About Us

Introduction

The Department of Developmental Disability Neuropsychiatry (3DN) was established by the Chair of Intellectual Disability Mental Health in 2009. The Chair is funded by Ageing, Disability and Home Care, Department of Family and Community Services NSW.
Acknowledgements/Declarations

Funding: Core
• Ageing Disability and Home Care | Family and Community Services NSW
• UNSW Medicine

Funding: Research and Projects
• NSW Ministry of Health & Related Organisations
  – MHDAO, MH Kids, HETI, ACI ID Network
• Australian Government Department of Health and Ageing
• Australian Research Council (ARC)
• National Health and Medical Research Council (NHMRC)
• NSW Institute of Psychiatry
• Autism CRC
Disclosures
Our Motivation

Please support us as we turn ugly and get moving for a good cause. Compared to men in the general population, men with intellectual or developmental disabilities:
- experience big barriers to accessing good health care
- have a higher risk of mental health problems
- are at risk of earlier death from preventable causes

Team 3DN want to raise awareness of these issues. For more information see http://3dn.unsw.edu.au/
Session 1
Intellectual Disability, Health and Ageing

ID and its causes
Health and ID
Mental Health and ID
Syndrome specific health issues
Ageing and ID
What is Intellectual Disability?

• Disorder with onset in the developmental period
  – Deficits in intellectual functions (Below average intelligence, IQ of <70, ie <2 SD below mean)
  – deficits in adaptive behaviours
  – onset before the age of 18

• Intellectual Developmental Disorder

• About 1.8% of the population
• About 400,000 Australians
• About 125,000,000 individuals worldwide
# Some Causes of Intellectual Disability

<table>
<thead>
<tr>
<th>Prenatal</th>
<th>Perinatal</th>
<th>Postnatal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chromosomal disorders</td>
<td>Intrauterine: placental insufficiency; prematurity; obstetric trauma</td>
<td>Head injury</td>
</tr>
<tr>
<td>Syndrome disorders</td>
<td>Neonatal: intracranial haemorrhage; respiratory distress; head trauma; kernicterus</td>
<td>Infections &amp; post-infectious</td>
</tr>
<tr>
<td>Inborn errors of metabolism</td>
<td></td>
<td>Degenerative disorders</td>
</tr>
<tr>
<td>Developmental brain abnormalities</td>
<td></td>
<td>Seizure disorders</td>
</tr>
<tr>
<td>Environmental factors</td>
<td></td>
<td>Toxic metabolic disorders eg lead poisoning</td>
</tr>
<tr>
<td>eg maternal malnutrition; placental insufficiency; fetal alcohol syndrome; varicella infection; irradiation</td>
<td></td>
<td>Malnutrition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Environmental deprivation</td>
</tr>
</tbody>
</table>
Classification

- Profound (IQ < 20/25) Affects 1-2%
- Moderate (IQ 35/40 – 50/55) Affects ~10%
- Severe (IQ 20/25 – 35/40) Affects 3-4%
- Mild (IQ 50/55 – 70/75) Affects ~85%

• Continuous, intensive assistance
  - communication, personal care, accessing services & facilities
• Expressive communication usually reliant on gestures, facial expression and body language
• Importance of objects & visual aids in communication
• Importance of relationships – recognise and form close bonds with key people

Historical Changes & Impacts

• Benefits of deinstitutionalisation
  – Social inclusion
  – Enhanced opportunities

• Other impacts of deinstitutionalisation
  – Historical separation of health and disability services
  – Erosion of expertise within health sector
  – Loss of educational and professional focus

• Subsequent service development
  – Enhanced behavioural support within disability services
  – Limited development of health services and policy
Impact of ID – Behaviour

• Higher rates of behavioural problems
• Relationship to communication difficulties
  – Behavioural change can be:
    • A communication of needs
    • A response to environmental and situational factors
    • A manifestation of a medical/psychiatric condition
    • Reinforced by learning

• Behavioural phenotypes
Impact of ID – Health

• Higher morbidity\textsuperscript{2,3}
• Lower rate of detection and treatment\textsuperscript{2,3}

Health Challenges – Physical Health

- Dental disease (7x)
- Vision impairment & eye disorders (7-20x)
- Hearing impairment
- Thyroid problems
- Epilepsy
- GERD
- Osteoporosis
- Hospitalisation (2x)
- Serious injury (2x)

- Mobility problems
- Multiple chronic complex disorders
- Polypharmacy
- Lifestyle related
  - Overweight & obesity
  - Constipation
  - Physical fitness
Why are People with an ID at Higher Risk of Ill Health?

- Multiple barriers to health care:
  - Less likely to express health concern or seek help
  - Reduced insight and awareness
  - Reduced supports and self advocacy
  - Communication
  - Lack of awareness of carers

- Atypical presentations
  - Signs of health condition or mental disorder are confused with a feature of ID or a behavioural problem

- Limited specialised services for people with ID
- Limited skills in general medical workforce
- Limited access to prevention, early intervention strategies
- ID specific health needs
Mental Health of People with an ID

• People with an intellectual disability experience an over-representation of mental disorders
  – Conservative estimates for adults/children with ID 2.5/3-4 x
• At any one time, an estimated 20-40% of people with an ID will be experiencing a mental disorder of some kind.
• Access to mental health supports and treatments is limited
• High impact for people with ID, families and carers
• Complexity
• Multiple vulnerabilities
Health Challenges—Mental Health

