

Dying well with an intellectual disability and dementia?

Kathryn Service, Karen Watchman, Mary Hogan, Matthew P. Janicki, Anna Beránková, Nicole Cadovius.

Kathryn Service is a nurse practitioner, formerly with the Massachusetts Department of Developmental Services, Mary Hogan is a family advocate, Matthew Janicki is associate research professor at the University of Illinois, Chicago, and Nicole Cadovius is a disability services consultant, all in the USA. Karen Watchman is a senior lecturer at the University of Stirling and Anna Beránková is a research assistant at Charles University in Prague.

Accepted for publication in *UK Journal of Dementia Care* published by Hawker Publications

As people with intellectual disability increasingly survive into old age, more are susceptible to age-related health conditions including diseases that result in dementia. Previously, most adults with an intellectual disability, particularly those with Down's syndrome, would not have lived to an age where death associated with dementia was considered an issue, whereas now adults with Down's syndrome may live beyond 60 years of age. Consequently, we now know of the increased prevalence of dementia among people with Down's syndrome, with estimates suggesting that approximately 60-75% of those older than 60 will be affected by dementia (Prasher *et al* 2017).

Many palliative care and end-of-life needs of adults with an intellectual disability and dementia are similar to those of the general population without an intellectual disability. Such similarities include the need for pain detection and management, identifying dementia as a terminal condition, more effective end-of-life care planning, and inclusion in the decision-making process (Marie Curie Cancer Care 2015). But the onset of dementia as a complicating factor does create some significant challenges for practitioners which may not be present when working with people who do not have an intellectual disability.

At a recent international summit on intellectual disability and dementia, we identified three areas where the added complexity of advanced dementia warrants particular attention around end-of-life services in people with an intellectual disability. These areas were ascertainment of advanced stage of dementia, place of care, and active support. All three issues are discussed here so as to

show the particular practice challenges that arise when someone with dementia also has an intellectual disability.

Two of these priority issues are pertinent to the dementia care field, for whom people ageing with an intellectual disability are a relatively new client group, while the third is an issue for intellectual disability services whose typical way of working and associated good practice initiatives are focused on self-determination and fostering autonomy across the lifespan. There is potential here for shared learning among palliative, older persons' care, and intellectual disability services with the aim of improved outcomes and mitigating undue pain or suffering. If differences in end-of-life care needs and provision are not recognised, this can negatively impact the wish for a "good death".

Expertise in the field of intellectual disability and advanced dementia is rare and our summit offered a series of recommendations including ongoing exchange of experiences and skills across professions, development of tools and scales that facilitate understanding of the progression of dementia, and more equitable access to palliative care and hospice services with increased and timely referral. We also recommended that intellectual disability services increased understanding of the fundamental dementia-related needs which complicate end-of-life care.

Intellectual disability is synonymous with learning disability as used in the UK, but we talk about intellectual disability because that is the internationally accepted standard. Adults with Down's syndrome generally make up 10-15% of the adult intellectual disability population and have the greatest risk for Alzheimer's disease.

Ascertainment

The first area of difference we identified is ascertainment of approaching end of life. Where someone has an intellectual disability, it is particularly hard to determine whether they have progressed to an advanced stage of dementia along with the possibility of death in the immediate future (McCallion *et al* 2017).

Deciding what stage a dementia has reached in the general population is typically based on measuring the impairment of self-directed daily activities (Cordell *et al* 2013, Holmerova *et al* 2016). But this is not always the case for people with intellectual disability as many may have a

pre-existing lack of independence in basic activities of daily living. This means that any decisions to change care due to advanced dementia must be informed by a more robust assessment of progression, one which signals a shift to the end-of-life phase of advanced dementia.

The first question is whether the individual has progressed to a point where dementia is shutting down bodily systems so that death is imminent. The second question is whether we can objectively determine this point. Instruments such as the Functional Assessment Staging Tool (FAST) (Reisberg 1988) may typically be used for this purpose. The FAST tool measures functional deterioration, originally in people with Alzheimer's disease but now more widely, in moderate to severe dementia at a point when standard tests can no longer identify the subtlety in changes. It has seven scales from stage 1 ("normal") to stage 7 ("severe dementia") with stage 6 ("moderately severe dementia") typically being the stage at which names are forgotten and increased assistance is required.

