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Prosopagnosia

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Abstract

Prosopagnosia refers to impaired recognition of faces. The article first describes the different etiological factors and clinical phenotypes of prosopagnosia: developmental and acquired prosopagnosia for the former and apperceptive, associative, amnestic, and multimodal prosopagnosia for the latter. This includes a review of the face and function specificity of the deficit. Subsequently, the results from neuroimaging studies are discussed. The article ends with a paragraph on neurodegeneration and prosopagnosia.

Introduction

Faces are attention-getters, and this is one of the main reasons why, in social interaction and in our thinking about it, the face occupies center stage. There may also be other, quantitative, reasons why the face plays such a central role. Indeed, very few, if any, natural objects carry as much information as does the face. Whether an observer needs to know identity or gender, is wondering about age or attractiveness, is listening to speech, or is making an evaluation of trustworthiness, the gaze turns to the other's face. If only for these reasons, one understands that there is more to learn from the face than from any other source of social signals.

Two assumptions are underlying the many claims about face specificity that are at the forefront of discussions on prosopagnosia: (1) that faces trigger perceptual processes, which have category-specific markers, and (2) that faces occupy a neurofunctional niche of their own, such that neural face representation coexists with (but does not overlap with) object representation – a view that in one sense or another, it is linked to modularity or more generally to the notion of special functional units in the brain. Clinical cases constitute critical tests for theoretical models, and patients suffering from deficits in face recognition (Bodamer, 1947) have long served as an important touchstone for models of face processing.

Acquired and Developmental Prosopagnosia

Imagine that if every time you encounter the face of an acquaintance, it seemed no more familiar to you than that of a total stranger. This peculiar symptom is known as prosopagnosia – and not surprisingly, it takes a heavy toll on the social life of the patients who suffer from it. Prosopagnosics can even have difficulties recognizing people whom they are very close to, such as immediate family members, and they have to rely on other cues to identify a person – clothing, for instance, or voice. In less-radical cases, prosopagnosics are helped by the context in which they routinely encounter individuals, but they fail to recognize them when they encounter them outside the usual context – for example, when they meet a colleague in the market.

Prosopagnosia may occur after a neurological antecedent, in which case it is known as acquired prosopagnosia (AP), and is often associated with lesions in the occipito-temporal brain regions, especially in the right hemisphere (Barton et al., 2002; Damasio et al., 1982; Landis et al., 1986; Levine and Calvanio, 1989; Meadows, 1974; Sergent and Signoret, 1992; Marotta et al., 2001; Sorger et al., 2007; Steeves et al., 2006; de Gelder et al., 2003) or following a head trauma but without evident brain lesions showing up on standard imaging (Hadjikhani and de Gelder, 2003; de Gelder et al., 2003).

Prosopagnosia can also be present without any neurological history. In this case, it is known as developmental prosopagnosia (DP). However, this does not imply the absence of structural brain abnormalities. For instance, there are strong indications that congenital hypoplasia of the cerebellar vermis can be associated with face recognition difficulties (Van den Stock et al., 2012b). Patients with DP usually report lifelong problems in recognizing people by the face. DP sometimes occurs in different generations of a single family, suggesting a genetic component, more particularly an autosomal dominant mode of inheritance (Grüter et al., 2007). The term 'congenital prosopagnosia' is sometimes used to refer to prosopagnosia without any neurological antecedents. However, the congenital nature of DP is a matter of debate, and there is no conclusive evidence that all of these cases have a genetic basis. The term 'developmental prosopagnosia' has a more descriptive connotation and stresses (next to the genetic hypothesis) the possibility of a developmental disorder – for instance, in the maturation and structural connectivity of different cortical areas, such as the fusiform gyrus (Behrmann et al., 2007; Thomas et al., 2009). The pattern of deficits in AP and DP is similar as far as some basic aspects of face processing are concerned, but different in other ways (de Gelder and Rouw, 2000a).

The prosopagnosia symptom centers on recognition of personal identity but not of facial expression, and such

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a dissociation has long been the cornerstone of the models of face processing in the neuropsychological literature of the last two decades, and is at the basis of the face recognition model of Bruce and Young (1986). Until recently, this dissociation was generally accepted – and as a consequence, researchers rarely investigated facial expression recognition itself, nor was much attention paid to the possibility of an interaction between the face identity and the facial expression recognition system. When problems with faces are defined as problems with identity recognition, then understanding face recognition may or may not be seen as a subspecies of object recognition. Substantial effort has gone into finding evidence for dissociations between face- and object recognition. In the 1980s and 1990s, the gold standard for supporting this kind of special or modular cognitive ability argument was to find a double dissociation. Patients with object-recognition deficits that were not prosopagnosic (as opposed to prosopagnosics without object-recognition problems) perfectly fit this bill. However such pure cases have been proved extremely rare.

