Understanding the cognitive mechanisms of developmental prosopagnosia

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Abstract

Developmental prosopagnosia (DP) is a condition associated with severe difficulties recognising familiar faces, which occurs in individuals with normal intelligence, typical low-level vision, and in the absence of manifest brain injuries. The neuro-cognitive origins of DP are still debated. Cognitive accounts have attributed face recognition deficits to reduced holistic processing of faces (i.e., whereby individual features of faces are integrated into a unified perceptual whole), and mnemonic difficulties, whereby prosopagnosics may be able to form accurate percepts, but are unable to maintain those percepts over time. At the neurological level, differences have been reported in the structural and functional connectivity of occipito-temporal regions which include face selective areas. Chapter 2 of this thesis investigated facial emotion recognition in DP and revealed widespread difficulties recognising facial emotion in individuals with apperceptive profiles of DP (i.e., DPs exhibiting difficulties forming view-invariant structural descriptions of faces at early stages of encoding). Chapter 3 explored body recognition in DP and found evidence of impaired body and object recognition in DP individuals. Moreover, the lack of relationship between observers' object and body recognition performances suggested that body and object recognition impairments in DP may co-occur independently. Chapter 4 investigated the susceptibility to the composite face illusion in two independent samples of individuals with DP and failed to show evidence of diminished composite face effects in both samples. Finally, Chapter 5 considered the contribution of perceptual encoding and short term face memory in DP using a delayed match-to-sample task and found that recognition impairments in prosopagnosics were insensitive to changes in retention interval and viewing angle, supporting an apperceptive characterisation of DP. The implications of these findings for the characterisation of DP and for understanding its underlying cognitive mechanisms, are discussed in Chapter 6.

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Authors contributions

Chapter 2: Study design was conducted by Richard Cook and myself. I created the stimuli and RC helped perfecting the stimuli and programming the experimental paradigm. I collected the data and ran the analyses, under the supervision of RC. I drafted the manuscript, which was then edited and finalised by RC.

Chapter 3: RC and I designed the experiment. Stimuli were created by RC and myself. RC programmed the experimental paradigm. I collected the data and analysed the results, under the supervision of RC. All authors were involved in the interpretation of the analyses. I drafted the manuscript for publication, which was edited by all authors.

Chapter 4: Esther Wu and I contributed equally to the study. RC and I designed, created the stimuli and programmed the experimental procedure for Experiment 1. I collected the data and conducted the analyses for Experiment 1, under the supervision of RC. EW, Hua Yang, Guo Jiahui and Brad Duchaine designed Experiment 2. EW, HY and GJ collected the data and conducted the analyses for Experiment 2, under the supervision of BD. All authors contributed to the interpretation of the analyses. EW and I drafted the manuscript, which was then edited by RC and BD.

Chapter 5: RC and I designed the experiment. Stimuli were created by RC and I. RC programmed the experimental paradigm. Katie Gray and I collected the data and analysed the results. All authors were involved in the interpretation of the analyses. I drafted the manuscript for publication, which was edited by all authors.

Declarations

This thesis is submitted to City, University of London in support of my application for the degree of Doctor of Philosophy. It has been composed by myself and has not been submitted in any previous application for any degree.

This thesis is composed by a series of published papers, with the exception of Chapter 5 which is currently in revision. The experimental chapters 2-4 are exact copies of the final version of the manuscripts accepted for publication, and are available as published journal articles on the relevant journal websites. As well as the introduction and discussion sections that are specific to each article, this thesis includes a general introduction, providing an overview of the literature relevant to the experimental chapters, and a general discussion of the main findings, interpretations, limitations and outstanding questions.

Table of contents

Table of tables

Chapter 2

Chapter 3

Chapter 4

Chapter 5

Table of figures

Chapter 1

Chapter 2

Chapter 3

Chapter 1. General Introduction

1.1 Background

The first cases of individuals with face recognition difficulties following a cerebral disease were reported at the end of the 19th century (Charcot, 1883; Wilbrand, 1892). The term *prosopagnosia* was proposed by Bodamer, who described the condition based on three brain-injured German soldiers who showed severe face recognition impairments after brain damage, which were not accompanied by broader object agnosia (Bodamer, 1947). These patients showed incapacity to recognise faces and used information such as voice, gait, clothes or accessories to aid recognition. Over time these symptoms were found to be associated with lesions of the right hemisphere, as face recognition difficulties were often accompanied by left visual field defects and visual-spatial agnosia, and cases of acquired prosopagnosia also emerged after surgical removal of brain portions in the right hemisphere (Hecaen & Angelergues, 1962).

In 1976, McConachie described the first case of developmental prosopagnosia (DP), in which the condition occurred in the absence of apparent neurological lesions and appeared to have a familial component (McConachie, 1976). Crucially, the lack of brain damage is critical to the distinction with acquired prosopagnosia, in which impairments stem from neural injuries in occipito-temporal areas underpinning face recognition (Bodamer, 1947). In her early observation, McConachie advanced the possibility that "perhaps the condition is more common than is presently thought [and] there may be a developmental aspect to prosopagnosia" (McConachie, 1976, p.81). In fact, DP was once thought to be extremely rare until the beginning of the 21st century, when the simplified access to information together with a growing attention of international media on the condition, revealed an increasing number of cases. The prevalence rate of DP has been estimated to be approximately between 1.9 and 2.47% of the general population (Kennerknecht, et al., 2006; Kennerknecht, Yee-Ho, & Wong, 2008). Both studies used large samples of participants (689 German students in Kennerknecht et al., 2006; 533 Chinese students in Kennerknecht et al., 2008) who had to complete self-report questionnaires and undertake interviews, but lacked of any objective assessment. A following prevalence study employed objective tests to screen 241 Australian adults, using a cut-off criterion of impairment defined as two standard deviations below the control mean (Bowles et al., 2009). The authors found a similar incidence rate to previous studies as between 2 and 2.9% of their sample exhibited face recognition impairments.

Today, DP is defined as a neurodevelopmental condition associated with severe difficulties recognising familiar faces and learning new identities, which occurs in individuals with normal intelligence and low-level vision, and typical social cognition (Susilo & Duchaine, 2013; Duchaine & Nakayama, 2006b; Behrmann & Avidan, 2005; Kress & Daum, 2003). Some authors prefer the term *congenital prosopagnosia* to indicate that the condition emerges from birth (e.g., Behrmann & Avidan, 2005). The use of the term *developmental prosopagnosia*, instead, refers to the possibility that in some individuals the condition may appear at some stage of neuro-development and not necessarily from birth.

People with DP develop compensatory strategies over time to cope with recognition difficulties. Familiar others are identified using non-facial cues such as voice, gait, clothing, hair-style or distinctive features, but also the context in which people are met aids identification (Cook & Biotti, 2016; Shah, Gaule, Sowden, Bird, & Cook, 2015). Consequently, recognition worsens when this type of information is not available or occluded (Shah, Gaule, Sowden, Bird, & Cook, 2015). Despite growing exposure, there is still insufficient awareness of the condition, especially compared to other neurodevelopmental disorders. Therefore, people with DP often blame bad memory or lack of attention as the source of their deficits. Some of them may report to an ophthalmologist, when problems are erroneously attributed to poor visual acuity, but rarely people will seek out neurological advice (Bate & Tree, 2017).

Recurrent failures recognising familiar others cause embarrassing situations, especially when those who experience lifelong difficulties are unaware of any organic cause that may account for their problems. Over time prosopagnosics tend to avoid social situations, resulting in detrimental interpersonal interactions. Crucially, occupational disability has also been reported in DP, whenever affected individuals struggle or fail to recognise their work colleagues, patients, students or employees (Yardley, McDermott, Pisarski, Duchaine, & Nakayama, 2008). In some cases, social anxiety and depression can occur as a consequence of social disability (Yardley et al., 2008; Duchaine & Nakayama, 2006b).

1.2 Diagnostic criteria

Identifying DP is not straightforward and standardised diagnostic criteria have to be defined (see Barton & Corrow, 2016; Dalrymple & Palermo, 2016; Shah, Gaule, Sowden, Bird, & Cook, 2015). In fact, DP is not recognised as a psychiatric disorder in the latest Diagnostic and Statistical Manual of Mental Disorders (DSM-5) (American Psychatric Association, 2013). Nevertheless, a developmental form of prosopagnosia is listed in the second edition of the Application of the International Classification of Diseases to Neurology (ICD-NA) (World Health Organization, 1997), and its signs may meet the criteria for a mental disability given the long-term negative impact on people's daily life (Lockwood, Henderson, & Thornicroft, 2012). Current standard batteries rely on subjective self-report measures as well as objective computerised tests.

1.2.1 Computerised tests

Objective instruments include an array of computer-based tests which have been validated and standardised on large samples and investigate different facets of face recognition. To justify inclusion in a DP sample it is conventional that authors present evidence that individuals score poorly on an array of computerised tests. The more evidence is collected, the stronger the case for individuals' inclusion.

1.2.1.1 Testing of unfamiliar face recognition

Traditional tests of face recognition were developed to study both typical participants and neuropsychological patients. The Recognition Memory Test (RMT; Warrington, 1984) requires participants to recognise targets paired with distractors. During the initial phase 50 target faces are presented for 3 seconds each. Subsequently, participants are presented with 50 forced-choice pairs consisting of one target face and one distractor face. This test is biased by several non-facial cues present in the pictures (e.g., hair, emotional expressions, photo artifacts, clothing). Therefore, individuals with DP can score normally on this test using such cues (Nunn, Postma, & Pearson, 2001; Duchaine, 2000). On the other hand, the Benton Facial Recognition Test (BFRT; Benton, Sivan, Hamsher, Varney, & Spreen, 1983) consists of a face matching test where observers have to match a target face with three of six simultaneously presented test faces. Performance at this test has been shown to rely greatly on feature matching strategy (Duchaine & Weidenfeld, 2003). Since target face and test faces appear on screen at the same time, participants with DP can use cues such as eyebrows or lips shape to perform within the typical range (Duchaine & Nakayama, 2004; Kress & Daum, 2003; Nunn, Postma, & Pearson, 2001). The Glasgow Face Matching Test (GFMT; Burton, White, & McNeill, 2010) provides an alternative to the BFRT to study face matching avoiding feature matching strategies. In fact, observers make same/different judgements on the identity of two simultaneously presented faces whose pictures are taken on the same day, using neutral poses only, under the same light condition, but using different cameras. Crucially, it has been widely shown that even typical participants find this task very difficult, with average error rates of 20% (see White, Rivolta, Burton, Al-Janabi, & Palermo, 2017). This can be explained by a well documented advantage of familiarity on face matching performance (e.g., Megreya & Burton, 2006; 2007; White, Burton, Jenkins, & Kemp, 2014). Crucially, some authors advanced the possibility that unfamiliar face matching does not rely on face-specific mechanisms (Megreya & Burton, 2006). In fact, DPs have been reported to score typically on the GFMT (White et al., 2017).

The Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006a) was created to address the limitations of previous tests. Currently, it is the most used test of unfamiliar face recognition and has both strong validity and high reliability (e.g., McKone et al., 2011; but see Bate & Tree, 2017). While keeping the memory paradigm of the RMT with multiple identities, the CFMT does not allow for the use of non-facial cues as this information is occluded. The test is divided in three phases with incremental levels of difficulty. Initially, observers have to memorise six unfamiliar male faces viewed from three different view-points. In the second phase the target faces are reviewed for 20 seconds. Subsequently, participants have to recognise the target among two unfamiliar distractors presented in 30 forced-choice trials. Finally, the third phase is similar to the second phase with the difference that images contain levels of Gaussian noise to make the task more difficult and to force participants to use mechanisms used in typical face recognition (Duchaine & Nakayama, 2006a). The final score is an average of the correct responses in the three individual phases (maximum score = 72). Norm data from the original paper, using 50 controls (M_{age} = 20.2), reported an average score of 57.9 (Duchaine & Nakayama, 2006a). Norm data coming from later studies found similar results (e.g., 58.9 in Garrido, Duchaine, & Nakayama, 2008; 59.6 in Duchaine, Yovel, & Nakayama, 2007; 54.6 and 55.3 in Bowles et al., 2009). Versions of the CFMT have been developed to match the face stimuli with ethnicity (e.g., the

CFMT-Australian by McKone et al., 2011; the Taiwanese Face Memory Test by Shyi, Cheng, Cheng, & Chen, 2015) and age (e.g., the CFMT-Children by Croydon, Pimperton, Ewing, Duchaine, & Pellicano, 2014) of participants.

The Cambridge Face Perception Test (CFPT; Duchaine, Germine, & Nakayama, 2007) provides a measure of face perception ability minimizing memory demands. Participants have to sort faces based on resemblance to a target face shown above. The six faces are created by morphing the target face with a distractor face and each one contains different proportions of the target image. The structure of this paradigm makes it prone to the use of compensatory strategies (e.g., looking for trivial details in the pictures); it has been shown to have poor reliability (Rezlescu, Susilo, Wilmer, & Caramazza, 2017; Bowles et al., 2009); and the necessity of using a mouse or laptop trackpad to move the items often interferes with execution, especially with elder participants. Therefore, good performances on this test are difficult to interpret as in some cases results could reflect its relative insensitivity rather than normal face perception.

1.2.1.2 Testing of familiar face recognition

There are substantial qualitative differences between unfamiliar and familiar face processing. Overall, familiar face recognition relies greatly on internal features (e.g. nose, eyes, mouth) and familiar faces are easily and effortlessly detected even in low-attention conditions. On the other hand, unfamiliar recognition is based more on external features such as hair and face shape, and it places higher demands on working memory (for a review see Johnston & Edmonds, 2009). Given these differences, tests have been developed to assess familiar and unfamiliar face recognition separately in DPs. The Famous Face Tests (e.g., Duchaine, 2000; Duchaine & Nakayama, 2005; Macquarie Famous Face Test 2008, Palermo, Rivolta, Wilson, & Jeffery, 2011) consist of a good estimate of familiar face recognition, as observers have to recognise faces of celebrities. However, performance on these tests is highly influenced by individual differences such as participants' interest in popular culture, age, and demographics. In addition, celebrities must be carefully selected based on the geographical area of observers. To minimise the effect of these variables, the standard FFTs use celebrities from various backgrounds (e.g. music, cinema, sport, politics), and the final score is calculated relative only to those stimuli participants are familiar with.

A familiar face matching test using famous faces has also been created to provide a more ecologically valid measure to the GFMT. In the Local Heroes Test (LHT; White, Rivolta, Burton, Al-Janabi, & Palermo, 2017) participants view pairs of images of either famous or unfamiliar people and have to establish whether the pictures depict the same identity. Unlike the GFMT, the LHT uses pictures taken under unconstrained conditions which should better represent everyday face perception. Given the advantage of familiarity in face matching tasks, it is expected that individuals with typical face recognition ability will perform significantly better when matching famous faces.

1.2.2 Self-report measures

Based on the assumption that people who struggle recognising familiar faces are aware of their difficulties, researchers have developed self-report instruments to provide an overall picture of the severity of self-reported problems (e.g., Shah, Gaule, Sowden, Bird, & Cook, 2015; Kennerknecht, et al., 2006). Self-report measures alone are not sufficient for a diagnosis of DP, but they represent an efficient screening tool (see Bate & Tree, 2017). For example, some authors have used semi-structured interviews to explore face recognition difficulties in everyday life (e.g., Grueter et al., 2007; Murray, Hills, Bennetts, & Bate, 2018), while studies with larger samples have benefitted from the use of self-ratings to recruit and select participants (e.g., *the Hereditary Prosopagnosia questionnaire* in Kennerknecht et al., 2006; *the self-rated face recognition questionnaire* in Bowles et al., 2009). However, these measures were purposely developed for individual studies, and lacked a formal validation procedure. The 20-item Prosopagnosia Index (PI20; Shah et al., 2015) was created to provide the first validated and standardised self-report test for DP diagnosis. The questionnaire consists of 20 items examining face recognition in everyday life, providing part-takers with hypothetical tangible scenarios to which they can easily relate. Strong correlations are normally observed between PI20 scores and objective measures of familiar and unfamiliar face recognition (i.e., the FFT, the CFMT, the CFPT), allowing the distinction between individuals with typical and atypical face perception. In addition, scores in the higher range (i.e. 65-100) can be broadly clustered into groups of severity (i.e., mild, moderate and severe). The predictive value of the PI20 found in Shah and colleagues (2015) is in contrast with the view that people may have little or no insight into their face recognition ability (e.g., Bowles et al., 2009; Laguesse et al., 2013; Palermo et al., 2017). However, the PI20 is a measure which has been created as a diagnostic instrument, with items constructed ad hoc to investigate prosopagnosic traits rather than general face recognition abilities (see Gray, Bird, & Cook, 2017). Therefore, any attempt to conceive PI20 results as an indication of face recognition ability *per se* would be misleading. Instead, the PI20 should be used to complement other diagnostic measures in order to provide additional evidence that bad performance on computer-based tests indicate actual atypical face processing rather than lack of motivation, boredom or fatigue effects. Moreover, an increasing number of authors are starting using the PI20 as a quick screening tool to sample DP participants, or when recruiting participants from online platforms.

1.2.3 Convergence of diagnostic evidence

On one hand, questionnaires are a useful screening tool enabling researchers to select putative prosopagnosics based on where their score falls compared to the norm. Though, these instruments alone cannot be regarded as the only diagnostic criterion. In fact, some people may be prone to either underestimate or overestimate their actual face recognition ability - in the typical population only little correlations were found between subjective assessments and objective scores (Bindemann, Attard, & Johnston, 2014; McGuin, Richler, Herzmann, Speegle, & Gautier, 2012). Therefore, it is common practice to integrate questionnaires with computerised measures for a precise assessment.

On the other hand, performance on computer-based tests is more objective but does not always generate reliable evidence. For example, it has been shown that the statistical sensitivity in identifying prosopagnosics is highly influenced by the composition of the control group, which must be matched for age, ethnicity, gender, and intelligence to produce reliable data (Bowles et al., 2009; McKone et al., 2011). Moreover, aspects such as lack of motivation, boredom, test anxiety and poor mouse control may also affect the execution of these tests (Butcher, Perry, & Atlis, 2000; see also Shah, Gaule, Sowden, Bird, & Cook, 2015). Coversely, existing impairments may not be always revealed by traditional tests, whenever individuals with a lifelong history of face recognition difficulites may be prone to apply compensatory strategies to execute diagnostic tasks.

Currently, the diagnostic approach of many researchers is leaning towards the use of different measures which can contribute to convergent evidence. For example, recent findings revealed that less than 1.5% of the general population score below 65% on the CFMT and more than 65 on the PI20 (Gray, Bird, & Cook, 2017). This data suggests that the use of multiple tests, including both objective and self-report measures, may be a particularly effective approach when classifying individuals with DP.

1.3 Perceptual and cognitive accounts of DP

Face recognition ability is distributed in the general population; the majority of people collocate in the middle of a bell-shaped curve, where at the two extreems are individuals with either exceptional skills (i.e. *super recognisers*, Russell, Duchaine, & Nakayama, 2009), or pronounced face recognition difficulties. It is rather tempting to describe DP on the basis of quantitative differences with the typical population. If this were the case, we would expect deficits to emerge merely from reduced functioning of the same mechanisms subtending face perception in neurotypical individuals. Conversely, prosopagnosics may rely on qualitatively different processes, which are less effective and forceful than those used typically.

Developmental disorders can vary on the degree to which cognitive deficits are widespread or specific to certain domains (Duchaine & Nakayama, 2006). In this respect, DP appears to be relatively restricted regarding its symptoms, the majority of which pertain to the visual domain. Despite this apparent simplicity, the cognitive characterisation of DP is not straightforward - after two decades of intense research, the causes of the condition remain unclear and our understanding is hindered by conflicting evidence. What we know for certain is that DP is a heterogeneous disorder which manifests with a variety of behavioural profiles (Susilo & Duchaine, 2013; Duchaine & Nakayama, 2006; Behrmann, Avidan, Marotta, & Kimchi, 2005).

1.3.1 Is DP due to a low-level visual deficit?

General low-level vision is thought to be intact in many DPs as authors infer normal low-level processing from typical performance in non-face conditions. Therefore, a very few studies have included measures of basic visual processing. Despite little attention, low-level visual properties have a significant effect on our perception of face stimuli (see Barton, Cherkasova, Press, Intriligator, & O'Connor, 2004). For example, luminance sensitivity can affect how we perceive subtle shades which provide important cues to facial contour and local shapes, and information about facial shape is conveyed by low and high spatial frequencies (e.g., Fiorentini, Maffei, & Sandini, 1983).

Nunn, Postma, and Pearson (2001) investigated an individual with DP who showed a selective impairment at face recognition despite normal basic visual skills (i.e., visual acuity, colour perception, line orientation, visual motor integration, visual matching and copying, perception of form, visuo-constructive ability and perceptual integration). Another single case was examined who showed normal low-level vision (i.e., visual fields, visual copying of figures and objects, visual matching of figures, length, size, and orientation, perception of overlapping figures) despite significant deficits at both face and object recognition (Duchaine, Nieminen-von, New, & Kulomaki, 2003).

A particular attention has been given to the role of high and low spatial frequencies in the perception of local and global features. It has been suggested that the perceptual advantage for global vs local structure found in neurotypical individuals may be mediated by low spatial frequency channels which operate at early visual processing (e.g., Lamb & Yund, 1993; Badcock, Whitworth, Badcock, & Lovegrove, 1990). To rule out the possibility that configural processing deficits commonly found in DP may be due to aberrant sensitivity to low spatial frequencies, Behrmann and colleagues (2005) tested the contrast spatial frequency sensitivity of 5 individuals with DP. Performances fell within the typical range. Furthermore, sensitivity to spatial relations between features was also tested and all DP individuals were able to derive contours from small Gabor patches whose stimuli varied in number, orientation and distance between each other (Behrmann et al., 2005).

