

## **SHORT REPORT**

### **Young onset dementia and the need for specialist care: a national and international perspective**

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**Abstract**

Receiving a timely and accurate diagnosis and gaining access to age-appropriate support for younger people living with dementia (YPD) remains a challenge both in the UK and internationally because the focus of most dementia services is primarily upon the needs of older people. The political case to improve services for YPD depends upon the establishment of an understanding of the clinical symptoms, an unequivocal evidence base about need and an accurate evaluation of the size of the population affected. This short report assesses the evidence base from international studies regarding service design and delivery. The goal is to raise awareness, advance best practice and galvanise the international community to address the serious underfunding and under provision of care for this marginalised group. The current evidence suggests that there are universal problems, regardless of continent, with delays to diagnosis and poor understanding of optimum models for service provision and long-term care.

***Keywords:*** Young onset dementia, younger people living with dementia, needs, neurodegeneration, service provision, Young dementia network, care pathway

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## **Background**

Younger people living with dementia (YPD) have been at the forefront of the growing campaign to challenge society's attitude to dementia. Despite their increased visibility in the UK, receiving a timely and accurate diagnosis and gaining access to age-appropriate support remains challenging for younger people because dementia services remain primarily focused upon the needs of older people. A recent UK-based survey of services for YPD highlighted the lack of age-appropriate post-diagnostic support, the lack of meaningful social and recreational activities and lack of respite and long-term care facilities (Rodda and Carter, 2015). The continued lack of international policy focused upon the needs of YPD results in enormous, social and psychological distress for individuals and high economic burden for the state. A recent US study (Nagaendran et al., 2015) emphasised a doubling of the economic cost for YPD compared to older counterparts as a result of the loss of socially and economically productive individuals to society.

## **The international perspective**

Despite the 2014 recommendations of the Alzheimer's COoperative Valuation in Europe project (ALCOVE), which included 24 countries and produced a set of evidence-based recommendations for policymakers on dementia, including young onset dementia, many countries do not offer any specific form of services or support for this under-represented group (Brooker et al, 2014).

The International Psychogeriatric Association has established a shared interest forum on YOD (Young Onset Dementia) to encourage further global action to address the need for services, research agenda and policy change. France, Norway and the Netherlands currently have specific measures to address services for YPD within their national dementia strategies. In the UK, the National Dementia Strategy (DoH, 2009) identified the importance of ensuring existing services for dementia are not restricted to those over 65, that services must meet needs of younger people and that inappropriate placement with older more physically frail people should be avoided. However, despite such recommendations, this model of best practice is not currently reflected in the majority of services in the UK. Of particular note, only 11% of respondents to the recent national survey could identify access to local age-appropriate respite care and only 14% could identify access to local age appropriate long-term care (Rodda and Carter, 2015).

## **Problems with epidemiological data**

The political case to improve services for YPD depends upon the establishment of an understanding of the clinical symptoms, an unequivocal evidence base about needs and an accurate evaluation of the size of the population affected. To date, there is a lack of clear epidemiological data as many international studies exclude people under 65 and the two main international studies using a case finding and case validation process have provided equivalent prevalence rates but identified very different clinical profiles (Harvey, Skelton-Robinson, Rossor, 2003; Ikejima et al., 2009). For example, the Harvey and Ikejima studies identified a population prevalence of 54.0/100,000 and 51.7/100,000 respectively for the 30-65 year age group but by contrast, a catchment area study conducted in Sydney produced a prevalence rate

of 68.2/100,000 (95% Confidence Interval: 54.9–83.4, (Withall, Draper, Seeher, K & Brodaty, 2014). An Argentinian study using data from a ‘closed hospital population’ with data extrapolated to the general population using national census data demonstrated an incidence rate of 11/100,000 (Sanchez-Abraham., 2015). The only UK study examining the incidence for all cases of primary dementia for the age range 45-64 years estimated it to be 11.5 cases per 100,000 person-years (95% CI 8.6-15.0, (Mercy, Hodges, Dawson, Barker, & Brayne, 2008)). Furthermore, the Ikejima study commented on the main aetiological subtypes of YOD in Japan and identified vascular dementia as the most common, whereas the Withall study from Australia identified alcohol related brain damage as the commonest cause.

