Chapter title: **Developmental prosopagnosia: Cognitive, neural, and developmental investigations**
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MJ, a seven-year-old boy, was an enigma to his mother. At home or when playing with unfamiliar children in public places, he was friendly and engaging. At school though he was a loner who tended to watch the other children play. His teachers regularly became exasperated with him when he failed to follow their instructions. MJ refused his teacher's request to take papers to a particular student, claiming that he didn't know who the student was. When told to stand next to a student named Casey, MJ stood next to the student he thought was Casey but his teacher became angry and she sent him to the end of the line. MJ's mother's claims that he was normal at home were met with disbelief by his teachers, and MJ's principal told his mother that his social problems were probably caused by her anxiety.

There were other oddities as well. MJ wasn't able to recognize the small number of neighbors they met regularly. He also confused his mother and his beloved aunt when they had similar hairstyles. When his aunt dramatically changed her hairstyle, MJ refused to believe that she was really his aunt, and he was mad at his mother for several days when she changed her hairstyle. In preschool, the only student MJ could identify was a Chinese girl -- the only non-Caucasian student in the class. His mother thought MJ seemed overly obsessed with body weight and skin color, and he embarrassed her several times by referring to a child as "the fat boy" or "the brown girl".

MJ's one friend in kindergarten was a boy named Jacob. Upon running into Jacob and his parents at a soccer game, MJ failed to recognize him. The adults laughed off MJ's inability to recognize Jacob, but MJ's mother could see the same confused look on his face she'd seen many times before -- a lack of recognition coupled with the look of someone who thinks they're being duped. Then, a couple of weeks later, MJ's older brother saw one of his former classmates at church. His name was also Jacob and he was roughly the same size and coloring as MJ's friend Jacob. Upon seeing his brother talking to this Jacob, MJ clearly thought this boy was his friend Jacob. His mother was astounded, andthe realization hit her like a ton of bricks. MJ could not recognize people!

Background

MJ suffers from developmental prosopagnosia (DP), a condition defined by severe face recognition problems resulting from a failure to develop the necessary visual recognition mechanisms. Although it has been long recognized that severe face recognition deficits can follow brain damage (Bodamer 1947; Wigan 1844), wider awareness of DP has only emerged in the last ten years. In the literature DP is also often called *congenital prosopagnosia*, but here I will refer to it as *developmental prosopagnosia* because it is unclear whether face recognition abilities in DPs are atypical at birth. Face processing problems are also seen in other developmental disorders such as autism spectrum disorder (Dawson *et al*. 2002) and Turner syndrome (Mazzola *et al*. 2006), but in DP intellectual function and social cognition is normal (Duchaine *et al*. 2010) and deficits to other abilities are limited.

Bornstein briefly described cases that appear to be DPs in a chapter on prosopagnosia (Bornstein 1963), but McConachie (1976) was the first to publish a case study of a DP. A.B. was an intelligent 12-year-old girl who reported having severe difficulties recognizing faces that she was not extremely familiar with. She found recognition of her uniformed classmates especially challenging. Despite her reported

difficulties, A.B. was able to hesitantly identify photographs of familiar faces and also scored normally on a test of unfamiliar face recognition. A.B. and her mother were unaware of any events that may have caused brain damage. Interestingly, A.B.'s mother also reported face recognition problems which suggested a possible genetic cause. In a follow-up study 15 years later, A.B. showed clear difficulties in tests of face recognition as well as deficits with the recognition of facial expressions and within-class object recognition (de Haan and Campbell 1991).

A few new cases of DP were reported in the 1990s (Bentin *et al*. 1999; Kracke 1994; Temple 1992), but it wasn't until the current decade that substantial numbers of prosopagnosics were investigated (Behrmann *et al*. 2005; de Gelder and Rouw 2000; Duchaine, 2000; Kress and Daum 2003). Awareness of the condition and research into it have benefitted greatly from the emergence of the internet. An email discussion group was created in late 1990s by a group of DPs, and Bill Choisser, an American DP, published an online book in 1997 about the condition and his experiences of it (Choisser 1997). The internet has also provided a means for researchers and DPs to make contact. Several groups have websites aimed at recruiting research participants, and more than 5200 DPs have contacted the website Ken Nakayama and I created in 2002 (http://www.faceblind.org). Approximately 95% of these self-reported prosopagnosics are unaware of suffering any brain damage so severe face recognition problems appear to be much more often due to developmental problems than brain damage in adulthood (Duchaine and Nakayama 2006). An estimate of the prevalence of DP based on self-report and interviews suggested that approximately 2% of the population experience significant face recognition difficulties in everyday life due to developmental problems (Kennerknecht *et al*. 2006). That the prevalence should be so high for such an ancestrally important ability is surprising, but it bears noting that modern environments place much greater demands on face recognition than ancestral environments did. Many people who experience difficulties recognizing the thousands of faces one encounters in modern life may not have experienced significant difficulties in ancestral environments.

Herein I will review recent research findings investigating DP. Like studies of acquired prosopagnosia, studies involving DP have addressed the cognitive and neural basis of face processing. However because DP is developmental in origin, it also holds promise as a means to better understand the developmental and the genetic basis of face processing. While our understanding of DP remains poor, the relatively rich cognitive, neural, and developmental theories of face recognition provide a framework that should allow rapid progress. In addition DP research may provide a model for investigating other selective developmental disorders. At present, only a handful of selective developmental disorders have been identified (Van Zandvoort *et al*. 2007; Garrido *et al*. 2009; Iaria *et al*. 2008; McCloskey *et al*. 1995; Ramus 2003; Bishop 2006; Temple and Richardson 2004), but the late recognition of a developmental deficit affecting an ability as critical as face recognition suggests that other, possibly many other, selective developmental deficits may exist.

