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UNIVERSITY OF BIRMINGHAM

Understanding behaviour in Smith-Magenis syndrome

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UK

Our starting point

- *Everyone is unique and each child and adult with Smith-Magenis syndrome is unique.
- In many, many ways children and adults with Smith-Magenis syndrome are just the same as everyone else and have the same needs, wants and rights as everyone else.
- In some ways children and adults with Smith-Magenis syndrome differ from people who do not have Smith-Magenis syndrome.
- We shall look at these differences but we will not forget the similarities or that everyone is unique.

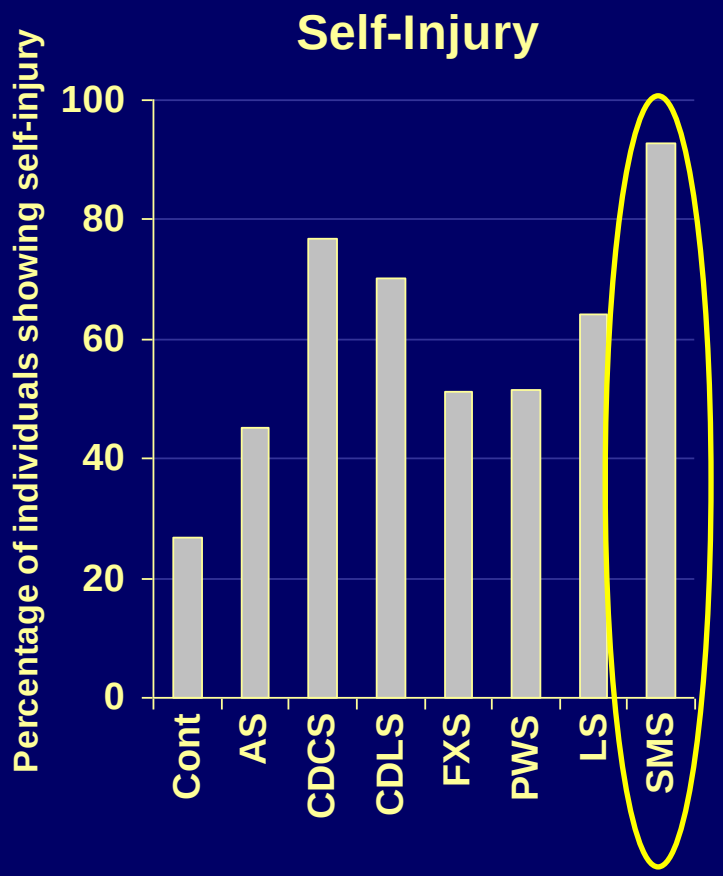
Engaging and sociable
Ability to form strong bonds
Memory and capacity to learn
Computer skills

Behaviour and Smith-Magenis Syndrome

- Self-injurious behaviour, aggression
- Impulsivity and temper outbursts
- Sleep disorder
- Autism (?) and social behaviour
- Resources

