

Assessment of dementia in persons with intellectual disabilities: challenges and opportunities

Introduction

The assessment and diagnosis of dementia in persons with Intellectual Disability (ID) is complex. Many of the difficulties are confounded by pre-existing intellectual impairment, additional physical and mental health co-morbidities and environmental and resource issues resulting in dementia often being quite progressed before any diagnosis is made. *The key to accessing appropriate dementia care and the basis for many of the therapeutic interventions is early recognition and diagnosis.* A decade has passed since consensus recommendations for screening and diagnosis of dementia in persons with ID were published (Alyward *et al.* 1995), however the development of memory clinics and routine screening are generally not part of the healthcare landscape for persons with ID. Recent research and clinical experience has contributed to a greater understanding on how dementia presents in persons with Down Syndrome (DS) and there has been success in the development of test instruments which are capable of detecting and measuring decline in persons with varying degrees of ID. *There is no cure yet for Alzheimer's disease, but no cure does not equate to no treatment,* and much can be done to improve quality of life and assist the person and their carers to cope more effectively with increasing disability—an inherent part of the disease. However, if persons with ID are to truly benefit from the advances in treatments and therapeutic interventions, dementia screening and early diagnosis are essential. This article will briefly explore some of the challenges in dementia screening and diagnosis and offer some tips and suggestions to assist families/carers and clinicians to understand and recognise change and be more proactive in eliciting help.

Why is dementia diagnosis difficult?

Pre-existing intellectual impairment

Initial accurate diagnosis of dementia is difficult for everyone, but a lack of scales with established cut-off scores, and factoring out the impact of underlying levels of ID, make it particularly difficult for persons with ID.

A clinical diagnosis of dementia in populations without pre-existing intellectual disabilities is based upon evidence of progressive deterioration in the person's memory, cognition, and day-to-day living skills (ICD-10) (WHO 1992) which is generally supported by widely accepted cut-off scores for dementia diagnosis using validated scales such as the Mini-Mental State Examination (MMSE) (Folstein *et al.* 1975). *There is no equivalent to the MMSE in diagnosing dementia in persons with ID. There is no quick 10-minute screen for dementia in persons with ID and comprehensive screening and assessment can take hours.*

For persons with an intellectual disability, ICD-10 criteria are still important, but many will have had lifelong impairments in cognition and learning making scales for the general population (like the MMSE) of limited value as a diagnostic tool, particularly in persons who function within the moderate to severe range of ID (Deb and Braganza 1999). Despite continued efforts and indeed much achievement in the development and validation of both informant-based (carer-rated) and objective test instruments (client-rated) *to date there is no agreed consensus on the optimal battery of test instruments to be used in detecting and diagnosing dementia in persons with varying degrees of intellectual disability, nor is there an established rapid assessment instrument with agreed cut-off scores.*

Instead, diagnosis of dementia in persons with ID remains a process of recognising change and decline in the person's previous level of functioning that must be understood and

evaluated in the context of a pre-morbid level of functioning, i.e. the person's capabilities and needs prior to the onset of perceived changes. *Also, in contrast to the generic population, changes in cognition and memory often appear not to be the most obvious initial presenting symptom of dementia in persons with ID. Changes in personality, behaviour and global day-to-day function are often more relevant as warning signs.* Identification of such changes is more likely if there is frequent and early screening for such changes; recommendations are for annual baseline screening for persons with DS over the age of 35 years, and for persons with ID from other aetiologies over the age of 50 years (Alyward *et al.* 1995).

Lack of routine screening

One of the major concerns is that annual screening for dementia is not part of general medical care, and memory clinics are not part of the landscape of ID services. Evidence from the generic population (Pitt 2001), and limited but emerging evidence from the field of ID in the US and UK (Chicoine *et al.* 1999; Hassiotis *et al.* 2003), suggests that the memory clinic model is effective in terms of supporting screening and consensus diagnosis and care management of dementia in people with ID. There are pilot efforts to establish a memory clinic in Ireland (McCarron *et al.* 2005), however, comprehensive baseline screening of persons with ID is often unavailable and generally not routine in many services in Ireland or, indeed, elsewhere.

