

# Down Syndrome

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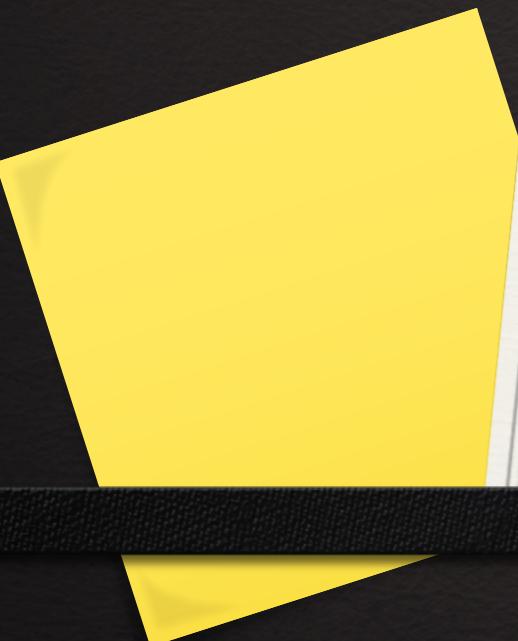
# Today's Session

## Part 1: Down Syndrome (DS), an overview

- Genotype of Down Syndrome
- Phenotype of Down Syndrome [Physical characteristics; Behaviour; Intellectual Functioning]
- Cognitive Profile [Strengths and Needs]
- Factors affecting the learning profile children with DS

## PART 2: Approaches and Strategies to support a child with Down Syndrome

- Other key Areas for support: Social Interaction and Language Skills; Memory & Attention; Motivation; and Handwriting



# Part 1: Down Syndrome (DS), an overview

# Down Syndrome



- ① Down Syndrome is the most common cause of intellectual disability (Sinet, 1999; Coyle, Oster-Granite & Gearhart, 1986)
- ② Evident in every ethnic group (Selikowitz, 1997)
- ③ Genetically determined syndromes associated with ID include: Down syndrome, Angelman, Cornelia de Lange, Fragile X, Lesch-Nyhan, Prader-Willi, Rett, Smith-Magenis, tuberous sclerosis complex, Velocario-facial and Williams syndromes (Carr & O'Reilly, 2007)

# Quality of Life

- Services for people with ID have progressed through a number of theoretical models over the decades
  - Separate institutions
  - Then separate schools
  - Progress towards inclusive models toward the end of the last century (Carr et al, 2007)

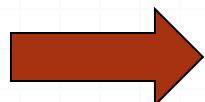
# Inclusion

Concept of inclusion as 3 generations of activity and practice (Turnbull et al. as cited in Wehmeyer & Lee, 2007)

1. Focus on the rights-base for inclusion (e.g. 'where' people with disabilities should be educated)
2. Focus on inclusive practice on the 'how' of inclusion (e.g. role-play, cooperative learning)
3. Focus on new perspectives that emphasise quality of life for individuals with ID (e.g. person-centred planning, maximising engagement/participation/social relationships/independence)

# Functional models and definitions of ID

- Emphasis in the literature on importance of identifying support needs, supportive environments and programme participation are recognised. How is this person around them? functioning in the world

 Shift in focus to strengths and needs of individuals.

- The AAMR-10 distinguishes between four different levels of support:

- 1) Intermittent
- 2) limited
- 3) extensive
- 4) pervasive

# *Down Syndrome*

## *Genotype*

# Types of Down Syndrome

- Down Syndrome is often referred to as Trisomy 21, however, the ICD 10 distinguishes between different forms of Down Syndrome:
  - Trisomy 21 (meiotic nondisjunction); 95% incidence rate among those presenting with Down Syndrome (Rubin & Strayer, 2007)
  - Trisomy 21 translocation; 4% of cases of Down Syndrome (Rubin & Strayer, 2007)
  - Mosaicism (Trisomy 21 mitotic nondisjunction); 1% of all cases of the condition (Rubin & Strayer, 2007).