Nevertheless, when tested on sensitive psychophysical tasks, 3 individuals with DP showed mixed results regarding their basic visual processing (Barton, Cherkasova, Press, Intriligator, & O'Connor, 2003). Impairments were found in spatial contrast sensitivity (3 DPs), particularly for high spatial frequencies, luminance discrimination (2 DPs), saturation discrimination (1 DP), spatial resolution (3 DPs), and dot displacement discrimination (3 DPs) (for a summary refer to Table 2 in Barton et al., 2003).

Mid-level vision is also been investigated in DP. The extraction of global form, where local elements are integrated into a coherent whole, is essential to face perception and sensitivity to global form can be studied using Glass patterns (1969) or a compound letter task (Navon, 1977). Glass patterns are created by overlaying two identical random patterns of dots; rotation of these patterns over a central axis results in the perception of different spatial patterns like spirals or circles. In fact, patterns rotation allows to introduce correlations between the two set of dots and our eyes are able to detect these spatial relationships in the form of concentric global shapes. Sensitivity to these global patterns is assessed by varying the ratio of signal dots (i.e. paired dotes) to noise dots (i.e. unpaired dots). Crucially, typical adults can detect global form even if carried by only 12% of signal dots (Lewis et al., 2004; Le Grand et al., 2006), and individuals with DP have been reported to show similar sensitivity to that of neurotypical observers (Le Grand et al., 2006). Prosopagnosics also exhibit typical perception of global motion (Le Grand et al., 2006), assessed by varying the proportion of noise dots moving in random directions to signal dots moving in the same direction.

Similarly, many DPs have been shown to perform typically on compound letters tasks (Duchaine, Germine, & Nakayama, 2007; Duchaine, Yovel, & Nakayama, 2007; Schmalzl, Palermo, & Coltheart, 2008), in which they have to identify compound letters either at global or local level (Navon, 1977). However, it must be mentioned that some authors found a local bias in prosopagnosics, resulting in slower reaction times for global judgements, especially when the global letter was inconsistent with the local letters (Behrmann, Avidan, Marotta, & Kimchi, 2005; Avidan, Tanzer, & Behrmann, 2011).

1.3.2 Is DP due to diminished acquisition of perceptual expertise?

The ability of typical adults to perceive and recognise faces so quickly and accurately may be explained by considering faces objects for which humans have developed a high degree of perceptual expertise (Diamond & Carey, 1986). This expertise is aided by the fact that all faces share a common arrangement of their features, and humans are particularly sensitive to this configuration (Gauthier & Tarr, 2002). Therefore, acquiring perceptual expertise means to be sensitive to the typical configuration of faces, which are perceived using holistic processing rather than a part-based one. As a result of perceptual expertise, compared to other objects faces are salient stimuli that can be detected quickly and efficiently, when they are in their typical upright configuration. Accordingly, people tend to automatically and preferentially orient their attention to face stimuli when they are present in a visual context (e.g., Rousselet, Macé, & Fabre-Thorpe, 2003; Ro, Friggel, & Lavie, 2007), and selective orientation of attention towards face-like configurations has been reported already in newborn infants (e.g., Morton & Johnson, 1991; Gliga, Elsabbagh, Andravizou, & Johnson, 2009), suggesting that some inherited preference for certain patterns may be present since birth.

Crucially, DP may result from a failure in developing perceptual expertise for faces. This idea has been explored in other develomental conditions that share face perception difficulties. For example, Schultz (2005) has proposed a compelling account of ASD as a breakdown in attention orientation towards face stimuli, in particular those with emotional salience, which affects the normal development of brain networks (i.e. amygdala-fusiform face area) involved in social perception. If this is true for DP, we might expect atypical behaviour at very early stages of face processing.

Face detection refers to "the process of finding a face in a visual scene" (Garrido, Duchaine, & Nakayama, 2008, p. 119) and it appears to be reduced by changes in the typical configuration of faces (e.g. inversion) or in their low-level visual properties (e.g. luminance, contrast, sharpness, hue) (e.g., Purcel & Stewart, 1986, 1988; Lewis & Edmonds, 2003, 2005; Rousselet, Mace, & Fabre-Thorpe, 2003; but see Garrido et al., 2008). Face detection is the very first step of face processing. Despite its relevance in computer sciences and machine learning, it has received only little consideration from vision scientists and its neuro-cognitive mechanisms are not covered by influentials models of face perception (Bruce & Young, 1986; Haxby, Hoffman, & Gobbini, 2000).

Whether face detection can be dissociated from other mechanisms of face processing, like face recognition for example, is a matter of debate. The inversion effects found in tasks of face detection are smaller compared to those normally observed in face recognition tasks (see Lewis & Edmonds, 2003, 2005). However, as suggested by Garrido and colleagues (2008), the presence of contextual cues in detection paradigms may reduce the size of the inversion effect. Evidence of dissociation with face recognition comes from findings of intact face detection in patients with acquired prosopagnosia (e.g., de Gelder & Rouw, 2000; Schiltz et al., 2006). Similarly, individuals with DP show typical face detection in tasks where they have to discriminate between a face and a scrambled face (e.g., de Gelder & Rouw, 2000; Le Grand et al., 2006; Duchaine, Nieminen-von, New, & Kulomaki, 2003; Duchaine, 2000; Duchaine, Yovel, Butterworth, & Nakayama, 2006). Nevertheless, when a more ecologically valid paradigm was used, in which participants had to locate face stimuli among distractors presented in a visual array, Garrido and colleagues (2008) found impaired face detection at the group level in 14 prosopagnosics.

1.3.3 Apperceptive accounts of DP

Historically DP has been conceived as a form of prosopagnosia which affects people's ability to encode the structure of faces (De Renzi, Faglioni, Grossi, & Nichelli, 1991; Behrmann & Avidan, 2005; Duchaine & Nakayama, 2006b). Specifically, the impairment was thought to occur early in the face processing stream, before the analysis of various face attributes (e.g., identity, emotion expression) segregates (Bruce & Young, 1986; Haxby, Hoffman, & Gobbini, 2000). Accordingly, face perception is not a monolithic function but is modulated by separate functional units which can act either in series or in parallel (Bruce & Young, 1986, Figure 1.1a; Haxby et al., 2000; Figure 1.1b). After an initial structural encoding which allows for a perceptual description, two relatively independent systems are responsible for the analysis of changeable (i.e. expressions, lip movement, eye gaze) and invariant (i.e. identity) aspects of faces. Depending on where the impairment occurs in this hierarchical model, deficits will likely emerge in all the subsequent processing stages, leading to a variety of impairment profiles.

In support of apperceptive accounts of DP, many prosopagnosics show difficulties when matching unfamiliar faces presented simultaneously (e.g., Duchaine, Germine, & Nakayama, 2007; Avidan, Tanzer, & Behrmann, 2011; Shah, Gaule, Gaigg, Bird, & Cook, 2015; White, Rivolta, Burton, Al-Janabi, & Palermo, 2017). Moreover, some DPs also present co-occurent impairments at categorising facial age and gender (Ariel & Sadeh, 1996), and emotion expressions (Burns, Martin, Chan, & Xu, 2017; Duchaine et al., 2007). The CFPT is been widely used to investigate apperceptive deficits minimising memory demands. However, its structure makes this test prone to compensatory strategies, and performance can also be influenced by aspects such as participants' visual acuity, anxiety when performing against the clock, or ability in using a trackpad or mouse. As a result, many DPs often fail to show impaired performance on the CFPT at the single case level (e.g., Bowles et al., 2009; Ulrich et al., 2017). However, differences at the group level that emerged in other studies (e.g., Duchaine et al., 2007; Shah et al., 2015; White et al., 2017) highlight the possibility of apperceptive deficits, which sometimes get undetected due to poor test-retest reliability.

1.3.3.1 Holistic processing

An influencial apperceptive account has tried to explain the deficits observed in DP as a consequence of aberrant visual processing, wherby faces cannot be perceived holistically. Crucially, faces are represented in a unique way compared to other visual stimuli: they are perceived as a perceptual whole rather than as a combination of the individual constituents (Farah, Wilson, Drain, & Tanaka, 1998; Maurer, Le Grand, & Mondloch, 2002; for a review see Piepers & Robbins, 2013). This representational style is known as configural processing, which refers to the natural tendency to i) being tuned to first-order relations, which correspond to the typical configuration of faces, ii) integrate individual facial features into a perceptual whole (i.e. holistic processing), and iii) being sensitive to second-order relations, which allow the distinction of different identities by extracting spatial distances between local features (Maurer, Le Grand, & Mondloch, 2002). This special modality of processing faces compared to other objects seems to develop as consequence of expertise (Diamond & Carey, 1986). The enormous amount of faces that people encounter throughout a lifetime makes it crucial to be able to differentiate and recognise them effortlessly and in different conditions. Therefore, a failure in configural processing appears to be associated with reduced face recognition (Rivest, Moscovitch, & Black, 2009), and some authors attempted to describe DP as a breakdown in configural processing (Avidan, Tanzer, & Behrmann, 2011; Carbon, Grüter, Weber, & Lueschow, 2007; Liu & Behrmann, 2014; Lobmaier, Bölte, Mast, & Dobel, 2010; Palermo et al., 2011). Accordingly, training prosopagnosics on holistic processing is being reported to improve their face recognition (DeGutis, Cohan, & Nakayama, 2014). Evidence of configural processing comes from phenomena such as the inversion effect (IE) (Yin, 1969), the composite face illusion (CFI) (Young, Hellawell, & Hay, 1987), and the part-whole effect (PWE) (Tanaka & Farah, 1993).

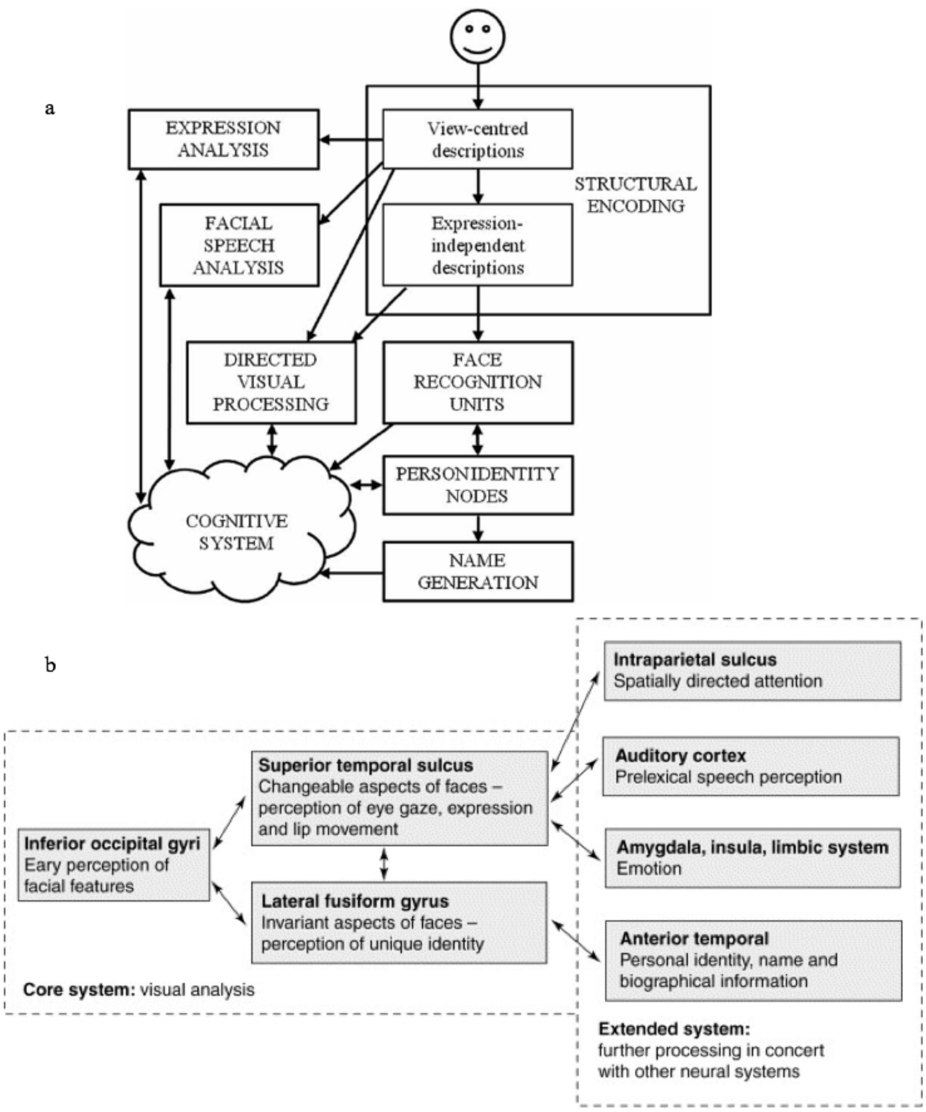

Figure 1.1. Neuro-cognitive models of face perception. (a) Bruce & Young, 1986, p. 312; (b) Haxby, Hoffman, & Gobbini, 2000.

1.3.3.1.1 The Inversion Effect

The IE refers to the disproportionate decline of perception when faces are viewed upside-down, and not in their normal upright configuration (Yin, 1969). This phenomenon occurs as people are sensitive to first-order relations (i.e. two eyes above a nose and a mouth) which trigger holistic processing. When faces are inverted, configural processing is disrupted and observers rely on featural processing, whereas local facial components are processed individually (Farah, Tanaka, & Drain, 1995).

Since non-face objects are generally perceived with a part-based approach, the IE is not as evident for other visual categories (Farah, Wilson, Drain, & Tanaka, 1998). Reports investigating the IE in prosopagnosics show mixed results, partially due to differences in sample sizes and tasks employed (Behrmann, Avidan, Marotta, & Kimchi, 2005; Duchaine, Yovel, & Nakayama, 2007; Garrido, Duchaine, & Nakayama, 2008; Nunn, Postma, & Pearson, 2001; De Gelder & Rouw, 2000).

1.3.3.1.2 The Composite Face Illusion

Holistic processing is also studied by looking at people's sensitivity to the CFI, where the perception of one face-half is inevitably influenced by the other half, when these are presented aligned and in the typical upright configuration (Young, Hellawell, & Hay, 1987). The perceptual illusion emerges when the top half of a face is aligned with the bottom of another, giving rise to the impression of a new facial identity (Young et al., 1987). Typically, individuals' perception is affected when composites are presented upright and aligned spatially, and people are unable to judge the target half disregarding the distractor, even when explicitly instructed to do so. However, recognition of the target half is preserved both in aligned and misaligned arrangements when faces are presented inverted (for a review see Murphy, Gray, & Cook, 2017). The traditional paradigms employed to investigate the CFI can be either simultaneous (e.g. Hole, 1994) or delayed (e.g. Goffaux & Rossion, 2006; Le Grand, Mondloch, Maurer, & Brent, 2004) matching tasks, where participants are required to make a same/different judgment of the top halves of two simultaneously or sequentially presented composites, which can be aligned or misaligned, upright or inverted. Holistic processing is revealed by the disproportionate detriment of performance when composites are presented aligned and upright compared to the other conditions. A compelling account of DP as consequence of impaired holistic processing stems from the idea that the process measured by the CFI is closely related to face recognition ability (DeGutis, Wilmer, Mercado, & Cohan, 2013; Maurer, Le Grand, & Mondloch, 2002; Piepers & Robbins, 2013). However, comparing people's sensitivity to the composite face effect and their face recognition produced mixed results in the general population (for a review see Murphy, Gray, & Cook, 2017). Whilst some authors found a positive association (DeGutis, Wilmer, Mercado, & Cohan, 2013; Engfors, Jeffery, Gignac, & Palermo, 2017; Wang, Li, Fang, Tian, & Liu, 2012), others only observed little or no correlation (Rezlescu, Susilo, Wilmer, & Caramazza, 2017; Konar, Bennett, & Sekuler, 2010). Interestingly, evidence from individuals with DP is also varied and a few studies have shown comparable CFI in both prosopagnosics and matched controls (Le Grand et al., 2006; Susilo et al., 2010; Schmalzl, Palermo, & Coltheart, 2008), whilst three studies concluded that DP is associated with reduced susceptibility to the illusion (Palermo et al., 2011; Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014).

1.3.3.1.3 The Part-Whole Effect

Another way of measuring holistic processing is through the part-whole effect, where facial components are perceived disproportionately better when they are englobed in the context of a whole face rather than as isolated parts (Tanaka & Farah, 1993). This phenomenon exposes the sub-component of configural processing which allows the detection of relative distances between face features, when these are presented canonically (i.e. "second-order relations" in Maurer, Le Grand, & Mondloch, 2002). In fact, the part-whole effect disappears with non-face objects and inverted faces (Tanaka & Farah, 1993). The standard task requires observers to memorise a target face and subsequently identify the whole-face paired with a distractor which differs in either eyes, nose or mouth, or identify the individual components of the target face shown in isolation with distractor components. This paradigm was employed to train a group of 5 DPs on other-race face recognition and pre-training data revealed a reduced, although not significantly different from controls, holistic advantage for whole faces in the DP group (DeGutis, DeNicola, Zink, McGlinchey, & Milberg, 2011). A later study using a very large sample of prosopagnosics ($N = 38$) observed a lack of holistic advantage for the eye region but not for the mouth (DeGutis, Cohan, Mercago, Wilmer, & Nakayama, 2012) posing interesting questions on the value of the eyes as critical source of information for face discrimination.

1.3.3.2 Domain-general configural processing

Taken together these results show a complex picture, where holistic processing appears to manifest as a variety of functions rather than as a monolithic phenomenon (for a review see Rezlescu, Susilo, Wilmer, & Caramazza, 2017). The inconsistency of results in DP may be partly due to the attempt of ascribing results in individual tests to holistic processing as a whole, whereas only certain processes of it may be impaired. Crucially, aberrant processing of configurations may also extend to non-face stimuli. Individuals with DP have been described to show a local processing bias with compound Navon stimuli, whereas local letter identification was faster than global identification (Behrmann, Avidan, Marotta, & Kimchi, 2005; Avidan, Tanzer, & Behrmann, 2011). Furthermore, Avidan and colleagues (2011) found a significant correlation between performance on a composite face task and a local bias index emerged in the global-local task. These results indicate that in individuals with face recognition deficits there might be an association between impairments in holistic face processing, measured with the composite task, and general configural processing of non-face stimuli, measured with the Navon task. However, this association does not always occur, and a case of acquired prosopagnosia showing clear dissociations between the two tasks has been reported (Busigny & Rossion, 2011), suggesting different cognitive mechanisms underlying the execution of these two tasks.

1.3.3.3 Norm-based coding

A compelling theoretical framework of face recognition postulates the existence of a cognitive map onto which each facial identity is represented as a point in this putative multidimensional face space (Valentine, 1991; for a review see Webster & MacLeod, 2011). The centre of this space consists of the average of all faces experienced hitherto and individual faces are coded based on the average norms. Distance and direction from the centred average determine distinctiveness and deviance along multiple dimensions of each perceived face, respectively. For example, distant faces will be judged as less typical than faces that collocate close to the centre. Faces are discriminated between each other based on where they are in this representational system – the closer two faces are together, the more similar they are perceived by the observer. Surprisingly, 6 individuals with DP were tested on different tasks where they had to judge face identities and showed typical representation of faces which was consistent with a face space model (Nishimura, Doyle, Humphreys, & Behrmann, 2010). The authors postulate that face recognition impairments in this population may originate from difficulties accessing mental representations. Specifically, aberrant holistic processing may prevent them from combining local features which can be perceived typically (Nishimura et al., 2010). In fact, DPs generally rely on distinctive features when identifying faces, with consequent detriment in performance when faces appear alike (Le Grand et al., 2006; Nunn, Postma, & Pearson, 2001). Conversely, typically developed observers may use salient cues when faces are enough distinctive, but take advantage of feature-binding mechanisms with more standard-looking faces (Nishimura et al., 2010; Mondloch, LeGrand, & Maurer, 2002).

The adaptation aftereffects reveal the unceasing updating of our face space, whereby extended exposure to a face biases perception in the opposite direction (Rhodes & Leopold, 2011). Palermo and colleagues (2011) employed an adaptation paradigm to examine whether face recognition difficulties may be associated to aberrant adaptive face coding in a group of 14 prosopagnosics. The DPs failed to show typical adaptation to face identity, whilst their aftereffects to facial shape was comparable to that of controls, suggesting that their mechanisms of face adaptation may be impaired, or at least different from those used by neurotypical individuals, only relatively to face identity.