Experience of Developmental Prosopagnosia

Having lost their face recognition abilities due to brain damage, it is usually apparent to acquired prosopagnosics that they have face recognition deficits. DPs however are

unable to appreciate first-hand that their face recognition abilities are deficient. Nearly all of the people who my laboratory work with were aware that they sometimes had trouble recognizing people but most did not attribute it to problems in their visual system. Many believed they were not trying hard enough or were not sufficiently interested in people. Jane Goodall, who has stated that both she and her sister are prosopagnosic, wrote:

I used to think it was due to some mental laziness, and I tried desperately to memorize the faces of people I met so that, if I saw them the next day, I would recognize them. I had no trouble with those who had obvious physical characteristics -- unusual bone structure, beaky nose, extreme beauty or the opposite. But with other faces I failed, miserably. Sometimes I knew that people were upset when I did not immediately recognize them -- certainly I was. And because I was embarrassed, I kept it to myself. (Goodall and Berman 1999)

Like Dr Goodall, many DPs tested in my lab didn't recognize that they were prosopagnosic until they were adults.

Many DPs are slow to realize that they have difficulties with faces because they compensate for their prosopagnosia by relying on the many other cues available for person recognition. DPs report using hairstyles, voices, body shape, gait, and even characteristic facial expressions. Context is especially important for many DPs. DPs might recognize co-workers in the office where they are expecting to see them but have little hope of recognizing them when meeting them in the grocery store. Although they have severe problems with facial information, most DPs report that they do use the face for person recognition and their scores on famous face tests, although far worse than controls, show they can perceive and store some facial information (Behrmann *et al*. 2005; Duchaine and Nakayama 2005; Duchaine *et al*. 2007b).

Not surprisingly, face recognition failures can create substantial difficulties for DPs. Their failure to acknowledge friends and acquaintances sometimes causes DPs to be seen as aloof or arrogant, and many of our DPs have discussed episodes in which their recognition failures have had major personal or professional consequences. For example, a DP who contacted our website stated:

I was a public high school English teacher for ten years. I made all of my students sit in assigned seats the whole year. When the school required all students to wear navy blue polo shirts and khaki pants, I was adrift in a sea of blue shirts. The students felt like I disliked them because I didn't know who they were in the hall, even if class had just let out. The lunchroom was social hell. I couldn't even recognize the other faculty members, so I usually stayed in my classroom and locked the door.

A recent study in which 25 DPs were interviewed found that many reported chronic anxiety about offending others and most reported fear and avoidance of social situations in which face recognition would be challenging (Yardley *et al*. 2008). Many reported that DP had significant occupational ramifications, but most of the DPs did not view their prosopagnosia as debilitating but saw it instead as taxing.

A number of other deficits are commonly seen in DP, but it is important to note that dissociations between face recognition and most of these associated conditions have been reported. These associated deficits are similar to those often seen in acquired prosopagnosia which suggests that the two types of prosopagnosia involve deficits to similar mechanisms (Duchaine and Yovel 2008). DP is defined by deficits with facial identity, but other aspects of face processing are also often impaired in DP including face detection (Garrido *et al*. 2008), expression recognition (Duchaine 2000; Duchaine *et al*. 2006; Garrido *et al*. 2009), gender discrimination (Duchaine *et al*. 2006), and trustworthiness judgments (Todorov and Duchaine 2008). Another type of social perception, biological motion perception, is also sometimes impaired in DP (Lange *et al*. 2009). DPs have not shown deficits with basic-level object recognition (Duchaine *et al*. 2006), but many have difficulties with individual item object recognition (often referred to as within-class recognition)(Behrmann *et al*. 2005; Duchaine *et al*. 2003a; Duchaine and Nakayama 2005). Although it has yet to be formally documented, many DPs report difficulties with large-scale navigation (Duchaine *et al*. 2003b).

I'm often asked what DPs experience when they view a face. Although this is not a question that can be answered confidently, their performance on face perception tasks relative to people with normal face perception supports one possibility. Figure 1 shows an example item and upright and inverted scores on a test requiring participants to sort simultaneously presented faces in terms of their similarity to a target face (Duchaine *et al*. 2007b). In Figure1B, the normal participants tend to cluster around the line which shows scores for which participants have made twice as many errors with inverted faces as with upright faces. The DPs however show a different pattern. Their scores are shifted primarily to the left because they made far more errors with upright faces than controls. Their scores are shifted primarily to the left because they made far more errors with upright faces than controls. Their scores are shifted primarily to the left because they made far more errors with upright faces than controls. Many DPs cluster around the line showing participants with equivalent errors with upright and inverted faces. The similarity of their scores with upright and inverted faces suggests that these DPs may process the upright and inverted faces with the same procedures (see also Behrmann *et al*. 2005; Nunn *et al*. 2001). In addition, the similarity of the DPs' upright scores and the normal subjects' inverted scores raises the possibility that DPs' experience of upright faces is similar to the percept that normal subjects experience when viewing inverted faces (See Figure 1C). For people with normal face perception, an inverted face is clearly a face, but the percept is not as rich as the percept of an upright face – the face's identity is not as apparent and its expression and attractiveness are more difficult to discern. If one imagines attempting to interact with a room full of inverted faces, it is easy to appreciate how challenging social situations can be for DPs.