# Aetiology

- Originally, it was believed that Down Syndrome occurred as a result of the presence of an extra copy of the twenty first chromosome (Patterson, 1987)
- Research (Korenberg et al., 1994; Delabar et al., 1993) indicates that Down Syndrome is caused by an extra amount of the critical portion of Chromosome21, i.e. a region around 21q22.2 with D21S17 and ETS2 as boundaries, on Chromosome21
- In Down Syndrome, the presence of an extra set of genes within the critical region leads to overexpression of the these genes

# Distinctive Characteristics

It is this overexpression, which researchers believe may contribute to the key characteristics of the Down Syndrome phenotype, e.g.

- an overexpression of the DYRK gene may result in **intellectual disabilities** (Galdzicki et al., 2001)
- the **skeletal abnormalities** evident in Down Syndrome may be attributed to an overexpression of gene ETS2 (Sumarsono, 1996)
- an overexpression of gene DSCAM may be accountable for **heart defects** in people with Down Syndrome (Kosaki et al., 2005)

# Down Syndrome Phenotype

*Range of distinctive characteristics*

# Think-Share

- Jot down 3 points that springs to mind when you think of supporting Individuals with:
  - Down Syndrome
  - General Learning Disability

A point to note on terminology

Intellectual Disability (Clinical Services)

versus

General Learning Disability (DES)



# Physical Characteristics

Down Syndrome is characterized by distinguishing facial and physical features, including:

- Characteristic head appearance - Small head and mouth, flat face with increased distance between the eyes and a flat nasal bridge. Marked depression in the posterior of the head.
- Small ears with anomalies of the folds.
- Hyperflexibility – an excessive ability to extend the joints.
- Broad feet with short toes and space between the first and second toes.
- Short, broad hands with a shortened, incurved fifth finger.
- Short, stocky body with short neck resulting from vertebral abnormalities

# Physical Characteristics

Down Syndrome also affects the skin and muscular structure and those with the condition can present with the following features:

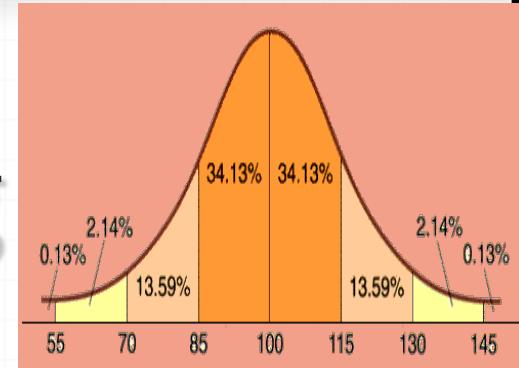
- ① Hypotonia - low muscle tone
- ① Ocular irregularities - Narrow and upward and outward slanting of the eyes. A ring of minute white spots the iris. Epicanthic eye-folds.
- ① A single transverse crease across the palm.
- ① A protruding tongue
- ① Excess skin on the back of the neck

# Physical Characteristics

In addition to the anomalies in physical appearance, people with Down Syndrome often experience a variety of medical conditions:

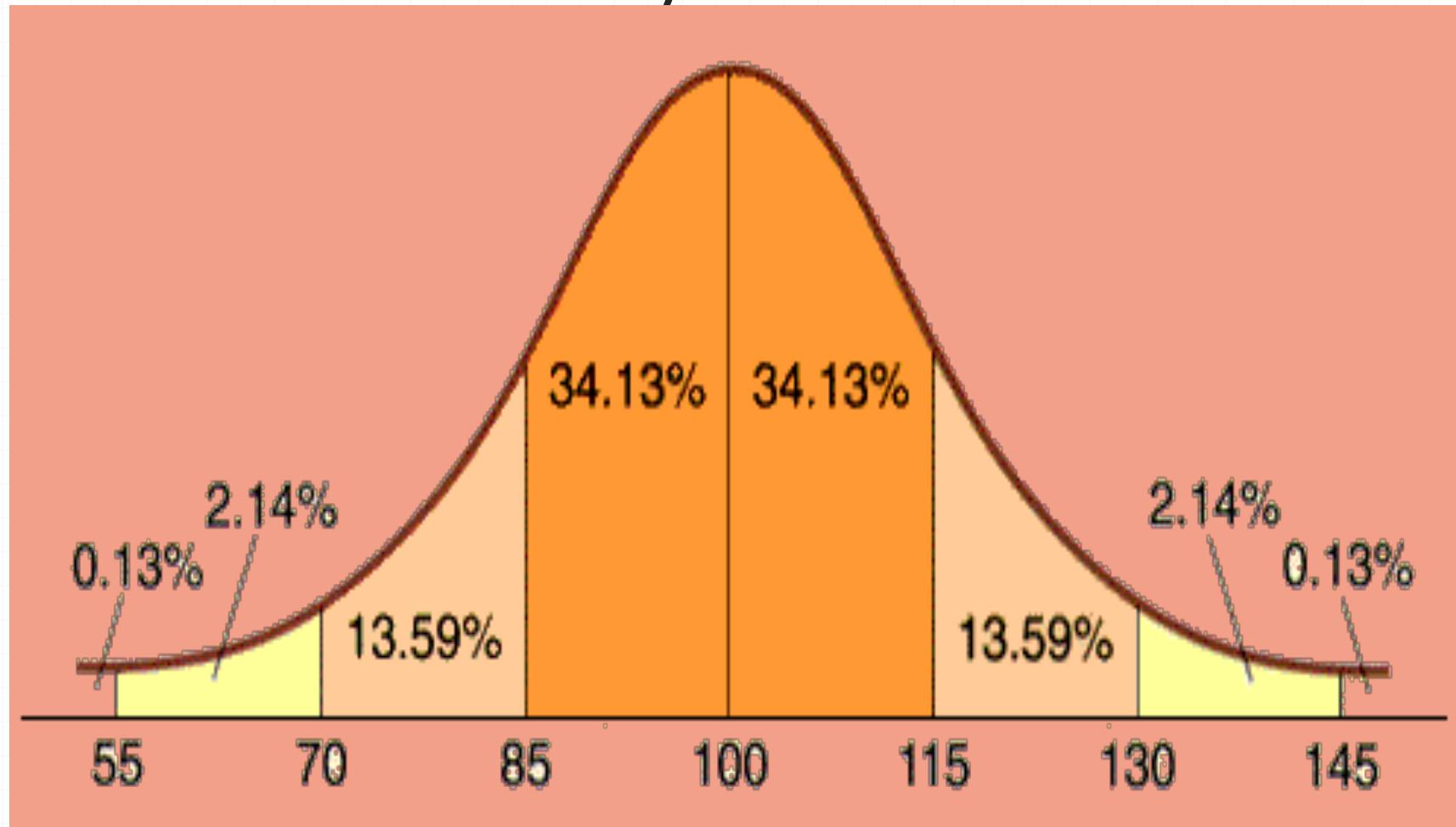
- congenital heart disease – 50% of babies with Down Syndrome (American Academy of Paediatrics – Committee on Genetics, 2001)
- gastrointestinal disorders
- hearing difficulties
- visual impairments, e.g. cataracts and glaucoma (Newman, 2004)
- increased risk of developing diseases such as Hirschsprung disease (Badner et al., 1990), leukaemia (Ugazio et al., 1990) and Alzheimer's (Masters et al., 1985)

# Intellectual Functioning



- Intellectual impairment is one of the most prominent features of Down Syndrome (Roubertoux & Kerdelhue, 2006; Fischler & Koch, 1991)
- The intellectual functioning of the majority of people with Down Syndrome is within the Mild to Moderate range of learning disability/ID (Vicari, 2004)
- However, people with Down Syndrome do not form a homogeneous group and there is a difference in the degree to which the syndrome impacts on an individuals' IQs and IQ scores can range from thirty to seventy

