



## Neural and genetic foundations of face recognition and prosopagnosia

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Faces are of essential importance for human social life. They provide valuable information about the identity, expression, gaze, health, and age of a person. Recent face-processing models assume highly interconnected neural structures between different temporal, occipital, and frontal brain areas with several feedback loops. A selective deficit in the visual learning and recognition of faces is known as prosopagnosia, which can be found both in acquired and congenital form. Recently, a hereditary sub-type of congenital prosopagnosia with a very high prevalence rate of 2.5% has been identified. Recent research results show that hereditary prosopagnosia is a clearly circumscribed face-processing deficit with a characteristic set of clinical symptoms. Comparing face processing of people of prosopagnosia with that of controls can help to develop a more conclusive and integrated model of face processing. Here, we provide a summary of the current state of face processing research. We also describe the different types of prosopagnosia and present the set of typical symptoms found in the hereditary type. Finally, we will discuss the implications for future face recognition research.

Faces are of utmost importance for human social life. Not only do they tell us the identity of other people, but they also inform us about their mood, age, health, and their gender. They are a major factor of sexual attractiveness. From the eyes, we can judge the direction of gaze with remarkable accuracy, and the observation of lip movements helps to understand speech.

Neurons tuned to faces are not limited to human brains, they have been found in the superior temporal sulcus (STS) of monkeys as well (Perrett, Rolls, & Caan, 1982). Recently, Tsao, Freiwald, Tootell, and Livingstone (2006) identified a face sensitive area in the macaque monkey brain, which is topographically homologous to the face sensitive area on the fusiform gyrus in humans (fusiform face area, FFA).

This indicates that face recognition is evolutionarily old and already existed in the last common ancestor of humans and macaque monkeys.

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Other face-related abilities are probably inherited as well: the gaze direction judgment is active from birth (Farroni, Massaccesi, Menon, & Johnson, 2007), and basic facial expressions as well as their recognition are universal among humans (Ekman *et al.*, 1987).

The function of the human brain's face identity recognition system cannot be substituted in full by other brain regions. Even if the system is damaged in early childhood, the plasticity of the juvenile brain may not be sufficient to recover the functionality (Barton, Cherkasova, Press, Intriligator, & O'Connor, 2003). The result is a life-long prosopagnosia, a selective face recognition deficit.

*Prosopagnosia* is a scholarly Greek compound word (from *prosopon* = the face and *agnosia* = non-recognition or non-knowledge). It was introduced by Joachim Bodamer (1947), who published a very thorough description of two soldiers with a markedly decreased face recognition after suffering severe brain injuries in the Second World War. In the following years, several case reports were published and prosopagnosia was regarded as a rare consequence of a damage to the temporal or occipital lobe.

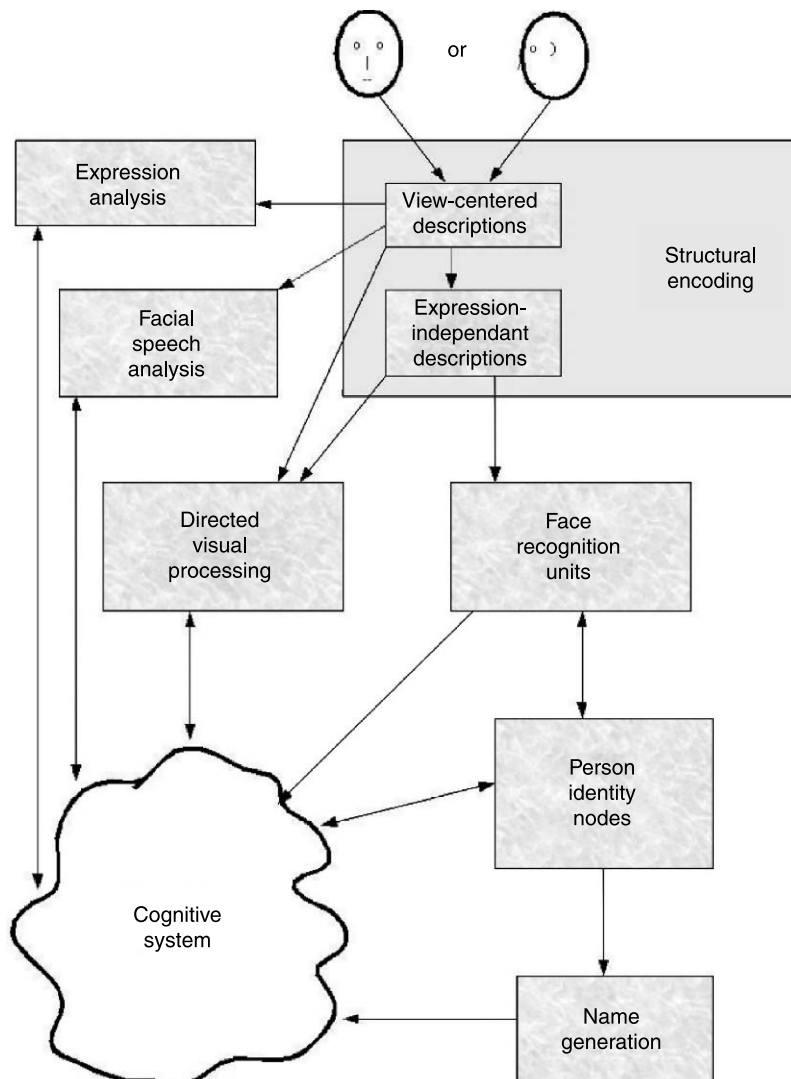
There is also a congenital type of prosopagnosia, which is present from birth and is not accompanied by any gross brain abnormalities, first reported by McConachie (1976). Recently, this type has been shown to be very common: about 2.5% of the Caucasian population could be affected, placing the prevalence of congenital prosopagnosia in the same region as dyslexia or dyscalculia (Kennerknecht *et al.*, 2006). The condition can be hereditary. De Haan (1999) found a family where the father and two daughters suffered from a markedly reduced face recognition ability. Grueter (2004) examined 38 people with congenital prosopagnosia in six families. Prosopagnosia diagnosis is difficult: there is no test which can establish the diagnosis of a face recognition dysfunction. Other than in the case of dyscalculia and dyslexia, face recognition skills are not taught in school and there are no age- and general intelligence-adjusted standards. The widely used Warrington Recognition Memory for Faces (RMFs) and the Benton Facial Recognition Test (BFRT) are not sufficiently face specific (Duchaine & Weidenfeld, 2003). Other tests have been proposed (Duchaine & Nakayama, 2006; Grueter *et al.*, 2007; Minnebusch, Suchan, Ramon, & Daum, 2007), but none of them can establish but only support a diagnosis. Prosopagnosia is not always obvious. In Bodamer's (1947) two cases, the patients never complained of their face recognition deficit, only Bodamer's observations, targeted questions and subsequent tests revealed their impairment. Rosler, Lanquillon, Dippel, and Braune (1997) tested 31 patients in the second week after a right (16) and left (15) temporal lobe infarction for face recognition. They found that most of them were impaired, some of them severely, but none complained of face recognition problems. The authors conclude that 'probably these deficits are easy to compensate for, because recognizing a certain person 'behind' a face is made easy by the perception of further visual and acoustic cues'. If this applies to patients with a recent stroke, how much more difficult may it be to identify people with a life-long face recognition deficit? We should always be aware that in the first place face recognition is a tool for person recognition and people with a defect will have a large reservoir of other tools to choose from.

