Visual deficits in a patient with 'kaleidoscopic disintegration of the visual world'

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We describe psychophysical, neuropsychological and neuro-ophthalmological studies of visual abilities in a patient who, following a right hemisphere stroke, had difficulty in combining parts of objects into a whole and in reading. Strikingly, her perceptual problems were accentuated when the objects moved or when she moved. Formal testing showed that her main deficits were in depth perception, various tasks of motion and object recognition of degraded stimuli. But low-level detection and discrimination of form and color were normal. Despite her deficits in visual motion and degraded static-object recognition, her visual recognition of 'biological motion' stimuli was normal. Structural magnetic resonance imaging revealed an infarct in the ventromedial occipito-temporal region, extending ventro-laterally and leading to a 'kaleidoscopic disintegration of visible objects'.

Introduction

The neurological literature abounds in reports of bizarre visual disturbances in patients with brain lesions. Sometimes, the patients' subjective complaints can be satisfactorily explained by a total or partial loss of specific visual abilities, reflecting focal damage to different cortical visual areas and characterizable by neurological, neuro-ophthalmological, neuropsychological and psychophysical measurements. But in other patients the pattern of visual disturbance is much less readily interpreted and can even lead to misdiagnosis. A case in point is the long and controversial history of simultagnosia, defined as 'the inability to apprehend the whole although the parts are well recognized.' Kinsbourne and Warrington (1962) and Wolpert (1924) reviewed this controversy when considering both the specific definition of the syndrome and its possible neurological basis as opposed to a more general 'intellectual deficit' (Weisenburg and McBride, 1935/1964) or a 'psychological loss on a high plane' (Nielsen, 1946).

The patient we describe here, Mrs BC, noted progressive worsening of her visual perception, exemplified by loss of ability to recognize faces, inability to read text or words and a failure to 'pull objects together into a whole.' She described her condition as a 'kaleidoscopic disintegration of visible objects', which both frightened and disturbed her. Her complaints led to her initial admission into a psychiatric service, and only later she was admitted for a neurological assessment. The patient's spontaneous description of her visual deficits together with some of the neuropsychological and psychophysical measurements pointed to simultagnosia. However, on neuroanatomical grounds and on the basis of the results of the psychophysical evaluation, the diagnosis of simultagnosia was less convincing. Instead, BC has a disorder in which she has difficulty in correlating parts of the visual scene when they move, or when she moves, and her disorder indicates that low-level information – especially about motion – is disconnected from mechanisms of higher-order form perception.

Case report

The patient (BC), a 45-year-old right-handed woman, was admitted into the acute care Neurology Unit for further assessment of progressive visual-perceptual deficits secondary to severe neurological complications and a stroke associated with a complicated endocrinological history.

Eleven years earlier BC was diagnosed as having progressive Cushing's disease, with severe complications, that required a transphenoidal resection of a pituitary tumor, radiation treatment 1 year later and adrenalectomy 9 years after that. The adrenalectomy resulted in Nelson's syndrome, which is characterized by rapid regrowth of the pituitary tumor. Nine months after the adrenalectomy, BC was seen again in the Neurology Unit for complaints of severe and unusual visual-perceptual deficits, headaches and blurred vision. A magnetic resonance imaging (MRI) of the brain

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confirmed recurrence of her pituitary tumor. A second transphenoidal resection was performed and 4 days after the surgery she developed left hemiplegia, slurred speech and left hemianopsia with macular sparing. However, her blurred vision recovered completely. She described her vision as again 'sharp.'

The patient underwent rehabilitation and made an excellent recovery within 2 months, walking with a left ankle and foot orthosis and a cane in her right hand. Her speech recovered to normal, but her left hemianopsia did not improve. Vision in the right visual field was clear, sharp and felt normal.

Two months later, BC noticed progressive deterioration of her vision of a kind she had not experienced before. She felt she was losing the ability to cope with the 'visual world', and this was devastating to her as she was artistic, and had superior visual abilities, on which she relied in her job as an interior designer. She was distressed that she had lost her mental imagery, one of her 'best qualities' and crucial to her job. She could no longer imagine familiar scenes, the spatial layout of her own house, and the faces of friends and relatives. However, she could imagine and describe accurately and even imitate, the gait of people she knew or of members of the laboratory who produced exaggerated and distorted gaits to determine to what extent she could identify or mimic them.

Her new symptoms were attributed to a reactive depression, presumably induced by the emotional response to her recent stroke. She was treated with antidepressants and psychotherapy. Her visual disturbances persisted and several weeks later she reported difficulties in seeing faces as a whole despite the ability to see and describe individual facial features. The severity and persistence of these visual symptoms eventually led to her admission to an acute-care hospital where she underwent detailed neurological evaluations. She was referred to one of the authors for assessment of her visual-perceptual performance. The results of all the neuropsychological examinations described in this study were obtained at Boston University in the Brain and Vision Research Laboratory during the subsequent 8 days. The patient agreed to participate in this study and signed the Informed Consent according to the regulations of Boston University Human Subject's Committee. Age-matched normal controls participating in this study for obtaining comparative results also gave informed consent according to Boston University Human Subjects' Committee requirements.

The patient's subjective reports

When first examined BC complained of seeing the world in pieces and having special difficulties in perceiving objects when they moved or when she moved. When walking she felt unsteady and had a sensation of a 'kaleidoscopic disintegration of visible objects.' She had reported difficulty in visually recognizing familiar people like her mother, sister or physician, instead recognizing them by their voice, hairstyle or characteristic facial features. She noted that she had no difficulties in identifying the presence of individual facial features, but that 'making them into a face is out of the question. I don't understand why, I always was a very visual person!'. Reading became impossible; she identified individual letters with no problems, but was not able to 'see the letters in a word.' Written words appeared to her to be '… garbled, just a long string of something. I can't make sense of them when I try to read. It's just too overwhelming.' Television images looked like 'masses of colors', and pictures on magazine pages became confusing when she flipped through them quickly. Overall she felt a disturbing inability to pull the world together into one piece. This was most severe when she attempted to walk, as it appeared that 'things were moving' around her as she was moving, including normally immobile objects. She described feeling as if she had no reference to where things were or how they related to her. She could not tolerate simultaneous conversations or more than two people in the room without feeling overwhelmed.

Neuroradiological studies

The infarct involved the right temporal lobe, including the medial temporal gyrus, and extending posteriorly into the lateral temporal occipital gyrus, and up into the inferior portion of the parietal lobe. The lesion also extended along the lateral margin of the trigone and occipital horn of the lateral ventricle. The relevant axial images are shown in Fig. 1. The temporal and parietal infarction was noticed in the first computer tomography (CT) scan but the infarcted area had increased in size in the intervening period, particularly in the more anterior portions of the temporal lobe. A sellar and right sided suprasellar mass was identified on the double echo sequence. A subsequent magnetic resonance angiogram showed signs of partial occlusion of the right internal carotid. An MRI study of the brain was made at the time of the present study.

Figure 1 allows better visualization of the cortical damage produced by the stroke with a 3D morphometric reconstruction of the brain, derived from the MRI data. The lesion involves Brodmann's areas 38, 21, 20, 37, and part of 39. Area 19 also appears to be slightly involved. The posterior view reveals the sparing of the medial aspects of the occipital lobe consistent with an infarct in the streambed of the middle cerebral artery.

