

## Foreword

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It is a great pleasure to be asked to provide a foreword for this excellent guidance on dementia and people with intellectual disabilities. The awareness of dementia has never been higher and the emphasis on providing support to people following a diagnosis is most important. We know that dementia attracts stigma and that is of at least equal if not more relevance in people with intellectual disabilities. Professionals and the public are generally more aware of the increased risk of developing dementia in people with intellectual disabilities.

The guidance is comprehensive across the pathway from diagnosis, post diagnostic support including assessment, through management and end of life care. Its strength is in its comprehensive nature, including, in the same breath, those practical and clinical suggestions which are so important in the day-to-day care of people as well as the evidence base for these actions. The multi-professional nature of the guidance is so important.

One of the things which has been achieved in dementia in the general population, particularly older people, is to convey a message that dementia is a challenge for everybody. Information and guidance has been key to this and the information provided in this guide will go a long way to reassuring people that issues regarding cognitive impairment and dementia in people with intellectual disabilities are not, in all cases and as a matter of course, the purview of the expert and the specialist.

Everyone knows that, thanks to advances in medical care and the social environment, people with intellectual disabilities are now living longer, a great success and a tribute to society, and that brings with it some challenges in terms of the need for more services for people with cognitive impairment and intellectual disabilities.

This guidance has been created carefully by the recognised authorities in the arena and has been woven together expertly. It is accessible and yet comprehensive, it is practical but not simplistic but most importantly it whets the appetite of the reader to learn more. Although concentrating on initiatives in England, an international audience could read it with benefit.

I certainly learnt more about dementia in people with intellectual disabilities in reading this than I have ever before.

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## Purpose of this guidance

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This report is a revision to the original joint British Psychological Society and the Royal College of Psychiatrists (2009) guidance on dementia and people with intellectual disabilities. It has been written by a joint working group of the DCP Intellectual Disabilities Faculty of the British Psychological Society and the Royal College of Psychiatrists.

The main purpose of the guidance is to enable those working in clinical and social care services to improve the quality of life of people with intellectual disabilities who develop dementia, by providing guidance to inform assessment, diagnosis, interventions and support. The guidance is aimed at clinicians in intellectual disabilities and older peoples' mental health services and services for younger people with dementia. The decision about which services provide which part of the care pathway is a local decision to be taken by commissioners and providers, but ensuring that all elements of this guidance are considered and in place for people with intellectual disabilities and dementia.

Since the original guidance was published there has been a far greater awareness about dementia in the general population and a proliferation of strategies and standards documents. However, dementia and people with intellectual disabilities has still received minimal focus. The current guidance has been updated using both the current research literature and the experience of senior clinicians working in the field. Some areas of the guidance have only needed minor changes, whilst others have had a more extensive re-write. Three completely new sections have been added.

As before, our main focus has been to highlight the key factors that we consider are the elements of an excellent service, and to help those working in services evaluate how they might help the increasing numbers of people with intellectual disabilities who are developing dementia given improvements in life-expectancy.

It is with these principles in mind that this report has been produced and has the following aims:

- To bring together available and relevant evidence-based practice with a consensus of clinical opinion and experience.
- To provide a framework for good practice and for the development of multi-agency care pathways.
- To promote effective and timely assessment, diagnosis, and interventions for people with intellectual disabilities suspected or confirmed as having dementia and to ensure quality support to them and their staff and other carers.
- To provide guidance for service providers, developers and commissioners.
- To provide a set of standards of good practice against which service provision can be benchmarked and audited.
- To provide a quality outcome measure to use to evaluate what the person with intellectual disabilities and dementia is experiencing in their care.
- To promote the development of comprehensive and effective local services and to reduce the number of individuals who are failed by current service provision.

- To provide a framework for training health and social care professionals and paid support staff and carers.
- To guide the future development of services.

The report was produced through the combined work of members of the working group and drew on the published evidence base and from the working group members' extensive clinical experience in this area. Although this report has been undertaken primarily by psychologists and psychiatrists, we recognise that people with dementia must have effective multi-agency and multi-disciplinary services. We believe that this report will therefore be relevant to anyone who has an interest in dementia and people with intellectual disabilities, including health and social care professionals, families, paid staff, advocates, service providers and commissioners.

We still cannot be prescriptive within the document, particularly about the choice of assessment tools, because the evidence is not available to support particular instruments. Decisions will still need to be made locally depending on local resources and configurations. However, the report is intended to highlight the specific issues that people with intellectual disabilities and dementia present, and to ensure that local services are timely, effective and ensure that the person continues to have a high quality, safe and person-centred lifestyle as the dementia progresses.

This is a rapidly developing area with new knowledge and practice developing all the time. Research with people with intellectual disabilities and dementia is increasing, and will add to the evidence base.

