Prosopagnosia

Eugene Mayer1 and Bruno Rossion2
1University Hospital of Geneva, Switzerland
2University of Louvain, Belgium

General Presentation of the Disorder

**Key points** Main characteristics of prosopagnosia

- Inability to recognize familiar person on the basis of visual perception, despite the absence of low level visual impairments or cognitive alterations
- Relative sparing of person recognition through other cues (essentially voice, but also gait, size, clothes, or facial features or accessories)
- Frequently associated with left hemianopia, achromatopsia and topographical disorientation
- Due to right or bilateral lesion in the inferomedial part of the temporo-occipital region (fusiform and lingual gyri)

Prosopagnosia is classically defined as an inability to recognize faces of people known to the patient on the basis of visual perception, despite the absence of low-level visual impairments, or cognitive alterations such as mental confusion or amnesia, with a preserved ability to recognize people through other cues: voice or other visual traits such as gait, size, clothes, or even facial features (moustache, scar, blemish) or accessories (ear-rings, eyeglasses). Prosopagnosics have also access to semantic knowledge concerning people.

According to Grüsser and Landis (1991), this condition seems to have been first described by Wigan (1844), in a book in which he expressed his views on the interaction of the two cerebral hemispheres. Wigan stated (pp. 128–9):

A gentleman of middle age, or a little past that period, lamented to me his utter inability to remember faces. He would converse with a person for an hour, but after an interval of a day could not recognise him again. Even friends, with whom he had been engaged in business transactions, he was unconscious of ever having seen (…) it was not till he heard the voice, that he could recognise men with whom he had constant intercourse (…) When I inquire more fully into the matter, I found that there was no defect in vision, except that his eyes were weak, and that any long continued employment of them gave him pain (…). He was quite determined to conceal it, if possible, and it was impossible to convince him that it did not depend solely on the eyes.
Early descriptions of prosopagnosia were also provided by Quaglino and Borelli (1867; a patient with right hemisphere stroke), Charcot (1883), Wilbrand (1887, 1892; stroke with bilateral posterior lesions). Frequently, prosopagnosia was found to be associated with other deficits such as achromatopsia and topographical disorientation (Quaglino and Borelli, 1867; Wilbrand, 1887, 1892), or even color and object agnosia (Charcot, 1883). A number of similar observations were published in the next 60 years (see Grüsser and Landis, 1991, for an historical review). Nevertheless, disturbance in face recognition was considered to be part of a complex visual agnosic disorder until the paper of Hoff and Pötzl (1937). These authors considered for the first time the inability to identify familiar faces as a specific form of visual agnosia. Ten years later, Bodamer (1947) isolated the failure to recognize familiar persons on the basis of facial appearance from other disorder of visual perception and coined the term prosopagnosia.

Patients with prosopagnosia are generally able to make fine visual discriminations. For example, they are able to read. They can also categorize a visual stimulus as a face. But prevented to use the voice, clothes, gait or specific attributes such as glasses, ear-rings or moustaches, they cannot identify a familiar person, and even themselves in some cases. For example, Charcot’s patient (1883) mistook his own mirror image for a stranger and apologized for blocking the way. Years ago, we filmed a woman who was prosopagnostic and we projected her own image to her on-line, without sound. We asked her to describe what she saw. She told us: “I see a woman, . . . she is speaking, . . . she is wearing a scarf.” Asked about the identity of this woman, the patient responded “it could be one of my neighbours.” This patient denied having any specific problem with face recognition even if she admitted not being physionomist. Nevertheless, absence of awareness of the disorder is not the rule. Other patients sometimes report that they do not recognize familiar people until they talk and base their visual recognition on clothes or hairstyle for instance.