Experimental studies of DP

The leading models of face processing (Bruce and Young 1986; Gobbini and Haxby 2007; Haxby *et al*. 2000) suggest that face processing is carried out by a hierarchically organized network of mechanisms, and recent neurophysiological work in macaques has definitely demonstrated that face processing involves a richly interconnected set of areas (Tsao *et al*. 2006; Moeller *et al*. 2008; Tsao *et al*. 2008; see Freiwald and Tsao this

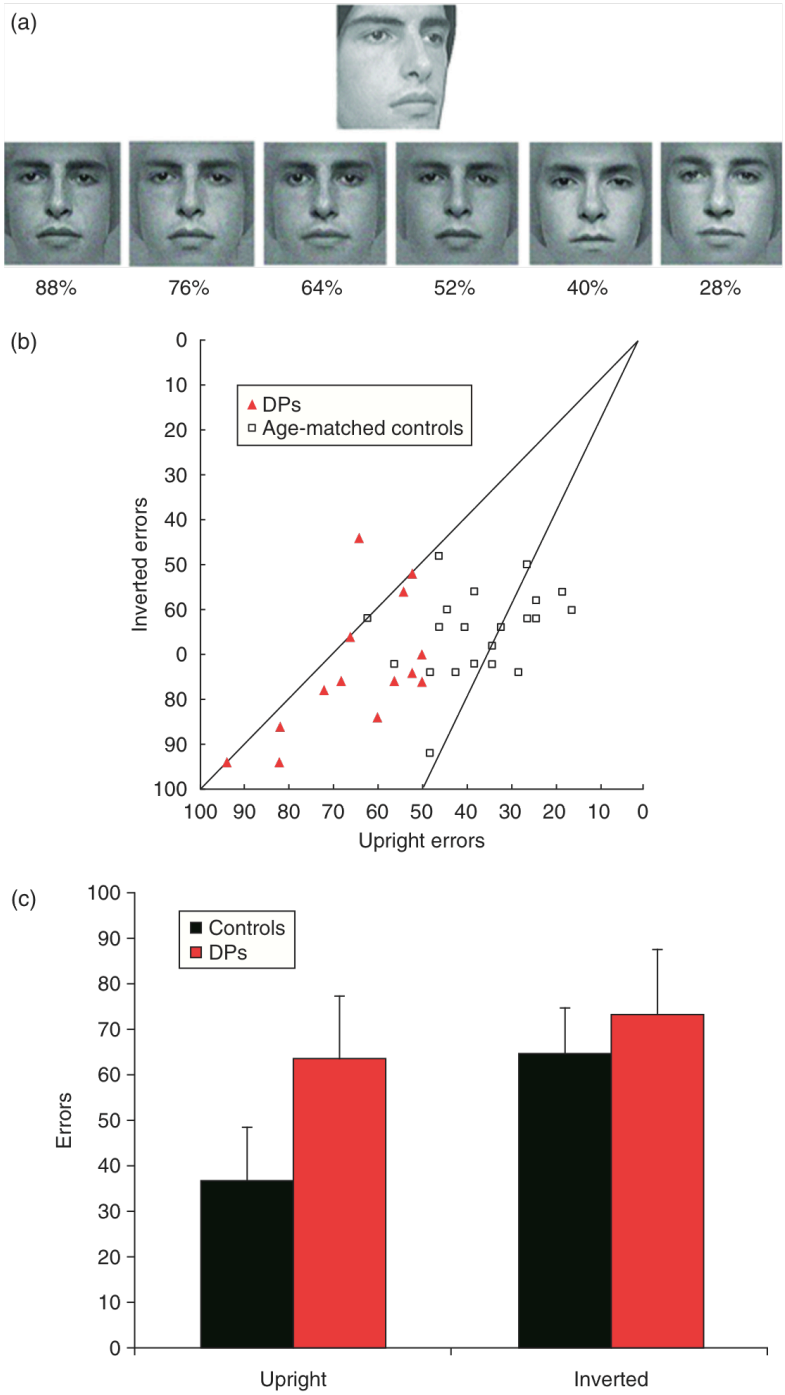

Fig. 42.1 Cambridge Face Perception Test (Duchaine et al., 2007b). (a) Example CFPT item. Participants have 1 min to sort the six faces in terms of their similarity to the target face above. Each participant sorted eight upright items and eight inverted items. (b) Individual scores on the CFPT for controls (black symbols) and DPs (red symbols). Each symbol represents the sum of errors for a participant on the upright items and the inverted items. Controls tend to cluster around the line on the right denoting scores for which inverted errors are twice the upright errors. Many DPs however cluster around the other line denoting equivalent upright and inverted errors. (c) Mean errors for controls and DPs on upright and inverted items. The DPs made as many errors with upright faces as the controls made with upright faces. Reproduced with permission from Social Cognitive Affective Neuroscience ©Oxford University Press. C42.F1

volume). Given the many areas and connections that play a role in face processing, there are undoubtedly many different types of potential face processing deficits. Developing a taxonomy of face processing deficits in DP and an understanding of the developmental events that lead to them is a major challenge, and studies investigating DP so far have addressed relatively simple issues.

Cognitive studies of DP

A range of cognitive issues have been addressed in DP in recent years including the long debated question of whether the brain contains face-specific mechanisms (Diamond and Carey 1986; Farah 1996; Moscovitch *et al*. 1997; Rock 1973; Yin 1969). Like many acquired prosopagnosics, many DPs have difficulties with challenging object recognition tasks (Behrmann *et al*. 2005; Duchaine and Nakayama 2005; Garrido *et al*. 2008). For example, eight out of 14 DPs in a recent study showed impairments on at least one of the seven old-new tests requiring discrimination of a set of similar objects from within a category and five of these DPs were impaired on four or more object tests (Garrido *et al*. 2008). Six of these DPs however showed no deficits on tests of object recognition, and other DPs have also shown dissociations between impaired face recognition and normal object recognition (Duchaine and Nakayama 2005; Nunn *et al*. 2001; Yovel and Duchaine 2006). The examples of apparently pure DP imply that the opposite dissociation may also exist in developmental cases, and my colleagues and I have recently reported a woman with no evidence of brain damage who has normal face recognition and impaired object recognition (Germine *et al*. in press).

