

SELECTIVE IMPAIRMENT OF DORSAL STREAM FUNCTIONS IN CHILDREN WITH WILLIAMS SYNDROME ?



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ABSTRACT

Evidence suggests that visual processing is divided into two pathways: the 'ventral stream' that is specialized for object recognition ("What") and the 'dorsal stream' that is specialized for visual guidance of action ("How"). We investigated the possible dissociation of the two cortical visual systems in children with Williams syndrome (WS), a rare genetic disorder that results in a unique cognitive profile including impaired spatial cognition, together with relatively spared language. Following Milner & Goodale (1995), we compared performance in an *action* task (Posting) to that in a *perception* task (Matching). Results suggest that WS children have deficits in both the action and perception systems as well as in visual short-term memory.

PROPERTIES OF PERCEPTION AND ACTION SYSTEMS

- "Ventral" stream responsible for representing the enduring aspects of objects, such as their shape^{6,7}
 - Important for *perceiving* objects
 - Uses *non-egocentric* coordinate systems
- "Dorsal" stream responsible for on-line visual action^{6,7}
 - Important for *reaching and grasping*
 - Uses *egocentric* coordinate systems

TWO HYPOTHESES

- Damaged "Action System"**
 - There are separate systems for action and perception tasks
 - *Action* system is impaired
 - *Perception* system is relatively preserved
- Damaged Visual Short-Term Memory (VSTM)**
 - Action tasks, such as drawing, make heavier demands on VSTM than do perceptual matching tasks

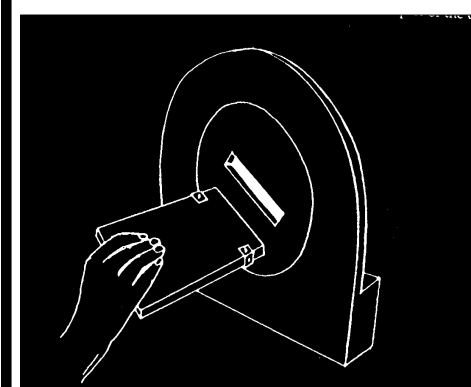
WILLIAMS SYNDROME (WS)



PHENOTYPIC CHARACTERISTICS^{1,2}

- Facial morphology
- Small stature
- Often accompanied by defects of organ especially the heart
- Brain volume reduced overall
- Mild to moderate mental retardation
- Unusual cognitive profile
 - Relatively spared language
 - Impaired spatial ability

EVIDENCE FOR THE DORSAL/VENTRAL DISTINCTION



- Patient DF could "post" a card in a slot but could not judge its orientation, suggesting a preserved dorsal system together with a damaged ventral system⁶.
- With a 5-second delay, DF failed at the posting task, presumably because of an absence of visual memory in the dorsal stream⁶.
- "Normal" subjects can still post with a 5-second delay suggesting they can "transfer" information from a VSTM in their intact ventral stream to the dorsal action system⁷.
- In normal subjects, context-based illusions (Roelofs effect) affect "perception" but not action.
 - With a 2-second delay, these illusions are also seen in the action task, suggesting again that with delay, the action system must rely on a VSTM located in the ventral stream⁸.

METHOD

PARTICIPANTS Twelve children with WS between the ages of 8;3 and 16;2 (Mean = 12) and 12 mental-age matched controls between the ages of 4;9 and 9;7 (Mean = 6;6). The WS children were recruited through the Williams Syndrome Association, and had all been diagnosed by a geneticist, using phenotypic characteristics, and in most cases, FISH tests. All children were tested using a standardized intelligence test, the Kaufman Brief Intelligence Test¹¹. The WS children were well matched to the normally developing children on both the Verbal component (Ms = 34, 35; Ranges = 23-46, 26-48, respectively) and on the Non-verbal component (Ms = 19, 20; Ranges = 13-24, 13-29, respectively).



Posting (Action) Task

Matching (Perception) Task



EXPERIMENT 1 - No Delay:

DESIGN & PROCEDURE Following Milner & Goodale's method, children faced a box with a slot (3.75" x .75") that could be oriented to 0 degrees, 90 degrees, 45 degrees left or right of 0 degrees. Participants were seated approximately 2' in front of the apparatus. In order to change the slot's orientation between trials, a black cloth was dropped in front of the slot so the child did not see the change until the new orientation was set.