### **UK figures**

The latest UK figures estimated that there are now 42,325 people with Young Onset Dementia (YOD) (nominally referred to as onset before the age of 65 years) in the UK (Dementia UK Update, Alzheimer’s Society 2014). This is a significant increase on the previous prevalence figure of 15,034 which was derived from two studies (Harvey et al, 2003; Ratnavalli et al, 2002) in which the prevalence was calculated as the number of cases known to local service providers divided by the total local population as enumerated in the census. Given that not all those with YOD will be known to local services this was an underestimate of the true prevalence. The 2014 figures were derived from a Delphi consensus study conducted with 13 senior academics, who were provided with the best UK and international data on prevalence and asked to use this as a basis for their own expert estimates. The steep rise in numbers expected to have a diagnosis of YOD can be attributed largely to higher numbers in the 60-64 year age group. Assuming constant prevalence, the number of people with YOD in the UK is projected to increase to 50,401 by 2025 and 50,979 by 2051, as the numbers of people in the relevant age groups rises, an increase of 20% over the next 38 years.

### **Needs of YPD**

Data from the Netherlands based NEEDYD study, a prospective cohort study examining the needs and characteristics of YOD has advanced our understanding of key issues which could help underpin service provision. This comprehensive study has identified that YPD are generally cared for at home for longer than those with late onset disease resulting in high carer burden (Bakker et al., 2013). YPD experience high levels of unmet needs in important areas of life, including daytime activities, social company, intimate relationships, eyesight and hearing/communication, memory, mobility, psychological distress, and needs for information. A statistically significant relationship exists between unmet needs and the higher rate of neuropsychiatric symptoms (Bakker et al., 2014). Despite these difficulties, carers of those with YOD are more likely to use informal care systems than statutory services, and have limited opportunities for respite (Bakker et al., 2013), mainly because such services are not age-appropriate, are costly or have inconvenient opening hours. Furthermore, the Dutch Beyond study for the first time reported data from YPD who reside in specialised age appropriate residential units. Among other characteristics, the sample of 230 individuals were reported to have high levels of behavioural problems and psychotropic drug use (Mulders et al., 2016; Mulders, Zuidema, Verhey, & Koopmans, 2014).

Further, due to the young age of onset, the stage of family life tends to differ for those with YOD compared with later onset dementia such that it has costly social, emotional, psychological and financial consequences for families. There is the impact on employment and hence family finances at a time when many are still paying off mortgage loans; the additional loss of the carer to the employment market as the partner reduces work hours to provide care; the changes dementia provokes in intimate and family relationships, including with children who may not yet be independent; and the realignment it causes in family roles, and the conflict this may engender (Allen and Oyebode, 2009; Rosness, Haugen, & Engedal, 2008; van Vliet, de Vugt, Bakker, Koopmans, & Verhey, 2010; Millenaar et al., 2014).

### **Heterogeneity in presentation and lack of operational criteria**

The differential diagnosis of Young Onset Dementia encompasses complex presentations of the common primary neurodegenerative diseases as well as autoimmune, inflammatory, late onset metabolic and hereditary/familial causes (Rossor, Fox, Mummery, Schott, & Warren (2010). Although age is clearly an unhelpful discriminator of dementia subtypes, it remains an important influence on both presentation and post-diagnostic needs, yet there is currently no international consensus on the definition of, or nomenclature about, YOD (Koopmans and Rosness, 2014). Any new definition needs not only to highlight differences that distinguish it from late onset disease, but also encompass the inherent heterogeneity with age within a younger population (eg there is likely to be more relevant difference between a person of 44 and 64, than between 64 and 67, (Kelley, Boeve, Josephs (2008)). Clarity regarding definition and nomenclature is also needed to improve the comparability of research in this field.

Any new criteria, must also overcome the traditional view, derived from the numerical dominance of late onset AD, that all dementia is associated with episodic memory loss and functional decline. Atypical presentations of the common dementias in younger people often result in clinical under-investigation, misdiagnosis and delays in obtaining a definitive diagnosis. It has been found, for example, that 1 in 3 people with young onset AD present to services with problems associated with posterior cortical atrophy, affecting object recognition and other visual abilities, rather than with the memory problems that are more typical of late onset AD (Crutch et al., 2012). Furthermore, due to the greater proportion of rarer diagnoses, early signs of change may not be recognized as connected with the onset of dementia. People with behavioural variant fronto-temporal dementia (bvFTD), for example, may start to show lack of empathy or concern for others, and social disinhibition, such as being over-familiar with strangers or acting on aggressive or appetitive urges, by swearing or over-eating sweet foods (Graham & Hodges, 2005). Changes in managing complex tasks may be shown up in the person becoming apathetic, perseverative or failing to plan ahead. Similarly, those with primary progressive aphasia (Gorno-Tempini, Hillis, Weintraub, & Kertesz, 2011) are likely to experience various problems with language, for example retaining meaning of words, producing or finding words. None of these difficulties would be well captured through most of the common cognitive screening tests for dementia, that focus predominantly on orientation and memory.

