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Italy, 2012

## Self-injury, autism and changes with age in Cornelia de Lange syndrome

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### Three main areas

- Self-injury
- Autism spectrum disorder
- Changes with age

Self-injurious behaviour in  
Cornelia de Lange Syndrome

A Guide for Parents and Carers

Chris Oliver  
Jo Moss  
Jane Petty  
Kate Arron  
Jenny Sioneem  
Scott Hall

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COMMUNITY FUND

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Understanding and Changing  
Challenging Behaviour in  
Cornelia de Lange Syndrome

Chris Oliver, Jo Moss, Jane Petty, Penny Tunnicliffe,  
Richard Hastings, Pat Howlin, Gemma Griffith,  
Leah Bull, Dorelle Villa and Michael Yip

Click on and watch...

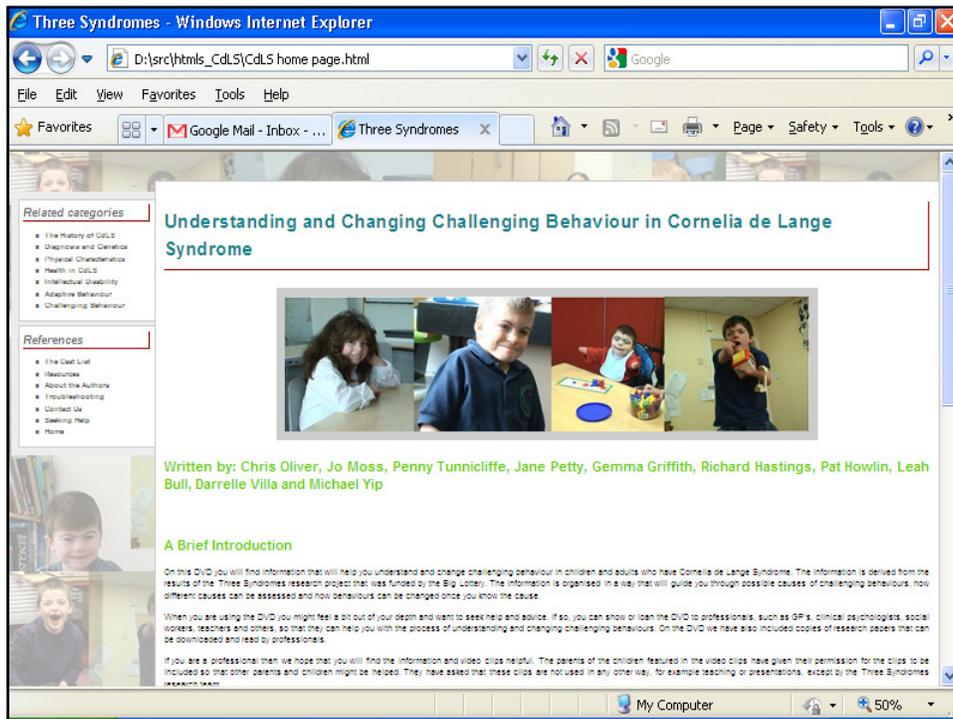
Understanding and Changing  
Challenging Behaviour in  
Cornelia de Lange Syndrome  
Cornelia de Lange Syndrome  
Awareness Foundation  
UK and Ireland and Chris Oliver,  
School of Psychiatry,  
University of Birmingham.