Indeed, while a number of cases of prosopagnosia have been reported over the last hundred years, only very few provide evidence for pure or genuine prosopagnosia, affecting face perception in the complete absence of any other object perception deficit. Regarding the question whether prosopagnosia is exclusively a face-identity recognition deficit or whether the face recognition deficit is simply the most dominant deficit, the debate has not been concluded yet. As shown in a well-known review of the literature published at the end of the 1980s, almost all of the reported cases have associated deficits in object recognition (Farah, 1990). Lively debates will undoubtedly continue as long as new ways of testing face specificity are being developed that reflect increasingly better understanding of face recognition.

Face and Function Specificity of Prosopagnosia

Standard procedures in prosopagnosia assessment must include at least a basic screening of the visual object-recognition abilities with standard clinical test batteries such as the Birmingham Object Recognition Battery (Riddoch and Humphreys, 1993) or the Visual Object and Space Perception Battery (Warrington and James, 1991). It is not uncommon that prosopagnosics score outside of the normal range on at least one of the tests (e.g., Duchaine, 2000). Standard clinical tests for evaluating face identity matching and memory of face identity include the Benton Facial Recognition Test (Benton et al., 1983) and the Warrington Recognition Memory Test (Warrington, 1984). The scores on these tests should thus be interpreted cautiously, especially when no data are available on reaction time (as is the case for the older studies).

More modern tests and batteries are available now, targeting specific aspects of face- and object processing (de Gelder et al., 1998; de Gelder and Rouw, 2000b; Duchaine and Nakayama, 2006; de Gelder and Van den Stock, 2011; de Gelder and Bertelson, 2009). These provide a more detailed analysis of the intact and anomalous aspects of face- and object processing. The Face Emotion Action Stimulus Test (de Gelder and Van den Stock, in preparation), for example, targets a range of specific abilities including face detection, emotional face identity memory, neutral face identity memory, facial emotion matching, facial identity matching, object identity matching, facial part-to-whole matching, object part-to-whole matching, and configural face and object processing.

Since normal face processing involves multiple hierarchical and parallel processes, impairments in different processes will result in different types of behavioral and neuroanatomical correlates. In developmental disorders like DP, heterogeneity is rather the rule than the exception and this is also reflected in the behavioral findings. Extensive evaluations of single cases or small groups of prosopagnosics have found different patterns of impairment on multiple aspects of face processing, including configural processing, modulation of the N170, within-object spatial relations, recognition of emotions, gender discrimination, recognition of famous faces, holistic face processing, and facial attractiveness ratings.

There have been some attempts at making a taxonomy of the heterogeneity in clinical phenotypes associated with prosopagnosia. An older and often ignored effort at getting underneath the surface of a behavioral deficit was to understand prosopagnosia as a form of agnosia using a historical distinction between two types of agnosia, described by Lissauer (1890): (1) apperceptive agnosia reflects impairment in consciously perceiving and recognizing stimuli and (2) associative agnosia refers to a relatively preserved ability to perceive objects, but an inability to interpret what is seen or match it with its associated meaning. Apperceptive prosopagnosia therefore relates to an inability to perceive a face as an invariant face and patients suffering from this type will be mostly impaired in tasks tapping into face *detection* and face *discrimination*; whereas, associative prosopagnosia is in the first place associated with the recognition of individual faces and involves a deficit in the kind of encoding that allows telling individual exemplars apart and accessing stored representations. It is the latter type that has been mainly reported and investigated as it is assumed that prosopagnosia arises at a higher-order recognition and cognitive level. Questions about intact face detection typically are not raised in prosopagnosia and intact face detection is typically not seen as possibly a separate and earlier stage in face perception (but see de Gelder and Rouw, 2000a; de Gelder et al., 2003; Righart et al., 2010). Formal neuropsychological evaluation of associative prosopagnosia requires the use of 'semantically unique items,' i.e., stimulus exemplars that carry stimulus-specific semantic associations. In other words, the stimulus needs to have unique semantic properties that are not present in any other exemplar of the same stimulus category. Typically, famous faces figure as the preferred stimulus category for this purpose, although idiosyncratically familiar (Van den Stock et al., 2012a) or experimentally familiarized (Stollhoff et al., 2011) faces have also been used.

