

Independence and navigation in individuals with Williams syndrome

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Structure of today's session

- Daily living, Problem Solving, Planning and remembering
- Spatial Navigation and Anxiety
- Discussion

Daily living



Leighanne Mayall

- » 50% of adults with WS require help with washing and dressing. Most adults with WS (80% to 94%) dependent on others for food preparation, cleaning and shopping (Davis et al., 1997).
- » 7-12% adults with WS in independent work (Stinton & Howlin, 2012).

Daily living

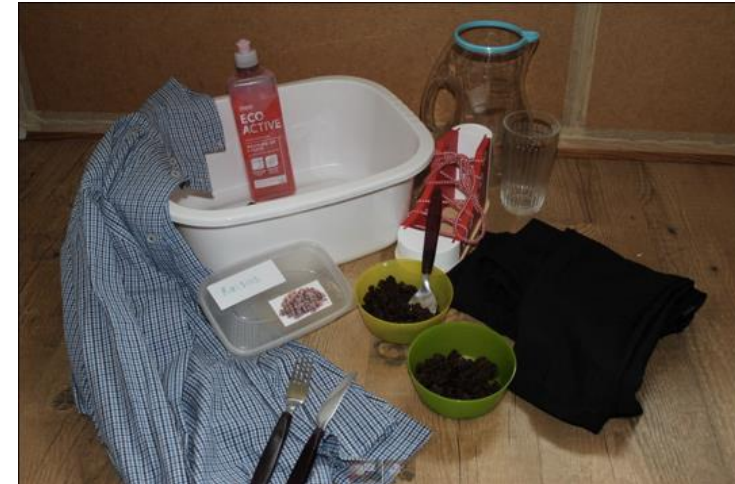


Leighanne Mayall

Previous studies are reliant on parent report

- » Participants: 21 individuals with WS aged 9-36 years and 18 individuals with DS aged 12-35 years.
- » Daily living ability was assessed in two ways.
 - Vineland Adaptive Behaviour Scale, second edition (VABS-II: Sparrow et al., 2005). Parent report.
 - Novel practical assessment of daily living ability (P-DLT). Participants were asked to complete a practical daily living assessment (P-DLT). These tasks were designed to have a strong motor element and be tasks that older children and adults would typically do everyday.

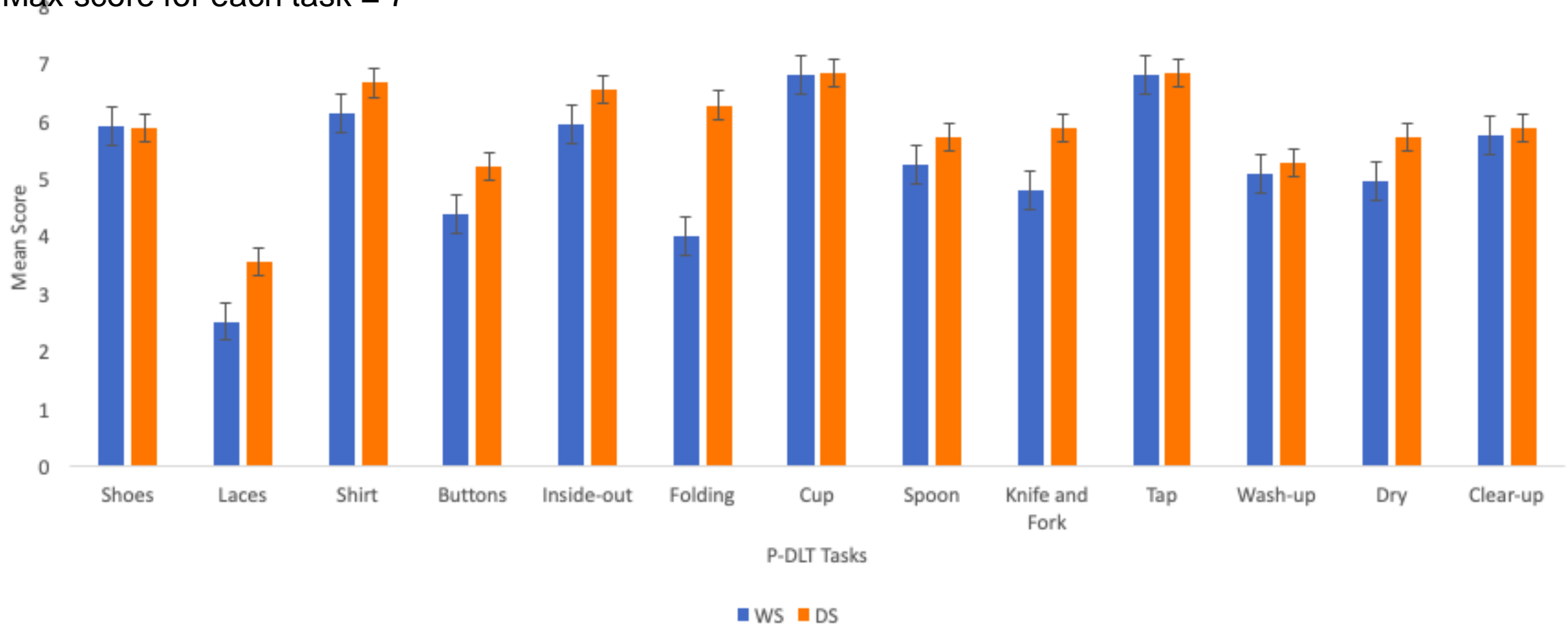
Putting on shoes (no laces)	Tying shoe laces	Putting on a shirt
Doing up buttons	Putting clothes the right way round	Folding clothes
Fill a glass with water from a jug	Use a spoon to pick up raisins and pour into a bowl e.g. as if weighing ingredients	Use a fork to hold soft food and a knife to cut it
Turn on and off a tap	Do the washing up	Dry items