• People with ID experience:
  – the full range of mental disorders seen in adults without ID
  – higher rates of mental disorder (~40%\textsuperscript{1} v ~20%\textsuperscript{2})
  – increased prevalence of mental disorder with increasing disability\textsuperscript{1}
  – psychopathology that varies with level of disability\textsuperscript{1}

1. Cooper et al., 2007; 2. Slade et al., 2009
Things that affect mental health

**Biological**
- Genes
- Changes in the brain
- Seizures & medications
- Chemicals/Hormones in Brain
- Physical health problems/pain

**Social**
- Relationships & Interactions
- Past events
- Living/Work/Schooling situations

**Psychological**
- Thinking style
- Coping skills
- Communication skills

- Physical Appearance
- Motor impairment
- Intellectual disability

- Sleep problems
- Intellectual Disability
- Temperament

School & other achievements; Communication & interpersonal skills
Health Challenges – eg Down Syndrome

- Visual impairment, cataracts
- Hearing impairment
- Hypothyroidism
- Epilepsy
- Congenital heart defects (40-50%)
- Atlantoaxial instability
- Skin disorders, alopecia, eczema
- Depression
- Alzheimer’s disease
- Sleep apnoea
- Increased susceptibility to infections
- Coeliac disease
- Blood dyscrasias
- Childhood leukeamia

1. Lennox & Eastgate, 2004. Adapted with permission from Nick Lennox
Health Challenges – eg Tuberous Sclerosis¹

- Retinal tumours
- Sleep problems
- Epilepsy
- Cerebral astrocytomas
- Rhabdomyomas
  - Eye
  - Bone
  - Liver
- Hypertension
- Kidney & lung hamartomas
- Polycystic kidneys
- Dental abnormalities
- Skin lesions
- Autism Spectrum disorders
- ADHD
- Anxiety Disorders

1. Lennox & Eastgate, 2004. Adapted with permission from Nick Lennox
Health Challenges – eg Fragile X Syndrome

- Visual impairment
- Hearing impairment
  - Recurrent ear infections
- Epilepsy
- Aortic dilation, Mitral Valve prolapse
- Connective tissue dysplasia
- Scoliosis
- Congenital hip dislocation
- Hernias
- Autism Spectrum disorders
- Anxiety Disorders
- Attention Deficit/Hyperactivity

1. Lennox & Eastgate, 2004. Adapted with permission from Nick Lennox
Ageing: an Important Issue for People with ID

• There is a rapid ageing of the ID population
  – does not extend to the ‘very old’ (cf general population)
  – Life expectancy varies with disorder and level of ID

• Ageing with ID carries specific health implications

• A small decline in cognition can translate to a large decline in function

• Impact on carers

• Service and cost implications
Current Status: Services and Supports

Services and supports for older people with ID are characterised by:

• Limited age-specific capacity in health and disability services
• Health and disability professionals with limited training
• Confused service models
• A growing demand for age-related services
• A growing demand for assessment of possible cognitive decline
Mortality in People with an ID

• Death as an outcome
• Health inequalities
• Preventable causes
Aims

• To compare age-standardised mortality rates (ASMRs), Comparative Mortality Ratios (CMRs) and Years of Productive Life Lost (YPLL) in people with an ID in NSW to the general NSW Population
• To determine underlying cause of death, including for preventable deaths, and compare this to the Australian population
• To determine the proportion of preventable deaths in people with an ID in NSW, Australia
Methods: Linkage and Mortality

- Linkage performed independently of researchers by CHeReL
  - De-identified linkage (SLK 581)
- Age-standardised Mortality Rates (ASMR)
  - [Number of deaths]/[total person years in that age range]
  - Adjusted to match age profile of Australian population
- Comparative Mortality Figure, (CMF_{ID/non-ID})
- Potential Years of Life Lost (PYLL)
  - number of years not lived because of death before age 75
Results: Demographics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>Number of people registered for disability services</td>
<td>73,674</td>
</tr>
<tr>
<td>Number of people with ID</td>
<td>42,204</td>
</tr>
<tr>
<td>Of those with ID:</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>23,328 (60%)</td>
</tr>
<tr>
<td>Female</td>
<td>18,876 (40%)</td>
</tr>
<tr>
<td>Median age (IQR) at Jun 2005 or at first contact the service, years</td>
<td>17 (8-36)</td>
</tr>
<tr>
<td>Number of deaths</td>
<td>953 (2%)</td>
</tr>
<tr>
<td>Number of deaths with known causes (percentage of all deaths)</td>
<td>758 (80%)</td>
</tr>
<tr>
<td>Number of male deaths</td>
<td>553 (58%)</td>
</tr>
<tr>
<td>Number of female deaths</td>
<td>400 (42%)</td>
</tr>
<tr>
<td>Median age (IQR) at death, years</td>
<td>46 (25-60)</td>
</tr>
</tbody>
</table>
Results: Age specific death rate per 1000 people for ID and Non ID
Age Standardised Mortality Rates (ASMRs) per 1000 people for ID and Non ID cohorts

<table>
<thead>
<tr>
<th></th>
<th>5-69 age groups</th>
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</thead>
<tbody>
<tr>
<td>ASMR ID</td>
<td>4.7</td>
</tr>
<tr>
<td>ASMR Non ID</td>
<td>1.8</td>
</tr>
<tr>
<td>CMF&lt;sub&gt;ID/Non ID&lt;/sub&gt;</td>
<td>2.6</td>
</tr>
</tbody>
</table>