But the use of such a tool with people who have an intellectual disability poses challenges as many would inherently meet the criteria for stage 6 of the FAST even without dementia, making progression to advanced dementia difficult to verify. They can have difficulty localising discomfort, pain, and other changes even before onset of dementia and their ability to self-report becomes more limited as dementia advances. The FAST may have utility for some individuals, but only if it is complemented with a clinical assessment that distinguishes between lack of function attributable to intellectual disability and that which may be attributed to dementia. Given the difficulty in using the FAST and lacking an intellectual disability-specific equivalent, the Prognostic Indicator Guidance contained in the Gold Standard Framework (Royal College of General Practitioners 2011) could offer an alternative means of ascertaining that adults with an intellectual disability might be nearing the end of life. This includes the "surprise question": "*Would you be surprised if this patient were to die in the next few months, weeks, days?*" The answer remains an intuitive one, pulling together a range of comorbidity, social, and other factors that give a whole picture of deterioration. If you would not be surprised, then measures should be considered to improve the patient's quality of life now and preparation made for possible further decline. It also recognises the impact of significant life changes such as a move

to a care home or experiencing a loss or bereavement, factors which may be subtler in their effect and harder to identify in people with intellectual disability.

Clinical assessment is a necessary component of ascertainment as it helps to identify other conditions that may be contributing to decline that may be life-threatening. While this is recognised as crucial for all people with advanced dementia (Alzheimer Scotland 2015), there are known health conditions associated with ageing and intellectual disability that often mimic or overshadow progression of dementia and confound the measures. New-onset seizures, for example, may occur in adults with Down's syndrome and contribute to rapid functional decline (Lott et al 2010).

A study in the south-west of England found that 42% of deaths among people with intellectual disabilities (not all with dementia) were considered premature and not anticipated, with a reasonable expectation that the person would have lived for at least one more year (Heslop *et al* 2013). Part of the problem is that many practitioners in dementia or older persons' care are uninformed about the range of conditions that can compromise health and function in older adults with intellectual disability (and especially those with Down syndrome) and so may miss causes of decline other than dementia. Given such diagnostic overshadowing, clinical assessment should also look carefully for these other factors, which may be confused with advanced dementia, and the correct treatment given to prevent premature death.

Place of care

Options for providing care will vary and will be dependent on living arrangements, which, along with other pre-existing circumstances, can be another point of difference with dementia more generally. As dementia onset often happens at a younger age among people with Down's syndrome than among adults in general, many remain living with parents or siblings. Given that the average age of onset for people with Down's syndrome is early 50s, their parents may find themselves with increased caring responsibilities at a time when they themselves are at the highest risk of dementia (Hodapp *et al* 2016).

So remaining in the family home may be a big challenge when older parents are the carers. When siblings become the primary carers, often because of the frailty or death of their parents, a

different set of issues arises. They may be in their 40s or 50s with other responsibilities such as child care and employment. In the general population, most dementia carers are a child or spouse of the person with dementia (Brodaty & Donkin 2009), but this is not the case for people with intellectual disabilities and dementia where there is a marked difference in the average age of carers and their relationship to the person concerned.

Although most people with intellectual disability in the UK and USA live with a family member, other places of care include shared accommodation with peers who have an intellectual disability, individuals living by themselves with outreach support, or increasingly with a partner or spouse who will also require support in their own right after a diagnosis of dementia in their partner. Support may be given by social care staff, inexperienced in dementia care, who have been employed to assist independent living and ensure people have ongoing choice and control over their lives rather than to adapt support for health and cognitive decline .

In such situations, advanced dementia-related physical care can be problematic, as the same staff are now faced with having to provide more extensive personal care. This requires care providers to rethink how they train staff to support people who have a terminal illness, wherever their home may be. For many people who live in shared group homes, paid carers have the same functions as family (Forbat & Service 2002). Yet, frequently they do not have legal authority for decision making, despite their long-term and detailed knowledge of the adult they support.