LOTTERY FUNDED

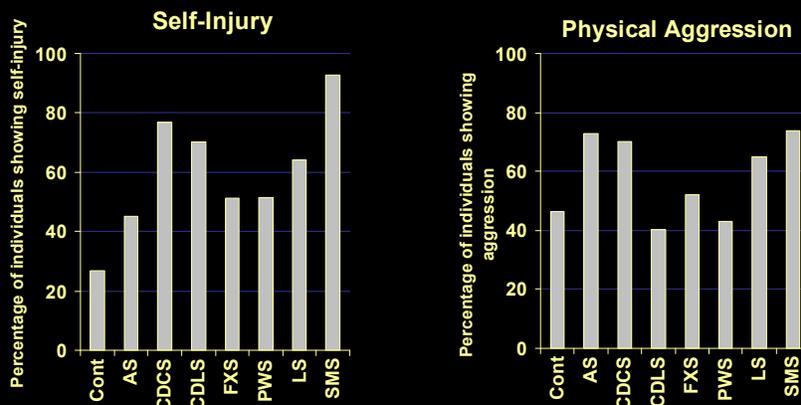
KING'S COLLEGE LONDON

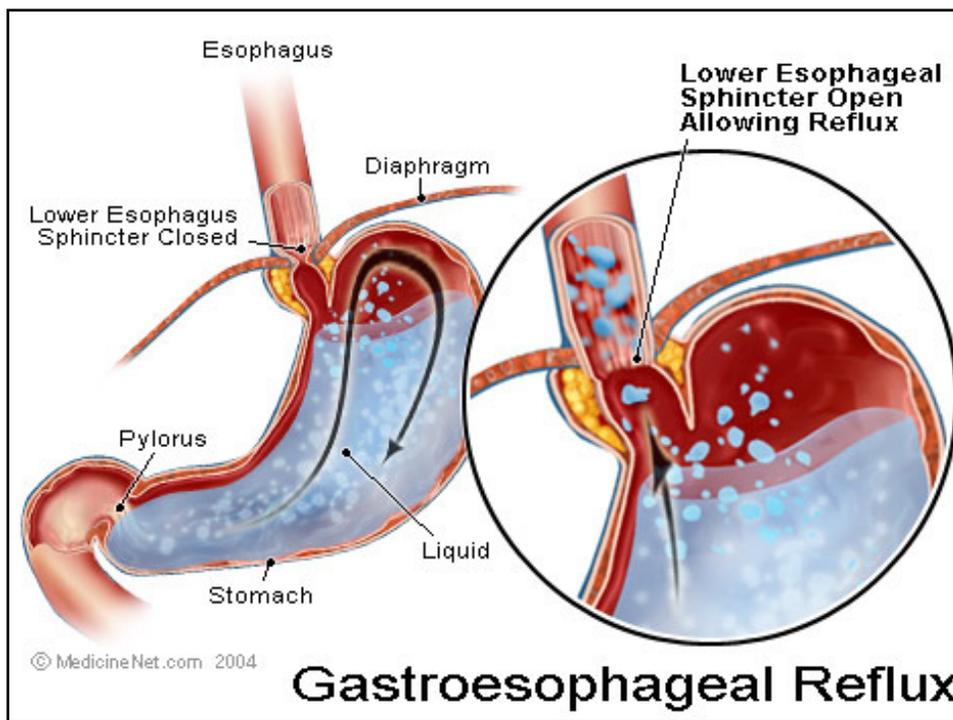
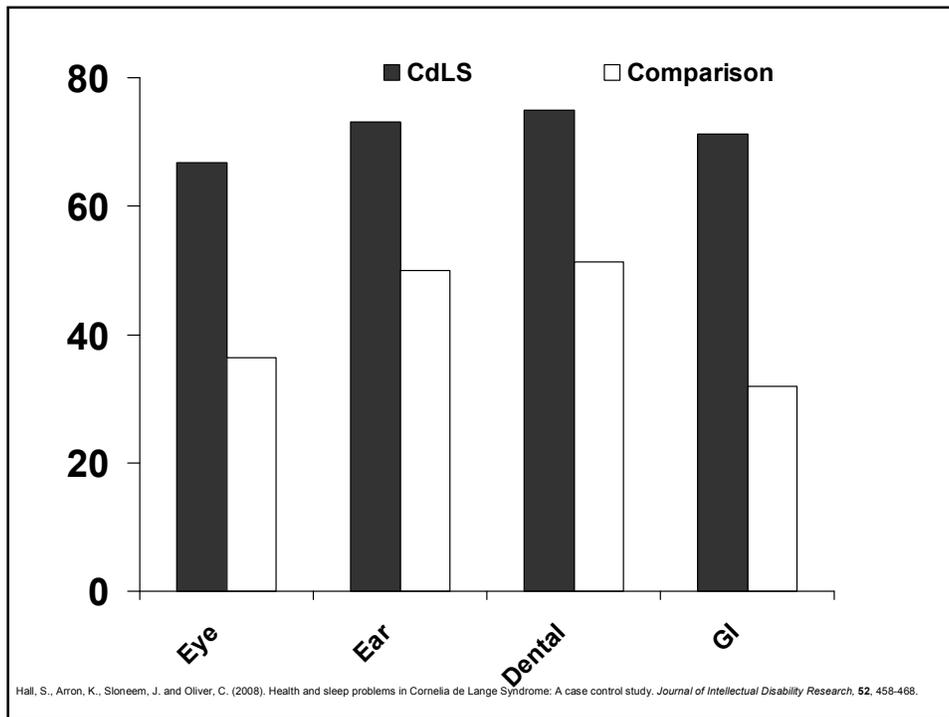
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## Prevalence of Self-Injury and Physical Aggression in Syndromes





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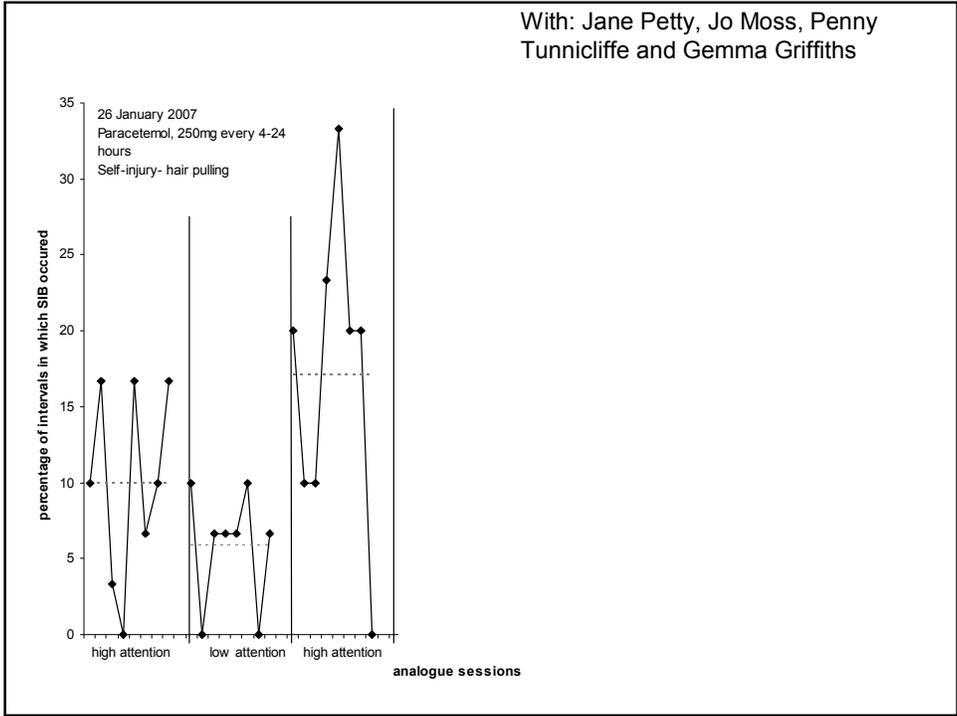
15% of all children bang their head between the ages of 1 and 4.  
50% of these children have a middle ear infection.

Arch his/her back	Cough/gag/regurgitate
Lie over object on stomach	Discomfort
Salivate excessively	Refuse food
Fidget/wriggle	Indecisive about food
Fingers in mouth	Wake during the night
Chew clothes	Sleep sitting or propped up
Grind teeth	Bad Breath
Scratch/rub chest/throat	Respiratory tract infections
Drink excessively	