ASMR for each cohort was calculated using direct standardised method based on Australian Standard population. CMF: Comparative Mortality Figure
Age specific death rates, Age Standardised Mortality Rates, per 1000 people, ID and Non ID

Male

Female

<table>
<thead>
<tr>
<th></th>
<th>5-69 yrs</th>
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<tbody>
<tr>
<td>ASMR ID</td>
<td>4.9</td>
<td>ASMR ID</td>
<td>4.5</td>
</tr>
<tr>
<td>ASMR Non ID</td>
<td>2.3</td>
<td>ASMR Non ID</td>
<td>1.3</td>
</tr>
<tr>
<td>CMF ID/Non ID</td>
<td>1.8</td>
<td>CMF ID/Non ID</td>
<td>3.4</td>
</tr>
</tbody>
</table>
## Leading underlying causes of death by ICD10 chapter (top 10)

<table>
<thead>
<tr>
<th>Underlying Cause of Death by ICD-10 Chapter</th>
<th>ID ABS Convention (N = 832)</th>
<th>ID Revised (N = 832)</th>
<th>Non ID (N = 312,693)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Rank</td>
<td>Frequency (%)</td>
<td>Rank</td>
</tr>
<tr>
<td>Nervous system</td>
<td>1</td>
<td>177 (21)</td>
<td>4</td>
</tr>
<tr>
<td>Neoplasms</td>
<td>2</td>
<td>123 (15)</td>
<td>3</td>
</tr>
<tr>
<td>Congenital malformations, deformations and chromosomal abnormalities</td>
<td>3</td>
<td>121 (15)</td>
<td>5</td>
</tr>
<tr>
<td>Circulatory system</td>
<td>4</td>
<td>118 (14)</td>
<td>2</td>
</tr>
<tr>
<td>Respiratory system</td>
<td>5</td>
<td>90 (11)</td>
<td>1</td>
</tr>
<tr>
<td>External causes of morbidity and mortality</td>
<td>6</td>
<td>46 (6)</td>
<td>6</td>
</tr>
<tr>
<td>Digestive system</td>
<td>7</td>
<td>37 (4)</td>
<td>7</td>
</tr>
<tr>
<td>Mental and behavioural disorders</td>
<td>8</td>
<td>35 (4)</td>
<td>8</td>
</tr>
<tr>
<td>Endocrine, nutritional and metabolic diseases</td>
<td>9</td>
<td>29 (3)</td>
<td>9</td>
</tr>
<tr>
<td>Symptoms, signs and abnormal clinical and laboratory findings not elsewhere classified</td>
<td>10</td>
<td>19 (2)</td>
<td>10</td>
</tr>
<tr>
<td>Genitourinary system</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>chromosomal abnormalities</td>
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<td></td>
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</tr>
<tr>
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<td>4</td>
<td>118 (14)</td>
<td>2</td>
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<tr>
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<td>90 (11)</td>
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<td>9</td>
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<tr>
<td>Symptoms, signs and abnormal clinical and</td>
<td>10</td>
<td>19 (2)</td>
<td>10</td>
</tr>
<tr>
<td>laboratory findings not elsewhere classified</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Genitourinary system</td>
<td>9</td>
<td>7569 (2)</td>
<td></td>
</tr>
<tr>
<td>Certain infectious and parasitic diseases</td>
<td>10</td>
<td>5395 (2)</td>
<td></td>
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</tbody>
</table>
Potentially Avoidable Deaths

- Using conventional cause of death coding: potentially avoidable deaths in PWID 26%
- Using revised coding: 34%
- General population 17%

<table>
<thead>
<tr>
<th>Cause of Death by ICD-10 Chapter</th>
<th>ID ABS Convention</th>
<th>ID Revised</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Rank</td>
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</tr>
<tr>
<td>Circulatory system</td>
<td>1</td>
<td>70 (8)</td>
</tr>
<tr>
<td>Infections</td>
<td>2</td>
<td>39 (5)</td>
</tr>
<tr>
<td>Other external causes of morbidity and mortality</td>
<td>3</td>
<td>36 (4)</td>
</tr>
<tr>
<td>Cancer</td>
<td>4</td>
<td>32 (4)</td>
</tr>
<tr>
<td>Respiratory system</td>
<td>5</td>
<td>16 (2)</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>219 (26)</td>
</tr>
</tbody>
</table>

Avoidable Deaths:
- people aged under 75 years:
- potentially preventable deaths: those amenable to screening and primary prevention, such as immunisation
- deaths from potentially treatable conditions: those amenable to therapeutic interventions
Session 2
Vulnerability to Cognitive Disorders in People with ID

Group task: Vulnerability to cognitive disorders in ID
Dementia in ID
Risk and compounding factors
Your Turn

• Working in your small groups (12 mins):
  – Appoint a scribe and a spokesperson (2 mins)
  – Individually, brainstorm factors that might increase vulnerability to dementia in people with ID & write one factor on each post-it note (5 mins)
  – As a group, share your ideas and order these from greatest to least impact on likelihood of developing dementia (5 mins)

• If time permits, groups will be randomly chosen to share their lists
Dementia in People with Intellectual Disability

**People with Down Syndrome**
- Dementia in DS 3-4 x that of general population (Strydom et al 2007)
- Dementia in DS
  - ~ 20% of persons with DS aged 45+
  - ~ 50% by 60-70 years
- greater risk of mortality for persons with DS and dementia or cognitive decline (Baird, 1988; Yang, 2002)

**People Without Down Syndrome**
- Prevalence & incidence figures vary
- Likely 2-3 X general population
  - 13% in those aged 60+ years (Strydom, 2007)
  - 6% in those aged 60+ years (Zigman, 2004)
  - 21.6% in those aged > 65 years (Cooper, 1997)
Why is Dementia More Common?