Finally, the report hopes to complement other publications and guidance in this area and to provide a way forward for supporting people with intellectual disabilities who develop dementia.

# Key Points from the guidance

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## Section 1 – Context

In March 2012 the Prime Minister, David Cameron, set out what he called ‘the Dementia Challenge’ (DH, 2012). Recognising the urgent need for change, he set targets for improvements by 2015 in three broad areas:

- 1) health and care,
- 2) the establishment of dementia friendly communities, and
- 3) improving dementia research.

This document has been revised with these challenges in mind, paying particular attention to how they apply to people with intellectual disabilities. It sets out standards of clinical practice in the areas of assessment, diagnosis and interventions for people with intellectual disabilities who develop dementia with an emphasis on how people with intellectual disabilities who develop dementia can be best supported in a manner that maintains that person’s dignity and his/her quality of life.

Although this document primarily references the context of dementia in England and the other UK nations, publications from other nations are equally as relevant and have the same underlying principles and practices outlined in them, for example, Australia’s *The National Framework for Action on Dementia* (2006) and Netherlands’ *National Dementia Plan* (Ministry of Health, 2004)’.

## Assumptions made within the guidance document

Within this document, a number of assumptions are made throughout. These include the need for effective partnership between all agencies involved in the care of people with intellectual disabilities and dementia as highlighted in reports cited in the previous section. The working party recognises that each area will be configured to meet local need, but effective care can only be provided when there is good partnership working within health services – between intellectual disabilities and older people’s services, and across statutory, private and voluntary agencies.

Regardless of how each service is configured, the working party has assumed that certain principles and ways of working are already integral to the delivery of services for people with intellectual disabilities, and that these will also be available to people with intellectual disabilities who develop dementia. It has been assumed that services already deliver care in line with *Valuing People* (DH, 2001) ensuring that everyone who wants one has a Person Centred Plan and a Health Action Plan, as well as an individualised care plan. It has also been assumed that services will be delivered in line with both their relevant mental capacity legislation, the Human Rights Act and relevant National Standards for dementia care (e.g. NICE, 2006, 2010, 2013).

## Section 2 – Epidemiology

### Key points:

- People with intellectual disabilities have a higher risk of developing dementia compared to the general population, with a significantly increased risk for people with Down's syndrome and at a much earlier age.
- Life expectancy of people with Down's syndrome has increased significantly and the number of older people with Down's syndrome has been increasing.
- The incidence and prevalence of Down's syndrome is relatively stable.

## Section 3 – Baseline assessment and monitoring

### Key points:

- It is recommended to assess every adult with Down's syndrome by the age of 30 to establish a baseline against which to compare future suspected changes in functioning.
- It is worth considering screening all adults with Down's syndrome over 40 regularly because of the increased risk of dementia and the prevalence of undetected but treatable illnesses. This should link to the person's health action plan.
- Once a person is suspected of having dementia, a programme of regular monitoring and reassessment needs to be established.

## Section 4 – Possible reasons for apparent decline in functioning in adults with intellectual disability

### Key points:

- The common differential diagnoses for individuals with Down's syndrome presenting with loss of skills are depressive illness, sensory impairments (hearing or visual), hypothyroidism, obstructive sleep apnoea and dementia.
- Sometimes conditions can co-exist.
- Do not forget about the following causes of apparent functional decline as these are often missed:
  - iatrogenic causes of cognitive impairment particularly when the individual is taking multiple medications;
  - impact of the environment, particularly in relation to occupational deprivation and under stimulation; and
  - impact of abuse on the individuals with ID.

## Section 5 – Clinical presentation of dementia

### Key Points:

- The course of dementia in people with Down's syndrome has been well studied, and may be atypical with early development of behavioural or personality change, though memory problems are also prominent.
- New onset of epilepsy or worsening of existing epilepsy in an older person with Down's syndrome should always raise the possibility of Alzheimer's disease.

- Diagnostic criteria are reliable in the intellectual disability population, but dementia is more difficult to diagnose in those with severe disabilities or comorbid problems and may require sequential assessment. Clinicians should also keep in mind the slightly atypical presentation of dementia in people with Down's syndrome
- A small number of young people with Down's syndrome seem to present with decline in their teens or early twenties, often with no clear aetiology.
- People with intellectual disabilities without Down's syndrome who develop dementia may have the same range of pathologies as the general population.

## Section 6 – Assessment

### Key points:

- Carrying out investigations may be difficult in some individuals with intellectual disabilities. In this situation, clinicians have to make a decision using information from the history, physical assessment and direct observation as to whether investigations are essential or not. There may be scope to work with the person to improve the chances of success in engaging with physical assessments (e.g. systematic desensitisation for blood tests, brain scans, eye-tests, etc.).
- Mental Capacity Act (2005) and its *Code of Practice* guidelines must be followed where investigation is considered and the person is unable to consent and cooperate.