# Distribution of scores in diagnosing ID/GLD



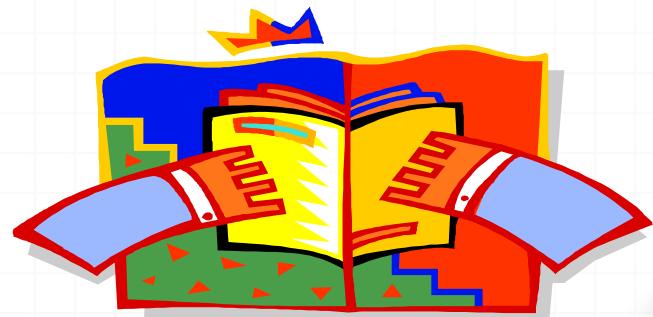
# Cognitive Phenotype

Strengths	Needs
<ul style="list-style-type: none"><li>o Visual awareness</li><li>o Visual learning skills</li><li>o Social Learning</li><li>o Social Skills</li><li>o Pragmatic Skills</li><li>o Kinaesthetic Learning</li></ul>	<ul style="list-style-type: none"><li>o Speech and Language Delay:</li><li>o Deficits in Memory:</li><li>o Delayed Motor Skills:.</li><li>o Poor concentration</li><li>o Poor thinking, reasoning and problem-solving abilities</li><li>o Sequencing Difficulties</li><li>o Generalising information</li><li>o Consolidating new information</li></ul>

# Cognitive Profile – Strengths

## Strong visual awareness & visual learning skills:

- Children with Down Syndrome display strengths in the area of visual perceptual skills.
- Their ability to process information presented in visual format exceeds their ability to process information presented orally.
- Strengths are also evident in the area of memory for visual information.



# Cognitive Profile – Strengths

- Social Learning (the ability to model behaviour of peers)
- The majority of children with Down Syndrome not only enjoy interacting with their non-disabled peers, but actually learn from this type of social engagement (Buckley & Sacks, 2001)
- Peer mentoring has been proven to be an effective method of assisting students with Down Syndrome to acquire academic and social skills (Lewis & Doorlag, 1987).
- Studies revealed that when children with Down Syndrome were paired with non-disabled peer tutors they achieved considerable gains in word recognition, oral reading and basic writing skills.



# Cognitive Profile - Strengths

## Social Skills:

- It is widely accepted that sociability and social skills are generally areas of **relative strength** for these children (Sadock & Sadock, 2007).
- Children with Down Syndrome usually develop age-appropriate social behaviour and conform to societal norms, if they receive encouragement and explicit instruction (Buckley & Sacks, 2001).
- However, it is important to note that children with Down Syndrome can miss non-verbal cues and, thus, their understanding of social situations can be impaired



# Cognitive Profile - Strengths



## Pragmatic Skills:

- Children with DS often adopt a pragmatic approach to problem solving (Selikowitz, 1997). Examples evident in their use compensating strategies, e.g. mimicry, gesturing, using actions, etc., in order to make themselves understood. Descriptions of a 'gesture-advantage' (Caselli et al., 1998).
- May 'overuse' their social skills to compensate for other weaker domains of functioning (Fidler, 2006).
- High rates of off-task social behaviour when children with DS are presented with challenging cognitive tasks (Kasari & Freeman, 2002). [reported in several studies]

# Cognitive Profile – Strengths

## Kinaesthetic Learning:

- In addition to visual strengths, and despite fine and gross motor difficulties, many children with Down Syndrome display a preference for the kinaesthetic learning style. These children benefit from engaging in practical, hands on activities (Hull Learning Services, 2004).



