The Cerebra Centre for Neurodevelopmental Disorders

Aston University

UNIVERSITY OF BIRMINGHAM

British Association for Community Child Health

Mental health difficulties in children with learning disabilities

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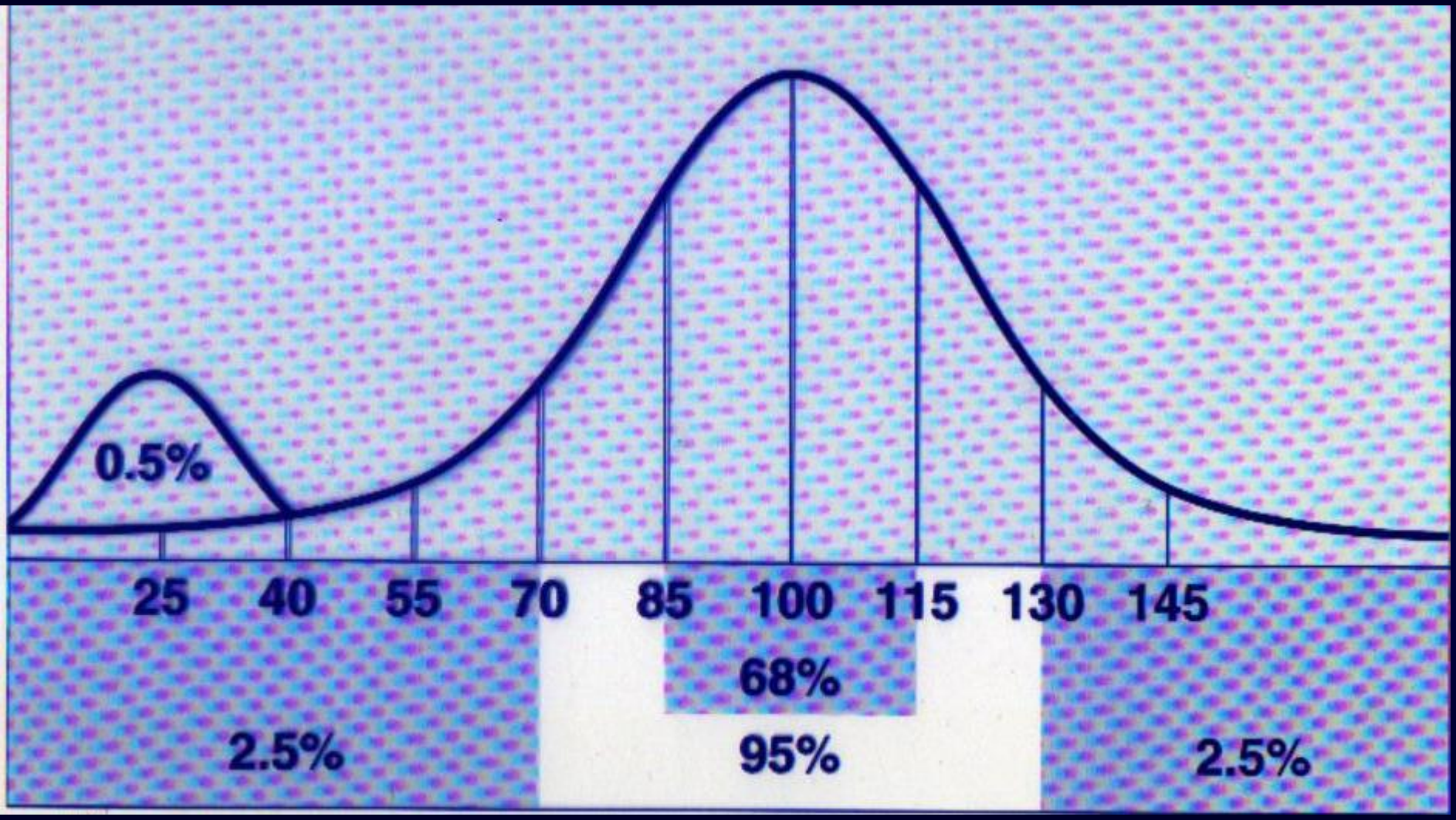
The problems

- 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)

- Aarskog
- Addison-Schilder
- Aicardi syndrome
- Alagille syndrome
- Allan-Herndon-Dudley
- Alpha thalassemia
- Alport
- Angelman
- Aspartylglycosaminuria
- Bardet-Biedl
- Beckwith-Weidemann
- Bertini
- Bickers-Adams
- Bloch-Sulzberger
- Brunner
- Cardiofacial
- Carpenter
- Cat eye
- CHARGE
- Christian syndrome
- Cleidocranial dysplasia
- Cohen
- Cornelia de Lange
- Cowchock
- Cri du chat
- Di George
- Down's
- Fragile X
- Fucosidosis
- Garcia-Lurie
- Goltz-Gorlin
- Greig-cephalopolysyndactyly
- Heterotaxia
- Hischsprung disease
- Hunter
- Hurler
- Kabuki make-up
- Kallmann
- Lesch-Nyhan
- Lowe
- Mandibulofacial dysostosis
- Marsidi
- Pateau
- Perlman
- Pitt-Rogers-Danks
- Prader-Willi
- Rett
- Richner-Hanhart
- Rieger
- Rubinstein-Taybi
- Rud
- Shprintzen
- Shprintzen-Goldberg
- Silver-Russell
- Smith-Magenis
- Snyder-Robinson
- Sotos
- Usher
- Watson
- Williams
- Wolcott-Rallison
- Wrinkly skin
- Zinsser-Engman-Cole