Tidy up

Results - p-DLT

Max score for each task = 7



Performance significantly correlated with motor ability

Only easy when you know how

For example...



Jo Camp

1. Fill the kettle
the right amount

2. Boil the water

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



3. Find a
(clean) cup

4. Locate the
teabags

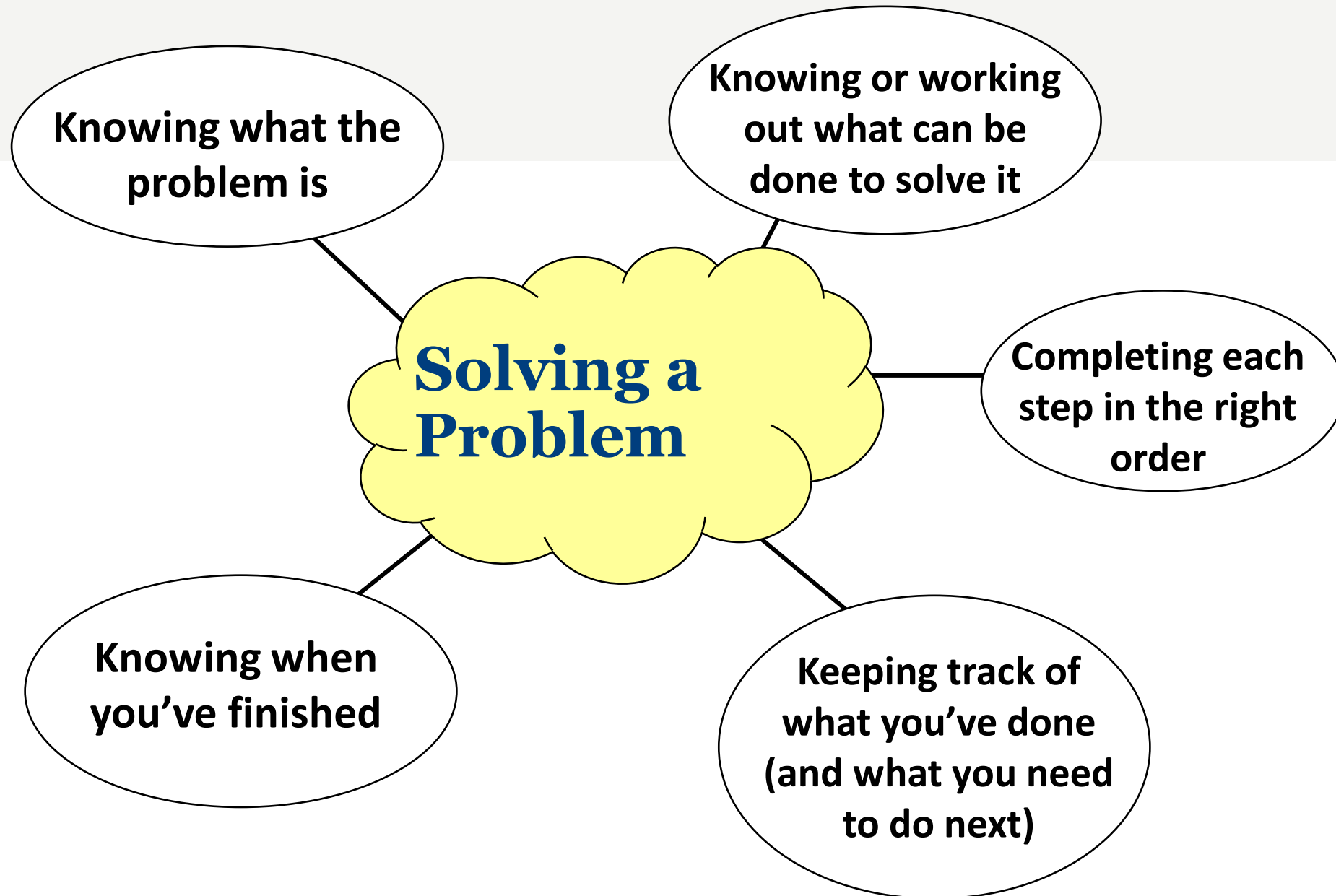
8. Carry to the
right place

5. Put the teabag
in the cup

7. Add milk
and/or sugar, if
required

6. Pour the right
amount of water
safely





Summary: Daily living and Problem solving

- » Limitations in planning skills and keeping items in memory can hinder problem solving progress in WS.
- » Performance on the practical daily living task and motor ability were related. This suggests that individuals with better motor ability will also be more skilled on many daily living tasks.
- » 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).

Daily living and Problem solving suggestions

- » If individuals are likely to be doing something that they are anxious about due to motor skills then practice them with them before to improve confidence.
