Mental health difficulties in children with learning disabilities

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The problems

• Substantial impairment of adaptive behaviour (day to day support for ADL) and limited speech\ expressive communication

• What is included under ‘mental health’
  – Challenging behaviour
  – ‘Comorbid’ ASD and ADHD
  – Impulsivity and repetitive behaviour

• The ‘value’ of psychiatric taxonomy
  – Grossly atypical development of CNS
  – Additional impairment

• Aetiology and psychiatric taxonomy
  – VCFS, TSC, PWS (UPD) but... ASD in FXS, RTS and CdLS, anxiety in WS

• Diagnostic criteria
  – Speech (language) and thought
  – Identify, label, report
  – Measure

• In combination these factors militate against identification of a mental health problem
Outline

• Cause matters
• Physical health (pain, discomfort, sleep)
• Learned behaviour
• ADHD and ASD
• Anxiety and low mood
The normal distribution of IQ scores and the basis to the two group approach
Some genetic syndromes associated with intellectual disability

Significant loss or change of genetic information caused by:

- Numerical chromosome abnormality (e.g. Down syndrome)
- Structural chromosome abnormality (e.g. Cornelia de Lange, Angelman, Prader-Willi syndromes)
- Single gene disorders (e.g. Fragile X syndrome)

<table>
<thead>
<tr>
<th>Aarskog</th>
<th>Cornelia de Lange</th>
<th>Perlman</th>
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<tbody>
<tr>
<td>Addison-Schilder</td>
<td>Cowchock</td>
<td>Pitt-Rogers-Danks</td>
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<td>Aicardi syndrome</td>
<td>Cri du chat</td>
<td>Prader-Willi</td>
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<td>Di George</td>
<td>Rett</td>
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<td>Allan-Herndon-Dudley</td>
<td>Down's</td>
<td>Richner-Hanhart</td>
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<td>Aspartylglycosaminuria</td>
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<td>Kabuki make-up</td>
<td>Usher</td>
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<td>Kallmann</td>
<td>Watson</td>
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<td>Carpenter</td>
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<td>Williams</td>
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<td>Cat eye</td>
<td>Lowe</td>
<td>Wolcott-Rallison</td>
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<td>CHARGE</td>
<td>Mandibulofacial dysostosis</td>
<td>Wrinkly skin</td>
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<tr>
<td>Christian syndrome</td>
<td>Marsidi</td>
<td>Zinsser-Engman-Cole</td>
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<tr>
<td>Cleidocranial dysplasia</td>
<td>Marsidi</td>
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# Complexity and effects on the environment

<table>
<thead>
<tr>
<th></th>
<th>Self-injury</th>
<th>Temper outbursts</th>
<th>Motivation for social contact</th>
<th>Sleep disorder</th>
<th>Impulsivity</th>
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<tbody>
<tr>
<td>Smith-Magenis</td>
<td>+++</td>
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<td>Angelman</td>
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<td>+++</td>
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<tr>
<td>Prader-Willi</td>
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<tr>
<td>Cornelia de Lange</td>
<td>++</td>
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<tr>
<td>Cri du Chat</td>
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</tbody>
</table>
Outline

• Cause matters
• Physical health (pain, discomfort, sleep)
• Learned behaviour
• ADHD and ASD
• Anxiety and low mood
Cornelia de Lange syndrome: Self-injurious behaviour, gastrointestinal disorders, middle ear infections, dental abnormalities and disorders

Reflux related behaviours in CdLS

(p<.01)
Pain and discomfort

- More pain behaviours in those showing self-injury?
- Higher prevalence of self-injury in those with health problems?
- More self-injury in those with suspected health problems

Comparison of pain behaviours in children with Tuberous Sclerosis Complex

(U=27; p<.001)

Kate Eden, Cerebra PhD studentship holder
Pain and discomfort

- More pain behaviours in those showing self-injury?
- Higher prevalence of self-injury in those with health problems?
- More self-injury in those with suspected health problems

Relative risk of frequent self-injury in children and adults with ASD given the presence of health problems

(99% CI)

Caroline Richards, PhD
Pain and discomfort

- The assessment of pain
  - FLACC (Merkel et al., 1997)
  - NCCPC (Breau et al., 2004)
  - QABF (Paclawskyi et al., 2000)

- Behavioural correlates of pain and challenging behaviour:
  - More pain behaviours in those showing challenging behaviour?
  - Higher prevalence of challenging behaviour in those with health problems?
  - More challenging behaviour in those with suspected health problems

- Social/operant causes and pain behaviour?