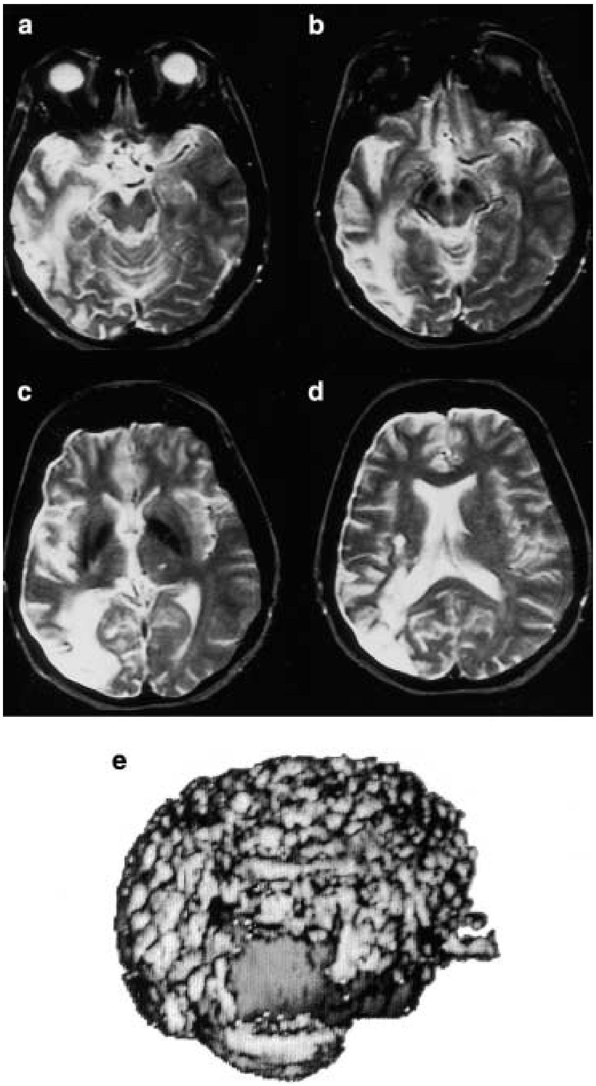

Figure 1 Axial slices of the magnetic resonance imaging (MRI) of BC's brain. (a) The infarct is seen in the right hemisphere (left side of the axial slice) extending throughout the temporal lobe ventrally, with widening of the CSF space along the right lateral margin of the suprasellar cistern and the temporal horn of the lateral ventricle. (b), (c) and (d) show tissue loss around the trigone of the lateral margin of the occipital horn into the lateral portion of the occipital and parietal lobes. (e) 3D reconstruction of the lesion from the axial images show its caudal and lateral extension into the vicinity of the motion area MT/V5. Brain images were acquired with a 1.5-T MR General Electric SIGNA System (GE Medical Systems, Milwaukee, WI, USA). A spin-echo, double-echo acquisition, covering the whole brain, was performed in the axial plane. Slice thickness was 5 mm; the field of view was 28 cm, and slices were acquired contiguously (no gaps) by combining two interleaved sequences in the individual acquisitions. Half-Fourier sampling (0.5 NEX) with 28 slice locations was acquired using 192 phase encoding steps, and echo time of 30 and 80 ms, with a repetition time of 2000 ms. Flow artifact was reduced with a gradient moment nulling flow compensation technique (Jolesz, 1990).

Neurological and neuro-ophthalmological examinations

The patient presented with some residual weakness and impaired coordination and function of her left extremities. Several cognitive and perceptual evaluations were initially conducted at bedside. The results indicate that short- and long-term memory were normal. On the letter cancellation task she performed slowly, but accurately. Design of a 3D cube, both copy and from memory were normal, as was her performance on the Rey-Osterrieth figure. She had no aphasia. Her performance was normal on the line bisection task (Bisiach *et al*., 1986). In this task, she was presented with 20 lines, one at a time, and requested to indicate the center of the line.

Eye examination was normal (20/30 OU without corrective lenses, and 20/20 with correction). Both saccades to command and finger smooth pursuit were normal and there was no evidence of nystagmus. Optokinetic nystagmus (OKN) was present, and normal to the left, but poor to the right. She had no optic ataxia or ocular apraxia. Color discrimination and color matching were normal. Formal visual field testing with Goldmann perimetry, repeated twice, showed a complete left homonymous hemianopsia without macular sparing (Fig. 2).

Neuropsychological evaluation

A brief neuropsychological evaluation was obtained to assess BC's perceptual and cognitive abilities. A short version of the Performance part of the Wechsler Adult Intelligence Scale-Revised (WAIS-R) was administered. Three tests in the Performance IQ set were administered. Her scaled scores were: 6 on Picture Completion; 5 on Block Design and 3 on Object Assembly, which is impaired compared with age-matched control subjects. Moreover, these scores indicate considerable impairment for a person whose visual-perceptual abilities as an interior designer were excellent prior to her stroke. Her performance on the Trail Making test (part B) was normal. In this test the subject has to draw a connecting line alternating between numbers (1–13) and letters (A–L) without taking the pencil off the page. The beginning and the end of the sequence are pointed out. The score records the time taken to complete the task together with the number of errors. BC needed 75 s to complete the task which, for her age, corresponds to roughly 50th percentile, indicating normal performance (25th percentile is the cut-off performance point suggestive of brain damage). This test measures planning ability as well as visuo-motor speed and concentration. On the Milner Faces Test (Milner, 1958), her performance was perfect for

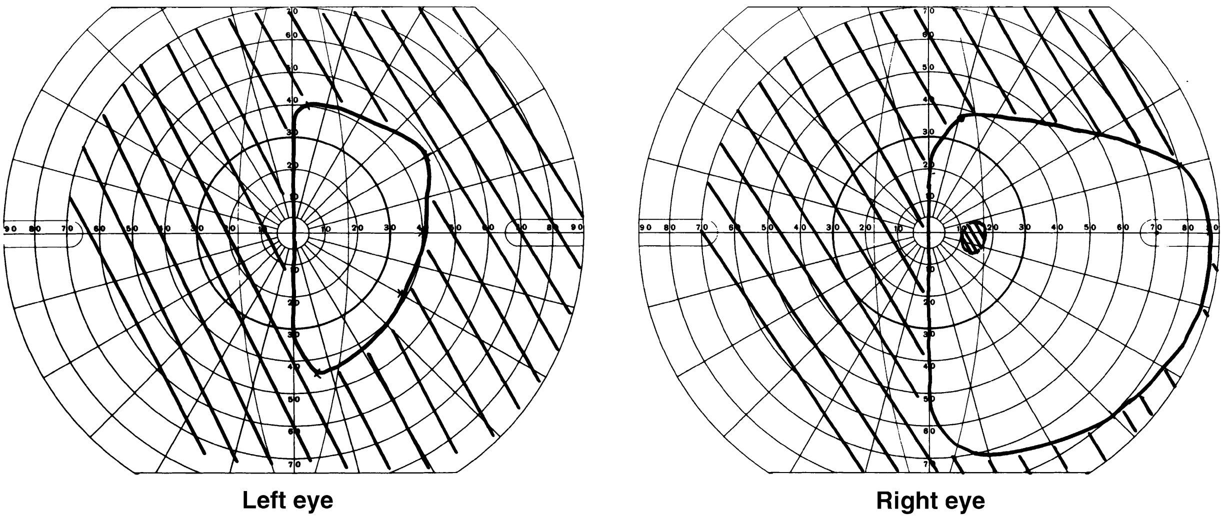

Figure 2 Visual fields of patient BC. Goldmann perimetry using a V4e test spot size shows a complete left homonymous hemianopia without macular sparing and with peripheral contraction of the field of the left eye.

matching (12/12) but below average for recall (8/12). However, she did both parts of the task very slowly. She correctly identified all faces of celebrities (e.g. Ronald Regan or Star Trek's Mr Spock).