• Association with Syndromes eg DS

• Interaction of known risk factors with specific types of ID
Why is Dementia More Common?

• Interaction between other health conditions and cognition
Why is Dementia More Common?

• Lifestyle factors
  – Diet
  – Overweight and obesity
  – Lack of exercise

• Cognitive Reserve Hypothesis
The Successful Ageing in Intellectual Disability Study (Sage-ID)
Basic Demographics of the SAge-ID Sample

• 117 people aged 40 – 76 years, mean 51.2 (±7.7)
• Living Situation
  – 51% residential care facility
  – 15% independent living
  – 32% living with family
• 97% single. 4% married or divorced.
• Employment
  – 28% currently employed
  – 17% previously employed, but no longer employed
  – 29% never employed
  – Production line/factory industry employed majority
• Skewed towards more severe level of adaptive behaviour and functioning
• Down syndrome 11%
Prevalence and Key Findings for Health Conditions

<table>
<thead>
<tr>
<th>Condition</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing and sight disabilities</td>
<td>28%, 20%</td>
</tr>
<tr>
<td>Chronic illness</td>
<td>51%</td>
</tr>
<tr>
<td>Epilepsy/Previous fit</td>
<td>38%</td>
</tr>
<tr>
<td>Thyroid problems</td>
<td>13%</td>
</tr>
<tr>
<td>Multimorbidity; when physical and/or sensory disabilities included</td>
<td>32%; 56%</td>
</tr>
<tr>
<td>Obesity was present (1.5 times the general population)</td>
<td>40%</td>
</tr>
<tr>
<td>Hypertension</td>
<td>27%</td>
</tr>
<tr>
<td>Hypercholesterolaemia</td>
<td>26%</td>
</tr>
<tr>
<td>Diabetes (2x rate of general population)</td>
<td>17%</td>
</tr>
<tr>
<td>Cardiovascular event (MI, Stroke, TIA)</td>
<td>9%</td>
</tr>
</tbody>
</table>
Psychotropic Medications

• 60% taking a CNS medication.
• 40% polypharmacy

• Most common medications: anticonvulsants (30%); antipsychotics (29%); antidepressants (21%).

• CNS medication associated with:
  – high cholesterol, particularly
  – Diabetes (antipsychotics)
  – Obesity (antidepressants)
Prevalence of Mental Disorders

- Lifetime diagnosis of mental disorder: 36%
- DBCA score above the cut off: 19%
- 45% of people had formal diagnosis and/or DBCA score over cutoff
- ASD: 15%

- Anxiety and mood disorders: 26%
- Psychotic disorders: 24%
- Multiple disorders: 17%
Comparison of Dementia Diagnosis

- Dementia
  - DCR-10: 7.3%*
  - DSM-IV: 6.1%*
  - DCLD: 7.3%*
  - DSM-5 Major ND: 4.9%*
- MCI (Winblad): 13.4%*; DSM-5 Minor ND: 14.6%*

<table>
<thead>
<tr>
<th></th>
<th>CAMDEX</th>
<th>DSQIID</th>
<th>ABDQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>DSM-IV</td>
<td>0.42</td>
<td>0.52</td>
<td>0.2</td>
</tr>
<tr>
<td>DSM-5</td>
<td>0.51</td>
<td>0.34</td>
<td>-0.11</td>
</tr>
<tr>
<td>DCR-10</td>
<td>0.093</td>
<td>0.44</td>
<td>0.16</td>
</tr>
<tr>
<td>DC-LD</td>
<td>0.35</td>
<td>0.22</td>
<td>-0.13</td>
</tr>
</tbody>
</table>

*82 eligible participants
What do we know about psychotropic prescribing patterns in people with ID?

Compared to the general population people with ID are more likely to be exposed to psychotropics (and their associated risks and benefits)

- Higher rates of mental illness (Cooper, Smiley et al. 2007)
- Psychotropics as treatment for challenging behaviour (Deb, Unwin et al, 2009)
- Commencement of psychotropics at a younger age (Matson and Mahan. 2010)
- Psychotropic polypharmacy (Deb, Unwin et al. 2014)
- Inadequate monitoring of psychotropic side effects (McGillivray & McCabe 2004)
Cardiometabolic Risk Factors and ID

While cardiovascular events are the leading cause of death amongst both the general population and people with ID (Patja, Mölsä et al. 2001) certain risk factors for cardiometabolic morbidity and mortality remain more prominent in people with ID.

Cardiometabolic risk factors that people with ID may be particularly vulnerable to include:

• Higher rates of psychotropic prescription
• Higher levels of physical inactivity and obesity
• Increased barriers to accessing quality healthcare
• Certain genetic syndromes
• Increased chance of socioeconomic disadvantage and stigma
Session 3

Clinical Manifestations and Diagnostic Criteria

Clinical manifestations
DM-ID working group adaptations to DSM 5 criteria
Limitations of diagnostic criteria
Clinical Manifestations: Symptoms of Neurocognitive Disorders in ID

Early
- Cognitive: Memory and language skills
- Non-cognitive: personality, behaviour and social skills changes (may proceed cognitive)
- Reduced capacity for complex activities of daily living eg personal hygiene, housekeeping and work skills

Mid
- Dyspraxias
- Decline in basic ADLs: toileting, dressing and eating
- Increase in challenging behaviour
- Impulsivity and personality changes

