College of Psychiatrists of Ireland

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A Guidance Document on Dementia in Persons with Intellectual Disability

A Paper by the Faculty of Learning Disability Psychiatry of the College of Psychiatrists of Ireland

Approved by Council
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Preface

The Faculty of Learning Disability Psychiatry of the College of Psychiatrists of Ireland commissioned a subgroup to write this paper.

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Executive Summary & Recommendations

This report was completed by the Faculty of Learning Disability Psychiatry to provide guidance to those working in clinical settings with persons with an Intellectual Disability and provide accessible information on patients for their families. The recommendations relate to the assessment and management of suspected or diagnosed cognitive decline or dementia.

Recommendations:

1. Each service for people with Intellectual Disability should have a plan or access to a plan (e.g. HSE) for recognition and management of dementia in persons with Intellectual Disability (ID).

2. People with Intellectual Disability should have baseline screening from the age of 35 years in people with Down Syndrome (DS) and 50 years in people with ID from other causes using a battery of carer rated tests and direct assessment. Repeat assessments should occur every 3–5 years. Where concern arises, repeat tests should occur 6-12 monthly.

3. A multidisciplinary team comprising of a psychiatrist, psychologist, specialist nurse, physician, social worker, occupational therapist, speech and language therapist and physiotherapist should be in place for the care and management of persons with both ID and dementia.

4. The team or the psychiatrist and psychologist working together should assess, agree and communicate the diagnosis of dementia.

5. Families and patients should be involved in discussion using accessible information.

6. Services should be comprehensive and allow the individual to age in place.

7. Palliative care should be available.

8. Trainees specialising in psychiatry of Learning Disability should have training in diagnosis and management of dementia.

Introduction

The prevalence of dementia for people with Intellectual Disability (ID) is increasing. People with ID are living longer due to advances in medical care and living circumstances. In Ireland there are 3,154 people with Intellectual Disability over the age of 55 & 10,725 over the age of 35 years. 41.1% of the total ID population is over the age of 35 (HRB, 2009).

People with Down syndrome also have an increased risk of dementia with 15 to 40% lifetime prevalence (Prasher and Krishnan 1996). This is due to the possession of a triple copy of Chromosome 21. In addition people with ID (without DS), also have an increased risk of dementia although the reasons are unclear (Cooper 1997). Services for people with ID in Ireland differ in their provision of expert clinical care and their underlying philosophy of care as to whether the specialist ageing services should be provided in-house or by using a generic service provider for the aged.

People with ID and dementia differ in a number of ways from the general population, including presenting at a much younger age. Based on the level of ID individuals may have significant associated communication difficulties making an accurate diagnosis challenging. In addition access to appropriate ageing services may be more difficult than the general population. Instruments designed to detect dementia in the general population assume a normal IQ whereas people with ID perform poorly with these instruments to detect dementia (Sturmey, 1991; Deb 1999). Various medical and social problems may mask the diagnosis and careful evaluation is necessary to establish a diagnosis of dementia.

The presentation of dementia in people with intellectual disability may be subtle and may go unrecognised. As in the general population, memory loss is an important feature but this may be masked. Loss of adaptive function may be the first sign of dementia and may be associated with slowing down in the workshop or the decrease of self-care skills in the home domain. A frontal lobe pattern of decline with apathy, poor motivation and behavioural changes may occur (Ball et al. 2006). As the disease progresses there is loss of speech, altered gait and finally dysphagia, immobility and incontinence. The development of epilepsy is an important feature of dementia in Down syndrome that is not seen to the same extent in the general population.

This document is intended to provide information to doctors working in the domain of intellectual disability services, however it may be also be of support to others that require guidance on this topic such as non-medical staff, family members or carers.

1. Diagnosis

There are inherent difficulties in making the diagnosis of dementia in people with Down syndrome and other causes of Intellectual Disability. Ideally, according to international recommendations, baseline cognitive testing on individuals should be performed in people with Down syndrome once they reach the age of 35 (IASSID). As each individual has a different baseline IQ and level of adaptive functioning, it is important to record this before development of disease. However, in practice such testing may be determined by the provision of local appropriate specialist care. At a minimum, each service should provide reactive screening in any individual in whom concerns have been raised. Following baseline screening, where there are no immediate concerns longitudinal follow up at regular intervals (3-5 years) should occur. If there is evidence of decline then the next assessment should occur between 6 and 12 months.

2. Assessment

The assessment should be multidisciplinary, however it is recognised that not all services have access to a full team. As yet no consensus is in existence agreeing a particular battery however IASSID (2009) and RCPsych/BPS (Dodd, 2009) have recommended a battery of tests that may be used depending on the clinicians preference. At present, there is no equivalent test to the Folstein Mini-Mental State Examination.