Complexity and effects on the environment

	Self-injury	Temper outbursts	Motivation for social contact	Sleep disorder	Impulsivity
Smith-Magenis	+++	+++	+++	+++	+++
Angelman	-	-	++	+++	++
Prader-Willi	+	+++	+	++	-
Cornelia de Lange	++	-	-	+	+
Cri du Chat	+	-	++	++	++

Outline

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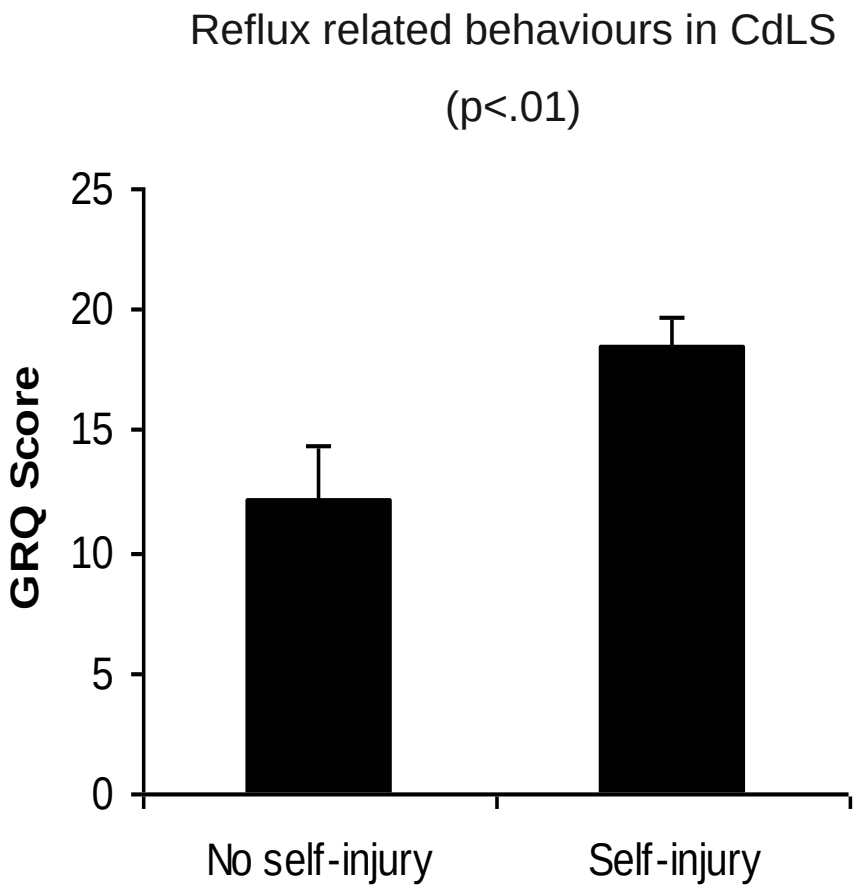
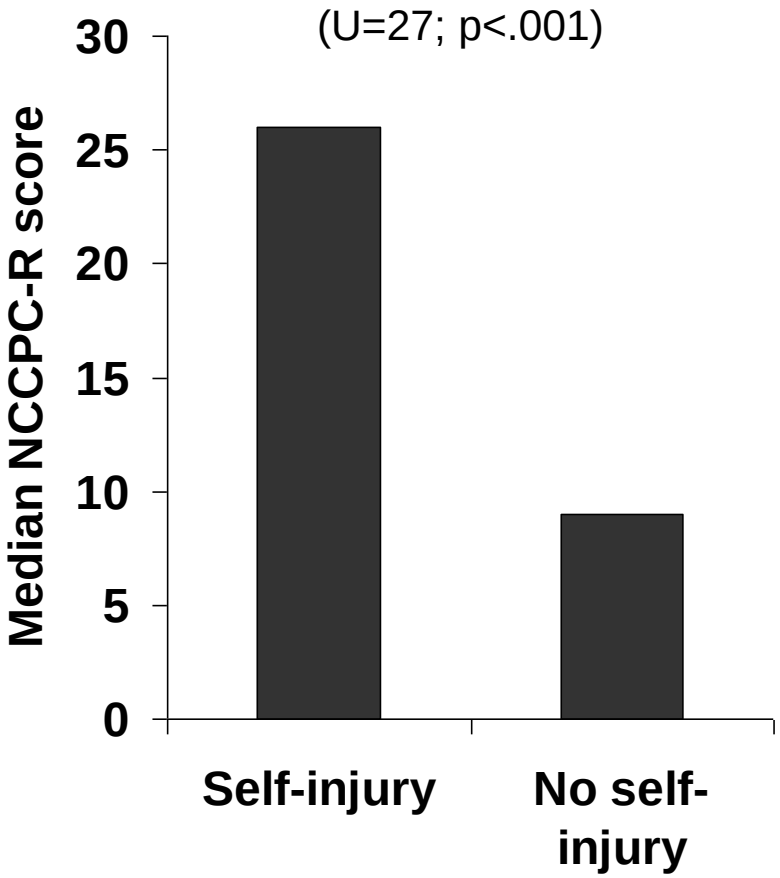
Cornelia de Lange syndrome: Self-injurious behaviour, gastro-intestinal disorders, middle ear infections, dental abnormalities and disorders