The simplest and still the best way to determine clinically whether a patient is prosopagnostic or not is to confront him with several persons, of the same sex and age, uniformly dressed, among which one is familiar. Ideally, the hair should be masked with a cap. The persons should remain completely silent and move as little as possible, without smiling. In this condition, a prosopagnostic will not recognize the familiar person. If this familiar person is immediately identified when she begins to speak, the diagnosis is made. Even though low-level visual impairments are likely to be present due to the extension of the lesions to early visual areas, the deficit must not be explainable by alterations of lower-level visual impairments, hence the importance of detailed assessment of the patient’s neuropsychological profile (see later).
A number of reviews of the anatomical basis and functional aspects of prosopagnosia and other face disorders have been published over the years (e.g. Barton, 2003; Behrmann and Moscovitch, 2001; Goldsmith and Liu, 2001; Grüsser and Landis, 1991). In line with the objectives of this volume, we have intended to provide a distinct account of this spectacular deficit, first by focusing on the assessment and rehabilitation of prosopagnosia, and second to highlight certain aspects of the deficit that are recent and/or relatively unknown. In the next section, the relationship between the localization of the lesions causing prosopagnosia and the neural basis of face processing in the normal brain will be discussed. Then we discuss recent findings illustrating how prosopagnosic patients can still process faces to some extent, something that may be critical to define for any attempt of rehabilitation of this deficit.

**Etiology and Lesion Localization**

**Key points** Strokes responsible for the disorder

<table>
<thead>
<tr>
<th>Stroke type</th>
<th>Territory/characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arterial Infarct</td>
<td>posterior cerebral artery: right or bilateral</td>
</tr>
<tr>
<td>Hemorrhage</td>
<td>infero-medial part of the temporo-occipital area</td>
</tr>
</tbody>
</table>

Prosopagnosia is extremely rare. For example, Zihl and Von Cramon (1986) did not find any pure prosopagnosics in a series of 258 patients suffering from posterior brain lesions of different origins (closed head injury, encephalitis, tumors, surgical lesions for epilepsy, hemorrhages and cerebral infarcts). If prosopagnosia is considered as one symptom among other visual and neuropsychological defects, the frequency is higher — for example 6% in the study of Hécaen and Angelergues (1962, 1963) concerning 382 patients with posterior cerebral lesions of different origins. **Prosopagnosia of vascular origin is due to posterior cerebral artery infarcts (PCAI).** This latter disease represents 5–11% of all strokes in the general population (Brandt et al., 2000). Among PCAI, prosopagnosia can be present in pure cortical infarct (4%), cortical and deep infarct (4%) and bilateral infarcts (7%) (Kumral et al., 2004). **If unilateral, the infarct must be on the right side in order to produce prosopagnosia** (Cals et al., 2002; De Renzi et al., 1994; Grüsser and Landis, 1991; Milandre et al., 1994; Uttner et al., 2002; Wada and Yamamoto, 2002). **Other etiologies include a carbon monoxide poisoning, temporal lobectomy, encephalitis, neoplasm, right temporal lobe atrophy, trauma, Parkinson’s disease, and Alzheimer’s disease.** The debate about the localization of lesions has for long been restricted to the question of whether a right hemisphere
Prosopagnosia

lesion was sufficient and/or necessary, or not, to cause the deficit (e.g. see Damasio et al., 1982; Michel et al., 1989). It is now widely admitted that a lesion of the right hemisphere is necessary to cause prosopagnosia, even though one case has been described with a lesion apparently restricted to the left hemisphere (see Mattson et al., 2000). The fact that there is only a single case of prosopagnosia following a left hemisphere lesion is remarkable, given the contribution of the left hemisphere during normal face processing, as evidenced by lateralized visual field studies (e.g. Hillger and Koenig, 1991), and more recently by neuroimaging (e.g. Sergent et al., 1992; Kanwisher et al., 1997) and event-related potential (e.g. Rossion et al., 2003a) studies. Face processes performed in the left hemisphere thus appear to complement a massively right localized function, and may not be critical for this function. Alternatively, a lesion in the left occipitotemporal cortex may cause only subtle face processing deficits that are usually not detected by conventional neuropsychological tests, perhaps because left unilateral brain-damaged patients present other, more spectacular, symptoms (e.g. alexia; see Farah, 1990).

