Dementia and people with Intellectual Disability

Dr Owen Doody
Dementia

• Alzheimer’s is the most common type of dementia with Lewy Body being the second most common type in ID
• Approximately 13% of individuals with ID will develop dementia under the age of 60
• Up to 18% over the age of 60 with ID will develop dementia
• Almost all individuals with Down’s syndrome will develop dementia
• Once diagnosed with dementia, may live anywhere from 3-10 years on average.
Misconceptions

• “It’s a symptom of normal aging”
  • As the disease gradually worsens, it takes away someone’s ability to think, eat, talk, etc.

• “It is a old person disease”
  • About 5% of people with the disease get symptoms in their 30s, 40s, or 50s

• “It won’t kill you”
  • It’s the sixth leading cause of death in the U.S. Most people live 8 to 10 years after they’re diagnosed.

• “There are treatments that stop the disease”
  • While certain treatments can help against Alzheimer's symptoms, there’s no current way to stop or slow the disease itself

• “It’s caused by aluminum, flu shots, silver fillings, or aspartame”
  • There’s no scientific evidence to support these claims
Rule out

- Vitamin B12 deficiency
- Vitamin D deficiency
- Hypothyroidism
- Medication changes
- Sleep schedule
- Depression
- Urinary tract infection or other infection causing fever
- Result of a fall
- Sudden or recent changes in schedule
Indicators

• Slowly progresses or occurs following a neurological incident
• Does not improve over time once other variables are accounted for
• Sleep-wake schedule changes
• Confusion/disorientation that does not change
• Wanders and seems to get lost

• Changes in personality
• Toileting changes (incontinence)
• Decline in daily living skills
• Lack of energy
• Difficulty ambulating
• Being uncooperative
Assessment

• Number of measurers to identify
Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)

- 53 items and requires 15 minutes to administer
- Require a reliable informant
- Assesses speech, daily living skills, and needed accommodations
- Rate each question
- Used qualitatively
- Pros-takes into account premorbid ability
- Con-normed primarily on individuals with Down’s syndrome
- Good sensitivity and specificity (e.g. rules out those with dementia and identifies those with dementia)
Dementia Questionnaire for Mentally Retarded Persons (DMR)

• 50 items and 8 subscales
• Used for adults with and without Down’s syndrome
• High degree of false positives (38.5%)
• May underdiagnose vascular dementia
Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE)

• 16 items; easy to administer and used qualitatively
• Shortcomings: requires informant to know individual 10 years prior; does not account for comorbid symptoms; inconsistent findings in research
• http://m.patient.media/pdf/9309.pdf
Early Signs of Dementia Checklist (ESDC)

• 37 total items

• Shortcomings: little research on the measure; does not account for premorbid functioning; questions can be misleading.
Additional tests

• Test for Severe Impairment (TSI)
• 24 items assessing memory, knowledge, and language
• Good reliability and validity

• Severe Impairment Battery (SIB)
• Short form and long form
• Six major subscales: attention, orientation, language, memory, visuo-spatial ability, and construction.
• Can identify severe dementia
• Good reliability and validity
• Developed for individuals without ID
Association with pain

• Baseline recordings should be part of each individual’s annual health assessment and aspects such as pain responses can be documented and reviewed for the benefit of unfamiliar healthcare professionals (Hoghton et al., 2012; Masterson, 2011; Robertson et al., 2010).

• Importance of knowing and recording the person’s individual behaviours and recognising behavioural and emotional patterns and changes requires close collaboration with a parent or caregiver to effectively assess (Davies, 2010; Dubois et al., 2010).
Association with pain

• Important that behavioural cues are used in combination with facial expressions to evaluate pain among people with intellectual disability along with considering the developmental stage, temperament, personality, environmental factors and previous pain experiences (Bajelidze et al., 2008; Hartman et al., 2008).

• Problem behaviours that develop gradually (e.g., rumination, pica, food refusal), need to be considered as potential pain behaviours in people with ID.
Association with pain

- As pain is a highly individualised and subjective phenomenon (Koyama et al., 2005), changes in physical behavioural signs (increased or decreased noises, change in eating habits, increased crying, etc.) together with an understanding of the individual’s typical abilities are potential indicators of pain (Herr et al., 2011).

- Physical, physiological and behavioural observations and atypical reactions recordings are essential if pain is to be recognised and treatment delivered (Dubois et al., 2010; Rattaz et al., 2013).
Assessment tools

- Evaluation Scale for Pain in Cerebral Palsy—ESPCP; Giusiano et al., (1995)
- Checklist of Nonverbal Pain Indicators— CNPI, Feldt (2000)
- Pain and Discomfort Scale—PADS, Bodfish et al., (2001)
- Pain Indicator for Communicatively Impaired Children—PICIC, Stallard et al. (2002)
- Paediatric Pain Profile—PPP, Hunt et al. (2004)
- Pain and Distress Scale—PDS, Bodfish et al., (2006)
Assessment tools

- Disability Distress Assessment Tool—Dis-Dat, Regnard et al. (2007)
- Revised face, legs, activity, cry, consolability—rFLACC, Voepel-Lewis et al., Tait (2005)
- Non-Communicating Adult Pain Checklist—NCAPC, Lotan et al., (2009)
- Chronic Pain Scale for Nonverbal Adults with Intellectual Disability, CPS-NAID, Burkitt et al., (2009)
Importance

• People with intellectual disability and dementia experience the same age related painful conditions as the general ageing population (Wilkinson *et al.* 2004; 2005; Kerr *et al.* 2006; 2011; Cleary and Doody 2017a), though at a younger age.

• People with intellectual disability also tend to experience more chronic health conditions which cause pain and discomfort than the general population including gastric reflux, osteoporosis and chronic constipation (Wilkinson *et al.* 2004; 2005; Kerr *et al.* 2006; Cleary and Doody 2017a).
Importance

The association of behavioural and psychological symptoms with dementia meant that nurses interpreted behaviours as challenging and due to the persons intellectual disability and/or dementia rather than considering the possibility of pain (Wilkinson *et al.* 2004; 2005; Cleary and Doody 2017b). Poor recognition of pain can lead to the inappropriate use of antipsychotics and sedatives which can mask painful conditions and further reduce the likelihood of identifying pain (Kerr *et al.* 2006; 2011).
Planning ahead

• Caring for people with ID and dementia is stressful (Wilkinson *et al.* 2005; Kerr *et al.* 2006; Cleary and Doody 2017a, 2017b) particularly when staff do not have good knowledge of dementia or ID.

• Training in dementia must focus on how dementia presents for people with ID, the likely progress of the condition, interventions that support the person with dementia (Wilkinson *et al.* 2004; 2005; Cleary and Doody 2017b).

• Training in older person must focus on ageing in ID, conditions associated with ageing (Wilkinson *et al.* 2004; 2005; Kerr *et al.* 2006; Cleary and Doody 2017a, 2017b).
Take home message

• It’s important to have a reliable collateral informant, preferably someone who has known and worked with the individual being assessed for years

• It’s important to get a good assessment of adaptive functioning (social, daily living, communication skills) and compare it to previous estimates

• It’s important to attempt some measure of cognitive abilities (language, memory, executive functioning, etc.)

• Remember that a diagnosis of dementia often requires both decline in cognition and adaptive abilities.
Recommendations

• 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).

• 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.

• 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.
Recommendations

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

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

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

• Palliative care should be available.

• Trainees specialising in psychiatry of Intellectual Disability should have training in diagnosis and management of dementia.
Thank you

Dr Owen Doody

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