Self-injurious behaviour and aggression

Prevalence of Self-Injury and Physical Aggression in Syndromes



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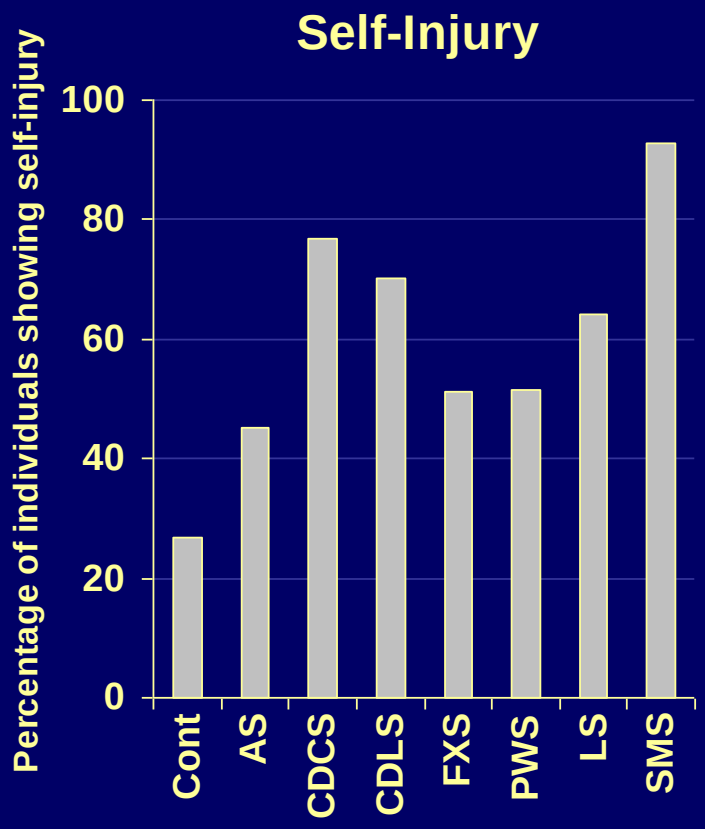
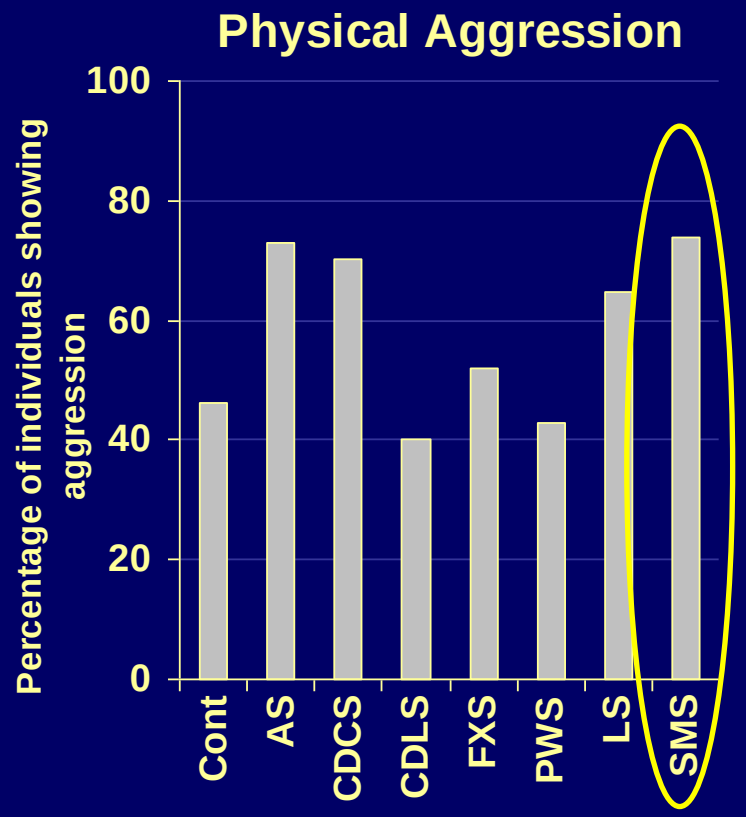
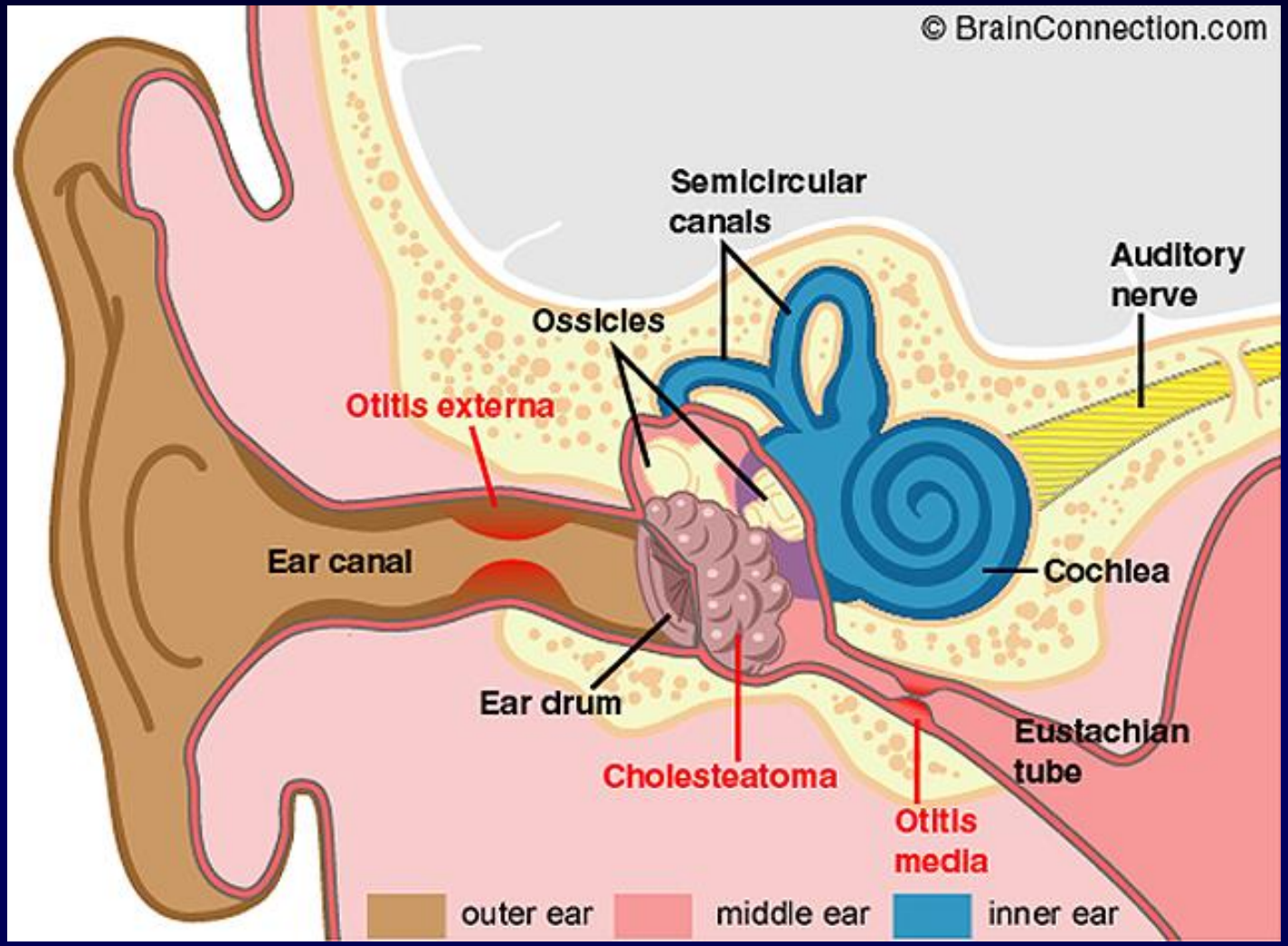
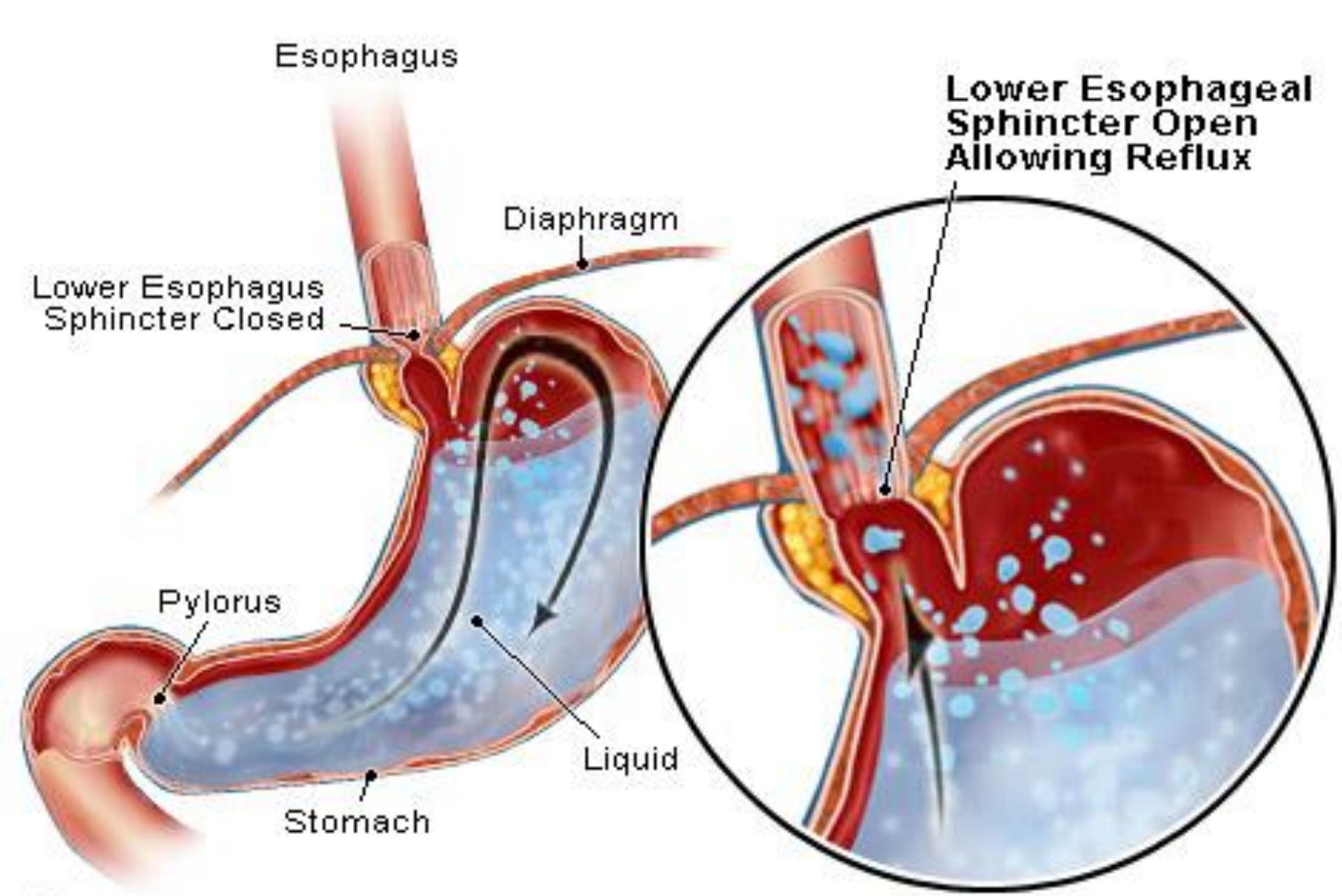