A *minimum reactive dementia assessment should include/consider:*

- Physical health check.
- Longer term and recent life events.
- Semi-structured interview exploring negative change in relation to functional skills, memory, behaviour, orientation, mood and consider potential alternatives for these changes.
- Mental state examination (may utilise a depression or anxiety measure).
- Objective assessment of cognitive and adaptive skills and abilities.

*Neuropsychological assessment:*

- Multi-disciplinary assessment is important.
- Assessments should include direct assessment of the person together with preferably, multiple-informant based questionnaire/assessments.
- Staff undertaking psychometric assessment as a part of the assessment process need to be suitably trained in psychometrics and receive appropriate supervision.
- Assessment for other co-morbid conditions is essential.
- Consideration should be given to practical issues in assessment (e.g. location).
- Consider test re-test issues (e.g. different informants/testers).

*Neuroimaging:*

- Neuro-imaging (CT/MRI) is not an essential investigation for the diagnosis of dementia in Down's syndrome.
- It may be of value where other brain lesions/vascular dementia are suspected.
- New generation CT scanners are as sensitive as MRI scanners and may be more acceptable for people with intellectual disabilities.

- Amyloid PET scans may become more available in the near future, but its value in the diagnosis of dementia in individuals with Down's syndrome who all have amyloid deposits in their brains remain uncertain

## **Section 7 – Establishing the diagnosis and breaking the news**

### **Key points:**

- People with intellectual disabilities, their families and carers need to be given opportunities to understand the nature of the intellectual disability and information about any associated health risks from an early point in their life and particularly from transition to adulthood onwards.
- People with intellectual disabilities need to be told about their diagnosis of dementia and given ongoing opportunities to understand their diagnosis and their experience of dementia.
- Family members and carers need to be informed about the diagnosis and involved as much as possible in support and management plans and, as appropriate, be given opportunities for education and training.
- The person's peers and friends are also important people to involve in giving information about the diagnosis and this will both help them cope and help them support the person affected by dementia.
- People with intellectual disabilities and their families and carers may need psychological interventions to enable them to feel emotionally supported and to begin to understand the diagnosis.

## **Section 8 – Additional health co-morbidities associated with dementia**

### **Key points:**

- The occurrence of seizures for the first time is very common in people with Down's syndrome who have developed dementia.
- The use of a single antiepileptic medication should be encouraged.
- Careful monitoring is required of seizure patterns and side effects of medication.
- Pain recognition and management for people with intellectual disabilities and dementia is often very poor.
- Diagnostic overshadowing is a frequent occurrence, and staff are often unaware of the range of painful health conditions that may present with increasing age.
- There are tools available to help staff and carers identify pain in people with intellectual disabilities.
- Sleep difficulties are commonly experienced in people with intellectual disabilities as they get older and/or develop dementia.
- Assessment should include ruling out co-morbid mental health problems, substance misuse, physical health problems and poor sleep hygiene.
- The management of sleep difficulties in dementia should be based on non-pharmacological approaches including good practice of sleep hygiene.
- Medication should only be used if other approaches have failed or risks are significant.

- NICE recommendations should be adhered to if pharmacological approaches are used.
- All people who have been diagnosed with dementia warrant a dysphagia assessment and should have eating and drinking guidelines in place.
- Eating and drinking guidelines should include the opportunity for ‘finger foods’ in order to allow the individual to maintain their ability to feed themselves.
- Nasogastric and PEG feeding are not associated with significantly improved outcomes and should be carefully considered.
- People with dementia have a high rate of infection and so any change in presentation should include an assessment for an infection.
- Infections and the causes for these need to be addressed and, where possible, changes should be made to prevent recurrence.
- Where possible, steps should be taken to prevent infections rather than to treat each as they arise.

## Section 9 – Conceptual understanding of the dementia process

### Key Points:

- The social model gives a conceptual model for staff and carers to understand dementia.
- Understanding what happens as dementia progresses and its consequences gives a framework to understand and respond appropriately.
- Think about what the person is actually experiencing, and use that to inform the care and support that is required.

## Section 10 – Philosophy of care

### Key Points:

- Staff and carers need to ensure that people with intellectual disabilities and dementia have stress-free, failure-free and consistent care.
- This approach leads to a more supportive environment and lower levels of behaviour problems and distress.
- Services need to review regularly the support given to people with intellectual disabilities and dementia, particularly the amount of staff support provided both during the day and through the night.

## Section 11 – Environments

### Key Points:

- Wherever possible, people with intellectual disabilities and dementia should remain in familiar environments.
- If the person has to move, then this should be within an intellectual disabilities service.
- Environments can be adapted to make them dementia friendly and enable the person with dementia to make sense of where they are living.
- It is important that environments provide stimulation for the person without putting them under any stress.