However, the notion that a single right hemisphere lesion is sufficient to cause prosopagnosia, or whether a bilateral lesion was always necessary, has proved more difficult to resolve. This question originated from the analysis of visual field defects in prosopagnosic patients (Hécaen and Anguerlergues, 1962; Meadows, 1974). The presence of a left visual field defect, in particular a left superior quadrantanopia, is the most common low-level visual defect observed in cases of prosopagnosia, but there are also prosopagnosic patients that have exclusive right visual field defects (Bouvier and Engel, 2005; Meadows, 1974; Goldsmith and Liu, 2001). In any case, a left visual field defect associated more often with prosopagnosia cannot be used as a reliable criterion to support the view that a right hemisphere lesion is sufficient to cause the deficit, since cases of prosopagnosia would not present visual field defects if posterior cerebral lesions spare the optic radiations and striate cortex (Damasio et al., 1982).

With the advent of structural imaging (CT scans, and then MRI; see Damasio and Damasio, 1989), it was clearly established that a right hemisphere lesion can be sufficient to cause prosopagnosia and several such cases have been reported (three cases in Barton et al., 2002; Landis et al., 1988; Marotta et al., 2001: case 2; Sergent and Signoret, 1992: cases 1 & 2; Sergent and Villemure, 1989; Uttner et al., 2002; Wada and Yamamoto, 2002), most of them following right posterior cerebral artery infarct. Furthermore, functional neuroimaging examination (positrion emission tomography, PET; or functional magnetic resonance imaging, fMRI) performed in some of these cases indicated a normal metabolism in the left hemisphere (see Sergent and Signoret, 1992).
The lesions causing prosopagnosia are generally located in the inferomedial part of the temporo-occipital area, in the fusiform gyrus, the lingual gyrus and the posterior part of the parahippocampic gyrus (Damasio et al., 1982; Grüsser and Landis, 1991). Within this large cortical territory, PET and mainly fMRI studies conducted on the normal brain have identified at least two small (a few mm$^3$) areas responding preferentially to faces, as compared to other object categories, or scrambled face stimuli. These areas are localized in the inferior occipital gyrus (IOG) and in the lateral part of the middle fusiform gyrus (MFG), (e.g. Kanwisher et al., 1997; Rossion et al., 2000, 2003b; Sergent et al., 1992; for a review see Haxby et al., 2000). They can be localized in individual subjects with a certain amount of interindividual variability in location, sensitivity and size (Rossion et al., 2003b). The area in the right midfusiform gyrus that responds most strongly to face stimuli has been termed by some the “fusiform face area” (FFA) (Kanwisher et al., 1997). However, isolated lesions in the vicinity of the right midfusiform that do not include occipital cortex are rare because the blood supply comes from both the posterior and middle cerebral arteries thereby protecting it from ischemic damage. In contrast, there are more cases of prosopagnosia with a lesion encompassing the right IOG and sparing the right MFG (Bouvier and Engel, 2005; Figure 16.1). A recent analysis of the lesions causing prosopagnosia has shown that the area of maximal lesion overlap concerned a small territory.
in the right IOG, exactly where a preferential response to faces is observed in the normal brain (Bouvier and Engel, 2005). From these observations, it can be concluded that a lesion of the right IOG is sufficient to cause prosopagnosia, but it is yet unclear whether an isolated lesion of the right MFG can also, in itself, cause a massive face impairment as found in prosopagnosia (see the discussions in Rossion et al., 2003b; Schiltz et al., 2005).

Finally, given that the face processing system is bilateral and widely distributed (Haxby et al., 2000), with a number of key regions in the right hemisphere, it is not strictly necessary to have a lesion in either of these two areas for prosopagnosia to occur. For instance, cases of prosopagnosia with more anterior lesions in the right temporal lobe due to closed head injury (Bukach et al., in press) or progressive right cortical atrophy (e.g. Evans et al., 1995; Joubert et al., 2003) may show a clear prosopagnosic deficit. Regions in the right inferotemporal lobe (i.e. Brodmann area 20) are rarely reported in fMRI studies of normal subjects due to magnetic susceptibility artifacts in the temporal lobe, but PET studies indicate that they play a role in the discrimination of familiar and unfamiliar faces and recollection of semantic information from faces (e.g. Leveroni et al. 2000; Rossion et al., 2001).