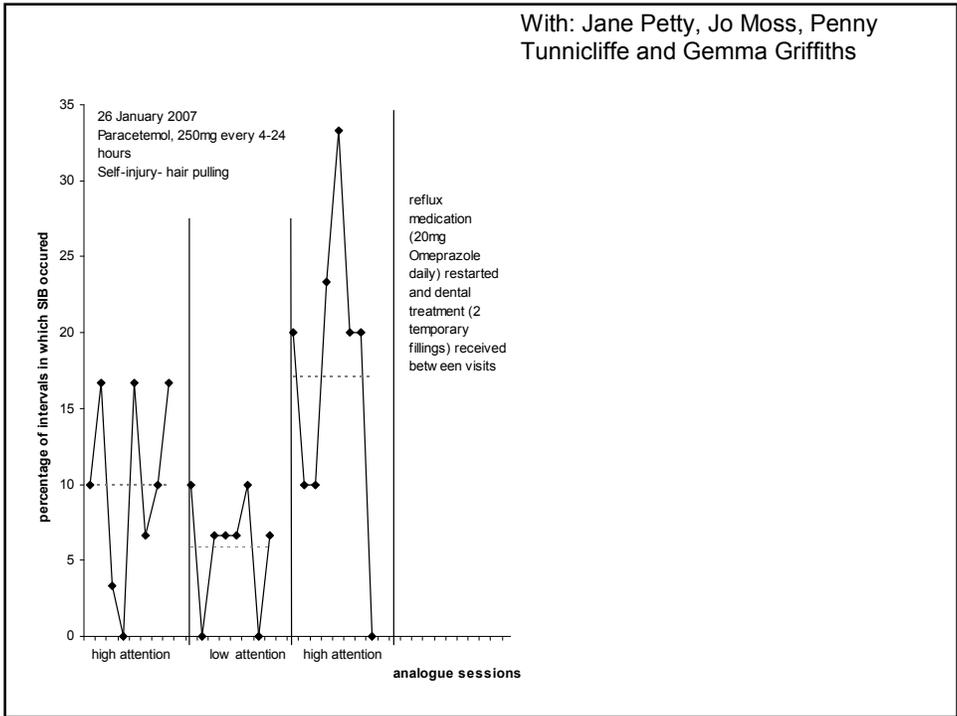
  

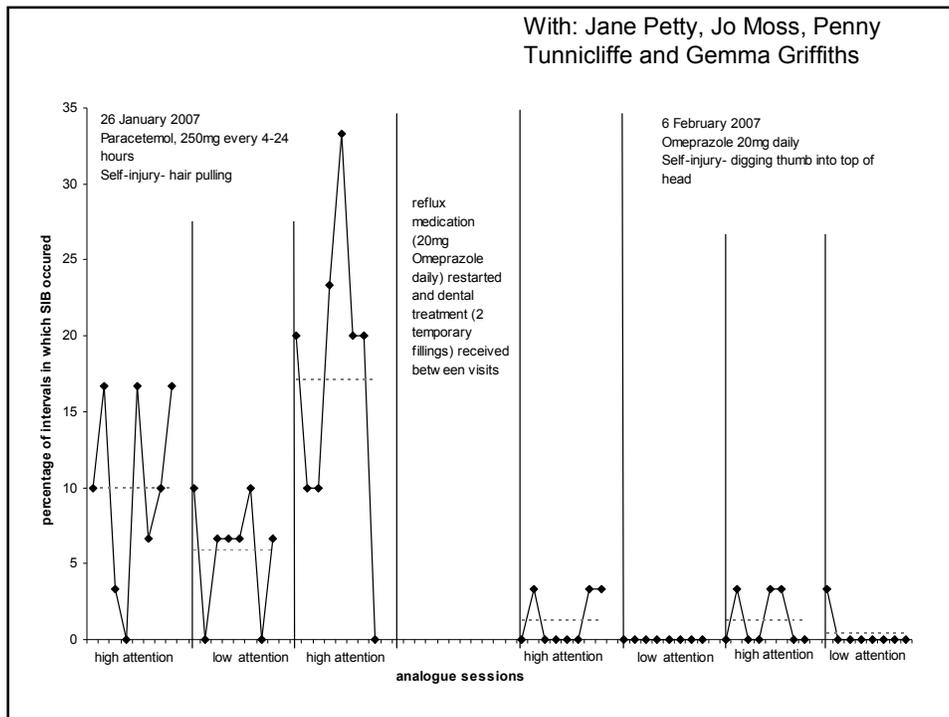
Behavior	No	Yes	p-value
Self-injury	~12	~18	0.004
Aggression	~13	~20	0.001
Prop. Dest.	~13	~19	0.001

With: Jane Petty, Jo Moss, Penny Tunncliffe and Gemma Griffiths



With: Jane Petty, Jo Moss, Penny Tunncliffe and Gemma Griffiths





## Pain signatures

- Face
- Legs
- Activity
- Crying
- Consolability

Kate Eden (PhD student, Cerebra Centre funded)