### **Delays to diagnosis**

Evidence regarding delays to diagnosis has identified that although 60% of young onset patients sought help within 12 months of symptom onset (Williams, 2001) it took an average of 3.3 years in a young onset AD group and 4.9 years in a young onset FTD group to receive a formal diagnosis (Rosness et al., 2008). More recent studies in 2013, indicated that the average time to diagnosis was 4.4 years in younger people for all-cause dementia compared to 2.2 years for late onset disease of comparable severity. Younger age and a diagnosis of fronto-temporal dementia were related to increases in time to diagnosis (Van Vliet et al., 2013). The INSPIRED study from Australia recently reported a time to final diagnosis of the type of dementia from first presentation as 4.7 years. Participants with younger age of onset had significantly longer time to first consultation and to family awareness of the dementia diagnosis. The time to dementia diagnosis was also significantly longer when the dementia was other than AD or FTD (Draper et al., 2016). Given the significance of changes in empathy and disinhibition often associated with FTD, delay in diagnosis can mean that close relationships break down prior to diagnosis or that people take considerable risks (Oyebode, Bradley and Allen, 2013).

Together these findings highlight the importance of a timely diagnosis for YPD and their families in order to access appropriate care, signposting to age appropriate services and specialist multidisciplinary expertise. Advancing this case, the new UK Memory Service National Accreditation Programme (MSNAP 5<sup>th</sup> edition) which sets standards of care for memory clinics in the UK, for the first time has included the requirement for a specific local care pathway for younger people with cognitive impairment and a named lead for YOD within the team.

### **International initiatives to improve care and services**

In the Netherlands, a national task force on YOD presented a ‘national YOD care program’ in 2004, based on best practices. The objectives of this national care program were to improve quality of care in YOD and to strive for social and financial economic acknowledgement. The special care needs of this group were recognised by the Dutch government in 2006 with the introduction of an extra financial compensation for YOD services and adaptation of recommendations for building regulations for more spacious care facilities. These policy developments stimulated care organisations to start or improve dedicated services for YOD, for example by offering specific daytime activities, creating special care units in long-term care facilities (LTCF) and improving team expertise by staff training. Several LTCF started to implement the ‘national YOD care program’ in their organisation. To date about 30 care-organisations in the Netherlands offer specialized care for people with YOD. Most care-homes have YOD specialised unit(s) where people with YOD live together, or have day-care centres. Most of them offer outreach care and advice or support groups for spouses and children. The service provision on the units and day-care centres differs from that provided on dementia special care units for people with LOD. There is more emphasis on hobbies suitable for physically active adults such as creating artwork, woodcarving, fitness and walking.

To ensure quality of care a quality mark has been established with quality indicators derived from the ‘national YOD care program’. In 2015 the Knowledge Centre for YPD published a specific ‘care-standard’ for younger people which was added to the national dementia standards.

A key European initiative, the RHAPSODY study, currently in progress, represents a co-ordinated investment across 6 different health and social care environments (France, Germany, the Netherlands, Portugal, Sweden and the United Kingdom) to assess the individual needs of YPD and their carers and how these are met. The information gathered will be used to develop an educational, web-based, interactive e-learning programme to support people with young onset dementia and their carers. The study will also provide information as to which services are best suited to meet the needs of this underserved group (Kurz et al., 2016).

### **UK initiatives to improve care provision**

In the UK, in response to the voices of YPD and their families, the Young Dementia Network steering group, hosted by the specialist charity Young Dementia UK has developed an optimum care pathway together with gold standards for best practice. The goal is to provide guidance to Commissioners of Care in local Clinical Commissioning Groups about best practice and to provide YPD with a blueprint for services that they should expect locally (Table 1 near here).

The international perspective continues to suggest that although there are pockets of excellent practice, in most countries support programs specifically designed for younger patients and their caregivers are largely lacking. Where exceptional services are established, this is often due to the commitment of local clinicians, and with an absence of recognised need by commissioners, well regarded YOD services can readily be eroded to achieve savings targets or due to key figures moving or retiring. The provision of age-specific units such as in the Netherlands may not be feasible for countries with larger land mass and more geographically dispersed populations so improved knowledge about optimum service delivery is essential. Finally, the need to define optimal palliative care in dementia is increasingly being recognised, but virtually no research has been conducted on the palliative care needs of YPD. It seems clear from personal experience that preferences regarding advance care planning, including the process of shared decision-making, treatment preferences of patients and family members, symptom prevalence and management in the final weeks of life, and palliative sedation may be unique and differ from those with late onset disease (Koopmans, van der Steen, & Bakker, 2015). Such issues require further exploration.