Ideally a clinical psychologist, clinical nurse specialist or other appropriately trained individuals (working with ID services or primary care teams if available) should carry out testing; however there are some services without access to such expertise and a psychiatrist or specialist nurse may use some of the following tests with the exception of the DSDS.

The assessment should be conducted with support from an individual very familiar with the patient. This may be a family member, carer or key worker. It is important that a variety of collateral histories be available to the assessor to cover the various domains such as home, day service, residential settings etc. In addition, the setting in which the testing is to be conducted should be familiar to the patient and the sensory environment be free of distractions such as noise, interruptions etc.

3. Test Battery

In practice a battery of tests including assessments of cognitive and adaptive function and a carer rated questionnaire should be used. The International Association for Scientific Study for People with Intellectual and Developmental Disability (IASSIDD) has recommendations for such tests (Burt and Aylward, 2000). Common carer rated tests used in practice are the Dementia scale for people with Mental Retardation DMR (Evenhuis, 1992, 1996) and the DSDS (Down Syndrome Dementia Scale (Gedye, 1995) or CAMDEX-DS (Ball et al, 2006).

A measure of adaptive function should be performed such as the Vineland adaptive behaviour scales.

If possible, direct cognitive tests using an instrument such as the Test for Severe Impairment or CAMCOG-DS (Ball et al, 2006) and also a test of memory (Fuld object memory evaluation tests) as recommended in the IASSID battery (Burt and Aylward, 2000) should be performed. See Table 1.

4. Investigations

In cases where dementia is suspected, possible reasons should be ruled out. In the differential diagnosis of dementia in those with an intellectual disability, Pary (1992) listed the following conditions that should be considered in functional decline in a person with Down syndrome: depression, hypothyroidism, infection, folate and B12 deficiency, hearing impairment, visual impairment, malignancy such as leukaemia, joint problems of neck, knee or hip and sleep apnoea. It should be noted however that the presence of a co- morbid disorder does not rule out the possible diagnosis of dementia or vice- versa.

A full medical evaluation including a comprehensive physical examination, hearing and visual assessments should be performed on each individual. Biochemical and haematological testing should include thyroid function, calcium levels, renal and liver profiles, full blood count, B12 and folate; erythrocyte sedimentation rate (ESR), fasting glucose and lipids, syphilis serology if indicated, an ECG and a psychiatric assessment to rule out the presence or absence of mental health problems and to establish the diagnosis. Other possible causes may include bereavement, abuse, environmental deprivation, or polypharmacy. Specialised investigations such as EEG, CT Scanning and MRI may be considered but are not necessary in uncomplicated presentations, except where another illness needs to be excluded or the diagnosis is unclear. Occasionally it may be necessary to consult a medical colleague for support and advice in particularly complex cases.

5. Management

Once the diagnosis has been made, the multidisciplinary team should communicate the diagnosis to carers and family as soon as is practical. Members of the multidisciplinary team should include a consultant psychiatrist in intellectual disability, a clinical psychologist, a social worker and specialist nurses with access to occupational therapy, physiotherapy, and speech and language therapy. However, as previously discussed many services are without such teams and the diagnosis may have to be made by either a psychiatrist or psychologist working alone. It is recommended that a meeting to reach a consensus on diagnosis and intervention be held to ensure a multidisciplinary approach to further management.

The individual also has a right to be informed as much as possible about their illness. There are some useful books for people with ID that deal with growing older and dementia published by the British Institute for Learning Disabilities (Dodd et al, 2005). The individual's GP and family should be informed of the diagnosis and management plan as they may be increasingly involved as the disease progresses. Others who share living arrangements or work space with the person may also need support to understand the changes in their friend or colleague. The 'Beyond Words' book series may be useful for those with an intellectual disability (Hollins et al., 2011).

A full multidisciplinary team approach to the care of individual and others should include supporting them as long as possible in familiar surroundings as the overriding philosophy of care should be that of 'Ageing in Place' so as to maximise the working memory for as long as possible. This can be further supported by the provision of community nursing, respite care and specialised day centre care. Ideally this should take place within an intellectual disability setting as users of generic settings may have different life experiences and people with ID may experience stigma in this situation. Non-pharmacological therapies such as reminiscence therapy, life books, and music and art therapy are strategies that can be used within a memory clinic setting.

6. Medication

The NICE Guidelines (2006) give clear advice on the use of medication in people with intellectual disability and dementia.

