RESEARCH REPORT

A DISCONNECTION SYNDROME DUE TO AGENESIS OF THE CORPUS CALLOSUM: DISTURBANCE OF UNILATERAL SYNCHRONIZATION

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Abstract

Recently, interhemispheric disconnection syndromes have been noted in patients with agenesis of the corpus callosum (ACC) during the performance of certain tasks. However, few studies have demonstrated an asymmetric disconnection syndrome. In this report, we present just such a syndrome in a patient with ACC, who manifested ambidexterity (but with a left-hand tendency) and had high intelligence, no neurological deficits, and no associated malformations. In a comparison with similar subjects (amateur musician), we studied her asymmetric deficits using four tasks: (1) simple reaction time for visual stimuli, (2) paced finger tapping in synchrony with visual or auditory stimuli, (3) paced finger tapping without an external reference, and (4) rhythmical finger tapping in synchrony with visual or auditory stimuli. While the comparable subjects displayed no significant difference between hands, and the patient showed no significant difference between hands in the auditory paradigm, her tapping performance deteriorated significantly when asked to synchronize the left hand with timed visual stimuli, irrespective of whether finger tapping was paced or rhythmical. We believe that this phenomenon constitutes a novel asymmetrical disconnection syndrome in an ACC subject; these results suggest that synchronization of multimodal temporal information was lateralized in the left hemisphere (in this case), which is something that the ACC patient could not compensate for.

Key words: agenesis of the corpus callosum, disconnection syndrome, finger tapping, synchronization, rhythm

Introduction

Investigations of patients with callosal damage have revealed a series of characteristic symptoms (Sperry et al., 1969; Bogen, 1985). Unlike patients with acquired callosal damage, congenitally affected patients, specifically those with agenesis of the corpus callosum (ACC), were thought to have few symptoms (Ettlinger et al., 1972). More recently, however, several interhemispheric disconnection syndromes have been identified in ACC patients (Lassonde, 1994). Interhemispheric disconnection syndromes in ACC patients appear to be bidirectional, i.e., behavioural impairment is manifested not only in the dominant hand (or visual field), but also in the non-dominant hand (or visual field). Apart from visuospatial orientation ability (Lassonde, 1994; Martin, 1985), there is little evidence of asymmetric disconnection syndrome in such patients, which suggests that compensation for visuospatial deficits might not be underdeveloped (or absent) in patients with ACC.

Most patients with ACC have associated malformations in addition to the corpus callosum. Therefore, it is not clear whether the cause of their behavioural dysfunction is the absence of commissural fibres or such malformations (Ettlinger, 1977). More recently, however, the development of imaging techniques, such as computer tomography (CT) and magnetic resonance imaging (MRI), has led to the observation that some patients with ACC have well-preserved intellectual functions, with no associated malformations (Sauerwein et al., 1994). Such patients are potentially valuable sources of information about the role of the corpus callosum in cognitive function.

In this report, we present a novel asymmetric disconnection syndrome in a patient with ACC whose intellectual functions were preserved, without associated malformations.

Case Report

Subject Y.M. was a 38 year-old ambidextrous female who had worked as a nursery school teacher for 18 years. She had normal grades in elementary, junior-high, and high school, and graduated from college with a degree in child education. After graduation, she passed a civil service examination and now works at a public nursery school as a local government employee. She began playing the piano when she was in elementary school. She uses both hands in her everyday musical activities, which include teaching children. Y.M. is highly intelligent and exhibits no

neurological deficits in her everyday activities; however, she does have various problems at work. Her co-workers have noted that she is occasionally inattentive to the schoolchildren, although she might be paying attention to one child in particular. For example, she was capable of supervising a single child who was bathing, but was apparently unaware of other children, even when they were in a potentially dangerous situation.

Consequently, Y.M. is unable to supervise a class independently. Despite this drawback, Y.M. has been a favourite with the children in the school, as well as with her colleagues. In addition, she is an exceptionally gifted writer, and has received awards for her writing. Since her talent appeared to be at odds with her ability in the classroom, in 1995 a co-worker arranged for her to be taken to Showa University Hospital for a CT scan, where ACC was diagnosed.