- » Inform keyworkers / teachers that verbal skills in WS are better than problem-solving skills; someone with WS might struggle with problem solving more than might be expected from listening to them speak.
- » Talking through the steps in routine, well-practiced problems might be beneficial, but more and different types of support might be required for less predictable problems.
- » Difficulty with planning ahead was related to solving puzzles in the lab. So, support with planning one or two steps ahead (which could also help reduce impulsive responding) might be a useful technique. It may be helpful for individuals to use visual help sheets to plan tasks with lot of steps.
- » Being able to keep emotions under control was related to being more likely to be able to solve everyday problems. Strategies which can help someone with WS to stay calm about a situation could be helpful during problem solving.

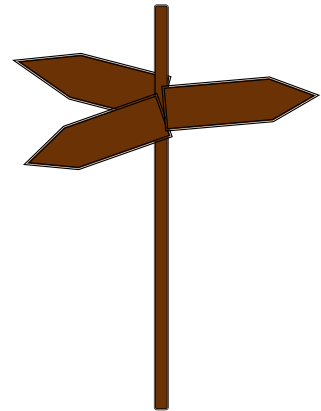
Independence and navigation

Independence and navigation

» 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.



Stages of route learning

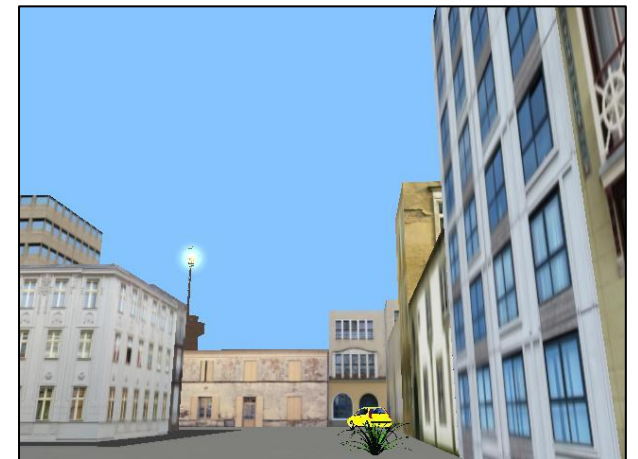
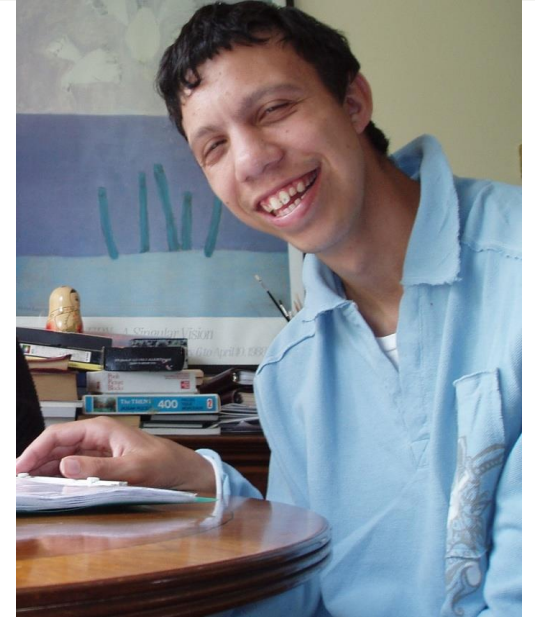
Siegel and White (1975)