1.3.4 Mnemonic accounts of DP

Evidence against an apperceptive characterisation of DP has emerged, as several findings show that DPs can encode face structure typically, resulting in unimpaired matching of simultaneously presented faces (e.g., Bowles et al., 2009; Dalrymple, Garrido, & Duchaine, 2014; Ulrich et al., 2017), accurate recognition of facial emotion (e.g., Humphreys, Avidan, & Behrmann, 2007; Palermo et al., 2011; Dobel, Bölte, Aicher, & Schweinberger, 2007), and typical susceptibility to the composite face illusion (e.g., Le Grand et al., 2006; Susilo et al., 2010; Ulrich et al., 2017). These findings have led some authors postulating the possibility that DP is caused by impaired short-term face memory (STFM), whereby DPs are able to derive an accurate structural description, but have difficulties maintaining the percept in memory over time (e.g., Dalrymple et al., 2014; Dalrymple & Palermo, 2016; Jackson, Counter, & Tree, 2017; Stollhoff, Jost, Elze, & Kennerknecht, 2011; Ulrich, et al., 2017). Accordingly, cases of DPs have been reported who show impaired performance on the CFMT, performing typically on tests with minimal memorial components such as the CFPT (e.g., Bowles et al., 2009; Dalrymple et al., 2014; McKone et al., 2011; Ulrich et al., 2017). Shah and colleagues (2015) systematically investigated the contribution of memory and perceptual deficits in DP. In a delayed match-to-sample paradigm, the authors manipulated the retention interval, maintaining constant the perceptual demand. Interestingly, although DPs were less accurate and slower compared to controls in both conditions, the long-interval condition did not disproportionally impair their performance, suggestive of perceptual deficits (Shah et al., 2015). Similarly, a recent study which manipulated memory load by increasing the number of target faces to memorise failed to show a disproportionate decrement of performance as a function of memory load in the DP group compared to typical controls (Jackson, Counter, & Tree, 2017). Taken together these findings suggest that problems at face matching observed in DP may stem from a perceptual deficit, rather than from aberrant retention.

1.3.5 A heterogeneous disorder

DP is a heterogeneous condition and it is often accompanied by difficulties recognising complex non-face objects (Behrmann, Avidan, Marotta, & Kimchi, 2005; De Haan & Campbell, 1991; Duchaine, Germine, & Nakayama, 2007; Duchaine & Nakayama, 2005; Ariel & Sadeh, 1996; McConachie, 1976; Lee et al., 2010; Todorov & Duchaine, 2008). A recent meta-analysis found that the 80.3% of the analysed data (N = 238 cases) consisted of DP individuals with concurrent object recognition difficulties, whilst only 19.7%[1] were pure cases of prosopagnosia (Geskin & Behrmann, 2017). Even when considering face recognition impairments alone, individual DPs vary regarding which aspects of face perception are affected by the condition, whether it is the perception of face structure and local features, or the retention of faces in short term memory.

1.3.5.1 Apperceptive and mnemonic profiles

Face processing difficulties observed in DP appear to have a different origin when considering individual cases. Some DPs (*apperceptive*) are unable to encode the structure of faces, resulting in deficits distinguishing unfamiliar faces (e.g., Avidan, Tanzer, & Behrmann, 2011; Behrmann, Avidan Marotta, & Kimchi, 2005; Duchaine, Yovel, Butterworth, & Nakayama, 2006; Shah, Gaule, Gaigg, Bird, & Cook, 2015; White, Rivolta, Burton, Al-Janabi, & Palermo, 2017; Fisher, Towler, & Eimer, 2017), and judging aspects of faces other than identity, like facial emotion (Duchaine et al.,

[1] This figure may underestimate the actual number of pure cases, as authors eliminated from the analyses all those studies which found normal object recognition but omitted to report RT data.

2006; Ariel & Sadeh, 1996; de Haan & Campbell, 1991; Duchaine, Murray, Turner, White, & Garrido, 2009; Minnebusch, Suchan, Ramon, & Daum, 2007; Schmalzl et al., 2008), facial age (Ariel & Sadeh, 1996), and facial gender (Ariel & Sadeh, 1996; de Haan & Campbell, 1991; Jones & Tranel, 2001). On the other hand, *mnemonic* DPs can succesfully encode facial structure and perceive emotion expressions (Dobel, Bölte, Aicher, & Schweinberger, 2007; Humphreys, Avidan, & Behrmann, 2007; Palermo et al., 2011), age and gender (Chatterjee & Nakayama, 2013) typically.

The heterogeneity observed in DP is consistent with the historical classification of prosopagnosia, which was based on acquired cases who presented either an apperceptive or an associative (or mnemonic) profile of impairment (De Renzi, Faglioni, Grossi, & Nichelli, 1991). In associative forms of prosopagnosia, a structural description can be extracted but the percept cannot be associated to its representation stored in face memory, resulting in impairments at face recognition tests (e.g., the FFT, the CFMT, the RMT). In acquired prosopagnosia, associative subtypes result from lesions in the right anterior temporal lobe and patients have been described to show impaired access to face memory despite preserved face perception (Pancaroglu et al., 2011). Furthermore, the recognition of familiar faces, tested using a variant of the FFT, results particularly impaired in patients with right and left anterior temporal lesions (Tsukiura et al., 2002; Tsukiura, Suzuki, Shigemune, & Mokizuki-Kawai, 2008; but see Gainotti, 2007).

The presence of apperceptive and associative profiles of impairment reflects the multifaceted nature of face recognition, which relies equally on *face perception* and *face memory*. While face perception refers to those processes contributing to the representation of face properties, face memory consists of those processes which allow the storing, retention and retrieval of identity information (Dalrymple et al., 2014). Evidence of dissociation between face perception and face memory emerges from studies of patients with acquired prosopagnosia, but also by looking at neurotypical development. Overall, perceptual impairments stem from lesions localised more posteriorly in occipito-temporal regions, whilst amnesic deficits arise from more anterior lesions (Barton & Cherkasova, 2003; Barton, Press, Keenan, & O'Connor, 2002; Damasio, Tranel, & Damasio, 1990; but see Dalrymple et al., 2014). On the other hand, anterior injuries will likely affect face memory (Busigny, et al., 2014; Davies-Thompson, Pancaroglu, & Barton, 2014). However, this distinction is not always clear-cut, and some case reports showed atypical face perception resulting from damage in the right anterior temporal lobe (Busigny et al., 2014; Williams, Savage, & Halmagyi, 2006).

Furthermore, face perception and face memory appear to segregate later in development, with face memory developing more slowly and reaching the peak of maturation after the first decade of life (Weigelt et al., 2014). For example, children with DP are equally impaired on perceptual and memory tasks, whereas adults show a dissociation (Dalrymple, Garrido, & Duchaine, 2014).

1.3.5.2 Non-face objects deficits

The extent to which DP affects face-specific mechanisms is still debated. Reportedly, some individuals perform in the typical range on standardised object recognition tests (e.g., Bentin, Deouell, & Soroker, 1999; Nunn, Postma, & Pearson, 2001). Duchaine and colleagues (2006) described a pure case of DP who showed good recognition of several complex objects including cars, guns, tools, horses and sunglasses, despite severe impairments in various components of face processing (i.e., recognition of identity, perception of gender, facial emotion and attractiveness). Interestingly, pure cases appear to be relatively common in DP (e.g., Duchaine & Nakayama, 2005; Garrido, et al., 2009; Susilo et al., 2010; Lee, Duchaine, Nakayama, & Wilson, 2010). These reports indicate the existence of forms of DP which can only affect face-specific processes, suggesting that mechanisms for the visual processing of face and non-face objects can follow independent developmental trajectories.

However, these results could be also explained by the fact that face recognition deficits are exacerbated by within-class discrimination, whereby several exemplars must be recognised within the same category, whilst such ability is not usually required in object recognition tests employed by several studies (e.g., the Birmingham Object Recognition Battery; Riddoch & Humphreys, 1993). Moreover, many of these studies only report accuracy data, which alone may not be sensitive to detect deficits when participants trade speed for accuracy (see Gauthier, Behrmann, & Tarr, 1999).

The degree of dissociability between face and object recognition poses interesting questions on the nature of the processes that regulate both. Evidence of face recognition impairments with spared object recognition supports the view that face processing relies, at least in part, on dedicated mechanisms. This idea has been supported by evidence gathered from neuropsychological cases (e.g., Farah, 1996; McNeil &

Warrington, 1993; Busigny & Rossion, 2010; Riddoch, Johnston, Brecewell, Boutsen, & Humphreys, 2008; Busigny, Graf, Mayer, & Rossion, 2010), neuroimaging (e.g., Kanwisher, McDermott, & Chun, 1997; Grill-Spector, Knouf, & Kanwisher, 2004; Yovel & Kanwisher, 2004), electrophysiological (e.g., Bentin, Allison, Puce, Perez, & McCarthy, 1996), and behavioural (e.g., Leder & Carbon, 2006; Young, Hellawell, & Hay, 1987) studies.

An alternative account postulates the existence of common, or partially overlapping, recognition mechanisms subserving the visual processing of both faces and objects (Tarr & Cheng, 1993; O'Toole, Jiang, Abdi, & Haxby, 2005; Haxby, et al., 2001; Gauthier, Skudlarski, Gore, & Anderson, 2000). In support, the neural substrates of face and non-face object recognition are located in adjacent brain areas, and a certain degree of overlapping has been found (e.g., Grill-Spector, 2003; Kanwisher, 2000). However, only little association (i.e., 13.6% of shared variance) was found in typically developed individuals between their perfomances on the CFMT and the CCMT (Dennett et al., 2011), suggesting that face and object recognition (cars in this case) may rely onto different mechanisms.

Remarkably, any comparison between the visual processing of face and general objects must take into account certain differences between the two categories. For example, the discrimination among different examplars of non-face objects, alike face recognition, relies greatly on local features (e.g., Farah, Levinson, & Klein, 1995; Farah, 1992). Greebles stimuli have been created to find a control class of objects whose within-class recognition could be closely comparable to that of faces (Gauthier, 1998). Greebles consist of a novel object category with a fixed spatial configuration and distinguishable small local components. Similarly to faces, they can only be discriminated by looking at their second-order properties (i.e., individual parts).

A compelling account challenges the assumption that a lack of association between face and non-face objects recognition may necessarily reflect separable neurocognitive mechanisms. In fact, the ability to recognise certain objects could be modulated by the degree of perceptual experience that individuals have with that class of objects (Gauthier et al., 2014; Wang, Gauthier, & Cottrell, 2016). Faces, for example, can be seen as a peculiar object for which people have developed a high degree of expertise due to constant exposure and to their critical evolutionary importance (Diamond & Carey, 1986; Gauthier & Tarr, 1997). In support to this claim, the Fusiform Face Area (FFA), a region showing selective activation for face stimuli compared to other objects

(Kanwisher, McDermott, & Chun, 1997; Grill-Spector, Knouf, & Kanwisher, 2004), strongly responds also to non-face objects of expertise (e.g., McGuin, Newton, Gore, & Gauthier, 2014; Xu, 2005; Gauthier, Skudlarski, Gore, & Anderson, 2000; Gauthier, Tarr, Anderson, Skudlarski, & Gore, 1999). Therefore, so called 'face-specific' mechanisms may also support the processing of other visual stimuli that share common perceptual characteristics with faces, for example a good amount of experience individuating within-class exemplars.

Interestingly, in support of the expertise hypothesis, Greebles have been used as control stimuli in a paradigm where subjects undertake a learning training until they become fast and precise at individuating different exemplars (for the procedure see Gauthier & Tarr, 2002). Evidence shows that Greeble expertise recruites the same neuro-cognitive mechanisms of face recognition, with selective activations of FFA (e.g., Gauthier & Tarr, 2002), face-like electrophysiological responses (e.g., Rossion, Gauthier, Goffaux, Tarr, & Crommelinck, 2002) and the employment of holistic processing (e.g., Gauthier, Williams, Tarr, & Tanaka, 1998).

Despite several perceptual similarities between faces and Greebles, a case of DP with intact recognition of Greebles has been reported (Duchaine, Dingle, Butterworth, & Nakayama, 2004), suggesting the existence of separable mechanisms for face and Greebles processing. However, the conclusions of this study have been challenged on the basis of the lack of a control task using face stimuli to be compared with the Greeble recognition task, and the unknown underlying causes of prosopagnosia in the DP individual tested, particularly those regarding the acquisition of expertise (see Bukach et al., 2012). Interestingly, Behrmann and colleagues (2005) found that a group of 5 individuals with DP and 3 acquired prosopagnosics were remarkably slower at fine-grained discriminations of Greebles compared to normal controls, indicating that impairments in non-face novel objects might be elicited by RT rather than accuracy. Similarly, a comprehensive investigation of Greeble expertise attainment in a case of acquired prosopagnosic showed that despite the patient could reach the typical expertise criterion, replicating Duchaine et al. (2004), his learning trajectory was slower and used qualitatively different mechanisms compared to normal controls (Bukach et al., 2012).

To date there are no definitive studies which can demonstrate whether faces are processed by domain-specific or domain-general mechanisms. Future studies using the DP population should include i) large samples, ii) comparable face and non-face objects tasks, iii) measures of speed/accuracy trade-offs, and iv) should take into account the contribution of expertise.

1.3.5.3 The co-occurrence hypothesis

Emerging theories have proposed alternative interpretations of the complex heterogeneity of DP. For example, Gray and Cook (2018) have advanced the possibility that different forms of developmental agnosia may co-occur as independent conditions (i.e., the Independent Disorders Hypothesis, IDH). This idea is strengthened by a factual observation that co-occurrence is a common phenomenon of many developmental disorders, and by the possibility that common genetic or environmental factors may play a role in the development of neural substrates involved in general visual processing (e.g., Kaplan, Dewey, Crawford, & Wilson, 2001).

Accordingly, for the IDH there is no such thing as DP disorder, but rather people inherit a predisposition to abnormal neural development of occipito-temporal areas (Gray & Cook, 2018) which are involved in the visual processing of both face and non-face objects (e.g., Grill-Spector, 2003; Kanwisher, 2000). For example, aberrant development can be observed as reduced integrity of white matter (WM) tracts connecting these areas (Thomas et al., 2009; Song et al., 2015), and it has been shown that depending on the specific location of WM alterations it is possible to predict perceptual impairments for non-face objects in subjects with DP (Gomez et al., 2015).

The IDH implies the existence of pure forms of developmental agnosia, which can only affect a specific category of visual stimuli, as the occurrence of one disorder simply increases the predisposition to develop another disorder. This is in clear contrast with the idea that co-occurrence of face and object recognition problems is due to common underlying domain-general mechanisms.

1.3.5.4 Non-visual impairments

1.3.5.4.1 Vocal recognition in DP

Less focus has been given to non-visual deficits in DP. There are many similarities between face and voice identity recognition. Leading neuro-cognitive models of voice recognition share many parallels with the Bruce and Young (1986) model of face perception (e.g., Ellis, Jones, & Mosdell, 1997; Belin, Fecteau, & Bedard, 2004). After initial structural encoding of the auditory stimulus, voices are analysed by independent systems which work in parallel and are responsible for the extraction of speech information, vocal affect information and vocal identity information (Belin et al., 2004; for a summary see Garrido et al., 2009).

Moreover, both face and voice recognition activate the right anterior temporal lobe, whose damage has been associated to associative forms of acquired prosopagnosia (Barton, 2008; Pancaroglu et al., 2011). Interestingly, patients with acquired prosopagnosia following lesions in the anterior temporal lobe have been reported to show impaired voice recognition only when damage was bilateral (Liu, Pancaroglu, Hills, Duchaine, & Barton, 2014). The authors advanced the hypothesis that a multimodal person recognition syndrome may occur when associative variants of prosopagnosia are accompanied by bilateral anterior temporal lesions, resulting in deficits recognising the identity of a person by their face and voice.

Along with congenital cases of face recognition impairment, cases of developmental phonagnosia have also been described showing severe voice recognition difficulties in the absence of brain damage (Garrido et al., 2009; Herald, Xu, Biederman, Amir, & Shilowich, 2014; Roswandowitz et al., 2014). This observation together with data from lesion studies, suggest that similar findings can emerge in DP. Given the existence of developmental forms of apperceptive and associative prosopagnosia, the latter may be associated with difficulties in voice recognition which could stem from a wider multimodal person recognition syndrome.

To date there are only a few reports of impaired voice recognition in DP (e.g., Liu, Corrow, Pancaroglu, Duchaine, & Barton, 2015; von Kriegstein, Kleinschmidt, & Giraud, 2006) and the picture that emerges from these studies reveals that voice-related deficits are quite rare within DP. For example, Liu and colleagues (2015) found that only 1 of their 12 prosopagnosics showed voice recognition impairments. In von Kriegstein and colleagues (2006) a single case of DP was examined who revealed a selective impairment at familiar voice recognition but normal unfamiliar voice perception. Further investigations found that alike controls DP participants failed to show a face-benefit effect when learning voices paired to facial identities (von Kriegstein et al., 2008). Overall, these results show that DPs tend to exhibit typical voice recognition in unimodal testing conditions (i.e. when participants have to learn the voice alone), but their vocal identity recognition is not enhanced by multisensory interaction, resulting in worse perfomances at familiar voice recognition (for a review see Maguinness & von Kriegstein, 2017).

1.3.5.4.2 Topographic processing in DP

Other non-visual deficits have been found in topographic processing. Often, individuals with DP report difficulties in spatial navigation (e.g., Duchaine, Parker, & Nakayama, 2003; Grueter et al., 2007). Formal testing focussing on landmark recognition and route learning found that DP participants show normal performance on these tests, contrary to anecdotal evidence (Corrow et al., 2016). However, when tested on topographic memory tasks which require participants to generate and apply metric representations of the environment, Klargaard and colleagues (2016) found that a group of DPs was less able to retain topographic information compared to typically developed individuals. Interestingly, the poor performance was not associated to visual short-term memory, as some participants with DP had preserved topographic memory despited impaired face memory. The authors suggest that while topographic memory impairments may co-occur with DP, the two conditions have to be taken as independent.

1.4 Neural accounts

1.4.1 Neural models of typical face perception

The processing of faces proceeds through several functional modules which are supported by dedicated brain areas in the bilateral ventral occipito-temporal cortex, forming the core system of face perception (e.g., Haxby, Hoffman, & Gobbini, 2000; Kanwisher, McDermott, & Chun, 1997; Grill-Spector, Knouf, & Kanwisher, 2004; Tsao & Livingstone, 2008) (Figure 1.1b). Neuroimaging studies have repeatedly observed that these areas show greater response for faces than non-face objects in typically developed individuals (e.g., Kanwisher et al., 1997; Gauthier et al., 2000; for a review see Duchaine & Yovel, 2015). Alongside neuroimaging evidence, lesion studies and brain stimulation experiments have supported the functional organization of the face-processing network. Cases of acquired prosopagnosia have been reported to occur after damage in the occipito-temporal cortex, particularly in areas such as the OFA and the FFA (e.g., Rossion et al., 2003; Sorger, Goebel, Schiltz, & Rossion, 2007; Barton, 2008). Transient prosopagnosia also emerges from temporary deactivation of core regions using transcranial magnetic stimulation (TMS) (e.g., Pitcher, Garrido, Walsh, & Duchaine, 2008; Pitcher, Walsh, Yovel, & Duchaine, 2007).

1.4.1.1 The Haxby, Hoffman, & Gobbini model

Haxby, Hoffman and Gobbini (2000) proposed a neural model of face perception based on the leading cognitive model by Bruce and Young (1986). In this model, each step of face perception is subtended by a dedicated brain area in the occipito-temporal cortex. The inferior occipital gyrus includes the occipital face area (OFA) and underpins the early visual processing, allowing for the structural encoding of face-like percepts. From this point, different aspects of face perception are analysed by dedicated areas: The fusiform gyrus encompasses the fusiform face area (FFA), which subtends the extraction of information that can be associated to a specific identity allowing for face recognition; whilst the superior temporal sulcus (STS) supports the analysis of changeable aspects of faces (e.g., expression, eye gaze, lip movement).

Together with the core system, an extended system is involved in those aspects of face recognition which involve access to semantic memory and the social cognition domain (see Gobbini & Haxby, 2007) (Figure 1.1b). For example, perceiving a familiar face will trigger episodic and biographical information associated to that identity, eliciting an emotional response towards that person. The extraction of personal knowledge is supported by the anterior paracingulate, which encodes personal traits and attitudes, the posterior STS/temporoparietal junction, which regulates those processes associated with Theory of Mind, the anterior temporal cortex, which retrieves biographical information, and the precuneus, which retrieves episodic memories. Finally, the emotional response is regulated by the amygdala, the insula and the striatum.

1.4.1.2 The Duchaine & Yovel model

Recently, a new neural core system of face processing has been proposed, based on emerging evidence that could not be explained solely by referring to the traditional models (Duchaine & Yovel, 2015). The newly proposed framework includes the three core areas of traditional models (i.e., the FFA, the OFA, and the STS) and three new areas which have been identified recently; The anterior temporal lobe face area (ATL-FA), the anterior superior temporal sulcus face area (aSTS-FA) and the inferior frontal gyrus face area (IFG-FA). These areas have been reported to function into two distinct pathways which interact with each other. On one hand, the ventral pathway, which includes the OFA, FFA, and ATL-FA, is responsible for the processing of information associated to the structure and the surface of faces, allowing the perception of identity, age, sex, and expression. On the other hand, the dorsal pathway, consisting of pSTS-FA, aSTS-FA, and IFG-FA, represents changeable aspects of faces, like expression, eye gaze, and mouth movements, and it is involved during the percepetion of dynamic faces.

Most research on the neural functional correlates of DP has focused on the functioning of areas in the core and extended systems. Below I provide an overview of the key findings within three main methodological areas.

1.4.2 Electrophysiological findings

Event-related brain potential (ERP) technique is a direct measure of the brain electrical response to a perceptual event. Research in face perception has focussed on the N170, a component which has been associated to the neural processing of faces. Compared to other visual stimuli, faces elicit greater negativity 140-200 ms after stimulus presentation in electrodes placed in occipito-temporal sites, particularly in the right hemisphere (e.g., Bentin, Allison, Puce, Perez, & McCarthy, 1996; Allison, Puce, Spencer, & McCarthy, 1999; Eimer, 2000). Specifically, N170 has been related to face structural encoding, in particular to early stages of face detection and perception of first-order relations (e.g., Ghuman, et al., 2014; Rossion & Jacques, 2008). Interestingly, given the close relationship of N170 with the analysis of face configuration, this component is highly influenced by face inversion, with larger and delayed N170 responses to inverted compared to upright faces (e.g., Bentin et al., 1996; Itier, Alain, Sedore, & McIntosh, 2007; Eimer, 2000). Leading interpretations of this phenomenon suggest that inverted faces might trigger additional neural processes involved in object perception (e.g. Rossion et al., 2000; but see Towler & Eimer, 2012). Alternatively, face-selective neural populations may increase their response to faces which are more difficult to perceive (Towler, Fisher, & Eimer, 2017).