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



Kate Eden, Cerebra PhD studentship holder

Pain and discomfort

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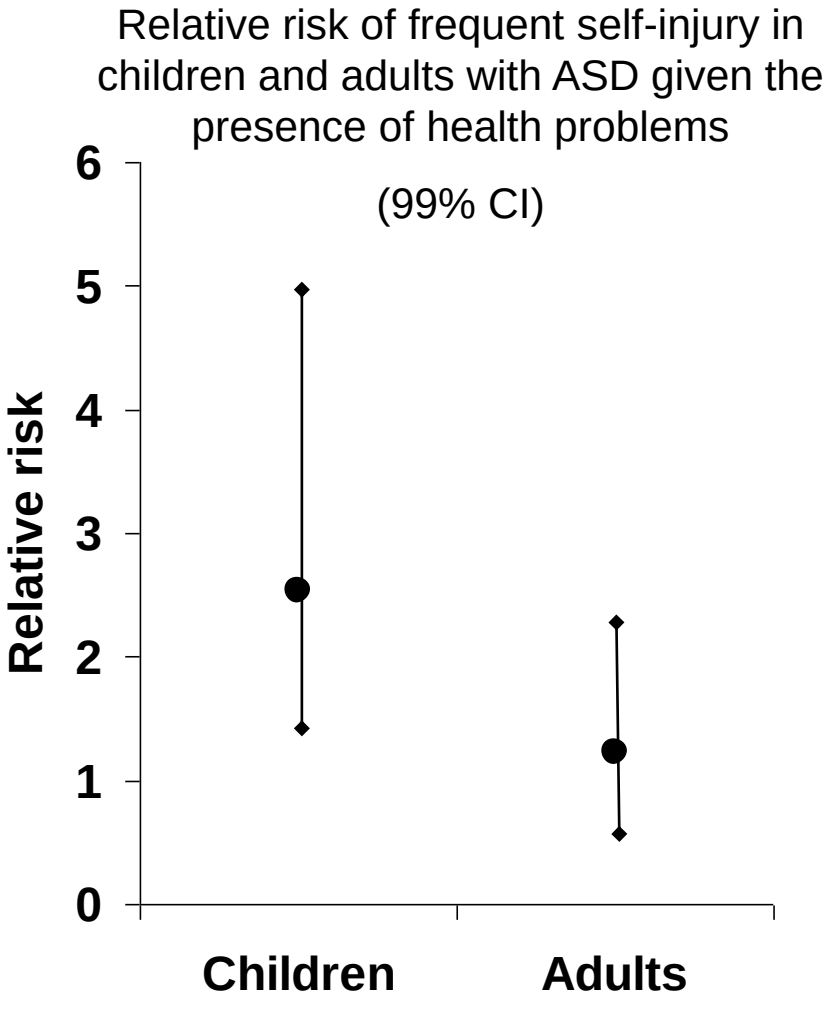
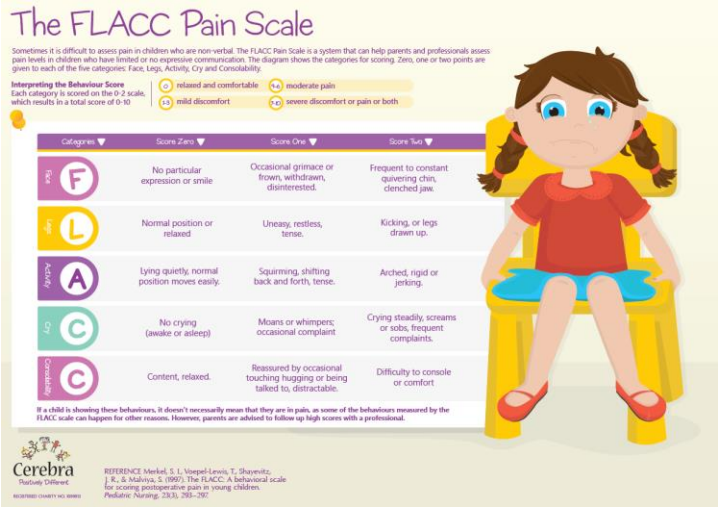


Pain and discomfort

- 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



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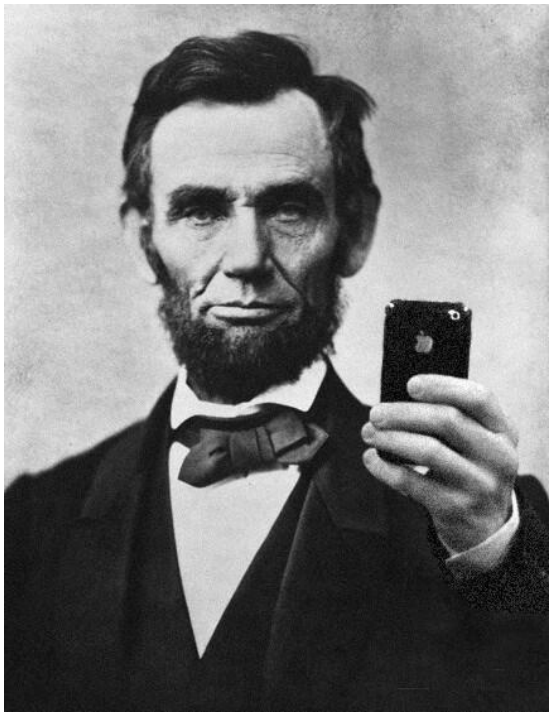


Other outrageously expensive smartphones are also available

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 <u>and it was behavioural or neurological. ……</u>

