Seeing, since childhood, without ventral stream: a behavioural study

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Summary
We report the case of a 30-year-old man (S.B.) who developed visual agnosia following a meningoencephalitis at the age of 3 years. MRI disclosed extensive bilateral lesions of the occipital temporal visual pathway (ventral stream) and lesions in the right dorsal pathway, sparing primary visual cortices. S.B. showed a severe visual recognition deficit (texture, colour, objects, faces and words), although movement and space perception were largely preserved. His remaining visual capacities illustrate the competence of an isolated dorsal system which essentially functions on the sole basis of magnocellular afferents (low spatial resolution, high sensitivity to low contrast and moving stimuli). Patient S.B. also shows remarkable visuomotor competences, despite his perceptual limitations. It is suggested that his perceptual capacities correspond to the visual processing limitations of the dorsal visual stream, which in this patient have become accessible to perceptual awareness.

Keywords: visual agnosia; ventral stream; dorsal stream; brain plasticity

Abbreviations: BORB = Birmingham Object Recognition Battery; PEGV = Protocole d’Évaluation des Gnosies Visuelles

Introduction
Two different streams for object vision and spatial vision were first inferred by contrasting the effects of inferior temporal and posterior parietal lesions in macaque monkeys (Ungerleider and Mishkin, 1982). Since this early work, multiple visual cortical areas in both monkeys and humans have been shown to be organized into two functionally and anatomically distinct streams—a ventral stream projecting to the inferior temporal cortex for object perception, and a dorsal stream connected to the parietal cortex for visuospatial control of movement. In an admittedly simplified way of summarizing the clinical and experimental data, one may attribute to the ventral stream the processing of ‘what’ and to the dorsal stream the processing of ‘where’ and ‘how’.

Neuropsychologists willingly accepted the proposal of Ungerleider and Mishkin. Indeed, since the beginning of the 20th century, the neuropsychological literature has distinguished syndromes related to occipitoparietal lesions (Balint’s syndrome, Holmes syndrome) and those related to occipitotemporal lesions (visual agnosia, prosopagnosia, alexia).

Some questions may arise from the relative independence of these two postulated streams: is object vision possible without any spatial vision? In other words, how can complex shapes or objects be identified without analysing the spatial relationships between their different parts? Functional neuroimaging evidence in any case indicates that visual object processing takes place in both the dorsal and the ventral streams (e.g. Kraut, 1997).

Milner and Goodale have argued that the ‘What–Where’ model cannot account for some new behavioural data (Milner and Goodale, 1993). Their neuropsychological investigations have shown that a patient with visual form agnosia remained able to grasp precisely objects of different size and orientation, while perceptively the patient could not appreciate size or orientation. This patient was able to process ‘what’ to a limited degree, but only when acting. In the proposal of Goodale and Milner, both streams are assumed to use information about objects and their locations, but each stream uses this information in different ways (Goodale and Milner, 1992). According to these authors, the ventral stream carries out transformations concerning characteristics of objects and their internal spatial relationships, therefore allowing the formation of long-term perceptual representations that are necessary to identify and recognize objects. In contrast, the dorsal stream carries out transformations that use moment-to-moment information about objects and their locations in

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egocentric frames of reference, thus mediating the visual control of skilled actions (such as pointing, reaching and grasping).

The present study reports the case of a patient (S.B.) with a pattern of visual agnosia that, to our knowledge, has not been described previously in the literature. After a meningoencephalitis at the age of 3 years, S.B. presented a severe visual deficit. The ensuing cerebral damage was extensive enough to destroy the right and left ventral streams as well as the right dorsal stream. At first glance, S.B. appeared to be blind, being unable to recognize colours, living and non-living objects, faces or letters. On the other hand, S.B. was strikingly good at many everyday visually guided actions. Facing such an apparently contradictory pattern, we wondered to what extent S.B.’s spared visual system, restricted to one dorsal stream, remained able to accomplish some recognition of the surrounding world. Accordingly, the purpose of this study was to investigate S.B.’s residual visual functions and to attempt an evaluation of the visual capacities of an isolated dorsal system. In other words, what is it like to see with one dorsal system only?

This case is peculiar not only because of the sites of the lesions but also because the cerebral damage happened when S.B. was a 3-year-old child. Thus, one must allow the possibility of an unusual maturation of some parts of the brain as well as of compensatory behavioural adaptations to the primary deficits.

Clinical status
S.B., a 30-year-old right-handed man, suffered from a meningoencephalitis at the age of 3 years. Between the ages of 6 and 16 years, he received education in a centre for Young Partially Sighted. Then he followed a professional training until he was 20 years old. When S.B. arrived in the centre, he was totally unable to identify familiar objects by sight, although he identified them readily by touch. His integration there happened to be difficult because he was surprisingly good at sporting activities necessitating vision, to a point where some teachers did question his blindness.