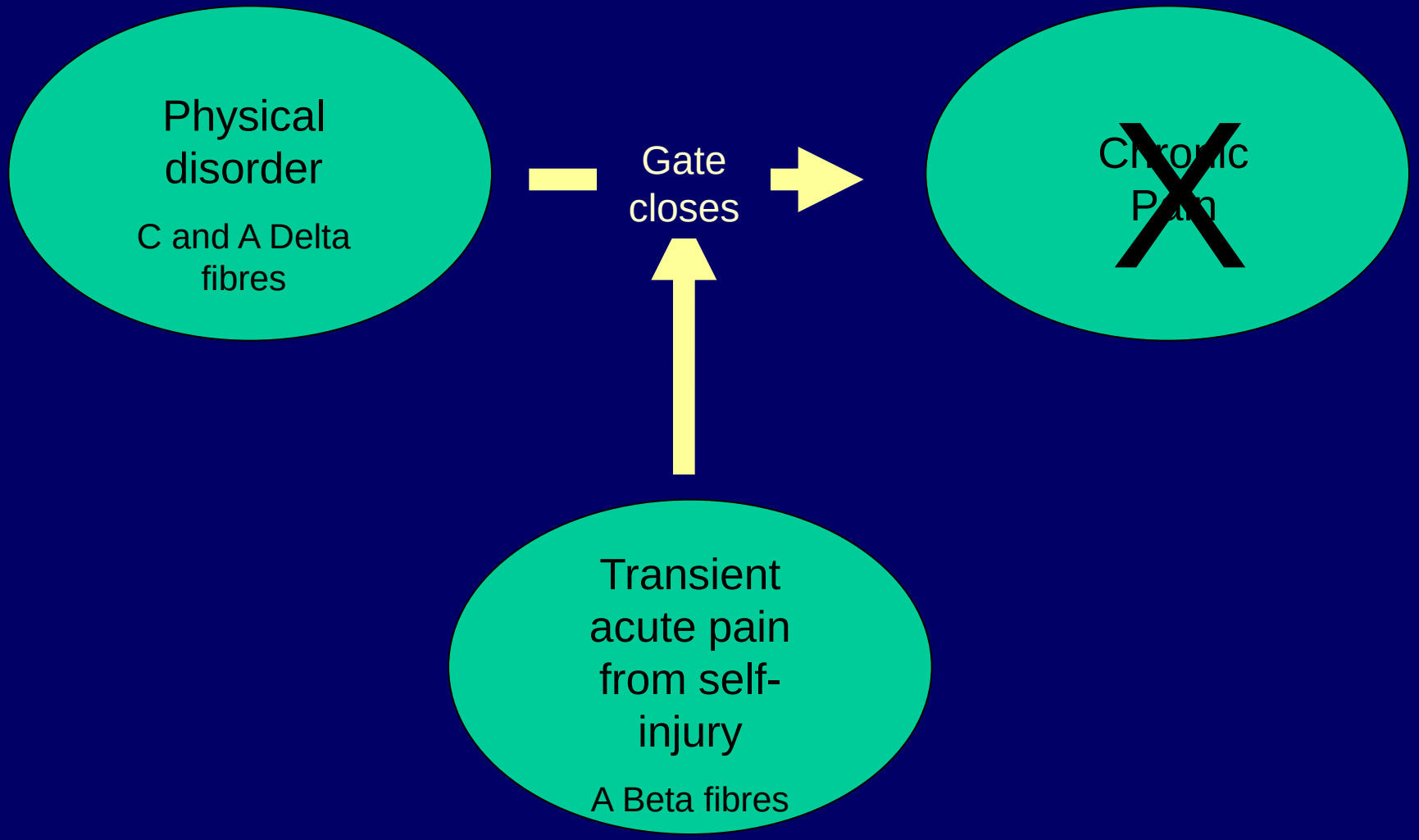




Recurrent middle ear infections are associated with headbanging



Gastroesophageal Reflux



Pain gate theory and learning to self-injure

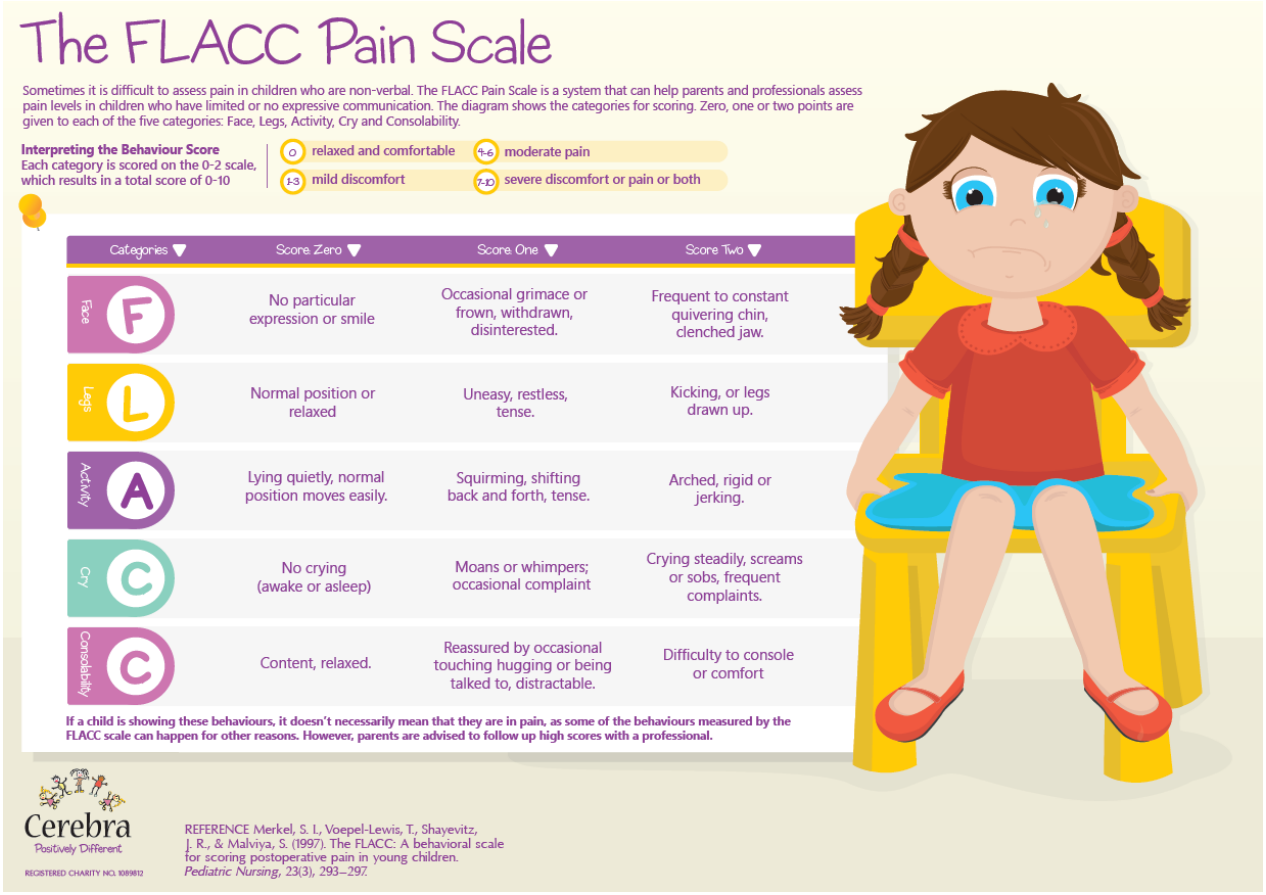
The pain cycle and self

1. Chronic or sharp pain is caus… …on or trauma
2. The child blocks the … …ing or hitting.
3. The hard rub… …ing leads to tissue damage …
4. Go t…

Early intervention and aggressive treatment

Pain signatures

- ***Face**
- **Legs**
- **Activity**
- **Crying**
- **Consolability**



The FLACC Pain Scale

Sometimes it is difficult to assess pain in children who are non-verbal. The FLACC Pain Scale is a system that can help parents and professionals assess pain levels in children who have limited or no expressive communication. The diagram shows the categories for scoring. Zero, one or two points are given to each of the five categories: Face, Legs, Activity, Cry and Consolability.

Interpreting the Behaviour Score
Each category is scored on the 0-2 scale, which results in a total score of 0-10

0 relaxed and comfortable
1-3 mild discomfort
4-6 moderate pain
7-10 severe discomfort or pain or both

Categories	Score Zero	Score One	Score Two
Face (F)	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested.	Frequent to constant quivering chin, clenched jaw.
Legs (L)	Normal position or relaxed	Uneasy, restless, tense.	Kicking, or legs drawn up.
Activity (A)	Lying quietly, normal position moves easily.	Squirming, shifting back and forth, tense.	Arched, rigid or jerking.
Cry (C)	No crying (awake or asleep)	Moans or whimpers; occasional complaint	Crying steadily, screams or sobs, frequent complaints.
Consolability (C)	Content, relaxed.	Reassured by occasional touching hugging or being talked to, distractable.	Difficulty to console or comfort

If a child is showing these behaviours, it doesn't necessarily mean that they are in pain, as some of the behaviours measured by the FLACC scale can happen for other reasons. However, parents are advised to follow up high scores with a professional.

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REFERENCE Merkel, S. I., Voepel-Lewis, T., Shayevitz, J. R., & Malviya, S. (1997). The FLACC: A behavioral scale for scoring postoperative pain in young children. *Pediatric Nursing*, 23(3), 293–297.

