Pergamon

PII: S0028-3932(97)00019-5

Neuropsychologia, Vol. 35, No. 8, pp. 1159–1173, 1997
Published by Elsevier Science Ltd.
Printed in Great Britain
0028-3932/97 $17.00+0.00

Isolation of a right hemisphere cognitive system in a patient with anarchic (alien) hand sign

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(*Received* 7 *August* 1996; *accepted* 8 *January* 1997)

Abstract—We report evidence of isolated conceptual knowledge in the right hemisphere of a woman with chronic anarchic hand sign after ischemic infarction of the central four-fifths of the corpus callosum. Limited visual information was available to the right hemisphere, access to medial temporal structures subserving memory was disrupted and disconnection from left hemisphere language structures was complete. Still, the right hemisphere could build mental representations of objects via tactile input and use them in cross-modal matching. These representations were not accessed consistently in auditory comprehension or naming tasks. This functional specificity and its pathoanatomical correlates demonstrate that the study of anarchic hand sign can illuminate not just motor control issues but may inform our understanding of the representation and lateralization of conceptual knowledge as well. Published by Elsevier Science Ltd

Key Words: tactile anomia; agraphia; corpus callosum; disconnection syndrome.

Introduction

Over the last three decades, the study of commissurotomy patients has yielded increasingly specific information regarding the role of the corpus callosum in the integration of lateralized cognitive functions. Whereas the classic studies of Gazzaniga, Sperry and colleagues relied on surgical reports to ascertain the extent of callosal transection (e.g., [18, 45]), magnetic resonance imaging (MRI) has since provided a means for the localization of partial callosal damage caused by naturally occurring lesions as well as therapeutic surgical ones. This capacity for *in vivo* localization, combined with the careful assessment of cognitive processes using tasks akin to those developed in experimental psychology, has, in turn, allowed a better understanding of structure–function relationships underlying interhemispheric transfer (for review, see [16]).

Studies of patients with partial (posterior) callosotomy or posterior cerebral artery stroke have established that the splenium mediates visual transfer [1, 16, 19, 23, 27, 36]. Direction-specific transfer of somesthetic information has been observed in one patient (EB) with a posterior section of the corpus callosum [16]. With respect to auditory transfer, MRI and dichotic tests of speech sound recognition in partial callosotomy patients and in a stroke patient indicate that the posterior body mediates the interhemispheric integration of phonemic and orthographic information [42, 47]. Preservation of transfer of semantic information in the absence of somesthetic transfer has also been documented after posterior section in one patient [43]. Another patient with inadvertent sparing of a limited amount of callosum has demonstrated very specific additive transfer of phonemic and orthographic information [19]. The present study of a patient with a naturally occurring lesion of the body of the corpus callosum that spares a portion of the splenium and the entire genu and rostrum presents an unusual opportunity to learn more about specific patterns of transfer and to confirm observations made with patients with a long history of neurological disease on a patient with normal neurological development.

This case of chronic anarchic hand syndrome was investigated 2.5 years post-onset. The patient suffered multifocal 'watershed' infarcts of the callosum and right hemisphere involving medial gray and white matter struc-

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tures in the frontal, posterior parietal, posterior temporal and occipital lobes. The extent of the callosal and hemispheric lesions are detailed via MRI, and specific perceptual and cognitive functions are examined using a variety of tasks to take advantage of this opportunity to learn more about interhemispheric transfer. Data reported here were gathered in a 6-month period in which AW was neurologically stable. Neuropsychological assessment and pilot naming experiments were initiated in October 1991. Experiments 2 and 3 were carried out in November 1991. Experiment 1, an elaboration of the pilot studies, was completed in March 1992. Experiment 1 examines the patient's ability to name objects based on a right hemisphere representation generated from tactile input. Experiment 2 examines her ability to use that information for cross-modal matching. Finally, Experiment 3 compares the patient's naming, auditory comprehension, tactile matching and memory ability using stimuli that are free of cues identified in prior experiments.

Case report

Neurological history

AW, a right-handed female born 22 October 1938, was admitted to the hospital on 7 August 1989 following a 2-week history of fluctuating neurological symptoms, including right occipital headache, unformed visual hallucinations (flashing white and colored lights), paresthesias and weakness of the left face and upper extremity, and dizziness. Her chief complaint on the day of admission was, "My left hand is not feeling like my own". The past history was notable for coronary artery disease requiring three-vessel bypass surgery in 1984, hypertensive vascular disease, hypercholesterolemia and heavy cigarette smoking. AW was employed as a laboratory technician prior to her stroke. She had completed 10 years of formal education.