S.B. was a very cooperative and intelligent young adult, trying hard to live a normal life. He refused to be considered a blind person. He could neither read nor write but was not illiterate since he read and wrote Braille fluently. Twenty-five years after the cerebral damage, S.B. retained a severe object, letter and face recognition deficit. He was able to describe object contours precisely, but usually could not reach an identification by vision. On the other hand, tactile identification of objects he had manipulated was immediate. Motion of objects helped visual identification. Recognition of objects was also easier when they were presented canonically and in an appropriate context. S.B. was totally unable to discriminate and name colours. On the other hand, he moved in space apparently without any difficulty. He could drive a motorcycle and was even able to practice some sports at a high level (for the disabled), such as goalkeeping, swimming and ping pong. To give a striking example of his surprising abilities, S.B. could easily catch two table tennis balls at the same time and juggle with them: yet he could not choose his food at the cafeteria.

Lesions
MRI scans (see Figs 1 and 2) performed in 1998 disclose lesions of occipitoparietal and occipitotemporal regions in the right hemisphere, and at the occipitotemporal junction in the left hemisphere. According to atlases from Talairach and Tournoux and Duvernoy (Talairach and Tournoux, 1988; Duvernoy, 1992), the lesions were distributed as detailed below.

In the right hemisphere, lesions involved most of the ventral visual areas, i.e. the posterior part of the inferior
temporal gyrus (T3), the inferior occipital gyrus (O3), the
fusiform gyrus (O4) and the inferior lingual gyrus (O5). In
functional terms, these lesions imply complete or partial
damage to the human counterparts of V2, V3, V4 and V5
(MT), as described in monkeys and delimited by functional
MRI (fMRI). As for the dorsal visual areas, large lesions are
observed that involve the middle occipital gyri (O2) and, to a
lesser extent, the superior occipital gyrus (O1), and in the
temporal lobe, the posterior part of the middle temporal gyrus
(T2) as well as a limited territory in the posterior part of the
Fig. 2 MRI axial contiguous slices (thickness = 1 mm) parallel to the bi-commissural plane (slice 0) in S.B. (left hemisphere shown on the right) showing the main lesions in this patient. These images confirm the quasi-isolation of the right occipital pole [including portions of the primary visual cortex (V1) and the posterior part of O4 and O5 (inferior aspects of V2)] from the visual output pathways, whereas the superior portions of the optic radiations are spared.
superior temporal gyrus (T1). Limited damage to the right inferior parietal lobule is also present in the region of the supra-marginal gyrus. The spared regions in the right occipital pole involve the polar parts of O4 and O5 (i.e. the posterior inferior part of V2), and the calcarine fissure (primary visual cortex, V1) at least in its rostral and superior aspects. Among the right dorsal regions, the greater part of the angular gyrus and a limited portion of the posterior part of T2 are spared.

In the left hemisphere, lesions are much more limited and concern mainly the ventral visual cortex, involving a complete destruction of the ventral part of the occipital–temporal junction (fusiform gyrus, O4). The lesion spreads further dorsally and partially involves the middle occipital gyrus (O2). In contrast to the right hemisphere, the parietal area (including V5 and its vicinity) is spared by the lesions.

In summary, in the right hemisphere, an isolated part of the primary visual cortex was spared, but reduced to a dead end. In the left hemisphere, the lesion disconnected the left spared ventral system anterior to the lesion from the occipital pole. Yet the connections between the occipital pole and the dorsal system were preserved.

Testing

Intelligence and memory

S.B.’s revised WAIS-R (Wechsler Adult Intelligence Scale) IQ tests were at an average level (verbal scaled score of 98). Memory was tested in the auditory modality only, by various means (Wechsler Memory Scale, verbal free recall and story recall), and showed average performances. S.B.’s working memory was not impaired, with a score of 6 on the digit span (Wechsler Memory Scale, test V), and his performance on the Brown–Peterson test (in the auditory modality) was globally correct with or without interference. S.B. had normal performances on the logical memory test of the Wechsler Memory Scale, test IV. He was able to learn, store and retrieve information from long-term memory (15 Rey’s words).

Gestural praxis

Testing of praxis was carried out in different stages. First we gave S.B. different tools to touch and manipulate, and we asked him to mime their use. Then S.B. had to recognize mimes performed by the experimenter. These mimes were of everyday actions and symbolic gestures. Finally, we asked S.B. to mime symbolic and concrete actions. The same tests were also performed by control subjects. A naive observer recognized the mimes done by S.B. S.B. knew how to use tools as long as he could touch them. For mimes depicting the use of objects, S.B. gave 10 out of 10 correct responses. He recognized 10 out of 10 concrete gestures (e.g. to cut with a knife, to brush one’s teeth, etc.) and four out of 10 symbolic gestures. He mimed nine out of 10 concrete gestures and four out of 10 symbolic gestures. Therefore, although S.B. was able to recognize and mime actions of everyday life, he was impaired at recognizing and miming symbolic actions. S.B. remarked that gestures without concrete meaning would not help sight-impaired persons.

Language

S.B.’s spontaneous speech was normal, without disorder of communication, articulation, prosody or phonological structure. S.B. was totally unable to read words or non-words. He could read only very few isolated graphemes in large, upper case and simple fonts. Yet, he read and wrote Braille fluently. His score on the Binois–Pichot vocabulary test, administered in the auditory modality, was 86, which would correspond to a verbal IQ of an average level. His verbal fluency was tested for animals, fruits, colours and nouns, beginning with the letter R or P. S.B.’s performances on this test were at an average level. On the whole, S.B. showed no language disorder, apart from a total alexia.

