
THE WILLIAMS SYNDROME COGNITIVE PROFILE: IMPLICATIONS FOR EDUCATION

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FIRSTLY, GREETINGS FROM EMILY FARRAN....



TALK OVERVIEW

- ◆ Examine the unique WS cognitive profile
 - A complex pattern of relative strengths and weaknesses

- ◆ Discuss recent research on some areas of the WS cognitive profile relevant to accessing education and development of independence...
 - Problem solving
 - Visuospatial cognition and navigation
 - Reading, writing and spelling
 - Early language development

- ◆ Implications for training and educational interventions

WS SCHOOLING (UK)

❖ **Primary school level**

- Many children with WS are in mainstream schools, with additional help
- Some children with WS attend schools for children with severe or moderate learning difficulties

❖ **Secondary school level**

- Many WS children move to special schools
 - ✓ More individualised teaching
 - ✓ Learning at a slower and less pressurised pace
- A few WS children continue in mainstream at the secondary level with support

EDUCATION AND INDEPENDENCE IN WS (IN THE UK)

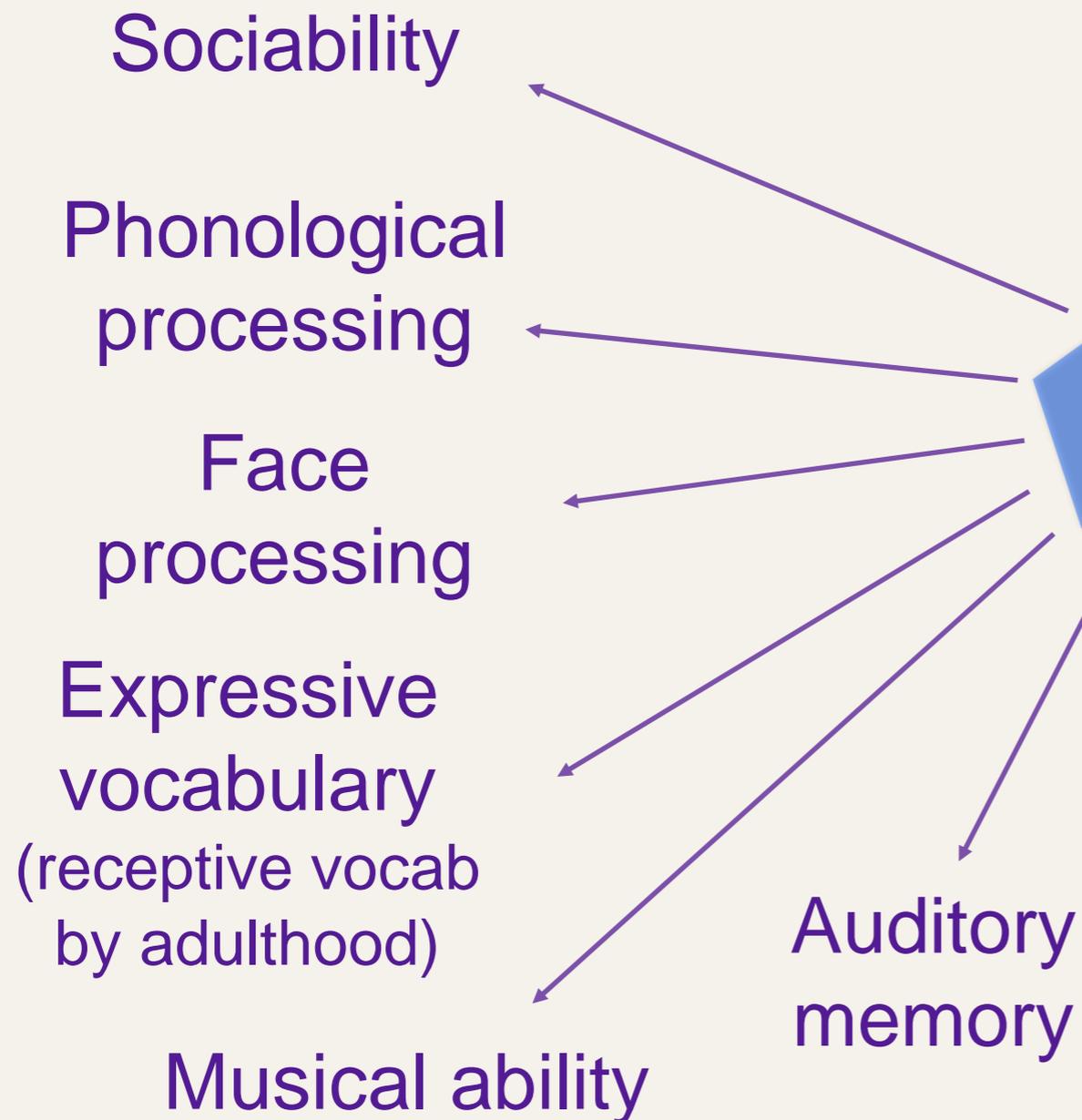
- ❖ WS associated with behavioural, social and cognitive difficulties that can effect education, independence and daily-living
- ❖ Children and young people with WS now more likely to attend mainstream school than before...
- ❖ However, education and employment attainments remain low: 7-12% adults with WS in independent work
- ❖ Ultimate goal of education in WS to develop independence, not just improve scholastically
- ❖ *'Independence is difficult to achieve for people with WS'*
(Stinton & Howlin, 2012)

