Age-related mental health needs in people with intellectual disabilities: understanding the complexity of the relationship and the extent of need

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Objectives

- To provide a research perspective on MH and ageing as it may affect people with ID;
- To consider the different conceptual models that contribute to an understanding of mental health and challenging behaviour in people with ID;
- To illustrate the above using PWS and DS as examples
The process of ageing

• Ageing
  • Chronological ageing
  • Biological ageing
  • Social ageing

• Distal and proximal effects on ageing

• Age-related illnesses

• Questions
  • Are these the same or different for people with ID?
  • Is the prevalence and the presentation and course of mental health problems and/or challenging behaviours different across the lifespan of people with ID?
What is ID?

• Core descriptive/diagnostic features
  – Intellectual and functional impairments
  – Onset during the developmental period

• Marked variation in the nature and degree of intellectual and social functioning;

• Different causes and varying developmental trajectories
  – Small effects of many genes
  – Major effects of single genes
  – Environmental effects in-utero and childhood

• Impairment/disability distinction
Mental Health and LD

Deb et al, 2001 (n=90 people with language)

13 (14.4% had evidence of psych disorder (ICD-10)

- Schizophrenia 4 (4.4%)
- Depressive disorder 2 (2.2%)
- Generalized anxiety disorder 2 (2.2%)
- Phobic disorder 4 (4.4%)
- Delusional disorder 1 (1%)

(prevalence of psychopathology increased with age and with severity of disability)
Glasgow prevalence study

- Point prevalence of mental ill-health (DC-LD)
- Mental ill-health of any type 35.2%
- Excl problem behaviour and ASD 19.1%
  - No clinical diagnosis 59.1%
  - One clinical diagnosis 29.1%
  - Two clinical diagnoses 9.2%
  - Three clinical diagnoses 2.4%
  - Four clinical diagnoses 0.2%
Glasgow prevalence study

• Point prevalence of mental ill-health (DC-LD diagnostic criteria)
  • Psychotic disorder 3.8%
  • Affective disorder 5.7%
  • Anxiety disorder 3.1%
  • OCD 0.5%
  • Organic disorder 2.1%
  • Alcohol/substance misuse 0.8%
  • ASD 4.4%
  • Problem behaviour 18.7%
Neurodevelopmental syndromes
Arron et al 2011 JIDR, 55,109

• Questionnaire data on SI, aggression, mood, hyperactivity, ASD, repetitive behaviour

• SI:
  • C de L, Cri-du-chat, Fragile-X, Lowe, PWS and SMS syndromes

• Aggression
  • Angelman and SMS syndromes

• Different underlying mechanisms associated with superficially similar behaviours
Questions

• In people with ID to what extent do adult MH/behaviour problems have their origins in different developmental phases across the lifespan?
  – Intra-uterine status (e.g., Barker hypothesis)
  – Neonatal period
  – Childhood
  – Adult life
  – Old age

• What are the mechanisms that mediate these risks?
  – Biological
  – Psychological
  – Social

• What are the implications for the development of new interventions?
PWS over the lifespan

**Intra-uterine (placental)**
- Poor growth
- Limited foetal movement
- High rates atypical births

**Infancy**
- Extreme hypotonia
- Failure to thrive

**Childhood**
- Developmental delay – intellectual disabilities
- Short statute – relative growth hormone deficiency
- Sexual immaturity – sex hormone deficiencies
- Over-eating - risk of severe obesity and its complications
- Scoliosis, respiratory disorders, maladaptive behaviours

**Adulthood**
- Increased risk of obesity (with greater independence)
- Age-related physical and psychiatric morbidity
Hypothesis: genes to behaviour in PWS
Woodcock et al 2009 JIDR, 53: 493-500

• Repetitive and ritualistic behaviours and temper outbursts cluster together;

• Children with PWS reported to show a preference for predictability with negative emotional behaviour and arousal following change (Woodcock et al, 2009);

• Repetitive questions focused on the future and occurred more frequently following change in routine;

• Change produces high demand on cognitive resources – in PWS specific deficit in task switching from one cognitive set to another (cognitive endophenotype) (Woodcock et al Cognitive neuropsychology)
Psychotic illness more common in mUPD than deletion; p<0.001, effect size 0.45

Soni et al 2008, Psychological Medicine, 38, 1505
Later life in people with ID

• Differential mortality rates depending on severity and cause of a person’s ID

• Risk of dementia

• The special case of DS and dementia
Morbidity and Mortality

Mortality in later life of those with severe LD:
  - Respiratory disorders from aspiration and gastro-oesophageal reflux (Patja et al 2002, JIDR, 45, 30-40)
  - Other significant health risk: epilepsy