<u>Is this pain tool going to be any use to use if no one listens?”</u>

Parent of a child with Angelman Syndrome

Behavioural Phenotypes and Genetic Determinism

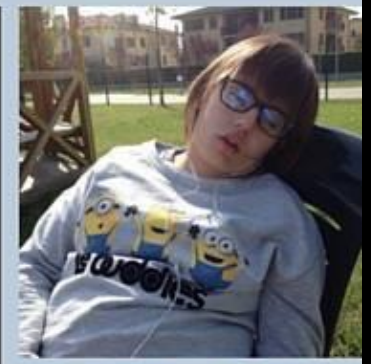
"There are at least 33 syndromes of learning disabilities where a behavioural phenotype has been reported……. the mechanism by which a genetic disorder could cause ……. behaviours is largely unknown, the ultimate pathway must be the structure and the function of the brain. Most of these behaviours are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

Psychiatry text published in 1996



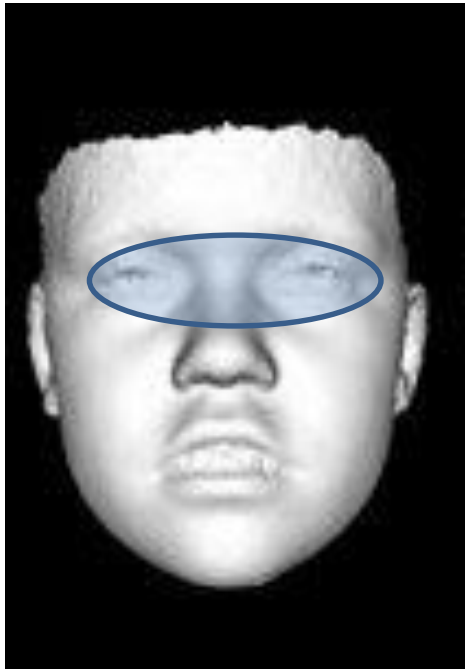

Smith-Magenis Syndrome does sleep

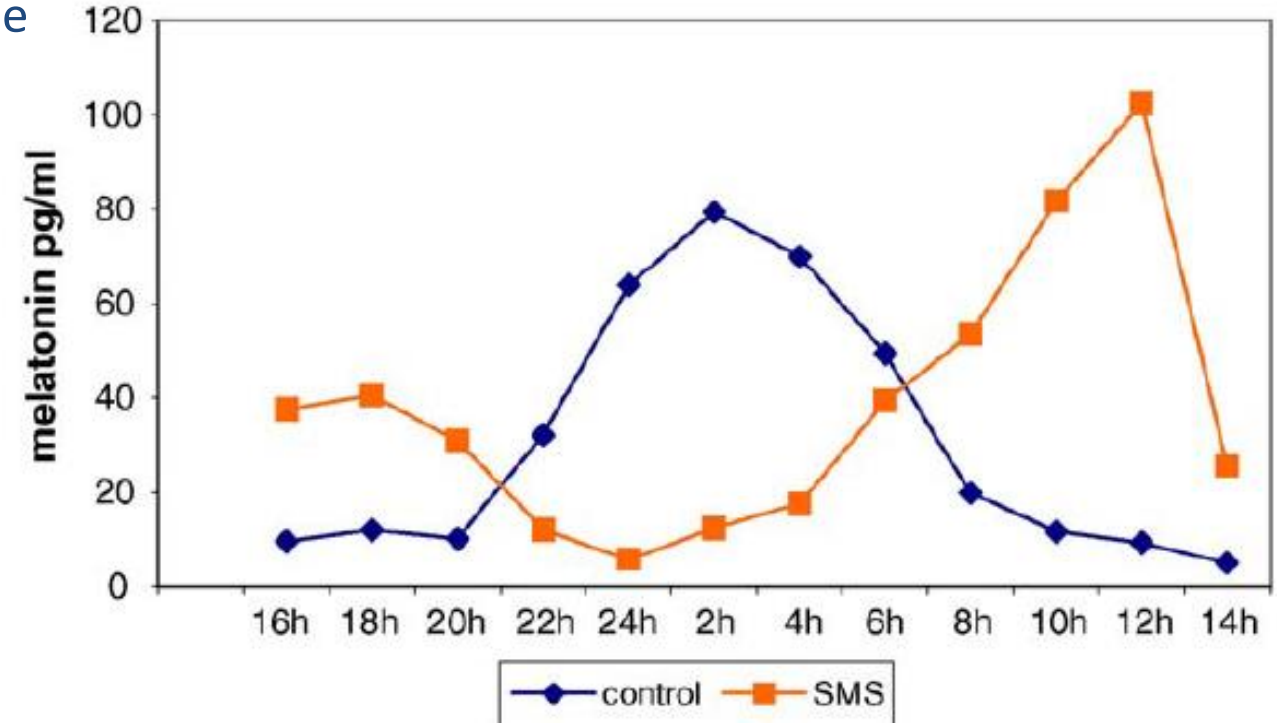
Just not when or where we want!






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

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

Outline

- Cause matters
- Physical health (pain, discomfort, sleep)
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Social Communicative Function of Challenging Behaviour: Positive Reinforcement

Need for others to do or give something

SIB

AVERSIVE!