1. Knowledge of the landmarks along a route: **landmark knowledge**
2. Knowledge of the sequential order of the turns and landmarks of a route: **route knowledge**
3. The development of a cognitive map (knowledge of the configural structure of an environment): **configural knowledge**

Navigation and Williams syndrome

- » Hippocampus and the dorsal visual stream are atypical in WS (Meyer-Lindenberg et al., 2004, 2005)
- » Some atypical use of landmarks (Farran et al., 2012; Nardini et al., 2008)
- » Can develop route knowledge, with repeated experience (Farran et al., 2010).
- » Configural knowledge is a challenge (Broadbent et al., 2014; Farran et al., 2015)

How does this translate to everyday difficulties? People with WS use fixed routes, but are unable to find alternative routes/ shortcuts and find it difficult to re-orient when lost.



Project ELSTRAD

Funded by ESRC and ANR

A series of studies over five years designed to compare navigation performance in TD children, WS and DS, using virtual reality.

1. Landmark types: landmarks near junctions, landmarks not near junctions (path landmarks), distant landmarks, non-unique landmarks
2. Sequential relationship between landmarks and turns
3. Configural knowledge and Cognitive maps: determining a short-cut
4. **Navigation and Anxiety**



Kerry Hudson



Harry Purser



Questionnaire measures: anxiety and wayfinding

Farran et al. (2022)

- » *Santa Barbara Sense of Direction Scale (SBSOD; Hegarty et al., 2002)*. Assesses interaction with the environment such as giving directions, navigation and understanding of space.
- » *Spence Children's Anxiety Scale (SCAS; Spence, 1997)*. Caregivers rate the frequency that their son/daughter experiences feelings such as 'I can't seem to get bad or silly thoughts out of my head'.
- » *Spatial and travel anxiety questions*. Questions related to anxiety during wayfinding. Such as:
 - Difficulties interacting with roads (crossing, traffics light, traffic)
 - Dealing with getting lost
 - Independent travel
 - Specific travel anxiety.

Anxiety and wayfinding

N=55 caregivers of individuals with WS.

N=42 caregivers of individuals with DS

SCAS: 53% elevated anxiety (WS)

35.7% elevated anxiety (DS)