Increasingly in the UK, we are seeing a residential or nursing care home for older people as place of care for someone with Down's syndrome, often unplanned and in a crisis situation. , Here, the resident with Down's syndrome and dementia can be anything between 20 and 40 years younger than other residents. Staff in such homes often express fears that these residents are somehow too "different" to have their needs met properly and stigma and isolation often result (Watchman 2016). Residential care staff are often inexperienced in caring for people with intellectual disability and there is a risk that care becomes task-focused rather than person-centred (Watchman 2008). These factors can conspire to reduce the likelihood that people are referred in a timely way to palliative care and hospice services.

Active support

Another area of difference is in the domain of “active support”, a care approach embedded in intellectual disability practice. This involves assessment of the person with intellectual disability followed by a person-centred plan with opportunities for individual choice and self-management. Such plans are regularly reviewed based on the needs and wishes of the person and generally contain three core components: promoting participation, developing activity plans, and recording what works well to facilitate future development. The underpinning philosophy is ongoing support to prevent loss of optimum functioning and to maximise the autonomy of the person over his or her lifespan (Service, Lavoie & Herlihy 1999).

But intellectual disability services should revise this approach for the purposes of end of life care. Indeed, there is a powerful case for shared learning here between the fields of older people’s care and intellectual disability. Passive activities, such as simply “being” with the person, are not always recognised as active support in intellectual disability services even though they are increasingly common in advanced dementia care with older people.

In view of the prevailing care philosophies and practices in dementia care, there is an argument for intellectual disability care to adopt a more flexible approach to end of life care. This would mean a person-centred focus on maintaining and maximising current abilities rather than developing new abilities. Passive activities would be recognised as equally, if not more, viable than active support as traditionally conceived, given the limitations that dementia poses on the development of new skills.

Summary

Collaboration between palliative care, dementia care, hospice provision, and intellectual disability services is required rather than any one of these systems working in isolation. This is necessary because mainstream ageing and dementia-specific determination tools and scales may have limited value in discovering whether an adult with intellectual disability has advanced dementia and is close to dying. It would also help intellectual disability providers to consider how “passive support” can actually be a means of active support for someone who also has dementia.

The challenges for those involved with end-of-life support include enhancing education and training on advanced dementia, which may be particularly urgent for intellectual disability care staff. Training in personal care of people with advanced dementia would be helpful, as would understanding signs of subtle decline that may indicate approaching end-of-life and ensuring individualised supports or interventions in various living situations to enable dying with dignity.

Education and training on intellectual disability would be valuable for staff in generic residential care settings for older people, wherever they are places of care for people with intellectual disabilities at end of their lives. As a minimum, this should include understanding individualised methods of communication, which may be non-verbal prior to dementia, and knowledge of the non-dementia related health implications of ageing with an intellectual disability, particularly Down's syndrome.

While many needs of adults with an intellectual disability at the end of life are the same as for others, critical differences have been highlighted. Failure to address these differences can lead to unrecognised and unmanaged symptoms, further decline and a heightened risk of diagnostic overshadowing. Ill-health or end-of-life indicators can be wrongly attributed to the intellectual disability or the dementia, resulting in delayed referrals to palliative care or hospice care.

Summit recommendations

Dementia-specific practice guidance for use alongside generic end of life and palliative care guidelines:

- Some people with intellectual disability will have always had limited independent living skills; this should not be used as a criterion to assess progression of dementia.
- Understand the importance of previous communication methods used by the person with intellectual disability before the onset of dementia, including how pain was reported, particularly if the person communicates non-verbally or appears to make “just noises”.
- Recognise passive care as active support.
- Recognise and draw on the potentially wider range of existing or previous professional relationships for people with intellectual disability.
- Recognise the role that support staff often fulfil; the closest relationships that the person with intellectual disability and dementia has had may not always be with family.

- Recognise that for family members, especially parents, having a caring role did not begin with the onset of dementia; it has been lifelong and may be continuing despite their own advancing age (recognition and support for this should be provided when the person with intellectual disability is dying or dies).
- Co-morbidities are more common in people ageing with intellectual disability, especially Down's syndrome; treatable medical conditions should not be neglected because of the progression of dementia.
- "Home" as place of death differs; it encompasses community-based options, living with family members, or living with peers who have their own different health or social care needs.
- People with intellectual disability are under-represented in both palliative and hospice care; this should be considered as part of advance care planning.
- Practice guidelines and education should be shared among intellectual disability, hospice, palliative care, and dementia support services.
- Further work is needed on the development of appropriate tools and scales to determine nearness to end of life, and to facilitate inclusion in research of people with an intellectual disability and dementia.

