The diagnosis and treatment of dementia in people with learning disabilities

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Learning Disabilities Services

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Most common form of dementia in people with DS is Alzheimer’s disease. It is known that the brain pathology of Alzheimer’s disease is almost universally found in later life in people with Down’s syndrome.

Vascular disease, and the risk of vascular dementia is rare.

Dementia in people with DS may present atypically with changes in behaviour and/or personality that can precede the full clinical picture of dementia by some years.

Dementia in people with Down’s syndrome may be associated with the onset of seizures for the first time in that person’s life (78 – 98%).
Atypical presentations in people with DS

- Anecdotal reports of people with DS in teens or early adult who deteriorate, often after a life event, and never recover, but plateau.

- Superficially resembles dementia or depressive illness but it neither seems to progress (as would be expected with dementia) or resolve (as would be expected with depressive illness with appropriate treatment).

- The clinical picture is of the development of a general slowness in mental and/or physical activity, apparent loss of interest in previous activities, and a lowered level of functioning.

- Unclear how such problems should be best conceptualised.
Current Prevalence and Survival

London Consortium study

- Median age of onset = 55.4 years
- 50% of those who develop dementia are diagnosed in their 50’s
- Median survival time = 4.1 years (95% CI: 3.6, 4.6)
- Age at diagnosis highly predictive of survival time
- Gender is not predictive

<table>
<thead>
<tr>
<th>Age of diagnosis</th>
<th>Median (95% CI)</th>
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<tbody>
<tr>
<td>Under 50</td>
<td>5.6 (2.7, 8.4)</td>
</tr>
<tr>
<td>50-59</td>
<td>4.4 (3.8, 5.0)</td>
</tr>
<tr>
<td>60+</td>
<td>2.6 (1.5, 3.7)</td>
</tr>
<tr>
<td>All</td>
<td>4.1 (3.6, 4.6)</td>
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</tbody>
</table>

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Comparison of dementia prevalence rates by age

- General population
- Intellectual disabilities
- Down's syndrome
There is more likely to be a range of pathologies resulting in dementia, as is the case in the general population, and for the same reason differences in presentation and the course of dementia.

The presentation and course of dementia among people with mild learning disabilities is likely to be similar to that which is observed in the general population.

The presentation and course of dementia in people with more severe learning disabilities may initially be atypical and present with changes in behaviour and for this reason dementia may not be suspected. As the illness progresses careful questioning of staff and carers may identify evidence of developing memory and functional impairments.
Issues in establishing premorbid functioning

Establishing pre-morbid skills, abilities and personality can be challenging in the intellectual disability population due to:

• variance in cognitive functioning and abilities,
• frequent poor record keeping from childhood
• the possible lack of consistent involvement of family or staff throughout the person’s lifespan.
What should an assessment consist of?

- Consent
- Physical investigations
- Direct testing with person
- Informant interviews
  - Health checklist
  - History gathering
  - Adaptive functioning
  - Social / Environmental issues
Physical investigations

Recommended routine investigations are:

- Full blood count.
- Urea & electrolytes.
- Blood sugar.
- Thyroid function tests.
- Liver function tests.
- B12 and Folate level.
- Lipid profile.
- Sensory screening – vision and hearing.

Optional tests are:

- Electro Encephalograph (EEG)
- Neuro-imaging
- Electro Cardiograph (ECG)
Neuroimaging

- The most consistent structural change in the early stage of Alzheimer’s disease is the atrophy of the medial temporal lobe.
- People with Down’s syndrome have medial temporal lobe atrophy even without dementia.
- Normative values have not yet been established, so neuro-imaging currently has limited value in the early diagnosis of Alzheimer’s disease in people with Down’s syndrome.
- Used mainly to rule out structural lesions other than atrophy (e.g. space occupying lesions). It should therefore be used only when the clinical picture suggests the possibility of such lesions.
- Some people with intellectual disabilities may be able to go through the procedure without any other interventions, others may benefit from the use of one dose of oral anxiolytic medication such as lorazepam or diazepam an hour beforehand, or buccal midazolam, which provides rapid and short-term sedation and therefore may be given immediately before the procedure.
Direct testing