THE WS COGNITIVE PROFILE

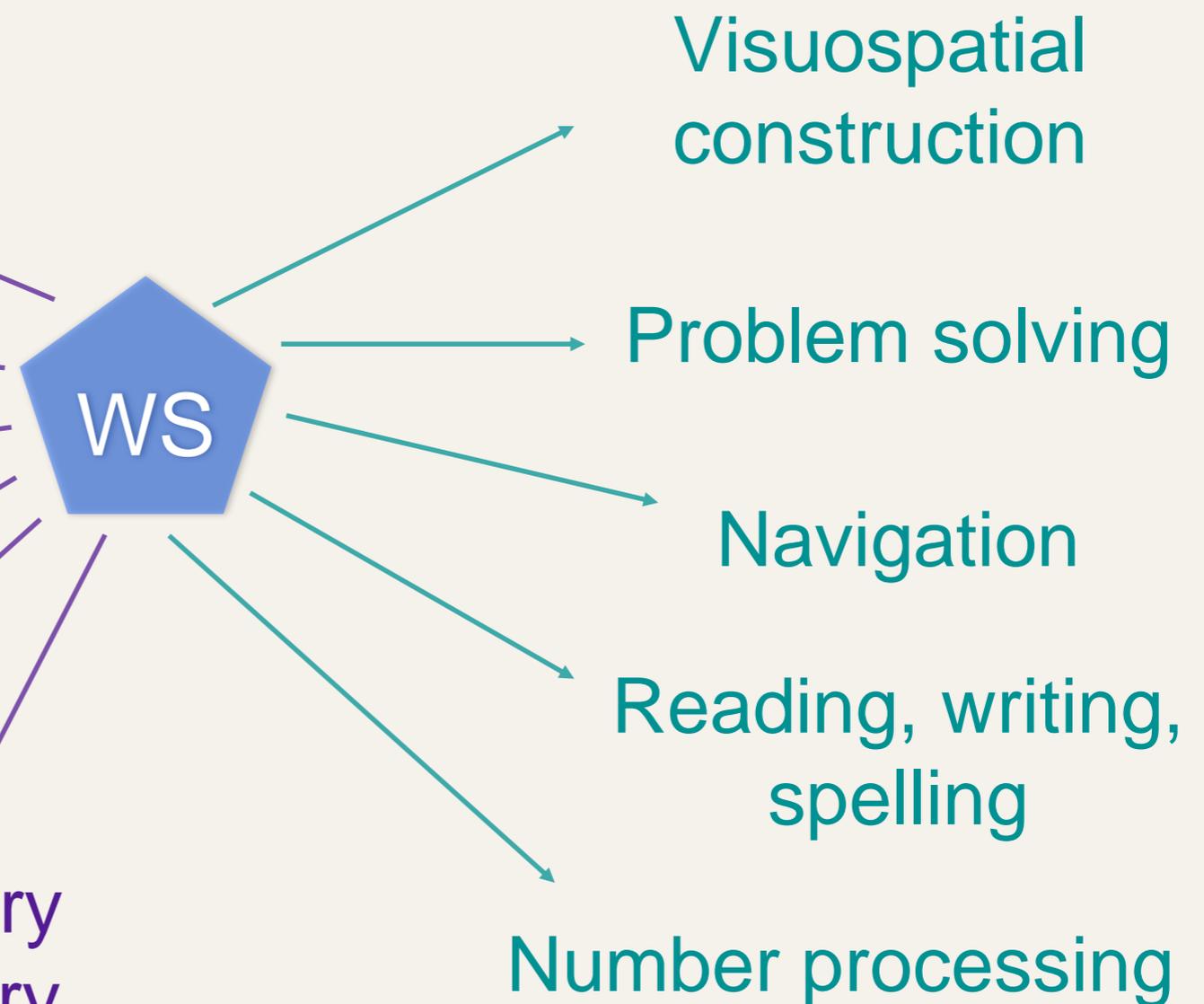
- ❖ IQ typically between 40 and 90 (Howlin et al.,2010)
- ❖ However, subtle pattern of strengths and weaknesses
- ❖ WS not the same across development, with changing cognitive profile across maturation
 - E.g., verbal abilities improve at a faster rate than non-verbal (Jarrold, Baddeley and Hewes, 1998)
 - Receptive vocabulary > expressive vocabulary in childhood, reverse by adulthood (Howlin et al., 2010)
- ❖ **Target education to training areas of weakness or compensation using strengths? What to focus on?**