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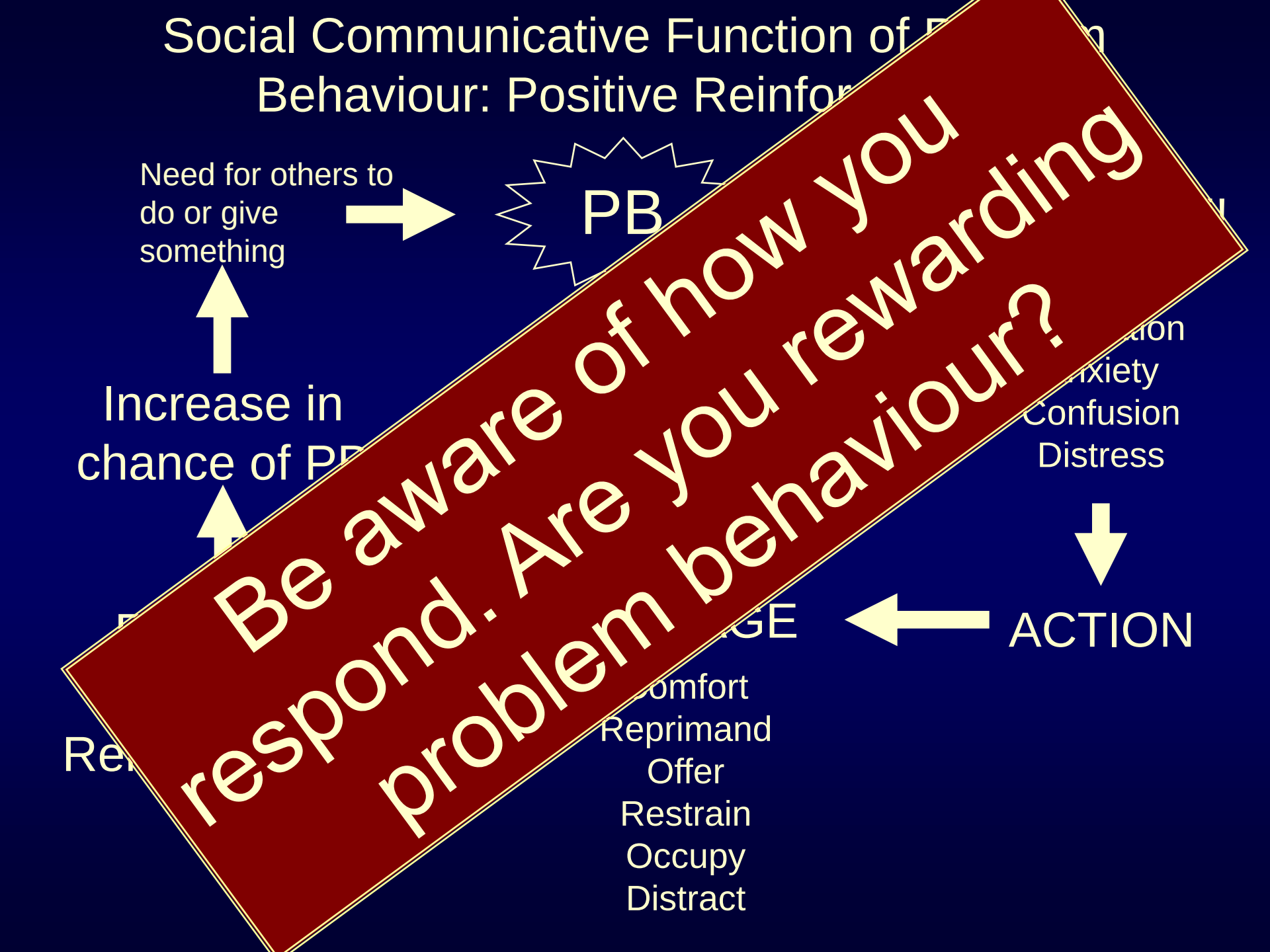
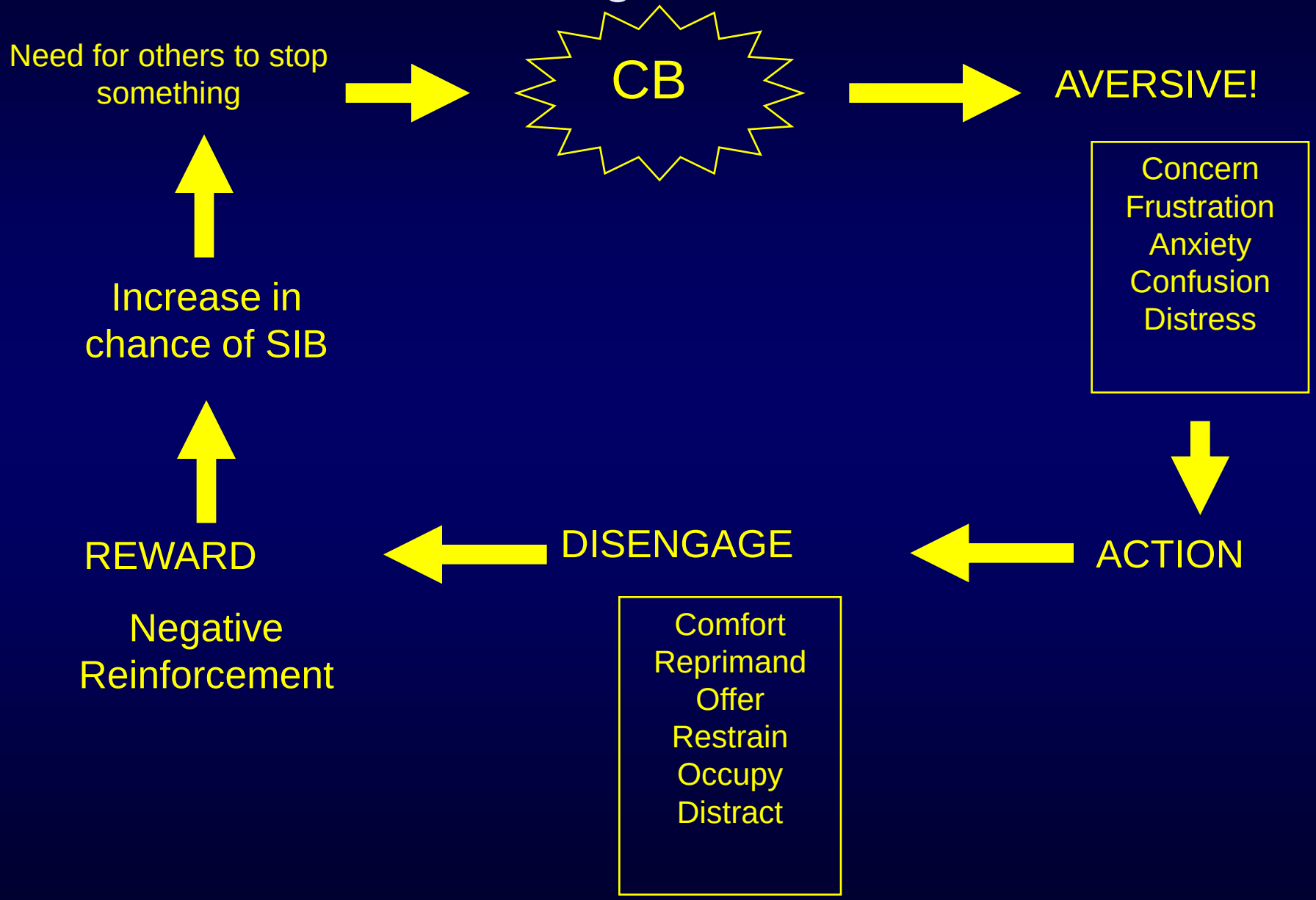
Development and Learning