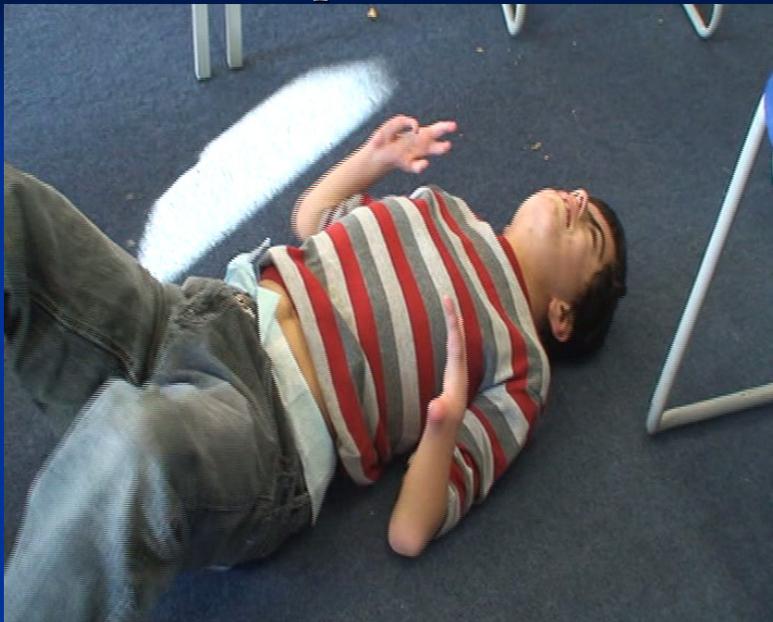
## Indicators of pain and discomfort



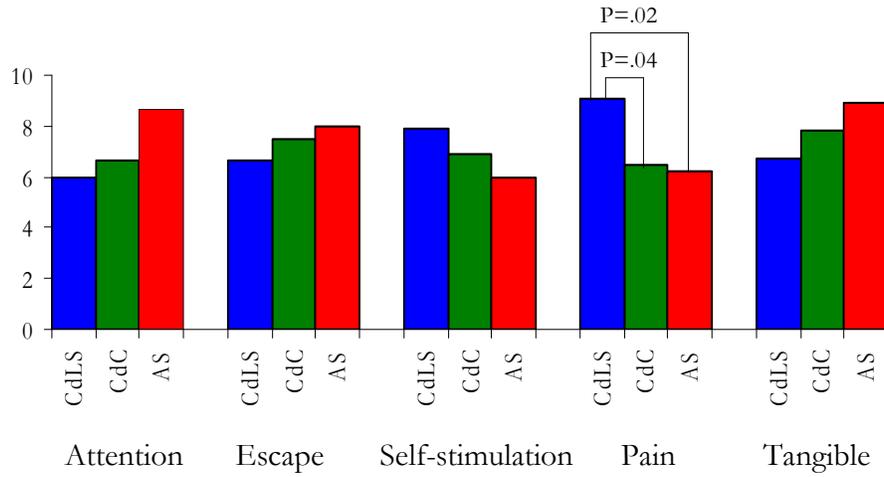
## Indicators of pain and discomfort



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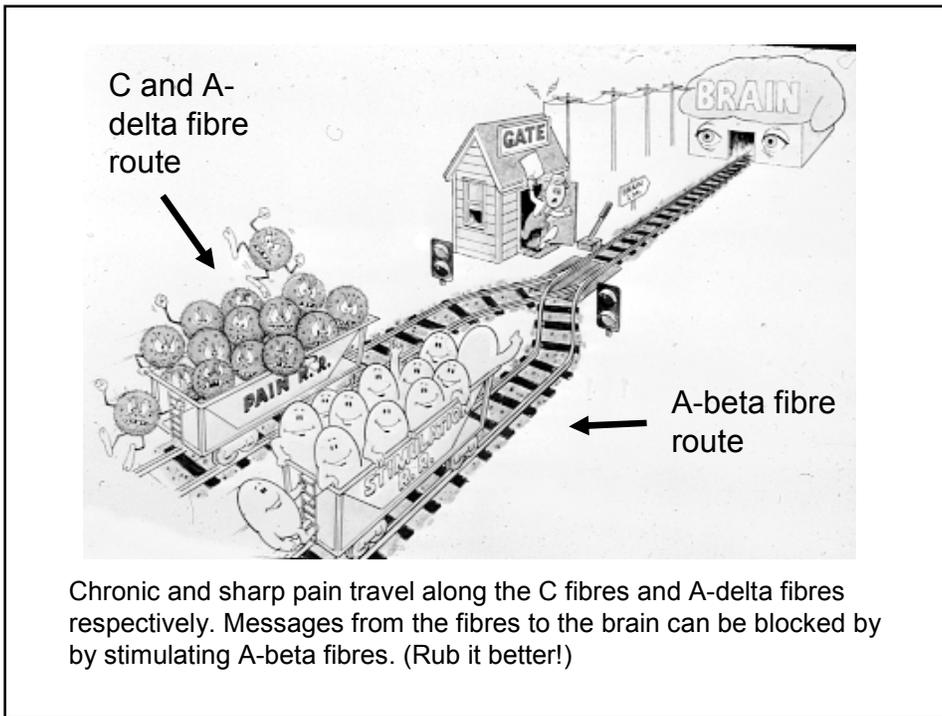
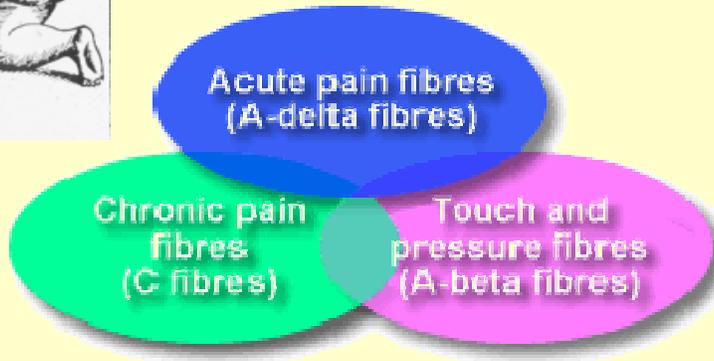
Comparison of causes of challenging behaviour in Cornelia de Lange, Angelman and Cri du Chat syndromes