THE WS COGNITIVE PROFILE

Areas of relative strength

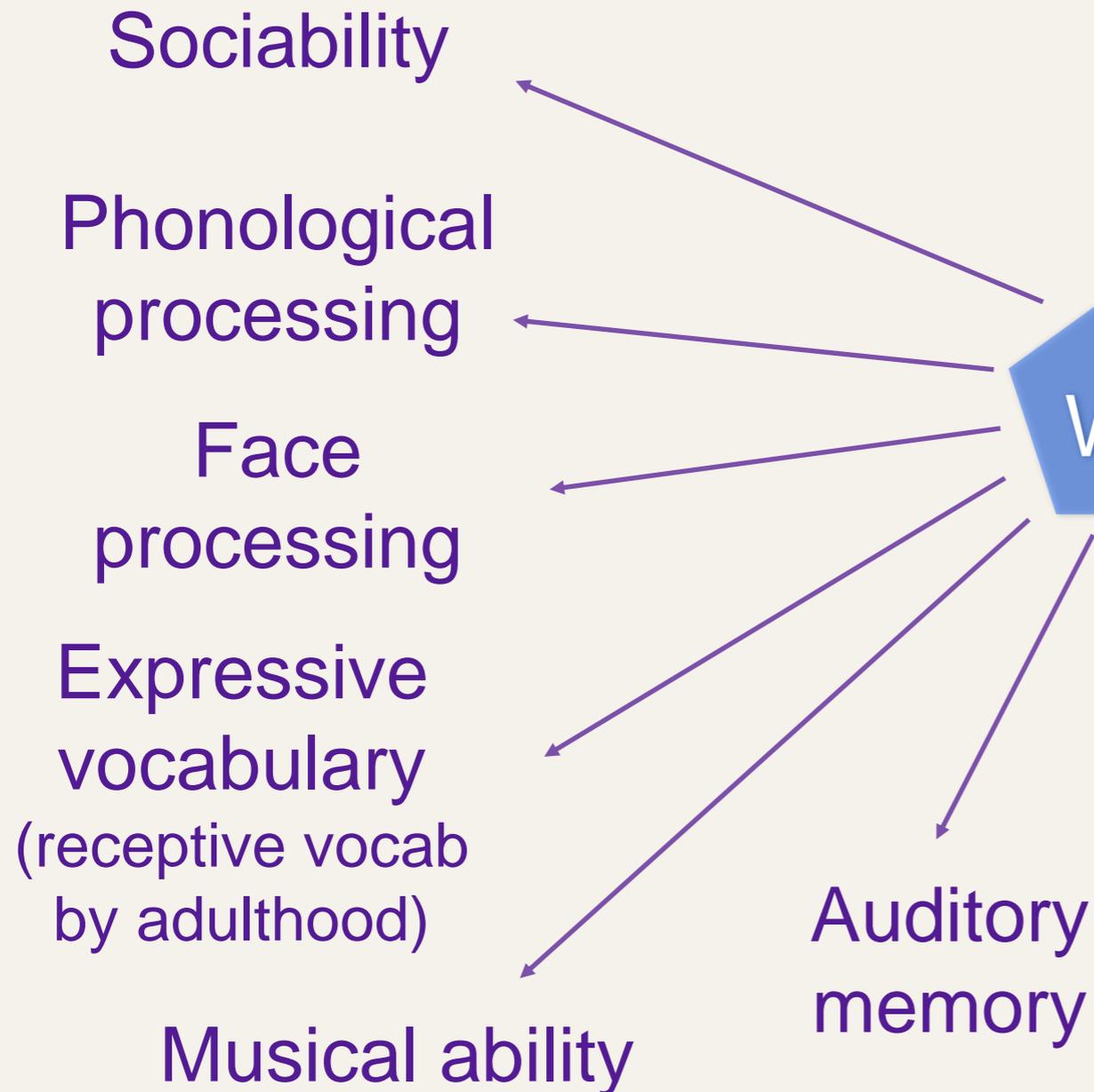


Areas of relative weakness

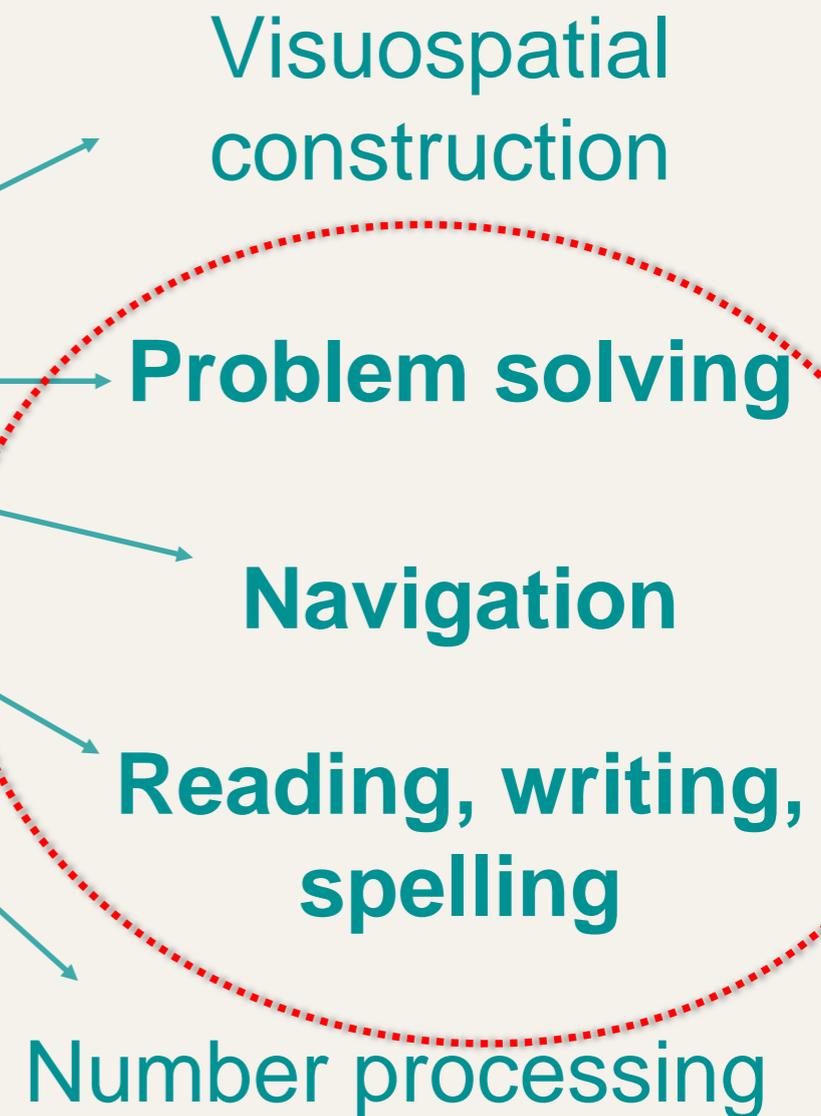


THE WS COGNITIVE PROFILE

