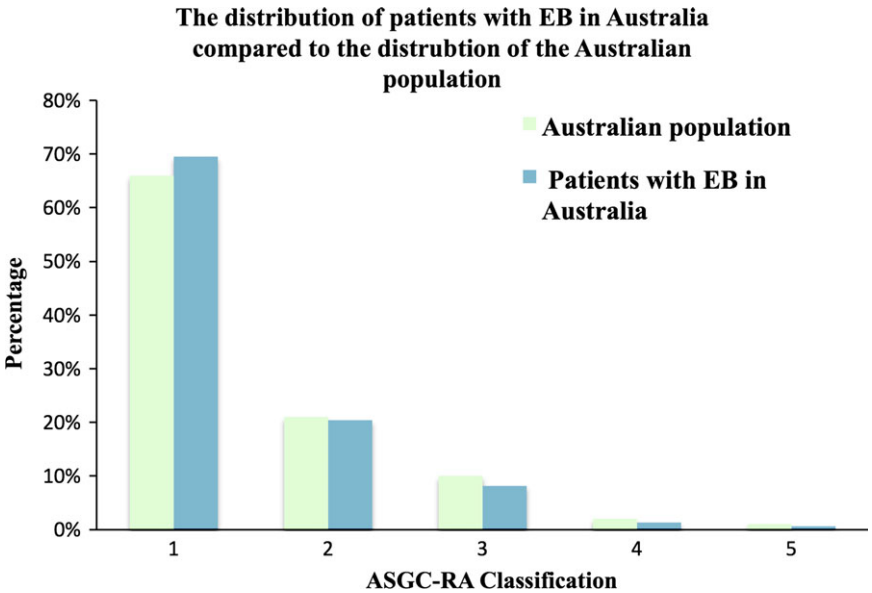

Figure 3 Comparison of the population distribution across each Australian standard geographical classification, remoteness areas category between patients with epidermolysis bullosa and the general population of Australia from the 2006 census. 1, major cities, 2, inner regional areas, 3, outer regional areas, 4, remote areas, 5, very remote areas

outcomes than those in metropolitan areas, there are no data to suggest this is specifically true of patients with EB.[2]

The Australian population is highly mobile, which places limitations on the data.[8] The postal code listed may have been the patients home address, but may not represent the postal code where the patient spent most of their time for reasons of employment or education. The categorisation of our patients was based solely on the postal code provided at the time of entry into the database or registry. These codes are not updated regularly and may have changed over time. They are also likely to change in the future and may be influenced by population mobility trends and migration patterns in Australia.[8]

One limitation of the ASGC-RA classification is that it averages the ASGC-RA category for the different points that exist within a particular postal code. This means that points with a high degree of remoteness may be nullified by other points with a lower measure of rurality or remoteness within the same postal code.[7] Despite this, the proportion of patients living in regional and remote areas is likely to be underestimated as these patients have reduced access to the hospitals and specialists participating in the Australian National Diagnostic Laboratory database and the Australasian EB Registry.

CONCLUSION

In Australia a significant proportion of patients with EB live outside major cities. Surprisingly, some patients with severe disease live in remote and very remote areas. Based on this geographical distribution, targeted strategies to improve access to EB-specific medical care, such as outreach clinics and travel reimbursements, may be needed for patients living in rural and remote areas. Services such as live video teleconferencing and teledermatology may also warrant further development to address this need. Other rare diseases may warrant similar investigation and data could be used to advocate for improved service provision to these patients in rural and remote communities.

ACKNOWLEDGEMENT

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