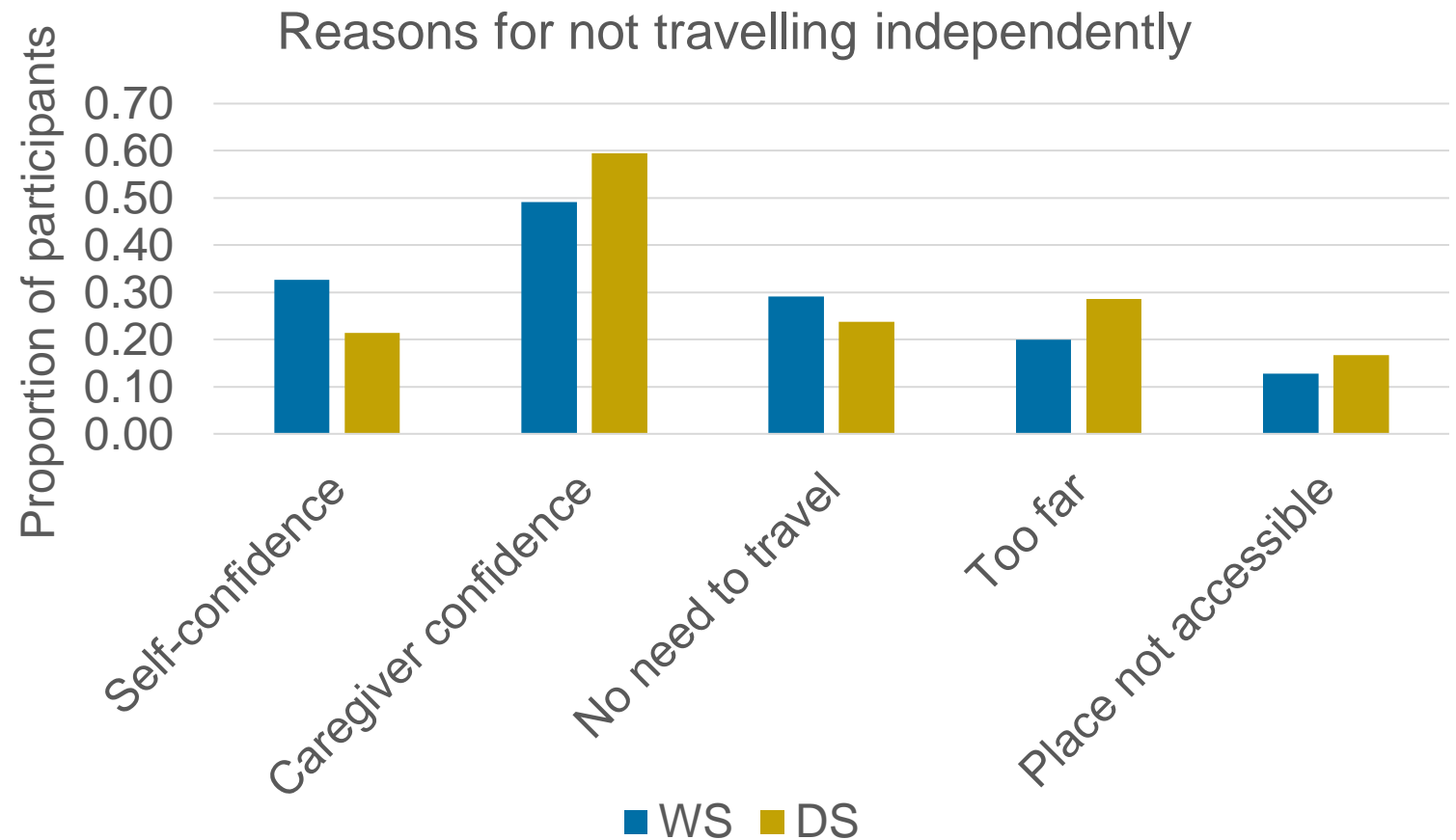
SOD: 100% 'poor' (WS and DS). WS stronger than DS