A less known third subtype has been proposed: amnestic prosopagnosia or prosopamnesia, which refers to the impaired ability to learn new faces. In case of acquired face recognition deficits, diagnostic criteria for a 'pure' variant would include preserved face perception, preserved memory for nonface visual material, and preserved recognition of premorbidly familiar faces (Tippett et al., 2000); whereas, for the developmental variant, the deficit would consist of abnormal processing of repeated unfamiliar faces, in combination with normal processing of familiar faces and familiar and unfamiliar nonface

visual materials (Williams et al., 2007). However, little is known about face familiarization and there is limited evidence for a clear distinction between associative and amnestic DP.

Finally, associative and apperceptive prosopagnosia are to be distinguished from impaired person recognition. There is anecdotal evidence of person recognition deficits that are not face selective and extend to voices and names (Tyrrell et al., 1990; Evans et al., 1995; Gentileschi et al., 2001; Gainotti et al., 2003). The modality specificity of the person recognition deficit would be among the main differentiating characteristics between prosopagnosia and person recognition symptoms. A multimodal person recognition deficit argues against a pure prosopagnosia. However, the majority of the studies on face recognition difficulties have only investigated the visual modality (Gainotti, 2013). In addition, cases of phonagnosia have been reported, showing impaired voice recognition, which may or may not be modality specific (Garrido et al., 2009; Hailstone et al., 2010; Van Lancker and Canter, 1982; Van Lancker et al., 1988; Van Lancker et al., 1989).

Functional Imaging Findings

The major focus of face perception research continues to be the so-called configuration-based processing or configural processing in short (Tarr, this volume). Configural processing refers to the mechanism of object perception that goes beyond the mere analysis of features or parts, but includes the analysis of spatial relations between the constituting features or parts of the object. Configural processing is most often measured by the inversion effect, which counts as the signature of intact face processing skills and it is commonly tested for in case of face recognition deficits. A few studies have reported that when a face perception disorder is present, the person or the patient does not show the normal inversion effect (de Gelder and Rouw, 2000a), but this phenomenon is also still a matter of debate (Huis in 't Veld et al., 2012). What is less clear is whether an anomaly in perceiving the face configuration and in showing the inversion effect, when confronted with neutral faces, does in fact generalize to faces where it is not the identity but the expression that matters. The available fMRI studies focusing on face perception in prosopagnosics have revealed inconsistent results (see Van den Stock et al., 2008 for an overview). In the first fMRI study with prosopagnosics, we studied activation in the occipital face area (OFA) and fusiform face area (FFA) of three prosopagnosia patients. None of them showed the normal higher activity to faces compared to objects in either region (or elsewhere). Furthermore, in two patients, there was similar activation for faces and objects in OFA and FFA (see Figure 1).

Few studies included the emotional dimension provided by a face. We investigated whether adding an emotional expression would normalize the face processing style of prosopagnosics with respect to the inversion effect. We presented neutral and emotional faces to patients with AP with lesions in

