Service provision for young-onset dementia in the UK

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In this review article, Drs Rayment and Kuruvilla look at the problems faced by younger people with dementia, the current state of care delivery in the country for young-onset dementia (YOD), guidelines for service provision and they suggest what could improve the availability of specialised YOD services in the UK.

In 2003 it was estimated that there were over 18 000 people in the UK with YOD, or 67 cases per 100 000 people aged 30-64 years. A more recent study by the Alzheimer’s Society suggests this was an underestimate and that the national prevalence may be over 40 000. Comparatively, the estimated national prevalence for all-age dementia is around 850 000 in 2015 increasing to one million by 2025.

The projected public health burden of dementia has led to increased awareness and government-backed drivers to increase diagnostic rates. Currently only around 44% of people in the UK with dementia get a diagnosis while they are alive. This unmet need is often called the ‘treatment gap’. This gap may be significantly wider for the 40 000 younger people with dementia.

Younger people with dementia have complex health and social care needs. They are more likely to be working, physically robust and active, looking after children or elderly relatives, and paying off mortgages or other loans. Their partners often become their carers, further depleting household income.

There are significant implications when someone is diagnosed with dementia as a younger person, which can often be very far reaching when compared with diagnosis in later life. The impact on patients and their families, and the needs of patients, can be very different from late-life dementia.

Dementia can present differently in younger people. There is often a higher level of mood and behavioural problems. It can be difficult to distinguish dementia from depression in the early stages of YOD, particularly in frontotemporal dementia (FTD), which may present with apathy, irritability, dietary changes and deteriorating self-care.

The prevalence rates of the dementia subtypes differ in younger people. While Alzheimer’s disease (AD) is still the most common subtype it accounts for only around a third of YOD, compared with around two-thirds in all-age dementia.

Conversely, FTD, which accounts for only around 2% of all-age dementia, makes up around 12% of YOD. Other YOD subtypes include vascular dementia, alcohol-related cognitive impairment and various neurological and systemic diseases more frequently encountered as causes of dementia in the under 65 year olds. These include Parkinson’s disease, Huntington’s disease, multiple sclerosis, corticobasal degeneration (CBD), progressive supranuclear palsy (PSP), and more rarely human immunodeficiency virus (HIV), and prion diseases like Creutzfeldt-Jakob disease (CJD).

YOD has a higher genetic burden compared with late-life dementia, examples being the dominantly-inherited variants of AD, cerebral autosomal dominant arteriopathy with sub-cortical infarcts and leucoencephalopathy (CADASIL), which is a genetic form of vascular dementia, and FTD. Hence genetic counselling of relatives of people with YOD arises more frequently clinically than in older people.

People with learning disabilities (LD) are more likely to develop YOD. Down’s syndrome is associated with young-onset AD due to over-expression of the amyloid precursor protein gene in trisomy of chromosome 21 leading to excess amyloid deposition. Again, people with LD who develop YOD have significantly different needs.

This wide range of diseases causing the YOD syndrome can make differential diagnosis particularly difficult, so it should involve close collaboration between neurologists, psychiatrists, radiologists, other physicians, psychologists, occupational therapists (OT), speech and language therapists (SALT) and other members of a multidisciplinary team. Sampson et al. and Rosser et al. detail the assessment and diagnosis of YOD.

Getting a diagnosis is often the key required to access support services. Once diagnosed, a person with YOD and their carers should receive a comprehensive care plan involving close collaboration between health services, social services, housing, reviewing...
work and pensions and the voluntary sector. Medical treatments may involve disease-specific licensed medication such as acetylcholinesterase inhibitors and memantine for AD, thiamine supplementation for Wernicke-Korsakoff syndrome and antiretroviral combination therapy for HIV related cognitive impairment. It may also include off-licence but evidence-based medication like serotonergic antidepressants for the apathy commonly seen in FTD. Psychosocial interventions may include psychological management of behavioural symptoms, meaningful occupational activity, appropriate financial assistance, carer respite, counselling for young children and safeguarding vulnerable adults.

Current state of care delivery
Current service provision for people with YOD in the UK is highly variable. In a few regions there are dedicated multidisciplinary specialist services. However, in the majority of regions there is no dedicated service with a single point of access and no clear referral pathways. Consequently, people with suspected YOD are often unsure of how to access help and general practitioners are unsure who to refer such patients to, contributing to the wide treatment gap.