- Assessment tools for dementia in the general population are not appropriate for people with intellectual disabilities.
- Currently no agreed battery of assessments and great variation in screening/assessment methods.
- Lack of research data to secure agreement in order for there to be better uniformity across services and for future data to be pooled and compared.
- The nature of the intellectual disability population requires each individual to be viewed independently in terms of their own functioning.
Minimum direct testing with the person

- A validated instrument for the cognitive assessment of dementia in people with intellectual disabilities e.g. NAID, CAMCOG-DS, SIB
- Prospective, short and long term memory (visual and verbal).
- Executive functioning.
- Orientation.
- Language (expressive and comprehension).
- Recording of evidence of new learning.
NAID (was Oliver & Crayton battery (1993, 1997)

Subtests
- Picture Naming
- Picture Identification
- Orientation
- Card sorting
- Action on Request
- Object Memory
- Picture Memory
- Memory for Sentences

Advantages
- Good depth – to see change over time
- Can administer some or all if needed
- Takes about 45 minutes
- Enjoyable for person
- Easy to graph and follow change over time
Card Sorting

- Sep-96: 117
- Mar-98: 106
- Nov-12: 127
- Jun-13: 144
- Apr-14: 163

Action on Request

<table>
<thead>
<tr>
<th></th>
<th>1st Prompt</th>
<th>2nd Prompt</th>
<th>Modelled</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sep-96</td>
<td>4</td>
<td>1</td>
<td>5</td>
<td>19</td>
</tr>
<tr>
<td>Mar-98</td>
<td>4</td>
<td>0</td>
<td>2</td>
<td>14</td>
</tr>
<tr>
<td>Nov-12</td>
<td>5</td>
<td>2</td>
<td>1</td>
<td>20</td>
</tr>
<tr>
<td>Jun-13</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>13</td>
</tr>
<tr>
<td>Apr-14</td>
<td>0</td>
<td>3</td>
<td>4</td>
<td>10</td>
</tr>
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More able people

- If people are at ceiling at baseline – consider additional direct testing.

- Use parts of CAMCOG-DS
  - Verbal fluency
  - Copying and drawing – circle, square, 3D house, clock
  - Counting to 20
  - Digit span
  - Draw a person
  - Write your name
People with more severe LD / additional issues

- Try parts of NAID – use any subtests that they can do.
- Visual impairment – can still do Actions on Request; Memory for Sentences; Orientation.
- Hearing impairment – depends on severity and their communication – try Picture Naming; Card Sort – use Makaton / signing
- More severe LD – depends on severity – will always be some people who cannot do any direct testing – but small number. If so need to have really good informant assessments
- Adapt – but record exactly what you did – so can repeat next time. Remember that each person is their own control.
Executive functioning

- Increasing importance of assessing executive functioning
- No agreement on what tests to try, but several under development / trial
- Can use individual tests e.g. verbal/category fluency, response inhibition tests and set/rule switching tasks such as card sorting tests
- Informant scales may be useful
New executive functioning tests

BADS-ID – adapted version of the BADS

- Simplified rule shift cards
- Simplified key search
- Modified 4 elements
- Supermarket map
Supermarket map

NAME:_________________________________________  AGE:_________________

NAME OF TESTER:________________________________  DATE:_________________

IN

OUT
Measure of everyday planning

The MEP comprises a grid of related but independent areas of functioning:

- **Initiation (volition):** the ability to recognise the need for action and to commit to taking that action
- **Planning / sequencing:** identifying and ordering the steps necessary for that action
- **Memory:** to remember that action needs to be taken and being able to hold the task and steps in mind long enough to complete the task
- **Attention & Concentration:** being able to focus on the task and steps sufficiently to see it through
<table>
<thead>
<tr>
<th>Task</th>
<th>Initiation</th>
<th>Planning/Sequencing</th>
<th>Memory</th>
<th>Attention/Concentration</th>
</tr>
</thead>
<tbody>
<tr>
<td>You’re at home and you feel thirsty, what do you do?</td>
<td>0 Do nothing.</td>
<td>0 Not able to plan / sequence steps required to make a drink</td>
<td>0 Can’t remember they are feeling thirsty long enough to take any action</td>
<td>0 Makes no attempt to get a drink.</td>
</tr>
<tr>
<td>1 Accepts offer of a drink when given by staff / carer e.g. at certain times of day only</td>
<td>1 Able to assemble some or all of the necessary items but requires prompting to get steps in the right order</td>
<td>1 May move to get a drink but will forget why they have moved, e.g. forgotten why they have walked to the kitchen.</td>
<td>1 Will take the first step to get a drink but will become distracted and not complete.</td>
<td></td>
</tr>
<tr>
<td>2 Will ask for a drink when feeling thirsty or will move to get drink if prompted.</td>
<td>2 Able to assemble the necessary items and complete most steps, but would miss out or mix up some steps if not prompted e.g. boiling kettle</td>
<td>2 Able to remember the first steps needed to make a drink but not able to complete / may repeat earlier steps</td>
<td>2 Able to complete several steps of making a drink before getting distracted by something else.</td>
<td></td>
</tr>
<tr>
<td>3 Is able to fetch a drink made by someone else</td>
<td>3 Able to assemble items and make a drink successfully given no problems e.g. all materials available</td>
<td>3 Able to remember the steps to successfully make a drink according to plan</td>
<td>3 Able to make a drink when there are no major distractions</td>
<td></td>
</tr>
<tr>
<td>4 Gets up and goes to suitable area e.g. kitchen and starts making drink whenever thirsty</td>
<td>4 Able to assemble items and make drink successfully, even when faced with a problem e.g. kettle not working</td>
<td>4 Able to successfully remember task and hold in mind long enough to solve a problem e.g. no milk for tea so what can I drink instead</td>
<td>4 Able to make a drink independently despite major distractions</td>
<td></td>
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Remarks: For a better life
Informant interviews

History and Information gathering

These should aim to cover those areas of function that are known to deteriorate with the development of dementia including:

- short and long term memory,
- general mental functioning,
- dyspraxia and dysphasia,
- daily living skills
- personality and behaviour

e.g. DLD, DSDS, DSQIID, ABDQ – or make your own

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Other measures

- Psychological / mental health e.g. PAS-ADD, Anxiety / depression
- Carer burden
- Life events
- Adaptive functioning e.g. ABAS-II, HALO, Vineland, ABS – important not just about quantitative change but qualitative change
Environmental assessment

- Quality of the person’s physical environment.
- Staffing levels (day and night).
- The mix of people with intellectual disabilities in the residential and day care settings.
- Quality and quantity of day activities.
- Staff characteristics: attitudes and competence, including consistency of approach.
- Scrutiny/review of historic/current support package.
Conditions that Mimic Dementia

- Physical Health issues
  - Uncontrolled Epilepsy
  - Nutrition – vitamin B12 & Folate
  - Electrolyte abnormalities
  - Hypothyroidism

- Sensory Impairments
  - Visual Impairments (recognition, lost skills)
  - Hearing Impairments (misunderstanding)

- Mental Health Problems
  - Depressive illness (pseudo-dementia)
  - Psychotic disorder (decline in function)
  - Chronic Anxiety (catatonia)

- Medication
  - Anticholinergics, psychotropics, AED’s, Pain meds, antihistamines and benzodiazepines
Conditions that Mimic Dementia II

- **Sleep Disturbance**
  - Sleep Apnoea or other sleep disturbances

- **Life Events**
  - Loss of parents, moves, day-care changes etc

- **Impact of poor social / physical environment**
  - Loss of skills and regression

- **Abuse**
  - Loss of skills and regression

- **Acute Organic Brain Syndrome**
  - Confusional state secondary to pain, chronic infection or head injury
Sharing the diagnosis

» Establishing the diagnosis is often a complex process

» 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 in a way that they can understand.

» 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.
Breaking the news to the person

- Generic breaking-bad-news models do not meet the needs of people with intellectual disabilities so we need to communicate information over time and based on “chunks”.

- A Person-centred approach aims to support the person to understand and cope with their changing experiences.

- We want to help them be involved as much as possible in decisions about their support and care and medical treatments, including future care.