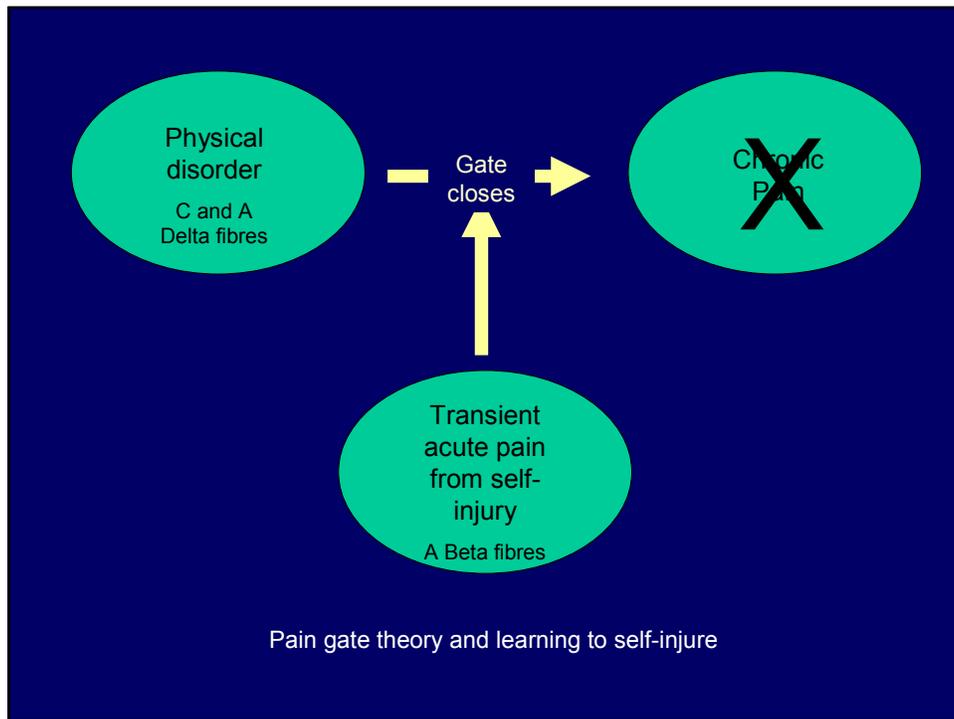
Pain gate theory



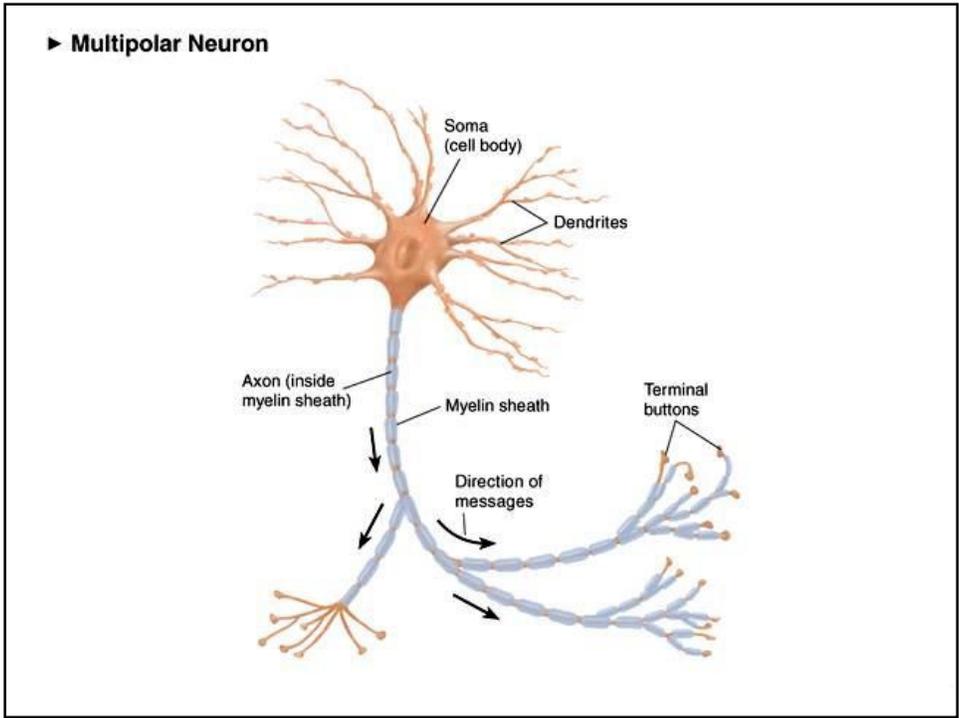
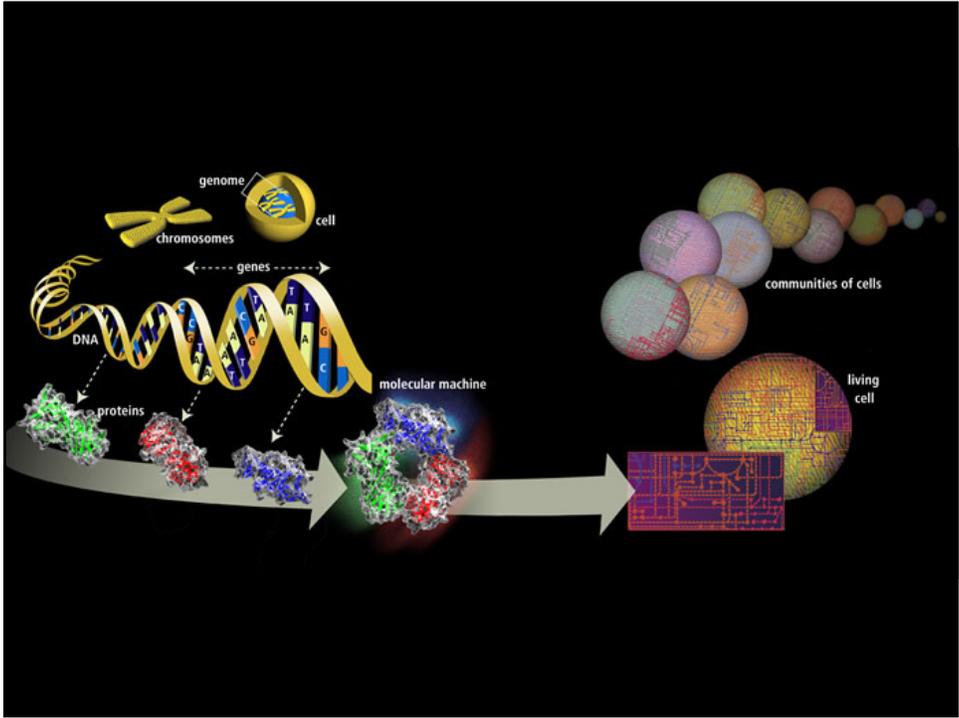
The experience of pain is influenced by many factors and there is no simple pathway from sensation to experience.  
Three types of nerve fibres are important