## Section 12 – Meeting changing needs/interventions

### Key Points:

- It is important to utilise the skills of the multi-disciplinary team to support the individual with intellectual disabilities and dementia and their network of support.
- A range of interventions should be available to meet the person's need as their dementia progresses.
- Other agencies as well as those in the statutory sector can help to support the person and their carers.

## Section 13 – Medication

### Key Points:

- Acetylcholinesterase inhibitors are used mainly in Alzheimer's type dementia. NICE guidance (2013) recommends their use in mild to moderate dementia to delay the progress of the illness. NICE has, however, restated that the difficulty in staging dementia in people with intellectual disabilities should not disadvantage them, thus introducing some flexibility for prescribers.
- Psychotropic medications have only a limited role in the management of neuropsychiatric symptoms in people with intellectual disabilities and dementia and should only be considered if other environmental/psychosocial approaches have produced only very limited or no benefit and the risk from the symptoms is assessed as high.
- Antidepressant medications are useful in the management of depressive symptoms in people with dementia and intellectual disabilities.
- Caution should be exercised in the use of antipsychotic medication in the context of the evidence of a high risk for cerebrovascular events and mortality.
- Where psychotropic medications are used, target symptoms should be clearly recorded; risks and benefits discussed with the person and/or carers and the minimum effective dose should be used for the shortest length of time.

## Section 14 – Safe eating and drinking

### Key points:

- Eating and drinking are basic human functions.
- It is vital that the person with dementia is assisted to make eating and drinking as enjoyable and stress free as possible.
- All eating and drinking difficulties must be taken extremely seriously and an urgent referral to a speech and language therapist.

## Section 15 – Palliative care and end of life issues

### Key points:

- Preparation for palliative and end of life care should be ongoing as the dementia progresses from diagnosis onwards and embedded in personalised plans for care and support at every stage.
- People with intellectual disabilities must be able to have the same end-of-life care planning and access the same palliative care services as everyone else.
- All care should be provided in accordance with the provisions of the Mental Capacity Act. Care includes medical care, physical care and psychological and emotional care of the person, and psychological and emotional care of family, friends and peers, and carers and staff, and this needs to be delivered with cultural sensitivity and where appropriate, religious support.
- Partnership working and close collaboration between professionals in intellectual disability and other health services, particularly palliative care, is very important in terms of ensuring appropriate access and timing to specialist support and appropriate symptom management. A mutual understanding of philosophies of care in these services needs to be developed.

## Section 16 – Capable commissioning for people with intellectual disabilities and dementia

### Key points:

- Each area should develop a dementia strategy and integrated care pathway to support the provision of high quality assessment, intervention and care for people with intellectual disabilities who develop dementia.

## Section 17 – Capable support

### Key points:

- Staff are key to ensuring that people with intellectual disabilities and dementia can ‘live well’ with the disease.
- Training must include developing a shared vision on which to build practice – this can only be achieved by training the whole staff team together
- Training is not a ‘one-off’ activity, but should be delivered on an ongoing basis as the person’s dementia progresses to ensure that staff are capable of meeting the person’s changing needs and achieving the best outcomes for the person.
- Capable dementia services need to be aware of, and meet the needs of, the person with dementia, those of staff and carers and other people with intellectual disabilities who live with the person, including those from BME communities.

## Section 18 – Outcomes

### Key points:

- Each area should use the Self-Assessment Checklist to measure the outcome of their services for people with intellectual disabilities and dementia, and to assist in the development of a local strategy document.
- Individual quality outcomes for each person with intellectual disabilities and dementia can be measured using the Quality Outcome Measure for Individuals with Dementia (QOMID).

## Section 19 – Future directions and research

### Recommendations:

- Staff in services seeing adults with intellectual disabilities and, specifically adults with Down's syndrome, where the diagnosis of dementia is being considered, should ensure that when a diagnosis of dementia is made the person concerned and those who support them are made aware of research projects being undertaken and permission requested to pass on their details to any approved and relevant research project.
- Services have the means to retrospectively identify any person with intellectual disabilities diagnosed as having dementia and specifically people with Down's syndrome either in the age at risk for dementia or with a diagnosis of dementia and be willing to be a point of contact with them if approved and appropriate research is being undertaken that is looking for potential participants.
- In specialist memory clinics and in services for adults with intellectual disabilities the use of standardised diagnostic and neuropsychological assessments for the diagnosis and monitoring of dementia, as it affects people with intellectual disabilities, is encouraged. This will ensure that comparison can be made across services and over time thereby providing consistent and reliable data on prevalence and incidence of dementia in this population and also enabling recruitment into future trials of any new intervention or treatment.