In the "Posting" or *action* condition, they were instructed to pick up a 5.75" x 3.25" plexiglass "dollar bill" from the table in front of them and "put it quickly into the slot of the piggy bank". They were tested at each of the 4 orientations (0, 90, 45 degrees left or right) for 6 trials each, for a total of 24 trials. Trial order was randomized over subjects.

In the "Matching" or *perception* condition, children viewed a mannequin "hand" attached to a pulley and lever-set-up, which allowed the experimenter to move it through a 180-degree rotation. The child was told that the hand would be moved by a third person (who was blindfolded during all trials), and that he or she should tell the person when to "stop" such that "Mr./Mrs. Hand" holding the dollar bill would be "just ready to put the dollar bill into the slot in the piggy bank". The same number of trials testing the same target orientations occurred in both the action and perception conditions.

EXPERIMENT 2 - Delay:

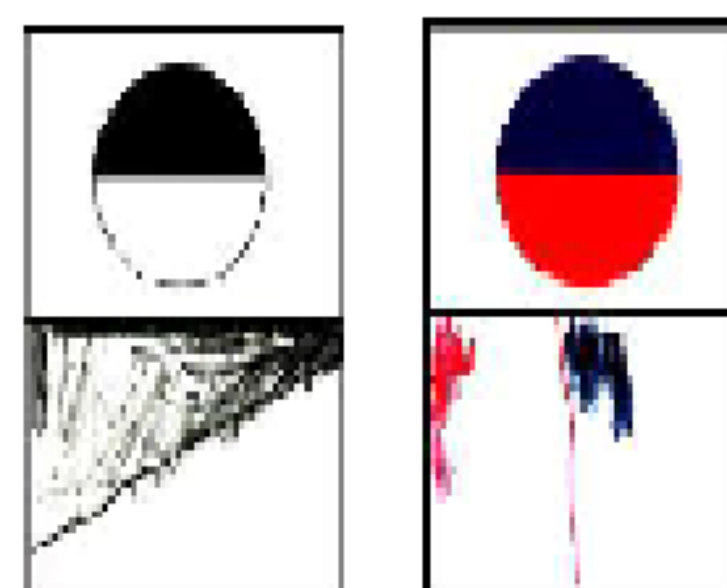
DESIGN & PROCEDURE Experiment 2 was identical to Experiment 1, but this time, a delay was introduced between the presentation of the slot and the subject's response. Children viewed the slot as in the above experiment, but then the black cloth was rolled down over its front, to conceal it from view. After a five second delay, the child carried out the *action* or *perception* task.

In the "Posting" or *action* task, they were asked to put the dollar bill through the slot (though it was covered, they could approach the panel and push the edge "through" the slot). In the "Matching" or *perception* task, they were asked to judge when the hand was holding the dollar bill in just the right position so as to post it through the slot (which was still covered by the opaque black cloth).

SPECIALIZATION OF SPATIAL COGNITION IN WS

Deficits are observed in *visual construction tasks*^{1,2}

Models



WS Drawings

Identification of objects is relatively preserved³



PRELIMINARY EVIDENCE THAT WS CHILDREN HAVE AN IMPAIRED DORSAL STREAM AND A PRESERVED VENTRAL STREAM...

- Atkinson et al. reported that WS subjects were impaired in the *posting* task but not in *perceptual matching*⁵.
- WS subjects were also impaired in detecting motion coherence (a "dorsal" task) but not form coherence (a "ventral" task)⁵.

...BUT WS CHILDREN ALSO EXHIBIT VISUAL SHORT-TERM MEMORY DEFICITS

- WS subjects perform significantly better on verbal short-term memory tasks and significantly worse on visual-spatial short-term memory compared to controls^{9,10}.
- WS subjects have poor visual short term memory for orientation (Hoffman & Landau, in preparation).

RESEARCH QUESTION

Is there a dissociation of visual *perception* and visually guided *action* in Williams syndrome?