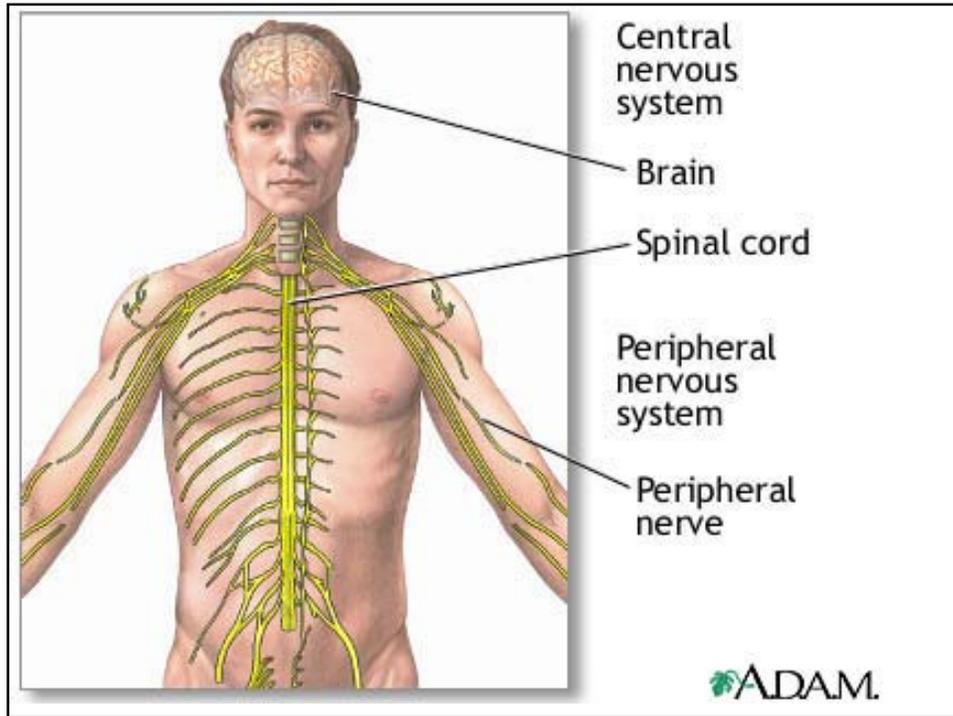


Chronic and sharp pain travel along the C fibres and A-delta fibres respectively. Messages from the fibres to the brain can be blocked by stimulating A-beta fibres. (Rub it better!)

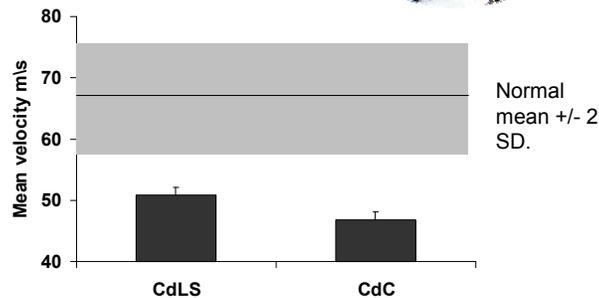


Disordered pain perception

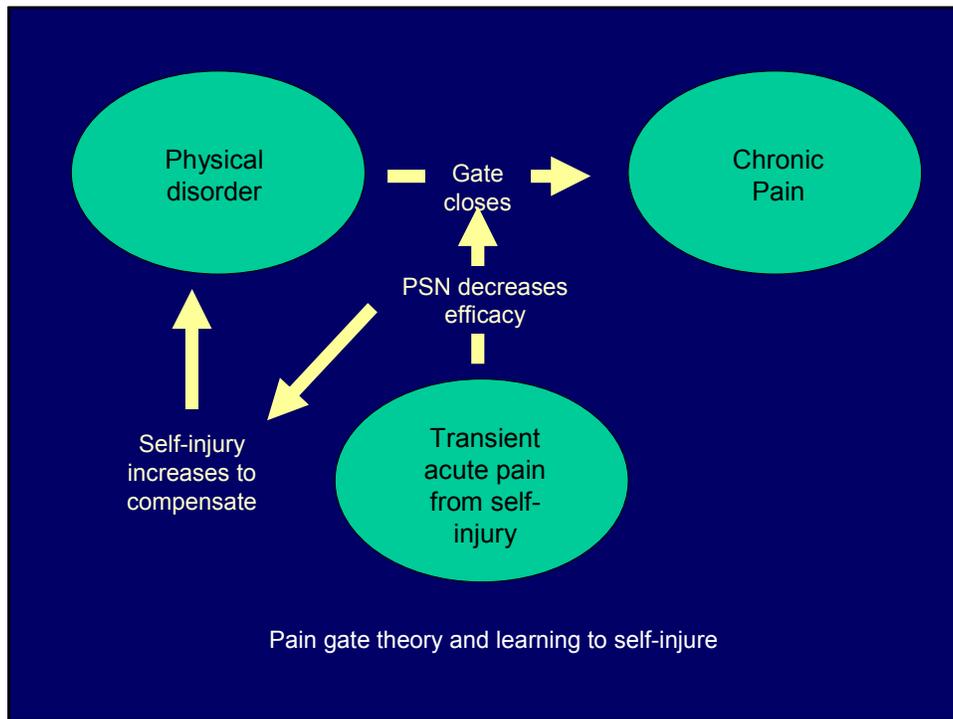




- Electrode stimulates the nerve with mild electrical impulse. Stimulus duration 0.1ms, frequency around 1Hz.
- Latency is time from the stimulus to the first positive peak of sensory nerve action potential (SNAP).
- Median (arm), medial plantar and sural (leg) nerves.
- Stable velocity at about 5 years old; median nerve mean = 67.5 m/s (SD 4.4)



Wilkie, L., Oliver, C., Seri, S., Freiss, S. & Moss, J. (2005). The Role of Nerve Function in Cornelia de Lange and Cri du Chat Syndrome *Proceedings of the Greenwood Genetic Center*, 24, 182.