Late
- Reduced response to the environment
- Reduced mobility, parkinsonism
- Loss of basic communication skills
- Incontinence
- Seizures
A note about Dementia in DS

- Personality change may occur early eg apathy, lack of motivation, and stubbornness
- Behavioural excess: irritability, aggression or self-injury
- Behavioural deficits: slowness, apathy, loss of interest, reduced social engagement.
- “frontal-temporal-like” in presentation
## Major Neurocognitive Disorder

| A. Evidence of significant cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition) based on: | Take care to determine previous level of performance  
Change in performance need to be clearly established  
Neuropsychological tools should have been designed for the intellectual population and/or validated in this population; including proxy-rated tools of cognitive performance |
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Concern of the individual, a knowledgeable informant, or the clinician that there has been a significant decline in cognitive function; and</td>
<td></td>
</tr>
<tr>
<td>2. A substantial impairment in cognitive performance, preferably documented by standardized neuropsychological testing or, in its absence, another quantified clinical assessment.</td>
<td></td>
</tr>
<tr>
<td>B. The cognitive deficits interfere with independence in everyday activities (i.e., at a minimum, requiring assistance with complex instrumental activities of daily living such as paying bills or managing medications).</td>
<td>Needs to be a change from previous level of functioning (i.e. measured against the person’s own premorbid baseline)</td>
</tr>
<tr>
<td>C. The cognitive deficits do not occur exclusively in the context of a delirium.</td>
<td>No change</td>
</tr>
<tr>
<td>D. The cognitive deficits are not better explained by another mental disorder (e.g., major depressive disorder, schizophrenia).</td>
<td>No change</td>
</tr>
</tbody>
</table>
## Adaptation to Diagnostic Criteria

### Mild Neurocognitive Disorder

<table>
<thead>
<tr>
<th>A. Evidence of modest cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition) based on:</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Concern of the individual, a knowledgeable informant, or the clinician that there has been a mild decline in cognitive function; and</td>
</tr>
<tr>
<td>2. A modest impairment in cognitive performance, preferably documented by standardized neuropsychological testing or, in its absence, another quantified clinical assessment.</td>
</tr>
<tr>
<td>Establishing change from previous level is crucial. Deterioration should be on basis of sequential assessment Modest impairment is difficult to interpret</td>
</tr>
<tr>
<td>B. The cognitive deficits do not interfere with capacity for independence in everyday activities (i.e., complex instrumental activities of daily living such as paying bills or managing medications are preserved, but greater effort, compensatory strategies, or accommodation may be required).</td>
</tr>
<tr>
<td>No change</td>
</tr>
<tr>
<td>C. The cognitive deficits do not occur exclusively in the context of a delirium.</td>
</tr>
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</tbody>
</table>
Difficulties in dementia diagnosis in people with Intellectual Disability – 1

• Difficulty in objectively defining cognitive impairment and decline in a population with pre-morbid deficit
  – Especially problematic for mild neurocognitive disorder,

• Formal neuropsychological testing:
  – Unpredictability (factors include contextual issues, emotional states, sensory problems, medical status, medications etc.); hard to optimise test results
  – Limited range of suitable tests, especially for those with more severe intellectual disability,
  – wide range of baseline abilities across different domains.
Difficulties in dementia diagnosis in people with Intellectual Disability- 2

• Tendency to rely on carer questionnaires to assess cognitive functioning
  – Questions often do not map well on to specific cognitive domains
  – The ‘context’ is not considered, e.g. whether a task is regularly ignored because the person is unwilling to do it rather than unable to do it or the familiarity of the person with his/her carers.
  – Questionnaires do not discriminate between ‘physical’ and ‘cognitive’ reasons for functional impairments
  – Reliability of informant reports of impairment or decline
Differential diagnosis and co-morbidity

• common co-morbidities in intellectual disability:
  – hearing or vision loss
  – thyroid function disorders (particularly in individuals with Down syndrome)
  – mental ill health esp. depression.
  – systemic illness, pain and constipation
Session 4
Dementia Assessment

General adaptations
Specific tools and approaches
Session 4
Dementia Assessment

Part 1: General
Top 10 Core Competencies

For working with people with ID

1. **PREPARE**- Take the time to prepare for working with a person with an intellectual disability by finding out about their strengths, and the support they may require to actively participate

2. **ADJUST COMMUNICATION**- Determine the person’s preferred communication style, and appropriately adapt your communication style to meet their needs

3. **ENGAGE SUPPORT NETWORKS**- Identify the person’s support network, and when appropriate to do so, and when consent to their involvement is given, works with them at all stages of service delivery

4. **FACILITATE SUPPORTED DECISION-MAKING**- Facilitate supported decision-making and give priority to the person’s expressed wishes, as far as possible

5. **MAKE INFORMATION ACCESSIBLE**- Provide information to the person with an intellectual disability, their family, and support networks in accessible formats, at all stages of the clinical process, acknowledging that the format may be different for different stakeholders
Top 10 Core Competencies  
For working with people with ID

6. **KNOW WHAT SKILL SETS ARE AVAILABLE** - Be aware of the different skills and approaches available in the mental health and disability sectors, and use this knowledge to facilitate collaborative work

7. **COLLABORATE ACROSS AGENCIES** - Work with partner organisations to deliver a seamless service to people with an intellectual disability, their families, and support networks

8. **SEEK SUPPORT** - Identify and actively seek support from specialist intellectual disability mental health professionals, when required