- Morbidity in later life for people with LD
  - Sensory impairments
  - Thyroid disorders
  - Other age related illnesses
  - Affective and anxiety disorders (12% >49yrs)
  - Dementia (12% >49yrs) (Moss and Patel 1993 and 1995)
  - Lifetime prevalence of mental-ill health 68% (Cooper, 1997)
  - Prevalence of dementia 20% (over 65 years) (Cooper, 1997)
Life expectancy and aged related morbidity in people with LD

• People with mild LD - similar factors in the general population influence age-related morbidity;

• In the LD population in general life expectancy also influenced by additional factors (e.g., severity and cause of LD & associated illnesses)

• Specific causes for LD associated with particular risks in later life (e.g., DS; PWS)
Dementia in people with LD (without DS)

Strydom et al 2007

- 222 people with LD 60 years or older
- 60 screened positive
- 29/60 positive for at least one set of diagnostic criteria
- 29/222 (13%) +ve for dementia

- Alzheimer’s; Lewy body; fronto-temporal dementias

- Presenting symptoms (N=26)
  - General loss of function (13/26)
  - Behavioural and emotional change (4/26)
  - Reported deterioration in memory (rare) (2/26)
Comparative Rates of Dementia - Down’s syndrome, L.D., General Population

Cooper, personal communication
Aetiology of AD in people with DS

• Amyloid cascade hypothesis
  – Relationship between the extent and distribution of amyloid distribution and cognitive function and diagnostic status with age

• Premature ageing hypothesis
  – AD is more than a disorder of the brain – risk increases with chronological and physical ageing
Down syndrome: Neuropathological Hallmarks
APP mutations/Down Syndrome → APP → Aβ42 → Tau Dysfunction → Cell Death

PS1/PS2 mutations → Plaques → Tau Dysfunction (?)

Tau mutations (FTDP-17) → Tangles (?)

AD: the Amyloid cascade Hypothesis Hardy and Higgins, 1992
Proof of principle study
[11C] Pittsburgh Compound-B to image brain fibrillar β-amyloid in people with DS

Questions?
What is the relationship between cerebral amyloid load and distribution and:

• Regional and generalised cerebral atrophy
• Cognitive function
• Diagnosis of dementia

• Is it safe?
• Is it acceptable to people with DS?
• Does it work?

Landt et al, (2011) using PET and carbon 11-labelled Pittsburgh Compound B to Image Brain fibrillar β-amyloid in adults with DS. Archives of Neurology
D’Abrera et al (in submission) JIDR
28-year-old participant, DS, no AD (1)
51-year-old participant, DS, AD (2)

For information about the project: contact Tony Holland at ajh1008@cam.ac.uk
Selected previous studies

- Martin, G.M. (1978) Genetic syndromes in man with potential relevance to the pathophysiology of ageing *Birth Defects Original Article Series*; 14, 5-39


- Schupf et al (2006) Bioavailable estradiol and age at onset of AD in postmenopausal women with DS *Neuroscience letters*; 406, 298-302

- Life-expectancy

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Age-related markers

• Physical
  • Skin wrinkles (‘crow’s feet’)*
  • Skin pigmentation (back of hands)
  • Frontal and crown baldness (males only)
  • Hand grip strength
  • Gait speed
  • Blood pressure

• Biochemical
  • Dehydroepiandrosterone (DHEA and DHEA-S)*
  • Cortisol and cortisol/DHEA ratio*

Age-related changes in DHEA levels in adults with DS and the risk of dementia, Landt et al 2011 J Neuroendocrinol., 23, 450-455
Conclusions

• Complex picture of age-related risks and atypical ageing processes;

• The importance of identifying mechanisms – informed prevention and intervention;

• Complexity of need
Theoretical models for understanding challenging behaviour

Wider physical and emotional environment

Applied behavioural analysis

Delayed or atypical development

Triggering events
Setting conditions

Co-morbidities

Autism spectrum conditions
Behavioural phenotypes

Physical illness
Psychiatric illness
Sequela of abuse

FORMULATION: biological, psychological and social factors predisposing to, precipitating or maintaining such behaviours or abnormal mental state
Formulation

- Accepted models of understanding
- Evidence-base for different interventions
- History
- Examination
- Investigations
- Observations

Reason for referral

Intervention

Good Clinical Practice

Evidence-base for different interventions

FORMULATION

Reason for referral

Intervention