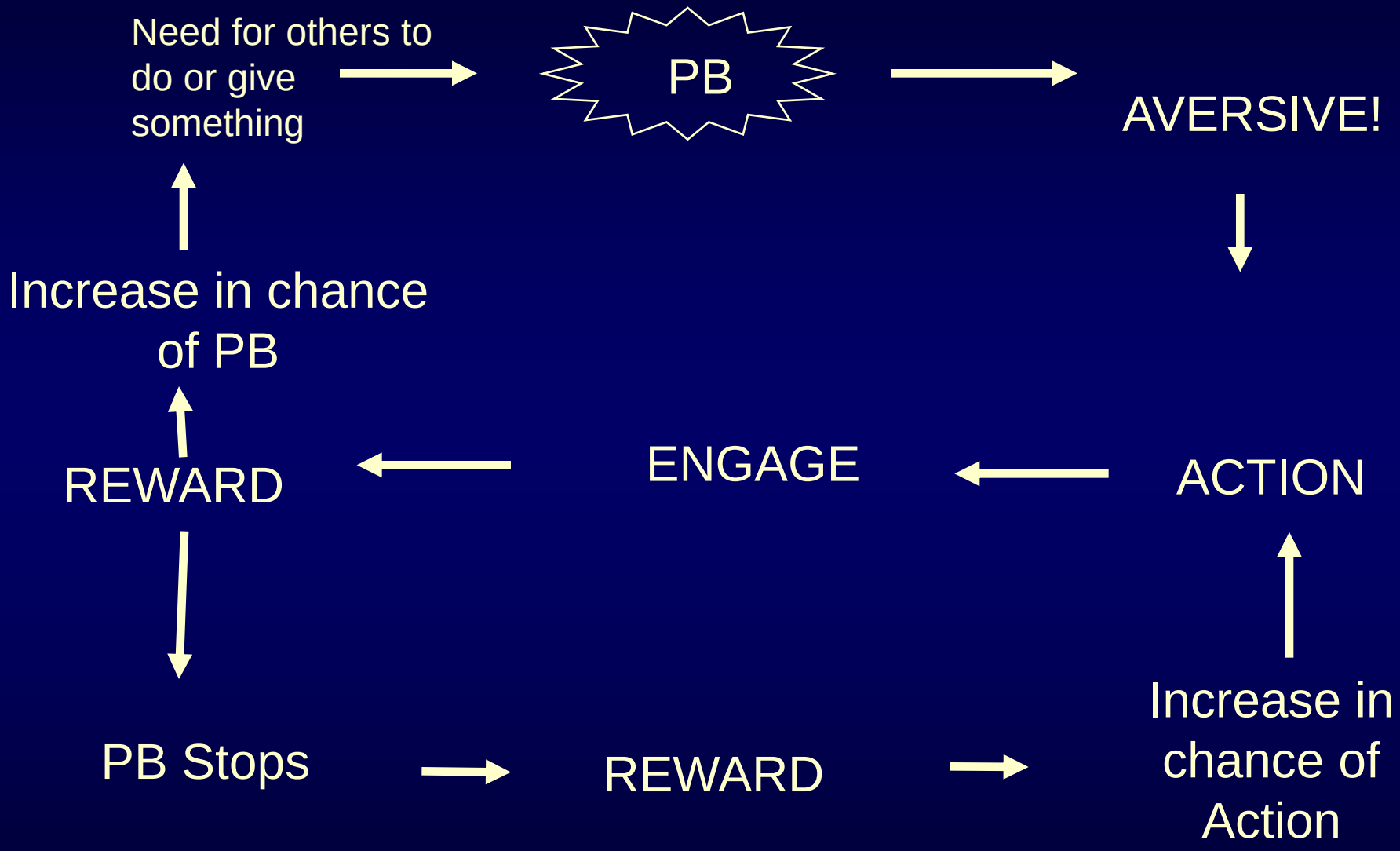
- Once a behaviour has occurred a number of times it can acquire a function
- The function can be to gain pleasant sensory stimulation (spinning) or to stop unpleasant sensory stimulation (scratch-itch)
- The function can become social or communicative. The behaviour can function to ‘say’:
  - “Come here and pay attention to me”
  - “Give me…..! I want…!”
  - “Stop! No! I don’t like that!”
- When behaviour has a social or communicative function it can get worse over time


Social Communicative Function of
Behaviour: Positive Reinfor
Need for others to do or give something
PB
Increase in chance of PB
ACTION
Confusion
Distress
Comfort
Reprimand
Offer
Restrain
Occupy
Distract
Be aware of how you respond. Are you rewarding problem behaviour?

Social Communicative Function of Challenging Behaviour: Negative Reinforcement



SIB: Mutual Reinforcement



An effective communication system is critical

Communicati of forms: speech, signs, objects, p to activate tapes. The most impor one can understand what is being 's

*E airment: get a communication system in plac en if language comes in later there is a window in whi ildren are ready to communicate but lack speech

Asking someone to do a task when they are tired, unhappy, unwell or in pain can make demand escape behaviours more likely to occur.

Self-injury and aggression summary

- We should be vigilant to the likelihood of self-injurious and aggressive behaviour
- Self-injury might be shown in response to pain – important to identify (e.g. FLACC) and treat pain which may not be expressed to due communication deficits
- Self-injurious and aggressive behaviour can gain a social/communicative function
- Responses to behaviour may be (inadvertently rewarding) – stop and think before you respond
- An effective communication system is essential

Studying sleep in children with Smith-Magenis syndrome

Jayne Trickett, Dr Caroline Richards, Mary Heald, Hayley Denyer & Prof Chris Oliver

Cerebra Centre for Neurodevelopmental Disorders

School of Psychology, University of Birmingham

Sleep quality in children with SMS

- Research studies: 100% of children had a sleep disturbance
- Night time wakings
- Early morning wakings (Greenberg, Lewis & Potocki, 1996; Smith, Dykens & Greenberg, 1998)
- Overall reduced sleep time relative to children of the same age (Gropman et al., 2006)
- Sleep debt- increased daytime sleepiness, daytime naps (Potocki, 2000; De Leersynder et al., 2006)

Actiwatch Study: Directly assessing children and parent's sleep quality- novel methodology

- School aged children with Smith-Magenis syndrome, Angelman syndrome and ASD and their caregivers & a typically developing contrast group

Child sleep characteristics

Actigraphy

Video recording of child sleep

Sleep diary on mobile app

Daytime behaviour

Behaviour ratings on mobile phone app- teachers & parents

Parental impact and wellbeing

Parents wearing Actiwatches

Sleep diary on mobile phone app

Saliva samples to assess parental stress

- The study is now very nearly complete with results being presented at a range of academic and family support conferences

Impulsivity and temper outbursts

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Impulsivity

Caregiver report

Primary problem behaviour associated with SMS: 10 most problematic behaviours included being **impulsive** & **demands must be met immediately**
(Clarke and Boer, 1998)

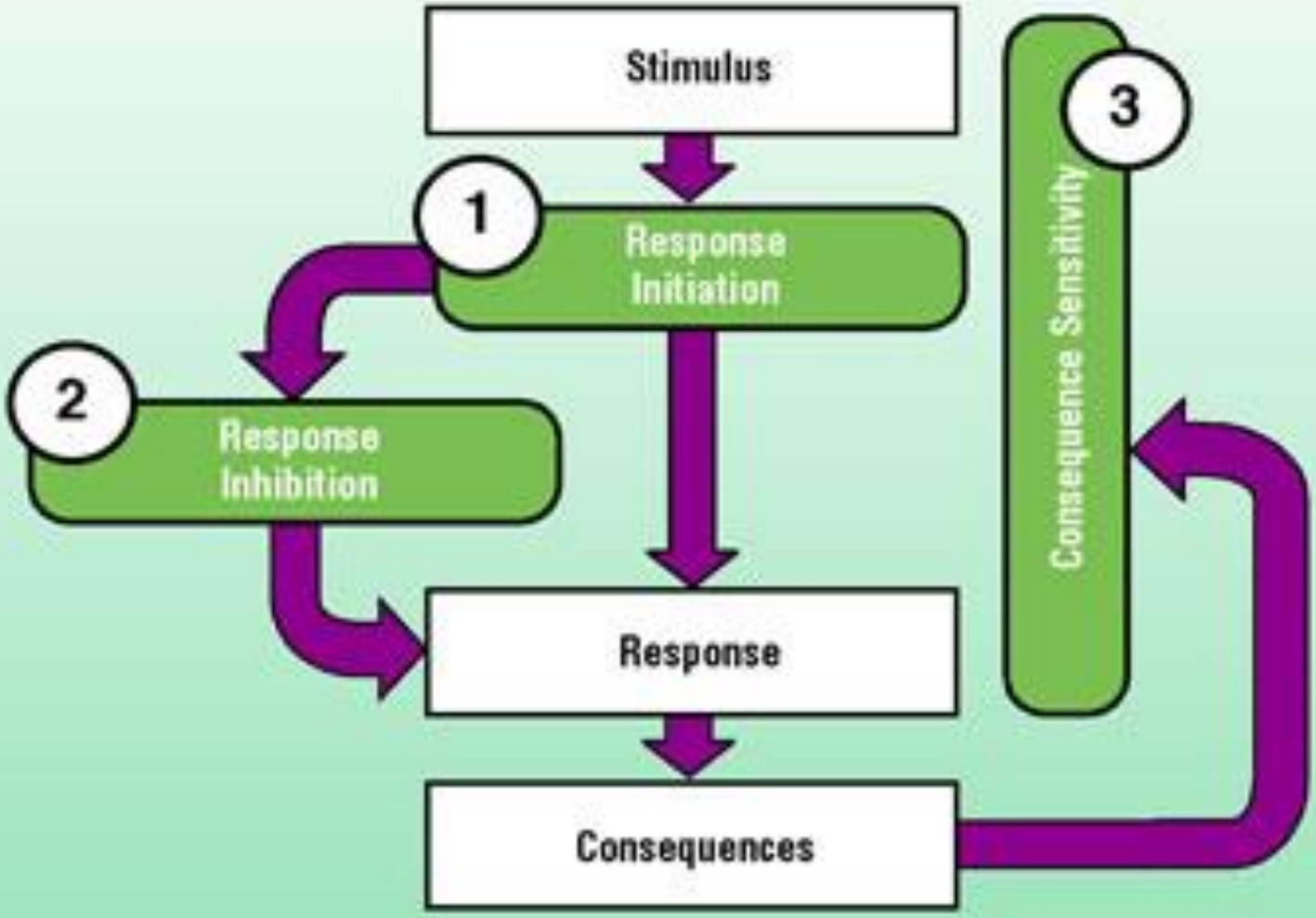
Present in over **80%** of individuals with SMS
(Dykens et al 1997 and 1998)

Uniquely high scores …significantly elevated in both children and adults with SMS (**40%** and **58.3%** respectively) compared to 7 other genetic syndromes
(Oliver et al., 2011)

* Aggressive behaviour severity strongly associated with impulsivity (Sloneem et al., 2011).