She was conspicuously impaired on the interpretation of complex pictures. Thus, for example, when shown the 'Telegraph Boy' from the Binet Scale (Fig. 3) she described it as follows: 'a forest, or a park, this old-fashioned car – an old picture, isn't it?, a hat, oh, a young man is catching his hat, probably someone threw it to him, it looks as if it fell from one of those trees – no, it couldn't have, the car is there in between so the trees must be further than they look. I always see things closer these days – of, there is also a bike, then here to the right a rock, or a shoe upside down, then a wheel, or something like wheel. Ah, the bicycle must be broken, because the wheel is on the ground. I see, this fellow has a broken bike, but he holds it with one hand, and in the other hand he catches the hat … ah, no, he is perhaps using the hat to stop the car for help. The car comes towards him.' This piece-meal way of describing a straightforward scene was consistent; she described magazine pictures similarly. She was aware that the description was sketchy, and that she had to reason in order to integrate visual information.

In order to control the effect of stimulus size on BC's inability to grasp the whole of the picture, we showed her differently sized black and white drawings of complex scenes (the basic pictures subtending 4 × 5 in. were either enlarged by 25%, and 50% or reduced by 25%, 50% and 75% with a photocopier). Size made no difference.

She was bothered by television, complaining that 'it is too much, I cannot put it all together'. Both on television and in her surroundings, she had difficulties making out what was going on in a scene especially when there was movement.

Object recognition

In contrast to her inability to recognize and understand multi-element scenes, especially when they moved, BC

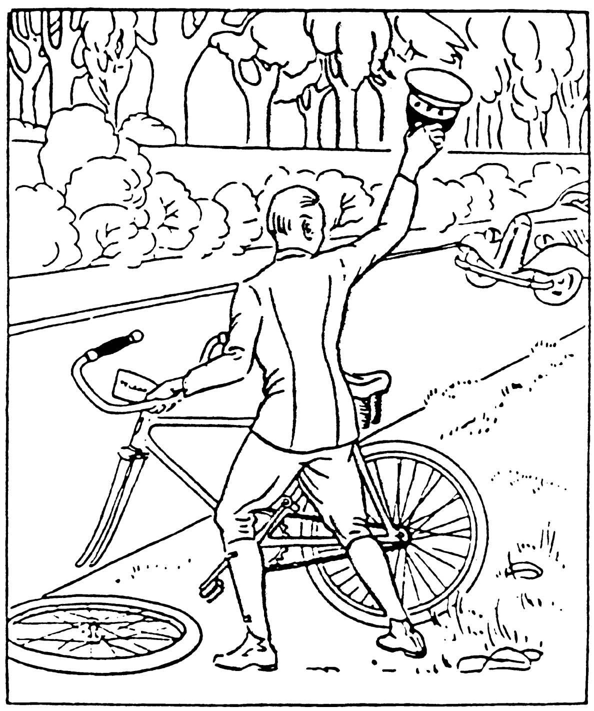

Figure 3 The picture of the 'telegraph boy' from the Binet scale. The picture was presented on a postcard size card at normal reading distance.

promptly recognized individually presented real objects or their photographs from a conventional front view. Given the evidence (McCarthy and Warrington, 1990) that complex or degraded pictures of common objects are more difficult to recognize than real objects or clear pictures, and that recognition can be poor when objects are photographed from unconventional views we used two tasks to assess BC's ability to cope with degraded or unusual information. She was severely impaired on the Unusual Views test adapted from Warrington and Taylor (1973). The test consists of black and white photographs of common objects shown from an unconventional viewing angle (Fig. 4). The test has two parts. First the subject is asked to recognize the objects presented in an unusual view, and then, if failing, to identify the same objects presented from a conventional front view. BC failed the initial part of the task (Table 1). For example, she described the picture of a clarinet as 'something round with shoe laces', the toaster, as a 'mailbox', goggles as a 'fire hose.' However, she recognized all the objects when shown in their prototypical, front view.

BC was also impaired on the Gollin Pictures test (Gollin, 1960), a series of incomplete line drawings of common objects. We presented 10 sets of five drawings each. Initially, a very fragmented drawing is presented, and the subject is asked to identify it. If the subject fails, then increasingly more complete versions of the drawings are shown (Fig. 5). The results are shown in Table 1.

Psychophysical measurements

Visual detection and discrimination of static stimuli

Spatial contrast sensitivity, evaluated with the Vistech 6500 chart (Ginsburg, 1968) at spatial frequencies of 1.5, 3, 6, 12 and 18 cycles/degree, was normal. Using computer-displayed horizontal sinusoidal gratings subtending 10 degree^2, we tested static and moving contrast sensitivity at two spatial frequencies (0.2 and 1 cycles/degree). The displays were generated and presented using a Macintosh Quadra computer and a black and white monitor augmented with the Pelli attenuator to reduce contrast (as described in detail in Saiviroporron, 1992). In the computerized test we used an adaptive staircase procedure starting at a contrast level of 10%.

Color discrimination

This was evaluated with the Farnsworth-Munsell 100 Hue Test (Farnsworth, 1943). This test actually has only 88 hue chips arranged in four groups of 22. A group of 22 is presented in a predetermined random order in a single row. The task is to sort them into an orderly progression of hues along the row between predetermined anchor hues at each end. BC's performance was normal, with only a few incorrectly positioned colors.

Table 1 Scores on the neuropsychological tests for BC and for control subjects

Performance IQ		Milner faces	
Picture completion	6	Matching	12/12
Block design	5	Recall	8/12
Object assembly	3		
Perceptual categorization	BC	Controls	
Unconventional views			
No correct	7/20	18/20 (SD = 1.05) ($N = 14$)	
Gollin pictures			
Error score	15/40	6/40 (SD = 2.45) ($N = 8$)	

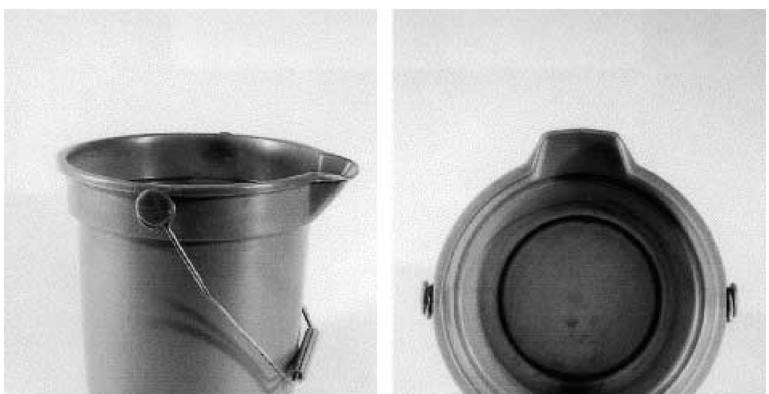

Figure 4 Examples of a water bucket from the conventional and unconventional views test, adapted from Warrington and Taylor (1973). The picture was of postcard size.

