

Younger Onset Dementia: A Literature Review

Executive Summary

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Executive Summary

Evidence underpinning services and supports for people with younger onset dementia is an emerging field. Much of the information available to date has been descriptive or based on expert opinion, with limited empirical studies regarding the efficacy of particular approaches. However, this is changing in response to the increasing recognition of the different aetiologies, trajectories and implications of the diagnosis for people who are aged less than 65 years at onset.

This Literature Review is the first part of a two-stage project funded by the Australian Government to inform the development of services and supports for people with younger onset dementia and their families. Preliminary findings have been used to shape the national consultations that were conducted as part of the needs and feasibility analysis.

Research methods

The review employed a rigorous search strategy to identify literature related to services and needs of people with younger onset dementia. It included a broad conceptualisation of ‘younger onset dementia’, and considered literature from within the disability, chronic health, business and volunteer sectors as well as the traditional aged care and mental health paradigms where the majority of dementia literature currently resides. Included for consideration have been the social, economic and environmental factors that enable and support people with younger onset dementia. The rationale for this approach was the potential of these other sectors to have developed models of care and support that could inform service development for people with younger onset dementia.

Likewise, we have broadened the definition of the term ‘younger onset dementia’ in our search terms, given the reviews to date have predominantly focused on Alzheimer’s disease and, to some extent, fronto-temporal dementia. Our search included groups that have neuro-cognitive deficits associated with illnesses such as HIV/AIDS, Parkinson’s disease, alcohol or drug abuse, Huntington’s chorea, Down syndrome and Multiple Sclerosis. The rationale for their inclusion was the potential for these groups to have developed service models that could also be appropriate for the broader younger onset dementia cohort. We have also included contextual factors such as living arrangements, with a particular search for information on those living alone.

This review was conducted between September and December 2013, including over 300 journal articles, reports, presentations and websites out of the 425 yielded from the search strategy.

Overview of Younger Onset Dementia

The introductory overview of the full Literature Review describes the different types of younger onset dementia and has noted the similarities and differences in the range of symptoms for these subtypes.

The overlapping of some symptoms across younger onset dementia types has made differential diagnosis complex and difficult. Problems experienced by people with younger onset dementia in obtaining a diagnosis, including the lengthy time to diagnosis and frequent initial misdiagnosis, is a common theme in this literature. Although this issue has been reported for many years it still remains a major issue for service improvement. Some studies have reported a greater presence of behavioural and psychological symptoms of dementia (BPSD) for some sub-types such as fronto-temporal dementia (FTD) but the evidence is as yet equivocal; however, behavioural and psychological symptoms appear to be relatively common for most types which add to the needs and care requirements for people with younger onset dementia.

As the onset of younger onset dementia commonly occurs in people aged between 40 – 65 years it occurs at an earlier period in the life cycle stage. This earlier onset raises a number of issues for people with younger onset dementia including loss or diminishment in roles such as provider, parent and spouse and the significant adjustment to those changes that is required. Many people consequently experience financial difficulties arising from the loss of income and additional care costs.

As people with younger onset dementia are younger they are often physically healthy and active and may not experience the co-morbidities that are associated with later onset dementia. Another common theme in this literature is the need for access to individually tailored, person centred services and the need for existing services to provide programs/services that are more age-appropriate for people with younger onset dementia.

Some types of younger onset dementia are more preventable. These include alcohol related brain damage or alcohol related dementias (ARBD), which recent prevalence studies indicate is more common in Australia than elsewhere. Recent findings concerning potent combination antiretroviral treatments for HIV/AIDS appear to be reducing the incidence of this form of younger onset dementia, reminding us that new treatments have the potential to affect both the incidence and the prevalence of younger onset dementia, highlighting the importance of ongoing biomedical research.

Huntington's disease, Down's syndrome and some forms of Alzheimer's disease are more strongly associated with genetic risk factors. Obviously for groups so affected provision of genetic counselling and clear information is of prime importance.

The limited literature available indicates there is quite a long period from onset of symptoms to permanent residential care placement for people with younger onset dementia (e.g. 6-9 years) and that the level of informal care provided is high which places a significant burden on these families. Although some studies indicate a relatively high use of institutional services for this group (e.g. hospital admissions, nursing home respite etc.) authors also report that community service use is relatively low for this group. Many authors suggest the need for a central gateway to services for people with younger onset dementia or the adoption of case-management and key worker approaches.