Figure 3

Diagram of How the Three Components of Impulsivity Interact to Affect Behavior



Source: Dougherty DM et al. (2005)

TANTRUM VS SMS MELTDOWN

Tantrum

- "Want"/ directed
- Goal/Control driven
- Audience to perform
- Checks engagement
- Protective mechanisms
- Resolves if goal is accomplished

SMS Meltdown

- Over-stressed/Overwhelmed
- Reactive mechanism
- Continues without attention
- Safety maybe compromised
- Fatigue
- Not goal dependent
- May require assistance to gain control

Impulsivity and temper outbursts summary

- Impulsive behaviour is very common
- Being very impulsive might be a risk marker for showing more severe aggression
- Different factors may affect impulsivity in SMS (ability to stop a response once it has started, how the person responds to rewards)
- ‘Meltdowns’ in SMS might differ from temper outbursts in other children, less goal directed
- Can not versus will not control behaviour?

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The autism question

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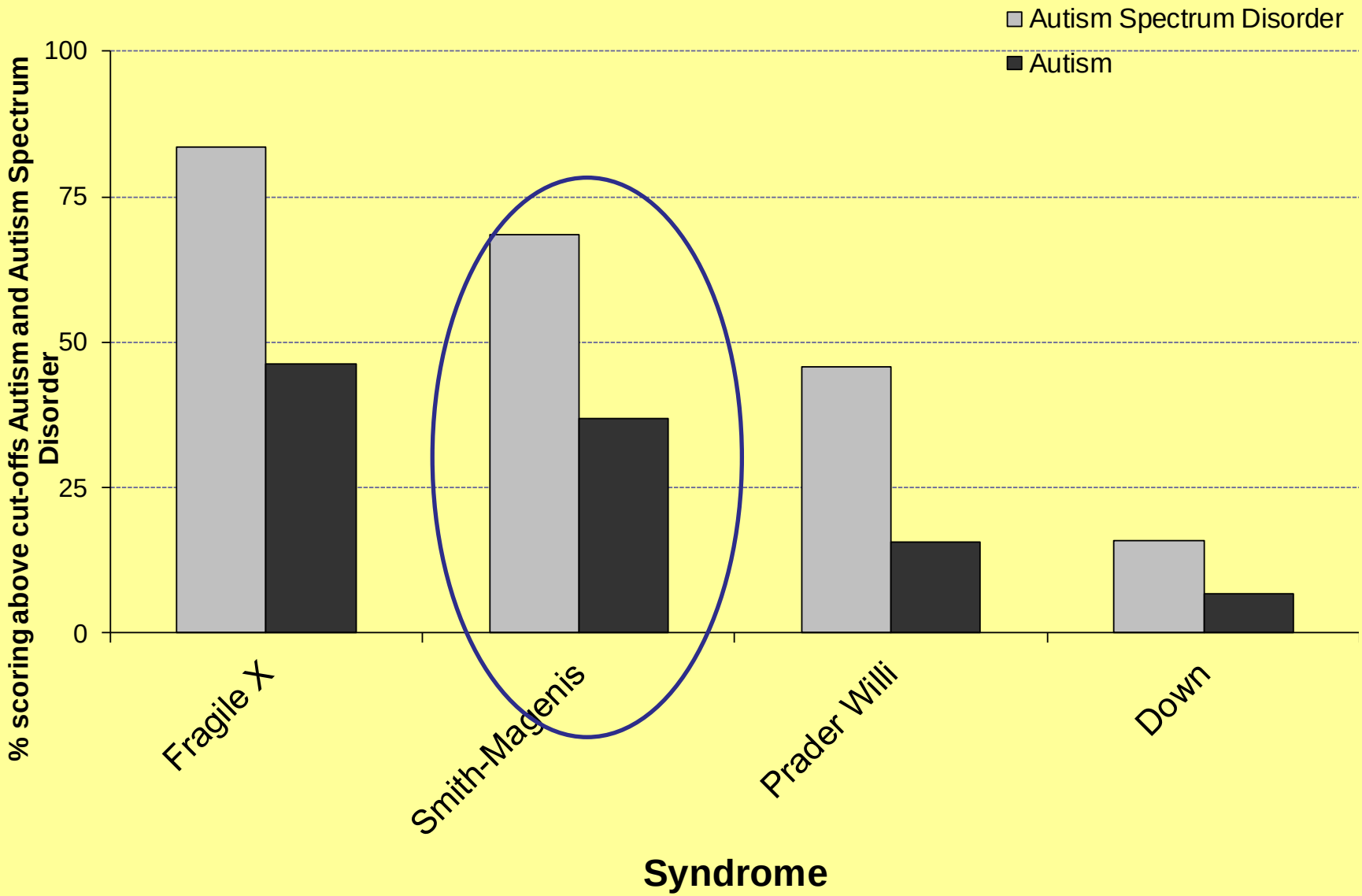
Autism spectrum disorder (ASD) (DSM 5)

Social communication & Social interaction.

+

Restricted, repetitive patterns of behaviour, interests or activities including sensory difficulties

Percentage of group scoring above cut-offs for Autism and Autism Spectrum Disorder on ASQ/SCQ



Oliver et al. 2011; Oliver et al., in preparation

Autism Screening Questionnaire

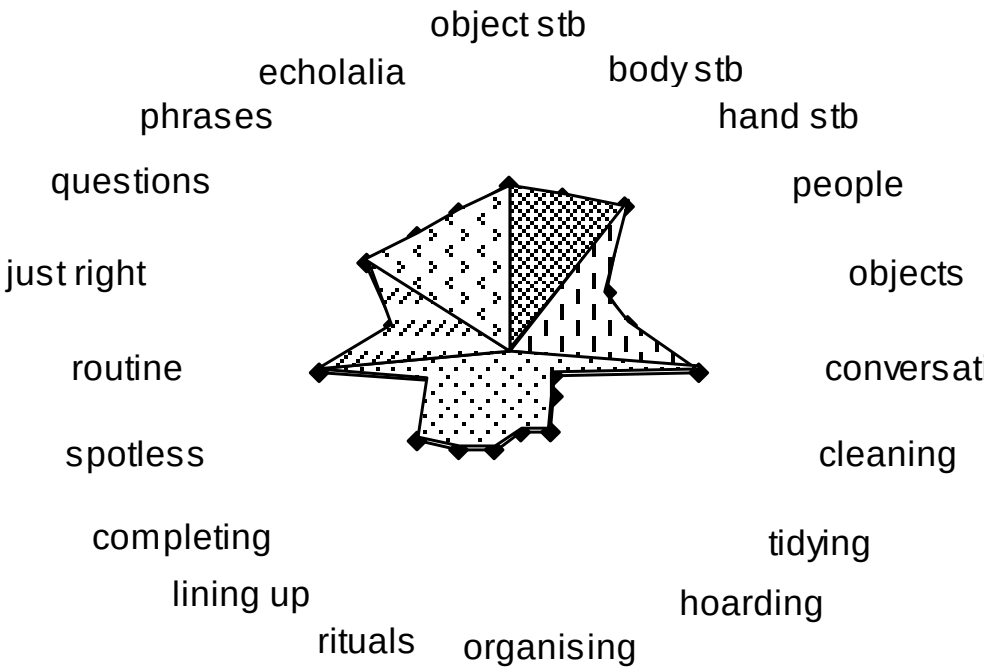
	% ASD	% Autism	Social	Comm.	Rep. Beh.
Angelman *(15q11-q13)*	66.3	17.8	++	+	-
Cri du Chat *(5p 15.2-15.3)*	40.0	8.0	----	-----	-
Cornelia de Lange *(5p 13.1)*	78.8	45.9	++	+	O
Fragile X *(Xq27.3)*	83.6	46.3	++	++	++++
Prader-Willi *(15q11-q13)*	45.8	15.5	----	-	--
Smith Magenis *(17p 11.2)*	68.4	36.8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