Wayfinding: WS stronger than DS

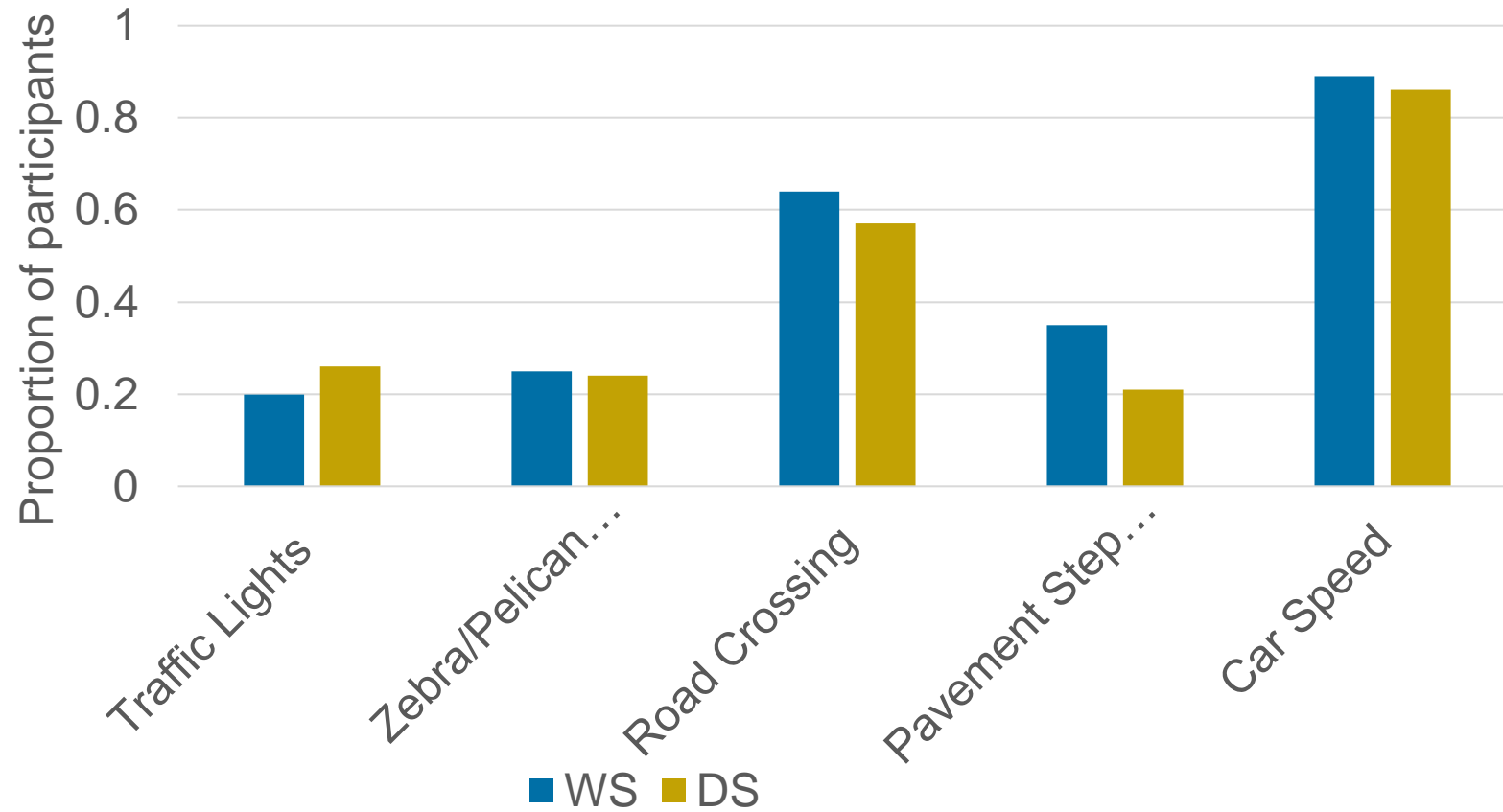
Wayfinding anxiety: WS more anxious than DS.

Anxiety and wayfinding

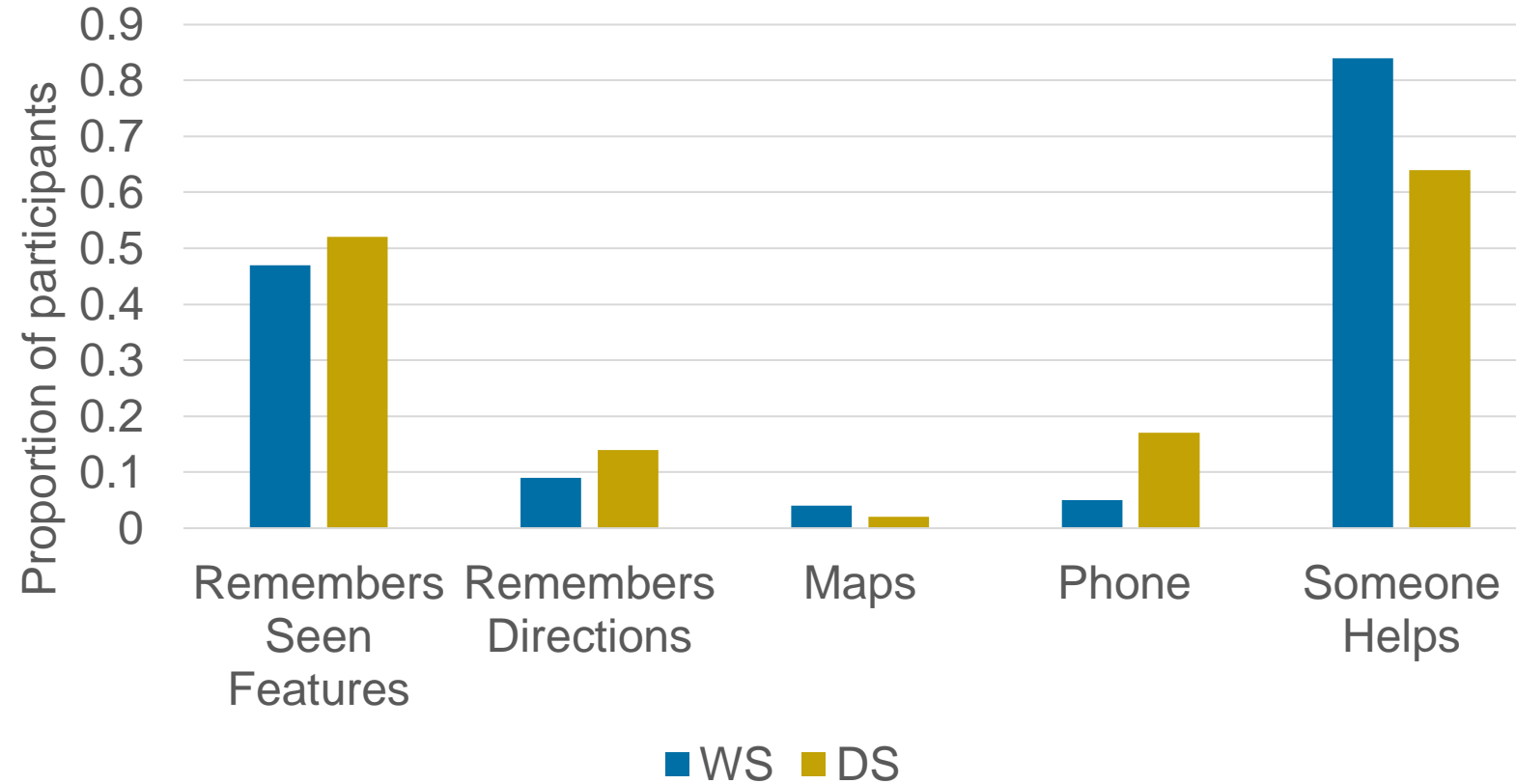
Travel independently:
40% (WS)
16.7% (DS)



