Dementia in People with an Intellectual Disability: Challenges and Opportunities in Care

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Celebration & Challenge of Ageing

A success story
Little known ageing
Promoting life long health
Maintaining independence
Postponing disability
Reorienting ID services
Mainstreaming agenda
Changing Demographic Profile of People with an Intellectual Disability in Ireland

Proportion of people with moderate, severe and profound ID: 1974 - 2014

Sources:
National Intellectual Disability Database, Health Research Board, 2014
Median Age at Death of Persons with Down Syndrome:

We now need to support a rapidly expanding older population with DS
Ensuring that older adults with an Intellectual Disability are part of the Public Health Agenda: (IDS TILDA)

• Identifying the principal influences on ageing

• Comparable with the general population study – TILDA

  • 753 participants
  • 55% Female; 45% Male
  • Age 41 – 90 years
  • All living circumstances
  • Two additional Wave 2 data elements:
    • Health Assessment
    • End of Life
  • Wave 3 about to begin
Characteristics of atypical ageing associated with Down syndrome

- Premature changes in skin and hair greying
- Increased frequency of senile cataracts
- Increased frequency of hearing loss

- Age-related increase in hypothyroidism
- Age-related increase in seizures
- Early menopause for women
- Increased risk of Osteoporosis

- Dramatically increased risk of AD
- Reduced life expectancy
Comparative Rates of Dementia Down’s syndrome, I.D., General Population

Holland 2011
Point Prevalence of dementia among Down syndrome IDS-TILDA

Prevalence of dementia among people with Down syndrome

WAVE 1: 15.8%
WAVE 2: 29.9%

The prevalence of epilepsy increased from 19.2% to 27.9% for those with Down syndrome
The Association between Down Syndrome and Alzheimer’s Dementia is not New

Reports of dementia in older adults with Down syndrome date back to the late 1800’s -

Early reports attracted a lot of academic interest and people with Down syndrome have contributed greatly to our overall understanding of dementia. However very few individuals lived long enough to be at risk.

In Ireland it is only recently that the public health significance of this association was appreciated and relevant issues began to attract “serious” attention - National Dementia Strategy 2015

‘The need for systems, structures and age appropriate services specifically to promote timely diagnosis of people experiencing early onset dementia, including people with Down Syndrome’ p21.
Why is there an association between AD and DS?
Down syndrome and APP

The gene coding for APP is on chromosome 21

Individuals with DS have 3 copies of this gene and produce excess APP

Over many years, this excess APP leads to β-amyloid plaque formation and eventual AD
DS (Age 65): Parahippocampus immunostained to show βA plaques

Source: Silverman et al 2016
Alzheimer’s Disease Associated Gross Pathology

Normal Aged Brain (1,300 gms)  Alzheimer Brain (560 gms)
(Courtesy of Dr. Jerzy Wegiel)
Neuropathology of Dementia and Adults with Down syndrome

Almost all individuals with DS have amyloid deposits in their brains consistent with a diagnosis of AD by the time they are 35-40 years of age.

Incidence of dementia increases in 50s; Mean age of onset 51 yrs.

Substantial individual differences

Why is there variability in the age of onset?
Diagnosing Dementia in people with DS is Highly Complex

Challenges
- Pre-morbid functioning
- Difficulties in using standardized tests
- Communication difficulties
- Improvised care environments
- Lack of base line
- Consequence of high staff turnover

Assessment
- Baseline assessment
- Annual follow-up DS > 35yrs
- Diagnostic work-up
- PCP
- Staff training
- Service redesign
- Policy inclusion

Memory Clinic Model
Well Established and Award Winning Dementia Specific Service At the Daughters of Charity Service

KEY COMPONENTS

1. Specialist Memory Clinic + comprehensive minimum data set to guide policy and service

2. Rolling tailored education program for staff, peers and family

3. Dementia Specific Day program underpinned by dementia specific PCP and Dementia Care Standards

4. Dementia Specific care homes to support people across the continuum of dementia also offering respite to family and community group homes

5. Research to guide Policy and Practice
Baseline Screening: Annual Follow up Consensus Dementia Ratings

- **No dementia**
  - Stable or age-related change

- **Clinical uncertainty**
  - All persons with DS >35 -40 years, annual follow up, classification established every 12-14 months based upon a comprehensive evaluations