### Associated Deficits and Functional Aspects of Prosopagnosia

#### Associated Deficits

As discussed previously, most prosopagnosic patients have a visual field defect, especially in the left upper quadrant (Bouvier and Engel, 2005; Goldsmith and Liu, 2001; Grüsser and Landis, 1991). As for low-level vision, impairments can be found in luminance discrimination, spatial resolution, curvature, line orientation or contrast at low spatial frequencies, but these deficits are rare and are unlikely to contribute to the prosopagnosic deficit (Barton et al., 2004). Contrast sensitivity at higher frequencies may be more frequent (Barton et al., 2004; Caldara et al., 2005; Rizzo et al., 1986) and directly related to the ability to extract fine-grained information about facial identity. One of the most frequent associations with prosopagnosia is achromatopsia — about 60% overlap in the meta-analysis of Bouvier and Engel (2005) — a deficit in color perception often associated with unilateral or bilateral lesions in the temporo-occipital junction, in the anterior part of the fusiform and lingual gyri (Zeki, 1990).

Another deficit frequently associated with prosopagnosia is topographical disorientation (see also Chapter XX). Two types of topographical disorientation are usually described (Landis, 2004). One of them concerns an impairment of vectorial orientation (troubles in angle and distance processing). The other is bound to a loss of environmental familiarity and difficulties in using landmarks. This latter pathology was described in association with a lesion circumscribed...
in the posterior part of the parahippocampal gyrus and the anterior part of the lingual gyrus of the right hemisphere (Habib and Sirigu, 1987).

Larger lesions in bilateral posterior regions can produce, in addition to prosopagnosia, shape and object agnosia, or even pure alexia (Farah, 1990). If prosopagnosia can be found without any deficit in word recognition, especially following unilateral right hemisphere damage, prosopagnosics have usually clear associated deficits in object recognition (e.g. Clarke et al., 1997; Gauthier et al., 1999). A few cases have been presented as having a deficit restricted to faces (e.g. De Renzi, 1986; Henke et al., 1998; Whiteley and Warrington, 1977), but they have been criticized on methodological grounds (e.g. Sergent and Signoret, 1992) because their apparent normal performance at object recognition could be explained by more general perceptual deficits and/or the possibility that a face recognition task is simply more difficult than an object recognition task (Gauthier et al., 1999). Even when the difficulty of face and non-face object discrimination is equated for normal controls and the same tasks are then used to assess the performance of a prosopagnosic patient, differential speed response between faces and non-face objects are usually not considered (e.g. Farah et al., 1995), whereas response times measures may reveal concurrent object recognition difficulties in prosopagnosic patients (Gauthier et al., 1999; Laeng and Caviness, 2001). Yet, certain brain-damaged prosopagnosic patients, such as the patient PS described in later sections, may show a deficit restricted to the category of faces even when response times are considered (Rossion et al., 2003b; Schiltz et al., 2005).

When there are clear impairments at object recognition, the deficits may concern object categories for which the patient had developed a visual expertise at recognizing prior to the lesion (e.g. flowers or cars; Clarke et al., 1997). For instance, Assal et al. (1984) described a farmer who in addition to his prosopagnosia could not recognize his cows anymore. In some cases, however, the deficit can spare the category of expertise, such as cars (Sergent and Signoret, 1992: case RM) or sheep faces (McNeil and Warrington, 1993). Finally, an extremely rare associated deficit is metamorphopsia, or the perception of a deformation of faces, first described by Bodamer (1947: case 3). In such cases, the deficit can also extend to familiar objects. For instance, Seron et al. (1995) described the case of a patient who suffered transient metamorphopsia following a small right occipitotemporal hemorrhage, in the area of the middle fusiform gyrus, and who, in addition to his prosopagnosia, perceived faces and objects such as cars as spatially distorted.