On admission, neurological examination revealed a clear sensorium with normal speech and language functions, a left homonymous hemianopia without hemi-inattention, a mild left hemiparesis affecting face, arm and leg, decreased pin prick sensation in the left hemibody, decreased proprioception in the left extremities, and inability to name objects placed in the left hand. Computerized X-ray tomography revealed a hypodensity in the right medial occipital and parietal lobes. Electroencephalography showed diffuse right hemisphere slowing and no paroxysmal activity. Cerebrospinal fluid analysis was normal. Carotid ultrasound revealed occlusion of the right internal carotid artery.

On her most recent follow-up at the clinic (8 October 1991), the patient's chief complaint was that her left hand did not obey her. For example, on several occasions while driving, the left hand reached up and grabbed the steering wheel from the right hand. The problem was persistent and severe enough that she had to give up driving. She reported instances in which the left hand closed doors the right hand had opened, unfolded sheets the right had folded, snatched money the right had offered to a store cashier, and disrupted her reading by turning pages and closing books. She stated that she could get her left hand to do things only after she watched the right hand do them. Neurological examination showed no left hemiparesis, severe left hand apraxia, which was worse with the eyes closed but still evident with the eyes open, a left homonymous hemianopia and left hemisensory loss. Her medications at the time of the present experiments included atenolol, diltiazem, triamterene, lovastatin, aspirin, dipyridamole and alprazolam.

Neuropsychological test battery

Standardized testing demonstrated average intelligence and memory skills. On the Wechsler Intelligence Scale (Revised), there was a 16-point advantage of verbal IQ over performance IQ, with mild motor slowing and difficulty with visual integration (FSIQ 93; VIQ 100; PIQ 84). Memory skills were in the average range on the Wechsler Memory Scale (Revised), but the Verbal Memory Index score was significantly above AW's Visual Memory Index score (General Memory 97; Verbal Memory 109; Visual Memory 87). Likewise, although her combined Attention and Concentration score was in the average range (104), Verbal Span was much easier for her than Visual Span (Forward 94th percentile, backward 90th percentile and forward 34th percentile, backward 36th percentile, respectively). The California Verbal Learning Test scores were generally at or above the mean for her age group. She did make many perseverative errors, however. In contrast, her performance on the Benton Visual Retention Test was in a range suggestive of impairment. She made many errors of rotation and distortion, but they were equally distributed over the right and left sides of the page. AW's score on the Benton Facial Recognition Test would place her in the borderline range. In summary, AW's verbal intelligence and memory are in the average range, but visuospatial skills, particularly visual memory function, were mildly impaired.

Signs of interhemispheric disconnection

AW continued to complain of an inability to control her left hand, the symptom that Brion and Jedynak [7] suggested was a clue to look for signs of callosal disconnection. Like most alien or anarchic hand patients, as pointed out by Della Sala *et al.* [11], AW recognized her left arm as her own, but complained that she could not control its actions. Her early report that her left hand had to observe the actions of her right to carry out commands is similar to behaviors reported in callosally sectioned patients [34]. This inability to control her left hand was intermittent and incidents occurred more frequently when she was fatigued. She found this symptom disturbing and irritating. In several sessions of standardized psychometric testing, the only evidence of left hand interference in activities was observed in the Picture Arrangement subtest of the WAIS-R, in which her left hand attempted to arrange the cards differently from the right hand.

Use of tasks known to elicit signs of interhemispheric disconnection in split-brain patients readily demonstrated her difficulties. Of the clinical features enumerated by Bogen [6] as potential callosal signs, AW demonstrated five. Left ear suppression on dichotic listening was demonstrated. She was mildly apraxic with her left hand. Her left hand was also agraphic (see Fig. 1). She was unable to consistently name common objects placed in her left hand despite having been tested extensively on those objects earlier. Each hand showed mild constructional apraxia on Kohs Blocks. AW's drawings also demonstrated some left-hand constructional apraxia, as well as perseveration (see Fig. 2). AW was unable to transfer somatosensory information between her right and left hands.

Brain imaging and lesion localization

On 6 November 1991, magnetic resonance images were acquired in the sagittal and coronal planes using a 1.5-Tesla