# Cognitive Profile- Needs

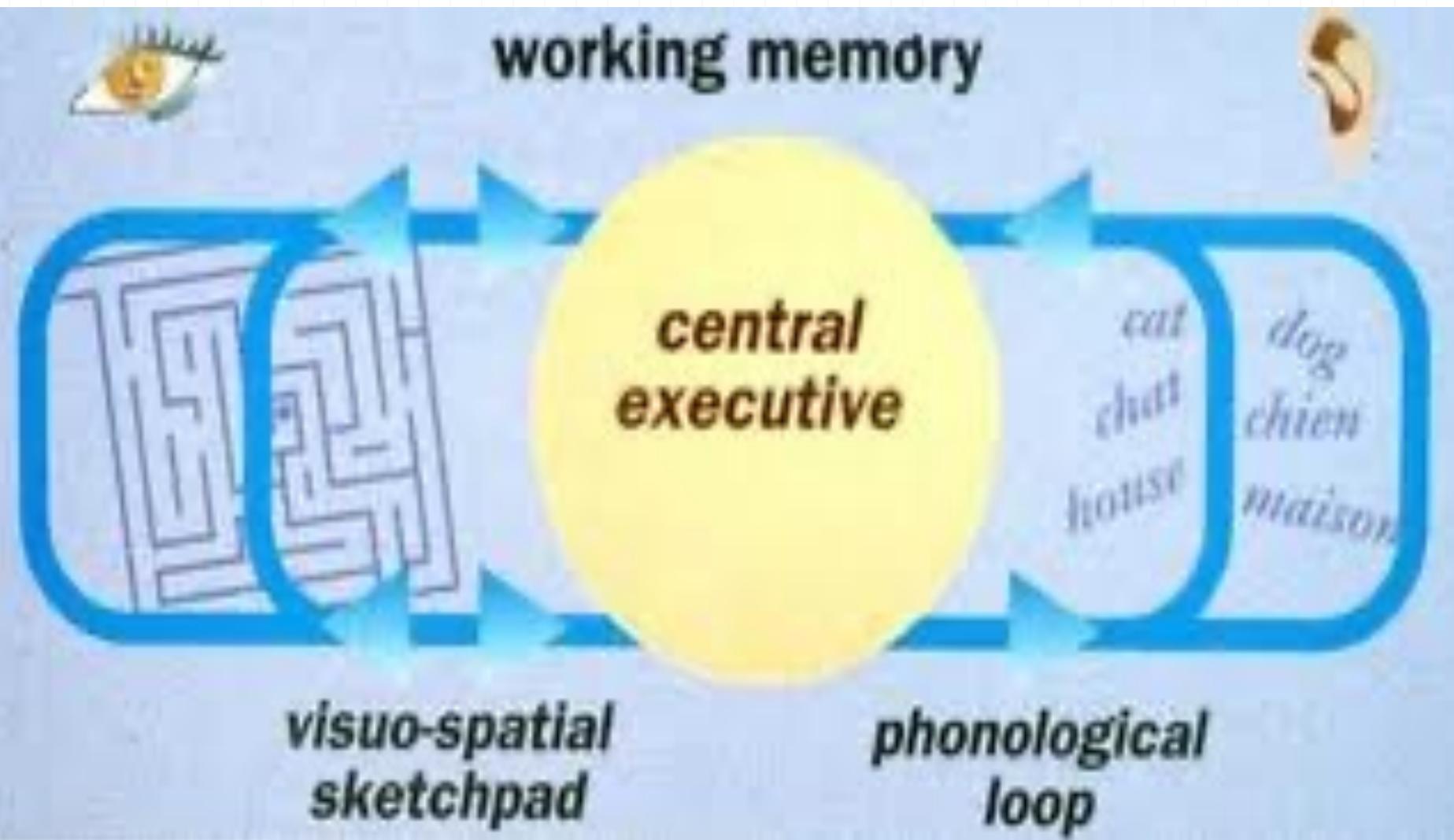
- Speech and Language Delay:
- Speech and language development is usually the area of most significant delay for children with Down Syndrome (Buckley & Sacks, 2001)
- Children with Down Syndrome present with an unique profile of speech and language characteristics which is characteristic of the syndrome and distinct from the language profile of typically developing children matched for nonverbal mental age (Chapman and Hesketh, 2001; Abbeduto & Chapman, 2005)

# Cognitive Profile – Needs

## Speech and Language Delay cont...

- The linguistic profile of children with Down Syndrome is marked by deficits in expressive language, which is most severe in expressive syntax (Chapman, Hesketh & Kistler, 2002). Deficits in motoric components of speech.
- The delay in language can be the culmination of a number of factors, such as physical (including hearing impairment), perceptual and cognitive.
- Receptive language and comprehension abilities usually exceed the individual's expressive language skills. Consequently, these children's understanding of language surpasses their ability to use language (Chapman, 1999; Sigman & Ruskin, 1999).