While these dissociations between faces and objects are consistent with the possibility that DP can result from deficits to face-specific mechanisms, they do not provide definitive support for it. Many cognitive accounts of prosopagnosia have been proposed over the years, and most of these hypotheses are compatible with certain dissociations between face and object recognition. To more thoroughly address the nature of the mechanisms impaired in DP, my colleagues and I tested the predictions of all the alternatives to the face-specific hypothesis in a case that had shown good performance on a number of non-face visual recognition tests.

Edward was a man in his early 50s with no history of brain damage. He recalls difficulties with faces dating back to childhood. Several aspects of Edward's face processing are impaired including identity, expression, gender, and attractiveness (Duchaine *et al*. 2006) which suggests that his deficits begin early in the face processing stream. An MRI showed no lesions or obvious abnormalities. Two separate fMRI sessions failed to find any face-selective voxels whereas all controls run with the same localizer scan showed areas of face-selective activation. Edward's normal performance on individual item object recognition showed that he did not suffer from a general problem involving within-class visual recognition (Damasio *et al*. 1982). Contrary to the predictions of an account proposing a general deficit with configural information (Levine and Calvanio 1989; Behrmann *et al*. 2005), Edward discriminated changes to the spacing of parts of houses normally while scoring near chance on a matched face task. He scored normally on two tests of inverted face matching yet was impaired when the faces were upright. His normal performance with inverted faces demonstrates that Edward's problems with upright faces are not due to an inability to represent stimuli with particular

properties such as non-decomposability (Farah 1990) or surface curvature (Kosslyn *et al*. 1995; Laeng and Caviness 2001). Expertise accounts of prosopagnosia suggest that face recognition deficits are due to problems with mechanisms that apply special procedures to stimulus classes with which an observer has substantial experience (Diamond and Carey 1986; Gauthier *et* al. 1998). One view of expertise proposes that expertise can be developed relatively quickly. To test whether Edward's prosopagnosia results from problems with the development of what we called rapid expertise we had him carry out a training procedure identical to those used to investigate rapid expertise. This training involved learning to identify 20 individual computer-generated greebles and also the family that each greeble belonged to (the five families were characterized by their body shapes). Edward's performance with greebles was comparable to the controls so his prosopagnosia does not appear to result from a rapid expertise deficit (Duchaine *et al*. 2004). Finally, to test whether he has general deficits with extended expertise (Diamond and Carey 1986), we tested Edward with sequential matching tasks involving faces and human bodies. Bodies were chosen because several studies have shown that bodies, like faces, show substantial inversion effects (Reed *et al*. 2003; Reed *et al*. 2006; Yovel *et al*. in press) which suggests that upright bodies receive special processing. Contrary to the predictions of the extended expertise view, Edward scored normally with the body task while showing a clear impairment with the face task. Taken together, Edward's results were inconsistent with all of the alternative account and so suggest that his prosopagnosia is caused by deficits to a face-specific mechanism (Duchaine *et al*. 2006). At present, Edward is the only prosopagnosic to have been tested in this manner so it will be important to see if future studies with acquired and developmental prosopagnosics find similar results (Duchaine and Garrido 2008).

Edward did not have general problems with configural information, but two papers found evidence that general configural deficits may give rise to DP. In global-local tasks, participants are presented with a stimulus that has information at both the global level and the local level (Navon 1977). One of the most common global-local stimuli consists of the same small letters (local) positioned to form a large letter (global). The local stimulus and global stimulus are usually one of two possibilities (e.g.-S or H), and on a trial, the local and global letters can be either consistent or inconsistent. Participants can be asked to respond to the stimulus at one level or the other, and normal participants usually respond faster on global decisions than local decisions. In addition, trials with consistent global and local letters are usually responded to more quickly than trials with inconsistent letters due to interference from the to-be-ignored letter. However in one study with five DPs, the DP group were slower with global decisions than controls, experienced greater interference from inconsistent local information, and less interference from inconsistent global information than controls (Behrmann *et al*. 2005). In contrast, their performance with local letters was normal. Similarly, KW, another DP, was faster with local than global decisions and also showed larger interference from inconsistent local information and no interference from inconsistent global information (Bentin *et al*. 2007). However atypical non-face global processing is not seen in all DPs. A group of 14 DPs showed no signs of deficits on a global-local task (Duchaine *et al*. 2007a), and eight failed to show problems with a global form task (Le Grand *et al*. 2006). In addition, eight DPs performed normally on a task requiring discrimination of spacing changes in houses yet failed a comparable task with faces (Yovel and Duchaine 2006).

This group showed comparable deficits for spacing and part discrimination in faces which indicates that their problems with faces extend beyond what is usually considered global processing.

Studies of DP have also examined performance on different aspects of face processing and have used these results to make inferences about cognitive organization within face processing. Any recognition system that includes face-specific processes requires a means to detect the presence of faces in a visual scene. This process of face detection has received considerable attention in machine vision (Viola and Jones 2004), but limited attention in human studies (Lewis and Edmonds 2005; VanRullen 2006). To investigate face detection in DP, 14 DPs were tested on two tasks requiring rapid detection of faces (Garrido *et al*. 2008). One task used stimuli consisting of a 5x5 array of greyscale images while the other used two-tone stimuli like the one shown in Figure 2a. Participants were asked to press a key as soon as they saw a face, and a substantial number of no-face trials were included in each task. Many of the DPs were less accurate and slower than controls, including several who showed deficits in both accuracy and response time. Figure 2B presents data for each DP and control on the task involving two-tone images. Note that, although many of the DPs were impaired, several scored normally and so appear to have normal face detection processes (Garrido *et al*. 2008). Eight DPs in another study also showed normal detection of Mooney faces (Le Grand *et al*. 2006).