Visual fields

Perimetry (see Fig. 3) was carried out by means of the Goldmann technique using a static and dynamic presentation of LEDs (light-emitting diodes) of variable diameter and luminance. This revealed a left lateral homonymous hemianopia, sparing the macula. It is fair to add, however, that the perimetric examination was made difficult by S.B.’s inability to maintain a precise central fixation.

According to a line crossing test, there was no left side neglect despite the right parietal lesion (see Fig. 4).

Visual acuity

The common clinical tests to assess visual acuity could not be used with S.B. because of his near inability to read letters. The ‘E’ test was also useless since S.B. could not report consistently the orientation of the visual stimuli. Therefore, we designed a same/different test with pairs of letters or forms (n = 140 pairs). S.B. had to say whether the items were identical or different. The contrast (items versus background luminance) and the apparent angle of the stimuli were the same as those used in the standard ophthalmologic material, i.e. black stimuli on a white background, with a decreasing size, presented at a viewing distance of 5 m. Visual acuity was estimated as 4/10 without ocular correction. Eye refractometry revealed a hypermetropia, and S.B. is now wearing glasses which allow him to recover 6/10 in visual acuity.

Perception of targets defined by luminance

The task was to detect square grey patches (targets) of varying luminance presented on a grey background. S.B. was
able to discriminate quickly and point accurately to the targets even at a difference as low as 1 cd/m² (the limit of our experimental equipment), thus reaching an excellent performance.

**Contrast sensitivity**

Contrast sensitivity was tested with static and dynamic sinusoidal gratings (see Figs 5 and 6). Both examinations were again difficult to carry out because of the left hemianopia combined with S.B.’s difficulty in directing his gaze precisely. In the static condition, a threshold of minimum contrast necessary to perceive the grids was calculated as the mean value of five trials for each spatial frequency tested. This evaluation has been done twice, and a good reproducibility was observed. In Fig. 5, one can notice quite low values of sensitivity compared with controls, at all spatial frequencies tested, especially for high spatial frequencies. The very low values above 8 c.p.d. confirm S.B.’s poor acuity as previously tested. In the dynamic condition, the same kind of evaluation was performed for different temporal frequencies. Each threshold was the mean value of four trials. S.B.’s performances for low spatial frequencies (<3 c.p.d.) are close to those of controls for values of temporal

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**Fig. 3** S.B.’s visual perimetry. The visual field for the right eye is shown on the right and for the left eye on the left.

**Fig. 4** Visual cancellation test exhibiting an absence of hemineglect.
frequencies varying between 1 and 10 Hz. However, they become much lower for higher spatial frequencies.

**Line orientation**

Orientation perception was tested in a task of detection of an oddly oriented line among a matrix of lines, all with the same orientation. S.B. did not seem to perceive any odd stimulus. Nevertheless, S.B.’s ability to match the orientation of an open slot does not seem to be impaired (it may be argued that this difference is due to a crowding effect present in the first test; alternatively, it could be due to his using a covert visuomotor strategy; see later).

**Brightness judgements**

S.B.’s perception of brightness was difficult to characterize. Indeed, S.B. would often call as brighter the darker stimulus, or the reverse. He apparently did not understand the meanings of the words darkness or brightness, just as he could not grasp the meaning of red and green. He was not really able to explain how he perceived brightness. It seemed that S.B. actually confounded brightness with contrast since he would call a black disc on a grey background bright. We made several attempts to explain the meaning of dark (‘dark is when the sun sets and the night is falling’) and bright (‘by a window on a sunny day . . .’), but that did not improve his evaluation. However, S.B. was able to point to the brighter card when only two grey cards were presented side by side (hit rate = 35 out of 39), though he failed when four cards with quite different brightnesses were presented (five out of 10). The classification of grey cards from the brightest to the darkest was near chance, a striking failure considering his remarkable capacity to detect small luminance differences (see Fig. 7).

**Colour perception**

S.B. was achromatopsic: he could not discriminate, order or name colours. Presented with so-called ‘Mondrian’ displays, he could not say if they were coloured or black and white. Moreover, he could not detect a shift from colour to black and white on a screen. He was totally unable to pass the Farnsworth-Munsell 16-Hue test (display with large cards;...
Farnsworth, 1947). Neither was he able to respond to the Goldstein–Scheerer test or the Ishihara test (Ishihara, 1974; version for illiterate observers). However, S.B. succeeded in pointing out a red square among 15 equiluminant green squares (nine out of 10) when the colours were quite saturated, but failed for low saturations. However, he commented: ‘red does not mean anything to me, but I simply see it better...’. In other words, S.B. had no idea about the subjective appearance (the quale) of a colour. This kind of performance (detection possible versus discrimination impossible), observed in some achromatopsic patients, may be attributed to border detection. Indeed, these results fit well with what is known of colour coding in the dorsal stream, i.e. whereas MT neurones in the macaque monkey are unselective for any particular wavelength, many of them continue nevertheless to respond to the orientation of a wavelength defined by a boundary, even at equiluminance (Saito et al., 1989).

**Texture perception**

Images containing an area differing in texture were presented to S.B. He had to report whether there was an area that was different. S.B. was not able to discriminate between two parts of the same surface with different textures, even when the boundary between both textures was shown to him.