Areas of relative strength



Areas of relative weakness



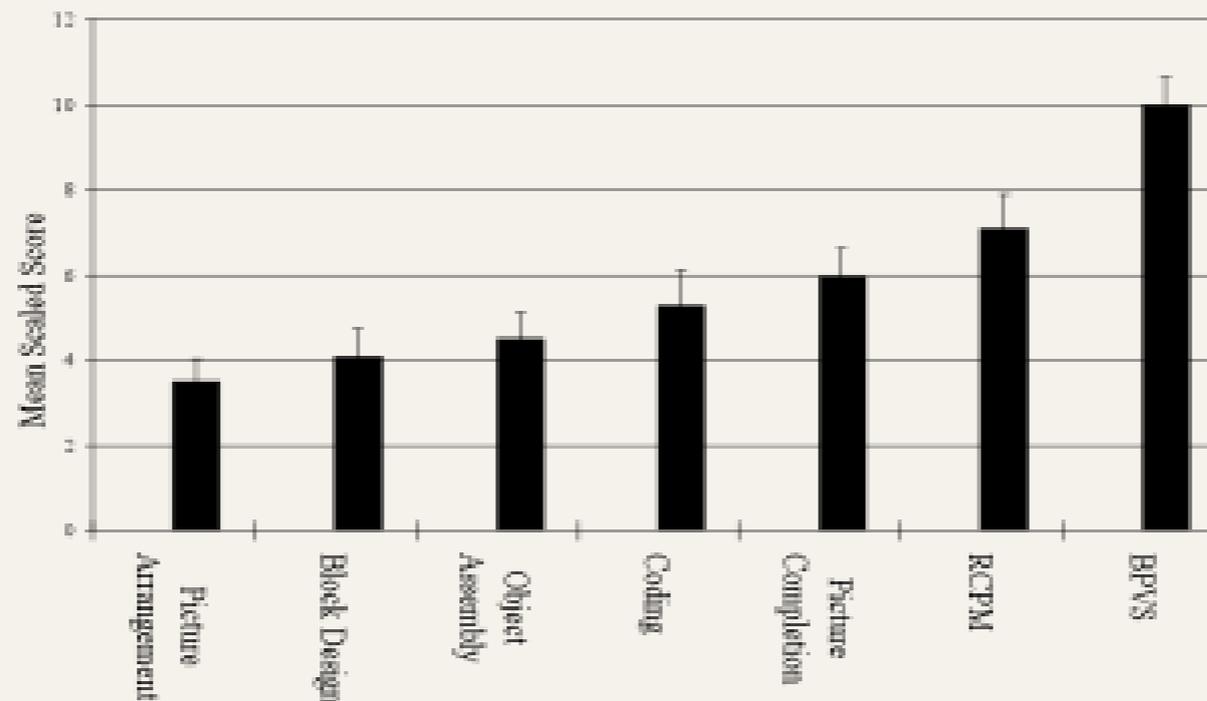
THE WS COGNITIVE PROFILE

RELATIVE VERBAL STRENGTHS CAN MASK SIGNIFICANT DIFFICULTIES IN OTHER AREAS.....