Research gaps

There are significant research issues and gaps relating to prevalence, incidence, life expectancy and the proportions of people that are diagnosed with the various sub-types of younger onset dementia. More accurate and up to date Australian estimates will be critical for service planning purposes. There are few studies that comprehensively examine service utilisation and the costs of illness for the younger onset dementia group or that include consideration of social and informal care costs. Likewise, there are relatively few studies that examine the cost effectiveness of both pharmacological and non-pharmacological for dementia overall, let alone for people with younger onset dementia.

Living with younger onset dementia

The experience, issues and stated need of people with younger onset dementia, their carers and families are explored in Section 4. There were quite a few studies reviewed that interviewed people with younger onset dementia directly about their own experience of living with dementia. Many could be characterised as qualitative thematic analyses of interviews usually based on very small sample sizes. The challenges faced were of both a personal and systemic nature.

Personal challenges related to adjusting to dementia and coping with role changes, such as loss of independence, loss of employment, loss of empowerment, and needing to rebuild and restructure one's life. Loss of empowerment was associated with the feeling that involvement in decision making was being denied to people with younger onset dementia, often by well-meaning carers or service staff. There was a desire expressed by many to remain involved and to get on with their lives as best they could.

Systemic challenges were problems associated with obtaining a diagnosis (length of time taken, earlier misdiagnoses), feeling stigmatised because of the dementia 'label', a lack of referrals to support services, falling between the cracks of service systems, a lack of access to age appropriate services and programs, and financial problems.

While some authors recommend further research on the 'experience' of people with younger onset dementia, we believe that 'more of the same' research is actually not required. What is required are a few well-funded studies with better research designs, larger sample sizes, a triangulation of methods of outcome assessment, and consideration of the control of extraneous variables/ confounding factors. The previous studies have provided valuable insights but the level of evidence is 'weak'. However, what is clear from the research is that the involvement and participation of people with younger onset dementia in service design, development and evaluation should become part of routine practice.

Despite explicit searching, no studies were found that considered the needs of people with younger onset dementia who lived alone or those that have no familial carer. Although it has been estimated that approximately one third of people with dementia live alone there is little data available in the younger onset dementia research literature and yet this is a group that is likely to have high service needs. It might well be that premature placement in

residential care facilities may be an issue for this group and this needs to be further explored. Similarly the issue of parental carers for people with younger onset dementia does not appear to have been explored.

The experience of spouses revealed similar themes to that of the person with younger onset dementia, along with some additional challenges. The familiar theme was the long quest to obtain a diagnosis and adjusting to that diagnosis. This is particularly important as spouses often report deterioration in their relationship arising from changed behaviours in the lead-up to the formal diagnosis.

In addition, issues included managing behavioural and psychological symptoms, changing roles, grief associated with the 'loss' of spouse (as they were) and their future plans, juggling the caring role with other family and daily life responsibilities including employment, and difficulties in making new plans for the future. Other studies report that social isolation may be quite common amongst spousal carers. These issues also need to be addressed by support services.

Some authors note there are different phases of the couple's relationship which can be characterised as progressing from sustaining their 'couplehood', through maintaining their involvement and in the end stages to 'moving on' (e.g. moving from a 'we' focus to an 'I' focus) which may actually be a necessary developmental step. Although many of the studies report negative effects on the spouse such as carer burden, many couples have a positive focus on their predicament and try to maximise their quality of life and to maintain their relationship. Some carers have reported developing higher self-esteem and assertiveness as a function of undertaking their caring role. Factors that need further investigation concern the nature of the couple's relationship at symptom onset and through the illness progression, the effectiveness of the coping strategies that spouses use and the identification of precursors of poor coping strategies in spousal carers.

Some carer studies have included mixed groups of carers in their studies (spouses, children, other relatives, formal carers, ex carers) although most of these studies include a large number of spouses/partners. However, studies that use mixed carer groups or combine both the person with younger onset dementia and carer feedback run the risk of ignoring particular issues for these groups. As noted above, although some of the issues raised are in common, each group is likely to have its own, slightly different perspective on each issue. An example of this is the area of unmet needs where people with younger onset dementia rate psychological distress as a more common unmet need than do carers or professionals.

