Developmental, Psychiatric & Psychological Aspects of Down Syndrome in Children & Young People

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Down Syndrome: current understanding

• intellectual disability
• characteristic personality & temperament
• relatively low rates of autism spectrum conditions & attention deficit hyperactivity disorders in childhood
• Depression from adolescence onwards
• Alzheimer disease from middle age onwards
Social Aetiologies (Emerson 2006)

- Mental health is socially determined
- Socio-economic position strongly associated with:
  - Child mortality
  - Adverse birth outcomes
  - Child physical health
  - Child mental health
  - Educational attainment
  - Life experiences & opportunities
- Poverty associated with mental disorder

• Investigation of prevalence & predictors of mental health needs & service use in adolescents with intellectual disabilities

• Prevalence of mental health needs increased from 51% by parental report to 67% as judged by clinical interview. Caseness associated with low adaptive functioning, autism diagnosis & family history of mental illness

• High mental health scores correlated with low adaptive functioning scores
Hassiotis & Turk, 2012: Conclusions

- Mental health challenges in adolescents with intellectual disabilities are
  - Common
  - Functionally impairing
  - Frequently unidentified & under-estimated
  - Frequently untreated
- They pose risk factors for further functional disadvantages, handicaps & social exclusion
DSM-5 Intellectual Disability

- Impairments of general mental abilities that impact adaptive functioning & determine how well everyday tasks are coped with: 3 domains
  - **Conceptual** domain: language, reading, writing, maths, reasoning, knowledge, memory
  - **Social** domain: empathy, social judgement, interpersonal communication skills, making & retaining friendships
  - **Practical** domain: personal care, job responsibilities, money, recreation, organising school & work tasks
- Diagnosis based on severity of deficits in adaptive functioning
Significant challenges in adaptive behaviours and life skills

• Self-care
• Self-occupancy
• Self-sufficiency
• Self-determination
• Safety
Intelligence

- Often minimal discrepancy in intellectual functioning in infancy
- Preschool: ↓ means-end thinking
- Infants: developmental challenges more evident
  - Mean IQ around 60, range approximately 25-70
- Early stimulation programmes may enhance development, at least temporarily
- Visuomotor skills & tactile discrimination particularly challenging
- ? Relative strengths in visual processing, receptive language & non-verbal social functioning
- ? Relative weakness in gross motor & expressive language skills
Parental Reports

• ↓ attentional focusing
• ↓ inhibitory control
• ↓ sadness
• ↓ repertoire of strategies for coping with frustration
• ↓ goal-directed strategies including assistance-seeking & cognitive self-soothing
School Years

• Visuospatial processing abilities exceed verbal ones

• Working memory, verbal short-term & long-term memory challenges

• ? Counter-productive strategies for novel problem-solving

• Association between IQ & parental academic & professional attainments
Childhood

• Levels of maladaptive behaviours less than expected
• Sibling emotional problems more common in presence of challenging behaviours
• Adaptive behaviours relatively preserved
Language

• Receptive skills exceed expressive ones
• Syntax particularly problematic
• High rates of pro-social behaviours
• Social strategies to engage conversational/interactional partners even to extent of distracting them from tasks in hand
Personality & Temperament

• Temperament = genetically inherited template
• Personality = interaction of temperament with social experiences
• Good-naturedness, contentedness, warm personality, interpersonal empathy
• Endearing yet sometimes challenging degrees of oppositionality, stubbornness & defiance
• Usually good mentalising & theory of mind abilities
• But emotional & behavioural challenges not unknown…
Emotions

• Rates of externalising behaviours low
• Internalising challenges increase through adolescence
• Increasing rates of depression
• But persisting good social skills
Attention Deficit-Hyperactivity Disorders

- Overactivity
- Poor concentration span
- Inattentiveness
- Restlessness
- Fidgetiness
- Impulsiveness
- Distractibility
- All-pervasive
ADHD DSM-5 Changes

• Recognises adult presentations
• Children: at least 6 items from inattention &/or hyperactive-impulsive group
• Adults: only 5 items needed
• Several symptoms must be present prior to 12 years
• No exclusion criteria for those with ASCs
• Nor indeed for those with intellectual disability
ADHD

• Reports of up to 40% i.e. probable substantial under-diagnosing

• Shorter & less frequent periods of sustained attention in toddlers

• Gross motor hyperactivity ↓ with age

• But marked poor concentration span, inattentiveness, impulsiveness & distractibility can persist
ADHD Medications

- Methylphenidate, dexamphetamine
  - Immediate & modified release
- Atomoxetine
- Alpha 2 partial agonists – clonidine, guanfacine
- In extremis exceptionally low-dose risperidone
- N.B. need to monitor cardiovascular status, weight & height
- N.B. sleep & appetite adverse effects, nervous tics, emotional lability, paradoxical excitation, social withdrawal, accentuation of ASC traits
ASD Core Diagnostic Criteria

- Impairments in **reciprocal social interaction**
- Impairments in **language & communication**
  - Receptive & expressive
  - Verbal & non-verbal
- **Repetitive & stereotypic behaviours & interests**
  - Gross motor
  - Abnormal obsessional interests
  - Insistence on routine & sameness
- **Lack of imaginary & symbolic skills**
- **Multiple sensory sensitivities: fascinations, aversions**
DSM-5 Autism Spectrum Disorder

• Single diagnostic term
• Communication deficits
  ▪ Receptive & expressive
  ▪ Verbal & non-verbal
• Social impairments
• Obsessional interests, behaviours, routines & insistence on sameness
• Features from early childhood even if not recognised until later
Autism Spectrum

- 10-20%

- ? Masked by intellectual disability (diagnostic overshadowing) & personality profile

- ? Odd & bizarre streotypies, anxiety & social withdrawal

- Risk Factors
  - Seizures
  - Early hypothyroidism
  - Post-cardiac surgery complications
  - Lower IQ
  - Family history of broad ASC phenotype
Mood Disorders

• Prevalence of depressive disorders ↑ with age

• Even when influences of adverse life events & daily hassles accounted for

• ? Biological & behavioural features more common than cognitive & emotional content
Sleep

• Frequently problematic especially with biological challenges (e.g. sleep apnoea) or developmental ones (ADHD, ASC)

• Parental stress, poor parent-child relationships, family cohesion fragmentation

• ↑ bedtime resistance, sleep anxiety, somnambulism, parasomnias, sleep disordered breathing, daytime somnolence
Carter 2009

- 66% rarely fell asleep in their own beds
- 55% always restless during sleep
- 40% woke at least once during night
- 78% tired during day
- Sleep hygiene measures & positive behavioural support
- Melatonin, clonidine
- Not a fan of sedating antihistamines
Interventions & support

• Psychoeducation, family support, DSA
• Functional behavioural assessments, positive behaviour support, applied behaviour analysis, adapted CBT
• Work on self-esteem, sense of self-worth, problem-solving & anger management repertoire
• Speech & language therapy
• Occupational therapy including sensory integration work
• Occasional judicious temporary use of low-dose medication