- ❖ Teachers report finding children with WS baffling and difficult to teach because their overall abilities have been overestimated
- ❖ Attention issues often missed (zoning out/blank face)
- ❖ Children with WS are sometimes placed in an inappropriate school in which they are unable to cope because of their lower abilities in non-verbal areas and their limited concentration
- ❖ However, in special education settings, focussing only on non-verbal abilities may have adverse effects on verbal and social cognition (especially also when placed with non-verbal children)

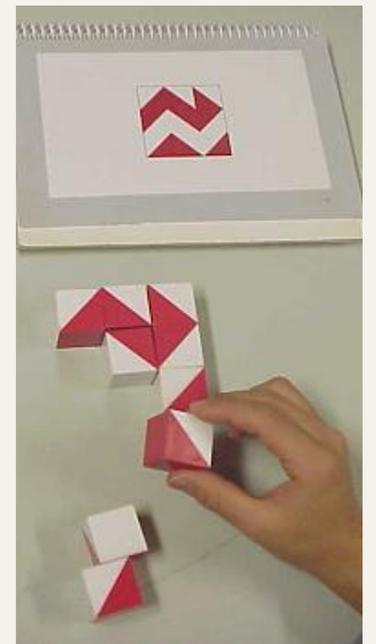
RELATIVE STRENGTHS - (NOT INTACT VS. IMPAIRED)

- ❖ E.g., in the verbal domain greatest strengths in concrete receptive vocabulary, whereas pragmatics and use of some forms of syntax and morphology are considerably weaker
- ❖ Maybe peaks and troughs within relatively impaired domain-specific processes, e.g., visuospatial cognition?



VISUOSPATIAL COGNITION IN WS

- ❖ Poor visuospatial construction in WS (small-scale tasks, e.g., block design)
- ❖ Able to learn a route in large-scale environment (in line with TD 5 yr olds) (Farran et al., 2010)
- ❖ Difficulties in 'allocentric/ global' coding of large-scale space for navigation (Broadbent et al., 2014)



VISUOSPATIAL COGNITION

- ❖ The ability to navigate is critical for independence and confidence

- ❖ For example:
 - ❖ knowing how to get to the local shop
 - ❖ the layout of a local supermarket
 - ❖ learning your way around a new school
 - ❖ knowing how to get to and from work
 - ❖ knowing what bus stop to get off at
 - ❖ learning strategies to help you find your way if you get lost

- **What strategies do individuals with WS rely on to navigate....?**

NAVIGATION STRATEGIES IN WS

- ❖ Previous findings suggest WS group do not use typical strategies to spontaneously navigate (Broadbent et al., 2014)

In a recent study....

- ❖ Used virtual environments (VEs) to examine ability to **learn a route with landmarks** and then ability to recall route once **landmarks removed**
- ❖ Examine the **reliance on landmarks** by TD and WS groups (do WS have ability to use sequential egocentric strategy when required?)
- ❖ Also test ability to learn route in VE with **no landmarks**
- ❖ Included a **visual-matching task**. Are individuals with WS able to correctly identify scenes from the maze like TD children?

OVERVIEW OF FINDINGS

- ❖ On 'landmark' route, individuals with WS in line with TD 5 year-olds (similar level of non-verbal ability) on number of trials and number of errors to learn route
- ❖ On 'landmarks removed' test, WS made significantly more errors than all TD groups
- ❖ On 'no-landmarks' route, WS required significantly more trials and made significantly more errors than all TD groups

OVERVIEW OF FINDINGS AND IMPLICATIONS

- ❖ Suggests a greater reliance on landmarks in WS than in typical development
- ❖ All groups performed well on visual matching task, no differences between any group
- ❖ Implications for training in WS.... Able to use a visual matching strategy to learn how to navigate
 - ❖ Are VEs useful for training?
 - ❖ Use of interactive web-technology (e.g. Google Maps)?
 - ❖ Web training programmes e.g., www.elstrad.eu
 - ❖ Use verbal strengths to support learning

PROBLEM SOLVING

ONLY EASY WHEN YOU KNOW HOW...