Service culture

There are additional challenges. Services for persons with ID are generally based on a culture of caring, and this can at times create additional challenges when it comes to dementia assessment, in that many of the environments may be sheltered, and sometimes un-stimulating. There may be few intellectual demands on the person, which often result in the more subtle memory impairments evident in early dementia being difficult to detect. For example, the person may have never had the opportunity to cook simple meals, choose their personal clothing, plan their day, etc. Thus, identifying problems in the areas of memory, judgment, and planning is more difficult, reducing opportunities to observe critical changes in behaviours and skills.

Additionally, carers working with persons with ID are very often accustomed to providing quite intensive supports to people, and so it does not cause alarm or concerns if a person requires help. Gradually too, over time the environments and levels of supports are adapted and increased to meet diminishing abilities without any real realisation that the individual is declining. This is further confounded by frequent staff changes and sometimes lack of prior knowledge of the person's previous abilities and needs. Furthermore, staff and family carers are often confused between the symptoms of normal aging and dementia in persons with ID. The cumulative impact is that there is an increased risk that the person may be at quite an advanced stage of dementia before appropriate referral and diagnosis are made.

Health co-morbidities

There are a host of other health co-morbidities which are common in aging persons with DS, some of which may mimic dementia, or on the other hand may often co-exist as part of dementia. *While the rates of AD increase with age in persons with DS, it does not necessarily mean that all people with DS will develop dementia. It is necessary to rule out the possibility that the experienced changes and symptoms observed have a*

different and perhaps more easily treatable cause. Some common conditions which may mimic dementia and need to be ruled out include:

- ◆ Depression or other mental illness
- ◆ Sensory impairment: vision/hearing
- ◆ Thyroid impairment
- ◆ Vitamin B12, folate deficiency
- ◆ Medical problems—acute/chronic illness, infection, pain, epilepsy, drugs-related concerns
- ◆ Sleep Apnoea
- ◆ Major life events: Separation, bereavement, environmental/living situation changes.

How is Alzheimer's dementia diagnosed in people with intellectual disabilities?

The most valuable information to assist with diagnosis is an accurate history of change by family/carers who have known the person for quite a long time.

Only if there is understanding about the person's past ability by family/staff who have known the person for a long time can a judgment be made about whether there has, or has not, been change.

It is important that family/carers note specific examples that demonstrate change, i.e. things that the person used to be able to do, but now is no longer able to do, or now requires more reminding to carry out tasks that previously they would have been able to do themselves.

Tests for dementia can only be considered positive if there is written evidence of a decline in memory and mental ability.

Although cognitive changes and memory are important for the diagnosis of dementia, changes in other aspects of behaviour are also relevant. For example, for individuals with Down Syndrome (especially those who are very low functioning) changes in personality, behaviour and mood, and general loss of interest in day-to-day events are also important. These sometimes are the first changes observed and reported by family/staff. It is critical to have some knowledge and some record of the person's previous personality, their interests and their general ability and level of independence and support required in day-to-day activities such as personal hygiene, dressing, eating, orientation, writing, reading.

Persons with ID without Down Syndrome

We have some understanding of aging and dementia in persons with DS, but we have limited understanding on aging and dementia in other people with ID. Cooper (1997) found that persons with ID from other aetiologies were at about five times greater risk of developing dementia compared to an aged matched cohort she drew from the generic population in a selected UK catchment area. Janicki and Dalton (2000), drawing from a state developmental disabilities dataset, found about equal risk. While dementia of the Alzheimer's type is most commonly seen in persons with DS, it is likely that persons with ID without DS will present with the greater range of the sub types of dementia also evidenced in the generic population, such as vascular dementia, lewy body dementia and fronto-temporal dementia, as well as Alzheimer's type dementia. In the generic population researchers are beginning to look at connections between dementia symptom presentation and other pathologies such as bipolar disorder (see, for example, Cavanagh *et al.* 2002). Similar answers are needed on whether, for example, dementia will present differently in persons with ID who are autistic, have had a longterm history of depression, behaviour problems, severe learning difficulties and limited education, and longterm history of medication use etc. A commitment to early and frequent screening will facilitate the development of longitudinal studies targeted at understanding dementia in different groups.

What are the main areas that decline/deteriorate in Alzheimer's dementia?