Subtypes of Prosopagnosic Deficits or Functional Aspects of Prosopagnosia

It has been proposed that there may be several subtypes of prosopagnosia (de Renzi et al., 1991; Damasio et al., 1990). Two broad categories are apperceptive
and associative prosopagnosia (de Renzi et al., 1991). In apperceptive prosopagnosia, the chief deficit is held to be impairment in generating an adequate percept of the face, such that it cannot be matched to stored representations of previously seen faces. In associative prosopagnosia, the process of generating the percept is intact but the percept cannot be matched to stored representations, either because of disconnection from, or destruction of, the facial memory stores. However, Farah (1990) and other authors (Davidoff and Landis, 1990) have cast doubts on the existence of pure associative visual (prosop)agnosia, claiming that when tested in detail, all visual agnosa patients, including prosopagnosics, show deficits at the perceptual level. For instance, a few prosopagnosics have been reported to perform in the normal range in face matching tasks on unfamiliar faces like the Benton face recognition test (Benton et al., 1983), but appear to take excessive time to do so (e.g. Bukach et al., 2005; Delvenne et al., 2004; Davidoff and Landis, 1990). All prosopagnostic patients may thus present perceptual deficits at the root of their face impairment, even when the lesion spares the right visual cortex and concern the anterior temporal lobe only (e.g. Joubert et al., 2003; Bukach et al., 2005). Yet, there is a large amount of interindividual variability between prosopagnostic patients in terms of their perceptual deficits (e.g. Barton et al., 2004; Sergent and Signoret, 1992; Schweich and Bruyer, 1993), such that distinctions between apperceptive and associative prosopagnosia are more a matter of degree of visual impairment than of kind. Recent studies suggest that prosopagnostic patients are impaired at extracting configural information on faces: the perception of the relationships between the features (i.e. metric distances) rather than the features (eyes, mouth, nose . . .) themselves would be impaired (Barton et al., 2002; Joubert et al., 2003) (Figure 16.2). Other studies have indicated a deficit at integrating the facial features into holistic representations (Boutsen and Humphreys, 2003; Sergent and Signoret, 1992; Sergent and Villemure, 1989; for a review clarifying the different kinds of configural information, see Maurer et al., 2002). This inability to integrate facial features into a whole representation and to extract metric distances between features is in line with the absence of a decrease of performance for faces presented upside-down generally found in prosopagnosia (e.g. Boutsen and Humphreys, 2003; Delvenne et al., 2004; Marotta et al., 2002): prosopagnostic patients appear to process faces in a piecemeal, analytical manner. Very recently, two independent studies showed that prosopagnosics may present a marked deficit at extracting diagnostic information to recognize faces on the upper part of the face, i.e. the eyes region (Bukach et al., 2005; Caldara et al., 2005). Caldara and collaborators used a learning and response classification method to show that their prosopagnostic patient PS rather used the mouth and the external features of the face, whereas normal controls relied on the eyes and the internal features in general (Figure 16.3). This specific loss of the ability to extract diagnostic
Figure 16.2 Diagnostic information to discriminate and recognize individual faces can be ‘featural’, i.e. concern a modification in the shape and surface properties of a local feature such as the eyes, or ‘configural’, i.e. concern a modification in the relative properties (metric distances) of the features rather than the features themselves. There is evidence that prosopagnosic patients are particularly impaired at extracting configural differences between faces (e.g. Barton et al., 2002).

Figure 16.3 Areas of the face stimulus used by a brain-damaged prosopagnosic patient (PS) with deficit restricted to faces (see Rossion et al., 2003). PS used mainly the mouth area to recognize faces, as identified by a response classification method. In contrast, normal participants extract diagnostic information to recognize faces from the eyes on average, with a left visual field bias (Caldara et al., 2005).
information from the eyes may be related to the impairment at processing configural information, which is heavily weighted in the upper part of the face (see Caldara et al., 2005).

Finally, it is usually stated that prosopagnosics are able to tell apart men and women, estimate age from face, or categorize facial expression on faces (e.g. Bruyer et al., 1983; Tranel et al., 1988). However, these categorizations are usually easier than individual face recognition, and the patients may use external features such as the hair, or local cues (e.g. smile). When external cues such as the hair are masked, the prosopagnosic patient’s performance is generally below normal range at these face judgments also (Sergent and Signoret, 1992).