Acknowledgements

This article derives from the International Summit on Intellectual Disability and Dementia held in Glasgow (13-14 October 2016), hosted by the University of Stirling and University of the West of Scotland, and funded by the RS MacDonald Trust, Scottish Government, and Alzheimer Scotland. Collaborating sponsors included the National Task Group on Intellectual Disabilities and Dementia Practices (NTG) and the University of Illinois at Chicago in the United States.

References

- Alzheimer Scotland (2015) *Advanced Dementia Practice Model: understanding and transforming advanced dementia and end of life care*. Edinburgh: Alzheimer Scotland.
- Brodsky H & Donkin M (2009). Family caregivers of people with dementia. *Dialogues in Clinical Neuroscience* 11(2) 217-228.
- Cordell CB, Borson S, Boustani M, Chodosh J, Reuben D, Verghese J, Thies W, Fried LB (Medicare Detection of Cognitive Impairment Workgroup) (2013) Alzheimer's Association

Recommendations for Operationalizing the Detection of Cognitive Impairment During the Medicare Annual Wellness Visit in a Primary Care Setting. *Alzheimer's & Dementia* 9(2) 141-150.

Forbat L, Service KP (2005) Who cares? Contextual layers in end-of-life care for people with intellectual disability. *Dementia* 4(3) 413-431.

Heslop P, Blair P, Fleming P, Houghton M, Marriott A, Russ L (2013) *Confidential Inquiry into premature deaths of people with learning disabilities*. Bristol: Norah Fry Research Centre.

Hodapp RM, Burke MM, Finley CI, Urbano RC (2016) Family caregiving of aging adults with Down's syndrome. *Journal of Policy and Practice in Intellectual Disabilities* 13(2) 181-189.

Holmerova I, Waugh A, MacRae R, Veprkova R, Sandvide A, Hanson E, Jackson G, Watchman K, Tolson D (2016) *Dementia Palliare Best Practice Statement*. Hamilton: University of the West of Scotland.

Lott I, Dierssen M (2010) Cognitive deficits and associated neurological complications in individuals with Down's syndrome. *The Lancet* 9(6) 623-633.

Marie Curie Cancer Care (2015) *Living and Dying with Dementia in Scotland: Barriers to Care*. Edinburgh: Marie Curie Cancer Care.

McCallion P, Hogan M, Santos F H, McCarron M, Service K, Stemp S, Keller S, Fortea J, Bishop K, Watchman K, Janicki MP and the Working Group of the International Summit on Intellectual Disability and Dementia (2017) Consensus statement of the International Summit on Intellectual Disability and Dementia related to end-of-life care in advanced dementia. *Journal of Applied Research in Intellectual Disabilities*, DOI: 10.1111/jar.12349 .

Prasher VP, Janicki, MP, Jozsvai J, Berg JM et al (2017) Alzheimer's disease and dementia: Implications for people with Down syndrome and other intellectual or developmental disabilities in Wehmeyer M, Brown I, Percy M, Shogren KA, Fung WLA (eds) *A Comprehensive Guide to Intellectual and Developmental Disabilities*. Baltimore, Maryland: Brooks Publishing Co.

Reisberg B (1988) Functional Assessment Scale (FAST) *Psychopharmacological Bulletin* 24(4) 653-659.

Royal College of General Practitioners (2011) *The GSF Prognostic Indicator Guidance*. London: RCGP.

Service K, Lavoie D, Herlihy JE (1999) Coping with losses, death, and grieving. In Janicki MP, Dalton AJ (eds) *Dementia, aging, and intellectual disabilities: A handbook*. Philadelphia: Brunner-Mazel

Watchman K (2008) Changes in accommodation experienced by people with Down's syndrome and dementia in the first five years after their diagnosis. *Journal of Policy and Practice in Intellectual Disabilities* 5(1) 61-63.

Watchman K (2016) Investigating the lived experience of people with Down's syndrome and dementia: overcoming methodological and ethical challenges to inform policy and practice. *Journal of Policy and Practice in Intellectual Disability* 13(2) 190-198.