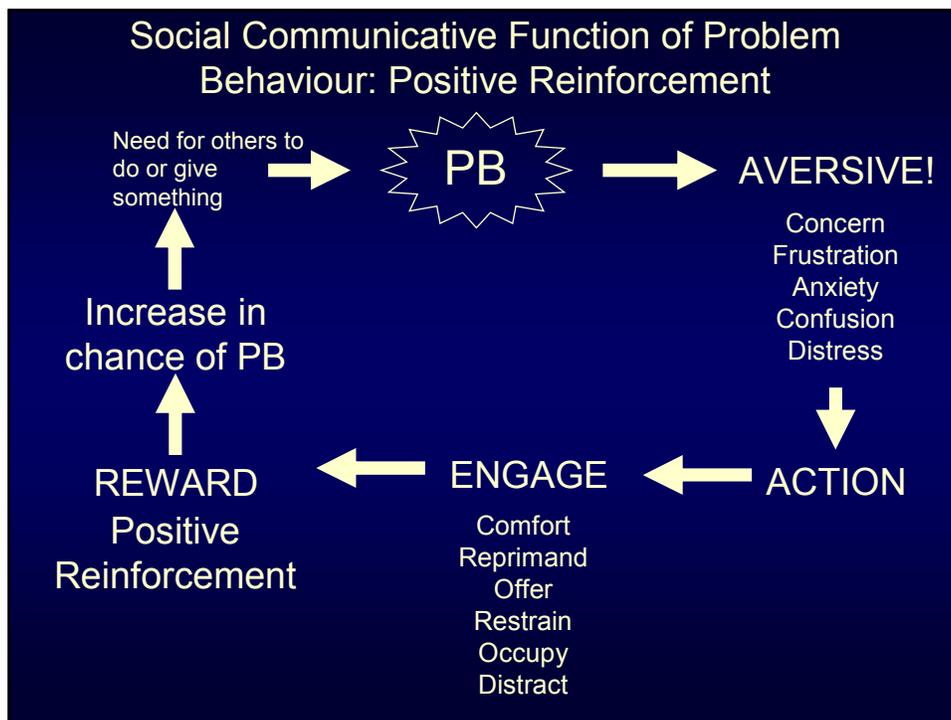
## The pain cycle and self-harm

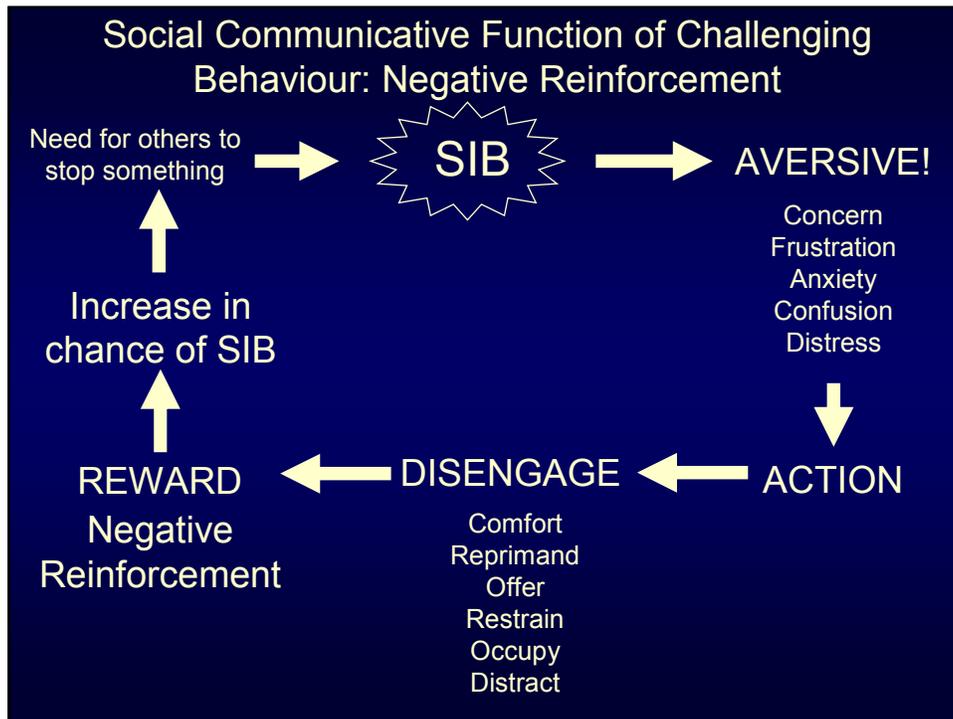
1. Chronic or sharp pain is caused by physical injury or trauma
2. The child blocks the pain by self-harm such as cutting or hitting.
3. The fibres carrying pain signals work very slowly so the child self-harms more and harder.
4. The more the child self-harms and hitting leads to tissue damage and sharp pain.
- 5.

**Early intervention and aggressive treatment**

## 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





Communication has a number of forms: speech, signs, or microswitches to activate tape recorders. The important thing is that everyone can understand what is being 'said'.

**An effective communication system is critical**

## Self-injury

- Reflux is not the only source of pain
- Know you child's pain signature
  - Face
  - Legs
  - Activity
  - Consolability
  - Crying
- Bursts of behaviour and then none
- 'Out of the blue'; unrelated to the environment
- BUT.....
  - The behaviour may become functional and so.....
  - Behaviour management strategies are helpful
- Skin picking on fingers, arms and legs may be anxiety related

## Three main areas

- Self-injury
- Autism spectrum disorder
- Changes with age

## What is Autism Spectrum Disorder?

- Impairments in Social Interaction
  - Difficulty making friends
  - Find social interaction unpleasant/uncomfortable
- Impairments in Communication Skills
  - Delayed/ no development of verbal and non-verbal communication.
  - Initiating and maintaining conversation
- Repetitive behaviours
  - Ritualistic behaviour
  - Compulsive behaviour
  - Restricted interests: people, topics, places

## Autism Spectrum Disorder or not?

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

## These are difficult because...

	<b>Associated problems</b>
<i>Rituals and stereotypies</i>	Limit acquisition of other skills. Interfere with other activities. May involve others. Can be highly disruptive. Dominate daily routines.
<i>Disruption of routines</i>	Distress, aggression, self injury; Lack of co-operation.
<i>Dislike of change</i>	Rigid and inflexible behaviour. Anxiety if the need for sameness cannot be met.

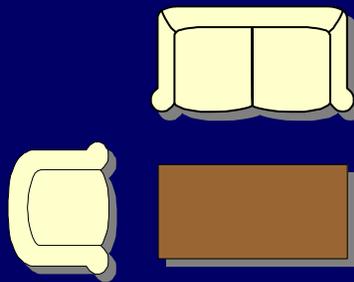
## Practical Implications: What can we do?

- Resources and approaches from the ASD literature may be helpful.
- Environment (e.g. classroom, college, residential placement) and level of social demand may need to be adjusted.
  - Environment with low social demands and high predictability.
  - Preparation and visual/picture schedule
  - Slow introduction to novel social situations.
  - Coping strategies e.g. Mobile phone/flash cards
- Important that the professionals working with your children are aware of these needs and sensitive to them.
- Specific interventions for repetitive behaviour

## Resistance to Change: Small changes: changing/moving furniture

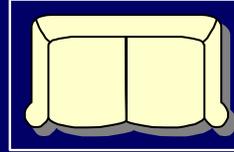
### Case study: Emily

Emily could not tolerate the smallest change in her physical environment. Everything had to be 'just so' and for years the furniture had been arranged in exactly the same way.