In this article, we will sum up the current research on face recognition and its dysfunctions with a special focus on the hereditary type of prosopagnosia, because with a prevalence of 2.5% this type is by far the most frequent one. We will propose a diagnostic checklist based on the clinical symptoms.

**Face recognition**

**Normal face recognition process**

Face recognition is a multistage process. First, a face is recognized as such; this stage is often described as detection phase. In a second step, individual and non-individual facial information (emotional expression, eye gaze, gender, age, and health) must be analysed. Individual face recognition includes a structural encoding and a comparison with stored mental images of faces. The 'classic' functional face recognition model of Bruce and Young (1986) depicts this process (Figure 1). It does not connect the tasks involved with any cerebral structures, though. A later model by Ellis and Lewis assumes conscious face recognition and the generation of a feeling of familiarity to be processed by separate modules. This dissociation can be shown in some cases of Capgras delusion, a rare psychiatric condition where the patients are convinced that



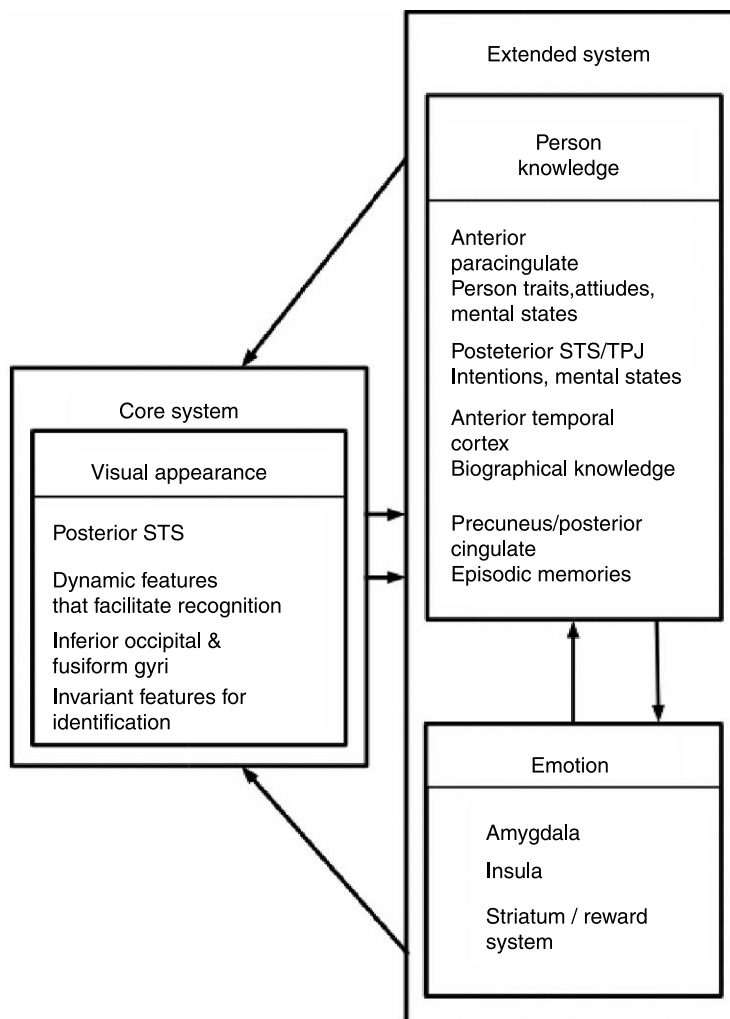
**Figure 1.** Functional model of face recognition by Bruce and Young (1986).