Some people with YOD are seen in memory services, the majority of which are old age psychiatry-based in the UK. These services may have practitioners skilled in the management of dementia in the elderly and who are competent at ruling out psychiatric disorders and there may be access to neuropsychology, OT and SALT thereby enabling good coordination of long-term care. However, the team may lack the expertise and resources to investigate for the broad range of neurological and systemic diseases that can cause the YOD syndrome. These investigations may include: structural neuroimaging with CT and MRI; functional neuroimaging with 99mTechnetium-hexamethylpropanolamine oxime single photon emission computed tomography (HMPAO-SPECT), fluorodeoxyglucose positron emission tomography (FDG-PET) and amyloid PET; cerebrospinal fluid (CSF) assay; electroencephalography (EEG); antibody titres; genetic assay, and other expensive tests.

An audit of an old age psychiatry-based memory service by O’Kelly et al. showed that the yield rate of a diagnosis of dementia in younger referrals is very low and they argued that it may not be cost effective to investigate these increasing referrals.

Some people with YOD are seen by neurologists who are proficient in neurological examinations and physical investigations but may lack expertise and resources for assessing mood and behaviour and in providing long term follow up after diagnosis. Other services where YOD patients are seen include general adult psychiatry, liaison psychiatry, neuropsychiatry, substance misuse, HIV, and LD services. The latter may be best placed to diagnose and manage YOD in people with LD as they are usually well known to the service.

Existing services based on informal arrangements without dedicated commissioning are not always able to build on sustained clinical experience and may not develop a good knowledge of support within the wider community. YOD patients and their carers report poor coordination between the different service providers, and highlight the problems of day care, respite services and residential care being designed around the needs of older people.

Some specialised YOD services may be at risk of being absorbed into old age psychiatry or even ‘age-less’ services in line with the current service reorganisation in the UK. Where specialised YOD services are available, they are highly valued by patients and carers.

Guidelines for service provision and sources of support
The Alzheimer’s Society began campaigning to raise awareness of YOD in the 1990s. It published its ‘Charter for younger people with dementia and their carers’ which stated that all people with YOD, their families and carers should have access to specialist services from diagnosis to long term care. The Alzheimer’s Society and the Royal College of Psychiatrists published a joint policy document on YOD in 1999, recommending that local commissioning bodies appoint one named individual responsible for planning YOD services and one consultant acting as a focus for referrals. These two individuals would form the hub of the service, liaising with other people already involved in the provision of care for YOD, and developing services appropriate to the region.

The National Service Framework for Older People (2001) directed the National Health Service and councils to ‘review current arrangements, in primary care and elsewhere, for the management of dementia in younger people, and agree and implement a local protocol across primary care and specialist services, including social care’, but it did not make any specific recommendations.

In 2006 NICE published its dementia clinical guideline, which was more specific in its
recommendations that: ‘younger people with dementia have special requirements, and specialist multidisciplinary services should be developed, allied to existing dementia services, to meet their needs for assessment, diagnosis and care’. In 2013 NICE published guidance on commissioning dementia care, which reiterated the above, also advising a single point of access and a single care coordination system.

The West Midlands regional forum for YOD has produced two strategic documents: commissioning guidelines for YOD dementia services and an integrated care pathway for YOD.

Alzheimer’s Society supports people with YOD at a regional level, offering practical information on dementia and services, and connecting patients and carers with others for mutual support.

It has an online discussion forum – ‘Talking Point’. It also maintains a national database of services which, in 2004, showed a significant increase in service provision, particularly community support, day centres, and carer groups. However, much more is needed, with respite and residential care remaining in short supply.

YoungDementia UK, formed in 1998, also keeps a record of services in different regions, runs forums and networks, and provides community services. The organisation commissions groups and research, and its sister charity, YoungDementia UK Homes, helps establish specialist residential support.

National support groups for rarer causes of YOD have emerged, including the Pick’s disease support group, the CJD support network and the PSP association. National conferences such as the recent ‘A life worth living: YOD services and support’ are useful for clinicians and commissioners involved in planning services.

Despite national guidelines, there is wide regional variation in service provision for YOD. Involving patients and carers at all stages of service development, along with persistent, committed pressure from patients and carers and other organisations, will help in the on-going development of dedicated YOD services and prioritisation of funding for YOD services, especially in the current economic climate.

Declaration of interests
No conflicts of interest were declared.

References