The models of face processing mentioned above include mechanisms specialized for different aspects of face processing. In Bruce & Young's model (1986), the perception of identity, expression, and facial speech (for lip-reading) depend on separate modules, and Haxby and colleagues' neurocognitive model (2000) proposes that invariants aspects of faces such as identity and gender are processed in the fusiform face area (FFA) and changeable aspects of faces such as expression and gaze are processed in the face-selective region of the superior temporal sulcus (STS). However evidence in support of these divisions is limited. An influential review suggested that no unequivocal support for the distinction between identity and expression processing has been reported (Calder and Young 2005), and support for the other divisions is even weaker. Although neuropsychological dissociations played an important role in the divisions of these models of face processing, many of the studies suffer from methodological limitations (Calder and Young 2005; Duchaine and Weidenfeld 2003).

For most face processing abilities that have been investigated, some DPs show normal performance whereas others show impaired performance. Above I mentioned that Edward has expression recognition difficulties, and other DPs have also shown deficits with expression (Duchaine 2000; Duchaine *et al*. 2003b). Several laboratories however have reported DPs with normal expression perception (Bentin *et al*. 2007; Duchaine *et al*. 2003b; Garrido *et al*., 2009; Humphrey *et al*. 2007; Nunn *et al*. 2001). Although a thorough investigation of gender discrimination in DP remains to be carried out, it appears that some DPs perceive gender normally (Behrmann *et al*. 2005; Nunn *et al*. 2001) whereas others do not (Behrmann *et al*. 2005; de Haan and Campbell 1991; Duchaine *et al*. 2006). Some DPs who have contacted my laboratory have complained of difficulty reading eye gaze, but discrimination of eye gaze direction and perceptual adaptation to left and right gaze was normal in six DPs (Duchaine *et al*. 2009). Similarly, some DPs were normal and others were impaired in a task requiring that they sort faces

based on attractiveness (Sadr *et al*. 2005), and two DPs made normal trustworthiness judgments whereas two DPs made judgments that were only weakly correlated with the judgments of controls (Todorov and Duchaine 2008).

The dissociations between identity and these abilities fits well with modular models of face processing, but our understanding of the relationship between the mechanisms carrying out these computations will benefit from a thorough examination of the dissociations and associations seen in a large group of DPs. Such an investigation may show, for example, that some closely related abilities (e.g.-gender and attractiveness) do not dissociate which would indicate that they rely on the same mechanisms. The dissociations between aspects of face processing may also provide some insight into the development of face processing mechanisms. The normal development of certain aspects of face processing in combination with deficits for the processing of facial identity indicates that the acquisition of these abilities depends on different developmental processes. However the unidirectional dissociations (impaired identity/normal ability with another aspect of face perception) identified so far leave open the possibility that the development of the mechanisms necessary for identity perception are more vulnerable to developmental disruption than the mechanisms that appear to be functioning normally. As a result, the identification of double dissociations in DPs would be valuable in that it would strongly suggest different developmental processes are involved in acquiring different face processing abilities.

A distinction between the perceptual representation of a face and its representation in memory is a common feature in most models and of facial identity recognition (Bruce and Young 1986; Duchaine and Nakayama 2006). Surprisingly little research has investigated this division in individuals with developmental face recognition deficits, probably because most DP research is motivated by questions about high-level vision and so requires that DPs have deficits to perceptual processes contributing to identity recognition. Dissociations between normal perception and impaired memory have been reported in acquired prosopagnosia (Tippett *et al*. 2000), and my laboratory has tested a number of participants who score normally with identity perception but have severe deficits with identity memory.