close relatives are replaced by identically looking imposters. Ellis and Lewis assume that in the Capgras delusion the conscious face recognition is preserved but no longer triggers the feeling of familiarity. In acquired prosopagnosia, the opposite effect might occur: while there is no conscious face recognition, a subconscious feeling of familiarity and an autonomous reaction, for example, measured by skin conductance response, can still be shown in some cases (Ellis & Lewis, 2001). The feeling of familiarity may also be distorted in a different way: Vuilleumier, Mohr, Valenza, Wetzell, and Landis (2003) reported the case of a young woman who felt an irritating hyperfamiliarity towards strangers after a left temporal brain lesion. Her conscious face recognition, though, was undisturbed. A similar case of hyperfamiliarity in an epileptic patient who suffered a large left occipito-temporal tissue damage in early childhood was reported by Murai, Kubota, and Sengoku (2000). Their patient showed hyperfamiliarity in combination with a paramnesia. Obviously, the feeling of familiarity is generated in a separate module which is triggered by the face recognition unit. While Ellis and Lewis hold a disconnection responsible for false feeling of unfamiliarity, we assume insufficient trigger input to cause a lasting uncertainty as observed in hereditary prosopagnosia. Instead, the false feeling of unfamiliarity in the Capgras delusion would stem from a damage in the familiarity feeling itself. Based on functional imaging research, Gobbini and Haxby (2007) proposed a face recognition model which ties functional units to certain brain regions. They distinguish between a core and an extended system (Figure 2). The core system comprises the occipital face area (OFA) in the inferior occipital lobe, the fusiform face area (FFA) in the middle fusiform gyrus, and the face area in the dorsal superior temporal sulcus (STS). In several fMRI studies, these areas showed an enhanced activation for faces as compared with other visual objects. The STS is supposed to process dynamic facial information (expression, eye gaze, and facial speech) while the OFA and FFA focus on invariant facial features for identification. The core system encodes the visual appearance of familiar faces while the extended system provides the person's information and the emotional response. The model is less explicit about the hierarchy of functions and regions than the earlier model by Haxby, Hoffman, and Gobbini (2000), which it supersedes, and proposes a tightly integrated network of face-processing areas.

### **Face recognition network**

Several studies have shown that visual face processing activates a whole network of brain regions (Gobbini & Haxby, 2007; Ishai, Schmidt, & Boesiger, 2005). The hierarchical model which postulated that the OFA is responsible for the initial processing of facial features and feeds the results into the FFA and STS is outdated.

Rossion *et al.* (2003) reported the case of a woman with an acquired prosopagnosia due to a major tissue loss in the right inferior lateral occipital lobe and the left medial temporal lobe, mainly in the fusiform gyrus. Though the right OFA was destroyed, the right FFA reacted to faces. A similar case was reported by Steeves *et al.* (2006). They found a face-related activation in the FFA and the STS in a prosopagnosic patient with a destroyed OFA. Obviously, the FFA reaction to faces is not necessarily missing in acquired prosopagnosia, though its quality may change. A second study with the first patient found a missing adaptation reaction to known faces in the FFA activation (Schiltz *et al.*, 2006). While in the controls, the FFA activation decreased when a face is shown repeatedly, the prosopagnosic patient showed no such adaptation.



**Figure 2.** Model of face processing by Gobbini and Baxby (2007).

A normal FFA activation has also been observed in congenital prosopagnosia (Avidan, Hasson, Malach, & Behrmann, 2005). Even in late-blind people, an FFA reacted when they touched a doll's face; no such activation was seen in congenitally blind people (Goyal, Hansen, & Blakemore, 2006). This may indicate an activation by the mental imagery of faces, which has indeed been observed (Ishai, Haxby, & Ungerleider, 2002). Usually, the right FFA reacts strong to faces than the left, but in some people, the left FFA dominates and in others, no face-specific activity could be shown in the fusiform gyri (Kanwisher, McDermott, & Chun, 1997).

Face-selective areas are also connected to voice recognition areas. In a cross-modal study von Kriegstein, Kleinschmidt, Sterzer, and Giraud (2005) showed that face-selective areas are activated by voices of familiar persons, if, and only if, the task emphasized speaker recognition over content recognition. During this task the FFA was coupled with the STS voice region but not with other cortical regions normally involved in person recognition.