Dominance in Y.M. is ambiguous. At the time of this study, Y.M. wrote with her right hand, although she had been left-handed before starting junior high school. Nevertheless, she is able to use either hand, according to circumstances. For example, she uses her left hand to eat, and uses her right hand to feed infants. One standard scale of handedness (Oldfield, 1971) suggested ambidexterity in Y.M., while another suggested dominance of the left eye and ear (Porac and Coren, 1981). In a dichotic listing task, Y.M. exhibited a left ear advantage (right/left correct = 90.0/98.9%) and her ear asymmetry index was – .05, indicating that the right hemisphere was dominant in verbal processing.

Primary perceptual and motor functions were estimated using the following tasks. Her visual field was assessed using a tachistoscopic presentation of Landolt rings. The visual angle of the ring was 1.0° in diameter, and it was presented for 66 msec at nine different positions (a central position and eight peripheral positions, i.e. up, down, right, left, upper right and left, and lower right and left) on a computer monitor (IBM ThinkPad 2681-KGJ). Each peripheral stimulus was 2.5° from the central fixation point. In the session, she was asked to name the direction of a gap in each ring. Her responses were very accurate; she scored 100% for each position. In the finger-tapping test (Shimoyama et al., 1990), she was asked to tap the plate as rapidly as possible with each index finger for 15 seconds; she was compared with three comparable right-handed subjects (30.7 ± 1.7 years). The plate functioned as a touch sensor that could detect a finger touch electrically; it was connected to a personal computer. Therefore, with this apparatus, there was no pressure intensity variance as with a keyboard. In all three comparable subjects, the right hand was much faster (5.99 ± .80 Hz) than the left hand (5.14 ± .81 Hz); in contrast, however, Y.M.'s left hand was faster (4.26 Hz) than her right hand (3.47 Hz). In the pegboard test, she was asked to insert a peg, in the form of an aluminium rivet (3 mm in diameter and 11 mm long), into 25 holes at 15 mm intervals; her performance was compared with that of three right-handed subjects (30.7 ± 1.7 years). In the other subjects, the right hand was much faster (65.7 ± 4.0 sec) than the left hand (78.7 ± 9.0 sec); however, Y.M.'s left hand was again much faster (70 sec) than her right hand (124 sec). As described above, she had no significant visual field defect and her left hand motor function was relatively preserved; however, her right hand was rather clumsy, despite the fact that she usually used her right hand for writing and another activities.

TABLE I

The results of various neuropsychological examinations. The score for the Wechsler Adult Intelligence Scale-Revised (WAIS-R) and Wechsler Memory Scale-Revised (WMS-R) are represented as a standard score. The result of the Benton Visual Retention Test (BVRT) is represented as a raw score. The age-matched average score was 100 for the WAIS-R and WMS-R scales, 10 for the 11 subtests of the WAIS-R scale, 8 for the correct score of the BVRT, and 2 for the error score. Y.M.'s intelligence and memory (excluding verbal memory) were well preserved

Wechsler Adult Intelligence Scale-Revised (WAIS-R)	
Full scale IQ	115
Verbal IQ	111
Information	9
Digit span	19
Vocabulary	8
Arithmetic	12
Comprehension	9
Similarities	13
Performance IQ	119
Picture completion	9
Picture arrangement	17
Block design	13
Object assembly	10
Digit symbol	15
Wechsler Memory Scale-Revised (WMS-R)	
General memory	86
Verbal memory	74
Visual memory	133
Attention/Concentration	115
Delayed recall	98
Benton Visual Retention Test (BVRT)	
Correct score	10
Error score	0

Table I shows the results of various neuropsychological examinations that were carried out on Y.M. According to the revised Wechsler Adult Intelligence Scale (WAIS-R), her intelligence (including verbal and performance IQ) was above average. Despite evidence of verbal memory deterioration, according to the revised Wechsler Memory Scale (WMS-R), Y.M. had no apparent problems with memory in everyday life. As in other cases of callosal agenesis (Ettlinger et al., 1972), she also had no signs of classical disconnection syndrome, such as tachistoscopic unilateral alexia, or deficits in her ability to draw, construct, name objects, and write with either hand.

Coronal MRI sections revealed ACC without any other malformations (Figure 1, left). The anterior commissure was visible in a sagittal MRI section (Figure 1, right).