Memory: One of the most common early signs of dementia is forgetting recent information. The extent of memory changes will vary depending on the person's level of intellectual disability. There may be some minimum memory loss, particularly of recent events. The person, for example, may forget that they have just had their dinner, yet may remember clearly an event that happened years past. Forgetting what they have just done, where they have been or who has just visited are often a major symptom of Alzheimer's dementia. The person may forget common day-to-day routines and places. They may also have difficulty in remembering names of familiar people that used to be known.

Disorientation to time and place: The individual may appear disorientated and confused. They may wander and have difficulty in locating familiar places, for example, the toilet or their bedroom. They may be unaware that they are entering or even in another person's room.

There is also often a loss of sense of time. The person may have difficulty in understanding the passage of time, which can be a source of stress and anxiety. For example, they may repeatedly ask what time it is and feel they are late for an event. There may also be disturbance to the 'internal clock' and the person's sleeping pattern may become disturbed. The person may wake during the night and feel it is morning, and may become agitated when guided/encouraged to return to bed. They may have difficulty understanding directions or instructions and become disorientated as to where they are or where they were going. As the disease progresses, memory losses become more pronounced.

Misplacing items: Everyone from time to time can temporarily misplace items. The person with dementia may often misplace and lose items, for example, their watch, glasses, etc. and may blame others. They may hoard items, put things in unusual places and may not remember where they have put them.

Language: There may be specific problems with language. The individual may experience difficulty in finding the right words to use during casual conversations. They may have difficulty naming objects or with maintaining a logical conversation. The person may appear to know what they want to say, but just can't say it.

Personality and mood changes: Severe changes in personality may become obvious, and social behaviour may be marked by suspiciousness and delusions.

Problems with day-to-day living activities: The person may begin to experience loss of self-care skills, regarding eating and use of the toilet. For those in a workshop or other job, work performance may begin to deteriorate. Finally, the disease will progress to the point where all abilities to function normally are lost, and affected individuals need total care. Such deterioration may occur over different time periods for different individuals, ranging from 12 months to 11 years approximately.

Some important considerations in the assessment process

As previously alluded to, there is *no gold standard* nor an agreed optimal battery for screening and detection of dementia in persons with varying levels of ID. However, there are a variety of validated informant-based and objective test instruments which have been recommended. There are two core tasks: screening and comprehensive diagnostic work-up.

Screening: We need to screen wisely, sparingly but nonetheless comprehensively to avoid over burdening persons with ID and their carers. Screening is only helpful if (1) it is repeated regularly, (2) it identifies persons at risk (requiring expert and timely

interpretation of screening results within the context of ICD-10 criteria), and (3) if when warning signs are identified they are followed up with a comprehensive diagnostic work-up.

Diagnostic Work-up: The components of a comprehensive diagnostic work-up are listed in Table 1. The range of tests and work-ups recommended reflects that there may be a variety of conditions that may result in symptoms similar to or accompanying dementia.

For comprehensive diagnostic work-ups to occur critical issues are commitment by providers to supporting this level of assessment activities, the availability of staff trained in the use of the recommended tests and instruments, a coordinated and sensitive approach to the assessment process, and the locating of informants. It has been acknowledged that this is best operationalised through the development of specialised memory clinics preferably at regional level (McCarron and Lawlor 2003).

Table 1: Components of a Comprehensive Diagnostic Workup

History: Family/personal/medical/psychiatric social/medication

Medical & Multidisciplinary Team Work-up

- Vital signs
- Full physical and Neurological examination
- Vision and hearing assessment
- Mental health screening
- Neuropsychological testing using both informant-based and objective instruments
- Sleep and weight graphs

Lab Work and other Tests

Required

Complete blood count- FBC, E/LFT, ESR, TFT, Folate & B12, Glucose, U&E, drug levels, Urinalysis, Electrocardiogram(ECG)

Recommended for select situations

Chest x ray, CT, MRI

Consensus Diagnosis

Determination of dementia or alternative diagnosis as a result of a multidisciplinary review of all testing and examinations

Some useful tips

Choice of instruments should be guided by the person's underlying level of ID and the availability of key informants who have known the individual for a considerable period of time (and most importantly prior to any perceived changes) to provide relevant background information and history.

To ensure consistency and reliability in the application of instruments, training is required either by someone who has had the relevant training and/or who has clinical skills and experience in their application. Attention to and adherence to the guidelines for a scale issued by the original authors are critical.

A combination of both informant-based and objective testing is preferable if at all possible.