The study of the N170 component in DP helps to clarify the nature of the underlying causes of impairment (for a review see Towler & Eimer, 2012 and Towler, Fisher, & Eimer, 2017). Since N170 reflects early structural encoding, any abnormality in this component may be reflective of apperceptive forms of DP, whereby the deficit occurs at early stages of perception. On the other hand, mnemonic subtypes of DP may not be revealed by simply looking at the N170, given that their deficits occur later on in the processing stream, and involve the association of the percept with identity representations stored in long term memory.

The heterogeneity found in behavioural studies reflects in the ERP findings where only some of the prosopagnosics tested showed reduced face-sensitivity of the N170, whilst others appeared to have typical neural responses (e.g., Bentin, Deouell, & Soroker, 1999; Kress & Daum, 2003; Harris, Duchaine, & Nakayama, 2005; Righart & de Gelder, 2007; Minnebusch, Suchan, Ramon, & Daum, 2007; Bentin, DeGutis, D'Esposito, & Robertson, 2007; Nemeth, Zimmer, Schweinberger, Vakli, & Kovacs, 2014; Rivolta, Palermo, Schmalzl, & Williams, 2012). A study using a sizeable group of 16 DPs found normal face-sensitive N170 responses to upright faces, but failed to show the typical enhanced N170 for the inverted faces in the DP group , suggesting that they may process similarly upright and inverted faces at the neural level (e.g., Towler, Gosling, Duchaine, & Eimer, 2012).

Alternatively, the typical N170 response in DPs may be triggered by the processing of the eye region which is known to elicit this component (Bentin, Golland, Flevaris, Robertson, & Moscovitch, 2006), and this interpretation is consistent with the idea that DP affects global rather than local processing (e.g., Avidan, Tanzer, & Behrmann, 2011; Carbon, Grüter, Weber, & Lueschow, 2007; Liu & Behrmann, 2014; Lobmaier, Bölte, Mast, & Dobel, 2010; Palermo et al., 2011). However, even when tested using Mooney faces, which prevent the engagement of feature-based processes, DPs show face-sensitive N170 responses, indicating that they may also rely on some global information at early stages of face perception (Towler, Gosling, Duchaine, & Eimer, 2016).

Despite normal activations of face-selective posterior areas within 200 ms from stimulus onset, prosopagnosics fail to show the typical pattern of enhancement and delay of N170 to inverted faces (e.g., Towler, Gosling, Duchaine, & Eimer, 2012) and scrambled faces (Towler, Parketny, & Eimer, 2016), suggesting that DPs may be less sensitive not only to face configuration but also to changes in the canonical spatial arrangement of local components. Towler and colleagues (2017) have suggested that the lack of modulation of N170 with non-canonical faces in DPs may reflect the fact that prosopagnosics recruit additional neural populations which respond to non-face objects. Therefore, they may treat faces and objects as similar perceptual stimuli. Alternatively, face-selective neurons that fire more with more difficult stimuli (i.e. inverted and scrambled faces) in neurotypical individuals, may respond similarly to both canonical and non-canonical faces in the DP population, suggesting that the difficulty does not vary sistematically for prosopagnosics.

1.4.3 Neuroimaging findings

1.4.3.1 Structural findings

Studies looking at the structural correlates of DP reveal a more striking picture, and white matter (WM) tracts appear a suitable candidate in the search for neural markers of prosopagnosia. White-matter tracts are bundles of myelinated axons which connect different brain areas. Reportedly, structural properties of the inferior longitudinal fasciculus (ILF), a large tract connecting occipital with temporal regions, have been associated with a variety of behaviours, including face recognition (e.g., Catani, Jones, Donato, & Ffytche, 2003; Gschwind, Pourtois, Schwartz, Van De Ville, & Vuilleumier, 2012; Pyles, Verstynen, Schneider, & Tarr, 2013; Saygin et al., 2012). Where high tract integrity facilitates information exchange between occipital and fusiform face areas, individuals may experience excellent face perception (e.g., Tavor et al., 2004). Conversely, where information exchange within this network is impaired, individuals may exhibit perceptual deficits for faces (e.g., Thomas et al., 2009; Gomez et al., 2015; Behrmann, Avidan, Gao, & Black, 2007).

Thomas and colleagues (2009) tried to functionally define ILF in relation to face-specific deficits observed in DP and found that reduced structural integrity in DPs correlated with errors in face recognition. Interestingly, more recent studies reported that local WM properties of ILF, rather than the entire fasciculus, accounted for category-specific behaviours (Gomez et al., 2015; Song et al., 2015). In Gomez et al. (2015) WM local properties of fibres within the ILF interconnecting face-selective regions correlated with performance on the Benton Face Recognition task; while integrity of the local structure of fibres in place-selective areas correlated with accuracy in a scene-recognition memory test. These findings extended and complemented previous research, showing that WM associated with specific regions may be a better predictor of behavioural performance. Crucially, a "local approach" in connectivity studies helps explaining the variability of ILF perturbation across different DPs revealed by Thomas et al. (2009). Since ILF interconnects areas in the visual cortex which mediate different functions, it is possible that different profiles of disruption may account for the individual differences widely observed in DP.

The evidence of differential WM properties in face-selective areas observed in DPs implicates that behavioural deficits associated with the condition may arise from aberrant neuro-structural development affecting the signal propagation through the core system of face perception (see Behrmann, Avidan, Gao, & Black, 2007). In fact, standard neuroimaging scans of DP individuals fail to show any apparent lesions or neurological anomalies of specific areas (e.g., Jones & Tranel, 2001; Kress & Daum, 2003; Duchaine & Nakayama, 2006; but see Behrmann et al., 2007). However, more sensitive techniques have shown differences in the microstructure of face-selective core regions in prosopagnosics. Reduced grey matter volume in areas such as the inferior temporal lobe, STS and FFA has been observed in DP, and these abnormalities are associated with face recognition performance (Garrido et al., 2009; Behrmann et al., 2007).

1.4.3.2 Functional findings

Activation studies using functional magnetic resonance imaging (fMRI) have repeatedly shown that the core regions of face perception of neurotypical adults generate stronger responses to faces than other objects (e.g., Kanwisher, McDermott, & Chun, 1997; Puce, Allison, Asgari, Gore, & McCarthy, 1996). Moreover, BOLD signal suppression can be observed in the fusiform gyrus and in STS following the repetition of the same identity or expression, respectively (e.g., Winston, Henson, Fine-Goulden, & Dolan, 2004; Fox, Moon, Iaria, & Barton, 2009; Rotshtein, Henson, Treves, Driver, & Dolan, 2005). In the DP population findings are more inconsistent: Some individuals show reduced or absent face-selective responses in the core regions (e.g., Hadjikhani & De Gelder, 2002; Minnebusch, Suchan, Köster, & Daum, 2009; Bentin, DeGutis, D'Esposito, & Robertson, 2007; Furl, Garrido, Dolan, Driver, & Duchaine, 2011; Dinckelacker et al., 2011), but the vast majority of DPs reveal typical activations (e.g., Hasson et al., 2003; Avidan, Hasson, Malach, & Behrmann, 2005; Bentin, DeGutis, D'Esposito, & Robertson, 2007; Zhang, Liu, & Xu, 2015; Avidan et al., 2014). A single case of DP has been reported showing typical activation of the left FFA, but reduced response in the right FFA to familiar and unfamiliar faces (von Kriegstein, Kleinschmidt, & Giraud, 2006). Furthermore, a few studies have shown typical repetition suppression to familiar faces in DPs (e.g., Avidan, Hasson, Malach, & Behrmann, 2005; Williams, Berberovic, & Mattingley, 2007).

Furl and colleagues (2011) suggest the possibility that the inconsistency found in functional studies may reflect the fact that face recognition is distributed in the general population, with DP representing the lower end of the distribution. Therefore, affected mechanisms may function less well than those of neurotypical individuals without necessarily being qualitatively different or absent. Consequently, neural responses in DPs would be reduced and related to behavioural performance, rather than completely non-existent. Accordingly, using a sizeable sample of 15 DPs the authors found an overall reduced face-selectivity in the bilateral FFA in the experimental group. Critically, the neural response of the right fusiform gyrus was positively related to face identity recognition performance, but not to expression or object recognition, in the whole sample, corroborating the prediction that heterogeneous functional results in DP indicate *quantitative* individual differences in this population (Furl et al., 2011).

An alternative account has been proposed recently by looking at the size of receptive fields (RF) of neuronal populations in the core face-selective regions of individuals with DP (Witthoft et al., 2016). Participants with DP showed smaller RFs in the core regions of face processing, but not in early visual cortex, compared to typically developed individuals. Crucially, this difference in size positively correlated with behavioural performance on a face recognition task. Since smaller RFs are associated with reduced spatial integration, the authors conclude that DPs may be able to perceive facial local information, but they might struggle integrating visual information from disparate face regions into a perceptual whole (Witthoft et al., 2016). The evidence provided by this study is in line with behavioural evidence of reduced holistic processing in DP (e.g., Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014; Palermo et al., 2011). Moreover, it helps unravelling the lack of functional differences in the face-selective regions of most DPs. In fact, face selectivity could purely emerge from neural responses to facial features irrespective of the extent of visual field that is actually attended by the observer.

Effective connectivity has also been investigated in DP (Lohse et al., 2016). Dynamic causal modeling (DCM) allows to estimate the directionality of the information flow within a pre-defined functional network. Lohse and colleagues (2016) found that face-related visual information modulates the coupling of regions in the face processing network. In particular, three main feedforward effective connections were found between the early visual cortex (EVC) and OFA, FFA and the posterior STS. Crucially, in the DP subjects the connection strength was reduced in EVC-FFA (bilaterally) and EVC-right pSTS connections, and this diminished connectivity was associated to reduced face selectivity in those occipitotemporal areas. These findings are in line with the hypothesis that deficits in DP may result from a failure in signal propagation from occipital to temporal regions (Behrmann, Avidan, Gao, & Black, 2007), and are further corroborated by structural evidence of major physical alterations in the ILF of DPs (e.g., Thomas et al., 2009).

1.5 Objectives

Taken together, the outcomes concerning the cognitive mechanisms of DP reported in the reviewed literature thus far have been controversial, yielding mixed results. Whether mixed findings are attributable to differences in methodology across studies, or to different compositions of experimental samples, is debatable. What does emerge is that the main theoretical accounts of DP seem to diverge around two main empirical open questions: the first concerning the type of mechanisms involved (e.g. apperceptive versus mnemonic), the second regarding the nature of the deficit (e.g. domain-specific versus domain-general). Apperceptive accounts of DP have attributed face recognition impairments to reduced holistic processing of faces and poor perceptual encoding. On the other hand, mnemonic accounts ascribed the deficits to difficulties maintaining face percepts over time. Regarding the specific nature of deficits, some studies found face-selective impairments, whilst others attributed co-occurring non-face object recognition problems to a domain-general impairment.

The present thesis sought to systematically assess these main open empirical questions using sensitive psychophysical tasks and sizable samples of individuals with DP, tested on several diagnostic measures and in controlled settings. Chapter 2 aimed to explore the apperceptive versus non-apperceptive debate by looking at facial emotion recognition in DP, an ability that is dissociated from face memory according to leading models of face perception. Chapter 3 sought to explore the specificity of deficit by investigating face, body, and object recognition using a task in which the three conditions were matched in cognitive demand. Chapter 4 re-examined the susceptibility of DPs to the composite face illusion using two independent samples of prosopagnosics and two slightly different versions of the composite face task. Finally, Chapter 5 sought to examine the contribution of perceptual encoding and short term face memory in DP using a delayed match-to-sample task where view-point and retention interval were systematically manipulated.

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Chapter 2: Impaired perception of facial emotion in developmental prosopagnosia

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Chapter 2: Impaired perception of facial emotion in developmental prosopagnosia

2.1 Abstract

Developmental prosopagnosia is a neurodevelopmental condition characterised by difficulties recognising faces. Despite severe difficulties recognising facial identity, expression recognition is typically thought to be intact in developmental prosopagnosia; case studies have described individuals who are able to correctly label photographic displays of facial emotion, and no group differences have been reported. This pattern of deficits suggests a locus of impairment relatively late in the face processing stream, after the divergence of expression and identity analysis pathways. To date, however, there has been little attempt to investigate emotion recognition systematically in a large sample of developmental prosopagnosics using sensitive tests. Interestingly, when questioned, many prosopagnosics report problems recognising expressions in their daily lives, raising the possibility that expression recognition difficulties may be more common in this population than currently thought. In the present study, we describe three complementary psychophysical experiments that examine the recognition of facial and vocal emotion in a sample of 17 developmental prosopagnosics. In Experiment 1, we investigated observers' ability to make binary classifications of whole-face expression stimuli drawn from morph continua. Psychophysical analyses revealed diminished ability to classify morphed facial expressions in our sample of developmental prosopagnosics, relative to typical observers. We replicated this group difference in Experiment 2 when observers judged facial emotion using only the eye-region (the rest of the face was occluded). In our third experiment, we examined the ability of observers to classify the emotion present within segments of vocal affect. Despite difficulties judging facial emotion, the prosopagnosics exhibited excellent recognition of vocal affect. Contrary to the prevailing view, our results suggest that many prosopagnosics do experience difficulties classifying expressions, particularly those with apperceptive profiles. These individuals may have difficulties forming view-invariant structural descriptions at an early stage in the face processing stream, before identity and expression pathways diverge.

2.2 Introduction

Developmental prosopagnosia[2] (DP) is a lifelong neurodevelopmental disorder associated with impaired face recognition, thought to affect as many as one in every 50 people (Kennerknecht et al., 2006; Kennerknecht, Ho, & Wong, 2008). Individuals with DP exhibit deficits recognising personally familiar faces as well as problems discriminating unfamiliar faces, despite normal intelligence, typical low-level vision, and an absence of manifest brain injury (Behrmann & Avidan, 2005; Duchaine & Nakayama, 2006b; Susilo & Duchaine, 2013). Due to characteristic problems with face recognition, individuals with DP often utilise cues derived from hairstyle, voice, and gait, for person recognition. Nevertheless, recognising familiar people encountered out of context or following changes in external appearance, can prove challenging (Shah, Gaule, Sowden, Bird, & Cook, 2015).

The precise origin of the face recognition deficits seen in DP remains unclear. Cognitive accounts have argued that, relative to typically developing (TD) individuals, DPs exhibit reduced holistic processing of faces – whereby individual features (eyes, nose, mouth) are integrated into a coherent unified whole – compromising the accuracy and efficiency of their face recognition (Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014; Palermo et al., 2011). At the neurological level, differences in cortical structure (Behrmann, Avidan, Gao, & Black, 2007; Garrido et al., 2009), structural (Gomez et al., 2015; Thomas et al., 2009) and functional connectivity (Avidan & Behrmann, 2009; Avidan et al., 2013) have been observed in inferotemporal regions including the fusiform gyrus, a region thought to be crucial for face processing (Kanwisher, 2000). Strikingly, DP often runs in families (Duchaine, Germine, & Nakayama, 2007; Johnen et al., 2014; Lee, Duchaine, Wilson, & Nakayama, 2010; Schmalzl, Palermo, & Coltheart, 2008), suggestive of a genetic component.

The characteristic deficits of facial identity recognition seen in DP have attracted substantial research attention (Susilo & Duchaine, 2013). However, there has also been considerable interest in the expression recognition abilities of individuals with DP. The facial expressions of others are a rich source of social information, conveying cues to affective and mental states (Adolphs, 2002; Frith, 2009; Parkinson, 2005). The ability

[2] We use the term Developmental Prosopagnosia in preference to Congenital Prosopagnosia to reflect the possibility that the condition emerges during development, and may not necessarily be present from birth.

to interpret facial expressions correctly is therefore important for fluent social interaction and wider socio-cognitive development. Moreover, the question of emotion recognition in DP also has important implications for neurocognitive accounts of the condition (Bate & Bennetts, 2015; Kress & Daum, 2003a). Where observed together, difficulties recognising facial identity and facial emotion are suggestive of apperceptive prosopagnosia (De Renzi, Faglioni, Grossi, & Nichelli, 1991); difficulties may arise early on in the face processing stream, leaving observers unable to form an accurate, view-invariant description of face shape (Bruce & Young, 1986; Haxby, Hoffman, & Gobbini, 2000). Alternatively, intact expression recognition despite impaired recognition of facial identity suggests a locus of impairment relatively late in the face processing stream, after the divergence of expression and identity analysis pathways (Bruce & Young, 1986; Duchaine, Parker, & Nakayama, 2003; Haxby et al., 2000).

Presently, difficulties recognising facial expressions are thought to be relatively uncommon in DP. Palermo et al. (2011) examined the performance of twelve DPs on three emotion recognition tests: *The Ekman 60 Faces Test,* in which participants label 60 greyscale images of prototypical basic emotions (Young, Perrett, Calder, Sprengelmeyer, & Ekman, 2002); *The Emotion Hexagon Test,* in which participants label expressions drawn from morph continua constructed from the six basic emotions[3] (Young et al., 2002); and *The Reading the Mind in the Eyes Test,* in which participants identify subtle social emotions from cues present around the eye region (Baron-Cohen, Wheelwright, Hill, Raste, & Plumb, 2001). Strikingly, the twelve DPs were unimpaired at both the group and single-case level, relative to aged-matched controls, on all three tasks (Palermo et al., 2011). Dobel, Bölte, Aicher & Schweinberger (2007) described intact emotion recognition in six DPs, having administered the Tübingen Affect Battery – a 4 alternative-forced-choice (AFC) emotion labelling task. Similar findings were reported by Humphreys, Avidan and Behrman (2007), having administered *The Emotion Hexagon Test* to three DPs[3], and Lee, Wilson, Duchaine and Nakayama (2010), having tested three DPs using *The Reading the Mind in the Eyes Test* and a 3AFC match-to-sample task. Several further studies of single cases have described intact emotion recognition in DP (Bentin, Degutis, D'Esposito, & Robertson, 2007;

[3] While expression stimuli were drawn from morph continua, psychophysical analyses were not employed (e.g., psychometric functions were not estimated). The authors' analysis was restricted to proportions of correct responses, defined through reference to the dominant emotion signal present in each stimulus.

Duchaine et al., 2003; Kress & Daum, 2003b; Nunn, Postma, & Pearson, 2001). Moreover, a study of four DPs indicated that they made typical judgements of facial trustworthiness (Todorov & Duchaine, 2008), an inference thought to be mediated by subtle emotion cues.

Nevertheless, many DPs report problems recognising facial expressions in their daily lives (e.g., Lee et al., 2010), and case studies have described individuals with DP, who *do* exhibit deficits of expression recognition (Ariel & Sadeh, 1996; De Haan & Campbell, 1991; Duchaine, Murray, Turner, White, & Garrido, 2009; Duchaine, Yovel, Butterworth, & Nakayama, 2006; Minnebusch, Suchan, Ramon, & Daum, 2007; Schmalzl et al., 2008). For example, Duchaine et al. (2006) described a 53-year-old male DP, Edward, who exhibited clear expression recognition impairments on *The Reading the Mind in the Eyes Test* and on a 3-AFC match-to-sample task. Similarly, De Haan and Campbell (1991) tested AB, the original case of DP first described by McConachie (1976), and found that as an adult she exhibited problems labelling prototypical basic emotions. Importantly, however, these reports are relatively infrequent (regarded as 'the exception' rather than 'the norm'), and no systematic investigation has found evidence for a group difference.

The present study sought to re-examine the expression recognition abilities of individuals with DP. As discussed above, this question offers critical insight into the locus of the perceptual difficulties seen in this condition. In particular, we sought to test systematically a large sample of DPs using sensitive tests. The ability of different tests to detect emotion recognition deficits varies widely. For example, Edward, the DP described by Duchaine et al. (2006), was substantially impaired on *The Reading the Mind in the Eyes Test* (4.1 standard deviations below the TD mean), but only mildly impaired on *The Emotion Hexagon Test* (1.4 standard deviations below the TD mean). In Experiment 1, we investigated observers' ability to make binary classifications of whole-face expression stimuli drawn from morph continua. In Experiment 2 observers judged facial emotion using only the eye-region (the rest of the face was occluded). In our third experiment, we examined the ability of observers to classify the emotion present within segments of vocal affect.

2.3 Neuropsychological testing

A group of 17 (11 females) individuals with DP participated in the study (Table 2.1). DP participants were recruited through www.troublewithfaces.org. All members of the DP sample described lifelong face recognition problems. None of the DPs had a history of brain injury or psychiatric disorder (e.g., Schizophrenia, Autism Spectrum Disorder). Convergent diagnostic evidence for the presence of DP was collected using the Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006a), the Cambridge Face Perception Test (CFPT; Duchaine, Germine, & Nakayama, 2007) and the Twenty-Item Prosopagnosia Index questionnaire (PI20; Shah, Gaule, Sowden et al., 2015). When administered in the upright orientation, performance on the CFMT correlated closely with scores on the CFPT (r = -.73, p < .001) and the PI20 (r = -.82, p < .001). There were also strong correlations between the PI20 and the CFPT (r = .61, p < .001). The prosopagnosics' scores on the CFMT and CFPT were compared with a comparison group of 35 age- and gender-matched TD controls. All but one of the DPs scored at least two standard deviations below the control mean (the remaining DP participant was 1.77 standard deviations below the TD mean). In addition to the face recognition tests, participants completed the Cambridge Car Memory Test (CCMT; Dennett et al., 2011) and the Cambridge Bicycle Memory Test (CBMT; Dalrymple, Garrido, & Duchaine, 2014) to assess their wider object recognition ability. In addition, the DPs were screened for colour blindness using Ishihara's Tests for Colour-deficiency (Ishihara, 1993).