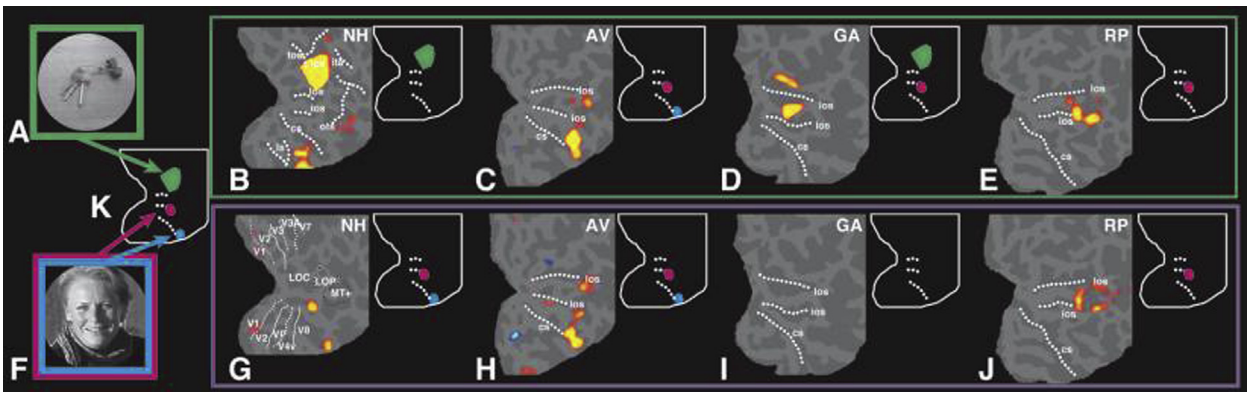

Figure 1 Activation obtained for objects (B–E) and faces (G–J) compared to control stimuli in the right hemisphere of a normal subject (B, G) and in three prosopagnosic patients (C–E, H–J). Data are represented on a flattened representation of the occipital cortex, with gyri appearing in light gray, whereas sulci are in darker gray. The dotted lines in (B) represent the major sulci (tos, transverse occipital sulcus; ips, intraparietal sulcus; its, inferior temporal sulcus; ots, occipito-temporal sulcus; los, lateral occipital sulcus; ios, inferior occipital sulcus; cs, collateral sulcus; ls, lingual sulcus). G shows the location of the different visual areas obtained by retinotopic mapping. (A) Object and (F) face show an example of the stimuli used in the experiment, and (K) is a diagram of the activation obtained for objects (in green) compared to scrambled objects and faces compared to scrambled faces (in blue and magenta) in a normal representative subject. On the upper right part of each panel, a diagram indicates the areas activated in each subject in comparison with the normal subject (K). In normal subjects, the viewing of objects compared to scrambled objects (B) elicited activation principally in the lateral occipital cortex (LO, in green in the diagram), whereas faces compared to scrambled faces activated two areas: the anterior part of the collateral sulcus and fusiform gyrus (fusiform face area (FFA), in blue) and the inferior occipital gyrus and sulcus (inferior occipital gyrus (IOG), in magenta). In patient AV, the developmental prosopagnosic, viewing both objects compared to houses (C) and faces compared to houses (H) activated the same region of the cortex, in the anterior part of the collateral sulcus and the fusiform gyrus (FFA). Objects did not produce activation in the expected LO region, and faces failed to activate the IOG area. In patient GA, injured at 18 months, faces failed to elicit any activation in the occipital cortex compared to scrambled faces (I), whereas objects compared to scrambled objects activated both the LO and the IOG area (D). Finally, patient RP, injured at 7 years, showed similar activation to both faces compared to houses (J) and objects compared to houses (E), situated in the IOG region. No activation was seen in the FFA area. None of the prosopagnosic patients showed activation in both the FFA and the IOG to face presentation. Instead they showed activation in either one or the other, or none at all.

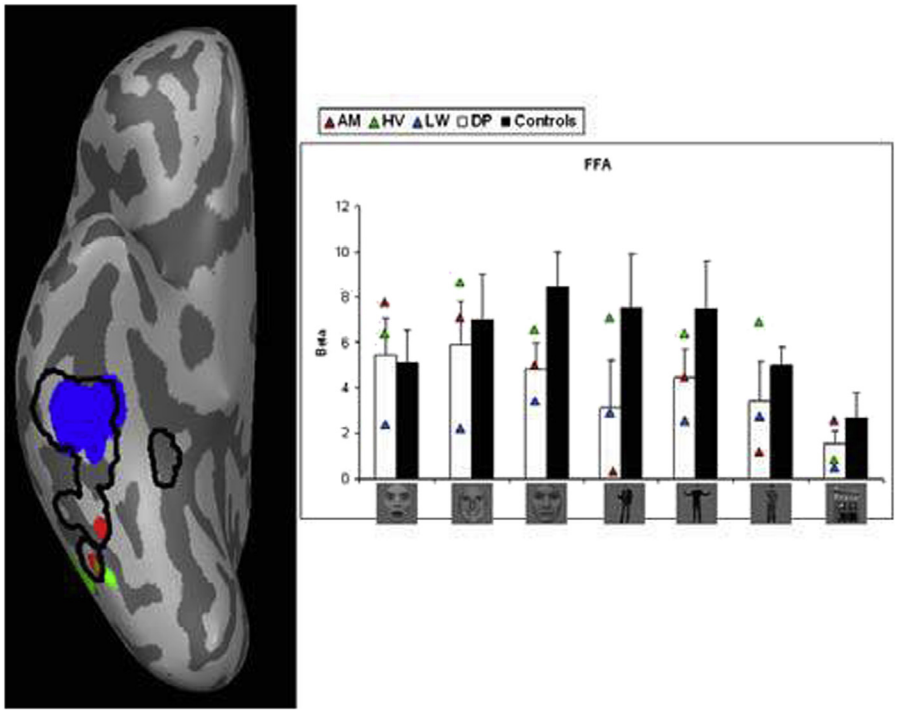

Figure 2 Face-specific activation in the right fusiform gyrus when comparing faces (fearful/happy/neutral) with houses. Left: Areas are shown on an inflated right hemisphere. Activation maps of the control subjects are collapsed and displayed by the black contours. Activation of the individual developmental prosopagnosia subjects (DPs) is plotted in color. Right: beta values by condition, group, and DP subject. Error bars represent one standard error of the mean. Conditions represent from left to right: fearful faces, happy faces, neutral faces, fearful bodies, happy bodies, neutral bodies, and houses. White columns display the average value of the three patients. Black columns show the average value of the controls. Triangles represent the individual values of the DPs.