- Specific resources: The Journey of Life and About Dementia; What is dementia.
Possible Interventions

- Philosophy of care
- Anxiety and stress reduction
- Life story work
- Reminiscence
- Reality orientation
- Validation
- Understanding behaviours
- Medication
- Dementia Friendly environments

For a better life
Establish a philosophy of care

Excellence in dementia care requires staff and family carers to:

» understand and know the person
» understand dementia and its consequences for the person
» and to consequently be able to think ahead and predict ‘stressors’.
They need to adapt their approach to ensure that the person with dementia has:

- **stress free**
- **failure free**
- **individualised care**
- **that is consistent**
- **without time pressures.**

needs to be incorporated into the person’s person centred plan.

can only be achieved by having a thorough knowledge and understanding of the person and their history. This can be aided by the ongoing use of Life Story work.

Need to maintain all the elements of normal daily living to retain skills for as long as possible.

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Positive and Proactive Care

Positive and Proactive Care is a framework for working with people with difficulties in a positive and proactive approach. It may also be called Positive Behavioural Support when working with people with behaviour that challenges.

Principles apply to people with dementia.

Emphasises person centred values, aims to enhance community presence, increase personal skills and competence and ensure dignity and respect is maintained for the person being supported.

When a diagnosis of dementia is suspected a shift in value from increasing skills to maintaining skills can be helpful in terms of reducing stress of the individual.
Anxiety and stress reduction

As people develop dementia, one of the first symptoms often seen, caused by the effects of disturbed encoding, is anxiety.

This can manifest itself in many ways e.g. The person may become very anxious about leaving familiar environments. The person may not remember where they are going or why, they may resist putting on their coat, or demand to return to their familiar setting.

Staff and family carers need to firstly recognise why the person has become stressed and anxious, and look at solutions to help reduce the person’s level of anxiety.

This can be achieved by a variety of techniques including:
- reassurance and verbal reminders,
- the use of visual aids to remind the person where they are going,
- the use of relaxation techniques that are already familiar to the person e.g. aromatherapy (Harrison & Ruddle, 1995), breathing exercises and imagery and visualisation exercises.
Life story work

Kerr & Wilkinson (2005) said of people with dementia that ‘if you do not know their past then you cannot understand their present’

- Need a thorough knowledge and understanding of the person and their history.
- Helps to maintain people’s sense of self esteem and identity.
  The process of talking to people about their memories, collecting objects and pictures are the important aspects, rather than the final end product.
- Engaging family members and friends in remembering and producing memories, anecdotes, photos and possessions for the life story is a key and beneficial aspect of the work.
- Use regularly to help reduce anxiety and give the person and staff a sense of the person and who they are
- The Life Story can help staff to understand what the person is referring to when they remember things from their ‘rollback memory’, and this helps them to respond more sensitively.
- For the person with a learning disability, a lifestory book would remind any new carers that behind the debilitating illness is a person who enjoyed certain things in life.

For a better life
Reminiscence

Staff and carers need to remember that people compensate for their deterioration in functioning by making greater use of remaining abilities (e.g. earlier memories).

This may mean that the person finds comfort in activities and objects from their childhood.

Reminiscence work can help the person with learning disabilities and dementia to find anchors with their past and to help steady and engage the person (Kerr, 2007).

Reminiscence Therapy is one of the most popular psychosocial interventions in dementia care with the general population, and is highly rated by staff and participants (Woods et al, 2006).

There is less written about its use with people with learning disabilities (Puyenbroeck & Maes, 2002; Gibson, 2006), but anecdotally has become valuable in its use with people with learning disabilities and dementia.
Reality orientation v validation

- In early stage dementia, reality orientation clearly has its place when people are ‘nearly oriented’.
- The use of cues, gentle reminders, photos and pictures can help the person to engage with the world around them, but this must be achieved in a meaningful and stress-free manner.
- Reality orientation needs to be used in a humanistic, sensitive and individualised manner. The home must be one in which it is worth being oriented e.g. diary of days events, photograph/ picture on bedroom door, arranging clothes in correct order for dressing.
- Reality orientation should enable people to retain control where possible, to reduce stress and facilitate their hold on reality, if that is in their best interests.
As the dementia progresses, the person with dementia does not remember events that have happened in the past as their memory ‘rolls back’ to an earlier time.