2.4 Experiment 1

Measuring individual differences in expression recognition ability is not straight-forward. In particular, tasks that require participants to label prototypical emotional expressions (e.g., happy, sadness, fear, disgust, anger, surprise) may lack sensitivity due to ceiling effects or noise introduced by differences in guessing base-rates (Ipser & Cook, 2015). In our first experiment, we sought to determine whether DPs are impaired at making binary categorisations of whole-face emotional expression stimuli drawn from morph continua. Psychophysical modelling of categorisation probability yields sensitive and reliable estimates of expression recognition ability. Previous studies suggest that this approach can reveal group effects that may go undetected by simple labelling paradigms (Cook, Brewer, Shah, & Bird, 2013).

Participant	Age	PI20	CFPT upright	CFPT inverted	CFMT%	CBMT%	CCMT%	CFMT z-scores	CFPT z-scores	PI20 z-scores
F1	43	79	26	58	62.50	83.33	62.50	−2.18	−.45	−4.16
F2	46	61	68	58	52.78	80.56	73.61	−3.13	−4.25	−2.35
F3	22	89	30	70	50.00	77.78	52.78	−3.40	.00	−5.16
F4	42	92	62	82	45.83	94.44	65.28	−3.81	−3.58	−5.47
F5	70	95	100	92	36.11	–	76.39	−4.76	−7.83	−5.77
F6	40	85	40	82	40.28	59.72	63.89	−4.35	−1.12	−4.76
F7	50	78	34	52	58.33	91.67	86.11	−2.59	−.45	−4.06
F8	21	59	30	64	63.89	75.00	–	−2.04	.00	−2.15
F9	29	68	32	58	61.11	–	63.89	−2.31	−.22	−3.06
F10	63	79	40	70	61.11	80.56	66.67	−2.31	−1.12	−4.16
F11	53	85	74	94	45.83	88.89	63.89	−3.81	−4.92	−4.76
F12	65	81	44	78	59.72	93.06	66.67	−2.45	−1.57	−4.36
M1	47	92	86	54	51.39	98.61	94.44	−3.26	−6.26	−5.47
M2	46	72	50	64	66.67	90.28	76.39	−1.77	−2.24	−3.46
M3	68	92	92	78	27.78	70.83	47.22	−5.57	−6.94	−5.47
M4	43	78	52	50	58.33	94.44	93.06	−2.59	−2.46	−4.06
M5	28	62	46	60	62.50	77.78	69.44	−2.18	−1.79	−2.45
TD mean	40.48	37.35	29.40	60.00	85.00	83.22	61.45			
TD SD	13.31	10.75	9.48	14.47	8.48	12.26	13.15			

Table 2.1. Scores of each Developmental Prosopagnosic on the Twenty-Item Prosopagnosia Index (PI20), The Cambridge Face Perception Test (CFPT), The Cambridge Face Memory Test (CFMT), The Cambridge Bicycle Memory Test (CBMT), The Cambridge Car Memory Test (CCMT). The mean and standard deviation of the comparison sample ($N = 35$) are provided below. The z-scores provided for the CFPT are based on performance in the upright condition.

Should individuals with DP exhibit subtle expression recognition deficits, we reasoned that a psychophysical approach may be most likely to reveal these problems.

2.4.1 Methods

The performance of the DPs was compared with a group of 23 TD controls (6 males; $M_{\text{age}} = 42.65$, $SD_{\text{age}} = 13.44$). All TD participants were screened for DP (Table 2.2). All participants had normal or corrected-to-normal visual acuity. Ethical clearance was granted by the local ethics committee and the study was conducted in line with the ethical guidelines laid down in the 6th (2008) Declaration of Helsinki. All participants gave informed consent.

	DPs		TDs (Experiment 1)			TDs (Experiment 2)			TDs (Experiment 3)		
	M	SD	M	SD	Contrast	M	SD	Contrast	M	SD	Contrast
CFMT	53.18	10.94	83.68	9.94	$p < .001$	83.13	10.25	$p < .001$	83.41	10.34	$p < .001$
CFPT	53.29	23.22	32.48	8.48	$p < .010$	32.67	7.39	$p < .010$	32.24	9.38	$p < .010$
PI20	79.24	11.52	37.05	9.52	$p < .001$	36.95	9.60	$p < .001$	38.05	9.88	$p < .001$

Table 2.2. Performance of the Developmental Prosopagnosics (DPs) and the typically developing controls (TDs) used in Experiments 1-3 on The Cambridge Face Memory Test (CFMT), The Cambridge Face Perception Test (CFPT), and The Twenty-Item Prosopagnosia Index (PI20).

Three morph continua (happiness-anger, disgust-sadness, fear-surprise) were produced by blending incrementally two greyscale photographs of emotional facial expressions, produced by a single actor, selected from Ekman and Friesen's (1975) *Pictures of Facial Affect*. Image morphing was performed using Morpheus Photo Morpher Version 3.11 (Morpheus Software, Indianapolis, IN). Each continuum consisted of seven stimuli which varied in emotion intensity between 20% and 80% in equidistant 10% increments. Stimuli were cropped to exclude external features (e.g., ears, hairline) and presented in greyscale (Figure 2.1a).

Participants completed a computer-based task written in MATLAB (The MathWorks, Natick, MA) using Psychophysics Toolbox (Brainard, 1997; Pelli, 1997). Experimental trials presented a single image centrally for 1200 ms. Each stimulus subtended approximately 6.5° × 4.0° of visual angle when viewed at 60 cm. Following stimulus offset, participants were asked to make a binary categorisation about the stimulus image (e.g., happiness or anger?). Each of the 21 expression stimuli (3 continua × 7 levels of morph intensity) was presented 20 times in a randomised order.

Participants completed 6 practice trials before starting the experimental task. No feedback was provided during the practice or experimental procedures. In total, the procedure consisted of 420 trials and took approximately 20-25 minutes to complete.

Participants' responses were modelled by fitting cumulative Gaussian functions to estimate separate psychometric functions for the three continua. Function fitting was carried out in MATLAB using the Palamedes Toolbox (Prins & Kingdom, 2009). The key parameter of interest, inferred from the psychometric function, was the estimate of categorisation threshold. The threshold estimate is a measure of the precision with which stimuli are categorized and was defined as the standard deviation of the symmetric Gaussian distribution underlying each cumulative Gaussian function (subject to a log transform to attenuate positive skewing). Threshold estimates are inversely related to the slope of the psychometric function; steep and shallow slopes are associated with low and high threshold estimates, respectively. Lower threshold estimates indicate that observers can perceive subtle differences in stimulus strength and vary their responses accordingly. Greater threshold estimates reveal that participants' responses are relatively invariant to changes in stimulus strength, indicative of imprecise categorization.

2.4.2 Results and discussion

The threshold estimates obtained for the DP and TD groups are shown in Figure 2.1b. Threshold estimates were analysed using ANOVA with Continuum (happiness-anger, disgust-sadness, fear-surprise) as a within-subjects factor and Group (TD, DP) as a between-subjects factor. The analysis revealed a main effect of Continuum [$F(1.46, 55.35) = 46.68$, $p < .001$, $\eta_p^2 = .55$]. Contrasts indicated that fear-surprise categorisations were associated with greater thresholds ($M = 2.81$, $SD = .58$) than happiness-anger ($M = 1.59$, $SD = .87$) [$t(39) = 8.15$, $p < .001$] and disgust-sadness ($M = 2.33$, $SD = .48$) categorisations [$t(39) = 5.74$, $p < .001$]. Disgust-sadness categorisations were also associated with greater thresholds than happiness-anger categorisations [$t(39) = 5.25$, $p < .001$]. Crucially, the analysis also revealed a main effect of Group [$F(1,38) = 4.19$, $p = .04$, $\eta_p^2 = .10$]. Collapsing across the three continua, the DPs exhibited higher thresholds ($M = 7.26$, $SD = 1.54$) than the TD controls ($M = 6.36$, $SD = 1.24$). No Continuum × Group interaction was observed [$F(1.46, 55.35) = 1.02$, $p = .33$, $\eta_p^2 = .03$].

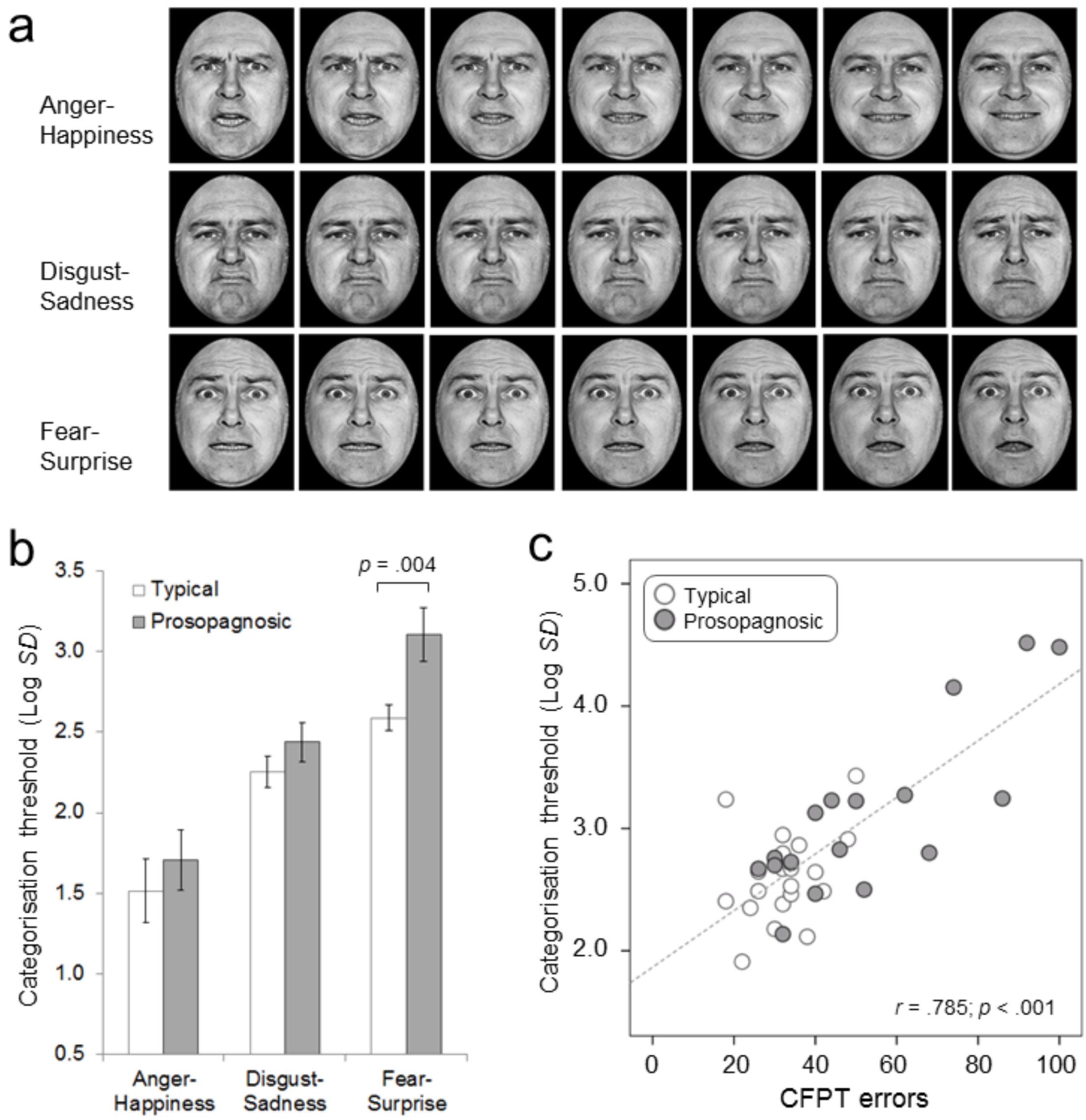

Figure 2.1. (a) Morphed expression stimuli used in Experiment 1. (b) Mean categorisation thresholds for the three continua exhibited by the typical observers and the developmental prosopagnosics. Error bars represent ±1 standard error of the mean. (c) Scatter plot of the relationship observed between participants' scores on the Cambridge Face Perception Test (CFPT) and their thresholds for the Fear-Surprise categorisations.

However, simple contrasts indicated a significant difference between the groups only in their fear-surprise thresholds, where the thresholds of the DP group ($M = 3.11$, $SD = .69$) were higher than those of the controls ($M = 2.59$, $SD = .37$) [$t(38) = 3.07$, $p = .004$]. Eight of the DPs scored at least one SD below the TD mean, and three (M3, F5, F11) were significantly impaired at single-case level (Figure 2.2a).

Clear correlations were observed between participants' categorisation thresholds for the fear-surprise continuum and their CFMT ($r = -.57$, $p <.001$) and PI20 ($r = .51$, $p =.001$) scores (Table 2.3). However, a striking correlation was found between participants' fear-surprise thresholds and their performance on the CFPT ($r = .78$, $p < .001$; Figure 2.1c).

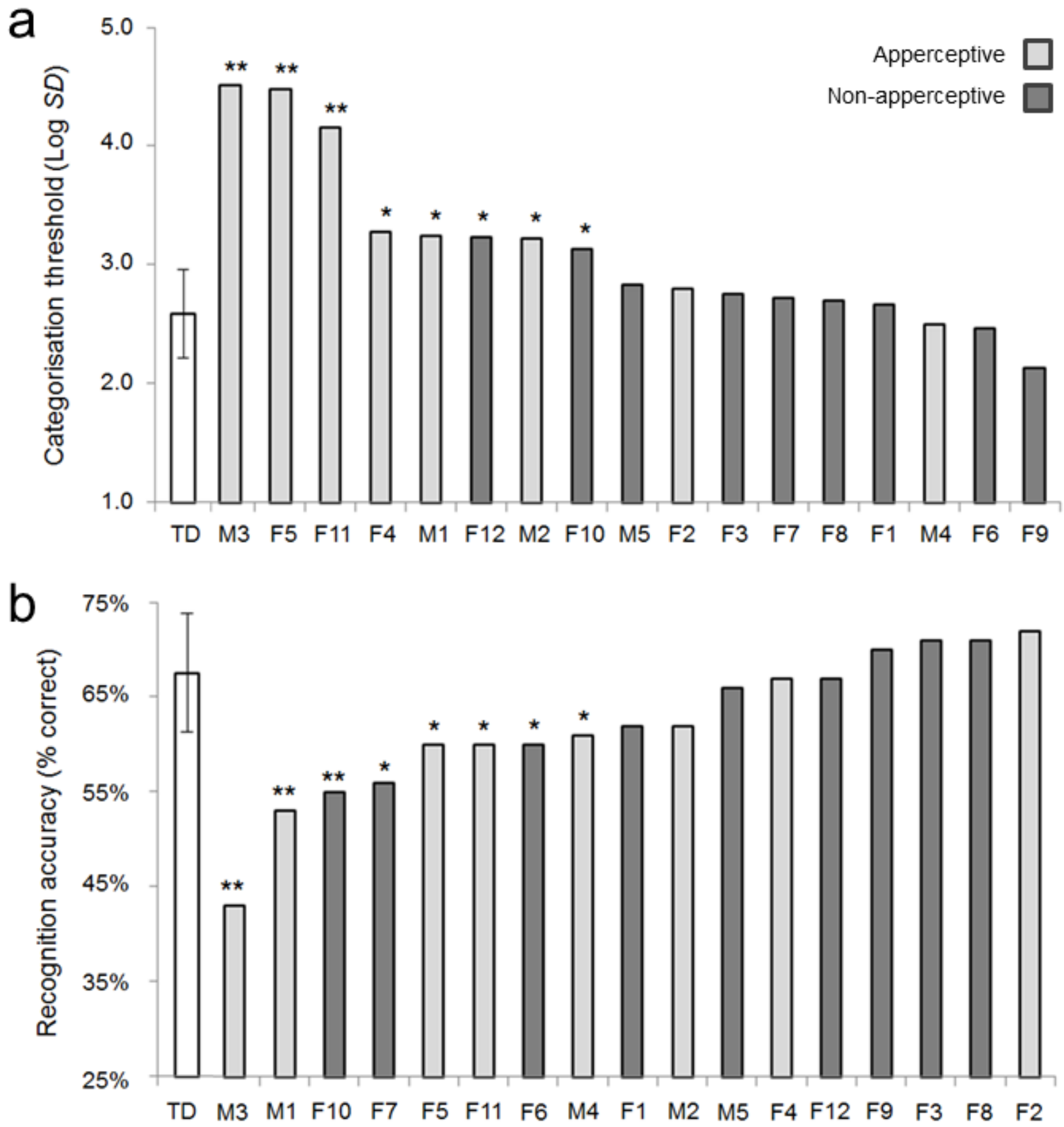

Figure 2.2. (a) Single-case analysis of the surprise-fear thresholds observed in Experiment 1. (b) Single-case analysis of the overall performance observed in Experiment 2. Error bars represent ±1 standard deviation. *denotes performance <1 standard deviation below the TD mean; **denotes performance at least 2 standard deviations below the TD mean.

To investigate this relationship further, the DP sample was split into two sub-groups based on their performance on the CFPT. Eight DPs who scored at least 2 SD below the control mean on the CFPT (Table 2.1), and the remaining nine DPs, were categorised as apperceptive and non-apperceptive, respectively. Simple contrasts revealed a significant difference in fear-surprise categorisation thresholds between the apperceptive subgroup (M = 3.54, SD = .73) and TD controls (M = 2.59, SD = .37) [$t(29)$ = 4.8, p < .001]. Interestingly, however, the fear-surprise categorisation thresholds of the non-apperceptive subgroup (M = 2.72, SD = .33) did not different significantly from the TD sample [$t(30)$ = .95, p = .35].

	CFMT		CFPT		PI20		CCMT		CBMT	
	r	p	r	p	r	p	r	p	r	p
Happiness-anger	−.23	.16	.24	.14	.08	.62	.00	.99	−.19	.29
Disgust-sadness	−.22	.19	.33	.04	.27	.09	.05	.80	.02	.92
Fear-surprise	−.57	<.001	.78	<.001	.51	<.001	.00	.98	−.03	.88
Mean threshold	−.46	.004	.61	<.001	.35	.03	.02	.92	−.13	.46

Table 2.3. Correlations between the expression categorisation thresholds observed in Experiment 1 and participants' scores on the Cambridge Face Memory Test (CFMT), the Cambridge Face Perception Test (CFPT), the 20-item Prosopagnosia Index (PI20), the Cambridge Car Memory Test (CCMT) and the Cambridge Bike Memory Test (CBMT).

The results of Experiment 1 suggest that our emotion categorisation task and the CFPT may tap very similar processes. The CFPT requires participants to rank order test faces according to their resemblance to a target face. Because the test and target faces are presented throughout each trial, the test is thought to measure observers’ ability to form perceptual descriptions of faces, under conditions of minimal working memory load. However, because the physical differences between test faces are subtle, the CFPT provides a demanding test of observers’ face encoding. Where perceptual descriptions are compromised, observers may be left unable detect and interpret subtle physical differences between stimuli, resulting in i) poor sorting performance on the CFPT and ii) judgements of expression that vary less closely with physical stimulus changes.

2.5 Experiment 2

The results of Experiment 1 suggest that relative to TD controls, individuals with DP are less able to categorise whole-face expression stimuli drawn from continua that morph emotional facial expressions. While our analyses suggest a trend for less precise categorisation overall, difficulties were particularly clear when observers were required to detect the subtle physical differences between stimuli drawn from the fear-surprise continuum. At least two accounts may be advanced to explain the group difference observed in Experiment 1. First, difficulties integrating information from disparate facial regions may prevent observers with DP forming unified perceptual descriptions of facial expressions. Consistent with this possibility, some observers with DP exhibit reduced composite interference for facial expressions (Palermo et al., 2011), suggestive of reduced holistic processing of facial emotion. Second, observers with DP may have a fundamental difficulty encoding the shape of local facial features. For example, cases of acquired prosopagnosia have been described who appear to have particular problems using information from around the eye region to discriminate (Bukach, Le Grand, Kaiser, Bub, & Tanaka, 2008) and recognise (Caldara et al., 2005) facial identities. Interestingly, problems using cues from the eye-region are thought to be associated with particular problems recognising facial expressions of fear (Adolphs et al., 2005).

In Experiment 2 we sought to distinguish these rival explanations by examining participants' ability to judge facial emotion using cues from the eye-region alone (i.e., a local region), using a variant of the *Reading the Mind in the Eyes Test* (Baron-Cohen et al., 2001). If the impairments observed in Experiment 1 arise from diminished integration of information from disparate facial regions, we reasoned that the DP group should perform typically on a task that does not require whole-face processing. However, if the impairment in emotion recognition is due to difficulties encoding the shape of local features, the group difference should still be evident.