Figure 5 Two examples from the Gollin's Picture test, showing a fish and a saloon motor vehicle. All the pictures were of postcard size.

Shape detection

The test was a figure-ground discrimination adapted from Warrington and James (1988). The stimulus (Fig. 6a) consists of a slightly noisy X or O superimposed on a background of denser random noise. Task difficulty was varied by adjusting the ratio of black to white in the figure texture relative to the background texture. In each trial subjects were asked to report whether they could detect the letter or part of it. There were 20 trials for each X or O. BC's ability to identify whether a letter was present in the display was errorless.

Shape discrimination

This is a computerized task closely replicating the Efron's 'square' test for shape discrimination (Efron, 1968; Warrington and James, 1988), in which observers discriminate between a square and an oblong subtending the same area but differing in the proportion of the horizontal and vertical dimensions (Fig. 6b). In the present task, the observer fixated a small fixation mark in the middle of the computer screen. For BC, stimuli were shown in her normal visual field, for 500 ms and one at a time. She was asked to indicate whether the stimulus was a square or an oblong. The stimuli were black on a white background and were either a square ($5° \times 5°$) or an oblong with dimensions: $5.25° \times 4.77°$ (the hardest discrimination), $4.6° \times 5.5°$, or $6.5° \times 4°$ (the easiest discrimination) (Fig. 6b). Ten target squares and 10 oblong distractors were presented in pseudorandom order for each level of difficulty. Normal observers were given exactly the same task to obtain comparative results. BC's performance was normal. The stimuli were then presented simultaneously with both figures displayed side by side in the patient's intact right visual field. Only the easiest discrimination was shown ($5° \times 5°$ vs. $6.5° \times 4°$). BC and 16 age-matched normal controls were asked to judge whether the shapes were the same or different. BC's score was at chance despite having scored 100% on the immediately preceding more difficult task. She reported spontaneously that this task was 'impossible.' Normal controls scored 97% correct (155/160 correct responses). To be sure that BC's failure to perform the task was not because of the fact that one of the stimuli was presented too eccentrically, we repeated this task but the stimuli were again presented one at a time at 10° eccentricity (the center of the stimulus). Her performance was 9/10 correct.

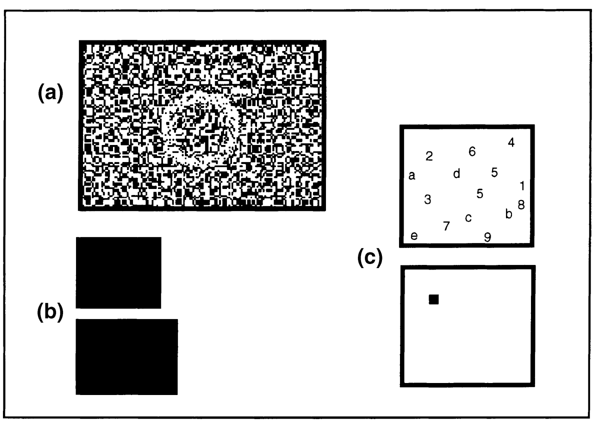

Figure 6 Examples of static stimuli: (a) shape detection; (b) shape discrimination; (c) spatial location.

Spatial localization

The stimuli, presented in a computer adaptation of MacQuarrie's Test (MacQuarrie, 1953), consisted of two large squares (Fig. 6c) each subtending $8° \times 8°$ displayed simultaneously one below the other. The top square contained randomly placed alphanumeric characters and the bottom, a small black square of 5.6 arcmin diameter. The subject has to identify the character in the top square corresponding to the position of the black mark in the bottom square. BC scored 52% correct (25/52 trials), which was below the 5% percentile scores obtained in an age-matched control group.

Binocular (global) stereopsis

The stimuli consisted of a series of static random dot stereograms from the series devised by Julesz (1971).

When viewed monocularly each target appears as a random array of small light and dark squares in which no form or depth is apparent. The two random dot patterns are identical except that in one pattern a contiguous cluster of dots in the central region has been displaced laterally with respect to the same region in the other pattern. Two disparities, corresponding to 4′ and 8′ were used. There are no monocular cues to depth in these patterns but with binocular fusion a central figure stands out in front of the surround. The patterns were printed in pale red and green ink. When viewed with a green filter over one eye and a red filter over the other, the central figure is seen in front of (or behind) the surround. We used five random dot stereograms. The subject's task was to indicate whether she saw a central figure standing away from the surround, and then to identify its shape. All the figures were simple geometric forms.

BC lacked any binocular stereopsis at both disparities. She saw 'just dots, no pattern or form at all' in the random dot stereograms. The loss of stereoscopic vision was recent, as it was normal in the previous neuro-ophthalmological examination 3 months earlier. Stereopsis was also absent when measured with the clinical Randot Stereotest (Stereo Optical Co., Inc., Chicago, IL, USA) based on random dot stereograms of various disparities printed on polarized cards and viewed through polarized glasses.

Depth perception (local stereopsis)

The apparatus (Howard, 1919) consisted of a box, 60 cm long and containing two illuminated parallel vertical rods (background illumination was 1 foot candle) about 1-cm thick and visible length of about 8 cm. The rods, positioned 4° apart laterally were attached to strings of a pulley and were viewed through a front opening in the box 12.5 × 7 cm. One rod was fixed whilst the other was positioned 3, 7, 10, 15, 22, 30, 35, or 40 mm either in front or behind the fixed rod. Subjects were dark-adapted to the experimental environment for 5 min, before judging binocularly, at a viewing distance of 4 m, whether the two rods were equidistant from their viewpoint. The position of the moveable rod on each trial was determined by a pseudorandom order. Subjects were instructed not to move their head and pull the strings until the rods appeared aligned. Trials where there was head movement were aborted and then repeated. There were 24 trials, three for each of the eight distances. BC was unable to reliably discriminate distances smaller than 35 mm between the rods, whereas all the age-matched normal control subjects ($n = 5$) were able to align them within 7 mm of each other.

Visual motion perception

The patient's complaints of being disturbed by moving objects and not being able to judge how fast they were coming or going, motivated the formal evaluation of her visual motion perception, using a battery of psychophysical tasks. We first tested the short-range process described by Julesz (1971) and Braddick (1974) by investigating whether BC could perceive form or contour from differences in motion across neighboring spatial regions. In three tests of increasing difficulty, we tested whether perceptual segregation could occur between a region moving past a stationary region, between regions differing in direction of motion, and between regions differing only in velocity magnitude. We also assessed her ability to discriminate motion speed. Next, we studied how BC might integrate motion information to extract global motion direction in the absence of local cues. Finally, we investigated the integrity of her higher-order motion abilities with three tests: 3D structure-from-motion cues alone, perception of long-range motion, and recognition of 'biological motion.