**Depth perception**

A series of stereograms were presented as anaglyphs for viewing through red/green filters. In the Lang test, S.B. had to localize objects on a piece of paper by pointing. He performed very well although he was not able to identify them. In the Wirt test, eight diamonds are composed of four circles among which one has a stereoscopic depth (the depth varying between the different diamonds). S.B.’s performance was perfect. On the whole, S.B. does not have any deficit in stereoscopic depth at the tested distance.

**Object recognition**

Standard sets of drawings of common objects [French `Imagier du Père Castor’ and the Birmingham Object Recognition Battery (BORB)] could not be identified at all. Nevertheless attempts to copy drawings produced quite good copies. In the case of the complex Rey’s drawing, the copy

![Fig. 7 Ordering of grey cards by luminance. In this test, S.B. had to order seven grey cards of various luminance, from the lightest to the darkest. The figure shows the comparison between the photometer ordering and S.B.’s setting: he made two striking errors.](image)

![Fig. 8 The original Rey’s figure (left) and S.B.’s copy (right). The order of the colours used was as follows: black, red, blue, green, maroon and purple.](image)
done by S.B. is rather good (score of 26) despite his perceptual deficits, though the time taken was very long. S.B. copied the figure feature by feature, with quite an accurate coordination between these details (see Fig. 8).

**Real objects**

A severe disorder of real object recognition was confirmed, with several errors being made with everyday items (e.g. a packet of cigarettes misidentified as a pack of tissues). S.B. was able to make intelligent guesses on the basis of specific features of the objects, although this process was time consuming and required a lot of effort. The identification of real objects evaluated in a 45 trial test (35 successes out of 45) was much better than identification of photographs of these objects (eight out of 45). S.B. managed sometimes to distinguish a manufactured item from a living object. He seemed to base his judgement upon physical characteristics such as legs, tails, etc. In all cases, the objects were readily identified tactually.

**Face recognition**

S.B. demonstrates a severe prosopagnosia and usually uses non-visual cues to recognize people, such as silhouette, gait and voice. With pictures, he was not able to discriminate well between human faces and non-face stimuli. It was only after a certain time that S.B. found out that the stimulus had parts representing the eyes, the mouth, hair, etc. S.B. is also unable to recognize facial expressions; nor can he distinguish famous faces from unfamiliar faces. On a decision task, different items were presented: normal faces, scrambled faces and objects. In the beginning, S.B. did not recognize the real faces at all. However, after several presentations, S.B. picked out a detail (such as the eyes) and adopted a new strategy of exploration of the items. Finally he succeeded in detecting real faces among other items. Using the same strategy, he was quite good in a same/different task, where he had to compare two faces. He had to give his responses within 2 s and had a score of 67 out of 94. In other words, S.B. was good at comparing details (ears, hair . . .) of two faces and at deducing whether they were identical or not.

**Protocole d'évaluation des gnosies visuelles (PEGV)**

A series of batteries was used to assess S.B.'s shape perception. The PEGV (Visual Gnosia Evaluation Protocol; Agniel et al., 1987) comprises four tests: two tests involving visual discrimination (one of identical figures and one of overlapping shapes) and two tests involving semantic processes (one of functional matching and one of categorical matching). The matching tests could not be done since S.B. was unable to recognize the drawings of the objects. On the other two tests, S.B.’s performances were characterized by a slowness that put him outside the normal range. Nevertheless, S.B.’s performances on the test of identical figures were above chance (eight out of 10), while the results for the test of overlapping figures were at chance level (five out of 10). For the test of identical figures, S.B. used a feature-by-feature analysis of the items presented. On the other hand, in the test of overlapping figures designed to assess figure–ground segmentation, S.B. was unable perceptually to segment one item from the others.

**BORB**

The BORB is composed of 14 tests designed to assess particular aspects of visual processing and visual object recognition. Some of the battery subtests were not given because of their difficulty, while others were modified slightly to make them feasible for S.B. Some of the subtests involving pre-categorical visual processing are designed to test the perception of basic characteristics of objects’ forms (including size, orientation, location and length). These tests include both copying and matching tasks. In the test of copying elementary shapes, S.B. performed better on copying simple two-dimensional items (triangle, square . . .) than three-dimensional items (cube, table . . .). In addition, the copy of overlapping figures requiring some degree of figure–ground segmentation was slightly impaired. The perceptual matching tests included matching of line length, of stimulus size, of line orientation and of the positions of gaps in two circles. S.B.’s performances on these tests were 23 out of 30 (77%), 27 out of 30 (90%), 18 out of 30 (60%) and 35 out of 40 (87.5%), respectively. These results show that S.B. was able to perform basic perceptual analysis. Effective object perception requires the ability to judge that an object is the same even when it is seen from different viewpoints. This means that the subject must achieve object constancy. This ability was assessed with BORB by means of two subtests: the ‘minimal feature view’, presenting the object in an unusual view which hides its main features, and the ‘foreshortened view’ where the main identifying features of the object are kept, while its overall shape is distorted by the foreshortening. S.B.’s performances were 18 out of 25 for the ‘minimal feature view’ task and 17 out of 25 for the ‘foreshortened view’ (matching tasks). The minimal feature view relies on overall shape perception while the foreshortened view relies on main feature perception. S.B.’s surprisingly good performances in the absence of main features show that, at least for these particular stimuli, he was able to use some global strategy.