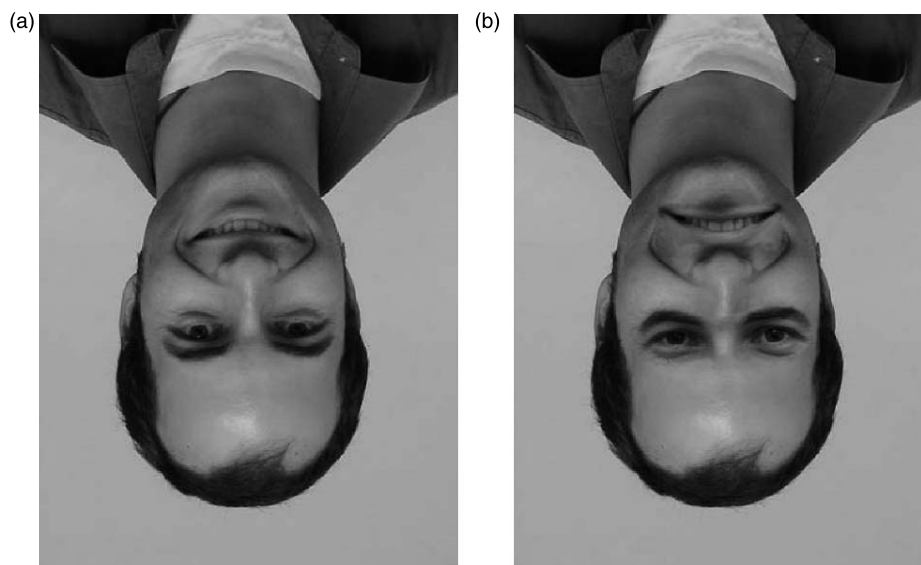
Face learning and face memory involve a widely distributed cerebral network (Ishai *et al.*, 2005) including the middle temporal lobe on both sides, hippocampus, parahippocampal gyri, amygdala, inferior frontal gyrus, and orbitofrontal cortex. Face recognition and learning are influenced by age, emotional expression, orientation, and attention (Aylward *et al.*, 2005; Carbon & Leder, 2006; Carbon, Schweinberger, Kaufmann, & Leder, 2005; Iidaka *et al.*, 2002; Lueschow *et al.*, 2004). Each processing stage is tightly integrated with and related to other tasks like voice recognition or expression evaluation (Ishai *et al.*, 2005). It is being debated, whether a specialized neural network is responsible for face recognition (McKone, Kanwisher, & Duchaine, 2007) or whether face recognition is just one task of a neural structure which processes the individual recognition of homogenous object classes (Buckley, Gauthier, & Tarr, 2006). In the latter case, face recognition would be an object recognition subtask. In order to prove the latter concept, Gauthier and Tarr (1997) constructed artificial objects called 'greebles'. Greebles are club- or column-shaped artificial objects with pins, beaks, or horizontal rings attached at different locations. Similar looking greebles form one family. Using this concept, Gauthier, Tarr and their co-workers defended their idea that face recognition is only an instance of the brain's ability to distinguish very similar objects at expert level (Gauthier, Skudlarski, Gore, & Anderson, 2000). If so, prosopagnosics should have problems to distinguish between similar objects. Duchaine, Dingle, Butterworth, and Nakayama (2004), though, found a normal performance in a prosopagnosic for a visual greeble learning and discrimination task. Also, if face recognition would only be an instance of expert level visual object discrimination, an object agnosia should always imply a prosopagnosia. This has been disproved: Carlesimo, Fadda, Turriziani, Tomaiuolo, and Caltagirone (2001) reported a patient with an acquired global visual object amnesia who could learn and recognize faces at a normal level. These double dissociate findings suggest independent neural structures for face and object recognition at least to a certain extent.

In the 1970s to 1990s, there was a debate whether a right-sided lesion could cause a prosopagnosia, or whether lesions in both occipital and temporal lobes were necessary to disrupt the face recognition system (De Renzi, Perani, Carlesimo, Silveri, & Fazio, 1994; Meadows, 1974). Since then, in several imaging studies, the right FFA consistently shows a stronger activation to faces than its counterpart on the left (e.g. Kanwisher *et al.*, 1997; Maurer *et al.*, 2007). This lateralization may be present from birth. In a PET study with six 2-month-old infants, Tzourio-Mazoyer *et al.* (2002) found that even at this early age, face-processing preferably activated structures in the inferior temporal gyrus on the right side. A recent study by Gonzalez, Anderson, Wood, Mitchell, and Harvey (2007) about the lateralization of memory deficits in children with temporal lobe epilepsy indicated a lateralization of memory deficits only for faces, but for no other verbal or non-verbal memory tasks. A comprehensive meta-analysis of Bouvier and Engel (2006) with 100 cases of acquired prosopagnosia listed 32% unilateral right-sided and only 3% unilateral left tissue defects as a cause of the disorder; 65% of the patients had bilateral damages. Together, these findings indicate that the right lateralization of face processing is present after birth and does not change with age. While both hemispheres can process faces and compensate for damage to the homologous area to a certain degree, the right hemisphere is dominant and a functional defect there may lead to a measurable face recognition deficit.



### Face recognition strategies

Though face recognition is obviously present in newborns already (Bushnell, Sai, & Mullin, 1989), the acquisition of face expertise takes several years (Mondloch, Maurer, & Ahola, 2006). Humans use different strategies to learn and recognize faces: they recognize the facial features (feature based; Carbon & Leder, 2005), the configuration of the features (configural; Leder & Carbon, 2006), and may recognize faces as a complete unit (holistic; Leder & Carbon, 2005; Tanaka & Farah, 1991). The configural recognition strategy works best for upright faces. It fails when faces are turned upside down, the strategy is disrupted (Carbon & Leder, 2006). Peter Thompson (1980) first demonstrated this dissociate face-processing effect with a picture of the former British Prime Minister Margaret Thatcher. By selectively turning the eyes and the mouth region upside down, a grotesque face emerges (Figure 3a and b). When Thatcherized faces are inverted (turned upside down), the alteration is hardly detectable and the perception of grotesqueness vanishes (Carbon, Grueter, Weber, & Lueschow, 2007; Carbon *et al.*, 2005). Another intriguing example of configural face processing was shown by Craig Mooney (1957). 'Mooney faces' are high-contrast photographs in which the face is formed of black shadows and white surfaces concealing any local or featural detail. By inverting them, they are hardly recognizable as faces at all (Figure 4). Valentine (1991) proposed that faces might be stored as vectors in multidimensional space representing the difference from a 'prototypical' face. This model would also explain the so-called other-race effect. Humans recognize faces of people from other races with less confidence than those of their own race. In face-space models, the faces of other races have similar vectors and are difficult to tell apart. The prototypical face is generated from the average of faces in memory. When more other race faces are stored, the standard face is adjusted and the recognition deficit decreases (Sporer, 2001).