A number of these studies have used more quantitative approaches, such as including the use of standardised scales to assess carer burden, stress, carer and patient unmet needs, the presence of psychiatric symptoms amongst carers, and health related quality of life and well-being. Generally these studies indicate high levels of stress and burden for carers, poorer quality of life, and unmet needs including social isolation, depression and anxiety.

Some recent studies have used person with younger onset dementia - carer dyads to explore these themes and the value of dyad studies is that data relating to the person (e.g.

severity, presence of behavioural and psychological symptoms etc.) can be directly related to carer findings and vice versa, which provides for a somewhat higher level of evidence. Similarly, some studies are now using comparator groups where the younger onset dementia group is compared to a later onset dementia group. Unfortunately many of these initial studies have poor control of confounding factors such as the duration of the caring period, age and the diagnostic composition of the comparator groups. Many studies are cross-sectional so do not address the course of the dementia and while recent studies from the Netherlands are addressing this research gap, there is a clear need for further longitudinal research.

Relatively fewer studies have interviewed or surveyed children of people with younger onset dementia with regard to their experience and in most cases the sample sizes are again very small which limits the degree to which these findings can be generalised. The literature on the effect on children having a parent with dementia makes mention of perceived stigma and associated shame/ embarrassment, bewilderment, family conflict, high care burden, the physical challenge of caring, psychological issues and problems at school. Many of these children are undertaking a demanding caring role (sometimes becoming a 'parent' to a parent) while also facing the usual development challenges of growing up. Coping strategies, family cohesion and security of attachment are raised as issues but little research has been conducted directly on these topics. Some children reported there positive effects of their caring role, such as maturation and the experience they have gained, despite the challenges. However, it is clear these children have substantial needs for support and due to the care burden placed upon them may have a potential for psychological and social disadvantage that needs to be further explored.

A few studies have focussed on issues concerning rarer forms of younger onset dementia. Some comparative studies have suggested that carer burden may be higher for carers of people with fronto-temporal dementia but it is not directly related to the number of behavioural and psychological symptoms as these have been found to be similar across comparator groups. The issue might not be the total number of behavioural and psychological symptoms the person displays but the type of symptoms and their frequency. These factors might also interact with carer and other socio-demographic variables and multivariate techniques might be required to gain a further understanding of these elements.

Some forms of younger onset dementia also have quite a different presentation as occurs with Down syndrome and dementia where the carer has experienced the caring role often since the childhood of the client. Although one study noted the provision of informal care was much greater for people with Down syndrome and dementia, another study suggested these carers were more settled in their caring roles and less prone to experience issues such as 'burn-out' or health related issues than for other younger onset dementia groups. This may require further exploration as the literature is sparse.

Despite the numerous attempts by organisations such as the Alzheimer's Australia to provide comprehensive information across a range of forums, including the internet, the

need for clear information and advice is still mentioned as an unmet need for carers and patients.

Service design

Issues concerning service design and development are addressed in Section 5. It provides both a policy context and an evidence base regarding how service providers, regardless of the source of funding, can build their capacity to deliver services to people with younger onset dementia. The fundamental premise for service provision for younger onset dementia is that services must be individually tailored due to the significant diversity across the younger onset dementia population. This section also explores the current, changing policy context for younger onset dementia service delivery in Australia.

A set of overarching principles for service design and development for delivering services to people with younger onset dementia have been described that can be applied to service provision for people with younger onset dementia in a variety of contexts such as health care, disability or aged care and community based services. These have been grouped into three main categories of principles of service design:

Individualised model of service:

- Listening to people with younger onset dementia and their carers
- Individualised service planning / a person centred approach
- Whole of family approaches
- Ongoing needs assessment
- Recognition of the diversity among the many younger onset dementia diagnostic groups and the special needs of individuals.

Staff attributes:

- Appropriately skilled and suitable staff
- An holistic approach
- An enabling and consumer centred approach
- Case management skills
- Effective communication
- Flexibility.

Organisational attributes:

- Access to integrated specialist diagnostic and ongoing symptom management services
- Capacity for organisational change

- Regionally based, integrated and coordinated interagency partnerships and pathways
- Timely service provision
- Dementia friendly environments
- Cost effective and flexible fees policy
- Cultural Safety
- Ability to cater for needs of people in rural and remote communities
- Individualised service planning / person centred approach
- Respect and consideration for clients and carers
- Effective risk management strategies
- Appropriate exit policies relating to the suspension and withdrawal of services.