Longitudinal use of selected instruments with each person at risk will greatly enhance the possibility of detecting change over time even in individuals who have always functioned at severe and profound levels of cognition.

Alyward *et al.* (1995, 1997) give some examples of recommended test instruments.

The need for training

Improved assessment will require that an urgent need is training in symptom recognition for families and staff at all levels. Additional professional training is also required for key groups of people (such as nurses, doctors, social workers, psychologists etc) to address the issues of screening and dementia care in this

population. In particular, the training should attend to optimising the skills of individual members of the multidisciplinary team in interpreting screening results and in the execution of comprehensive diagnostic work ups.

In countries like Ireland and the UK, intellectual disability nurses are more likely to be at the forefront of care delivery and hence have a pivotal role to play in recognising change in those areas known to decline in AD. Again training is critical and Ireland is at the forefront of efforts to create specialised training for ID nurses with the creation of the first Higher Diploma in Specialist Nursing in Intellectual Disabilities and Dementia being offered at Trinity College Dublin. This effort to create a cadre of trained specialists will likely be an important step in addressing assessment needs.

Conclusion

Dementia symptoms are a new and growing concern for people with ID. The situation is even more difficult because of a lack of good instrumentation, difficulties in diagnosis, the masking effect of life long cognitive impairments, and the possibility that other conditions may be influencing symptoms. Perhaps the greatest difficulty is the lack of experience and preparedness by ID services providers and professionals. A concerted and organised effort is needed which at the very least requires a comprehensive approach to training staff in both identifying persons at risk and in undertaking comprehensive and competent diagnosis. Instituting routine, regular and early screening, establishing memory clinics and developing a cadre of trained staff are therefore challenges to be faced by ID providers. Researchers and practitioners must also continue work on establishing an agreed assessment protocol with recommended instruments and established cut-off scores.

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References

Aylward, E., Burt, D., Thorpe, L., Lai, F., Dalton, A. 1995 *Diagnosis of dementia in individuals with intellectual disability*. Washington DC. American Association on Mental Retardation.

Aylward, E., Burt, D., Thorpe, L., Lai, F. and Dalton, A. 1997 Diagnosis of dementia in individuals with Intellectual disability. *Journal of Intellectual Disability Research*. 41, 152 –64.

Cavanagh, J.T.O., van Beck, M., Muir, W. and Blackwood, D.H.R 2002. Case-control study of neurocognitive function in euthymic patients with bipolar disorder: An association with mania. *British Journal of Psychiatry* 180, 320–26.

Chicoine, B., McGuire, D. and Rubin, S.R. 1999 Speciality clinic perspectives. In M.P. Janicki and A.J. Dalton (eds). *Dementia, aging, and intellectual disabilities*. USA. Taylor and Francis, pp.278–91.

Cooper, S.A. 1997 High prevalence of dementia among people with learning disabilities not attributable to Down's Syndrome. *Psychological Medicine* 27, 609–16.

Deb, S., and Braganza, J. 1999 Comparison of rating scales for the diagnosis of dementia in adults with Down's Syndrome. *Journal of Intellectual Disability Research* 43 (5), 400–07.

Folstein, M.F., Folstein, S.E. and McHugh, P.R. 1975 Mini-Mental State: A practical method for grading the cognitive state for the clinician. *Journal of Psychiatric Research* 12, 189–98.

Hassiotis, A., Strydom, A., Allen, K. and Walker, Z. 2003. A memory clinic for older people with intellectual disabilities. *Aging and Mental Health* 7 (6), 418–23.

Janicki, M.P. and Dalton, A.J. 2000 Prevalence of dementia and impact on intellectual disability services. *Mental Retardation* 38 (3), 276–88.

McCarron, M. and Lawlor, B.A. 2003 Responding to the challenges of ageing and dementia in intellectual disability in Ireland. *Ageing and Mental Health* 7 (6), 413–17.

McCarron, M., McLoughlin, M. and McCallion, P. 2005 A memory clinic for persons with intellectual disabilities. Alzheimer's Europe Conference, Killarney, Ireland.

Pitt, B. 2001 The history of memory clinics. In Z. Walker and R. Butler (eds) *The memory clinic guide*. London. Martin Dunitz, p.4.

World Health Organization (WHO) 1992 *The ICD-10 classification of mental and behavioural disorders, clinical descriptions and diagnostic guidelines*. Geneva. WHO.