Neuropsychological Assessment

Key points Assessment of prosopagnosia

Tests aimed to assess other cognitive or visual deficits:
– Wechsler adult intelligence scale (WAIS III)
– Wechsler memory scale (WMS III)
– Birmingham object recognition battery (BORB)
– Visual object and space perception battery (VOSP)
Tests aimed to evaluate facial perception and recognition:
– Benton face recognition test, with measure of accuracy and response times
– Warrington recognition memory test

As stated in the introduction, it is relatively straightforward to define prosopagnosia in a brain-damaged patient, based on the behavior of the patient in a normal life situation. However, the question at stake is not simply to be able to classify the patient as being prosopagnosic, but to characterize the deficit (at which functional level it takes place) and define associated visual and memory impairments. Given the extension of the lesions, it is almost impossible to encounter a patient without any visual and/or memory deficits concerning non-face material. The question will be whether these deficits are directly related (i.e. explain) to the prosopagnosic deficit, or can be considered separately. For instance, achromatopsia — the loss of color perception — is most often (Bouvier and Engel, 2005), but not always associated with prosopagnosia, because the lesions causing prosopagnosia often encompass the visual area V4/V8 in the ventral pathway, critical for color processing in man (Zeki, 1990). Even though color information may play a role in face recognition, it is not thought to be critical for the ability to discriminate and recognize faces. In any case, visual and mnesic impairments will have to be defined extensively, as they will help to determine the kind of tests that can be proposed.
to the patient to assess his/her face processing abilities. A patient with visual short-term memory deficit cannot be tested in delayed face picture matching tasks for instance. Below, we provide a list of tests that could or should be used during the detailed assessment of prosopagnosia.

A first step should exclude a confusional state or a general intellectual impairment. The Wechsler Adult Intelligence Scale (WAIS III) is one of the best tools for this purpose. Furthermore, it allows the examiner to observe how the patient behaves on a wide array of tasks and can elicit cognitive dissociations (for example, better verbal than visual—spatial processing). The role of memory is crucial in the recognition of faces. In this optic, the Wechsler Memory scale (WMS III) evaluates verbal, visual, short- and long-term memory and, once more, can be useful in showing some specific impairment (for example, visual short-term memory deficit). The copy and recall of the Rey-Osterreith figure (Spreen and Strauss, 1998) can give useful informations about visuo-constructive and/or visual memory impairments.

The evaluation of low-level visual capacities is made by a neuro-ophtalmologist. It comprises generally a test of visual acuity (Snellen or Monoyer’s scale), an evaluation of contrast sensitivity (Ginsburg, 2003; Nadler et al., 1990), a dynamic (Goldmann) or static (Octopus) visual field measure, an analysis of stereoptic vision (Buser and Imbert, 1987), color (Farnworth-Munsell; Ishihara) and movement perception (Cavanagh, 1992).

Some neuropsychological tests may also be useful in testing low-level visual capacities. For example, subtests two to five from the Birmingham Object Recognition Battery (BORB) (Riddoch and Humphreys, 1993) are suited to elicit pre-categorical visual processing impairments. These tests (matching tasks) are directed solely at the perception of basic properties of object forms — size, orientation, location and length. Computerized versions of some tests of low-level vision (contrast sensitivity, spatial resolution, etc.) have been developed (Barton et al., 2004). The “shape detection screening test” and the “incomplete letters test,” two subtests from the Visual Object and Space Perception Battery (VOSP) (Warrington and James, 1991), as well as subtest six of the BORB (“the overlapping figure test”), can help to detect difficulties in figure/ground perception. More elaborated tests would involve determining whether three-dimensional figures are structurally possible or impossible (Farah, 1990; Delvenne et al., 2004).