2.5.1 Methods

The performance of the DPs was compared with a group of 23 TD controls (7 males; $M_{\text{age}} = 44.26$, $SD_{\text{age}} = 13.59$). All TD participants were screened for DP (Table 2.2). All participants had normal or corrected-to-normal visual acuity. Ethical clearance was granted by the local ethics committee and the study was conducted in line with the ethical guidelines laid down in the 6$^{\text{th}}$ (2008) Declaration of Helsinki. All participants gave informed consent.

The original *Reading the Mind in the Eyes Test* requires observers to recognise complex 'social emotions' (e.g., concerned vs. unconcerned, sympathetic vs. unsympathetic), and may therefore tax both mentalizing and perceptual processes. To minimize any mentalizing demands, our novel variant included different exemplars of four commonly encountered facial emotions. Stimuli were constructed from six Caucasian identities (3 females) selected from the Radboud Faces Database (Langner et al., 2010). For each identity, we produced four morph continua by blending images of the actor exhibiting a neutral expression, with images of the same actor expressing happiness, anger, fear and sadness[4]. The expression morphs containing 30%, 50% and 70% of each emotion (corresponding to low, moderate and high intensity) were cropped

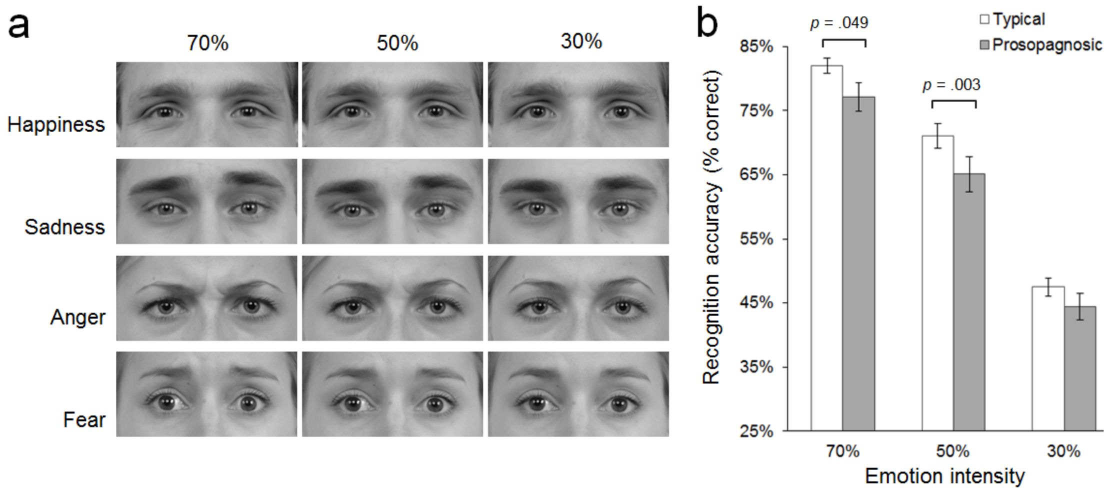

Figure 2.3 (a) Examples of the eye-region stimuli used in Experiment 2. (b) The mean recognition accuracy exhibited by the typical observers and the developmental prosopagnosics in the three emotion intensity conditions. Error bars represent ±1 standard error of the mean.

so that only the eye-region was visible, and presented in greyscale (Figure 2.3a). The position of the eyes in the resulting 72 images (6 identities × 4 emotions × 3 levels of emotion intensity) was standardised to ensure similar cues were available in each stimulus. Stimulus images subtended approximately 2.5° × 6.5° of visual angle when viewed at 60 cm.

Experimental trials presented a single stimulus centrally for 1200 ms, followed by a prompt to make a 4-AFC response (happiness, anger, fear, or sadness). The 72 stimuli were presented three times each, in a randomised order, yielding a total of 216 trials. The experiment was preceded by 6 practice trials. No feedback was provided during the practice or experimental procedures. The task lasted approximately 20 minutes. The experimental program was written in MATLAB using the Psychophysics Toolbox (Brainard, 1997; Pelli, 1997).

[4] Pilot testing of a 6-AFC procedure (happiness, anger, disgust, sadness, fear, and surprise) revealed that typical participants were unable to reliably distinguish i) angry and disgusted eyes, and ii) fearful and surprised eyes. One expression in each of the two problematic pairs was therefore dropped (i.e., disgust and surprise).

2.5.2 Results and discussion

The performance (% correct responses) of the DP and TD groups in the three intensity conditions is depicted in Figure 2.3b. Results were analysed using ANOVA with Intensity (30%, 50%, 70%) as a within-subjects factor and Group (TD, DP) as a between-subjects factor. The analysis revealed a main effect of Intensity [$F(2,74) = 453.95$, $p < .001$, $\eta_p^2 = .92$]. Fewer correct responses were provided in the 30% condition ($M = .47$, $SD = .07$) than in the 50% ($M = .69$, $SD = .10$) [$t(38) = 18.39$, $p < .001$] and 70% conditions ($M = .80$, $SD = .08$) [$t(38) = 27.20$, $p < .001$]. The 50% condition was also harder than the 70% condition [$t(38) = 10.60$, $p < .001$]. The analysis also revealed a main effect of Group [$F(1,37) = 6.49$, $p = .01$, $\eta_p^2 = .15$], indicating that the DP group ($M = .62$, $SD = .07$) correctly identified fewer emotions than the TD group ($M = .68$, $SD = .06$), when performance was collapsed across emotion intensity. Interestingly, however, Intensity interacted significantly with Group [$F(2,74) = 4.43$, $p = .01$, $\eta_p^2 = .12$]. Simple contrasts indicated that the TD group ($M = .82$, $SD = .05$) outperformed the DP group ($M = .77$, $SD = .09$) in the 70% condition [$t(37) = 2.04$, $p = .04$]. A similar difference was seen between the DP ($M = .64$, $SD = .09$) and TD ($M = .73$, $SD = .08$) groups for the 50% condition [$t(37) = 3.24$, $p = .003$], but not for the 30% condition [$t(37) = 1.14$, $p = .26$]. Eight of the DPs scored at least one SD below the TD mean, and three (M1, M3, F10) were significantly impaired at single-case level (Figure 2.2b).

Significant correlations were found between participants' overall performance (collapsing across Group and Intensity) and their scores on the PI20 ($r = -.48$, $p = .003$), the CFMT ($r = .48$, $p = .002$) and the CFPT ($r = -.58$, $p < .001$) (see Table 2.4). Once again, the DP sample was split into apperceptive and non-apperceptive sub-groups based on their performance on the CFPT. Simple contrasts revealed a significant difference in emotion recognition ability of the apperceptive subgroup ($M = .60$, $SD = .09$) and TD controls ($M = .68$, $SD = .06$) [$t(28) = 2.74$, $p = .01$]. Interestingly, however, the performance of the non-apperceptive subgroup ($M = .64$, $SD = .06$) did not differ significantly from the TD sample [$t(29) = 1.48$, $p = .15$]. The inability of the apperceptive DPs to judge facial emotion using cues from the eye-region alone does not appear to be a product of diminished integration of information from the eye and mouth regions (Palermo et al., 2011), or to a strategic failure to use information from the eye region (Adolphs et al., 2005).

	CFMT		CFPT		PI20		CCMT		CBMT	
	r	*p*	*r*	*p*	*r*	*p*	*r*	*p*	*r*	*p*
70% (high intensity)	.39	.01	−.55	<.001	−.44	.007	−.01	.97	.02	.90
50% (moderate intensity)	.49	.002	−.52	.002	−.56	<.001	.01	.95	.10	.58
30% (low intensity)	.36	.03	−.46	.005	−.29	.12	−.05	.77	.17	.35
Overall performance	.48	.002	−.58	<.001	−.48	.003	−.02	.93	.11	.54

Table 2.4. Correlations between the expression recognition accuracies scores observed in Experiment 2 and participants' scores on the Cambridge Face Memory Test (CFMT), the Cambridge Face Perception Test (CFPT), the 20-item Prosopagnosia Index (PI20), the Cambridge Car Memory Test (CCMT) and the Cambridge Bike Memory Test (CBMT).

2.6 Experiment 3

The results of the first two experiments indicate that DP individuals are less able to categorise ambiguous facial expressions than TD controls. In Experiment 3 we sought to determine whether this affect recognition deficit was specific to faces, or whether these difficulties extend to other domains. Crucially, aberrant limbic functionality may leave observers unable to interpret emotion *per se* (Calder & Young, 2005). For example, individuals with developmental alexithymia – a neurodevelopmental condition associated with problems interpreting emotional experiences and other forms of interoceptive sensation (Bird & Cook, 2013; Brewer, Happe, Cook, & Bird, 2015) – exhibit a range of emotion recognition difficulties, including problems categorizing facial (Cook et al., 2013), vocal (Heaton et al., 2012), and musical affect (Allen, Davis, & Hill, 2012). To determine whether DPs exhibit face-specific emotion recognition difficulties, we examined their ability to recognise vocal affect. Typical performance on this task would suggest that the poor categorisation exhibited by the DP group in the first two experiments is a product of face, not emotion, perception deficits.

2.6.1 Methods

The performance of the DPs was compared with a group of 22 TD controls (8 males; $M_{\text{age}} = 42.86$, $SD_{\text{age}} = 12.89$). All TD participants were screened for DP (Table 2.2). All participants had normal or corrected-to-normal hearing. Ethical clearance was granted by the local ethics committee and the study was conducted in line with the ethical guidelines laid down in the 6th (2008) Declaration of Helsinki. All participants gave informed consent. All participants spoke English as first language.

The stimuli employed in Experiment 3 were short (< 3000 ms) audio sequences of British actors (2 males, 2 females) uttering 3-digit numbers ("two-hundred-and-fifty-five" and "five-hundred-and-twenty-eight") with different emotional inflections (happiness, disgust, fear, sadness, anger and surprise). Stimuli were recorded in a soundproof studio. Having cropped the audio files, and removed background noise using Audacity sound-editing software (http://audacity.sourceforge.net/), stimuli were validated in an online rating study. To create exemplars with varying degrees of ambiguity, we sought to manipulate the pitch of the stimuli, a vital component of vocal affect (e.g., Scherer, 1986). Different amounts (0%, 30%, 60%) of jitter – variability in pitch over the course of the sound – were added to the audio tracks using the 'Raspiness' function in *Praat* (Boersma & Weenink, 2015). In total, 144 stimuli were employed (2 exemplars × 6 emotions × 4 actors × 3 levels of degradation).

Experimental trials presented a single audio clip, followed by a prompt to make a 6-AFC response (happiness, anger, disgust, fear, sadness, or surprise). Each stimulus was presented once, in a randomised order, yielding a total of 144 trials. The task lasted approximately 15 minutes. Twelve practice trials (all with 0% jitter) preceded the experimental procedure to help familiarise participants with the actors' voices. No feedback was provided during the practice or experimental procedures. The experimental program was written in MATLAB using the Psychophysics Toolbox (Brainard, 1997; Pelli, 1997).

2.6.2 Results and discussion

The performance (% correct responses) of the DP and TD groups was analysed using ANOVA with Jitter (0%, 30%, 60%) as a within-subjects factor and Group (TD, DP) as a between-subjects factor. The analysis revealed a main effect of Jitter [$F(2,74) = 13.04$, $p < .001$, $\eta_p^2 = .26$]. As expected, greater pitch degradation was associated with poorer recognition: Fewer incorrect responses were provided in the 60% condition ($M = .61$, $SD = .09$) than in the 30% ($M = .65$, $SD = .10$) [$t(38) = 3.21$, $p = .003$] and 0% conditions ($M = .68$, $SD = .09$) [$t(38) = 4.95$, $p < .001$]. The 30% condition was also harder than the 0% condition [$t(38) = 2.29$, $p = .03$]. Crucially, however, we observed no main effect of Group [$F(1,37) = 1.90$, $p = .18$, $\eta_p^2 = .05$], nor a Group × Jitter interaction [$F(2,74) = .99$, $p = .38$, $\eta_p^2 = .03$], indicative of similar recognition accuracy in the TD and DP groups.

These results support the view that the emotion recognition difficulties exhibited by the DP group in the first two experiments are face-specific, and are not indicative of broader emotion processing impairments. The ability of the DP sample to interpret vocal signals accurately accords with anecdotal evidence that DPs often recognise familiar others using their voice (Cook & Biotti, 2016). We note, however, that recognition of vocal identity and vocal affect are thought to dissociate; for example, cases of developmental phonagnosia have been described who appear to exhibit broadly intact recognition of vocal affect, despite striking difficulties recognising vocal affect (Garrido, Eisner, et al., 2009; Garrido, Furl, et al., 2009).

2.7 Discussion

Despite severe difficulties recognising facial identity, emotion recognition deficits are thought to be relatively uncommon in DP (Bate & Bennetts, 2015; Humphreys et al., 2007; Palermo et al., 2011). Contrary to this view, however, we find evidence for widespread deficits in this population. In Experiment 1 we tested observers' ability to make binary classifications of whole-face expression stimuli drawn from morph continua. Psychophysical analyses revealed diminished ability to classify morphed facial expressions in our sample of DPs, relative to TD observers. We replicated this group difference in Experiment 2 when observers categorised facial emotion using only the eye-region. In our third experiment, we examined the ability of observers to classify the emotion present within segments of speech. Despite their difficulties judging facial emotion, the prosopagnosics exhibited excellent recognition of vocal affect, suggestive of a face-specific difficulty.

In our first two experiments, we observed striking correlations between expression classification accuracy and performance on the CFPT (Duchaine et al., 2007). The CFPT is thought to provide a demanding test of face encoding – observers' ability to represent and discriminate facial shape – in the absence of substantial demands on visual memory. Poor performance on this test is suggestive of an apperceptive form of prosopagnosia (Dalrymple, Garrido et al., 2014; Duchaine et al., 2007; Shah, Gaule, Gaigg, Bird, & Cook, 2015). Strikingly, when the DP sample was split into apperceptive and non-apperceptive subgroups based on CFPT performance, only the apperceptive subgroup exhibited impaired recognition of facial emotion. DPs with an apperceptive profile may have difficulties forming view-invariant structural descriptions of faces at an early stage in the face processing stream, before the divergence of identity and expression processing (Bruce & Young, 1986; Haxby et al., 2000). Inaccurate descriptions of local feature shape may result in imprecise expression categorisation as well as severe problems recognising facial identity.

To our knowledge, these findings are the first evidence of impaired recognition of facial emotion in DP, at the group level. Importantly, our results suggest that the ability to detect emotion recognition difficulties in this population may be extremely sensitive to the procedure used. In our first experiment, the clearest group difference was observed when observers were required to categorise expressions containing different degrees of surprise and fear. Typical observers also found these categorisations more demanding, and the increased difficulty may be responsible for the clear group difference observed. Alternatively, DPs with an apperceptive profile may have particular problems encoding the shape of the eye-region, variation crucial for distinguishing emotions, notably fear and surprise (Adolphs et al., 2005). In our second experiment, a clear group difference was observed only when judging the eye-region stimuli containing intermediate emotion intensities. All three levels of emotion intensity (30%, 50%, 70%) yielded recognition performance comfortably above chance (floor) and below 100% (ceiling) when typical observers were tested. However, stimuli either side of the 50% 'sweet-spot' may i) contain sufficiently obvious cues to be detected by observers with apperceptive deficits, or ii) be difficult for some typical observers to categorise reliably.

In light of these results, we recommend that authors demand a high standard of evidence before concluding that cases of DP exhibit intact emotion recognition. With respect to methodology, task sensitivity is a crucial issue. Modelling the categorisation of stimuli drawn from morph continua, by fitting psychometric functions, offers a precise means to estimate perceptual sensitivity independently of response bias[5]. Where morph continua are employed, the use of 7 levels of stimulus intensity affords greater sensitivity than the 5 stimulus levels present in the 'morph hexagon' used previously (Humprhreys et al., 2007; Palermo et al., 2011). The use of longer presentation durations and ambiguous expression stimuli may have also increased sensitivity in the

[5] In previous studies employing the morph hexagon, the authors have selected particular levels and analysed % correct. Fitting psychometric functions may yield more accurate measures of perceptual precision that allow for individual differences in response bias.

present study. With respect to sample size and composition, it is important that group studies have sufficient power to detect impairments. As awareness of DP increases, it should be easier to run group designs with reasonable sample sizes. Our results also suggest that studies with larger numbers of apperceptive DPs may be more likely to find expression recognition deficits. Where samples include relatively few DPs with an apperceptive profile, authors may also consider qualifying their conclusions accordingly.

Problems recognising facial identity – the defining feature of DP – can impact substantially on the social development and behaviour of sufferers. DPs often avoid social situations experiencing feelings of guilt and shame about actual or imagined offense caused to others (Davis et al., 2011). Long-term consequences can include reduced social circle, loss of self-confidence and limited work opportunities (Dalrymple, Fletcher et al., 2014; Fine, 2012; Yardley, McDermott, Pisarski, Duchaine, & Nakayama, 2008). In severe cases, DP can also contribute to the development of depression and anxiety (Yardley et al., 2008). Where observed, problems recognising the expressions of interactants will likely exacerbate these difficulties. Reduced ability to detect the emotional and mental states of others may prevent DPs responding appropriately and hinder social interaction, particularly in situations where vocal cues are unavailable. At present, relatively little is known about the impact of DP during childhood (Dalrymple, Corrow, Yonas, & Duchaine, 2013). The present results suggest the possibility that reasoning about the mental states of others ('theory of mind') may sometimes develop atypically in DP.

In summary, having tested a group of 17 DPs on complementary emotion recognition tasks, we find evidence of widespread difficulties recognising facial affect. These findings are contrary to the view that emotion recognition deficits are relatively uncommon in this population (Humphreys et al., 2007; Palermo et al., 2011). Deficits were apparent when observers were asked to categorise emotion using cues from the whole-face or from the eye-region only, and thus do not appear to reflect diminished integration of information from disparate facial regions (i.e., aberrant holistic processing). Instead, individuals with apperceptive forms of DP appear to have difficulties encoding facial shape, at an early stage in the face processing stream, before the divergence of identity and expression pathways. More broadly, these findings serve to illustrate how existing theoretical frameworks can be used to make sense of the heterogeneity seen in this population.

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Chapter 3: Impaired body perception in developmental prosopagnosia

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Chapter 3: Impaired body perception in developmental prosopagnosia

3.1 Abstract

Developmental prosopagnosia (DP)[6] is a lifelong neurodevelopmental disorder associated with difficulties recognising and discriminating faces. In some cases, the perceptual deficits seen in DP appear to be face-specific. However, DP is known to be a heterogeneous condition, and many cases undoubtedly exhibit impaired perception of other complex objects. There are several well-documented parallels between body and face perception; for example, faces and bodies are both thought to recruit holistic analysis and engage similar regions of visual cortex. In light of these similarities, individuals who exhibit face perception deficits, possibly due to impaired holistic processing or aberrant white matter connectivity, might also show co-occurring deficits of body perception. The present study therefore sought to investigate body perception in DP using a sensitive delayed match-to-sample task and a sizeable group of DPs. To determine whether body perception deficits, where observed, co-vary with wider object recognition deficits, observers' face and body matching ability was compared with performance in a car matching condition. Relative to age-matched controls, the DP sample exhibited impaired body matching accuracy at the group level, and several members of the sample were impaired at the single-case level. Consistent with previous reports of wider object recognition difficulties, a number of the DPs also showed evidence of impaired car recognition. Interestingly, however, we observed little or no relationship between observers' car perception ability and their body perception. We speculate that forms of developmental agnosia affecting the perception of faces, bodies, and objects may be best thought of as independent neurodevelopmental conditions.

3.2 Introduction

Developmental prosopagnosia (DP) is a lifelong neurodevelopmental disorder associated with difficulties recognising familiar faces and deficits of unfamiliar face discrimination. The condition occurs in people with normal intelligence, typical low-level vision, and with no apparent brain lesions (Behrmann & Avidan, 2005; Duchaine

[6] We use the term developmental prosopagnosia instead of congenital prosopagnosia to indicate the possibility that in some cases the disorder may appear during development and not necessarily from birth.

& Nakayama, 2006b; Susilo & Duchaine, 2013). As many as one in every 50 people are thought to experience lifelong face recognition difficulties severe enough to disrupt their daily lives (Kennerknecht et al., 2006; Kennerknecht, Ho, & Wong, 2008). Individuals with DP identify others using non-face cues, including hairstyle, voice, and gait. Consequently, DPs often experience great difficulty when familiar people are met in unusual contexts or when they alter their appearance (Cook & Biotti, 2016; Shah, Gaule, Sowden, Bird, & Cook, 2015). In addition to problems recognising facial identity, some DPs also exhibit problems perceiving facial emotion (Biotti & Cook, 2016; Duchaine, Yovel, Butterworth, & Nakayama, 2006).

DP frequently runs in families, indicating that the condition has a genetic component (Duchaine, Germine, & Nakayama, 2007; Johnen et al., 2014; Schmalzl, Palermo, & Coltheart, 2008). However, the origins of DP remain poorly understood. From a cognitive perspective, reduced holistic processing – whereby information from disparate facial regions is integrated into a unified perceptual description – may underlie the face recognition difficulties seen in DP (Avidan, Tanzer, & Behrmann, 2011; DeGutis, Cohan, Mercado, Wilmer, & Nakayama, 2012; DeGutis, Cohan, & Nakayama, 2014; Palermo et al., 2011). At the neurological level, studies have revealed reduced grey matter volume in occipitotemporal cortex of individuals with DP (Behrmann, Avidan, Gao, & Black, 2007; Garrido et al., 2009), and have suggested atypical functional connectivity in high-level visual areas (Avidan & Behrmann, 2009; Avidan et al., 2013; Lohse et al., 2016). In addition, recent studies have revealed striking white matter differences in the occipital and temporal lobes of DPs (Gomez et al., 2015; Song et al., 2015; Thomas et al., 2009). Reduced density and coherence of the inferior longitudinal fasciculus (ILF) may impair information exchange within the face processing network.