Concern
Frustration
Anxiety
Confusion
Distress

ACTION

ENGAGE

Comfort
Reprimand
Offer
Restrain
Occupy
Distract

REWARD

Positive Reinforcement

Increase in chance of CB

Outline

- Cause matters
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Autism Screening Questionnaire

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

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

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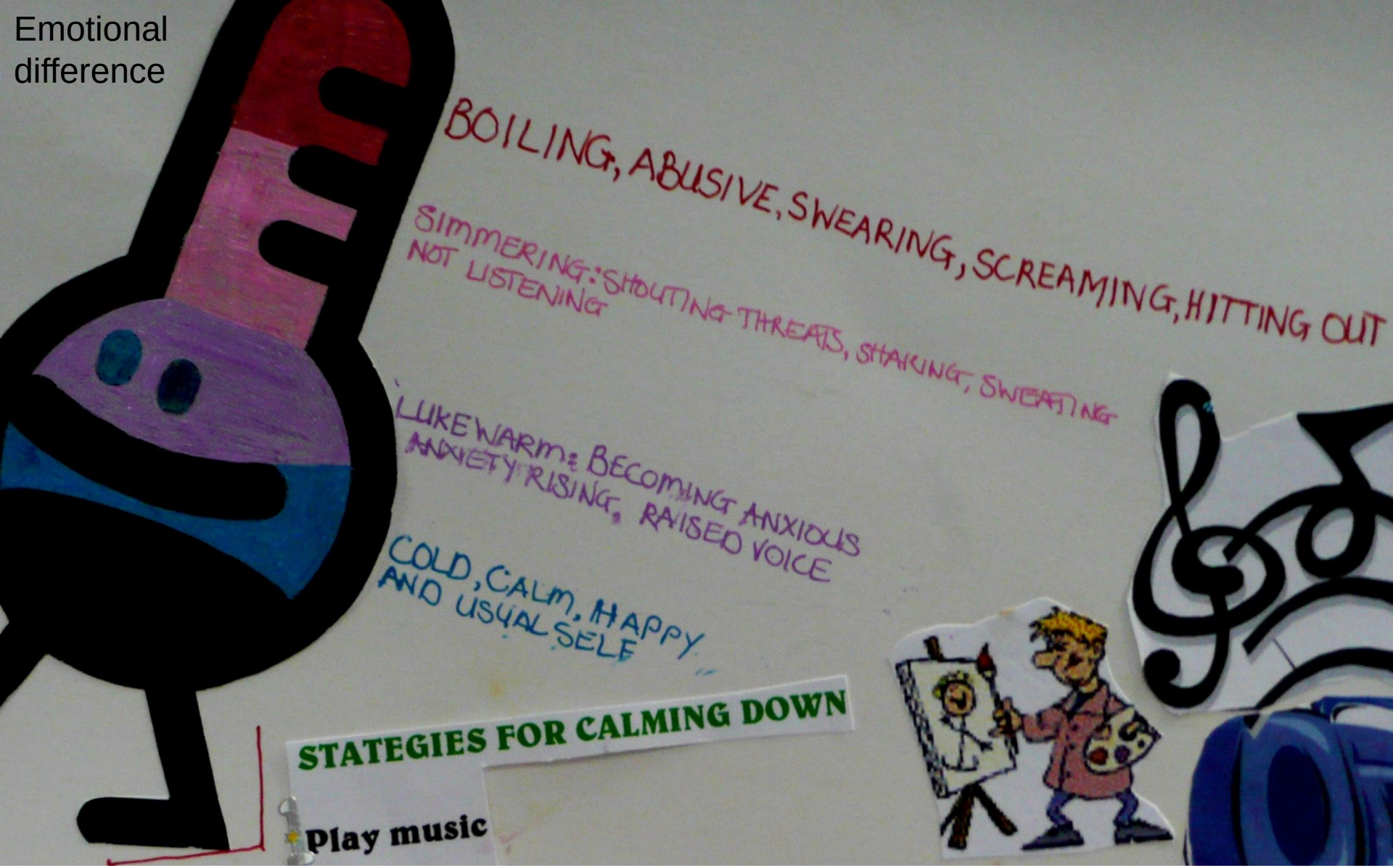
Difference of emotion

Emotional difference



Thornton, F. and Matthews, P. (2008). Addressing the balance. 1st Asia Pacific Prader-Willi syndrome conference. Wellington, New Zealand.

Outline

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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.

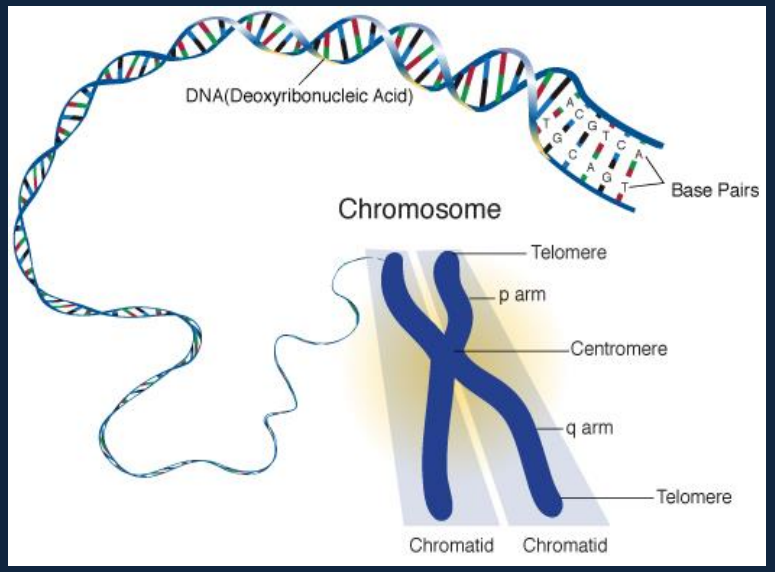
3-22% of children with intellectual disabilities have an anxiety disorder (Reardon et al., 2015).