# Cognitive Phenotype – Needs



# Cognitive Profile – Needs

## Deficits in Memory:

- People with Down Syndrome experience considerable deficits in memory. The syndrome impacts on working memory, long-term memory and auditory memory (Nadel, 1996; Brown et al., 2003)
- There is a predominant delay in working memory development (Buckley & Sacks, 2001) with children experiencing particular difficulty with verbal short-term memory which leads to auditory processing difficulties.
- As working memory plays a pivotal role in the areas of thinking, problem solving and reasoning, children with Down Syndrome experience difficulty completing tasks that draw on these skills.
- Conversely, memory for visual information is a relative strength for children with Down Syndrome

# Cognitive Profile- Needs

## Delayed Motor Skills:

- o Both fine and gross motor disturbances are characteristic of the Down Syndrome profile and may occur as a result of hypotonia (Shumway-Cook & Woollacott, 1985).
- o Delay in the development and refinement of motor skills can impede an individual's ability to acquire self-help skills, to manipulate the school utensils, etc.



# Cognitive Profile- Needs

- o Additional Difficulties:**
- i. Poor concentration
- ii. Poor thinking, reasoning and problem solving abilities
- iii. Sequencing Difficulties
- iv. Difficulty generalising information, i.e. applying knowledge to new situations
- v. Difficulty understanding abstract concepts
- vi. Difficulty consolidating new information



# Behavioural Profile

- o Traditionally, terms such as 'lazy' and 'stubborn' have been used to describe children with Down Syndrome. More recently, may be physical & biological reasons for stubbornness observed.

Children with DS display fewer behavioural problems in childhood than individuals with other forms of ID but more than those observed in developing children (Dykens, 2007).

- o In children and adolescents with DS: Low rates of severe aggression (6%) and high rates of low-level aggression (e.g. disobedience) were measured on the Child Behaviour Checklist (Dykens et al., 2002).
- o Mood and anxiety disorders more prevalent than other psychiatric diagnoses (Patti & Tsioris, 2006).

# Behaviour Profile

o Considerable variability in the profile of children with DS  
(Dykens, 2007)

Protective Factors	Risk Factors
Enriched environments (EEs): neural plasticity?	Sleep problems
Relative strengths in sociability	Low serotonin/ Obesity/hypothyroidism/cardiac complications
babyfaced' craniofacial appearance	
Caregiving/parenting environment	

# Learning profile: How Down syndrome affects learning

Clumsiness

Handwriting difficulties

Dressing difficulties

Poor metacognition

Poor rehearsal skills  
e.g. silent voice

Poor executive functioning skills

Fine motor problems

Gross motor difficulties

Language delay

Cognitive difficulties

Weak auditory memory

CHILD

Short attention span

Hearing problems

Difficulties with phonics

Difficulties in number skills

Problems being understood

Word finding difficulties

Difficulties in following instructions

Misunderstanding school rules

Immature social skills

# Factors affecting the learning profile children with Down syndrome (DS)



- Children with DS tend to have:
  - ❖ Strong visual awareness and visual learning skills.
  - ❖ The ability to learn and use sign, gesture and visual support.
  - ❖ The ability to learn using written word
- The desire and ability to learn from their peers, to imitate and take their cue from them

BUT:

- Delayed motor skills, fine and gross, leading to clumsiness and manipulation difficulties
- Auditory and visual impairments leading to hearing and sight loss
- Speech and language delay. Problems with articulation , comprehension and expression
- Poor short term auditory memory. Problems with consolidation and retention
- Short concentration span. Difficulties generalisation, thinking , reasoning i.e metacognition

**NB. Strong visual learners and poor auditory learners**

## **Part 2 : Approaches and Strategies to support a child with Down Syndrome**

# The teacher and SNA team: Key areas for support

- Social Interaction and Language Skills
- Memory & Attention
- Motivation
- Handwriting

# Interacting with a Child with Down Syndrome

- Always face the child
- Make eye contact with the child
- Try to speak to the child at the same level – your lip patterns will be perceived better when you are face to face
- No need to raise your voice or slow down your speech
- Repetition will help the student internalise this behaviour, which will soon turn into an automatic response



# Language

## Language

- o Speech, language and communication skills are practised everyday through interacting with others. Therefore, it is essential to ensure that children with Down Syndrome have maximum opportunity to interact with others.
  