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

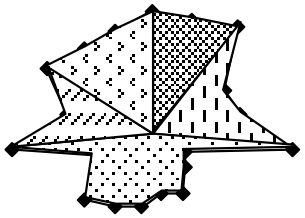
Autism



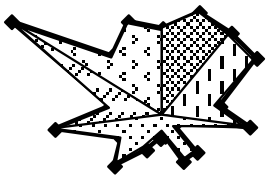
Profiles of repetitive behaviour across syndromes

Moss, J., Oliver, C., Arron, K., Burbidge, C. and Berg, K. (2009). The prevalence and phenomenology of repetitive behavior in genetic syndromes. *Journal of Autism and Developmental Disorders.* **39,** 572-588.

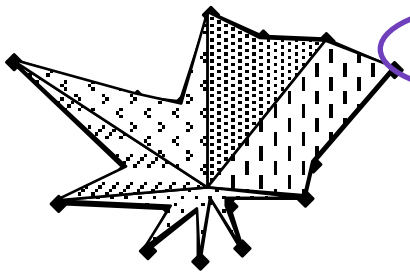
Autism

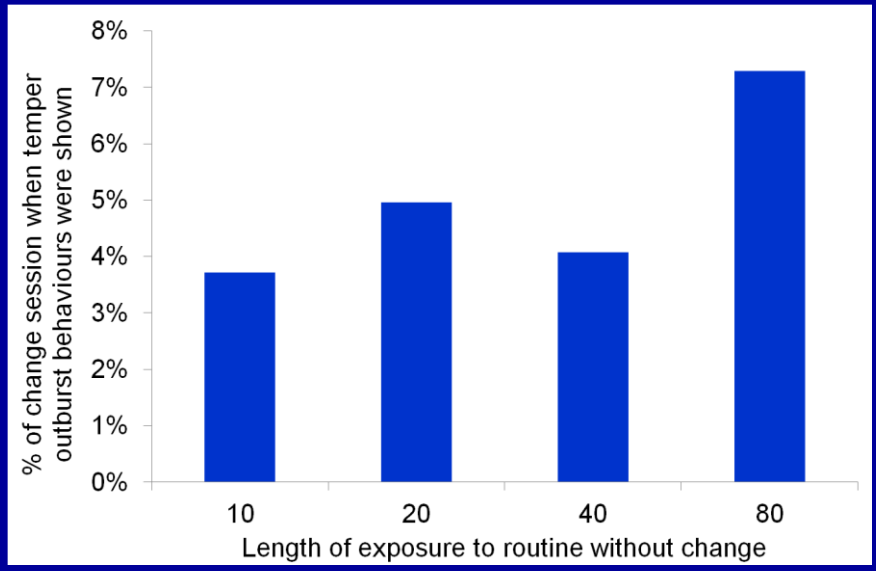


HID



SMS




8%
7%
6%
5%
4%
3%
2%
1%
0%
% of change session when temper outburst behaviours were shown
10
20
40
80
Length of exposure to routine without change

Autism Spectrum Disorder or not?

- Behaviourally defined, ‘shopping list’ of criteria
  - Attaining cut-off scores but with different profiles
  - Scoring on an item for different reasons
- Unusual features
  - Profile of repetitive behaviours
- Is the diagnosis helpful?
  - Services
  - Good advice from Autism materials

Autism

- A high percentage of people with SMS reach cut offs for autism/ASD
- This might be more to do with repetitive behaviour than difficulties with social interaction or communication
- Repetitive behaviours in SMS (routines, people, questions) might differ from those in ASD
- Introducing ‘fuzzy’ routines might help reduce difficult behaviours when routines are changed
- Pursue a diagnosis of autism/ASD if this would be useful to access services/support
- Autism/ASD resources may be useful (repetitive behaviour/sensory issues)

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Attention seeking

Anecdotal accounts

“Students with SMS tend to be **very adult-oriented**, demanding an **inordinate amount of individualized attention from adults**; when this is denied, aggressive and self-injurious outbursts are frequently the result”

Haas-Givler, 1994

"If I could provide **constant, unwavering, one-on-one attention** to this child throughout the day, she might never have another tantrum."

Teacher

Caregiver report

Attention seeking problematic for 80% of individuals with SMS
(Dykens et al., 1997)

94-100% of those with SMS **“demand a lot of attention”**
(Dykens & Smith 1998)

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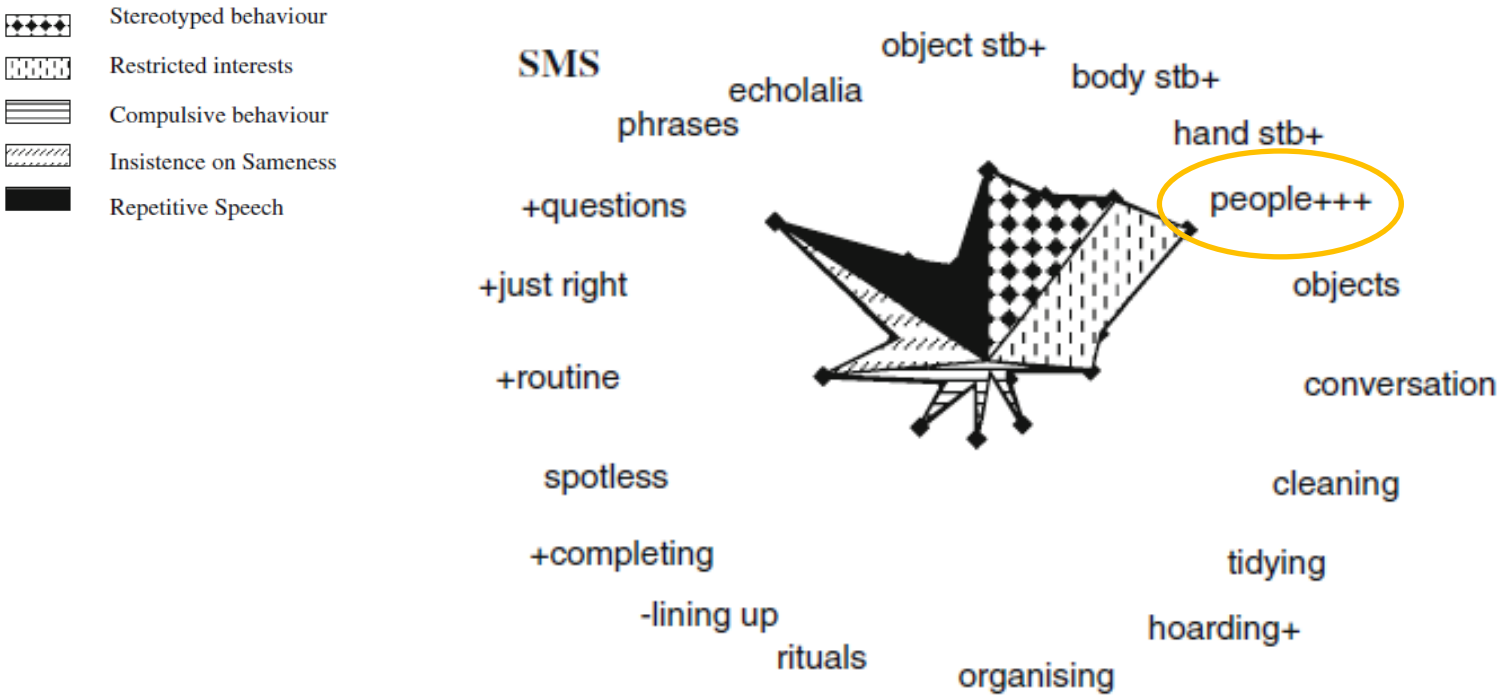
Repetitive behaviour in SMS



Suggests uniquely strong drive to see, contact or speak to particular favourite people in SMS

Direct observations

- Reduced adult attention/adult demand *precedes* self-injurious and aggressive/disruptive outbursts, a*fter* the outburst the probability of the child receiving attention from an adult increases (Taylor & Oliver, 2011)
- Greater preference for directing attention and looking towards adults (over peers) across different conditions in SMS than DS (Wilde, Silva & Oliver, 2013).
- Compared to DS, comparatively high social initiations when attention is low compared to high attention, increased preference for mothers than strangers (Wilde, Mitchel & Oliver, 2016)

Sensitivity to reduced attention; some difficult behaviours may 'function' to access adult attention.