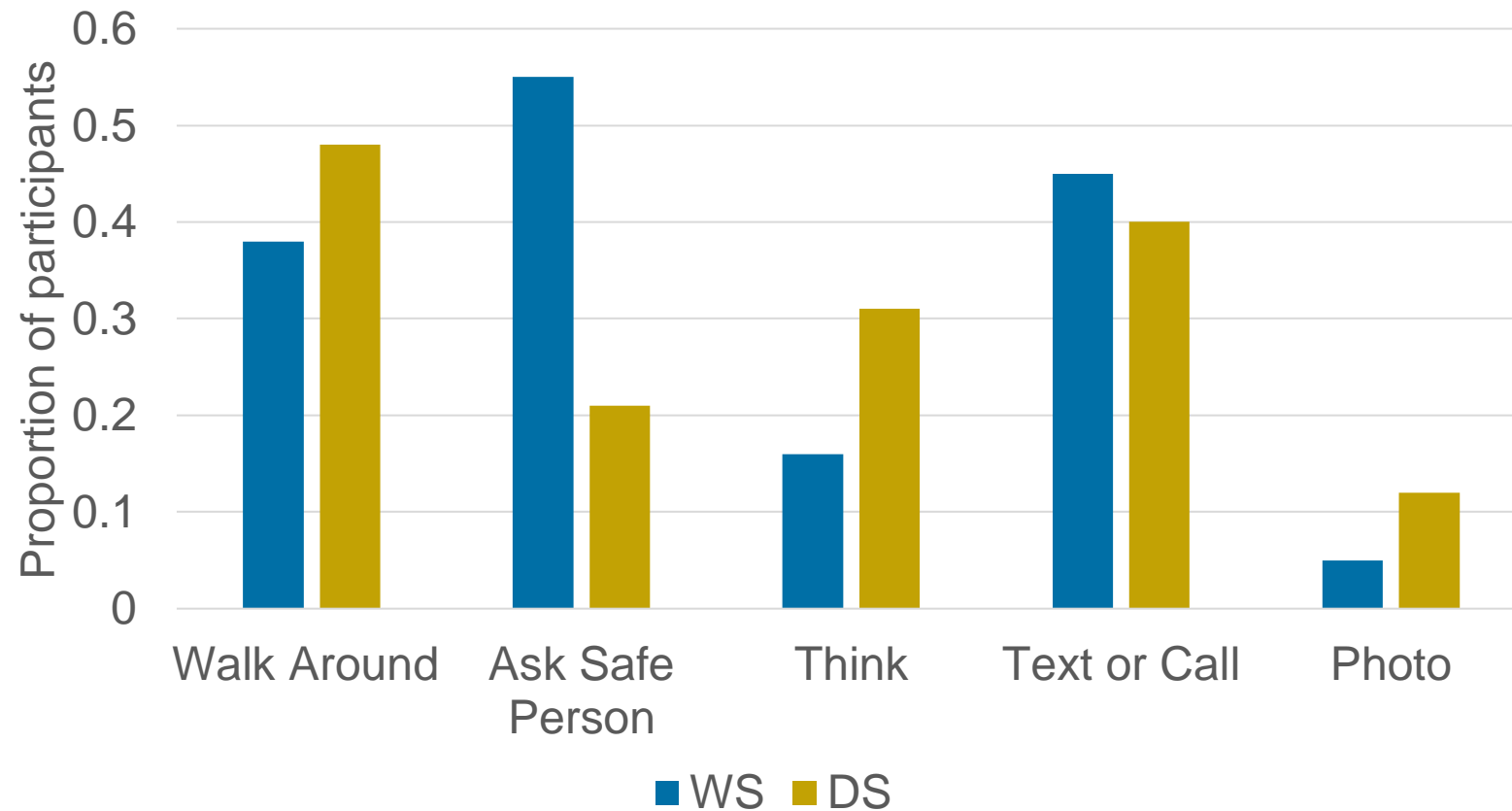
Wayfinding: What do people find difficult?