9. **LEARN & INTEGRATE KNOWLEDGE INTO PRACTICE** - Learn about intellectual disability mental health and use your new knowledge to improve practice

10. **REFLECT** - Reflect on how your personal beliefs, and emotional reactions towards people with a disability might influence your clinical practice
Adaptations to assessment
Before the assessment session

**Top 10 Core Competency**

**PREPARING**: Take the time to prepare for working with a person with an intellectual disability by finding out about their strengths, and the support they may require to actively participate

- Find out as much as possible about the person **before the appointment** eg preferred communication methods, communication aids they may need, mobility or sensory needs, if a support person will accompany them
- Read any **previous assessments/records** you have consent to access
- Allow **extra time** for the assessment; it may need to be conducted over a few shorter sessions
- Simplify appointment and referral letters using **Easy English**; make reminder phone calls
Adaptations to assessment

Clinic room/assessment environment

- If possible, see the person with ID in a **setting that is familiar** to them e.g. home, day centre
- Arrange room to **accommodate sensory/mobility needs** e.g. fluorescent lights can be stressful for individuals with autism
- Choose quietest room with fewest distractions if possible
- **Try to be punctual**; waiting for appointments can cause particular anxiety for PWID
- If an individual has to wait and is anxious, check whether they would be more comfortable waiting in a quiet room away from the waiting room. **Try to avoid long waiting times in high stimulation environments**
Adaptations to assessment

Communication

Top 10 Core Competency

ADJUSTING COMMUNICATION: Determine the person’s preferred communication style, and appropriately adapt your communication style to meet their needs.

- Communication skills vary; mild ID - may have necessary receptive and expressive speech to conduct assessment; moderate to severe ID - may use methods other than speech as primary communication
- Use reflective approach - ensure PWID has understood you, and you have understood them; allow more time for individual to answer
- Communication > verbal eg gesturing, smiling, nodding, showing pictures.
- Some individuals will use augmentative and alternative communication methods, or ACC
- Write instructions/factsheets/documents in Easy English where possible.

Example AAC techniques

<table>
<thead>
<tr>
<th>Unaided</th>
<th>Aided</th>
</tr>
</thead>
<tbody>
<tr>
<td>Facial expression</td>
<td>Written text</td>
</tr>
<tr>
<td>Natural gesture/sign</td>
<td>Real objects</td>
</tr>
<tr>
<td>Key Work Signing eg Makaton vocab</td>
<td>Pictures/symbols eg Picture Exchange Communication System (PECS)</td>
</tr>
<tr>
<td>Pointing</td>
<td>Photos</td>
</tr>
<tr>
<td>Finger spelling</td>
<td>Electronic talking device</td>
</tr>
<tr>
<td>Signed language</td>
<td>Computer systems</td>
</tr>
</tbody>
</table>
Adaptations to assessment

During the assessment

- **Greet the person with ID first**: speaking directly to them. If a support person has come along, check with the PWID if it’s OK for you to ask the third party about them.
- **Adapt communication** to support the individual’s engagement.
- **Assess capacity to consent**: PWID may be able to consent for simple decisions, but not complex ones (→’person responsible’/appointed guardian); this may change over time.
- Consider **medical and developmental complexity** (comorbid medical/physical/sensory conditions, other disabilities, polypharmacy, other DD e.g. ASD, ADHD).
- Be aware of **diagnostic overshadowing** - symptoms/behaviour inappropriately attributed to ID rather than to a mental health disorder.
- Investigate **baseline functioning** as this will vary and be unique to the individual; central to interpreting current functioning.
- Be aware of **specific/appropriate assessment tools** for use with PWID.

*Top 10 Core Competency*

Facilitating **SUPPORTED DECISION-MAKING**: Facilitate supported decision making and give priority to the person’s expressed wishes, as far as possible.
Adaptations to assessment

Involving others

- **Identify support networks**- before and during assessment
- With consent of PWID, **seek information** from family/support person who has known the individual long-term; determine if symptoms/behaviours are new or ongoing; identify coping strategies
- Provide **psychoeducation/factsheets** to support persons
- Family carers/support persons can assist with **monitoring and seeking follow-up**; create a support plan
Session 4
Dementia Assessment

Part 2: Specific Adaptations
Dementia Assessment in ID

- There is no gold standard
- Important things to consider are:
  - Administrative: Flexible age cuts for clinics
  - Screening: Proactive screening for an at risk population > 40
  - Assessment:
    - Establishing severity of ID and baseline functioning
    - Individual’s history (subjective cognitive complaints)
    - Reliance on carer reports
    - Functional status (IADLs/ADLs): sensitivity to environmental changes, changes in physical status
    - Structured assessment of cognitive function, tailored to level of ID but note lack of robust cognitive assessment tools
    - the ‘bounce’ phenomenon
    - Effect of medical and psychiatric comorbidities
  - Follow-up assessment needed
Areas to assess – ideal scenario

- Pre-morbid IQ
- Attention
- Processing Speed
- Visuospatial skills
- Language and communication
- Executive Function
- Fine motor
- Mood
- Motivation
Carer Reports

CHECKLISTS
DSQIID/NTG-EDSD

Dementia Screening Questionnaire for Individuals with Intellectual Disability (DSQIID):
• Three sections:
  – Best function
  – Declines in ADLs/Adaptive Behaviours
  – Overall observed changes in mood, skills, etc.
• All levels of ID
  – But severe-profound not tested separately in establishing the cut-off score
• Administer: Once, with cut-off score
• Pros:
  – Free, quick
  – Reasonable data, though more is needed
• Cons:
  – Sample restricted to DS
  – 53 items on one A4 page: needs retyping for older carers