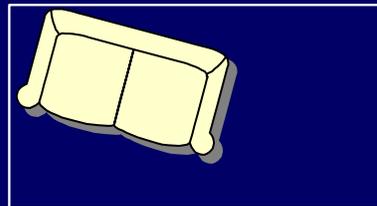
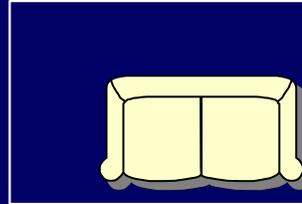


## Successful solution:

**Step one:** They used tape to mark an outline round the sofa and explained to Emily that the sofa had to be within these limits.



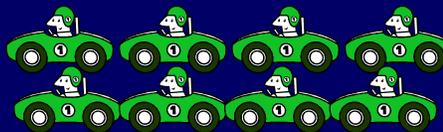
**Step two:** At a small distance at a time, her parents moved the tape, enlarging the area in which the sofa was located. Each time they repositioned the sofa within these limits.



Example from Whitaker (2005). Challenging behaviour and autism

## Lining up objects

- Step one: gradual reduction of number of cars from 50 to 20



- Step two: Further reduction in length of lines to 4 cars only



- Step three: Pairs of cars only allowed in house, thought these scattered in various rooms



Howlin (2005) Children with Autism and Asperger Syndrome

## Repetitive Behaviour in CdLS

- Tidying up and lining up or other compulsive like behaviours.
- Strategies that might help can be found in the ASD literature.
- Key points to remember:
  - Work to *modify* the behaviour not eliminate it.
  - Make *gradual* rather sudden changes in routine and the environment.
  - Take small steps.
  - Can take a long time before you see results.
- Deal with one behaviour at a time.
- Keep an eye out for any ‘new’ behaviours.
- Young children: prevention is better than intervention.

## Three main areas

- Self-injury
- Autism spectrum disorder
- Changes with age

## Planning ahead for person environment fit

“Hi Chris

I have had quite a lengthy phone call from .... whose son .... is having quite severe behavioural problems. .... (He) will be 18 in a couple of weeks and the problems are getting worse. It is a nightmare to get him out of the house ..... and if things don't go exactly as he expects them to he will throw quite a violent tantrum. i.e. one particular carer may usually take him to the shops but if somebody else has to do it instead he will become aggressive.

They are hoping to get (him) into a new college soon and also are hoping to get a week's respite but they don't know how they are going to achieve this with his current behaviour.

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# The value of recognising shared characteristics

## Story 1: 18 year-old male with CdLS.

- Eye contact poor, keen preference for routine and various repetitive and stereotyped behaviours and speech. Selective mutism.
- A move to college, where the emphasis was on flexibility and student choice led to significant deterioration in his mood and behaviour.
- The college were not able to/willing to modify their programme.
- Increasingly tearful and withdrawn, stopped taking part in his usual daily activities, refused to go to college.
- Since received a formal diagnosis of ASD.

## Story 2: 18 year-old male with CdLS

- Period of regression in his late teens.
- Recognition by teaching staff and other professionals early on that he showed many characteristics of ASD, led to his being transferred to specialist autism provision, resulting in significant improvements in his mood and behaviour.

Moss & Howlin (2009), *JIDR*, 53, 852-872

## BOX 6 EXAMPLE OF A SUCCESSFUL TRANSITION FROM SCHOOL TO WORK

After visiting two mainstream colleges for typically developing young adults, Craig felt that he did not want to attend these colleges when he left school. Consequently, his father secured him a part-time job at his work place, a steel works company. Craig was gradually integrated into the workplace over a long period of time. Initially Craig worked two hours per week at the steel works and this has gradually increased to three days a week, working four hours on each day. Over time this will increase until Craig works there full time.

During his time at the steel works company, Craig's responsibilities have also increased. Initially, Craig's job mainly involved sweeping the shop floor. Craig now has a wide range of responsibilities such as, shredding paper, processing timesheets of other employees and tidying up.

Craig's work colleagues feel that he has increased in confidence since starting to work there. Although Craig was quite quiet when he started work, he has become much more sociable over time, greeting other members of staff when he sees them around the workplace and also enjoying their company at work. Craig often says that he really enjoys working there.

The key to this successful move for Craig was listening to Craig's concerns, gradual change and making sure there was predictability and routine.



Craig Ryder happy to work with people (above) and get on with the job (below).



## We think.....

- For some people with CdLS there are changes in behaviour around late adolescence/early adulthood (approximately):
  - Becoming less sociable with unknown people (preferring own company)
  - Becoming more reliant on routine and (and disliking unexpected change to routine, perhaps avoiding new experiences)
  - Becoming more impulsive (emotional outbursts)
  - Becoming more anxious (especially around new experiences and change)
- These behaviours are related to some specific cognitive abilities:
  - Working memory: holding recent information in a store and using it
  - Flexibility: being able to plan when change occurs, being able to switch from one thing to another
  - Inhibition: being able to stop yourself from doing something and controlling your emotions
- This may explain why ASD like behaviours seem to emerge
- We do not know:
  - What is causing these changes
  - The age they might occur at and who might experience them
- We are fairly sure that:
  - The environment someone is in effects how they cope with any of these changes
  - Medication is unlikely to be helpful

## So.....

- Well rehearsed, well known activities, people, places (predictability) are preferred
- Being able to do well rehearsed things may give an illusion of being good at everything
- Need more help with new (unpredictable) activities, people, places.
  - Gradual and long introduction (short periods), same thing every time for the first few times then increase variability
- Unexpected disruptions to routine can cause major 'upset'
  - Support and understanding at these times (not won't but can't)
  - Everything back to 'normal' asap
- Working memory may become shorter
  - One phrase at a time, repeating is OK
  - more trials to learn new things
  - schedules for the future
- Time to speak and respond



**Core Funding**

Cerebra

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- Research Autism
- NE Essex PCT
- Birmingham Children's Hospital
- Angelman Syndrome Foundation (USA)
- Newlife
- National Autistic Society
- ESRC
- Tuberous Sclerosis Alliance

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