FFA, inferior occipital gyrus, or both. The results showed that the patients more strongly activated other face-sensitive areas like the superior temporal sulcus or amygdala when they perceive facial expressions (de Gelder et al., 2003). Furthermore, they were more accurate and faster in processing emotional faces compared to neutral faces, consistent with other studies (Duchaine et al., 2003; Jones and Tranel, 2001; Nunn et al., 2001). Since the patients in de Gelder et al. (2003) had lesions in the ventral occipito-temporal cortex, we wondered how these brain areas respond to emotional information in DP patients. Therefore, we presented a group of DPs and matched controls with fearful, happy, and neutral faces (and bodies). The results showed normal activation in FFA for emotional faces (fearful and happy), but lower activation for neutral faces, compared to controls (Van den Stock et al., 2008) (see **Figure 2**).

Prosopagnosia and Neurodegeneration

Neurodevelopmental syndromes can reveal important insights into normal cognitive functioning as selective deficits may arise in the course of neurodevelopmental syndromes, including neurodegeneration. In addition, the change in symptomatology associated with increased regional brain atrophy following progression of the neurodegenerative disease process can provide information regarding the regional involvement in cognitive functioning. While the results from studies targeting face perception in, for instance, autism spectrum disorders are heterogeneous (Barton et al., 2004), there have been recent significant advances in the characterization and categorization of dementia syndromes, for instance, concerning both language (Gorno-Tempini et al., 2011) and behavioral (Rascovsky et al., 2011) variants of frontotemporal lobar degeneration. The behavioral and neuroanatomical findings from neurodegenerative syndromes grossly mirror the apperceptive-associative-amnestic-multimodal classification of prosopagnosia. For instance, there is evidence that the neurodegenerative syndrome, known as 'posterior cortical atrophy,' is associated with apperceptive prosopagnosia and neurodegeneration of primarily occipito-temporal regions (Meek et al., 2013). There is consensus that semantic dementia, a syndrome characterized by progressive semantic deficits and anterior temporal lobe abnormalities, is associated with associative prosopagnosia (Snowden et al., 2004). A recent structural neuroimaging study in a group of neurodegenerative patients with progressive atrophy of primarily the frontal and temporal lobes provided additional evidence for the role of the anterior temporal lobes in famous face recognition: the results revealed an association between left anterior temporal lobe atrophy and famous face naming, whereas famous face recognition was related to bilateral anterior temporal lobe atrophy (Gefen et al., 2013). These results are extended by the selective progressive prosopagnosia symptomatology of an anatomical

subtype of frontotemporal neurodegeneration, characterized by predominantly right temporal lobe abnormalities as evidenced by several case reports (Evans et al., 1995; Gainotti et al., 2003; Joubert et al., 2003; Grossi et al., 2012).

However, there have been an increasing number of case reports on atypical presentations of neurodegenerative syndromes, including face perception symptomatology. For instance, we recently reported two atypical cases of Alzheimer's disease. One patient (MV) showed a mirror sign (inability to recognize one's own image in the mirror) (Van den Stock et al., 2012) and her performance on in-house custom-designed face and object processing tests was indicative of face-selective hyperfamiliarity processing with relatively intact identity recognition. The second patient (IS) displayed a particular delusion: she carried around a toy doll, of which she was convinced it was her own living grandchild (Van den Stock et al., 2013) and her neuropsychological face and object processing abilities also indicated face-selective hyperfamiliarity processing but also impaired identity recognition. These findings illustrate the heterogeneity in clinical phenotypes regarding different aspects of face and object processing.

As there is an increasing body of evidence on reports of prosopagnosia, in both scientific and nonscientific media (Della Sala and Schweinberger, 2013), the future decade holds promise regarding a better understanding of normal and abnormal face perception. But this will require putting the debate on face perception in the broader context of object recognition on the one hand, and of affect perception on the other. It is fair to conclude that for the time being, specialized studies on the localization of one, two, or more face areas in the brain have not yet moved forward our understanding of prosopagnosia.

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