- o During these interactions it is of paramount importance to talk to the child. Even if the child does not respond or his response is limited, always speak in full sentences as exposure to language helps children to acquire language.

# Language cont.....

## Language

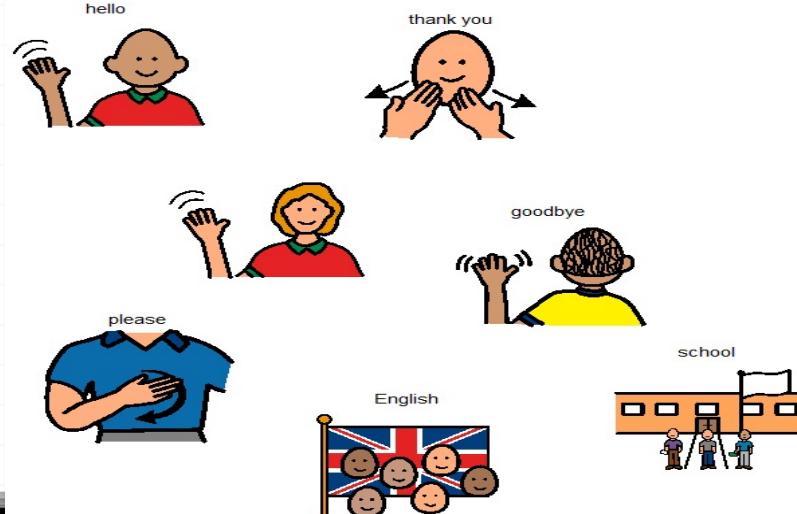
○ Grammar development begins when children can understand approximately 250 words. To optimise grammar development the following activities may prove useful:

- Reading
- Sentence games, i.e. ordering words
- Pacing boards
- Sequencing cards
- Story recall
- Focused personal books, i.e. individual book about the child.
- Open ended questions, e.g. “Tell me more....”

# Language cont.....

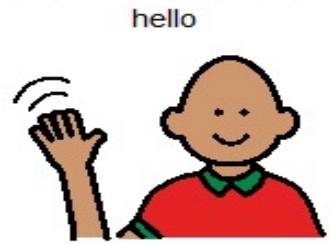
## Language

0 Conversation diary, i.e. ask the child about their day. Then transcribe what he says into grammatically correct sentences. NB: Ensure that the sentences are accompanied by pictures or photos as this will reinforce learning.