The goal for this short report is to raise awareness, advance best practice and galvanise the international community to address the serious underfunding and under provision of care for this marginalised group. The current evidence suggests that there are universal problems, regardless of continent, with delays to diagnosis and poor understanding of optimum models for service provision and long-term care.

Table 1.

A young onset dementia journey

|                                                                                                         | <b>Person with young onset dementia and their family carers</b>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                         | <b>Services to support these needs</b>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
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| <p><b>Positive contact with my GP which reduces the time taken to receive the correct diagnosis</b></p> | <p>My GP listens to me and doesn't discount dementia because of my age.</p> <p>I can talk to my GP about how young onset dementia could affect me, or my family.</p> <p>My GP knows about the referral process for young onset dementia and it is explained to me. I'm able to record these conversations so I can refer to them later.</p> <p>I'm told about the National Young Onset Dementia Network, and local young onset support services who can support me.</p>                                                                                                                                                                                 | <p><b>GP aware of young onset dementia and appropriate local referral routes</b></p> <ul style="list-style-type: none"> <li>• GP knowledgeable about young onset dementia and the variety of symptoms which can present, which may differ from those with later onset dementia</li> <li>• GP has a low threshold for referral if there are 'red flags'</li> <li>• GP listens to the person and family members to make thorough assessment of symptoms including baseline investigations, physical examination, assessment of activities of daily living and a brief cognitive assessment</li> <li>• GP excludes a potentially treatable illness or reversible cause of the 'dementia'</li> </ul> <p>Referral made to the <b>locally agreed young onset referral care pathway</b></p> <p><b>Access to pre-assessment counselling</b></p> <ul style="list-style-type: none"> <li>• Pre-diagnostic review/support and counselling</li> </ul> <p><b>Information is provided, including</b></p> <ul style="list-style-type: none"> <li>• Reason for referral</li> <li>• Expectation / possible outcome / consent</li> <li>• Implications for other areas of life</li> </ul>                                                                                                                                                                                                                                                                                                                                                                |
| <p><b>A clear, collaborative and specialist diagnostic process</b></p>                                  | <p>I know who my young onset dementia lead is, and am kept updated about the diagnostic process.</p> <p>I'm offered the same support as older people who are going through the process of being diagnosed with dementia. For example, I might need a family member or close friend to attend appointments with me.</p> <p>If I'm supporting someone through diagnosis, I can speak to someone who understands my concerns and can answer questions I may have about what a dementia diagnosis at this age might mean for my family.</p> <p>I can participate in counselling tailored to my age and situation, in preparation for the journey ahead.</p> | <p><b>Making the diagnosis in specialist services</b></p> <p>Clinicians based in memory clinic or specialist services/departments where diagnosis takes place are <b>knowledgeable and skilled regarding young onset dementia and the impact the diagnosis can have on the person diagnosed and their family.</b></p> <p>There are <b>designated young onset dementia leads</b> in each diagnosing service</p> <p><b>Comprehensive assessment is performed by a commissioned multi-disciplinary team</b> who have opportunities for multidisciplinary case discussion.</p> <p>Pre-assessment counseling is routinely provided to establish informed consent, find out what someone wants to know about their diagnosis, who else they would like to be present when it is given, and if there is someone else they would like to be informed.</p> <p><b>Key elements of diagnostic assessment</b></p> <ul style="list-style-type: none"> <li>• Thorough history taking</li> <li>• Speaking to someone who knows the person well</li> <li>• Neurological examination</li> <li>• Blood screen for rare cause</li> <li>• Neuroimaging</li> <li>• Advanced cognitive assessment / neuropsychology</li> <li>• Consider EEG, CSF analysis, genetic testing</li> </ul> <p><b>Feeding back diagnosis - Diagnosis is given in a confidential setting and is delivered sensitively</b></p> <p>There should be a recognition that the person diagnosed with young onset dementia and their family may still be in work, have dependent young</p> |