**Figure 3.** a) Does one of these pictures look bizarre? Due to disruption of configural face processing, this is difficult to decide when faces are upside down (Carbon *et al.*, 2005). b) When the faces are shown in the normal upright position (please turn Figure 3 upside-down), the left one looks bizarre.



**Figure 4.** Mooney faces: While the left picture appears, more or less, as meaningless black and white areas, the left one is a face. This also depicts the disruption of configural face processing at unusual viewing angles.

### **Dysfunctions of face recognition**

Any neural tissue damage in the face recognition network of the brain can cause a prosopagnosia, a condition which can best be defined as a selective impairment of the visual learning and recognition of faces. As of yet, there is no generally accepted classification. A prosopagnosia caused by an accident or stroke in adulthood is mostly called ‘acquired’ prosopagnosia or simply ‘prosopagnosia’. More confusing are the classification attempts for the early onset prosopagnosia. Prosopagnosia can be inherited or acquired by an early brain tissue damage. The term ‘developmental prosopagnosia’ has been used for both conditions in the past (Barton *et al.*, 2003; Kress & Daum, 2003a). In a study of three patients who acquired prosopagnosia at an early age, Barton *et al.* found that ‘developmental prosopagnosia is similar to the adult-onset form in encoding deficits for the spatial arrangement of facial elements’. Therefore, it is somewhat doubtful if a discrimination between prosopagnosia acquired before or after adulthood makes sense. There is another type of prosopagnosia which has no defined onset and is not accompanied by any visible brain damage (Behrmann & Avidan, 2005). This type has also been called ‘developmental’, while we would prefer the attribute ‘congenital’ to show that it was not acquired at some defined point in life (cf. Behrmann & Avidan, 2005). A detailed discussion of the classification problem can be found elsewhere (Grüter *et al.*, 2007). Hereditary prosopagnosia is a type of congenital prosopagnosia which runs in families, as already documented by De Haan (1999). It is quite common among the congenital cases (Kennerknecht *et al.*, 2006).

For practical purposes, we would also like to define another type, the ‘symptomatic’ prosopagnosia which is part of other diseases like a pervasive developmental disorders (PDD), psychiatric diseases, dementia or congenital impairments of vision (Elgar & Campbell, 2001; Geldart, Mondloch, Maurer, de Schonen, & Brent, 2002; Serra *et al.*, 2003). This classification is not disjunctive and complete in a mathematical sense.



For example, a hypoxia at birth can cause a congenital and acquired prosopagnosia at the same time. Still, the classification covers the vast majority of cases.

### **Acquired prosopagnosia**

An acquired prosopagnosia should not necessarily be considered a disease, but as one of several visually selective residual impairments after a brain tissue damage. It can be defined as a dissociation of face and object recognition impairment to the disadvantage of face processing. The individual range of impairments depends on the extent and the location of the damage, causing a unique pattern of symptoms in each patient. An acquired pure prosopagnosia is rare, only very few cases have been published (Kleinschmidt & Cohen, 2006; Rossion *et al.*, 2003). The widely differing patterns of impairment make it difficult to identify the face-processing brain regions. In a comprehensive meta-study of 100 cases of acquired prosopagnosia, Bouvier and Engel (2006) found a high lesion overlap near the OFA while the FFA and the STS were less often affected. Instead, they identified an area medial to the FFA which was damaged in a number of cases. They cautioned, though, that the image slices used to identify the extent of the damage did not always cover the ventral cerebral surface with the FFA. Therefore, a damage to the FFA may be more frequent than they could demonstrate.

Acquired prosopagnosia is quite rare, most neurologists have told us that they never saw a single case during their professional career. De Renzi *et al.* stressed that 'the inability to recognize familiar faces is a rare disorder, not manifested by the majority of patients with right temporo-occipital injury'. Hier, Mondlock, and Caplan (1983) examined 41 patients within 7 days after a right-sided stroke and diagnosed prosopagnosia in 19 of them, mostly accompanied by more dramatic symptoms like visual field defect, hemineglect, anosognosia (denial of illness) or weakness of arm and leg. Therefore, a temporary prosopagnosia after a right-sided stroke may well be hidden by more apparent symptoms and not be recorded at all.

In some cases of acquired prosopagnosia, a residual autonomic (covert) response to familiar faces has been reported (e.g. De Haan, Bauer, & Greve, 1992; Tranel & Damasio, 1985). Some authors therefore postulated a second pathway for the covert, unconscious recognition of faces (De Haan *et al.*, 1992). Farah, O'Reilly, and Vecera (1993) offered a different explanation which they based on a neural network simulation of face recognition. Stone and Valentine (2003) argued that emotional preference rather than individual recognition may trigger an autonomic response and confound the study results (see also Schweinberger & Burton, 2003).