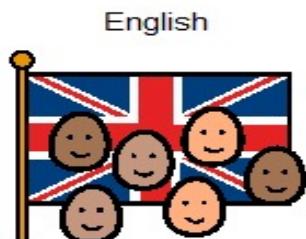


# Understanding Instructions and Visual Supports

## Boardmaker Software

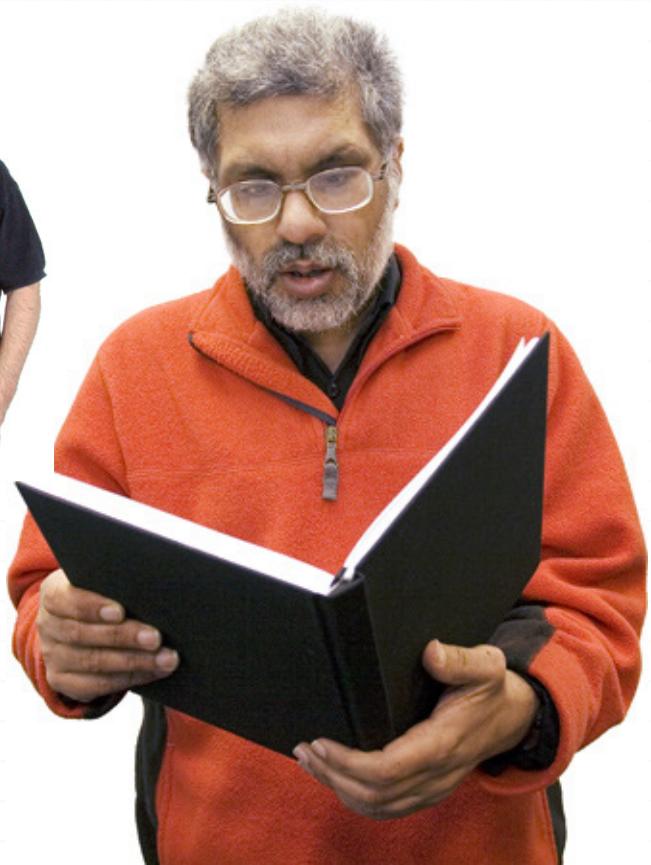


goodbye



# Understanding Instructions and Visual Supports

## Photosymbols



A concrete and visual Supports for Numeracy...

# Numicon



- Numicon is a multi sensory teaching approach designed to give children the understanding of number ideas and number relationships through the use of structured patterns, i.e. Numicon shapes.
- Numicon shapes provide an accurate visualisation of numbers
- Number shapes clearly show:
  - That each number is one more equal unit
  - The relative size of different numbers.
- Thus, the child develops his own mental imagery as they combine and compare the shapes.





# Motivation

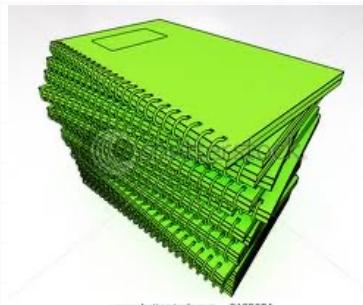
- o ALWAYS provide opportunities for the child to make choices as this will provide him with some sense of control over his day.
- o Develop a reward system to promote positive behaviour, e.g. reward chart, magic pot, sparkles, etc.
- o Provide positive role models or operate a 'buddy system' so that the child can learn from his peers. Modelling is an area of strength for children with Down Syndrome.
- o Provide sufficient warning and preparation time for transitions, changes or new demands.
- o Ensure that the child is facilitated to appropriately communicate his difficulties should any arise. It may be necessary to teach the child to articulate how he is feeling, e.g. "help", "I need more time", "I'm busy", "Go away, please", etc. It may be an idea to provide the child with cards containing these phrases so that he can give them to others when appropriate.
- o Provide the child with small jobs and responsibilities as this will boost his self-esteem

# Handwriting



① **Pencil Grip** – Consider whether it might be useful to provide the child with a moulded pencil grip. A number of different types are available through [www.thinkingtoys.ie](http://www.thinkingtoys.ie) or other educational supply stores.

② **•Multi-Sensory Approach** – Use multi-sensory approach that provides the child with opportunities to look, copy, trace, picture, cover, write and check.



# Memory & Attention cont...



## Instructions

- Children who have working memory difficulties can struggle to remember instructions and, therefore, they should always be encouraged to repeat instructions in order to ensure that s/he has understood them.
- The number of instructions given at any one time should be kept to a minimum. Instructions and directions should be short and succinct.
- The 4S strategy - *go Slow and Show, Say less and Stress* may be useful to ensure the child follows instructions. It might be useful to repeat any information given to the whole class, individually to the child. The child should also be encouraged to tell his teachers when he has not understood.

# Memory & Attention cont...



## Instructions cont...

- **Cued Listening** – Prepare students so they understand when it is time to listen. Visuals and gestures can be used to cue students to listen and look (i.e. point to your ear / eye, use Lamh signs). Using intonation in voice can also be used to emphasise key points of words can cue and focus listening. A discrete visual cue (i.e. touching one's ear) might be agreed between the child and his teachers to cue listening without undue disruption to the class or attention being drawn to him.
- **Repeat** – Repeat any information given to the whole class and if needed individually to the child.

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