General methods and procedures

A Macintosh IIcx computer with an extended 8-bit video card was used to generate and present all stimuli (except the 'biological motion test') as well as to collect and analyze responses. The stimuli were presented at the center of a Macintosh RGB monitor at a resolution of 640 by 480 pixels and a vertical scanning frequency of 66.7 Hz. Random dots were used to minimize familiar position cues and to isolate motion mechanisms (Nakayama and Tyler, 1981). Each screen pixel subtended 1.8 × 1.8 arcmin at the viewing distance of 65 cm. The background was black and the random dots were white. Viewing time was 2 s per trial. All the tests, except motion coherence, employed the method of constant stimuli. The stimuli in the motion coherence test were generated by an interactive staircase procedure driven by the subject's responses (Vaina *et al.*, 1990c). The control group consisted of age-matched volunteers with no known ophthalmological, neurological or psychiatric disorders. Most were spouses and friends of other patients who had been tested in the laboratory. It was the subjects' first experience of psychophysical testing. All subjects had corrected-to-normal vision.

Before each experimental session, the subject was familiarized with the task through examples and feedback and was dark adapted for 5-min before each experimental session, following which no feedback was provided. The subject started each trial by pressing a designated key. The room illumination was maintained at a low photopic level, and the subjects were instructed to restrict fixation on the square fixation mark, which was placed 2° to the left of the center of the imaginary boundary of the stimulus. This assured that the entire stimulus was within BC's intact visual field. The same arrangement was used with the normal controls.

Boundary from relative motion

As BC had intact static 2D form discrimination, we used the form-from-motion task introduced by Julesz (1971) to evaluate early motion processing. Psychophysical studies of motion segregation have demonstrated that the human visual system is capable of detecting motion discontinuities when the background is static (Julesz, 1971) or when the figure and the background move at different velocities (Baker and Braddick, 1982). The extraction of the objects' boundaries appears to be mediated by a short-range process (Braddick, 1974), a visual mechanism which matches up corresponding local pattern elements of the same luminance polarity in successive time frames and operates over short time intervals and spatial separations.

Two-dimensional form-from-motion in a static background

The sensation of a moving textured planar surface was elicited by a patch of contiguous random dots uniformly displaced from one frame to the next in a translational motion across a random dot stationary background (Fig. 7a). The moving shape was defined solely by the relationship of the displacement between each moving patch and the static surround. The moving shape had one of the following outlines: square, circle, triangle, cross or oblong (orientated horizontally or vertically). The square, circle and the cross had roughly the same area, and the oblong was half of the square area. Static black silhouettes of the moving shape were shown at the bottom of the display and numbered from one to six.

The display subtended $10° \times 10°$ and the moving patch covered approximately $2.2° \times 2.2°$ of visual angle and moved at roughly 3°/s. In a six-alternative forced-choice, the subject was asked to identify the moving shape by reporting the number corresponding to the shape on the bottom of the screen or by orally naming the shape. Out of 30 trials BC scored 95% correct (Fig. 7b). This excellent performance was not surprising given earlier reports (Vaina, 1988, 1989; Vaina *et al.*, 1990) which showed that the task of perceiving a moving shape against a stationary background does not distinguish motion-deficient from normal subjects. But it is important in showing that BC's form perception is not impaired by motion *per se*.

Boundary localization by relative motion

In this experiment, the two halves of the display moved either in different directions or in the same direction at different speeds. The stimuli (similar to those used by Hildreth, 1984) were dense dynamic random dot fields subtending $12° \times 8°$ of visual angle. In each trial, a sequence of 50 frames was constructed in such a way that there was a vertical boundary of discontinuity in the velocity field (Fig. 7c,e). Located on right side of the boundary was a $1.4° \times 1.4°$ notch (protrusion of one field into the other), whose distance from a central black mark (0.5×0.5 degree^2) varied along the vertical from trial to trial but remained within 2° of visual angle above or below it. The imaginary vertical boundary and the notch were entirely defined by the difference in velocity between the moving dot fields and were invisible in any static frame.

The discontinuity was obtained in two ways: (i) by direction differences between the two regions (Fig. 7c), using four angular differences (18.4, 37.1, 90 and 180°); (ii) by speed differences between the two regions (Fig. 7e), using three speed ratios (1.5, 2 and 3). The subject was instructed to maintain fixation on the black mark at the center of the display. The experimental session consisted of 20 trials for each condition using a two-alternative forced-choice (2-AFC) task in which the subject had to decide whether the notch was above or below the fixation mark.

Figure 7d,f shows that these tasks were relatively easy for the normal control subjects. BC's scores were normal on the direction-defined boundary test, but slightly impaired on the speed-defined boundary test. On the boundary discrimination task, she had a perfect score for the larger angles, and scored 80% correct on the smallest angle (18.4°). Her score on the boundary-by-speed test (Fig. 7f) demonstrated that for all ratios tested her performance was either at chance or she performed significantly worse than the age matched normal observers (for ratio 1.5, $Z = 3.4$; for ratio of 2, $Z = 3.1$ and for ratio of 3, $Z = 2.8$).

Encoding average speed and coherent motion in dynamic displays

Experiments conducted so far employed stimuli in which local motion was highly coherent. In the next two experiments the stimuli were very different, consisting of rapidly fluctuating dynamic dot displays. Here the observer is confronted with motions that are locally incoherent and where the task is to judge the average