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

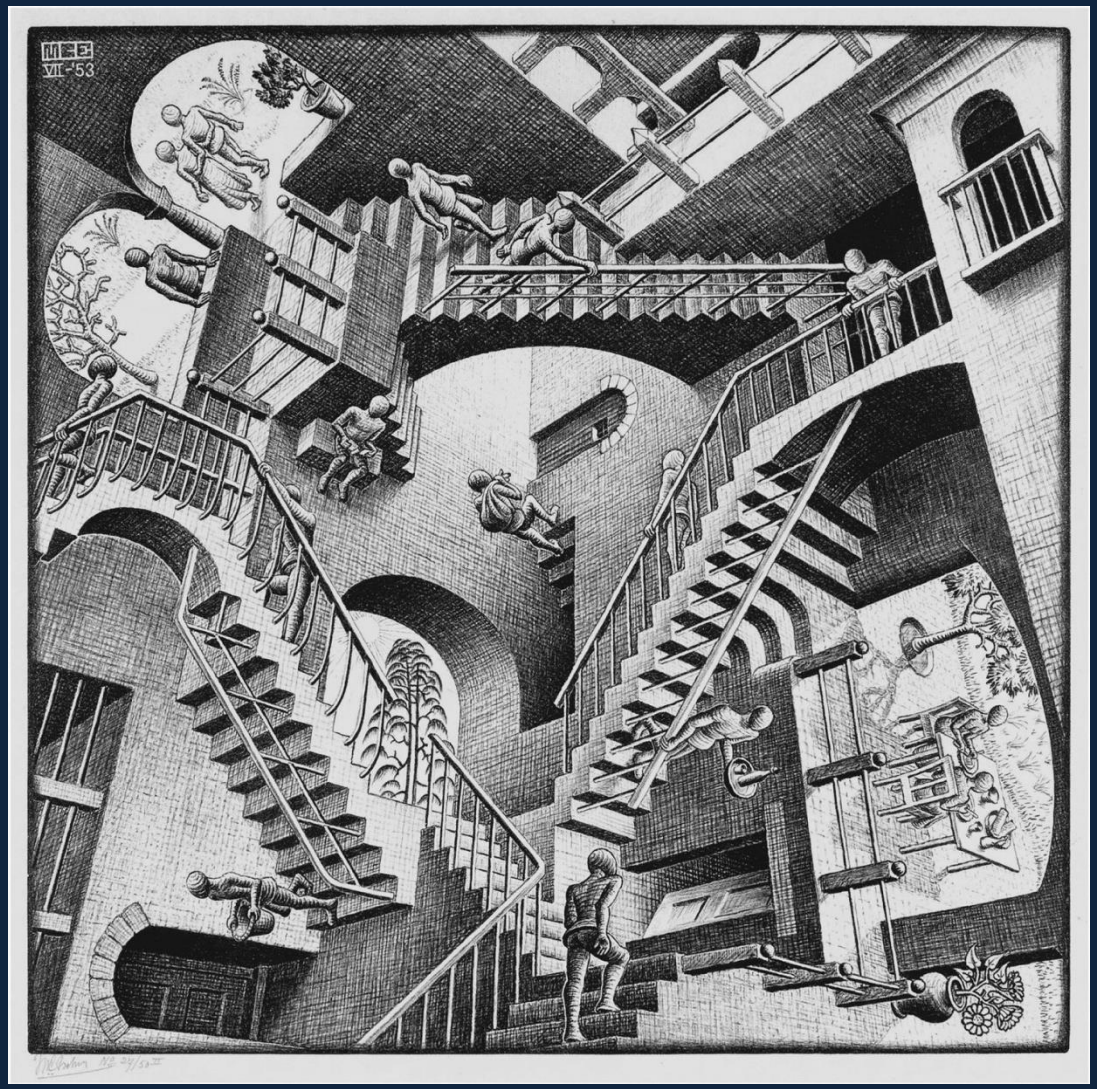


Altered processing of information i.e. attention to more threatening stimuli.

Fewer cognitive resources in the face of stressors

Hout et al., 2009; Taylor & Knapp, 2003

Intolerance of uncertainty.....