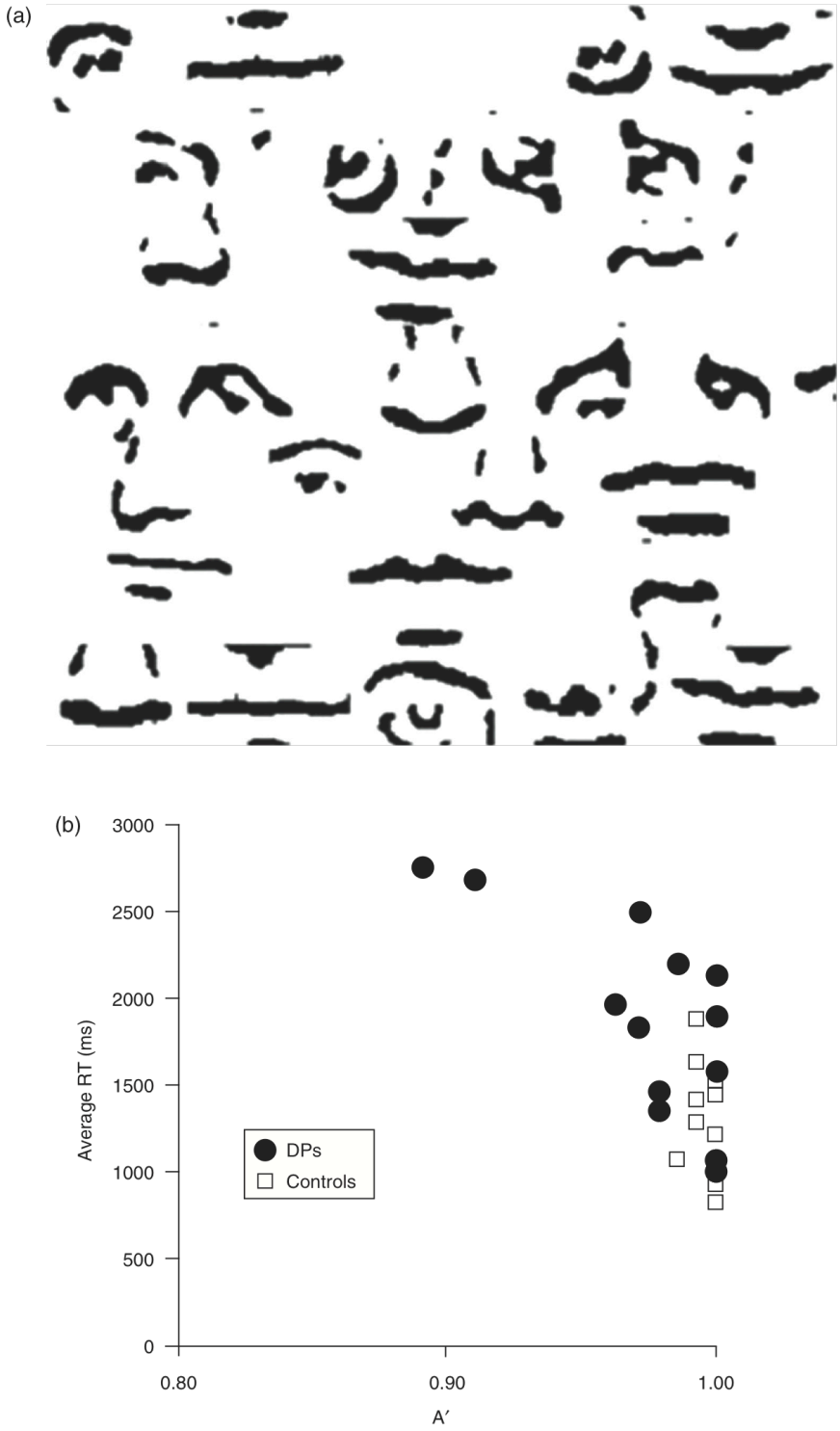

Fig. 42.2 Face detection (Garrido et al., 2008). (a) Example of a face present item in the two-tone face detection task. Face absent stimuli contained the same elements but the black regions creating the face were scrambled. The importance of orientation in face detection can be experienced by viewing the stimulus upside-down. (b) Each symbol shows performance for individual DPs (black circles) and controls (open squares). Reproduced with permission from *Journal of Neuropsychology*

Neural studies of DP

Research into the neural basis of face processing has flourished in recent years, and neuroimaging and neurophysiology have identified several face-selective responses (Bentin 1996; Kanwisher *et al*. 1997; McCarthy *et al*. 1997). Nevertheless identifying atypical neural markers in DP has been challenging because the neural differences between DPs and controls are often not apparent. One reason for this difficulty is that participants with normal face processing show varied fMRI responses and event-related potentials (ERP) to faces so substantial differences are necessary to reveal abnormalities in small samples of DPs. The larger samples in several recent papers provide increased power for group comparisons, but it would be advantageous if methods were developed that allowed stronger inferences from single cases.

In the first fMRI paper published with a DP, YT showed a normal response in left fusiform face area (FFA), right FFA, and right occipital face area (OFA) (Hasson *et al*. 2003). However in the left lateral occipital cortex, which houses the left OFA as well as an object-selective area, YT showed reduced selectivity for faces in the face area and for objects in the object area. Two of the paradigms used in the YT paper as well as two new paradigms were used to assess face-selective activity in four DPs (Avidan *et al*. 2005). In all paradigms, the DPs showed much stronger activation to faces than objects in the fusiform gyrus and also showed normal reduction of this response when faces were repeated (repetition suppression) in face-responsive regions in the fusiform gyrus and the lateral occipital cortex. Given the important role that these face-selective activations in ventral visual cortex are believed to play in face processing, these results were considered a surprise by many and suggested that this behaviorally striking developmental deficit results from either subtle neural abnormalities to these areas or impairments in other areas. Several single case studies however have found functional differences between DPs and controls. Edward, the DP discussed above, failed to show any face-selective voxels in two separate face localizer scans (Duchaine *et al*. 2006), and KW also showed no face-selective voxels in ventral temporal cortex (Bentin *et al*. 2007). Interestingly, these two DPs show very different behavioral abilities. Edward fails with a wide range of face processing tasks whereas K.W. performs normally with expression matching despite severe deficits with identity. SO showed a normal right FFA, but a weaker response to faces relative to objects at the left FFA and the temporal poles bilaterally than controls (von Kriegstein *et al*. 2006). Although Avidan *et al*. (2005) found normal adaptation to faces in four DPs, a recent paper did find atypical adaptation in one DP (Williams *et al*. 2007). This DP participant, referred to as C, shows an unusual pattern of abilities. She was impaired on tests of identity perception and unfamiliar face memory, but was able to successfully identify famous faces and reported that she was able to recognize faces after considerable exposure. When scanned she showed normal FFAs bilaterally and also bilateral parahippocampal place areas (PPA). Like controls, C showed adaptation to repeated familiar and unfamiliar places in PPA and also to repeated familiar faces in FFA. However consistent with her behavioral results, she failed to show a weaker response to repeated unfamiliar faces.