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|                                                                                 |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                | <p>children or other care responsibilities, so may have different questions and responses to the diagnosis. Enough time should be allocated in order to answer these questions.</p> <p>Depending on the persons' wishes, they will receive full feedback of the results of their assessment and an explanation about how the diagnosis was reached as a result and should be provided with prompt follow-up</p> <p>Where a diagnosis is provided in a national or regional centre, <b>there must be a clear 'exit plan' which links the person back into their local services.</b></p> <p><b>Introduction to Key Worker</b></p> <ul style="list-style-type: none"> <li>• Key Worker works in collaboration with the diagnostic team and introductions can be made at the time of diagnosis or when appropriate for individual</li> <li>• Clear information is provided about next steps and how they can be contacted</li> <li>• Working together with clinicians to ensure a joined up service</li> </ul>                                                                                                                                                                                                                                                                                                     |
| <p><b>Specialist support and information which meets our changing needs</b></p> | <p>I'm contacted by a Key Worker who, if I choose, meets with me to discuss what dementia at my age will mean for me and my family. I have the opportunity to ask about the impact on my job, mortgage and other financial commitments, childcare and any other concerns I might have.</p> <p>I have the opportunity to learn more about my type of dementia, which because of my age, is more likely to be a rarer form.</p> <p>If I'm supporting someone with dementia, the Key Worker also supports me.</p>                 | <p><b>A regular review with the person, their family, the clinical team and others involved in their care is established.</b></p> <p>The individual and family should have access to:</p> <ul style="list-style-type: none"> <li>• Information about the diagnosis</li> <li>• Evidence based treatment and psychological support e.g. pharmacological eg Cholinesterase inhibitors/and or memantine, cognitive stimulation therapy, counselling, relationship or family-centered work, Information and advice about social, recreational and meaningful activities and Peer support</li> <li>• Admiral nurses (specialist dementia nurses) who can work intensively with families who are facing a difficulty managing the effects of the diagnosis.</li> </ul> <p><b>Key Worker engages with individual and their family to understand their needs</b></p> <ul style="list-style-type: none"> <li>• Discuss life, achievements, abilities and aspirations pre and post diagnosis</li> <li>• Discuss preferred / usual ways of coping and their potential support needs</li> <li>• Gain understanding of employment status, financial situation, care responsibilities and family circumstances</li> <li>• Meet family carers and support circle to understand their perspective and circumstances.</li> </ul> |
| <p><b>Living with young onset dementia</b></p>                                  | <p>My Key Worker helps me prepare for what lies ahead, providing information about practical issues like financial and care planning, as well as helping me to talk about my condition with my children, family and employers.</p> <p>I know I can contact my Key Worker and they can point me in the direction of other helpful services.</p> <p>I want to continue my social life, relationships and activities and hobbies. My Key Worker helps me to adapt so I can keep doing the activities that I am interested in.</p> | <p><b>Regular contact and joint reviews with Key Worker and Clinical team with access to specialist nurses, Occupational Therapy, Psychology and Social Work support as necessary</b></p> <p><b>Review response to medication</b></p> <p><b>Support Carers and assess needs of YPD and family</b></p> <p><b>Signpost to meaningful activities</b></p> <p><b>Key Worker tailors a plan with the individual and their family which is appropriate for their stage in life.</b></p> <ul style="list-style-type: none"> <li>• Outline goals and aspirations and route to achieving these</li> <li>• Facilitate access to community services, both dementia specific and otherwise</li> <li>• Provide or facilitate access to information about young onset dementia and rare forms of dementia and how to communicate</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                   |

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|                                       | <p>I'm introduced to new activities and social contacts/groups that are more specific to dementia when the time is right for me.</p>                                                                                                                                                                                                                        | <p>the diagnosis to children, family, employers and friends.</p> <ul style="list-style-type: none"> <li>• Provide or facilitate access to specialist advice and support about young onset specific needs e.g. employment, mortgage and financial obligations and future financial planning.</li> <li>• Provide advice and emotional support to family carers and others closely involved in care.</li> <li>• Link to health and social services to ensure plans are aligned.</li> <li>• Advanced care planning</li> </ul>                                                                                                                                                                                                                                                          |
| <p><b>Towards the end of life</b></p> | <p>I'm supported to think about and prepare for the end of my life, including talking about what I do and don't want.</p> <p>The support I get is tailored to me, depending if I live alone or with my family.</p> <p>The family and friends of the person with dementia are given information and support to help them to prepare for the end of life.</p> | <p><b>Regular reviews are adapted as the person reaches the end of their life and support is tailored to provide:</b></p> <ul style="list-style-type: none"> <li>• Key worker provides support for advanced care planning</li> <li>• Facilitate access to short term respite opportunities (including day and residential)</li> <li>• Acute care is provided in dementia friendly wards which comply with John's Campaign so a carer can stay with the person during their stay</li> <li>• Age appropriate long term residential care within a reasonable distance from home</li> <li>• Support to stay in the home if this is what the person wants</li> <li>• Age appropriate palliative and end of life care</li> <li>• Bereavement support for carers and families.</li> </ul> |

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