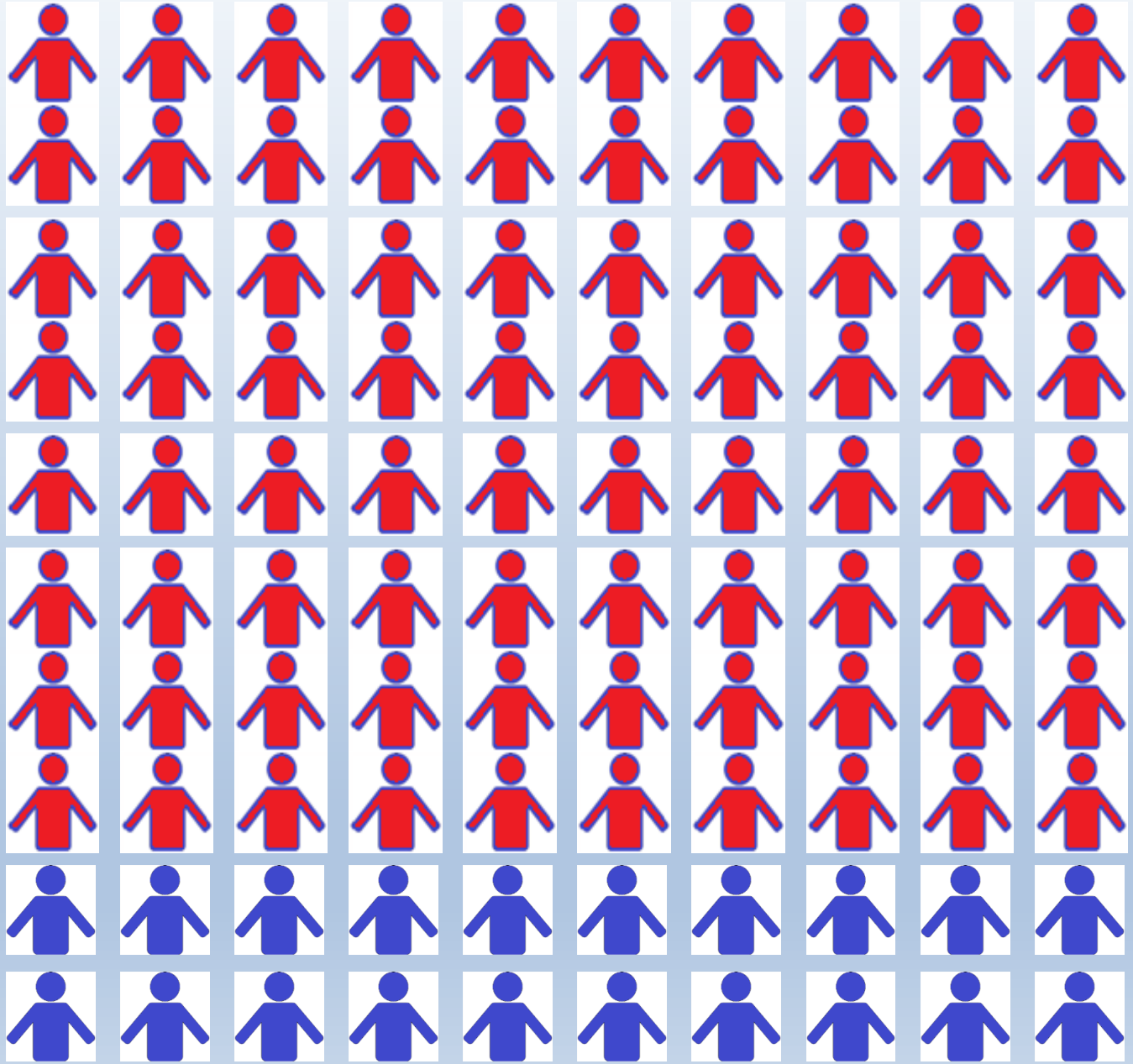


Interconnected set of neurobiological and psychological processes

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

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

Pain rather than depression

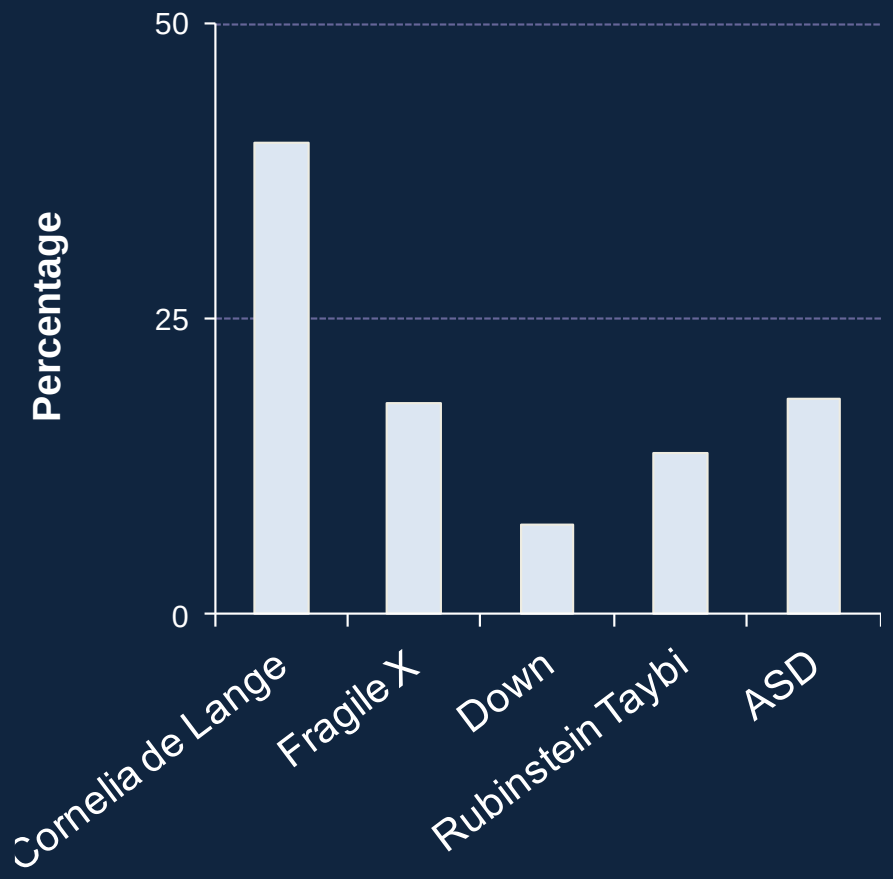
Different interests and desires, rather than depression

Inaccurate identification works both ways!

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.**

Prevalence of Selective Mutism



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)

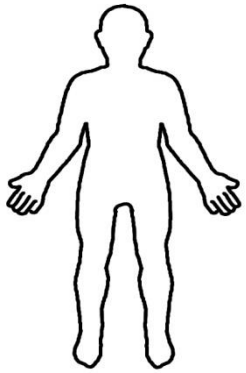
Even less appropriate for people with severe intellectual disability

Do not interrogate behaviour change

Do not rule distinguish physical and emotional distress

CHANGE IS KEY

Anxiety



Changes to the body:	Changes to thoughts/ thinking patterns:	Changes to emotions	Changes to behaviour
• fast and irregular heartbeat • sweating • tiredness • muscle tension • dizziness • trembling • pale complexion • stomach aches • nausea	• inability to concentrate • repetitive thoughts about perceived threat • concerns about losing control • inability to relax	• irritability • feeling worried • distress • crying	• avoiding situations • fidgeting/ moving more than usual

Depression

Activity Levels

Eating

Range of facial expressions

Social engagement

Sleeping

Emotional expression

Interest and pleasure

Self care skills/adaptive skills

Change???