- Visual *perception* appears relatively intact
 - Possible exception: WS subjects may be impaired in perception of line orientation^{1,4}
- Visually guided *action* is impaired⁵

PREDICTIONS

Experiment 1: No-Delay performance (visual information continuously present)

Posting: WS < Controls Reason: Deficient dorsal stream in WS
 Matching: WS = Controls Reason: Intact ventral stream in WS

Experiment 2: Delay Performance (assuming information is stored in VSTM and passed to the dorsal stream as per Bridgeman)

Posting: WS << Controls Reason: Deficient dorsal stream + damaged VSTM in WS
 Matching: WS < Controls Reason: Damaged VSTM in WS

Figure 1A
ACTION VS. PERCEPTION BY GROUP
NO DELAY

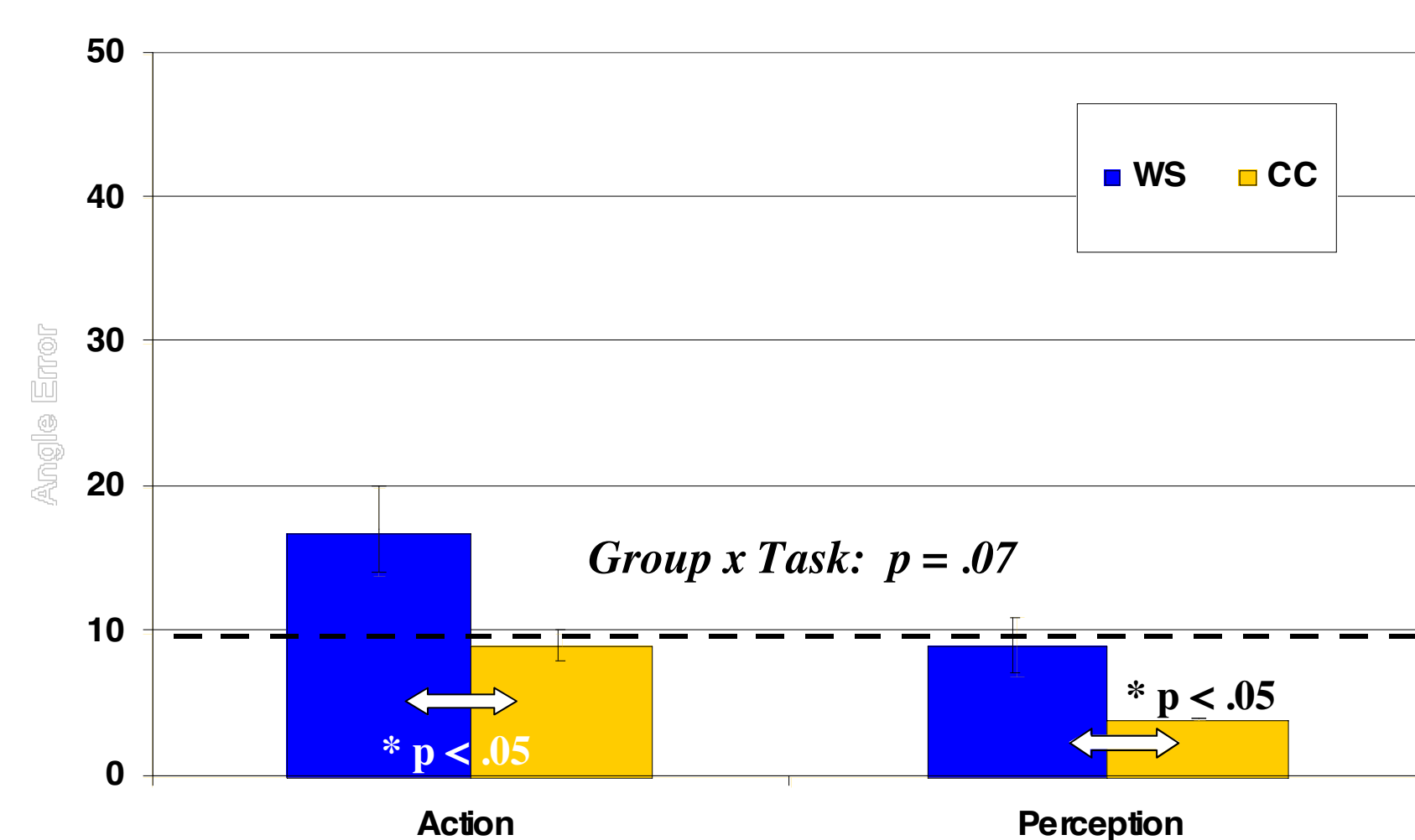
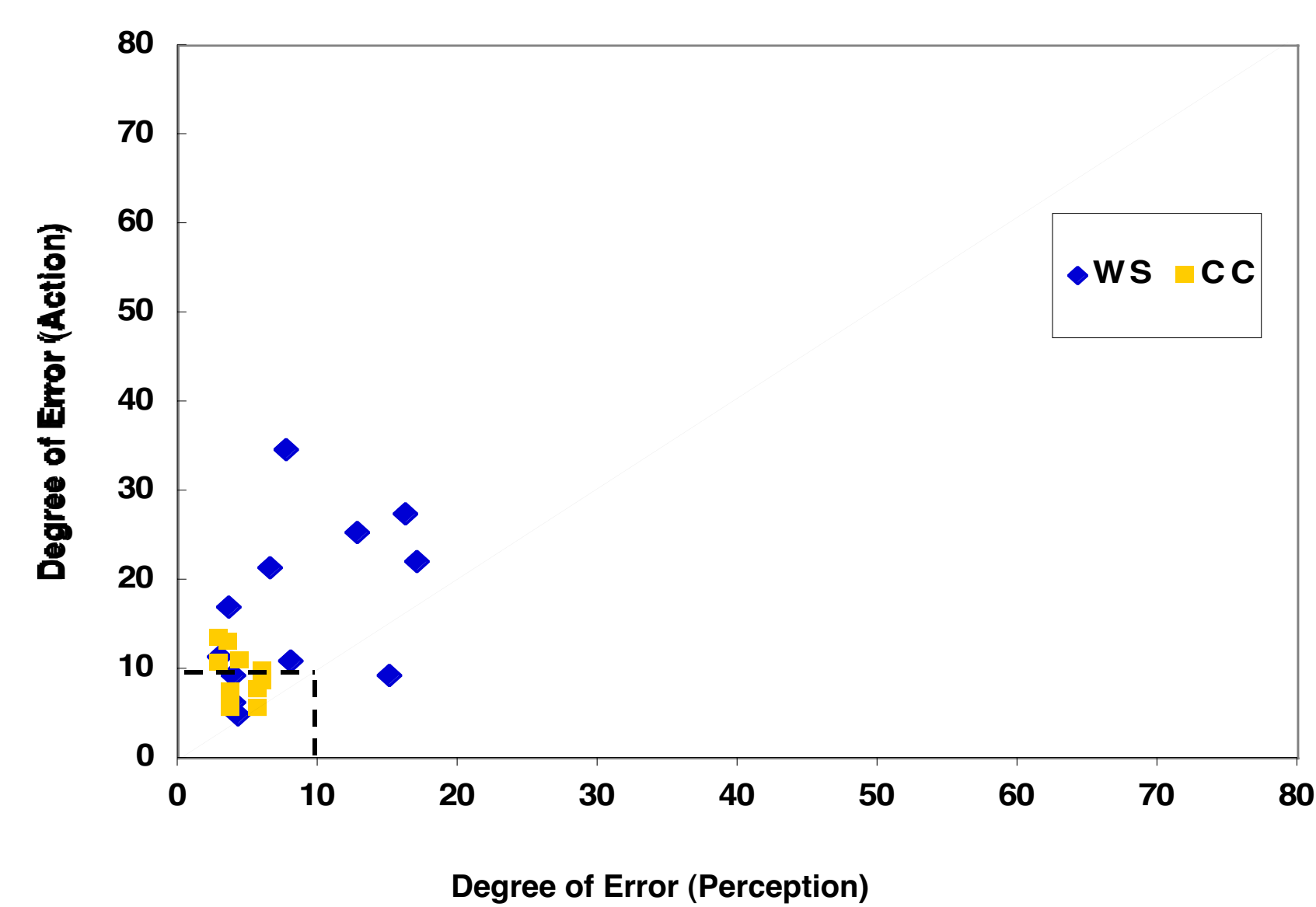