### **Congenital and hereditary prosopagnosia**

McConachie was the first to report a case of prosopagnosia without any history of brain tissue damage (retested by De Haan & Campbell, 1991). Twenty years later, Ariel and Sadeh (1996) published a second case. By 2002, less than a dozen cases of congenital prosopagnosia were published worldwide, three of these had been found in one family (De Haan, 1999). Other researchers also mentioned affected family members (Behrmann, Avidan, Marotta, & Kimchi, 2005; Bentin, Deouell, & Soroker, 1999; De Haan, Young, & Newcombe, 1991; Duchaine, 2000; Galaburda & Duchaine, 2003; McConachie, 1976; Temple, 1992), but did not assess the formal genetics. From about 2002, prosopagnosia received more public interest. An internet mailing list for prosopagnosics was started by affected persons and some web sites were dedicated to

the condition (e.g. [www.faceblind.org](http://www.faceblind.org), [www.prosopagnosie.de](http://www.prosopagnosie.de)). Duchaine and Nakayama (2005) wrote that within 2 year after they set-up their web site, more than 175 self-diagnosed prosopagnosics contacted them. Our research group systematically investigated the relatives of people with a congenital prosopagnosia and found seven pedigrees with 38 cases of congenital prosopagnosia (Grueter, 2004; Grueter *et al.*, 2007). All pedigrees were in accordance with a simple autosomal dominant mode of inheritance. A survey by Kennerknecht *et al.* (2006) found a prevalence of 2.5% in the German population (17 out of 689) for congenital prosopagnosia. In 14 cases, first-grade relatives were affected as well, three prosopagnosics did not want the researchers to interview family members. This indicates that the hereditary type is quite frequent if not prevailing among the cases of congenital prosopagnosia. It should be noted that hereditary prosopagnosia is the first identified hereditary disorder to affect the central visual cognition system. The familial segregation of hereditary prosopagnosia in a simple autosomal dominant mode suggests that it may be caused by a single mutation in one or more genes (point mutation). This would imply that at least within families, but probably also across families, the underlying defect is the same. This is supported by the quite uniform scope of clinical symptoms in hereditary prosopagnosia. To our knowledge, the responsible gene has not been identified yet.

Though the affected people have a lifetime to compensate for their deficit, a careful interview with a set of standard questions showed a set of typical symptoms (Table 1). The table summarizes the results of semi-structured interviews with 54 people with hereditary prosopagnosia (45 females, 9 males, mean age: 45.0, range 19-92) and 56 controls (35 females, 21 males, mean age: 47.4, range 16-78). The interviews lasted between 1 and 2 hours. The interviewer asked open questions from a questionnaire with at least three or four questions regarding each diagnostic item. Interviewers were held to embed the questions into the conversation and to make sure that questions about the same diagnostic item were not asked in a row. All interviews were done by experienced physicians. This proved to be helpful because questions about face recognition deficits can be embarrassing and most interviewed people played the symptoms down. The diagnostic procedure is also described in Kennerknecht *et al.* (2006). It was validated by face recognition tests in two samples with a total of 21 prosopagnosics (Carbon *et al.*, 2007; Grueter & Grueter, 2007). The test results confirmed the diagnosis of hereditary prosopagnosia in each case without exception. The leading symptom of hereditary prosopagnosia is a lack of confidence about the familiarity of faces. The affected people do not complain that all faces look unfamiliar, but that they cannot determine the familiarity on a valid basis. Therefore, they not only overlook familiar people, but also confuse strangers with familiar persons (T. Grueter & Grueter, 2007). This symptom is found throughout and therefore should be regarded as a diagnostic hallmark. People with hereditary prosopagnosia also take longer to recognize familiar faces and to learn new faces. They cannot remember an onset of the problem. Nearly, all of them find gaze contacts unnecessary for social communication – as did 8 out of 56 controls. Eye-tracking experiments showed a peculiar scan pattern for faces in hereditary prosopagnosia. The gaze was more dispersed and more directed to external facial features (Schwarzer *et al.*, 2007). The development of an adaptive behavioural pattern (avoiding critical situations, a ready set of excuses, etc.) indicates a long-standing perceptual deficit. Nearly, all hereditary prosopagnosics admitted problems with the visual recognition of objects and scenes. This is consistent with the observation by Behrmann *et al.* (2005) in five congenital prosopagnosics with affected family members. They all had problems with the judgment of global

configurations from simple visual stimuli. As configural processing is of foremost importance in face processing (Behrmann *et al.*, 2005; Carbon & Leder, 2005, 2006), a dysfunction will especially impair face recognition. Not all studies, though, confirmed a (general) configural processing problem in congenital prosopagnosia (Duchaine, 2000; Le Grand *et al.*, 2006). In the latter publication, the authors did not find a uniform pattern of visual impairments to accompany the face recognition problem and stated that congenital prosopagnosia is a heterogeneous condition. Of the eight prosopagnosics tested reported in these publications, only one is reported to have affected family members. Therefore, some of them may suffer from congenital prosopagnosia other than the hereditary type.