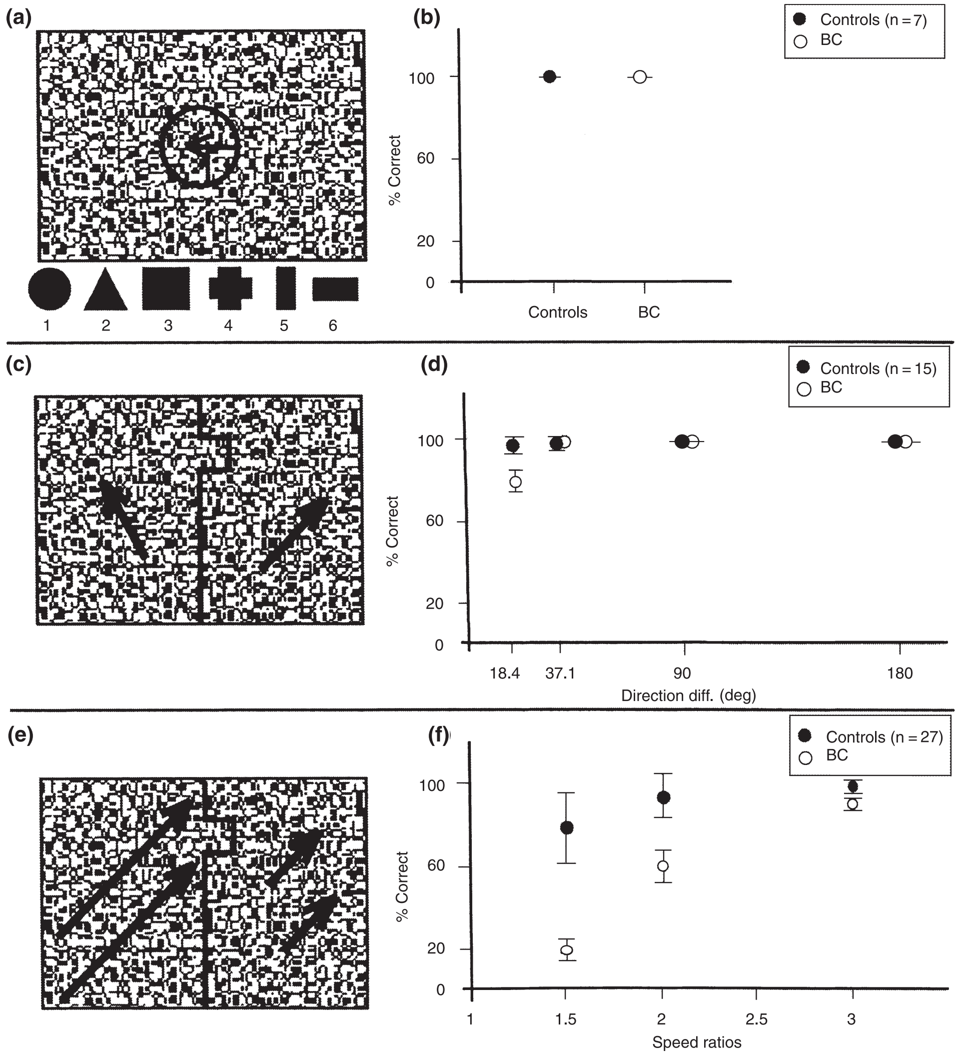

Figure 7 Examples of motion tests and BC's results compared with those of normal control subjects: (a) and (b): form-from motion in a static background; (c) and (d): boundary location from direction differences (as indicated by the black arrows); (e) and (f): boundary location from speed differences.

speed of the overall cluster of moving dots or to detect motion of a small number of coherently moving dots within this dynamic field.

Local speed discrimination

The previous psychophysical task indicated that BC was profoundly deficient in seeing 2D form from differences in velocity magnitude. To examine her possible 'speed' deficit more directly using a very different technique, we measured her ability to judge the average speed of a cluster of random dots. The stimuli consisted of two sparse dynamic random dot kinematograms each comprising 20 computer-generated dots. The kinematograms were displayed in two rectangular apertures shown one after the other (with 250 ms interval), each subtending an area of $4° \times 2.5°$, thus giving a dot density of 2 dots/degree^2 (Fig. 8). In any single trial each dot took an independent, 2D random-walk of constant step size defined by the speed. The direction in which any dot moved was independent of its previous direction and also of the displacements of the other dots. Each frame was on for 66 ms with no interframe interval. To maintain constant density a 'wrap-around' scheme was used, in which dots displaced beyond the boundary of the aperture reappeared on the opposite boundary in the next frame. Each kinematogram was shown for 990 ms. The speed of the dots, defined as a function of the distance a dot was displaced between successive frames, was uniform within a box and was assigned independently for each box. A base speed of 3°/s was always compared with

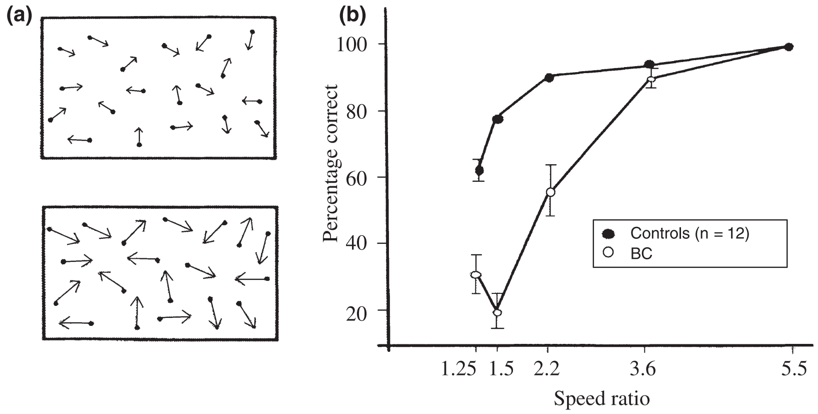

Figure 8 (a) A schematic representation of the stimuli employed. The display consisted of two sparse random dot fields each displayed in a rectangular aperture subtending 4° × 2.5°. Within each aperture the dots, plotted here as vectors, move in randomly distributed directions with constant speed. The apertures were displayed sequentially. The observers' must decide in which of the two apertures the dots move faster. (b) Results for BC and normal controls. The graph plots the percentage of correct answers as a function of the speed ratios between the two sequentially presented apertures. The data present the mean correct responses and the standard error.

five other speeds, giving five speed ratios of 1.25, 1.5, 2.2, 3.6 and 5.5. The assignment of the higher speed to the top or bottom aperture was pseudorandomly selected. Subjects had to indicate which of the two apertures (the first or the second) contained the faster moving dots. A two-temporal alternative forced-choice (2-TAFC) procedure was used for measuring the subject's ability to detect difference in speed.

In comparison with the age-matched control group, who were performing almost perfectly for 2 : 1 speed difference, BC was severely impaired. She failed to discriminate reliably speed ratios smaller than 3.6 and was performing at near chance when normal observers were performing better than 90% correct. BC's performance on this task is consistent with her results on the Boundary from speed differences test described above.

Motion coherence

The task was adapted from Newsome and Paré (1988). The stimuli were dynamic random dot kinematograms with a correlated motion signal of variable strength embedded in motion noise (Fig. 9a). The strength of the motion signal, i.e. the percentage of the correlated dots moving in the same direction, varied from 0 to 100%. The remaining dots moved in random directions providing masking motion noise. The algorithm by which the dots were generated is described in Vaina *et al.* (1990c).

The aim of this task was to determine the percentage motion coherence at which a subject could reliably discriminate in a 4-AFC paradigm the direction of motion (up, down, left or right). The dynamic random dot display was presented in a square 10° × 10° aperture situated at 2° left or right of a white fixation mark. The subject was instructed to maintain fixation on the fixation mark throughout each trial and to verbally report the global direction of motion in the display. Dot density was 2 dots/degree^2. The speed of motion, defined as a function of the distance a dot is displaced between successive frames, was 3°/s. BC was significantly impaired on the Motion Coherence task (Fig. 9b). Her coherence threshold direction discrimination was 35% for stimuli presented in her 'intact' right visual field and, as expected, little different when the stimuli were presented with free fixation and when we used different dot densities. The mean of the motion coherence threshold for the normal controls was 6.5% ($N = 16$). BC's performance on this task demonstrates an impaired motion mechanism which spatially integrates local velocity measurements.

Three-dimensional structure-from-motion: the rotating cylinder

This experiment examined BC's ability to create a 3D percept from the relative motions of elements in a changing 2D image. The display (Fig. 9c) consisted of two dynamic random dot kinematograms presented simultaneously, one below the other, each subtending 3 × 3 degree^2. Each kinematogram was composed of 128 dots with an average point density of 14 dots/degree^2, which was optimal for a normal control group. At the end of its lifetime the dot disappeared and was replotted at a new random location within the boundary of each display and began a new trajectory. One kinematogram consisted of dots painted at random locations on the orthographic projection of an imaginary transparent rotating cylinder, and the other