Figure 1B
ACTION VS. PERCEPTION BY GROUP
NO DELAY
(Individual Subjects)



FINDINGS

Experiment 1: No-Delay performance (visual information continuously present)

Posting: WS < Controls Conclusion: Deficient dorsal stream in WS
 Matching: WS < Controls Conclusion: Deficient ventral stream in WS

Figure 2A
ACTION VS. PERCEPTION BY GROUP
DELAY

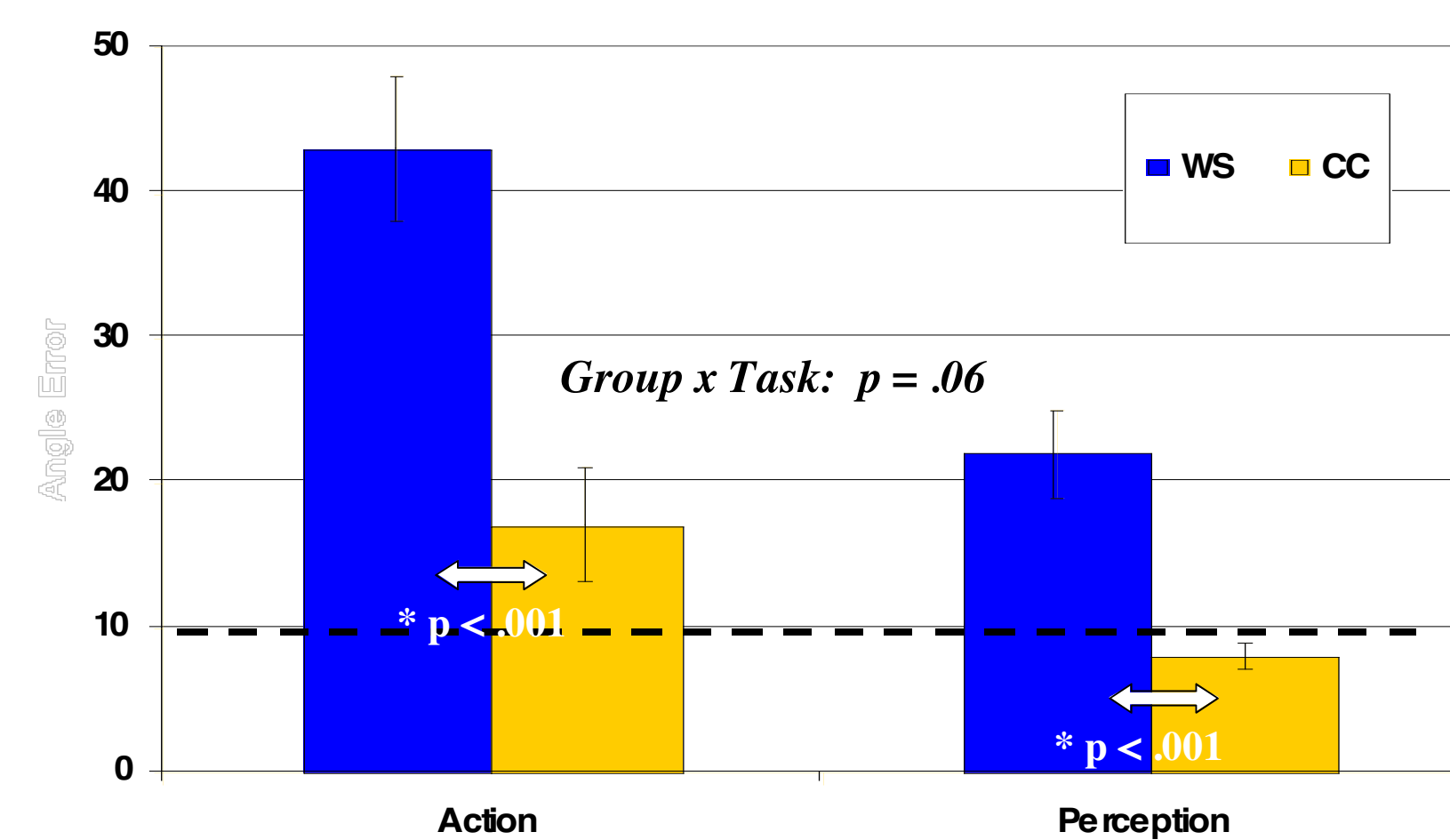
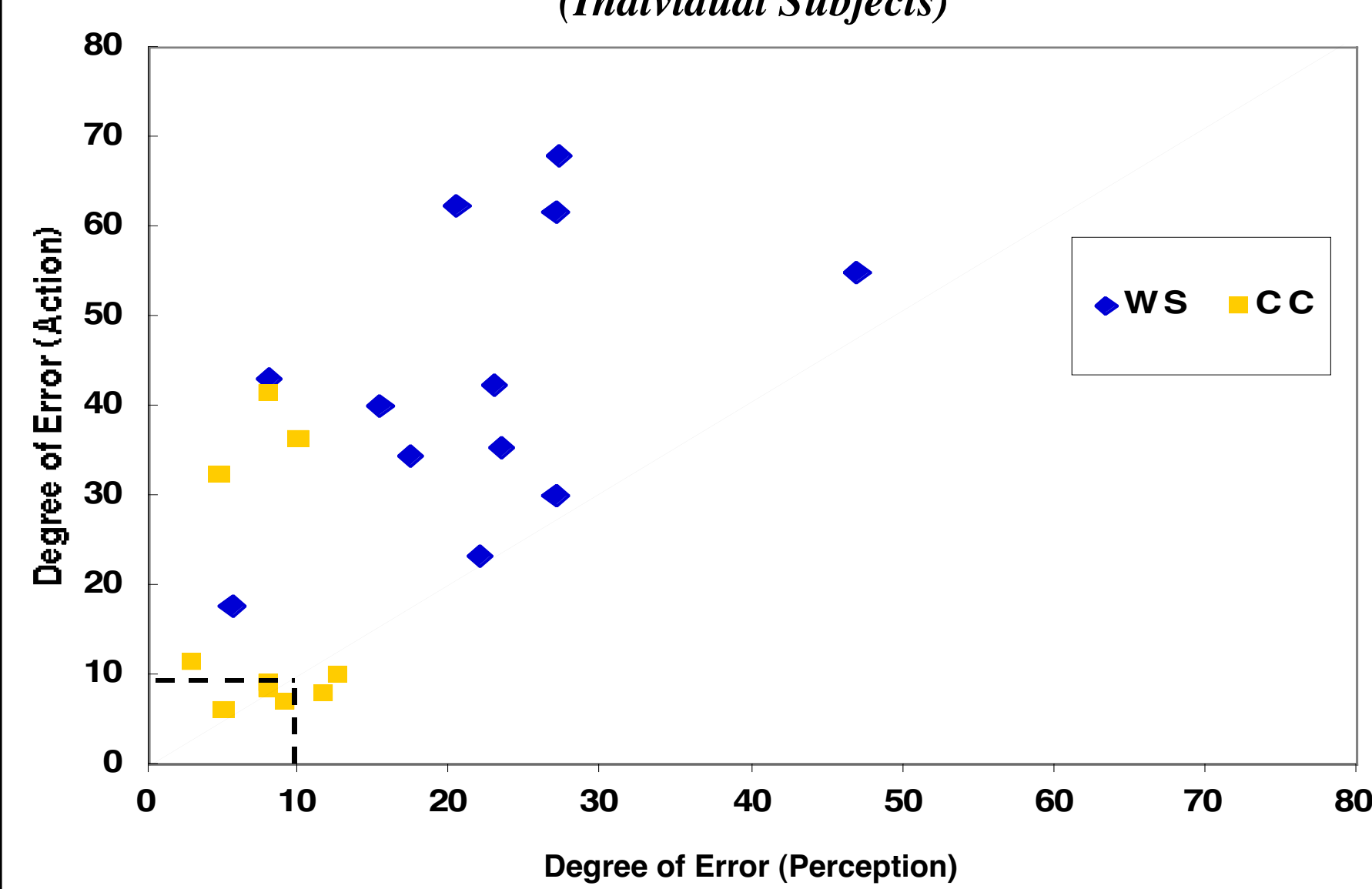


Figure 2B
ACTION VS. PERCEPTION BY GROUP
DELAY
(Individual Subjects)



FINDINGS

Experiment 2: Delay Performance (assuming information is stored in VSTM and passed to the dorsal stream as per Bridgeman)

Posting: WS << Controls Conclusion: Deficient dorsal stream + damaged VSTM in WS
 Matching: WS < Controls Conclusion: Damaged VSTM in WS

SUMMARY

- WS children were less accurate than controls in posting a card in a slot (*action task*) as well as in judging its orientation (*perception task*).
- This difference between groups was pronounced when children had to rely on their memory for the slot's orientation.
- These results suggest that WS children have deficits in both the **action** and **perception** systems as well as in **visual short-term memory**.

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