In some cases, the perceptual deficits seen in DP appear to be face-specific; many individuals achieve perfect or near-perfect performance on standardised object recognition batteries (e.g., Bentin, Deouell, & Soroker, 1999; Nunn, Postma, & Pearson, 2001). For example, Duchaine and colleagues (2006) described Edward, a 53-year old male, who exhibited a pure case of DP. Despite severe face recognition difficulties, Edward showed typical recognition of a range of objects including cars, tools, guns, horses, and sunglasses. Moreover, Edward was able to discriminate houses either on the basis on of elemental or configural differences, and showed typical learning and individuation of Greebles. However, DP is known to be a heterogeneous condition, and many cases undoubtedly exhibit impaired perception of other complex objects (Behrmann, Avidan, Marotta, & Kimchi, 2005; Dalrymple, Elison, & Duchaine, 2016; De Haan & Campbell, 1991; Duchaine et al., 2007). For example, of seven siblings with DP tested by Duchaine and colleagues (2007), five were significantly impaired at car perception, and 3 showed significant gun perception deficits. The extent to which cases of DP are face-specific or extend to other classes of object, may depend on the nature and extent of an individual's aberrant white matter connectivity (see Gomez et al., 2015).

There has been much interest in potential similarities between the visual processing of faces and bodies in typical observers (de Gelder et al., 2009; Minnebusch & Daum, 2009; Peelen & Downing, 2007; Slaughter, Stone, & Reed, 2004). Like faces, bodies are salient stimuli that capture attention when other classes of object go undetected (Downing, Bray, Rogers, & Childs, 2004; Stein, Sterzer, & Peelen, 2012). Faces and bodies both appear to preferentially engage regions of visual cortex. Strikingly, two areas thought to play a crucial role in body perception, the extrastriate (EBA; Downing, Jiang, Shuman, & Kanwisher, 2001) and fusiform (FBA; Peelen & Downing, 2005) body areas, are spatially adjacent to the occipital (OFA; Pitcher, Walsh, & Duchaine, 2011) and fusiform (FFA; Kanwisher & Yovel, 2006) face areas, respectively, suggestive of parallel networks (Peelen & Downing, 2007). Similar event-related brain potentials (ERPs) are elicited by both faces (N170; Bentin, Allison, Puce, Perez, & McCarthy, 1996; Eimer, 2011) and bodies (N190; Stekelenburg & de Gelder, 2004; Thierry et al., 2006). Both the N170 and N190 components are delayed and increased when stimuli are presented upside-down, however their origin appears to be distinct and dissociable (Sadeh et al., 2011).

There has also been great interest in whether or not bodies recruit holistic processing similar to that engaged by faces. Composite effects, whereby the presence of an aligned task-irrelevant region alters observers' perception of a target region, provide direct evidence of holistic face processing (Murphy, Gray, & Cook, 2016; Rossion, 2013). Interestingly, similar composite effects have recently been reported with expressive body postures (Willems, Vrancken, Germeys, & Verfaillie, 2014), but not for body shapes in neutral poses (Bauser, Suchan, & Daum, 2011). Sizeable inversion effects, often cited as an indirect measure of holistic processing, are seen for both faces and bodies (Cook & Duchaine, 2011; Robbins & Coltheart, 2012a). Inversion effects are particularly strong when participants are required to match sequentially presented body postures (Reed, Stone, Bozova, & Tanaka, 2003; Reed, Stone, Grubb, & McGoldrick, 2006). It is unclear, however, whether these effects reveal holistic body processing; for example, the magnitude of the posture inversion effect is disproportionately affected by the presence and position of the head (Yovel, Pelc, & Lubetzky, 2010).

Where individuals exhibit deficits of face perception, possibly due to impaired holistic processing or aberrant white matter connectivity, one might therefore expect co-occurring deficits of body perception. Consistent with this intuition, Righart and de Gelder (2007) found that the N170 marker of body processing exhibits atypical modulation following orientation inversion in three observers with DP. Nevertheless, many DPs report using body shape and bodily motion cues to recognise others (Biotti & Cook, 2016), and several empirical results suggest that body perception may be broadly typical in this population. For example, a recent study found no differences in torso matching accuracy when a sample of 11 DPs were compared with matched controls[7] (Rivolta, Lawson, & Palermo, 2016). Similarly, a sample of 16 DPs exhibited typical discrimination of hands – stimuli known to elicit strong responses in EBA (see Peelen & Downing, 2007) – in a match-to-sample procedure (Shah, Gaule, Gaigg, Bird, & Cook, 2015). Typical body matching has also been described in individual cases of DP (Duchaine et al., 2006). DPs and matched controls show broadly similar responses to body stimuli in core areas of the body processing network, including EBA and FBA (Van den Stock, van de Riet, Righart, & de Gelder, 2008), and multi-voxel pattern analysis (MVPA) suggests that distributed neural representations of body stimuli in inferotemporal cortex are largely typical (Rivolta et al., 2014).

The present study sought to investigate body perception in DP through the use of a sensitive identity matching task of headless torsos, using a sizeable group of DPs (N = 20). Individual differences in body matching ability were compared with performance in comparable car and face matching conditions. In light of the equivocal literature on body perception in DP, we anticipated a range of abilities in our sample. However, we were interested in the possibility that co-occurring deficits of body perception, where observed, may co-vary with wider object recognition deficits described previously (Behrmann et al., 2005; Dalrymple et al., 2016; De Haan & Campbell, 1991; Duchaine et al., 2007). In neurotypical individuals, the perception of faces, bodies, and objects is

[7] While prosopagnosics and controls did not differ in body matching accuracy, the prosopagnosics responded slower.

thought to rely on functionally and spatially distinct networks (Pitcher, Charles, Devlin, Walsh, & Duchaine, 2009). However, the diffuse white matter differences seen in some cases of DP (Gomez et al., 2015; Song et al., 2015; Thomas et al., 2009) may predispose individuals to a range of perceptual deficits.

3.3 Method

3.3.1 Participants

Participants were 43 right-handed adults, 20 with (6 males; M_{age} = 38.04 years, SD_{age} = 13.05 years) and 23 without DP (9 males; M_{age} = 40.30 years, SD_{age} = 14.38 years). The groups did not differ significantly in age [$t(41) = .54$, $p = .593$] or proportion of males [$X^2(1) = .09$, $p = .760$]. Ethical approval was granted by the local ethics committee. The study was conducted in line with the ethical guidelines provided by the 6th (2008) Declaration of Helsinki. All participants provided informed consent.

3.3.2 Diagnostic testing

DP participants were recruited through www.troublewithfaces.org. All members of the DP sample described lifelong face recognition difficulties. None of the DPs had a history of brain injury or psychiatric disorder (e.g., schizophrenia, autism spectrum disorder). Diagnostic evidence for the presence of DP was collected using the Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006a), the Cambridge Face Perception Test (CFPT; Duchaine et al., 2007) and the Twenty-Item Prosopagnosia Index (PI20; Shah, Gaule, Sowden et al., 2015). The prosopagnosics' scores on the diagnostic procedures were compared with a group of 56 age-matched controls (M_{age} = 40.25 years, SD_{age} = 13.71 years, 24 males). All DPs scored at least two standard deviations below the mean of the comparison sample on the PI20 (Table 3.1). All but one of the DPs scored at least two standard deviations below the comparison average on the CFMT; the remaining DP scored -1.86 standard deviations below the comparison average. Thirteen of the DPs also scored two standard deviations below the comparison average on the $\text{CFPT}_{upright}$. In addition to the face recognition tests, participants completed the Cambridge Car Memory Test (CCMT; Dennett et al., 2011), to assess their wider object recognition ability, and were screened for colour blindness using Ishihara's Tests for Colour-Blindness (Ishihara, 1993).

Participant	Age	PI20	CFPT upright	CFPT inverted	CFMT %	CCMT%
F1	21	59*	30	64	63.89*	51.39
F2	22	80*	66*	98	40.28*	56.94
F3	48	85*	60*	70	63.89*	59.72
F4	66	79*	40	70	61.11*	66.67
F5	49	82*	62*	68	62.50*	68.06
F6	48	78*	26	64	58.33*	61.11
F7	23	82*	38	48	65.28*	80.56
F8	56	91*	30	60	54.17*	55.56
F9	23	84*	52*	70	66.67*	100.0
F10	47	75*	46	78	50.00*	72.22
F11	46	92*	72*	78	50.00*	47.22
F12	29	83*	66*	84	51.39*	62.50
F13	54	85*	74*	94	45.83*	63.89
F14	25	70*	74*	68	44.44*	54.17
M1	30	85*	48*	84	59.72*	65.28
M2	58	82*	52*	56	68.06	59.72
M3	56	82*	54*	78	66.67*	79.17
M4	36	77*	44	42	40.28*	68.06
M5	26	82*	54*	48	65.28*	66.67
M6	43	83*	48*	54	40.28*	59.72
TD mean	40.25	37.75	29.57	60.00	84.82	64.98
TD SD	13.71	9.16	9.24	14.47	9.02	14.42

Table 3.1. Scores of each developmental prosopagnosic on the Twenty-Item Prosopagnosia Index (PI20), the Cambridge Face Perception Test (CFPT), the Cambridge Face Memory Test (CFMT), and the Cambridge Car Memory Test (CCMT). Higher scores on the CFPT and PI20 indicate poorer face recognition. The mean and standard deviation of the comparison sample (N = 56) are provided below. Asterisks indicate performance at least 2 standard deviations below the comparison sample.

3.3.3 Stimuli

Each category (faces, bodies, cars) comprised fifty exemplars (Figure 3.1). Individual categories were further organised into five subsets of ten exemplars based on approximate similarity. Each exemplar was depicted twice: once in frontal view, once in 3/4 view. When viewed at 57 cm, the face and body stimuli subtended 11° of visual angle vertically; the cars subtended 8° vertically. Face stimuli (male Caucasian faces) were created using FaceGen Modeller Version 3.3 (Singular Inversions Inc.). Body stimuli (Caucasian male torsos) were created with Poser 7.0 (e frontier America, Inc.). Car stimuli (black saloon / sedan cars and SUVs) were generated through www.3dtuning.com. The use of torsos prevented observers employing simple limb-matching strategies and allowed us to present body stimuli a scale that accentuated 3D shape variation. We note that torsos, unlike other body parts, elicit strong responses both the EBA and FBA (Taylor, Wiggett, & Downing, 2007).

3.3.4 Procedure

Testing took place at City, University of London. Trials started with a fixation point (750 ms), before a single target stimulus was presented centrally (400 ms). Targets were always shown in frontal view. A retention interval (3000 ms) followed target offset. A mask image – constructed by recombining regions cropped from other target images from the same category – was presented throughout the retention interval. An array of four test items followed the retention interval. The array comprised the target and three lures from the same within-category subset, all shown in 3/4 view[8]. Test arrays were visible until a keypress response was registered. Participants were asked to locate the target item with speed and accuracy. All stimuli appeared as a target once, yielding 150 experimental trials, which were preceded by six practice trials. Trial type (Face, Body, Car) was interleaved within each mini-block. No feedback was provided during the procedure. The task lasted approximately 30 minutes and included three short breaks. The matching task was programmed in MATLAB using the Psychophysics Toolbox (Brainard, 1997; Pelli, 1997).

[8] The requirement to identify exemplars across different viewpoints prevents the use of simple image matching strategies; instead, observers must form a view-invariant 3-dimensional representation of the target (e.g., Longmore, Liu, & Young, 2008).

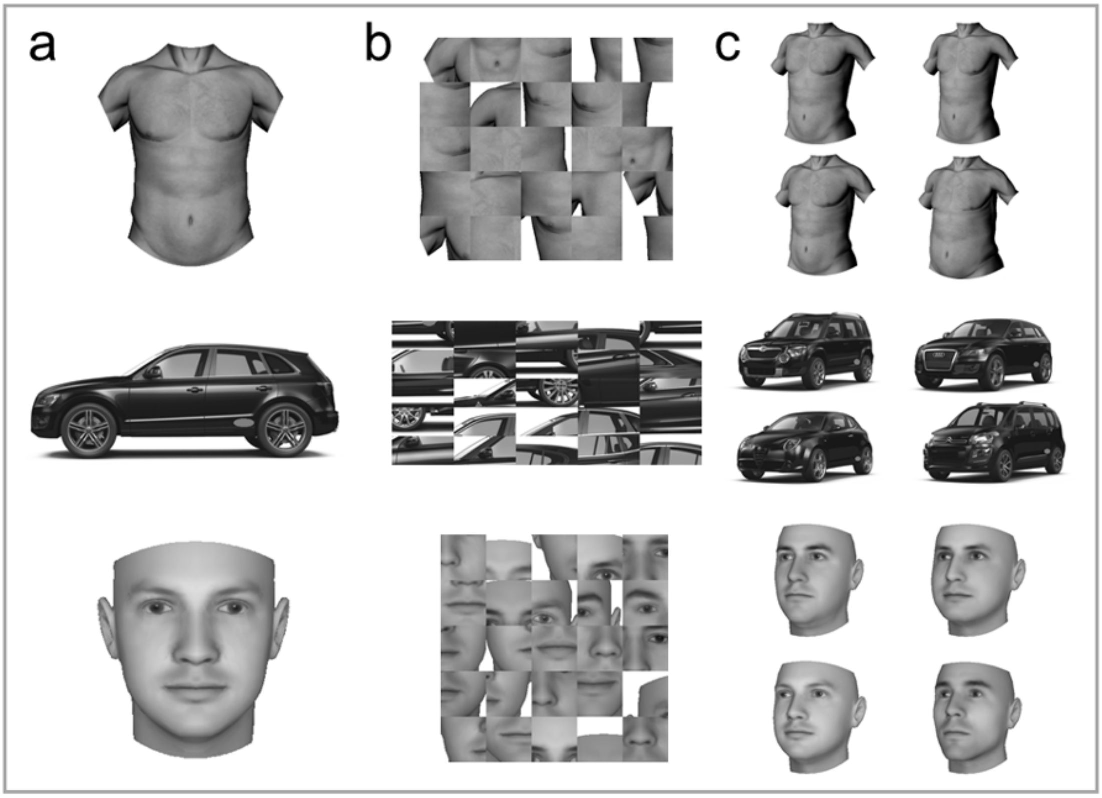

Figure 3.1. (a) Target stimuli were presented in frontal view for 400 msec. (b) During the retention interval (3000 msec) a mask was presented, constructed by recombining regions cropped from other target images from the same category. (c) Test arrays presented the target and three lures in 3/4 views.

3.4 Results

3.4.1 Group analyses

Matching accuracy (% correct; Figure 3.2a) was analysed using ANOVA with Category (bodies, cars, faces) as a within-subjects factor and Group (DP, TD) as a between-subjects factor. The analysis revealed significant main effects of Category [$F(2,82) = 5.29$, $p = .007$, $\eta^2 = .11$], and Group [$F(2,41) = 24.03$, $p < .001$, $\eta^2 = .37$], as well as a significant Group × Category interaction [$F(2,82) = 5.46$, $p = .006$, $\eta^2 = .19$]. As expected, simple contrasts indicated that the DP group ($M = .45$, $SD = .13$) was less accurate at face matching than the TD group ($M = .66$, $SD = .15$) [$t(41) = 4.97$, $p < .001$]. Crucially, however, the TD group also outperformed the DP group when matching bodies (TD: $M = .54$, $SD = .09$; DP: $M = .46$, $SD = .09$) [$t(41) = 2.69$, $p = .01$]

and cars (TD: M = .61, SD = .14; DP: M = .53, SD = .10) [$t(41) = 2.16$, $p = .036$]. The relative performance of individual DPs in the three conditions is shown in Figure 3.3.

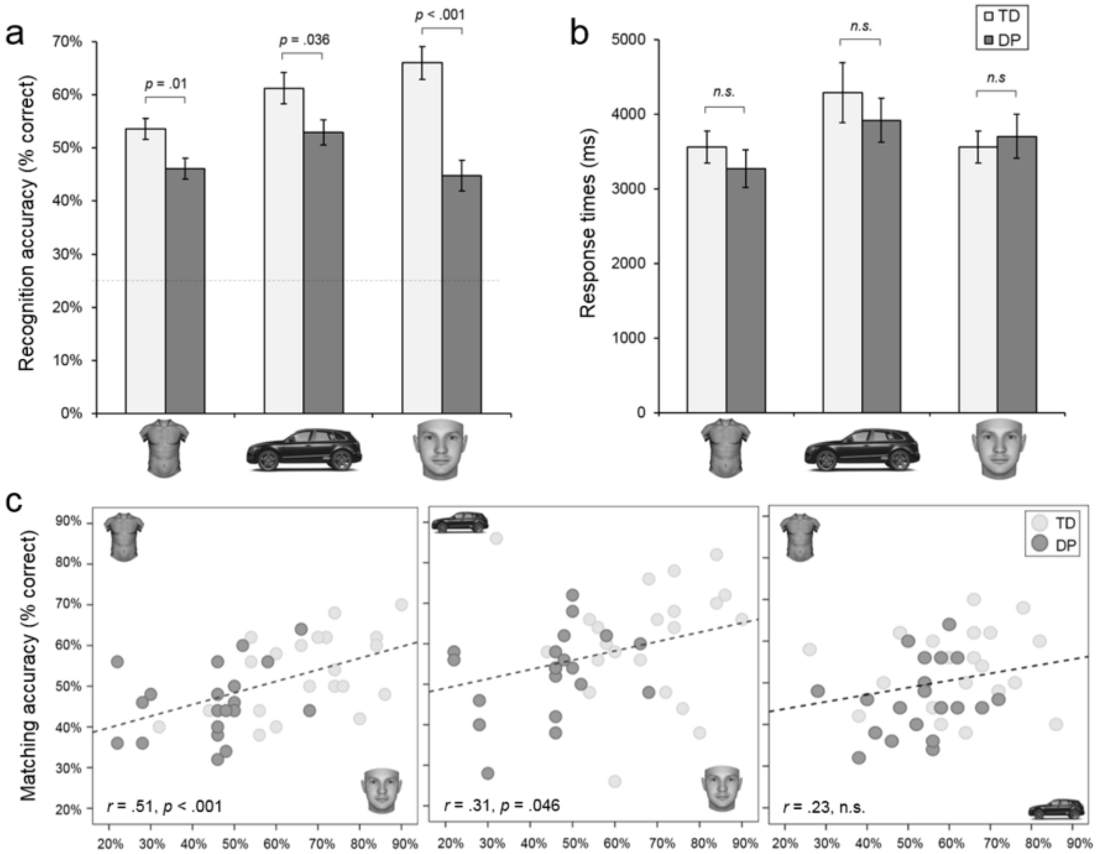

Figure 3.2. (a) Mean matching accuracy and (b) mean response times for the typically developing (TD) controls and the developmental prosopagnosics (DPs) in the three conditions. Error bars denote ± one SEM. (c) Scatter plots comparing participants' matching accuracy in the three conditions.

The analysis of response times (RTs; Figure 3.2b) revealed a main effect of Category [$F(2,82) = 9.66$, $p < .001$, $\eta^2 = .19$]. Generally participants were slower when matching cars (M = 4118, SD = 1662), than faces (M = 3629, SD = 1169) [$t(42) = 2.35$, $p = .024$] or bodies (M = 3428, SD = 1069) [$t(42) = 4.34$, $p < .001$]. RTs were also significantly slower on face trials, than on body trials [$t(42) = 2.16$, $p = .036$]. The analysis revealed no main effect of Group [$F(2,41) = .22$, $p = .638$, $\eta^2 = .005$], nor a Group × Category interaction [$F(2,82) = 1.5$, $p = .229$, $\eta^2 = .035$].

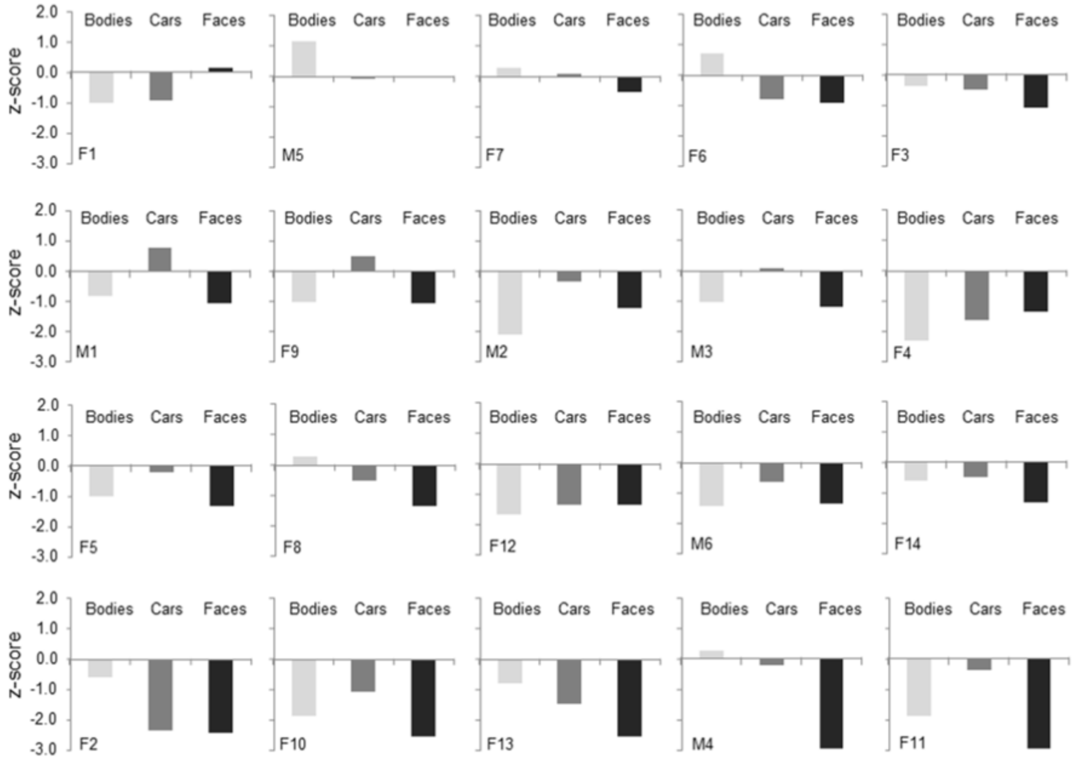

Figure 3.3. Single-case data for the members of the DP group. For ease of comparison, cases are ordered by performance in the face matching condition. A z-score of zero denotes performance comparable with the mean of the control group.