- **Definite dementia**
  - Substantial declines of extended duration

- **Unknown status**
  - Circumstances prevent determination
Memory Clinic – Current Dataset

177 Persons with Down syndrome Assessed
Differential diagnosis of functional and cognitive decline

Consider other physical or mental health problems:

– Depression or other mental illness
– Sensory impairment: vision/hearing
– Thyroid impairment, B12, folate deficiency
– Medical problem- drugs, acute, chronic, infection, pain, epilepsy
– Major life events: Separation, bereavement, environmental changes

These conditions are common in people with Down syndrome and mimic dementia
Understanding Cognitive Decline: Diagnostic Workup

**Physical**
- Vital signs
- Full physical examination
- Complete blood count - FBC, E/LFT, ESR, TFT, Folate
- & B12, U&E, drug levels etc.
- Mental Health
- Urinalysis
- Vision, Hearing

**Neuro-imaging**
- CT, MRI (depending on feasibility)

**Neuro-Psychological Testing**
- Informant and objective based measures
Well Defined Clinical Characteristics of Dementia in people with DS

DSM-IV Definition of Dementia

Changes in Mood and Behaviour
Decline in social, community and daily living skills.
Memory and Cognition – disorientation in time and place

Motor Function

New onset epilepsy

Severity of deficits increase dramatically over time
Mental health

A prospective 14-year longitudinal follow-up of dementia in persons with Down syndrome

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### Findings

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<tr>
<th>Percentage</th>
<th>Description</th>
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<tr>
<td>89.6% (n=69)</td>
<td>developed dementia</td>
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<td>Age of onset: 55.41 years (SD 7.14)</td>
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<td>88.52% (n=54/61)</td>
<td>persons with moderate ID developed dementia</td>
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<td>93.3% (n=15)</td>
<td>Persons with severe ID developed dementia</td>
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Cognitive and Physical Functioning Measures

TSI, DSMSE, DLSQ and DMR Vs Years since dementia diagnosis

Years since dementia diagnosis

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Years since dementia diagnosis

Trinity College Dublin, The University of Dublin
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Risk trajectory according to age
Mortality

Survival curve

Cumulative Survival

Time in years

Median
Co-morbidities profile of people with and without dementia
Some Key Conclusions

Substantial increased risk of dementia >50 years
Rate of progression seems slightly increased, but, nonetheless:

Mortality risk is low during the first few years following diagnosis.

Rate of progression varies substantially among individuals.

Anecdotal reports of adults with Down syndrome “falling off a cliff” reflect unusual cases.

High risk of new onset epilepsy

Increased survival at advanced dementia
Huge Gaps in Knowledge

All adults with DS in their mid-30s or older have key neuropathological hallmarks of AD, and yet many will not present with the clinical features of dementia until they are much older.

Raises the question that current “gold standard” for diagnosis of AD is perhaps meaningless for this population?

Why some individuals with Down syndrome are far more vulnerable than others?

We need to urgently develop skills in dementia recognition and assessment for people with DS; valid diagnostic standards, and we need to determine what specific neuropathology actually causes dementia.

Most critically we need to develop responsive and humane services to respond to the changing needs of this increasingly at risk population with dementia.
Experience of dementia is unique to the individual and depends upon the interaction of five key factors:

\[ D=P+B+H+NI+SP \]

- **P** = The persons Personality
- **B** = Their Biography
- **H** = Physical Health
- **NI** = Neurological Impairment
- **SP** = Social Psychology of the environment

*Kitwood (1996)*
Developing Standards to Guide Care

http://www.docservice.ie/includes/documents/Dementia%20Publication%202011.pdf
Moving Ideas into Action: A Dementia Specific Home

Designed to create opportunities for persons with dementia to succeed and use their retained abilities by:

- maximising ease in finding their way
- Familiarity
- promote a feeling of empowerment
- freedom and control
- while at the same time reduce opportunities for potential failure and feeling trapped and/or alienation.
Specialist Care Centre of the Year
Daughters of Charity Disability Support Services