Neuroimaging has also been used to examine whether DPs have structural differences from controls. Volumetric MRI analysis with YT, one of the DPs who showed a normal fMRI response to faces in the ventral occipito-temporal cortex (Hasson *et al*. 2003), found that his temporal lobe was significantly smaller than controls (Bentin

et al. 1999). A study with six DPs also found temporal lobe abnormalities (Behrmann *et al*. 2007). This analysis measured the volume of regions in the temporal lobes and found that the anterior fusiform gyrus was smaller in DPs than controls. The FFA is seen in regions of the posterior and mid-fusiform gyrus, so it is posterior to anterior fusiform (Behrmann *et al*. 2007). Interestingly, both the anterior and the posterior middle temporal gyrus were larger in DPs than controls. A recent study using voxel-based morphometry (VBM) found that DPs had less grey matter than controls in a number of regions that show face-selective responses (Garrido *et al*. 2009). Analyses of the T1-weighted images revealed that the 17 DPs had reduced grey matter volume in right anterior inferior temporal lobe and in the STS/middle temporal gyrus bilaterally. A separate analysis of the segmentation of magnetization transfer images also showed less grey matter volume in DPs in right middle fusiform gyrus and inferior temporal gyrus.

Diffusion tensor imaging (DTI) done in the six DPs in Behrmann *et al*. (2007) identified deficiencies in white matter fibers that connect ventral occipito-temporal regions with more anterior regions (Thomas *et al*. 2009). Using deterministic tractography, the authors identified the fronto-occipital fasciculus and the inferior longitudinal fasciculus for each DP and 17 controls. They found that the number of fibers and the number of voxels through which they passed was reduced in DPs when compared to controls. The mean fractional anisotropy in these tracts was also significantly lower in DPs than controls, possibly due to problems in the micro-structural integrity of the IFOF and ILF. Using the same dependent measures to compare callosal tracts, Thomas *et al*. (2009) did not observe any differences in the forceps minor, but did find a reduced number of voxels in the forceps major in DPs. This study indicates that deficits in the integrity of white matter tracts that pass through the temporal and occipital cortex are associated with DP.

Studies measuring ERPs in participants with normal face processing consistently find a negative component approximately 170ms (N170) after stimulus presentation that is much stronger for faces than for other stimuli (Bentin *et al*. 1996; see Eimer; Schweinberger this volume), and measurement of the N170 provides a means to examine whether DPs have deficits in early face processing. YT, the DP discussed in the fMRI and the MRI paragraphs above, had a non-selective N170 in that his response to faces and non-face objects were comparable. Interestingly, the non-selectivity resulted from a stronger than normal response to non-face objects rather than a weaker response to faces. This led to suggestions that YT's face recognition systems processed both face and non-face representations due to early filtering deficits. Two DPs also showed a non-selective N170 due to a strong response to non-face objects (Kress and Daum 2003), and MZ, who we will discuss further below, also showed a non-selective N170 due to a strong response to watches prior to face training (DeGutis *et al*. 2007)(See Figure 3). Some DPs however show normal face-selectivity in their response, which suggests that their prosopagnosia results from later processes. Five DPs were presented with faces and houses while their response was measured with magnetoencephalography (MEG) (Harris *et al*. 2005), a technique that also has high temporal resolution and measures many of the same neural sources as ERP (Hämäläinen *et al*. 1993). Three DPs had non-selective M170 responses similar to those discussed above, but two DPs had normal M170s. ERPs were recorded in these two DPs and they also manifested a face-selective N170 (Harris *et al*. 2005). Hence, as with fMRI, DPs show variability in their early response to faces and this

heterogeneity indicates that DP results from impairments to different mechanisms in different individuals. In summary, neural measures have identified a number of neural abnormalities in DPs, but an integrated picture of the neural basis of DP remains to be worked out.

Developmental and genetic studies of DP

Given that DP is a failure of development, it is ironic that little research has investigated the developmental course of DP. At present, only a few reports of children with DP have been published and they only provide a snapshot of DP in childhood and no information about its developmental trajectory. Several parents with children with DP have told me that they noticed that their child was having difficulties with faces as early as two years of age. Often these individuals were aware of their own prosopagnosia and so were likely to be especially sensitive to their child's prosopagnosia. For most parents, identification of prosopagnosia in a child is challenging.

Marked differences between children with DP and adults with DP have not been identified (Jones and Tranel 2001; Schmalzl *et al*. 2008), but this