- New directions in assessment
  - Temporal relationships

www.findresources.co.uk

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Pain and discomfort

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- Social\operant causes and pain behaviour?
- New directions in assessment
  - Temporal relationships

Other outrageously expensive smartphones are also available

Kate Eden, Cerebra PhD studentship holder
“(……..) only has challenging behaviours when in pain. This results in a complete change in personality, ripping lumps of hair out so massive patches are missing. screaming like a banshee.

But we are not believed at hospital and just get sent home as they see no fever, no infections, ears, eyes, teeth, skin, joints. And refuse to do anything even basic bloods or x-rays. We then have to go to our community consultant who found that acid reflux had burned her severely and finally got meds needed. The hospital telling us that she had nothing wrong and it was behavioural or neurological. ……..

Is this pain tool going to be any use to use if no one listens?”

Parent of a child with Angelman Syndrome
“There are at least 33 syndromes of learning disabilities where a behavioural phenotype has been reported…….. the mechanism by which a genetic disorder could cause ……. behaviours is largely unknown, the ultimate pathway must be the structure and the function of the brain. Most of these behaviours are not curable…………”

Psychiatry text published in 1996
Smith-Magenis Syndrome does sleep

Just not when or where we want!
Smith Magenis syndrome

- Prevalence estimates of 1 in 25,000 births (Greenberg et al., 1991) to 1 in 15,000 (Laje et al., 2010)
- Deletion chromosome 17 p11.2 (Greenberg et al., 1991; Smith et al., 1986) or mutation (gene RAI1) (Slager et al., 2003)
## Effects of treating reflux on self-injury, Mood, Pain & Sleep

<table>
<thead>
<tr>
<th></th>
<th>Pre-Treatment 1</th>
<th>Post-Treatment 1</th>
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</thead>
<tbody>
<tr>
<td><strong>Challenging Behaviour Interview</strong></td>
<td>31/55</td>
<td>28/55</td>
</tr>
<tr>
<td><strong>Frequency</strong></td>
<td>Daily</td>
<td>Daily</td>
</tr>
<tr>
<td><strong>Worst Effect</strong></td>
<td>Moderate injury (bruising, cuts, abrasions)</td>
<td>Moderate injury (bruising, cuts, abrasions)</td>
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<tr>
<td><strong>Mood, Interest and Pleasure Questionnaire</strong></td>
<td>30</td>
<td>36</td>
</tr>
<tr>
<td><strong>Gastroesophageal Distress Questionnaire Total Score</strong></td>
<td>45</td>
<td>39</td>
</tr>
<tr>
<td><strong>FLACC (average across 5 days)</strong></td>
<td>4</td>
<td>0.6</td>
</tr>
<tr>
<td><strong>Total Sleep Time (average across 5 days)</strong></td>
<td>06:49:24</td>
<td>07:15:00</td>
</tr>
<tr>
<td><strong>Number of Night Wakings (average across 5 days)</strong></td>
<td>1.8 (range = 1-4)</td>
<td>1.4 (range = 1-2)</td>
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<tr>
<td><strong>Total Waking Duration</strong></td>
<td>01:35:00 (range = 00:10 – 06:05)</td>
<td>00:15:00 (range = 00:05 – 00:30)</td>
</tr>
</tbody>
</table>
Outline

• Cause matters
• Physical health (pain, discomfort, sleep)
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• ADHD and ASD
• Anxiety and low mood
Social Communicative Function of Challenging Behaviour: Positive Reinforcement

Need for others to do or give something → SIB → AVERSIVE!