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

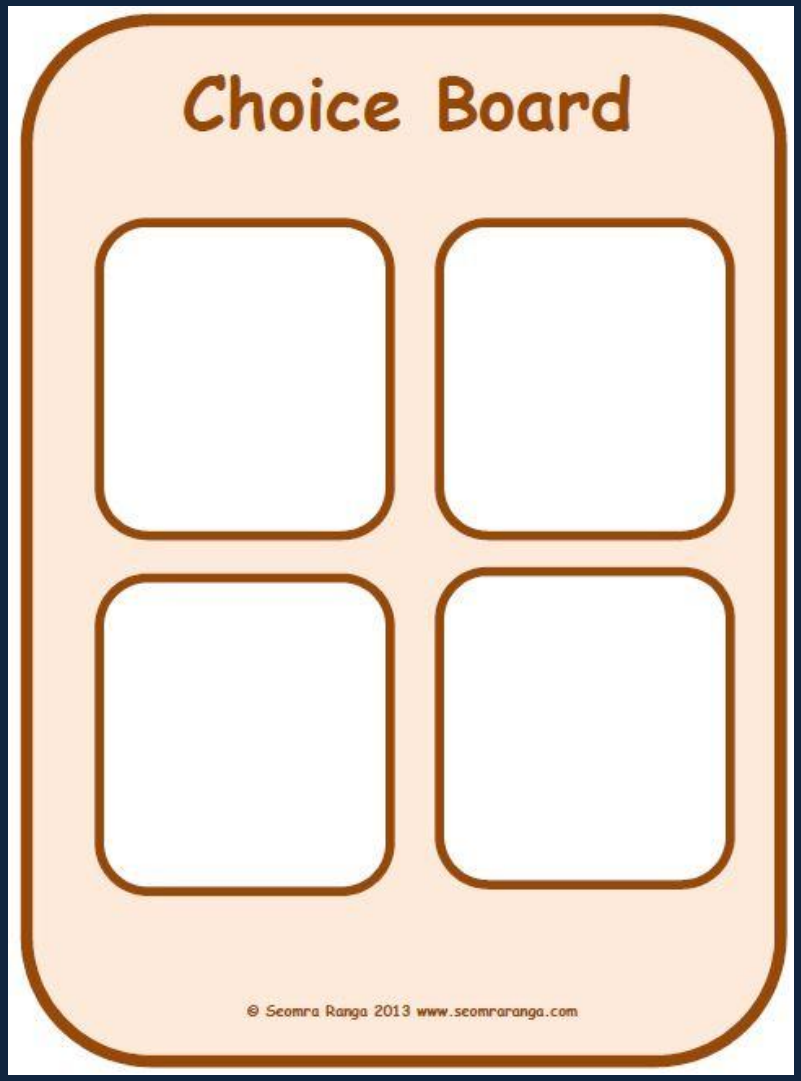
Improving quality of life

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

All anxiety has an avoidance component
There is a balance between managing anxiety and avoidance

lerance to
uilding

-Scheduling something unpredictable
-Introducing subtle changes
-Skills to cope with stress

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?)

The Cerebra Centre for Neurodevelopmental Disorders

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Core Funding

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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
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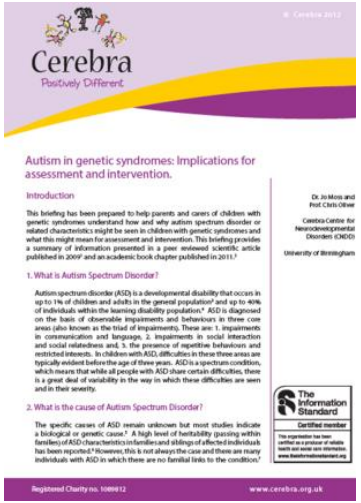

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Autism in genetic syndromes: Implications for assessment and intervention.

Introduction

This briefing has been prepared to help parents and carers of children with genetic syndromes understand how and why autism spectrum disorder or related characteristics might be seen in children with genetic syndromes and what this might mean for assessment and intervention. This briefing provides a summary of information presented in a peer reviewed scientific article published in 2009[4] and an academic book chapter published in 2011.[5]

1. What is Autism Spectrum Disorder?

Autism spectrum disorder (ASD) is a developmental disability that occurs in up to 1% of children and adults in the general population[6] and up to 40% of individuals within the learning disability population.[4] ASD is diagnosed on the basis of observable impairments and behaviours in three core areas (also known as the triad of impairments). These are: 1. impairments in communication and language, 2. impairments in social interaction and social relatedness and, 3. the presence of repetitive behaviours and restricted interests. In children with ASD, difficulties in these three areas are typically evident before the age of three years. ASD is a spectrum condition, which means that while all people with ASD share certain difficulties, there is a great deal of variability in the way in which these difficulties are seen and in their severity.

2. What is the cause of Autism Spectrum Disorder?

The specific causes of ASD remain unknown but most studies indicate a biological or genetic cause.[7] A high level of heritability (passing within families) of ASD characteristics in families and siblings of affected individuals has been reported.[8] However, this is not always the case and there are many individuals with ASD in which there are no familial links to the condition.[7]

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