Uniquely strong preference for interacting with adults

Unusually strong social motivation, preference for familiar adults

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What does this ‘look’ like?

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Impact on family

Parents of children with disabilities experience higher levels of stress (Hodapp, Fidler & Smith, 1998; Olsson & Hwang, 2001)

Caregivers of people with SMS report higher stress levels than those of children with ID of heterogeneous origin (Hodapp *et al.* 1998)

Stress likely to be exacerbated by attention-seeking behaviour towards caregivers (Isles, 2011),

particularly given association between reduced attention and challenging behaviour

‘Attention seeking’ summary

- ‘Attention seeking’ is very commonly described
- Desire for high levels of attention (one to one) with difficult behaviours when attention is unavailable
- Strong preference – for favourite people, adults, familiar adults/caregivers
- When this behaviour is focussed on caregivers this might be hard to manage and increase stress

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FIND

Further Inform Neurogenetic Disorders

A sustainable web based resource for dissemination of research findings: Jane Waite

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E·S·R·C ECONOMIC & SOCIAL RESEARCH COUNCIL

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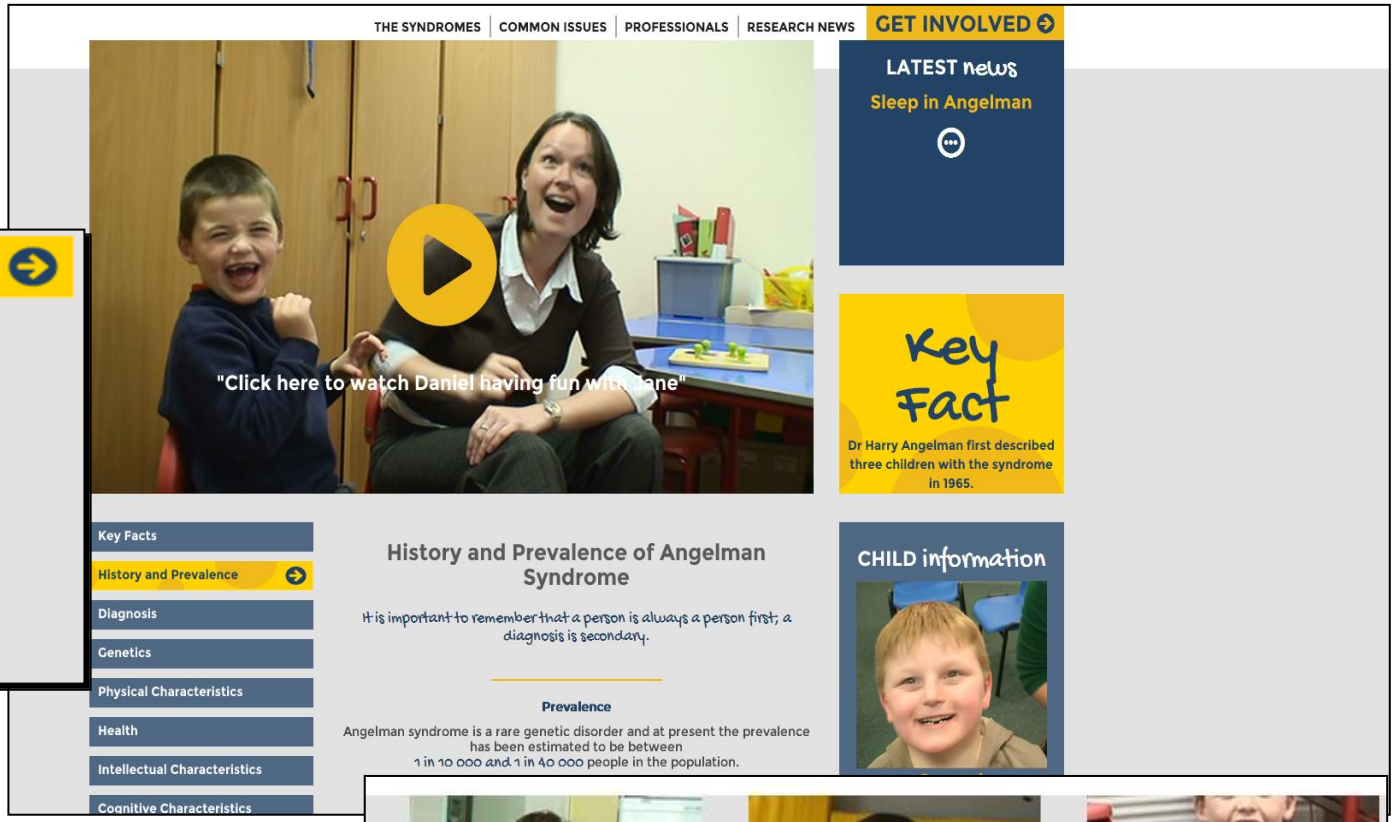
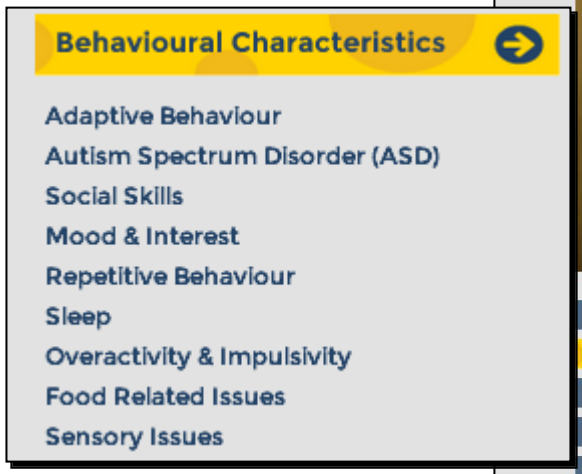
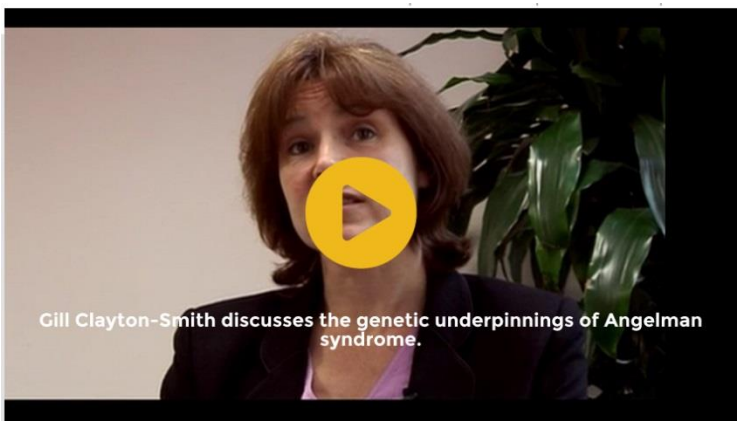
FIND

Further Inform Neurogenetic Disorders







www.findresources.co.uk

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Self-injurious behaviour in children with an intellectual disability

Prof. Chris Oliver
Dr Louise Davies
Dr Caroline Richards

This briefing has been written to help parents and carers of children with an intellectual disability to understand what self-injury is, what the causes are and which interventions are effective. The briefing focuses on children

Some thoughts

- These things are not inevitable
- Start now
- Key points
  - Communication
  - Health
  - *Sleep routines and management
  - Response to behaviours
  - Managing ‘temper outbursts’/*impulsive behaviour

Core Funding

Cerebra

Grant Support

Medical Research Council
The Big Lottery
Baily Thomas Foundation
Cornelia de Lange Syndrome Foundation
Research Autism
NE Essex PCT
Birmingham Children's Hospital
Angelman Syndrome Foundation (USA)
Newlife
National Autistic Society
Smith-Magenis Syndrome Foundation
ESRC

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