A range of programs were identified in Section 6 for people with dementia, their carers and family members. The majority of programs were aimed at the person with dementia and/or their carer and only a few programs were aimed at other family members. The majority of studies that evaluated programs generally provided only a low level of evidence. However, there are some types of programs that were identified that can be considered helpful or that may warrant further research.

Programs that have been shown to be helpful include:

- tailored physical activity programs;
- Support programs such as peer support and carer support groups, as well as those that include both the person with dementia and the carer. While studies usually involved support programs that were face to face, the facilitation of support groups through communication technology, such as email and videoconferencing, showed promise;
- Information and education services such as the Living with Memory Loss program provided through Alzheimer's Australia and,
- Cognitive stimulation programs.

Likewise, programs that provide active meaningful participation, including horticulture, volunteering or supported workplaces and creative expression programs also warrant further study with people who have younger onset dementia.

While the need individually tailored support programs such as case management and key workers is clear, their mode of operation and purported outcomes require further research.

In Section 7 the service needs of special needs groups are discussed. Australian Commonwealth legislation (Aged Care Act 1997, as amended 2013) recognises a range of people as having special needs with regard to access and equity which must be considered

as an integral part of service planning, development and delivery. These include people from:

- Aboriginal and Torres Strait Islander communities
- non-English speaking backgrounds
- rural and remote areas;

and people who are:

- financially or socially disadvantaged
- veterans
- homeless or at risk of becoming homeless
- care leavers (people who had been raised in care homes), or
- Lesbian, Gay, Bisexual, Transgender and Intersex (LGBTI).

When considering these groups it is imperative to acknowledge that there are people living with dementia that may be classified under multiple special needs groups and therefore may experience disadvantage on a number of fronts.

In summary, the literature in relation to dementia and special needs groups was sparse and generally not specific to people with younger onset dementia. The identified literature ranged from emerging practice and expert opinion to mainly qualitative research with small sample sizes. While the issues identified are similar to those of the general population, the key messages for these groups is to ensure the cultural, linguistic and geographic factors are adequately identified and addressed in the planning, funding and delivery of services. Barriers identified included: access to culturally appropriate diagnostic services; denial of dementia within some cultural groups; privacy and confidentiality concerns - including the use of personal information; transport - especially in rural and remote areas; and, having suitably qualified staff in some cultural and linguistic specific services, e.g., translation services.

Additionally, the literature identified particular needs for some of these groups. From a service provision perspective living in a rural or remote region is likely to be disadvantageous across the board because mainstream services may be scarce or non-existent, restricting both choice and access. Since there is very small number of people living with younger onset dementia it is highly unlikely that appropriate services for this group with special needs would be available, particularly in rural and remote areas.

Although the research literature about younger onset dementia relating to the defined special needs groups is scant the available literature, including grey literature, emphasises that the needs of these groups are complex, multifaceted and dynamic and become more so with the onset of dementia, reinforcing the call for person-centred, culturally appropriate, flexible service options.

Next steps

Throughout the report a large number of research gaps have been identified and a number of recommendations for future research have been identified. Likewise recommendations concerning service provision and service delivery have also been identified for further consideration by government. Despite the limitations regarding strength of evidence for many of the programs and services reviewed, there are some clear lessons emerging about optimal service system design. These are summarised in the following three key points:

Focus on the person with younger onset dementia

Fundamental is a person-centred focus with people with younger onset dementia involved in the development, design and the evaluation of services and supports they may need. This is important to ensure services are tailored to the individual needs, goals and aspirations, and relationships of the person with young onset dementia, and their surrounding support network.

Service attributes

At the individual service level, the focus needs to be on practices that are delivered with respect and consideration, by staff who are appropriately skilled, in an environment that is enabling and 'dementia friendly' in its design.

The delivery of services needs to be timely and responsive to the progression and complexities of young onset dementia, within the context of a risk management framework that appropriately balances the likelihood that younger people with dementia may want to take part in activities where there is a higher level of risk involved.

Integration of services

To improve the coherence of what can be an extremely fragmented service system, the focus needs to be on policies and models that facilitate service integration, such as multi-disciplinary teams, service linkages, care pathways and case management and coordination.

The complete report, including references, can be found at:
<http://ahsri.uow.edu.au/chsd/projects/yod/index.html>