**Mooney figures**

Mooney figures were poorly recognized. S.B. was not able to recognize a bottle, a spoon, a cup, a foot, a rabbit, a comb and
a truck, but he recognized a car by the wheels. When we revealed what the object was, S.B. could sometimes name some details of the item (e.g. the car’s windscreen, the cup’s handle . . .). He was disturbed by the frame around some figures and by the background, which he tried to analyse as being part of the object.

Visual memory

Benton’s visual retention test (drawing version)
The test was administered with a free time allowance that gave S.B. >10 s to memorize the drawings. S.B. was able to reproduce from memory six drawings out of 10. He succeeded in reproducing some drawings that he had seen before for <10 s.

Rey’s figure
S.B. used a slavish strategy, copying step by step and taking much more time than a normal subject. He ended up with a score of 26 (inferior to the 10th centile). More surprising was the reproduction from memory, which required 13 min of drawing. Indeed, S.B. ended up with a score of 24 (60th centile), i.e. above the adult average, attesting to his good spatial memory (see Fig. 9).

Visual imagery
When asked to compare mentally the size of different objects and animals (e.g. a violin and a guitar, a dog and a cat), S.B. performed well (39 out of 40). Another test (see Mehta et al., 1992) consisted of mentally comparing a series of three items (objects or animals) and of finding the two items that were similar (i) according to their global contour for the objects (e.g. a hammer, an axe and a saw) and (ii) according to their head for the animals (e.g. a horse, a cow and an elephant). S.B. performed well for the objects (11 out of 13) but was impaired for the animals (five out of 12). S.B.’s performances for describing objects from memory was relatively good as well. For example, he defined a hammer as a wooden shaft with a triangular piece of iron at one end; an umbrella as a shaft with cloth fixed to the ribs on the top of it.

Drawing from memory (see Fig. 10) was performed with 10 items that S.B. knew from their encyclopaedic definition (e.g. cloud, snail, cactus) and 10 items that he knew from their manipulation (e.g. fork, ashtray, hammer). The drawings were fair for objects which he knew from manipulation (compare glasses and cactus).

S.B. knows most alphabet letters in their upper case form and can write them down. S.B. performed well at a test requiring him to state from memory whether upper case (24 out of 26) and lower case (25 out of 26) letters contained any curved lines. To test his visual imagery on this restricted repertoire, we also designed a test consisting of mentally modifying letters via global rotation, feature deletion or addition, as well as changes of feature orientation. S.B. had to identify letters resulting from these mental modifications (e.g. for the letter ‘V’ we proposed: ‘what is the letter if you put it upside down and add an horizontal bar in the middle ?’ S.B. readily gave the correct response ‘A’). Performance was quick and accurate for the seven letters used in this test.

Executive functions
Since S.B. is achromatopsic, the Wisconsin Card Sorting Test was administered in a non-conventional way. The sorting rule regarding the colour of the cards was disregarded. Despite his recognition deficit, S.B. could find out the rule concerning the form and the number, again exhibiting strikingly good performances based on processing a limited number of graphic features (cf. Benton’s Visual Retention Test).
Motion processing
Opto-kinetic nystagmus was easily obtained with a chequerboard that was moved in front of his eyes.

S.B. reported no difficulty at all in estimating the speed of walking persons or moving cars. When required to follow visually an examiner walking down a street and evaluate verbally his/her distance upon stopping, S.B.’s performance was strikingly accurate, without practice, from a few metres to hundreds of metres.

A test was performed with numerous coherently moving dots that were displayed via a video projector, on a screen as large as ~50°. S.B. had to report the direction of the movement: left or right, up or down, forward or backward. He decided rapidly and accurately, until the coherence of the dots was reduced to 35% (a performance similar to the performances of several healthy observers during the session).

A motion discrimination experiment using drifting gratings was also conducted, using a 21 inch LCD computer screen. Sine wave gratings (black and white bars) were presented in peripheral vision either in the normal or the ‘blind’ visual hemifield on a white background. S.B. was placed 2 m away from the screen and had to fixate a cross while discriminating the direction (either to the left or to the right) of the moving gratings. We used two different velocities for the movement of the gratings (slow and fast). S.B.’s performances for stimulation in the normal field were 100% correct, while his responses for stimulations in the blind field ranged between 45 and 80% (see Table 1).

The results show that S.B. is better for low spatial frequencies than for high spatial frequencies and when the gratings were moving towards the left (away from the fixation cross). We did not observe any significant difference regarding the velocity of the drifting gratings. It seemed, therefore, that S.B. could perceive movement in his blind visual hemifield, but this point requires a more comprehensive investigation since, again, it was not certain that he fixated the centre of the screen correctly.

On the other hand, S.B. had a clear abnormality in the perception of apparent motion. Since S.B. was hemianopic, all the stimuli were presented parafoveally in his intact right visual hemifield. The test consisted of two or four illuminated discs that gave the impression of appearing and disappearing simultaneously (flicker) or moving from one site to the other (phi phenomenon). S.B. could not decide whether it was a flicker or a movement.

S.B. was also tested with the presentation, on a 50° wide screen, of dynamic light points defining moving people (Johansson, 1973). For ‘a man walking’ or ‘two persons throwing a ball to each other’ displays, both immediately recognized by control subjects, S.B. needed >1 min to name them eventually, after several misidentifications.