Increase in chance of CB → REWARD

Positive Reinforcement

ENGAGE

Comfort
Reprimand
Offer
Restrain
Occupy
Distract

Concern
Frustration
Anxiety
Confusion
Distress

ACTION

Increase in chance of CB → REWARD
Outline

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<table>
<thead>
<tr>
<th>Syndrome</th>
<th>ASD</th>
<th>Autism</th>
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<tbody>
<tr>
<td>Angelman</td>
<td>66.3</td>
<td>17.8</td>
</tr>
<tr>
<td>(15q11-q13)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cri du Chat</td>
<td>40.0</td>
<td>8.0</td>
</tr>
<tr>
<td>(5p 15.2-15.3)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cornelia de Lange</td>
<td>78.8</td>
<td>45.9</td>
</tr>
<tr>
<td>(5p 13.1)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fragile X</td>
<td>83.6</td>
<td>46.3</td>
</tr>
<tr>
<td>(Xq27.3)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prader-Willi</td>
<td>45.8</td>
<td>15.5</td>
</tr>
<tr>
<td>(15q11-q13)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lowe</td>
<td>71.2</td>
<td>34.6</td>
</tr>
<tr>
<td>(Xq26.1)</td>
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<td></td>
</tr>
<tr>
<td>Smith Magenis</td>
<td>68.4</td>
<td>36.8</td>
</tr>
<tr>
<td>(17p 11.2)</td>
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</tr>
</tbody>
</table>

Age range 4 to 54

+ indicates score higher than 1 other group, - indicates score lower than 1 other group, O indicates no difference from any other group.

Oliver, C. et al. (2011). *JADD*, 41, 1019-1032
Autism Spectrum Disorder (ADHD) or not?

• Behaviourally defined, list of criteria
  – Attaining cut-off scores but with different item level profiles
  – Scoring on an item for different reasons
  – Communication problems
• Is the diagnosis helpful?
  – Services
  – Good advice from Autism (ADHD) materials
Outline

• Cause matters
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• Anxiety and low mood
Difference of emotion
BOILING, ABUSIVE, SWEARING, SCREAMING, HITTING OUT
SIMMERING: SHOUTING, THREATS, SHAKING, SWEATING
NOT LISTENING
LUKE WARM: BECOMING ANXIOUS
ANXIETY: RISING, RAISED VOICE
COLD, CALM, HAPPY, AND USUAL SELF

STATEGIES FOR CALMING DOWN

Play music

Outline

• Cause matters
• Physical health (pain, discomfort, sleep)
• Learned behaviour
• ADHD and ASD
• Anxiety and low mood
Prevalence of anxiety and depression in people with intellectual disabilities

24% of young people and children with ID experience mental health difficulties

Anxiety and depression and mixed affective disorder are the most common diagnoses.

3-22% of children with intellectual disabilities have an anxiety disorder (Reardon et al., 2015).

Four to six times more likely to have an affective disorder (Taylor et al., 2004).
What increases the prevalence?

**Person characteristics**

- Information processing (rate and complexity).
- Reduced executive functioning: memory, inhibition, flexibility, problem-solving and planning, predicting the future.
- Low levels of adaptive skills

Altered processing of information i.e. attention to more threatening stimuli.

Fewer cognitive resources in the face of stressors

Hout et al., 2009; Taylor & Knapp, 2003
Intolerance of uncertainty.....

Interconnected set of neurobiological and psychological processes

Cognitive capacity to manage uncertainty is reduced

Boulter et al. (2014); Wigham et al. (2015)
Rare genetic syndromes (e.g. Williams, fragile-X or Cornelia de Lange syndromes)

Royston et al., 2016
Why are anxiety and depression more prevalent?

**Environment factors around the person**

- Boredom
- Loneliness
- Lack of opportunity to exert control over own life and the future
- Lack of meaningful friendships and relationships
- Stressful family circumstances
- Stigmatisation and bullying
- Being asked to complete tasks that are too difficult and opportunities being removed
- Unemployment
- Debt
- Chronic poverty

Matorell et al., 2009; MacMahon & Jahoda, 2008
Anxiety and depression remain undetected, and hence untreated

Therapeutic Disdain

Psychological therapies are ineffective with people with ID

Lack of clinical research in this area
Therapeutic overshadowing

Social withdrawal being seen as a lack of social skills rather than depression or anxiety.

Crying as an indicator of pain rather than depression.

Lack of engagement with activities as being due to intellectual disability.

Ethos of services focusing on challenging behaviour above mental health.
Therapeutic overshadowing

Inaccurate identification works both ways!