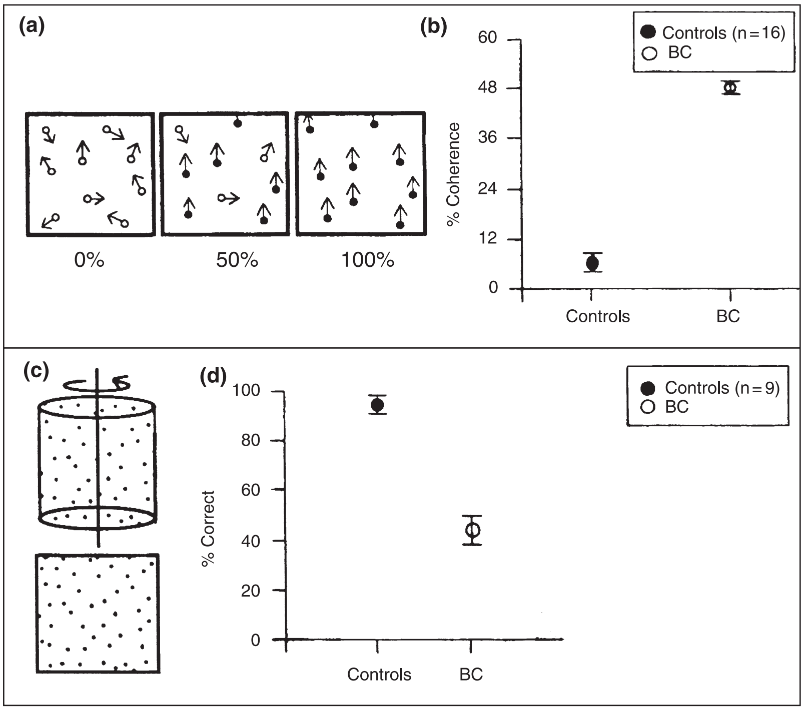

Figure 9 Global motion tests. (a) Motion Coherence. Left: 0% correlation between the individual dot motions. Right: 100% correlation (all dots move in the same direction). Middle: a stimulus with 50% correlation in which 50% of the dots are always plotted at random locations within the aperture whilst the other 50% of the dots are plotted in correlated motion. Dots maintain their motion direction throughout the stimulus display. Most of the trials consisted of stimuli intermediate between 0 and 100% correlation.
(b) Results on the motion coherence task from 16 age-matched control subjects and BC. The *y*-axis represents the percentage coherence. (c) 3D structure-from-motion in the two-alternative choice task.
(d) Results from BC and nine normal control subjects on the 3D structure from motion task.

showed a pattern of scrambled velocities. The angular velocity of the cylinder rotating about its vertical axis was 30°/s. The maximum distance travelled by a dot between two consecutive frames was 4.3 arcmin. Dots had a limited life-time of 400 ms and all dots were projected onto the imaginary surface of the rotating hollow cylinder (100% structure). The spatial positions (top vs. bottom) of the structured field and the unstructured field were randomly assigned. Subjects were asked to report which of the two fields of dots (top or bottom) portrayed a clearer cylinder. There were 20 trials.

BC was unable to do this task (Fig. 9d). She could not perceive a rotating cylinder; all that she reported seeing was, 'dots, flickering around, there is no difference between the top and the bottom. Am I really supposed to see something?' Worried that her deficit on this task might be because of her possible simultagnosia, we repeated the test but with only one stimulus. This is a generic structure-from-motion task (Vaina, 1989; Vaina *et al.*, 1990b,c). She was asked to report whether she perceived a rotating cylinder. She repeated that she could not see 'anything but moving dots, no cylinder at all.' She became frustrated with her inability and testing was curtailed.

Long-range motion

The perception of long-range motion, adapted from Green (1986) was measured by asking the subjects to judge the direction of rotation in a display composed of two pairs of 2D Gaussian-modulated sinusoidal gratings (Gabor patches) arranged at the four corners of an imaginary square. The stimuli consisted of four separate frames, displayed twice in succession for a total of eight frames in each trial. Figure 10 shows schematically that for each of the four frames only the position of the Gabor patches changed, not their orientations. The separation between centers of identical Gabor patches was 3.6°. For a 45° rotation, each Gabor traversed a distance of 1.4° of visual angle, well within the realm of long-range motion. Gabor patches of five different central spatial frequencies were used, 1.0, 1.7, 3.0, 5.0 and 10.0 cpd, with 1.7 cpd being the reference spatial frequency. In each trial eight frames were displayed, interleaved with seven interstimulus blankings, in one of two sequences corresponding to clockwise or counter-clockwise rotation. A 2-AFC procedure (clockwise or counter-clockwise rotation) was used with the method of constant stimuli. Subjects had to report whether the four Gabor patches appeared to rotate clockwise or counter-clockwise. Each combination of spatial frequencies was presented 12 times.

BC found the task easy and scored within the normal range (Fig. 10b). This might seem surprising both in view of the suspected simultagnosia and in view of her motion deficits. However, we have shown in previous studies (Vaina *et al.*, 1990c; Vaina *et al.*, 1993b) that performance on long range motion can be dissociated (both ways) from performance on other motion tasks.

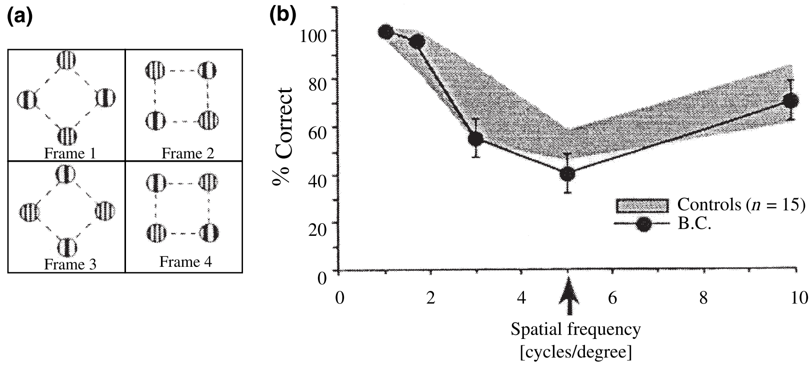

Figure 10 (a) Schematic representation of the frames used in the long range motion display. Circles labeled with the same letter represent Gabors of the same spatial frequency. The separation between centers of similar Gabor patches is 3.6°. For a 45° rotation, each Gabor patch traverses a distance of 1.4° of visual angle, putting this display well within the realm of long-range motion. Gabor patches of five different central spatial frequencies were used: 1.0, 1.7, 3.0, 5.0 and 10.0 cycles/degree, with 1.7 cycles/degree the reference spatial frequency. Clockwise rotation is produced by presenting the frames in the order 1, 2, 3, 4. Counter-clockwise rotation is produced by presenting them in the order 1, 4, 3, 2. (b) Results from normal controls and BC. The shaded gray area represents the mean results from the normal controls on each spatial-frequency pair ±1 SD, to indicate the range of the controls' data.

Biological motion recognition

A very different yet compelling 'structure from motion' demonstration was introduced by Johansson (1973). He called it 'biological motion' and defined it as the pattern of movement generated from the evolving pattern of dots placed at the joints of a moving human actor. Johansson demonstrated that the motion of the lights alone was sufficient to enable observers to perceive unequivocally the activity of the human actor. Such biological motion patterns are sufficient for the perception of specific actions, the perception of gender (Kozlowski and Cutting, 1977) and the perception of sign language (Poizner *et al.*, 1981), yet the only information available in the display is provided from the dynamic source alone. The pattern of the organization of the dots, and the pattern of their spatial modification over time is the only clue for recognition.

In our experiment, the stimulus was presented on a videotape using scenes from the original Johansson's movie. In the display one sees only the pattern of lights attached to the joints of the actor during the performance of some prototypical actions: walking, stair-climbing, riding a bicycle, push-up, two men walking, shaking hands and hugging. When static, the pattern gives no clue as to the identity of an object or the activities concerned. However, just from the motion of the pattern of lights, one swiftly has a vivid impression of a person performing specific actions.