1. Fill the kettle the right amount

2. Boil the water

3. Find a (clean) cup

9. Wait for it to cool for the right amount of time

8. Carry to the right place



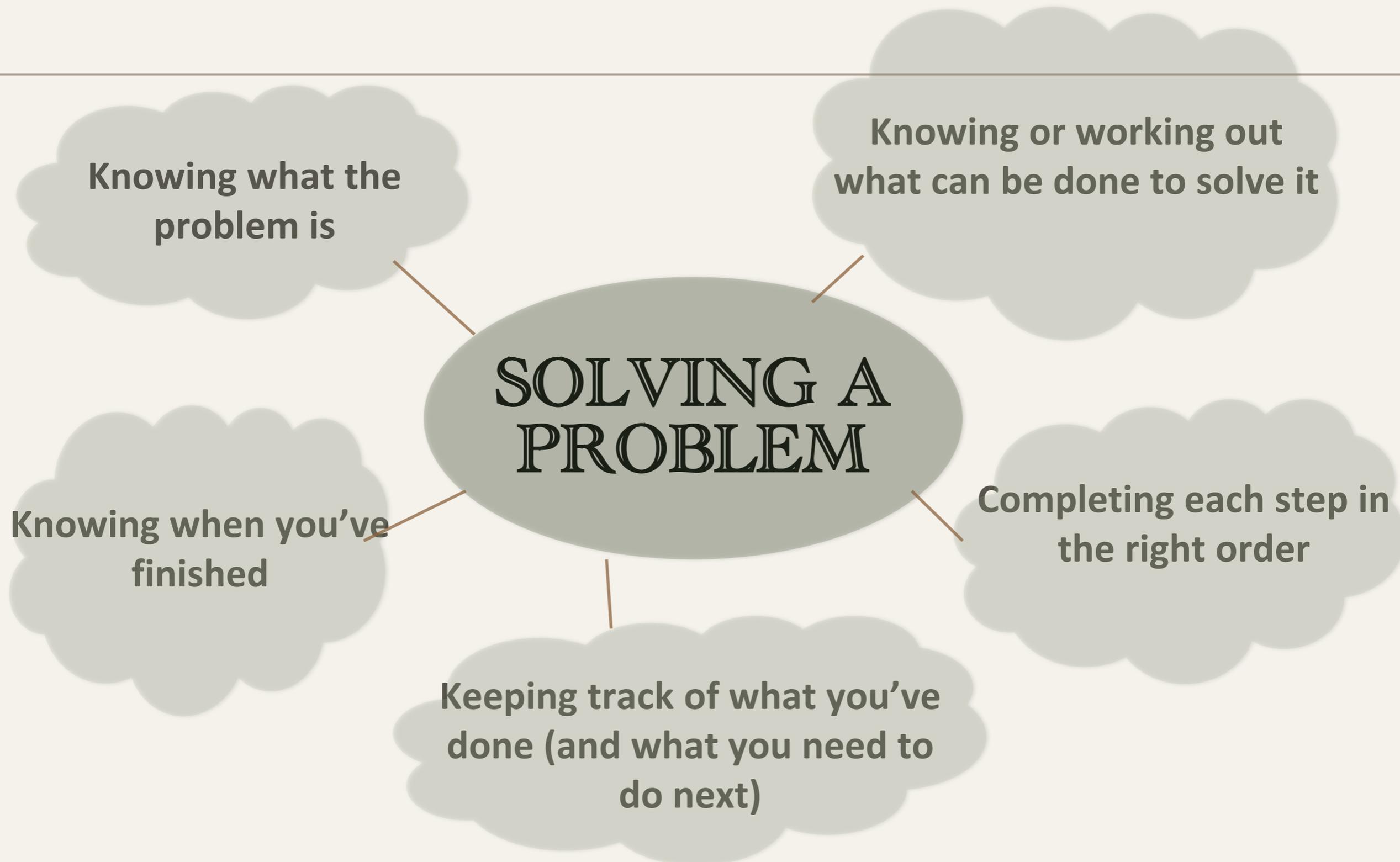
4. Locate the teabags

5. Put the teabag in the cup

7. Add milk and/or sugar, if required

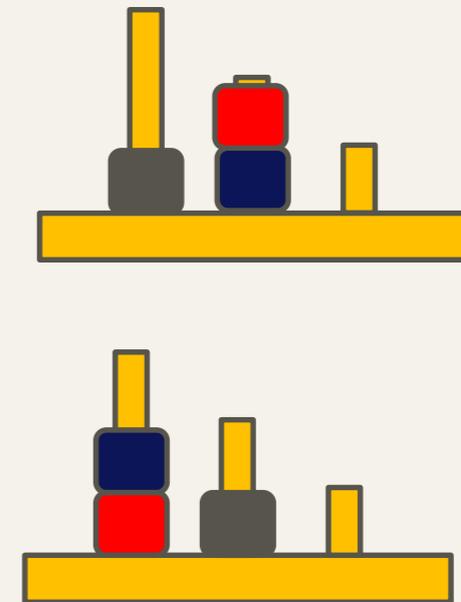
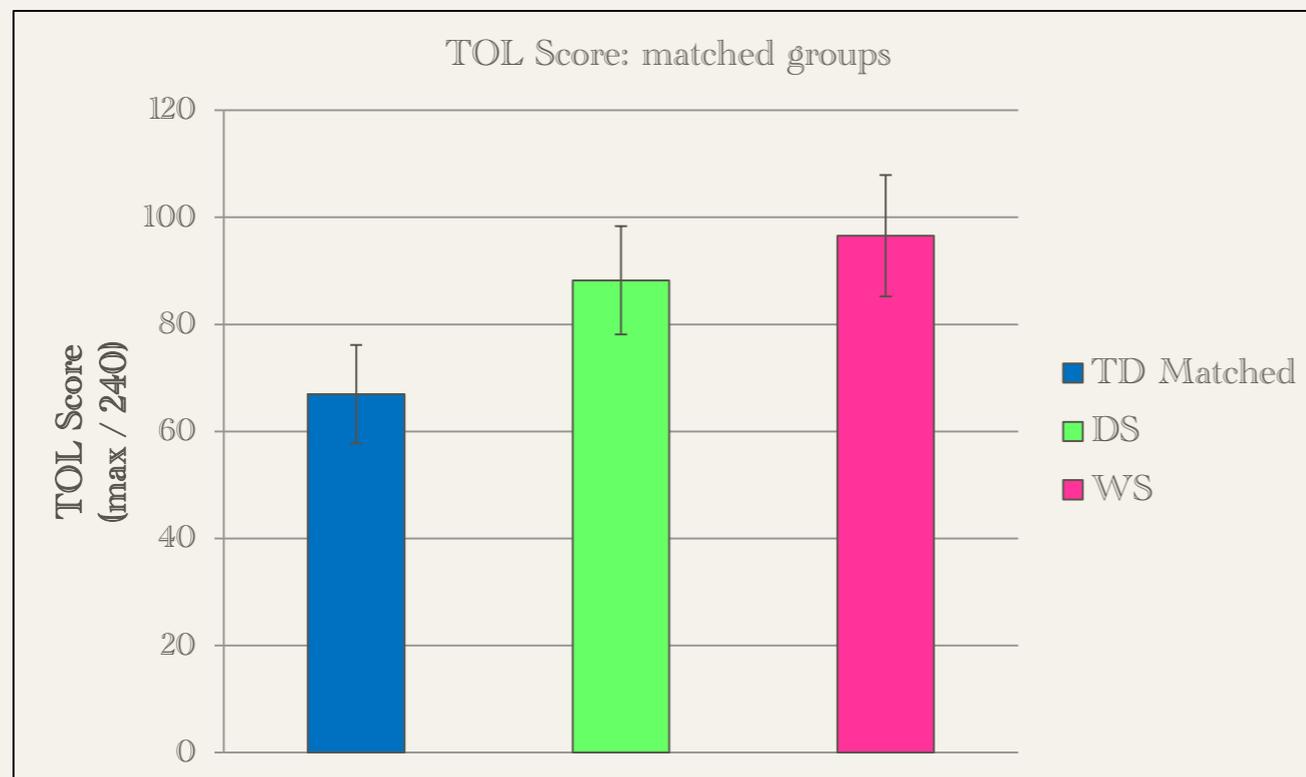
6. Pour the right amount of water safely

PROBLEM SOLVING



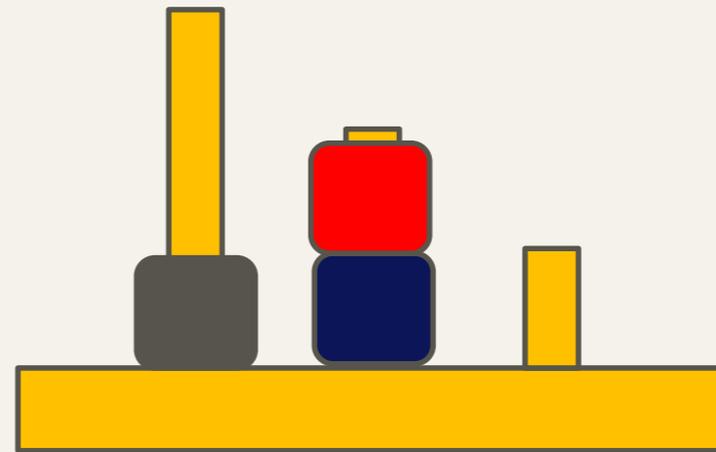
PROBLEM SOLVING IN WS

- ❖ Lab-based tasks: Tower of London
- ❖ $WS < TD$ (matched for non-verbal ability)



PROBLEM SOLVING STRATEGIES AND ERRORS

- ◆ Lab-based task



- ◆ Individuals with WS made a lot of perceptual errors
- ◆ We also know from other research that people with WS do not check their solutions as much as typical groups (e.g., Hudson & Farran, 2013)

WHY IS PROBLEM SOLVING DIFFICULT IN WS?

- ❖ Potential contributors: planning, rule changes, impulsiveness, verbal memory, non-verbal memory, verbal ability, non-verbal ability.....

In WS:

Problem solving is limited by:

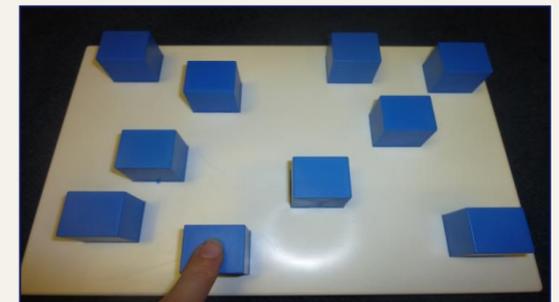
- planning
- spatial memory

Problem solving is facilitated by:

