Inspiring action: Australian team at forefront of young onset dementia research

Australia’s INSPIRED Study is leading young onset dementia research internationally, with the aim of improving services and support for this previously under-researched group. Adrienne Withall explains

The foundations of the INSPIRED Study began in 2007 after I was fortunate enough to not only be offered a three-year postdoctoral fellowship at the Dementia Collaborative Research Centre: Assessment and Better Care (DCRC: ABC) at UNSW Australia, but also afforded the opportunity to choose my research focus.

I had observed that more people younger than 65 seemed to be presenting with cognitive complaints to our Academic Department of Old Age Psychiatry at Prince of Wales Hospital, Sydney. Professor Brian Draper agreed to partner with me to investigate this further. When we started to research young onset dementia (an onset of symptoms under the age of 65; at that time also known as presenile dementia or early onset dementia), it soon became evident that there was very little known about this group. Further, people under the age of 65 were not really factored into calculations about the need for dementia services as there was an assumption that that this population was so small as to be inconsequential. Often they fell into the gap between disability and aged care services.

The INSPIRED Study (Improving Service Provision for Early Onset Dementia) was thus born out of the desire to establish how many people had young onset dementia in Australia so that a robust, evidence-based argument could be made to establish adequate services for this group.

Prevalence in Australia

Dementia in young people is more common than most people think. Our prevalence study, funded by an Alzheimer’s Australia Research grant, indicated that approximately one in every 750 people aged 45-64 are diagnosed with young onset dementia. Additionally some people are very young at the onset of their dementia symptoms; in particular, five people in our study were aged less than 40 years at the time of the study. Overall, it is currently estimated that there are 25,000 Australians with young onset dementia, accounting for 8% of all dementia cases; certainly not an insignificant group.

More clinically diverse

Young onset dementia is much more clinically diverse than dementia in older people, so you are more likely to see a greater range of dementias and also rarer causes of dementia.

Some dementias tend to have an earlier age of onset, such as frontotemporal dementia, and are seen more often in this group. There are also more people with autosomal dominant, or genetically inherited, dementias although the rate is still only around 15-20% of young onset cases. This means that the majority of dementias in younger people are not inherited but are instead primarily due to environmental, lifestyle and/or other (as yet unknown) risk factors.

Within diagnostic groups, such as Alzheimer’s disease, there does not seem to be great differences between young and older people with the condition. In fact, when Alois Alzheimer first described the neurodegenerative illness that would later be named for him, he believed it to be a ‘presenile’ condition; his first patient had an onset of symptoms in her forties and the second in his fifties.

Importantly there are a number of young onset dementias with potentially preventable causes, such as dementia secondary to alcohol abuse and/or head injury.

Unexpected hurdles

We did not expect the significant level of stigma regarding the term ‘dementia’. This initially created a barrier to some groups participating in our study and caused us to rethink our terminology. We eventually used the term ‘young onset memory and related disorders’ and people seemed happy to endorse this.

We also learnt that people assisting with...
the care of a person with young onset dementia wish to be known as ‘supporters’ rather than ‘carers’. This term better reflects the range of people in our study, who were not always family members, and also shows that many people regard themselves as being in supportive partnerships as opposed to dependent relationships. I see it as a very positive term.

**Translating into practice and policy**

One of the strengths of the INSPIRED study is that we used a mixed methods approach. This means that both quantitative (eg severity scores on dementia scales) and qualitative data (eg people’s attitudes, care pathways) were collected, and were used in an iterative approach. Not only did we generate a lot of data about each participant, but the data we collected is very rich and meaningful. This can help to promote the translation of research findings into policy and practice.

It is wonderful to see the National Young Onset Dementia Key Worker Program working so well across Australia and helping so many people and I like to think that The INSPIRED Study helped to play a role in that.

We also recently presented data from our study at the 2015 Senate inquiry into the adequacy of residential care available for relatively young people with severe physical, mental or intellectual disabilities in Australia. Our quantitative prevalence data gave the panel a feel for the size of this group but the qualitative data helped us to convey the lived experiences of our participants. The panel members found this very powerful and it helped them to grasp the key issues and challenges for young people with dementia going into aged care facilities, either for respite or more permanently.

Our study has led to recognition of the diversity of support systems for people with young onset dementia and their unique needs. Some are living alone and can be quite isolated, some are cared for by older parents or by siblings, some have young children who also need support, and many have partners who are still working.

There is no ‘one size fits all’ model of service provision or support. We know that people need services that are flexible, tailored to their needs and interests, and allow them to be meaningfully engaged. Although services for young people with dementia are still relatively limited, we have had the ability to advocate for a consideration of this group within the development of dementia service frameworks.

INSPIRED data is also being used to help guide health professionals in their diagnosis and management of this group. One of our published papers has examined care pathways for people with young onset dementia, with a focus on the diagnostic process. We found that there was often a delay of more than two years from the person and / or their supporter first noticing symptoms before presenting for their first consultation with a doctor; this was more noticeable with our youngest participants.

Nearly half of our participants were initially given a non-dementia diagnosis; predominantly depression or mild cognitive impairment. The median time to diagnosis for the group was three years from the onset of symptoms.

Importantly, increased recognition is needed of the importance of referring a person for specialist cognitive assessment if there is behavioural change or treatment-resistant depression with an onset in the 40s or 50s. Clinicians also need to use the term ‘mild cognitive impairment’ with caution, as this is a research and not clinical diagnosis and creates confusion for their patients and supporters.

**International importance**

Internationally, there are relatively few groups focusing on the experience of dementia in young people. Australia, and the investigators of the INSPIRED Study, are at the forefront of young onset dementia research.

We reported the first Australian prevalence data for young onset dementia, and this work is only one of a handful of international studies examining the epidemiology of dementia in young people.

Comparison of our data with the other main comprehensive studies conducted in the UK and Japan has shown differences in the types of dementia that are more common in each country. This means that different geographical regions might have different risk factor profiles and may require individualised interventions tailored to their country.

The International Psychogeriatric Association (IPA) Young Onset Dementia Shared Interest Forum, of which I’m a member, recently met during the IPA Congress in San Francisco. We are currently planning an international Delphi consensus study to establish consistent terminology, principles of care, harmonise assessments, and establish research priorities. The findings of the INSPIRED Study will help to inform this project.

**What’s ahead?**

It is an exciting time for the INSPIRED Study. Our PhD student Monica Cations, who is funded by an Alzheimer’s Australia Dementia Research Foundation scholarship, has been focusing on determining modifiable lifestyle and environment risk factors for young onset dementia.

Monica recently presented her findings at The Alzheimer’s Association International Conference in Toronto and her pilot work has particularly indicated the importance of early and mid-life factors. In our planned larger study (INSPIRED-II), for which we are about to seek national funding, we are therefore taking a lifestyle approach to understanding dementia risk, including factors such as childhood trauma and stress. Importantly we are investigating modifiable factors that accelerate the young onset of dementia, thus creating opportunities to intervene to reduce a person’s risk profile.

There is also much to learn about the impacts of concussions on later risk for cognitive decline. We believe that there may be an overlap between the behavioural and cognitive signs observed in a relatively small proportion of sportspeople exposed to multiple head knocks over the course of their career and frontotemporal dementia, particularly in association with Parkinsonism. We will also consider the impact of susceptibility genes such as ApoE4, which can modify a person’s recovery from a head injury by promoting neuroinflammation, for example.

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**References**