Wayfinding strategies



Strategies when lost



Anxiety and wayfinding

- » Anxiety (SCAS) associated with Sense Of Direction and wayfinding scores in WS and DS.
 - WS: significant associations for Social Phobia and Panic
 - DS: significant associations for OCD, Panic, Separation Anxiety, Fear of Physical Injury

Anxiety and wayfinding: We asked what training would be beneficial.

General

bus training

road safety/ crossing roads

learning how to buy tickets

help with money and time awareness

sign recognition

safety tips



Independent navigation is complex. Not just spatial. Not just anxiety related.

Anxiety and wayfinding: We asked what training would be beneficial.

Anxiety related

“Help to be aware of what to do if lost and NOT to panic. She has shown erratic behaviour e.g. running into roads after missed buses. Gets really upset if things go wrong.”

“We have tried to teach my 21 year old to cross the road for about 10 years. She knows how to do it but when left to do it alone gets panicky. Because she is worried she will get run over and killed.”

“I do not think ANY amount of training will conquer the high levels of anxiety and lack of confidence she now experiences in the mornings/ going somewhere. “



Anxiety often overrides spatial skills.

Navigation conclusions

- » Individuals with WS can learn a fixed route.
- » Difficulty with detours from a fixed route.
- » Strategies: prefer to rely on help from others than own spatial abilities (“Knows how to do it.... but gets panicky”)
- » ANXIETY is a big stumbling block for independent navigation in WS

Navigation suggestions



- » Individuals with WS need significantly more practice than typically developing children when learning a route. Repetition is key.
- » When practicing the route, ask your son/ daughter to actively lead you from point A to point B, correcting their errors as you walk along. This active engagement helps with focus and concentration.
- » During route learning, encourage your son/ daughter to point out and verbalise useful landmarks. For example, “There is a postbox at this corner” Verbally labelling useful landmarks draws on their strengths.
- » Break the route up into sections, and ask your son / daughter to identify the landmarks within each section. This makes it easier for them to associate each landmark with an action: e.g. “At the bakery I turn left, then at the chemist’s I turn right”
- » Teach your son / daughter about which landmarks are most useful, and why. Why? As individuals with WS show a strong reliance on landmarks, it is important that they know how to select useful ones.

Navigation suggestions



- » Make sure your son/ daughter has strategies to use if they should become lost, e.g. phone or text for help, store pictures of landmarks along the route on their phone, ask a safe person to help. This might help with associated anxiety.
- » Be aware that the return route should be learned as well. It will look different, so it is difficult to visually match the environment to the one learnt for the outgoing route.
- » Make other caregivers aware of the impact of anxiety on navigation in WS. Provide a safe and supportive environment in which to practice.
- » Use pedestrian crossings wherever possible. This is because issues with depth perception are common in WS, and so judging the speed of oncoming traffic is difficult.
- » Using a virtual environment could be a safe way to practice exploration and navigation skills. This can be as simple as walking the route using a streetview app on a desktop computer. Potential hazards such as traffic, uneven ground, etc are not present in a virtual environment. Learning in a virtual environment can improve real-world performance.