The next step is to test for the presence of a visual agnosia for non-face objects. Subtests seven and eight of the BORB point on difficulties in object recognition across different viewpoints. However, these may be achieved successfully through slow feature matching, even in visual agnosic patients (Delvenne et al., 2004).
A complete testing of normal object perception should include the simultaneous matching of novel (non-existent) objects and familiar objects presented in the same and in different viewpoints (Delvenne et al., 2004; Rossion et al., 2003a; Sergent and Signoret, 1992). At the associative level, tests nine to fourteen in BORB assess whether patients have intact stored knowledge required to recognize and name visually presented objects. Another visual agnosia battery often used in the French community is the Montréal–Toulouse battery (Agniel et al., 1992). Object naming can be tested with the Snodgrass and Vanderwart’s (1980) set of line drawings, for which norms have been collected in several languages, and which are also available in gray level and colorized versions (Rossion and Pourtois, 2004). As indicated in the previous section, accuracy measures are not sufficient to exclude a deficit in object recognition since prosopagnosic patients may achieve normal performance at object recognition when given unlimited time to respond (Farah, 1990). Response time measures may reveal concurrent object recognition difficulties in prosopagnosic patients (Delvenne et al., 2004; Gauthier et al., 1999; Laeng and Caviness, 2001). Since brain-damaged patients can be generally slowed down (Benton, 1986), it may be important to assess phasic reaction times (RT), or other baseline measures of RT. Alternatively, the performance of the patient can be compared to brain damaged controls without visual deficits.

Concerning face processing, the most popular test is the Benton face recognition test (Benton et al., 1983), which is particularly well adapted to the detection of perceptual impairments. It requires matching unfamiliar faces that do not contain external face features, across changes of viewpoint and lightning and the face pictures. This test is highly diagnostic of prosopagnosia. Some authors have criticized this test because in rare cases the prosopagnosic patients can perform in the normal range, or because it would be possible to match faces on local features such as eyebrows (Duchaine and Weidenfeld, 2003). However, these criticisms are unjustified: when such strategies are used, RT measures will complement the accuracy scores and reveal that prosopagnosic patients take unusually long times to respond (e.g. Bukach et al., 2005; Delvenne et al., 2004). The recognition memory test (Warrington, 1984) evaluates non-verbal recognition memory with a facial recognition (old/new) test. A shorter version of this test is obtainable in the Camden Memory Battery (Warrington, 1996). The patient can also be tested with face photographs of personally familiar or famous people, but such tests should be developed on an individual basis. Other face processing functions such as gender categorization, facial expression and age assessment can be performed with face stimuli without external cues (hair, earrings, etc.) again considering both accuracy rates and RT measures.
Prognosis and Management of the Disorder

Key points Management and treatment

- Spontaneous recovery: unusual
- Revalidation:
  - in selected patients
  - strategy adapted to the deficit (internal features . . . )
  - decrease the consequences of prosopagnosia in everyday life

Prosopagnosia due to vascular disease is always associated with posterior cerebral artery territory infarcts (PCAI). In the series of PCAI, death is rare, varying from 0% (Brandt et al., 2000), to 3% (Yamamoto et al., 1999), 5% (Pessin et al., 1987), and 7% (Kumral et al., 2004; Milandre et al., 1994). Mortality and major disability were found to be strongly associated with cardioembolism and atherosclerotic large-artery disease of vertebrobasilar artery and PCA (Brandt et al., 2000; Kumral et al., 2004). Outcome is usually good (Brandt et al., 2000).