National Task Group Early Detection Screen for Dementia (NTG-EDSD):
• *Almost* every item of the DSQIID plus more
• Unscored
• Suggest use over time
• Further evaluation is proposed by the task group.
Adaptive Behaviour Dementia Questionnaire (ABDQ)

- 15 Items of the Adaptive Behavior Scale where declines over time associated with AD in DS
- All levels of ID
- Administer: once, with cut-off score
- Pro’s:
  - Face validity and statistical approach to development
  - Good properties – for AD in DS
  - Free
  - Quick
- Cons:
  - Sample for test development was still small (n = 150)
  - No data regarding other forms of dementia/non-DS ID
Dementia Questionnaire for Persons with Learning Disabilities (DMR/DLD)

- 50 items in 8 subscales, clustered into 2 subscores:
  - Cognitive
  - Social
- All levels of ID
- Administer:
  - Sequentially
  - Previously: singly with different cut-offs for different levels of ID
    - No longer recommended due to poor psychometric data
- Pros:
  - Validation sample included people with all levels of ID, and non-DS ID
  - Good face validity
  - Reasonable results for validity administered sequentially
- Cons:
  - Very expensive
  - Sequential administration means
  - Hence impractical as checklist for screening
    - But potentially useful supplement to an assessment in clinical setting
Carer reports

INTERVIEWS
Dementia Scale for Down Syndrome

• Structured interview
• Better for moderate – profound ID
  – Recommendations for mild ID vary
• Pros:
  – Some good data regarding moderate-profound ID
• Cons:
  – Needs 2 informants, one knowing person for 2+ years
  – Restricted to masters-level psychologists
  – Some conflicting data on psychometric properties
    • May miss some early cases of dementia
CAMDEX-DS

- Semi-structured carer interview. Essentially a clinical tool.
- Assesses declines *and their time-frame*
- Accompanies the CAMCOG-DS test
- All levels of ID
- Aligns items of interest with DSM-IV, ICD-10 and CAMDEX criteria
  - Including AD subtyping
  - Flags for potential differentials, with algorithm
- Administer: Once.
- Pros:
  - Psychometric data are good
- Cons:
  - Data based a small sample (n=74/56) and only DS
  - Takes 45 – 60 minutes
Direct Assessments

OMNIBUS TESTS
CAMCOG-DS

• Adaptation of the CAMCOG and Severe Impairment Battery
• Administer:
  – Sequentially to track changes over time
  – Authors say it is appropriate for most with DS
  – Severe ID – floor effects still likely.
• Pros:
  – Accompanies the CAMDEX-DS Carer interview
  – CAMDEX-DS + The CAMCOG together have good properties
• Cons:
  – Properties established on a small sample (n=56)
  – UK-centric items
  – May miss some cases
Neuropsychological Assessment of Dementia in Individuals with Intellectual Disabilities (NAID)

- Conglomerate of bits and pieces from various tests
- Domains and Subscales:
  - Early signs:
    - Memory for Objects
    - Memory for Pictures
    - Memory for Sentences
  - Late signs:
    - Picture Naming
    - Picture Identification
    - Action on Request
  - Total Score
- Scores suggest: normal, of interest or of concern.
NAID Continued

- Mild and moderate ID only
- Administer:
  - Sequentially - reliable change index
  OR
  - Once – cut-offs stratified according to either:
    • Vineland Adaptive Behavior Scale
    • British Picture Vocabulary Scale (BVPS - like the Peabody).
- Pros:
  - Validation data on over 400 people with Down Syndrome
    • Most impressive data of any test in this field so far
  - Capacity to assess at single time and sequentially
- Cons:
  - Copyright issues are tricky – need to make test yourself
  - Validation group – only DS
  - BVPS is hard to find in Australia, Peabody more common. The Vineland version used for norming is outdated.
Severe Impairment Battery

- Designed for use in the general population with severe dementia

- Subscales:
  - Attention; Orientation; Language; Memory; Visuo-spatial ability; Construction

- Administer: Sequentially
  - For Moderate – Severe ID
  - Mild may get ceilings
  - Profound: floor effects likely

- Pros:
  - Availability

- Cons:
  - Not a lot of data on its usefulness in ID
  - Expensive to buy
Test for Severe Impairment

- Designed for general population with floor effects on MMSE.
- For moderate - severe ID:
  - Ceiling effects in mild ID unless already declining
  - Floor effects still a risk with severe & profound ID
- Administer:
  - Sequentially to detect declines
  - A single administration cut-off score may become available
- Pros:
  - Quick, easy
  - Free
- Cons:
  - Limited range of applicability
  - no reliable change index or cut-off score as yet.
Prudhoe Test of Cognitive Function

• Designed for use with people with ID
• Many items adapted from the MMSE
• Covers:
  – Orientation; Recall; Language; Praxis; Calculation
• Short form available
• For Mild ID
  – Relies on verbal comprehension and responses
  – Floor effects likely for lower moderate/severe/profound ID
• Administer:
  – Sequentially, 6+ months apart
  – Authors suggest a 50% decline in scores indicates dementia.
• Pro’s:
  – Brief to administer, to track over time
  – Face Validity and Test-Retest reliability established
  – Free, downloadable from https://research.ncl.ac.uk/pcft/
• Cons:
  – Diagnostic accuracy yet to be established.
Direct Assessments