Table 1 S.B.’s correct responses (out of 20) when drifting gratings were presented in his ‘blind’ visual field

<table>
<thead>
<tr>
<th>Leftward (out of 20)</th>
<th>Rightward (out of 20)</th>
</tr>
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<tbody>
<tr>
<td>Fast Slow</td>
<td>Slow Fast</td>
</tr>
<tr>
<td>At low spatial frequency (SF = 0.5 c.p.d.)</td>
<td>16 14 10 10</td>
</tr>
<tr>
<td>At high spatial frequency (SF = 1 c.p.d.)</td>
<td>11 11 9 11</td>
</tr>
</tbody>
</table>

Fig. 11 S.B. immediate grasping. S.B.’s ability to grasp (his maximum grip aperture is reported on the y-axis) rectangular shapes of different widths (indicated on the x-axis). These data show that S.B. is able to adjust his finger–thumb grasp to object size during natural grasping movements with a precision well within the normal range.
Identification of motion-defined shapes
Learned forms (a motorcycle, a dog, a bike, a camel) were presented either as still black silhouettes or moving silhouettes all defined by coherently moving dots against a stable background. S.B. could not identify these forms despite an immediately prior teaching.

Vision for action
Although S.B. has difficulties in perceiving and discriminating shapes, he is far from being impaired in visuomotor control. For example, he performed as well as controls when he had to orient his wrist while reaching to pass his hand through a slot placed at different orientations (like patient D.F. in Milner et al., 1991).

In formal testing (see Fig. 11), we measured S.B.'s ability to grasp rectangular shapes of different widths (see Goodale et al., 1991). The six shapes used were all 1 cm thick and of 25 cm² surface area, with widths varying between 2.5 and 5.0 cm in steps of 0.5 cm. They were presented singly at a distance of 25 cm from a starting point located at 12 cm from the near edge of the table, both target and starting point placed in alignment with the subject’s mid-sagittal axis. The objects were presented in a lateral orientation for S.B. to reach out and grasp them width-wise (i.e. front-to-back).

The positions of the index finger and thumb were sampled using a Minibird magnetic movement recording system (Ascension Technology Ltd) at rates of 86.1 and 103 Hz. These data allowed us to compute the MGA (maximum grip aperture) during each reach, and this measure served as the dependent variable. Analysis of variance revealed a highly significant main effect of object width \[ F(5,36) = 9.86, P < 0.0001 \]. The apparent high degree of visual grip scaling (Fig. 11) is confirmed by a Pearson correlation between width and MGA of \(+0.72 (P < 0.001)\). These data show that S.B. is able to adjust his finger–thumb grasp to object size during natural grasping movements with a precision well within the normal range, just like the visual form agnostic D.F. (Goodale et al., 1991).

Spatial visual attention
Attentional pursuit
Attentional pursuit was tested with stimuli containing several black discs randomly moving in a square. Some of the discs (one or more) become white for a few seconds before returning back to their original colour. The task of the subject is to track the discs that became white and point to them on a final still display after they have recovered their original colour. The tracking of several discs at the same time cannot be but attentional. S.B. was completely unable to do the task with two target discs. With only one target disc among three distracter discs, his performances were slightly above chance, while controls would show near perfect performances. Strangely enough, S.B. reported that the change of ‘colour’ (black to white and vice versa) did not ‘pop out’.

Subitizing
A square of \(\sim 10°\) filled with 100 small discs (10 \(\times\) 10) was presented on a computer screen for 5 s, then replaced for 300 or 700 ms by one to five discs of the same size (0.5°), then replaced by the original square of 100 discs. The presentation was directed to the good visual field. The task was to count the number of discs presented during the short intermediary presentation. A normal subject shows ceiling performances. S.B. showed abnormally poor performances (see Fig. 12).

Gestalt perception
Although S.B. used a feature-by-feature visual recognition strategy, he did not seem to be able to perceive independently local forms embedded into a global form. S.B. did not detect illusory contours (Kanisza figures) and failed to perceive
geometrical illusions (Müller–Lyer, Titchener, Ponzo figures). For hierarchical stimuli (Navon, 1977), S.B. easily recognized large letters and forms (the ones he knew) whereas he was at chance level for the small letters and forms. Evidently, when global and local processing compete with one another, a deficit in the perception of high spatial frequencies is revealed whilst the perception of low spatial frequencies is less impaired.

Corsi block test
S.B.’s performances for this test were within the normal range (spatial span of 5 ± 1).

Topographic imagery
S.B. had no difficulty with topographic imagery. Living on his own, S.B. had mental references like those of a blind person. For example, he knew where common objects (e.g. kettle, coffee . . .) are kept in his home. S.B. answered accurately questions about the topography of the hospital (e.g. where is the cafeteria with respect to the laboratory?).

Discussion
To our knowledge, the case of S.B. is unique in the neuropsychological literature. This very singular patient has learned to see and act in the surrounding world with a visual system restricted to one dorsal stream. Observing S.B. in his daily life is a surprising experience since one is confronted by a person who alternately seems blind or visually normal, depending on the behaviour required. This contradictory behaviour naturally raises the question: does S.B.’s case represent a case of visual agnosia, and, if so, which type of agnosia?