The examiner gave no information about the nature of the display; the subject was only told that she would see a bunch of dots moving in a certain overall direction and that the task was to first report the direction of motion and then to describe what it looked like. Surprisingly, BC had no difficulties on this biological motion task. In every trial she quickly and correctly recognized that the point-light display portrayed a person carrying out a specific activity. In the biological motion stimuli the motion overall is non-rigid, but there are, however, local rigidity constraints amongst the pattern of dots.

Discussion

We first discuss BC's performance on the psychophysical motion tasks. Secondly, we consider whether she has simultagnosia in the context of her overall performance on the neuropsychological and psychophysical studies and in relation to the anatomical locus of her lesion.

A deficit of motion integration?

Table 2 presents a qualitative summary of BC's results on the static and motion psychophysical tasks. Her hugely elevated threshold on the motion coherence task reveals a major difficulty in computing global motion. The latter requires the spatial integration of local motion measurements, a processing stage which is beyond the simple detection of motion direction. We suggest that BC's poor performance on this task reflects a deficit in integrating the output of the local directional motion system, particularly in the presence of masking motion 'noise.'

Why did BC fail to perceive 3D structure from motion even at 100% coherence despite her ability to correctly identify all the human activities in the Johansson's 'biological motion' videotape and the

Table 2 Qualitative summary of results on the static and motion psychophysical tasks

Qualitative summary of psychophysical scores			
Static tasks			
Contrast sensitivity	Normal	Shape detection	Normal
Color discrimination	Normal	Shape discrimination	Impaired
Binocular stereopsis	Absent	Spatial localization	Impaired
Depth perception	Impaired		
Motion tasks			
Two-form from motion	Normal	Motion coherence	Impaired
Boundary from relative motion		3D SFM	Impaired
By direction	Normal	Long range motion	Normal
By speed	Impaired	Biological motion	Normal
Speed discrimination	Impaired		

direction of the point-light walker? In view of the fact that BC's discrimination of speed and direction were imprecise it is not surprising that she could not perceive 3D structure-from-motion cues. One common theoretical assumption is that 3D structure-from-motion computations are derived from earlier, metrical measurements of local motion (Longuet-Higgins and Prazdny, 1980; Lawton, 1983; Koenderinck and van Doorn, 1986).

However, we have previously argued that the perception of 3D structure-from-motion does not necessarily require precise low-level motion computations, as patient AF (Vaina *et al*., 1990b,c) whose visual motion perception was much like BC's, was able to do this task even in the presence of a small amount of noise.

Another way to compute 3D structure-from-motion is described by Ullman's Incremental Rigidity Scheme (Ullman, 1984), which requires the comparison of positional information in successive frames. Yet another theory (Ando, 1991; Hildreth *et al*., 1991; Treue *et al*., 1991) suggests that observers use global rather than local motion cues for deriving 3D surface from motion, and that the visual system integrates information over space and time by computing a 3D surface of the object. unfortunately, as neither BC nor AF is available for further and refined testing of motion perception, we cannot resolve the conundrum of what useful cues were selectively available to AF and not to BC.

In contrast with the poor performance on low-level motion tasks and failure to perceive 3D structure from motion, BC's excellent performance on the long-range motion test and her flawless performance on Johansson's biological motion task is both unexpected and remarkable. In the long-range motion task the impression of apparent motion depends on establishing a correspondence between elements with the same characteristics (here, spatial frequencies). The subject must match a displaced element with the nearest preceding element having the same features. Thus, it is possible that even with simultagnosia, BC succeeds in this task because matching involves sequential not simultaneous processing of discrete form features. The 'motion blind' patient LM (Zihl *et al*., 1983) and the severely motion-impaired patient AF (Vaina *et al*., 1990c), whose symptoms supported the idea of cortical areas specialized for motion processing, were also able to perceive human figures and their actions in Johansson-type displays (McLeod *et al*., 1996). However, BC's performance differs from LM's because the latter could recognize the actions portrayed in the biological motion display, but not the direction of motion of the human actor. McLeod *et al*. (1996) suggested that LM had access to an object-centered representation of the biological motion stimuli, but that she failed to access and recognize the viewer-center representations of the actions. Unlike LM, BC (like AF) was also able to correctly report the direction of the movement.

The processing of these stimuli involves both local groupings between subsets of points and the perception of relative motion amongst the groupings. Furthermore, the test involves the construction of global figural coherence, that is, the organization of the points into a human frame or shape. Indirectly, BC's normal performance on the Long Range Motion Task suggests that she can sequentially match such local groupings. Moreover, as form-changes characterize biological motion, it is possible to achieve recognition by integrating form over time, and not motion pattern. A recent fMRI study by Vaina *et al*. (2001) suggests that biological motion recognition can be achieved by using either form or motion cues.

Is BC's deficit a form of simultagnosia?

Tables 1 and 2 qualitatively present BC's results on a set of neuropsychological and psychophysical tasks. Her detection and discrimination of contrast sensitivity, color, form and boundaries based on directional difference were normal. However, she was severely impaired on tasks of spatial relations, spatial discrimination and depth perception. In the previous section we suggested that her deficits on motion coherence and 3D structure from motion could be interpreted as a deficit of visual integration of motion stimuli in the presence of noise (induced both by the short point life time of the dots and by signal/noise distribution of the dots).

BC was impaired on the performance subtest of the WAIS-R. When asked to describe complex figures (e.g. the picture of the 'telegraph boy') she described the parts accurately, but only through painstaking inference could she eventually arrive at a description of the whole picture. A deficit on this task is a classic example of simultagnosia. She also could not read beyond the letter level, another characteristic of simultagnosia. BC was impaired on neuropsychological tasks of object recognition, in which the stimuli were either degraded, presented as incomplete drawings (Gollin pictures) or in unusual views. Warrington and Taylor (1973) were the first to use the Gollin pictures test to assess object recognition ability in patients with localized unilateral cerebral lesions. They found that right hemisphere lesions affected performance more than left hemisphere lesions, and that the most impaired patients had right parietal damage. The unusual views test is particularly sensitive to parietal and temporo-parietal lesions. Based on several studies, Warrington and Taylor (1973) defined these deficits as a syndrome of perceptual categorization which is characteristic of right hemisphere lesions, especially right parietal.

Conclusion

The major deficits exhibited by BC can be grouped into four classes: (i) depth and spatial localization; (ii) motion integration in the presence of noise; (iii) perceptual categorization; (iv) the inability to grasp a whole (scene or word) although the parts are apprehended. The first three classes of deficits are characteristic of damage to the posterolateral parietal–temporal–occipital area, which is involved in BC's lesion. The deficits in Class d are like those of classic simultagnosia, specifically the integrative type of simultagnosia (Grüsser and Landis, 1991). A patient suffering integrative simultagnosia can discriminate single components of an object, but is unable to integrate them into a meaningful whole. The lesions in cases of 'integrative simultagnosia' described in the literature are either bilateral or, if unilateral, they involve the infero-medial occipital temporal area. BC's lesion was restricted to the right hemisphere but did invade the latter region ventrally. Her clinically bizarre symptoms presumably arise because the lesion leading to simultagnosia also involved lateral areas involved in motion processing, making it even more difficult to analyze a visual scene when either it or she moved.

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