SPECIFIC DOMAINS
Executive Function

Tower of London
• Others have found this a useful test of early signs of dementia in DS
• Mild ID only
  • Our study: floor effects even in mild ID in a modified version (TOLDx)
  • Administer: Sequentially, as norms will be useless

Scrambled Boxes Test
• Tests Executive Memory
• Mild – Moderate ID
• Administer: Sequentially
• Pros:
  – Varying degrees of difficulty
    = minimise floor effects
  – Maintains rapport & motivation
• Cons:
  – DIY test materials
  – No change index/cut off score as yet
Memory

Cued recall test:
• Association between picture and category (= encoding)
• Both free and cued recall then tested – only for items named
• Mild & upper moderate ID
• Administer: once-off, with cut-off score
• Make the test materials yourself

FULD object memory test (modified by Sano 2005):
• Mild to moderate ID
• Sequential assessment
• Some subscales may be useful for detecting declines
Praxis:
Dyspraxia Scale Down Syndrome

• 62 items
• Items administered in up to 4 ways:
  – 1. Request
  – 2. Additional verbal cues
  – 3. Model
  – 4. Physical prompt (e.g. hand over hand) – check if responsive
• Moderate – Profound ID
  – Or milder but with more advanced dementia
• Pros:
  – Free (if you want to buy a very good book – Prasher 2009)
  – Reasonable data on reliability and validity
  – Little training required to administer
• Cons:
  – Takes around an hour
• 20 item version = The Brief Praxis Test
  – Administer sequentially, expected changes per year are given by Sano 2005
Adaptive Behaviour

- Monitor to detect functional declines over time
- Almost all are informant interviews or checklists
- Similar to IADL's and ADL's
- Where many scales of IADLs and ADLs yield floor effects, measures of adaptive behaviour may be useful
- One element in establishing level of ID (pre-declines)
- Suitable for all levels of ID
  - Psychometrics not well established at the severe-profound end though
- Many people with ID will already have pre-decline measures in their files, but note:
  - Different scales are not comparable
  - Some age-related decline can be expected, and/or due to other conditions
- If you’re going to choose one to use over time, choose the Adaptive Behavior Scale (ABS).
Mood/Behaviours

• High level of behavioural/psychiatric problems in ID without dementia
• Changes in behaviour = an important early sign
  – DS and AD: a FTD-like prodrome has been suggested
  – Non-DS ID: less is known
• Important issue for:
  – Management
  – Carer wellbeing
Language and Communication

• Evaluating language may be useful to track changes over time:
  – Grammatical structures
  – Simple and complex syntax
  – A sample of their use of spoken language – track declines in richness, fluency.

• Single-word receptive vocabulary:
  – one of the last skills to decline
  – correlated ~ 0.7 with IQ
  – good to include if IQ unknown
  – e.g. PPVT.

• Boston naming test – some data on this in ID (Palmer 2006)
Motivation

• Prone to fatigue – give breaks
• “Assessment” can have negative connotations for this group
• Maintaining rapport is important
• Consider undiagnosed sensory disabilities
• Ask carers about changes in motivation
Session 5
Clinical Scenarios & Approaches
Case 1

You are a clinical neuropsychologist in an older person’s health/mental health service. You are contacted by a local GP asking your service to assess a 45 year old woman (Belinda) with Down syndrome associated with mild intellectual disability. Her parents have become concerned about changes in cognition and behaviour, which have been obvious for about 2 months. She seems to be forgetful, slower, and withdrawn. She refuses to go to her day placement, and is more easily prone to tearfulness and displays of irritability.

1. What are the most likely things that could possibly explain this presentation?
2. Outline the key steps which will need to followed in order to ensure a thorough assessment.
3. Are there any specific tools related to people with intellectual disability may be of use in this assessment?
4. Under what circumstances (if any) would detailed neuropsychological assessment be warranted?
Case 2

You are a clinical neuropsychologist working in a neurology service. You are asked to consult regarding a 26 year old man (Max), with moderate intellectual disability and recent functional decline, who has just been admitted to hospital for assessment. There have been major changes in Max’s ability to carry out his IADLs over a 3 month period. He has also been noted to be increasingly vague and forgetful. One month ago he had his first ever seizure, and was started on anticonvulsants. He has been sleeping poorly since this event. He has become unsteady on his feet, but it is unclear if this is related to his anticonvulsants or something else. He has a history of a psychotic illness, which is probably schizophrenia, and is also taking olanzapine and lithium.

1. What would the role of a neuropsychologist be in this situation?
2. What factors could be contributing to the apparent decline in cognition and functioning?
Case 3

You are a clinical neuropsychologist in an older person’s health/mental health service. You are contacted by a local disability service asking you to assess a 55 year old woman (Liz) with Down syndrome associated with mild intellectual disability. The key worker reports changes in language and memory, which have been subtly worsening over at least 12 months. Liz has also become more impulsive, moody and uncharacteristically aggressive.

1. What may explain Liz’s symptoms?
2. What assessments are needed and who should perform them?
3. Under what circumstances would detailed neuropsychological assessment be warranted?
Case 4

You are contacted by a proactive team leader of a large NGO disability service provider in your location. The NGO has a particular focus on services for older people with intellectual disability. Following a diagnosis of Alzheimer's disease in two clients, the team leader (a registered psychologist) has done some reading about dementia in people with intellectual disability. The team leader would like to set up a screening and monitoring process for people over the age of 40 with ID.

• How would you proceed?
Session 6
Resource Review; Q & A