Certainly many of his deficits (achromatopsia, prosopagnosia, alexia and object agnosia) resemble those typically presented by visual agnostic patients.

Like many visual agnics, S.B. focused on parts of objects rather than on objects themselves, whatever the objects, including faces. However, he performed quite well on matching meaningful and meaningless objects as well as copying drawings, relying on a feature-by-feature analysis of the objects, which proved both time consuming and often unsuccessful. Being quite often unable to identify the whole object, he would nevertheless be able to assign properties or functions to its isolated parts. Unlike some patients classified as apperceptive agnosics or visual form agnosics (Benson and Greenberg, 1969; Campion, 1987), S.B. could process local aspects of shape but failed to integrate and link them to global aspects. A dissociation between the two stages or processes would result in a feature integration deficit. The consequence of this impairment leads to an inadequate segmentation of the viewed stimuli, an impairment in integrating local visual shape information into perceptual wholes. Such a failure has
been labelled as ‘integrative agnosia’ (Riddoch and Humphreys, 1987). Patients showing this type of agnosia are therefore unable to perform tasks requiring the integration of local features, although they remain able to do simple physical matching and copying of (unrecognized) objects through a feature-by-feature visual strategy.

S.B. shows some similarities to H.J.A. (see Table 2 for a comparison between the two cases), who has bilateral damage to the occipitotemporal regions of the cortex (Riddoch and Humphreys, 1987).

Both patients show difficulties in reading, recognizing colours and faces, and both use a feature-by-feature visual strategy in matching and copying tasks, and focus on details to identify objects. In both patients, bilateral damage to the dorsal stream leads to a limitation in the parallel grouping of visual forms, impairing object recognition. Therefore, as well as other cases reported in the literature, these patients attempt to name objects according to a serial identification of their parts (Grailet et al., 1990; Butter and Trobe, 1994; Shelton et al., 1994; Humphreys, 1999).

S.B. also shows some similarities with patient D.F., whose case has been studied extensively (Goodale et al., 1991; Milner et al., 1991; Goodale and Milner, 1992; Goodale et al., 1994). D.F. suffered from a severe recognition deficit involving especially discrimination of shapes and orientation. Yet this patient could use this visual information to orient and shape hand movements during reaching and grasping. MRI showed that the effective cortical damage disrupted large parts of V2, V3 and V4, leaving V1 largely intact. Milner and Goodale suggest that D.F.’s brain suffered a disconnection such that visual form information could no longer reach the ventral stream, and could only be processed within the dorsal stream. As a result, she became unable to report on the shape or orientation of objects, while still remaining able to act upon them using that same shape and orientation information.

S.B.’s cerebral damage is less selective than that of D.F., since the ventral system is not more or less damaged, but simply absent, and that since the early years of life. Thus, even though both patients seem to depend largely (D.F.) or exclusively (S.B.) on the dorsal system, they clearly differ in several aspects. Contrast sensitivity curves are the opposite, D.F. perceiving high spatial frequencies well while S.B. only sees low frequencies: hence D.F. sees fine textures that S.B. ignores. D.F. (like other patients with visual form agnosia) shows remarkably well-preserved colour discrimination, while S.B. is totally achromatic. These contrasted capabilities are easily explained: D.F. can make some use of her ventral system while S.B. is limited entirely to one dorsal system. It is well known that the primary input to the dorsal stream arises from the geniculocortical magnocellular channel, which carries little or no information about colour or fine texture. These qualities depend on the parvocellular channel, particularly on the ‘parvo-blob’ subdivision, which projects entirely to the ventral stream, and which is thought to have remained relatively intact in D.F., despite her loss of form information processing in the ventral stream.

Visual agnosia is a deficit acquired by a previously normal person, while S.B. has suffered from an extensive cortical damage very early in life. He has learned to compensate for the apparently complete absence of the ventral system, while adult agnosics usually retain some use of their ventral systems. Therefore, it seems hazardous to classify S.B. as a special case of visual agnosia. S.B. has not lost an acquired visual competence but rather he has learned to see with very impoverished cerebral visual structures. S.B. represents a unique case which must be described on its own, without an attempt to locate it in a comprehensive classification of visual agnosias.

S.B. offers the quite unusual opportunity of evaluating what it is to see without a ventral system, or more precisely to see through a dorsal system on the basis of magnocellular information alone.

The first most striking observation is that S.B. can easily move inside and outside, even with poor lighting, even in non-familiar environments, mastering landmarks to direct his approach of a goal. His facility to use movement information is also striking.

The second interesting feature of S.B.’s case is his capability to deal with the world of moving objects. It is surprising to see this quasi-blind patient directing both hands at two table tennis balls at the same time and catching them easily. His egocentric space seems to be very well preserved, at least compared with his allocentric space.

Another very peculiar feature is S.B.’s apprehension of the grey world in which he lives. S.B. is bothered by high luminance levels; he prefers to move within a low luminance world (dawn, night), somewhat as a nocturnal ‘animal’, presenting a sort of ‘night sight’, or nyctalopia. His difficulty in appreciating luminance differences subjectively, those he uses so remarkably when moving and acting, suggests that the parvo system is necessary to interpret and name the qualia of the greys. We may propose the label ‘aphotopsia’ as a mirror deficit of achromatopsia, but this time for achromatic features. Interestingly, a less severe version of this difficulty in perceiving different shades of grey was also found at an early stage of D.F.’s disorder (Milner and Heywood, 1989).