This frequently results in the person asking for people who either are no longer alive, or part of their lives.

Some staff and professionals believe that we should not ‘collude with false reality’ and that we should always tell people the truth, regardless of how painful that is for the person.

**e.g.** the person with dementia asks staff repeatedly when their mother is coming, even though in reality she has died.

- To remind the person of the truth reminds them of their loss.
- The person will forget what they have been told, and will ask again therefore repeatedly experiencing feelings of loss.
- Staff need to consider what has been gained for the person by repeatedly reminding them of their loss.

Kerr (2007) who writes that ‘To tell someone with dementia the truth when it is going to cause immense stress and grief is cruel and unacceptable’.

Barbara Pointon recommends that if the person is unable to enter our world, then we must enter their and affirm it.
Medications

- Acetylcholinesterase inhibitors are used mainly in Alzheimer’s type dementia.

- Although there is no conclusive evidence that these drugs are effective in people with learning disabilities, the available evidence suggests that they may improve the quality of life for the person and their carers.

- Antidepressant medications are useful in the management of depressive illness in people with dementia and learning 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.

For a better life
Behaviour that challenges

Physical Health

Mental Health

Social Environment

Physical Environment

Get to know the person

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Understanding behaviours

The problem behaviour may be transitory to the current stage of the person’s dementia and not need an intervention. Similarly, it may be possible to withdraw interventions if/when the person’s skills and behaviour change again.

The situation should be viewed through the eyes of the person with dementia, i.e. their current reality.

Continual correction by staff/carers of a false reality (e.g. person with dementia asking when a dead parent will visit) will not reduce their immediate confusion and distress.
The behaviour should be viewed as an attempt by the person to communicate or to make sense of a bewildering environment (e.g. GP practice waiting room is confused with an airport lounge).

The behaviour may be an exacerbation or return of previous behaviours. If the person had difficult behaviours/personality traits previously, these may return/re-occur with roll back memory.

The behaviour may be caused by a return to a long term memory that is now inappropriate, e.g. wanting to walk home from their day service.

There may be an underlying neurological change, e.g. taste bud changes leading to food fads and a liking for stronger flavours, refusing baths resulting from problems with depth perception and/or stepping into the bath.
Managing behaviours

» Simple and practical solutions may work, e.g. a net to catch items thrown out of the window.

» Behaviours may reduce by reducing anxiety and stress. Introduce relaxation, avoid conflict and confrontation. Ask yourself: ‘Does it matter?’

» Simple environmental alterations may alter the behaviour e.g. covering a mirror.

» Use usual methods of observing, monitoring, and understanding the function of the behaviour to decide on the best intervention, alongside thorough risk assessment and crisis management plans.
Most environments where people with learning disabilities live are not dementia enabled.

Adaptations will need to be made to ensure that the environment does not add more stress to the person with dementia. Many of the adaptations required are not too expensive but can have a very positive effect on the quality of life for the person with dementia.

Environments may need to be altered to be:

- **Calm** – noise (internal and external), colour
- **Predictable and making sense** – cues, signposting, no surprises
- **Familiar** – homely, long term memory
- **Suitably stimulating** – noise, views
- **Safe and risk assessed** – access, stairs, hazards
Possible solutions

- Problems are created by changing floor colourings, steps and stairs, patterned carpets, mirrors, lights that cause shadows, modern fixtures and fittings that are not within the person’s long term memory.

- Many of the problems can be solved by the use of appropriate colour e.g. red toilet seats, painting toilet doors red to aid orientation, or cues.

- As the dementia progresses, staff need to be aware of safety issues, especially in relation to access to the kitchen, roads, hot water and electrical appliances.

- Creative solutions can cost very little, but make a huge difference to the person.

- Care needs to be taken when refurbishing a home for a person with dementia to ensure that it is equipped with furnishings and equipment that are familiar to the person and their roll back memory.
Questions and Discussion?
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