As discussed before, prosopagnosia is due to lesions in the inferomedial part of the temporo-occipital area, and the outcome of prosopagnosia depends on the size of infarction in these regions. Cognitive rehabilitation of prosopagnosia is rarely described. In fact we found only four publications on this subject (Beyn and Knyazeva, 1962; Glowic and Violon, 1981; Ellis and Young, 1988; Polster and Rapcsak, 1996). For the two first studies, the therapy methodology is poorly described. On the contrary, descriptions of Ellis and Young (1988) concerning the training of a young girl in face recognition are well-detailed (mostly face matching tasks), but the conclusions are dramatic. After more than two years of daily training, no improvement was observed. The authors concluded that any attempt at improving face recognition was vain, and that the patient should be trained to recognize persons by the mean of others cues such as the voice. Polster and Rapcsak (1996) tried to teach new faces to their prosopagnosic patient RJ. They concluded that semantic cueing (such as asking to associate the photograph of a face with a story concerning the person or with personality traits of this person) was more efficient than structural cuing (for example to determine the salient traits of a face). Nevertheless, the improvement was restricted to a specific viewpoint of the photograph and no generalization to other presentation of the photography could be made. Consequently, the techniques used to improve learning of new faces were inefficient in daily life, where faces are always seen from different viewpoints. We tried to improve face recognition in the patient PS, a state nursery school teacher who suffers from a particularly pure and severe prosopagnosia (Caldara et al., 2005; Mayer et al., 1999; Rossion et al., 2003b;
Prosopagnosia

Schiltz et al., 2005). For example, this patient did not recognize her daughter or her husband out of context when they kept silent. The revalidation strategy was based on the patient’s spontaneous descriptions of known persons. We noted that the patient face’s descriptions were based on the hair (length, color, presence of a fringe) and the global form of the face (round, lengthened, oval) as well as on visually derived semantic representations (“she looks Portuguese”). She never described the eyes, the nose or the mouth. We made the hypothesis that the patient presented a deficit in the processing of internal features of faces, as confirmed later (Caldara et al., 2005). The processing of internal features, in particular the eyes, is essential to recognize familiar faces (Ellis et al., 1979), and increases with face familiarity (O’Donnell and Bruce, 2001). Therefore we chose to train PS in the processing of internal traits. Rehabilitation was four months long with two sessions per week. First we established with the patient a list of terms related to faces features (for example, almond eyes, turned-up nose, shaggy eyebrows). Then we trained PS to analyze internal features of faces (the nose, the mouth, eyes and eyebrows). Three types of face with increasing difficulty were presented: (1) personality caricature, (2) unknown faces of adults, and (3) unknown faces of children. Two tasks were proposed: first, PS had to sort face pictures by means of a given criterion (for example, the length of the mouth); then PS had to describe the internal feature of face photographs. Finally, the last sessions were devoted to establish a description in a card index of the prominent features of all the children in PS’ class. PS had to fill those cards herself. Then, the descriptions were presented to the therapist, and sometimes modified after discussion. The card had to be learned by heart by PS, who should have been able to give one or more prominent features when a child’s name was given.

Two measures were employed in order to evaluate the rehabilitation efficacy:
1. The first consisted in testing the effective improvement of PS in the recognition of her class children from certain features (internal versus external) by comparing her scores to those of a colleague.
2. The second was a questionnaire based on subjective changes in the manner of processing faces (for more details see Mayer et al., 1999).

Results showed that the improvement of PS was significantly better in the recognition of faces composed of internal features than in recognition of faces composed of external features. Moreover, the questionnaire showed a subjective improvement in the item concerning the identification of children with a hat, which indicate that PS was less impaired than before when she could not use the hair for identification. Among the cues spontaneously used for children identification, we noticed the apparition of the mouth and the eyes, even if the hair and the face outline remained the main cues used by PS. The consequences of PS’ improvement in analyzing faces were important: before training, she could
not go outside the school alone with her pupils; after training, she felt sufficiently secure to stay alone outside with the pupils. New training strategies may be developed based on the findings that PS, as well as other prosopagnosics, can use the mouth area to recognize faces, but is markedly impaired at the level of the eyes (Caldara et al., 2005).

In conclusion, rehabilitation does not treat prosopagnosia — that is, it does not restore the possibility to recover the familiarity of a known face. Nevertheless, it can diminish the consequences of such deficits in everyday life. This aim corresponds to the recent approach of neuropsychological rehabilitation, as described by Barbara Wilson for example:

The main purposes of neuropsychological rehabilitation are to enable people with disabilities to achieve their optimum level of well being, to reduce the impact of their problems on everyday life and to help them return to their most appropriate environments. (Wilson, 2003, p. 1)
331 Prosopagnosia


Prosopagnosia


Prosopagnosia