Once the diagnosis is made, individuals with ID should be considered for a trial of anticholinesterase medication such as Donepezil, Galantamine or Rivastigmine. The evidence suggests that there is some benefit however there are few robust trials (Prasher et al. 2003, Lott et al. 2002). People should be monitored regularly whilst on these medications using either the DMR, CAMDEX- DS or the DSDS, as referred to above.

The use of Memantine should be under the care of a consultant psychiatrist though a recent multi-centre trial has recently not shown any benefit (Hanney et al, 2012).

Other medications for behavioural correlates of dementia such as transient psychosis, sleep disorder, and aggressive behaviour should only be considered if there is a clear risk benefit analysis and a lack of response to other interventions.

Side effects may outweigh the benefits, and the aim should be to analyse the social and environmental domains before medication is considered. The exception to this is depression. Depression should be treated with one of the newer generation antidepressants as the anticholinergic and adrenergic effects from older medication can impair cognitive function.

Due to the increased risk of CVA the use of atypical antipsychotics should only be considered after careful diagnosis, analysis of risk and benefit and full discussion with carers, and the lowest possible dose of a drug should always be used.

In sleep disorder, a review of sleep hygiene practices may be sufficient to guide interventions however if medication is required Trazadone may be useful at a low dose.

Physician advice may be sought for treatment of epilepsy and other complications. Epilepsy is common in the dementia of those with Down syndrome with generalised seizures, myoclonic jerks and atonic seizures presenting as the disease progresses.

7. Care Environment

Although 'Ageing in Place' should be the first option, as the disease progresses the need for specialised service increases. Respite and home supports may no longer be sufficient despite environmental adaptations. The need for long term placement may arise and if required should occur within the service of a specialised service which will best meet the need of a client with ID who may not have behavioural difficulties but who will certainly have mobility and swallowing problems. As in the general population this should be near and accessible for family and friends to visit. Standards of care for services are needed and a template for these has been drawn up by the Daughters of Charity Service (McCarron & O'Reilly 2010).

In late stage dementia, there are issues regarding continued feeding, recurrent chest infections and pain relief and best practice palliative care may be needed to support dignity and quality of life. On-going liaison with the individual's family and carers is important to support decisions on management issues before they arise and to educate as the disease progresses.

8. Training Requirements

As people with ID are living longer, age related problems such as dementia are increasing. It is recommended that each trainee in the psychiatry of learning disability receives appropriate training in the area of dementia, either as part of an attachment in the Psychiatry of Learning Disability or by an attachment to Psychiatry of Old Age. Some ID services are now using generic services to deliver care and it is essential to maintain expertise in this area as the psychiatrist in intellectual disability is best placed to understand the multiplicity of issues for the individual with both ID and dementia.

The support and education of family, carers and staff is crucial to ensure that best quality care is given to the individual. The psychiatrist individually or as part of a team has a role to ensure this need is addressed.

Appendices

TABLE 1

LIST OF INSTRUMENTS THAT MAY BE USED IN DIAGNOSIS

DIRECT ASSESSMENT OF COGNITION
Test for Severe Impairment + FULD object memory tests Or CAMCOG **CARER RATED (at least one of)** DSDS (Dementia scale Down syndrome); only valid if administered by a trained clinical psychologist Or DMR (Dementia in persons Mental Retardation) or Camdex DS **ADAPTIVE BEHAVIOUR SCALES** Vineland ABS

Please note that this list is not exhaustive and that consultation with a clinical psychologist on the most appropriate instruments is always advisable.

Definitions

Intellectual Disability

The definition of Intellectual Disability used in this paper is synonymous with that of Mental Retardation in the ICD-10 and DSM-IV, Classification of mental and behavioural disorders.
It is a condition of arrested or incomplete development of the mind which is especially characterised by impairment of skills manifested during the developmental period. Adaptive behaviour is always impaired. The degree of cognitive impairment is measured by an IQ test. Individuals are grouped according to their intellectual level.

Levels of Intellectual Disability

Data received from ICD-10 and DSM-IV

	IQ Range
Mild	50-70
Moderate	33-49
Severe	20-34
Profound	<20

Individuals that lie within the mild range of intellectual disability have minimal impairments in adaptive functioning. The majority of this group do not require specialist mental health services in Intellectual disability.

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Further Reading - Resources for Families and carers

Dementia and Down Syndrome –Factsheet www.dementiaweb.org.uk

Learning Disabilities and Dementia. Alzheimers.org.uk website

Alzheimer's Dementia: what you need to know, what you need to do. Max Neill
www.intellectualdisability.info

Down Syndrome and Dementia: a resource for carers and support staff
Karen Dodd, Vicky Turk and Michelle Christmas. BILD publications