Social withdrawal being diagnosed as depression when it is Autism Spectrum Disorder

Pain rather than depression

Different interests and desires, rather than depression
Social anxiety in CdLS

- High levels of anxiety associated with social situations.
- Strong preference to observe rather than participate.
- Increased withdrawal when social demands become heightened.
- Motivation for social contact appears to be intact.

Moss et al., 2008. *AJMR 113*, 278-291;
Richards et al., 2009, *JADD*, 39, 1155.
Reid, Nelson, Moss & Oliver, In preparation
Inadequate assessment measures

Based on the general population. Different behaviours? (Fydrich et al., 1998; Rodgers et al., 2012)

Even less appropriate for people with severe intellectual disability

Do not interrogate behaviour change

Do not rule distinguish physical and emotional distress
<table>
<thead>
<tr>
<th>Changes to the body:</th>
<th>Changes to thoughts/thinking patterns:</th>
<th>Changes to emotions</th>
<th>Changes to behaviour</th>
</tr>
</thead>
<tbody>
<tr>
<td>fast and irregular heartbeat</td>
<td>inability to concentrate</td>
<td>irritability</td>
<td>avoiding situations</td>
</tr>
<tr>
<td>sweating</td>
<td>repetitive thoughts about perceived</td>
<td>feeling worried</td>
<td>fidgeting/moving more than usual</td>
</tr>
<tr>
<td>tiredness</td>
<td>threat</td>
<td>distress</td>
<td></td>
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<tr>
<td>muscle tension</td>
<td>concerns about losing control</td>
<td>crying</td>
<td></td>
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<tr>
<td>dizziness</td>
<td>inability to relax</td>
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<tr>
<td>trembling</td>
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<td>pale complexion</td>
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<td>stomach aches</td>
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<tr>
<td>nausea</td>
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</table>
Depression

- Activity Levels
- Social engagement
- Interest and pleasure
- Range of facial expressions
- Emotional expression
- Change???

- Eating
- Sleeping
- Self care skills/adaptive skills
The immediate environment of the person is a legitimate target for intervention

- Making environment more sensitive to the individual’s needs
- Increasing social engagement and engagement (or reducing this if it is not reinforcing to the person)
- Increasing engagement in meaningful activity that provide natural reinforcement
- Increasing an individual’s repertoire of functional skills
- Working with families and staff team to develop their awareness of mental health

Improving quality of life

Jones & Dowey, 2013
Increase controllability and choice!

Focus on communication

Give a choice

Carr et al. (2009)
Increasing certainty!
• Predictable routines
• Visual timetables
• Using a cue card when change occurs

Tolerance to uncertainty (building skills!)
- Scheduling something unpredictable
- Introducing subtle changes
- Skills to cope with stress

All anxiety has an avoidance component
There is a balance between managing anxiety and avoidance
Psychological therapy is not just ‘talking therapy’

- Relaxation Training

- Skills development (ways of coping a difficult situation)

- Graded exposure (Modelling/Rewards) for anxiety

- Behavioural activation for depression

Although Cognitive Behaviour Therapy may be appropriate for some people with ID

Chalfant *et al* 2007; Dagnan *et al*., 2005
Summary

Anxiety and depression are often overlooked in people with intellectual disability.

Individuals with intellectual disability experience more adverse life events and may be equip with fewer skills to manage these difficulties.

Assessment remains problematic due to difficulties with:
- Self-report and parental-report
- Confounded measurement tools (ASD, challenging behaviour, pain)

Assessing change from baseline is key.

A goal of psychological intervention is to increase access to activities and environments that encourage well-being, social contact and meaningful friendships.
Closing thoughts

- Cause matters (check syndrome information)
- Physical health (pain, discomfort, sleep)
- Learned behaviour
- ADHD and ASD (a pragmatic approach)
- Anxiety and low mood (underestimated?)
Core Funding
Cerebra

Grant Support
Medical Research Council
The Big Lottery
Baily Thomas Foundation
Cornelia de Lange Syndrome Foundation
Research Autism
Birmingham Children’s Hospital
Angelman Syndrome Foundation (USA)
Newlife
National Autistic Society
Economic and Social Research Council
Jerome Lejeune Fondation
Tuberous Sclerosis Association
NIHR
Leverhulme

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