**Table 1.** Most discriminative symptoms for the diagnosis of prosopagnosia

Symptoms	Prosopagnosics	Controls
Lasting and irritating subjective uncertainty of face recognition	54/54	0/56
Face recognition deficit especially in crowded places or out-of context encounters	54/54	0/56
False-negative and false-positive face recognition events	54/54	0/56
Face recognition time longer than socially accepted	54/54	0/56
Prolonged face learning time longer than socially accepted	54/54	0/56
Onset in childhood	54/54	0/56
Development of adaptive behaviour	54/54	0/56
No gaze contact necessary	53/54	8/56
Use of explicit learning strategies for visual person recognition	53/54	0/56
Impaired visual recognition of objects and scenes	54/54	2/56

In hereditary prosopagnosia, there is no subjective or objective perceptual deficit in the recognition of facial emotions (Duchaine, Parker, & Nakayama, 2003; Humphreys, Avidan, & Behrmann, 2007). While in acquired prosopagnosia, a central achromatopsia and a quadrantanopsia (loss of vision in one quarter of the visual field) is quite frequent (Bouvier & Engel, 2006), it has not been observed in hereditary prosopagnosia. The affected persons also reported that their judgments of attractiveness and gender are not different from other people. They had no problems to recall semantic information about persons and recognized other people easily from non-facial clues (Table 2).

The functional and anatomical peculiarities in congenital prosopagnosia remain unclear to date. A single case study by Bentin *et al.* (1999) showed an unusual N170 wave in the EEG. Normally, the N170 is particularly face sensitive, but in the studied person, it was also strongly elicited by objects. Kress and Daum (2003b) reported a similar lack of N170 face specificity in two more prosopagnosic patients. While Bentin *et al.*'s patient had an unusually small right temporal lobe, both patients described by Kress and Daum (2003b) had no structural brain abnormalities. In an fMRI study with four congenital prosopagnosics, Avidan *et al.* (2005) could not demonstrate conclusive differences in the anatomy or in the FFA activation to faces. A recent structural imaging study by Behrmann, Avidan, Gao, and Black (2007) with six prosopagnosics found them to have a significantly smaller anterior fusiform gyrus, though.

**Table 2.** Cognitive symptoms usually not impaired in prosopagnosics (two prosopagnosics and one control were not asked about facial attractiveness, one control was not asked about gender recognition)

Symptoms	Prosopagnosics	Controls
Normal recognition of facial emotions or emotions in general	45/54	52/56
Unimpaired recognition of gender from faces	51/54	54/55
Normal colour recognition	54/54	56/56
Normal field of vision	54/54	56/56
Normal judgment of facial attractiveness	51/52	54/55
Normal semantic memory for persons	53/54	55/56
Recognition of persons from non-facial clues	53/54	55/56

### Comparison of hereditary and acquired prosopagnosia types

The hereditary and acquired types have a completely different aetiology. The acquired type is the consequence of a localized tissue damage, while the hereditary type is more probably caused by a defective neural development. As most cases of acquired prosopagnosia are caused by occipito-temporal tissue losses, adjacent anatomical structures are quite often damaged as well, though they are not functionally linked to face recognition. This explains the frequent coincidence of acquired central achromatopsia and prosopagnosia. Also, parts of the lateral geniculate nucleus may be damaged, causing a quadrant- or hemianopsia (Bouvier & Engel, 2006). While emotion recognition is always spared in hereditary prosopagnosia, it is frequently defective in acquired prosopagnosia (Sergent & Signoret, 1992; Takahashi, Kawamura, Hirayama, Shiota, & Isono, 1995). Hereditary prosopagnosics show a frequent impairment in the visual recognition of objects and scenes, indicating a general impairment in visual recognition most prominent in face recognition. While acquired prosopagnosia causes a loss of familiarity feeling (Spillmann, Laskowski, Lange, Kasper, & Schmidt, 2000; Verstichel, 2005), people with hereditary prosopagnosia suffer from a lasting lack of confidence (Kennerknecht *et al.*, 2006). They cannot judge familiarity, all faces seem vaguely familiar or vaguely unfamiliar. In her report on the prosopagnosic, woman Dr S. Temple (1992, p. 200) stated about their second meeting: 'She failed to recognize me... until I said her name. She would also report that as she knew I had blonde hair, she had moved expectantly towards several other people before I arrived, thinking them to be me'. Obviously, seeing faces did not trigger the familiarity decision at all in Dr S. Temple. She cannot even reliably classify unknown faces as unfamiliar. This may lead to false recognition of unknown people as familiar, and indeed did all hereditary prosopagnosics report those events in our interviews. Their familiarity feeling *per se*, though, is not impaired, it can reliably be triggered by familiar voices, for example. False recognition is not common in acquired prosopagnosia. Rapcsak and his colleagues (1996) studied a group of 21 patients with right hemispheric damage and found no significant association between false recognition and prosopagnosia severity.

Nearly, all hereditary prosopagnosics told us that they do not need no gaze contact in social interaction. Many added that they use gaze contact nevertheless because they noticed that other people expect it. Still, the unusual gaze behaviour is among the most striking symptoms for the interviewer. Many prosopagnosics remember having been criticized by schoolteachers for not listening, because they did not look at the teachers. We could not find any data about unusual gaze behaviour in acquired prosopagnosia,

though. No paper mentioned a sudden change of gaze behaviour in acquired prosopagnosia. As the symptom is quite obvious, we would assume that it would have been mentioned somewhere.