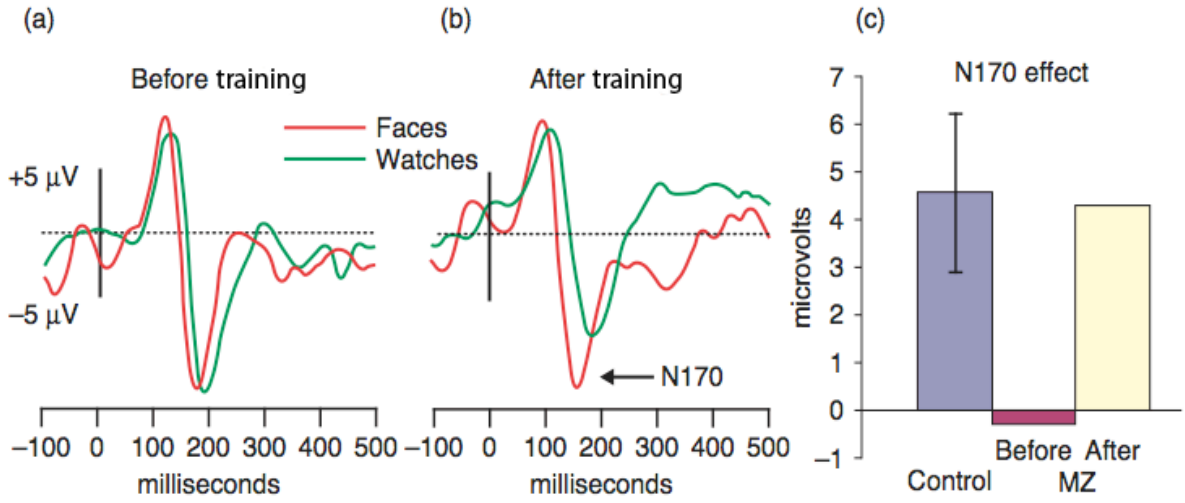

Fig. 42.3 The N170 in MZ before and after training (DeGutis et al., 2007). (a) Prior to face training, MZ's N170 was not selective for faces, because the amplitude to faces and watches is similar. (b) Following training, MZ's N170 is stronger to faces than to watches. Comparison with (a) shows that this selectivity emerged because the response to watches was weaker after training. (c) Difference in microvolts between the response to faces and watches. The difference in MZ's response after training is comparable to the average difference for controls. Errors bars represent the 95% confidence interval of the control mean. Reproduced with permission from Joseph M. DeGutis, Shlomo Bentin, Lynn C. Robertson, and Mark D'Esposito, 'Functional Plasticity in Ventral Temporal Cortex following Cognitive Rehabilitation of a Congenital Prospagnostic', *Journal of Cognitive Neuroscience*, **19**:11 (November, 2007), pp. 1790–1802. (c) 2007 by the Massachusetts Institute of Technology.

comparison rests on limited findings. The youngest child tested so far was a 4-year-old girl, K, who showed perceptual deficits with faces (Schmalzl *et al*. 2008). Prior to face

training, K made a large proportion of fixations to regions other than the inner face, with especially poor attendance of the eye region. Poor attention to the eyes has also been noted in acquired prosopagnosia (Rossion *et al*. 2009) and autism spectrum disorder (Dalton *et al*. 2005). K's results fit well with a hypothesis suggesting that DP may sometimes originate in a failure to attend to the face in early childhood (Johnson 2005). Infants and toddlers direct considerable attention to the face (Goren 1975; Johnson and Morton 1991; see de Haan; Lee *et al*. this volume), and diminished attention to the face may affect the tuning of face processing mechanisms. While this hypothesis provides a straightforward account of DP, it is also possible that DP children fail to attend to faces normally because of pre-existing high-level face processing deficits. Current evidence does not allow us to discriminate between these possibilities and it is likely that DP results from a variety of developmental disruptions. The presence of object recognition problems and navigational problems in some DP cases indicates that the prosopagnosia seen in these cases does not have its origins in lack of attention to faces (Duchaine and Yovel 2008).

Although little is known about the developmental course of DP, recent studies have demonstrated that it sometimes runs in families. Many of the early papers hinted at this possibility (Bentin *et al*. 1999; Duchaine 2000; McConachie 1976), but de Haan (1999) was the first to test multiple members of the same family. Recent years have seen many families with multiple DPs reported (Duchaine *et al*. 2007a; Grueter *et al*. 2007; Kennerknecht *et al*. 2006; Schmalzl *et al*. 2008) so it now well established that DP can run in families. This finding fits well with a twin study in the normal population indicates that face processing ability is heritable (Wilmer *et al*. in press). Given the complexity of the face processing system, it seems likely that DP will result from multiple genetic deficits but no genes associated with DP have been identified yet. Studies that have relied on self-reports of face recognition ability have found segregation patterns consistent with a dominant autosomal inheritance (Kennerknecht *et al*. 2006). There have been some suggestions that DP always runs in families (Kennerknecht *et al*. 2008), but in a group of 19 DPs in studies in my laboratory, only 58% were aware of genetic relatives who shared their prosopagnosia (Duchaine 2008).

Training studies with developmental prosopagnosics

Not surprisingly, the first question that most DPs ask when contacting me is whether treatment exists that will improve their face recognition. At present no proven treatment methods are available, but this is an area of active research and several studies have found encouraging results. The studies with DPs carried out involve training with behavioral tasks, but recent studies with normal participants showing that the neuropeptide oxytocin improves face memory (Rimmele *et al*. 2009; Savaskan *et al*. 2008) and increases fixations to the eye region (Guastello *et al*. 2008) also hold promise for DP.

Two DP children, one who was eight years old and another who was four, were taught to recognize familiar faces using the inner facial features rather than external information (Brunsdon *et al*. 2006; Schmalzl *et al*. 2008). In both cases, recognition of the familiar faces improved following training. Importantly, this training appeared to transfer to other faces. Recognition of unfamiliar faces improved in the eight-year-old

(Brunsdon *et al*. 2006) and the four-year-old increased her fixations of an unfamiliar set of faces (Schmalzl *et al*. 2008).

Training with MZ, an adult DP, involved discrimination of spacing differences between faces with similar features in an effort to improve her face configural processing (DeGutis *et al*. 2007). MZ engaged in several cycles of training followed by no training, and she reported that her everyday face recognition was markedly improved during training periods. MZ was tested with the same face recognition tasks before and after training, and her performance on them improved. Neural measures also indicated that her face recognition after training was carried out in a more typical fashion than prior to training. Before training she had a non-selective N170 due to a strong non-face response, but after training she showed a face-selective N170 because the amplitude of the non-face response had diminished (See Figure 3). In addition, her right OFA and right FFA showed increased functional connectivity following training (DeGutis *et al*. 2007).

Summary

DP holds promise as a means to investigate a range of issues in face processing and may provide a useful model to better understand the development of neurocognitive mechanisms. Cognitive studies of DP provide support for the existence of face-specific processes, and dissociations between different types of face processing in DPs are consistent with leading models of face processing that propose separable mechanisms for various aspects of face processing. Research on the neural basis of DP has found abnormalities in a number of occipital and temporal regions that show face-selective responses in people with normal face processing, and so provide additional evidence that the integrity of these areas is necessary for face recognition. However, despite the progress made in recent years, much work remains to be done and connections between cognitive, neural, developmental, and genetic levels of explanation remain to be worked out.

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