- verbal abilities



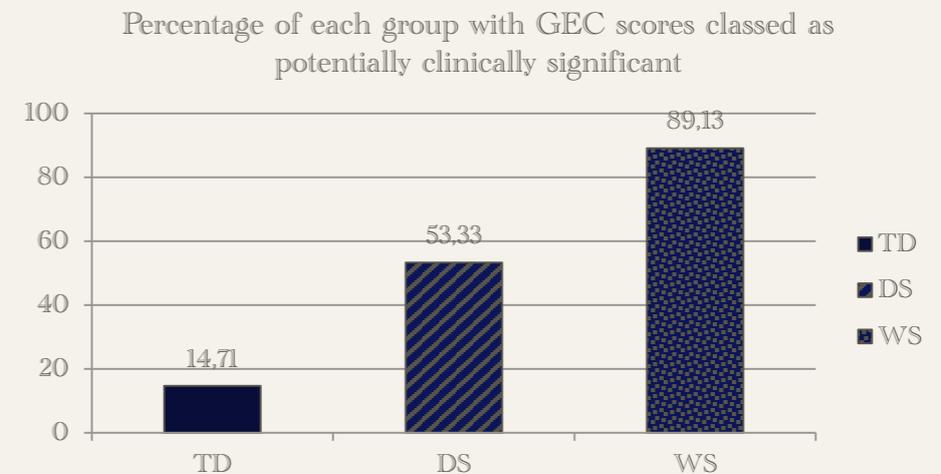
Planning



Spatial
memory

PROBLEM SOLVING CONCLUSIONS

Examining performance on lab-based tasks AND parent-reported problem-solving questionnaires....



- Limitations in planning skills and keeping items in memory can hinder problem solving progress in WS.
- Individuals with WS have a tendency to become emotional when they encounter a problem; the better they are at dealing with this emotional response, the more successful they are at solving problems.
- The relatively strong verbal skills observed in WS benefit everyday problem solving, particularly if this is a routine problem (brushing teeth, or getting dressed).

READING SKILLS IN WS

- ❖ Adults with WS have an average reading age of 8 years 8 months
- ❖ Individuals with WS rely on a **phonological** reading strategy (Levy et al., 2003; Laing et al., 2001) – contrast with DS reliance on sight-word reading strategy
- ❖ As yet, little research into the development of reading abilities in WS and development of phonological-awareness skills
- ❖ Currently no research on reading intervention programmes in WS
- ❖ Research suggests children with WS would benefit from combined oral-language-based and phonology-based reading training

TRAINING READING SKILLS IN WS

So....

- Approaches to teaching reading which rely on auditory skills and which emphasize a phonetic approach may be more effective
- Difficulties with reading may also be related to poor attention and distractibility....
- Standard beginning reading books (detailed pictures, colours, words on the same page) may be distracting. Books should have a few simple pictures or outline drawings which are less likely to distract

WRITING AND SPELLING IN WS

- ❖ Writing and spelling is poorer than reading
- ❖ Problems with forming the letters and spacing out and aligning words
- ✓ Exercises to improve tactile perception, motor control and eye-hand coordination, e.g. Tracing letters first
- ✓ Laborious task – tap into child's interests
- ✓ Encourage child to use speech to talk themselves through the exercises. Use the same verbal cues each time (e.g. "b goes down, then back up and around")
- ✓ Later stages, e.g. writing unfamiliar words. Encourage greater reliance on the auditory channel. E.g. by sounding and then spelling out the word verbally

EARLY LANGUAGE DEVELOPMENT

Child development and learning difficulties unit (CDLD)

- ❖ Although language is a relative strength, children with WS develop at a slower rate
- ❖ Use other strengths to support language development, e.g., Music
- ❖ Encouraging development of oral and vocal tract muscles
 - ❖ Pulling faces in the mirror,
 - ❖ Blowing candles or toys,
 - ❖ Licking things to develop tongue and facial muscles
- ❖ Although vocabulary a relative strength, poor grammatical skills
 - ❖ Teach grammatical structures by acting out sentences (e.g., “the dog is chasing the cat”) with toys, and then changing the structure (“the cat is being chased by the dog”)

INTERVENTIONS AND TARGETING EDUCATION IN WS

- ❖ Large-scale visuospatial cognition - training using strengths in visual-matching
- ❖ Problem solving - Planning, memory difficulties, and poor emotional regulation inhibit performance. Use of verbal strengths can support solving of problems
- ❖ Reading - interventions to target areas of relative strength, e.g., phonics reading approach preferable to whole-word approach, (Mervis & John, 2010)
- ❖ Language – although a relative strength, recommendations for targeting weaknesses using interests and strengths in other areas (e.g., music, face processing)

CONCLUDING POINTS

- ❖ WS cognitive profile characterised by a complex pattern of strengths and weaknesses, even within specific domains of functioning
- ❖ This has implications for training and educational interventions that need to be targeted to the WS profile
- ❖ At each stage of development, different strengths and weaknesses in the profile point to need for differing emphasis on education
- ❖ Need for individualised education plans
- ❖ Further research is needed on the impact of interventions that target specific weaknesses compared to those that focus on developing cognitive strengths in WS
- ❖ Aim of education in WS is for the development of independence, not just academic achievement

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