Hereditary prosopagnosia is accompanied by a well-defined pattern of symptoms not always related with 'pure' prosopagnosia. False recognition, a familiarity decision deficit, no need for gaze contact in social interaction, and a general visual recognition impairment define this condition, while other capabilities like emotion recognition are not affected. Functionally independent but anatomically adjacent structures are never affected, ruling out a blood vessel malformation or a birth trauma as a cause of the condition (Table 3). Point mutations have been shown to cause pervasive functional disorders of the brain (Lai, Fisher, Hurst, Vargha-Khadem, & Monaco, 2001). The unique symptom pattern and the simple autosomal way of inheritance in all reported hereditary prosopagnosia cases suggest a functional brain development disorder caused by a point mutation. This offers a twofold opportunity: firstly, finding the gene (or the genes) would increase our knowledge about the genetics of brain development. Secondly, as the underlying defect is the same at least within families and probably across families, a comprehensive study with a large number of hereditary prosopagnosics promises new insight into cerebral face processing.

**Table 3.** Difference between acquired and hereditary prosopagnosia

Symptoms	Hereditary PA	Acquired PA
Feeling of familiarity	Not triggered (lasting uncertainty)	Lost, no feeling of familiarity
Colour blindness	Not associated	Frequently associated
Quadrantanopsia	Not associated	Frequently associated
False-negative and false-positive recognition events	Always present	Inconsistent, rare
Emotional expression recognition	Normal	Inconsistent
Gaze contact necessary	No	No data
Impaired visual recognition of objects and scenes	Always present	Inconsistent

### **Prosopagnosia in autism spectrum disorders (ASDs) and social developmental disorders (SDDs)**

Prosopagnosia has been suggested to be linked to ASD in two ways. ASD may cause prosopagnosia because the lack of social interest may prevent the acquisition of facial experience. In this case, the face recognition structures themselves would work at a normal level, but not be employed sufficiently. On the other hand, a failure to extract sufficient information from faces may interfere with normal social development. The same could happen, if the brain cannot build up the normal facial attention preference levels, because the corresponding structures are abnormal (Sasson, 2006). Schultz (2005) suggested that a defective function of the fusiform face area and the amygdala may represent a 'core mechanism' for the pathobiology of autism.

This view is highly controversial. Teunisse and de Gelder (2003) studied face recognition abilities in a group of 17 high-ability autistic adults and found that they formed a normal configuration-based face representation. People with a congenital

prosopagnosia are known to have a deficit there (Carbon *et al.*, 2007). In a comprehensive study, Barton *et al.* studied the face recognition performance of 24 people with SDD in comparison with neurotypical and prosopagnosic people. Their results showed, as they wrote, that ‘SDD subgroup membership by face recognition did not correlate with a particular SDD diagnosis or subjective ratings of social impairment’. They concluded that SDD does not necessarily cause a face recognition impairment (Barton *et al.*, 2004). Hadjikhani and her colleagues (2004) compared the FFA activation in normal and autistic adults. They assumed that the earlier described FFA activation deficit in autistic persons (Schultz *et al.*, 2000) might be caused by an attentional deficit and used a fixation point to make sure that the faces received attention. With this experimental setting, they found no significant differences in FFA activation to faces between autistic and neurotypical study participants. It should also be noted that the prevalence of congenital prosopagnosia (2.5%) is much higher than that of ASD (<0.5%; Yeargin-Allsopp *et al.*, 2003).

As of today, a causal dependency of SDD or ASD and face recognition abnormalities cannot be considered as established. The aetiology and pathogenesis of ASD remain elusive (for a review see Santangelo & Tsatsanis, 2005).

## Conclusions

Face processing plays a decisive role in effective social interactions. As it has not been designed or engineered, it is highly efficient, but not necessarily structured in a simple or logical way. Current research results show that a complex interplay between different highly face-responsive brain areas is involved in encoding, integration, and interpretation of the huge amount of different information we draw from a face (Gobbini & Haxby, 2007). Some structures in the occipital (OFA) and temporal lobes (STS, FFA) have been found to be specialized on face recognition. Tissue damage in these regions lead to a lasting face-processing deficit, called (acquired) prosopagnosia. In contrast to many other visual abilities, the brain’s functional plasticity cannot compensate for this deficit. While the acquired type is rarely observed, the congenital type without any gross brain abnormalities is quite frequent. In this condition, deficits in face processing are apparent from early childhood on while other sensory and intellectual functions are preserved and macro-spatial brain abnormalities are discreet or absent (Behrmann & Avidan, 2005). Only recently, a hereditary type of congenital prosopagnosia (De Haan, 1999) was identified. The segregation is compatible with a simple autosomal dominant way of inheritance suggesting a point mutation as a cause of defect (M. Grueter, 2004). First studies indicate a very high prevalence (2.5%) of hereditary prosopagnosia (Kennerknecht *et al.*, 2006). The condition is characterized by a great homogeneity of clinical symptoms, including deficits in learning and recognition of faces, while other facial information is processed normally (e.g. expression and gaze). Obligatory symptoms are, for instance, lasting and irritating subjective uncertainty of face recognition, time-inefficient recognition strategies, and false-positive and -negative face recognition events (see Table 1). In the light of such impairments, apologetic, compensating, and preventing strategies are typically and intuitively developed by people with prosopagnosia. The high prevalence of hereditary prosopagnosia demonstrates its societal importance. Face research will have to analyse the neurocognitive bases and develop better diagnostic criteria. This will create the pre-condition for educational and training programs in a sensitized society. Face recognition is a complex and highly integrated task employing large parts of the brain. A unitary and



common impairment of face perception like the hereditary type of prosopagnosia opens a big window of opportunity to improve our knowledge about face processing and its genetic base.

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