S.B. showed surprising capacities of mental imagery, being able to draw objects quite well. Moreover, as demonstrated by the letter rotation test, S.B. was able to generate and manipulate spatially stored representations that are essentially visual in normal subjects.

One of the major issues in literature about mental imagery and perception is to know whether perception and imagery share common mechanisms relying upon common neural substrates. Half of the studies have described cases of patients showing preserved perception with impaired imagery, and vice versa. Several observations from agnostic patients (Riddoch and Humphreys, 1987; Behrmann et al., 1992; Jankowiak et al., 1992; Servos et al., 1993; Young et al., 1994; Servos and Goodale, 1995; Bartolomeo et al., 1998; Riddoch et al., 1999) suggest that visual perceptual processes and visual imagery processes may be independent. On the
other hand, unlike the patients cited above, the patient H.G., reported by Grailet et al. (1990) showed poor performances in visual imagery.

That early visual areas are not essential for retrieval from memory has also been illustrated by reports of patients with cortical blindness who were able to form visual images (Chatterjee and Southwood, 1995; Goldenberg et al., 1995). Results from investigations in normal subjects using fMRI have shown that, contrary to earlier reports (Farah et al., 1988; Goldenberg et al., 1989; Kosslyn et al., 1993), activation in primary visual areas is not systematically observed in mental imagery tasks, depending on the nature of the processes elicited by these tasks (Kreiman et al., 2000; Mellet et al., 2000; Trojano et al., 2000).

In spite of profound and early installed visual perceptive deficits precluding building up of a normal store of mental images, S.B. has preserved some visual imagery processes, like other patients with visual agnosia (Riddoch and Humphreys, 1987; Servos and Goodale, 1995; Bartolomeo et al., 1998; Humphreys, 1999). His imagery resources, at least partially spared, have been built up by the occipitoparietal system, on the basis of spatial and motor references (Servos and Goodale, 1995; Trojano et al., 2000).

Conclusions

Patient S.B. presents a completely unique case: he was deprived of the right dorsal system and of both occipitotemporal visual systems as the result of an encephalitic episode at the age of 3. Therefore, S.B. has learned to see the world (except for the first 3 years of his life) with one dorsal system only. What was learned before the age of 3 is impossible to know. Nevertheless, he has no idea of what a colour is and he can hardly draw a face. Indeed, S.B. cannot identify colours, shapes, drawings, objects, letters or faces in a natural environment. Yet he can find his way, move easily in familiar environments, reach for objects, use them in an appropriate way, play fast ball games and even ride a motorcycle. In other words, he appears at different times both as a normal and as a blind person.

However, S.B. demonstrates some perceptual abilities, allowing him to identify certain familiar objects and to reproduce simple drawings, through mentally piecing together their elementary features. For these basic perceptual abilities, he has to rely only on luminance or motion contrasts, since texture and colour contrasts are unavailable to him. Such residual but significant visual competences obviously must depend on the spared portion of his visual system, i.e. the left dorsal, occipitoparietal pathway. His ability for elementary perception therefore supports the view that the magnocellular-dorsal stream pathway can suffice for basic ‘what’ processing. We believe that, as in the case of Patient D.F., dorsal stream mechanisms for analysing elementary features have remained intact, at least in one hemisphere. The difference is that unlike D.F., S.B. has direct perceptual access to these features, which D.F. can only respond to in an automatic ‘visuomotor’ fashion.

This interpretation is consistent with current ideas proposing that the dorsal stream is involved in some kind of elementary form analysis up to the level required for action (Jeannerod et al., 1995; Milner and Goodale, 1995; Faillenot et al., 1997; Kraut et al., 1997). However, in our patient, the dorsal visual pathway may be recruited for perception as well as for the visual control of skilled actions. Unlike most of the patients described in the literature, S.B. acquired his agnosia in his early childhood. Some cases of visual agnosia (e.g. Wilson and Davidoff, 1993) have recovered some partial visual abilities, especially for the recognition of real objects. Even in D.F., who suffered her brain damage during adulthood, some improvement in performance on perceptual tasks has been observed in recent years. This may be due to her use of self-cuing strategies (Dijkerman and Milner, 1997), by which she seems to be able to ‘short-circuit’ her visuomotor system internally without the need to perform overt actions. S.B. presumably can do this even more effectively, after a lifetime’s opportunity to acquire such strategies; however, in his case, neurological plasticity during childhood seems somehow to have rendered such self-cuing available to conscious experience.

This neurological compensation may have grown hand-in-hand with S.B.’s well-developed haptic skills. For example, Sadato et al. (1996) demonstrated in Braille readers, blinded in early life, that the primary and associative visual cortices can receive information coming from other sensory modalities. In their subjects, tactile discrimination tasks, such as Braille reading, induced activation in the primary visual cortex. In S.B.’s case, it is possible that the tactilo-kinaesthesic information, so important for blind people, has developed privileged connections with the dorsal visual system. This sensory motor information could have played a decisive role in S.B.’s surprising capacities to draw objects, especially those he knows by touch.

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