3.4.2 Correlational analyses

In addition to the group-level analyses, we also examined the individual differences seen on our matching task using correlational analyses (Table 3.2). For the purposes of these analyses, we collapsed across the control (N = 23) and DP (N = 20) groups to yield a combined sample of 43. It is clear, however, that correlations described with larger sample sizes are associated with greater power and increased stability (e.g., Schönbrodt & Perugini, 2013).

We began by confirming that performance in our face and car conditions correlated with our diagnostic measures of face and car perception. Strong correlations were observed between participants' matching accuracy in the face condition and their scores on the CFMT (r = .77, p < .001), PI20 (r = -.66, p < .001), and CFPT (r = -.71, p < .001). Performance in the cars condition also correlated with scores on the CCMT (r = .57, p < .001).

Next, we sought to compare matching accuracy for bodies and cars with measures of face perception. Body matching accuracy correlated with scores on the CFMT (r = .46, p = .002), PI20 (r = -.31, p = .04), and CFPT (r = -.41, p = .006). Moderate correlations were found between car matching accuracy and both CFMT (r = .39, p = .009) and CFPT scores (r = -.37, p = .015). Matching accuracy for bodies (r = .51, p < .001) and cars (r = .31, p = .046) correlated with performance in the face condition (Figure 3.2c).

Finally, we sought to compare our measures of body and car perception. Interestingly, we failed to find significant correlations between body and car matching accuracy, either in the combined sample (r = .23, p = .14), in the TD sample (r = .04, p = .86), or in the DP sample (r = .25, p = .28). We also failed to find any relationship between CCMT scores and performance in our body condition in the combined sample (r = .16, p = .30), in the TD sample (r = .07, p = .75), or in the DP sample (r = .06, p = .79).

To compare the strength of correlations observed in the combined sample we used Steiger's (1980) modification of Dunn and Clark's z (1969), implemented using the 'cocor' package (Diedenhofen & Musch, 2015) in R. There was some indication that the strength of the face-body correlation exceeded that of the body-car correlation [z = 1.7, p = .045 (one-tailed)]. However, the strength of the face-body [z = 1.2, p = .12 (one-tailed)] and body-car correlations [z = .52, p = .3 (one-tailed)] did not differ significantly from the face-car correlation.

	CFMT		CFPT		PI20		CCMT	
	r	p	r	p	r	p	r	p
Bodies (% correct)	.46	.002	−.41	.006	−.31	.041	.16	.29
Cars (% correct)	.39	.009	−.37	.015	−.29	.057	.57	<.001
Faces (% correct)	.77	<.001	−.71	<.001	−.67	<.001	.16	.30

Table 3.2. Correlations observed between the diagnostic tests and matching accuracy for bodies, cars, and faces. None of the correlations with response times reached significance.

3.5 Discussion

The present study sought to determine whether body perception is impaired in DP. Relative to age-matched TD controls, the DP sample exhibited impaired body matching accuracy at the group level, and several members of the sample were impaired at the single case level. These results provide the clearest behavioural evidence of impaired body perception in DP reported to date. Previous findings suggest that, in some cases of DP, ERP markers of body processing fail to show typical modulation by stimulus orientation (Righart & de Gelder, 2007). At the behavioural level, however, typical body matching accuracy has been described (Duchaine et al., 2006; Rivolta et al., 2016; Shah, Gaule, Gaigg et al., 2015). Our use of a larger DP sample and a sensitive task likely helped reveal body perception deficits. Consistent with previous reports (Behrmann et al., 2005; Dalrymple et al., 2016; De Haan & Campbell, 1991; Duchaine et al., 2007), a number of the DPs in the present study also showed evidence of wider object recognition difficulties. At the group level, the DPs were less accurate in the car matching condition than TD observers, and several DPs showed signs of impairment on the CCMT.

Evidence of body perception deficits in DP accords with well-documented parallels between body and face perception (de Gelder et al., 2009; Minnebusch & Daum, 2009; Peelen & Downing, 2007; Slaughter et al., 2004). For example, the EBA-FBA and OFA-FFA networks for body and face processing, respectively, recruit spatially adjacent regions of occiptotemporal cortex (Peelen & Downing, 2007). Indeed, the FFA and FBA partially overlap in some observers (Peelen & Downing, 2005). Given the diffuse white matter differences described in occipitotemporal regions of some DPs (Gomez et al., 2015; Thomas et al., 2009), entirely typical body perception would be surprising. Similarly, the accurate perception of face and body shape may depend on holistic processing (Murphy et al., 2016; Robbins & Coltheart, 2012a, 2012b). Should cases of DP result from atypical holistic processing (Avidan et al., 2011; DeGutis et al., 2012; DeGutis et al., 2014; Palermo et al., 2011), one might therefore expect problems perceiving both faces and bodies.

While the incidence of body agnosia may be higher in DP than in the typical population, body perception deficits do not appear to be a universal feature of DP. At the group level, the deficits do not appear to be as strong as for faces, and only 4 of the DPs exhibited significant body perception deficits at the single-case level. Again, this is not surprising given previous evidence that the perceptual processing of bodies and faces appears to dissociate. For example, neuropsychological patients have been described who exhibit severely impaired body perception, but spared face perception (Moscovitch, Winocur, & Behrmann, 1997). Conversely, other patients exhibit severe face recognition, but typical body perception (Susilo, Yovel, Barton, & Duchaine, 2013). The application of transcranial magnetic stimulation to EBA and OFA also appears to selectively impair the perception of bodies and faces, respectively (Pitcher et al., 2009).

In light of co-occurring deficits of body and car perception, it is tempting to conclude that DPs have a domain general perceptual deficit. Interestingly, however, we observed little or no relationship between observers' car perception ability and their body perception. We speculate that forms of developmental agnosia affecting the perception of faces, bodies, and objects may be best thought of as independent neurodevelopmental conditions. A key feature of neurodevelopmental conditions is that they co-occur; for example, the incidence of several conditions is elevated in ASD relative to the typical population (Baron-Cohen et al., 2013; Bird & Cook, 2013; Dziuk et al., 2007; Jones et al., 2009; Matson & Shoemaker, 2009; van Steijn et al., 2014). Though often overlooked by vision scientists, it is widely recognised in psychiatry that genetic or environmental factors that predispose an individual to one developmental condition, often increase their risk of developing others (Bishop & Rutter, 2008; Gilger & Kaplan, 2001; Kaplan, Dewey, Crawford, & Wilson, 2001; Rutter, 1997; Rutter et al., 2011). Observers predisposed to developing DP may therefore be at risk from developing body agnosia, and wider object recognition difficulties.

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Chapter 4: Normal composite face effects in developmental prosopagnosia

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Chapter 4: Normal composite face effects in developmental prosopagnosia

4.1 Abstract

Upright face perception is thought to involve holistic processing, whereby local features are integrated into a unified whole. Consistent with this view, the top half of one face appears to fuse perceptually with the bottom half of another, when aligned spatially and presented upright. This ‘composite face effect’ reveals a tendency to integrate information from disparate regions when faces are presented canonically. In recent years, the relationship between susceptibility to the composite effect and face recognition ability has received extensive attention both in participants with normal face recognition and participants with developmental prosopagnosia. Previous results suggest that individuals with developmental prosopagnosia may show reduced susceptibility to the effect suggestive of diminished holistic face processing. Here we describe two studies that examine whether developmental prosopagnosia is associated with reduced composite face effects. Despite using independent samples of developmental prosopagnosics and different composite procedures, we find no evidence for reduced composite face effects. The experiments yielded similar results; highly significant composite effects in both prosopagnosic groups that were similar in magnitude to the effects found in participants with normal face processing. The composite face effects exhibited by both samples and the controls were greatly diminished when stimulus arrangements were inverted. Our finding that the whole-face binding process indexed by the composite effect is intact in developmental prosopagnosia indicates that other factors are responsible for developmental prosopagnosia. These results are also inconsistent with suggestions that susceptibility to the composite face effect and face recognition ability are tightly linked. While the holistic process revealed by the composite face effect may be necessary for typical face perception, it is not sufficient; individual differences in face recognition ability likely reflect variability in multiple sequential processes.

4.2 Introduction

In recent years, research has revealed substantial individual differences in face processing ability. Whilst 'super-recognisers' make up the upper tail (Russell, Duchaine, & Nakayama, 2009), the lower-end of the distribution is composed of individuals with developmental prosopagnosia[9] (DP). DP is a neurodevelopmental condition characterised by difficulties recognising facial identity, despite normal intelligence, typical low level vision, and no history of brain damage (Behrmann & Avidan, 2005; Cook & Biotti, 2016; Duchaine & Nakayama, 2006b). DP was once thought to be extremely rare (McConachie, 1976), but one in every 50 people are now thought to experience lifelong face recognition difficulties severe enough to disrupt their daily lives (Kennerknecht, Ho, & Wong, 2008; Kennerknecht et al., 2006). Individuals with DP typically utilise non-face cues including voice, gait, and hairstyle to recognise others. Consequently, they often experience great difficulties when non-face cues are unavailable or changed, or when familiar people are encountered out of context.

Numerous papers have suggested that diminished holistic face processing may underlie the difficulties seen in DP (Avidan, Tanzer, & Behrmann, 2011; Carbon, Grüter, Weber, & Lueschow, 2007; DeGutis, Cohan, Mercado, Wilmer, & Nakayama, 2012; DeGutis, Cohan, & Nakayama, 2014; Liu & Behrmann, 2014; Lobmaier, Bölte, Mast, & Dobel, 2010; Palermo et al., 2011). Typical face perception appears to involve a rapid parallel analysis, whereby local features are integrated into a unified whole (Farah, Wilson, Drain, & Tanaka, 1998; Maurer, Le Grand, & Mondloch, 2002; McKone & Yovel, 2009; Piepers & Robbins, 2013). Evidence of holistic face perception is provided by the composite face effect, where the top half of one face appears to fuse perceptually with the bottom half of another, when the two halves are aligned and presented upright (Hole, 1994; Young, Hellawell, & Hay, 1987). The resulting illusion-induced interference disrupts observers' ability to judge the identity (Young et al., 1987), physical resemblance (Hole, 1994), age (Hole & George, 2011), gender (Baudouin & Humphreys, 2006), and attractiveness (Abbas & Duchaine, 2008) of constituent face halves (for reviews see Murphy, Gray, & Cook, 2017; Rossion, 2013). When face halves are inverted, observers show little or no interference (McKone

[9] We use the term developmental prosopagnosia instead of congenital prosopagnosia to indicate the possibility that in some cases the disorder may not be present at birth.

et al., 2013; Susilo, Rezlescu, & Duchaine, 2013). Importantly, the composite effect reveals a tendency to integrate feature information from disparate regions when faces are presented canonically, consistent with holistic theories of face perception (Farah et al., 1998; Maurer et al., 2002; McKone & Yovel, 2009; Piepers & Robbins, 2013).

The suggestion that DP results from disrupted holistic processing is closely related to the view that the whole-face binding process measured by the composite face effect contributes to face recognition ability (DeGutis, Wilmer, Mercado, & Cohan, 2013; Farah et al., 1998; Maurer et al., 2002; Piepers & Robbins, 2013). However, studies comparing observers' susceptibility to the composite face effect and their face recognition ability have yielded mixed results (Murphy et al., 2017). In cases of acquired prosopagnosia (AP), individuals are left with face recognition difficulties following brain injury. While some APs exhibit reduced composite face effects relative to matched controls (Busigny, Joubert, Felician, Ceccaldi, & Rossion, 2010; Ramon, Busigny, & Rossion, 2009), others exhibit typical susceptibility to the original matching procedure (Finzi, Susilo, Barton, & Duchaine, 2016; Rezlescu, Pitcher, & Duchaine, 2012). Where composite face effects and face recognition ability have been compared in samples drawn from the general population, some authors have observed positive associations (DeGutis et al., 2013; Engfors, Jeffery, Gignac, & Palermo, 2017; Richler, Cheung, & Gauthier, 2011), whilst others have found little or no correlation (Konar, Bennett, & Sekuler, 2010; Rezlescu, Susilo, Wilmer, & Caramazza, 2017; Wang, Li, Fang, Tian, & Liu, 2012).

The literature is also inconsistent with respect to the relationship between individuals' susceptibility to the composite face effect and other putative markers of holistic representation, including the part-whole (Tanaka & Farah, 1993) and face-inversion effects (Yin, 1969). For example, some authors have found associations between susceptibility to the composite face effect and the part-whole effect (DeGutis et al., 2013). However, other studies have found no association between susceptibility to the composite face effect and the part-whole effect (Rezlescu et al., 2017; Wang et al., 2012), or between composite face effects and perceptual decrements induced by face inversion (Rezlescu et al., 2017). These findings cast doubt on the view that a unitary process underlies holistic face processing. Where different measures of holistic processing are unrelated or weakly correlated in the typical population, neuropsychological dissociations might also be seen in the DP population.

Although studies have described a number of individuals with DP who exhibit composite effects comparable with those of matched controls (Le Grand et al., 2006; Schmalzl, Palermo, & Coltheart, 2008; Susilo et al., 2010), three studies have concluded that DP is associated with reduced susceptibility to the composite face effect at the group level (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011). Nevertheless, the case for diminished composite effects in DP remains unconvincing. In at least one study, inspection of single-case data suggests that previously reported group results have been strongly influenced by the presence of outliers in DP samples (Palermo et al., 2011). In other studies, DP samples perform poorly in the baseline 'misaligned' condition making it hard to interpret putative differences in composite effect susceptibility (Liu & Behrmann, 2014).

Given the uncertainty about the functional significance of the holistic processes revealed by the composite face effect (Finzi et al., 2016; Konar et al., 2010; Rezlescu et al., 2017; Wang et al., 2012) and the popular view that DP may be caused by diminished holistic representation (Carbon et al., 2007; DeGutis et al., 2012; DeGutis et al., 2014; Lobmaier et al., 2010), obtaining a better understanding of composite face effects in DP is theoretically important. It may also have implications for interventions aimed at improving face recognition in DP (e.g., DeGutis et al., 2014). The present study therefore sought to confirm that DP is associated with reduced composite face effects at the group level. We describe two experiments employing independent samples of DP participants collected in the UK and the USA ($N = 16$ and $N = 24$) and complementary paradigms (simultaneous and sequential matching). Contrary to previous group studies (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011), we find no evidence for diminished composite face effects in DP.

4.3 Experiment 1

In our first experiment we compared the composite face effects of DPs and matched controls using a simultaneous matching procedure (Hole, 1994). Composite effects seen with upright faces were compared with those seen with inverted faces. Whereas strong effects of alignment are seen when composite faces are presented upright, interference is greatly reduced when composites are constructed from inverted faces (Susilo et al., 2013). This comparison is useful as it addresses the possibility that effects of misalignment found with upright faces are due to general factors rather than face-specific processes (McKone et al., 2013; Rossion, 2013). We also examined composite effects for pseudo-words which resemble the effects found for upright faces (Anstis, 2005). For the sake of brevity, however, details of the procedure and results for pseudo-words are provided as supplementary material.

4.3.1 Methods

4.3.1.1 Participants

Two groups of observers completed the procedure; 16 individuals with DP (M_{age} = 43.56 years, SD_{age} = 15.09 years, 3 males), and a control group comprising 16 neurotypical adults (M_{age} = 39.81 years, SD_{age} = 12.95 years, 10 males). All observers were resident in the UK. Ethical approval was granted by the local ethics committee and the study was conducted in line with the Declaration of Helsinki. All participants provided informed consent prior to testing.

4.3.1.2 Diagnostic testing

DP participants were recruited through www.troublewithfaces.org. All members of the DP sample described lifelong face recognition difficulties that affected their daily lives. None of the DPs had a history of brain injury or psychiatric disorder (e.g., Schizophrenia, Autism Spectrum Disorder). Diagnostic evidence for the presence of DP was collected using the Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006a) the Twenty-Item Prosopagnosia Index (PI20; Gray, Bird, & Cook, 2017; Shah, Gaule, Sowden, Bird, & Cook, 2015), and a Famous Face Test suitable for use with UK residents (FFT_{UK}). Scores on the CFMT were compared against data from 50 typical observers reported by Duchaine & Nakayama (2006a). Participants also completed the Cambridge Face Perception Test (CFPT; Duchaine, Germine, & Nakayama, 2007) to determine whether face recognition deficits had an apperceptive origin (De Renzi, Faglioni, Grossi, & Nichelli, 1991). While participants were not selected on the basis of these scores, the DP sample was impaired at the group level [$t(22) = 2.34$, $p = .029$]. Scores on the CFPT and PI20 were compared with a group of 56 controls (M_{age} = 40.25 years, SD_{age} = 13.71 years, 24 males). Comparison data for the FFT_{UK} was collected from a sample of 20 controls (M_{age} = 30.4 years, SD_{age} = 10.27 years, 9 males). When tested on the CFMT, all DPs scored at least 1.53 standard deviations below the mean performance of the comparison sample. All DPs tested[10] also scored at least 2 standard deviations below the mean of the comparison samples on the FFT_{UK} and the PI20. Diagnostic information is presented in Table 4.1.

Participant	Age	PI20	FFT_{UK} %	CFMT %	CFPT upright [errors]
F1	21	59 (–2.3)	25 (–4.2)	62.50 (–1.6)	30 (–.0)
F2	22	89 (–5.6)	–	50.00 (–2.8)	30 (–.0)
F3	25	87 (–5.4)	41 (–2.9)	63.89 (–1.5)	44 (–1.6)
F4	28	68 (–3.3)	48 (–2.3)	61.11 (–1.8)	32 (–.3)
F5	35	85 (–5.2)	34 (–3.4)	43.06 (–3.4)	46 (–1.8)
F6	42	92 (–5.9)	18 (–4.8)	45.83 (–3.1)	62 (–3.5)
F7	50	78 (–4.4)	30 (–3.8)	58.33 (–2.0)	34 (–.5)
F8	53	85 (–5.2)	42 (–2.8)	45.83 (–3.1)	74 (–4.8)
F9	55	85 (–5.2)	–	58.33 (–2.0)	36 (–.7)
F10	65	79 (–4.5)	14 (–5.1)	61.11 (–1.8)	40 (–1.1)
F11	65	81 (–4.7)	25 (–4.2)	59.72 (–1.9)	44 (–1.6)
F12	48	78 (–4.4)	45 (–2.5)	58.33 (–2.0)	26 (+.4)
F13	48	85 (–5.2)	37 (–3.2)	63.89 (–1.5)	60 (–3.3)
M1	28	62 (–2.6)	44 (–2.6)	62.50 (–1.6)	46 (–1.8)
M2	54	88 (–5.5)	48 (–2.3)	58.33 (–2.0)	66 (–3.9)
M3	58	92 (–5.9)	5 (–5.9)	44.44 (–3.3)	68 (–4.2)
DP mean		80.81	32.57	56.26	46.12
DP SD		9.97	13.53	7.66	15.31
Comparison mean		37.75	75.35	80.4	36.7
Comparison SD		9.16	12.00	11.0	12.2

Table 4.1. Scores for each developmental prosopagnosic in Experiment 1 on the 20-Item Prosopagnosia Index (PI20), the Cambridge Face Memory Test (CFMT), the Cambridge Face Perception Test (CFPT), and the Famous Faces Test (FFT_{UK}). *Z*-scores are shown in parentheses. Negative *z*-scores denote performance worse than the typical mean. The mean and standard deviation of the comparison samples are provided below.

4.3.1.3 The composite task

Face composites were constructed from images of emotionally neutral faces taken from the Karolinska Directed Emotional Faces database (Lundqvist, Flykt, & Öhman, 1998). Faces were cropped to exclude external facial features (e.g. ears, hairline). Face halves containing the eyes were used as target regions. Face composites subtended 8° of visual angle, vertically. The to-be-judged regions subtended 4°. In the misaligned conditions, the horizontal offset corresponded to approximately 25% the width of a face.

In total, 40 face composites were employed. Each composite was allocated a partner arrangement of the same type with which it would be presented simultaneously. For half the composite pairs, the target regions were identical, for half the pairs the target regions differed. Following the standard composite design (also referred to as the

[10] In Experiment 1, two DPs did not complete FFTUK. In Experiment 2, two DPs did not complete the FFTUS and two did not complete the ONFRT.

original design; Murphy et al., 2017; Rossion, 2013), the distractor regions within each pair were always different. The two target regions appeared at the same vertical position in the display (the lower edge of each target region was aligned to the vertical midpoint of the display). Two dashed guidelines were imposed over the arrangements to clearly delineate the stimulus regions to be judged. Example displays are presented in Figure 4.1a.

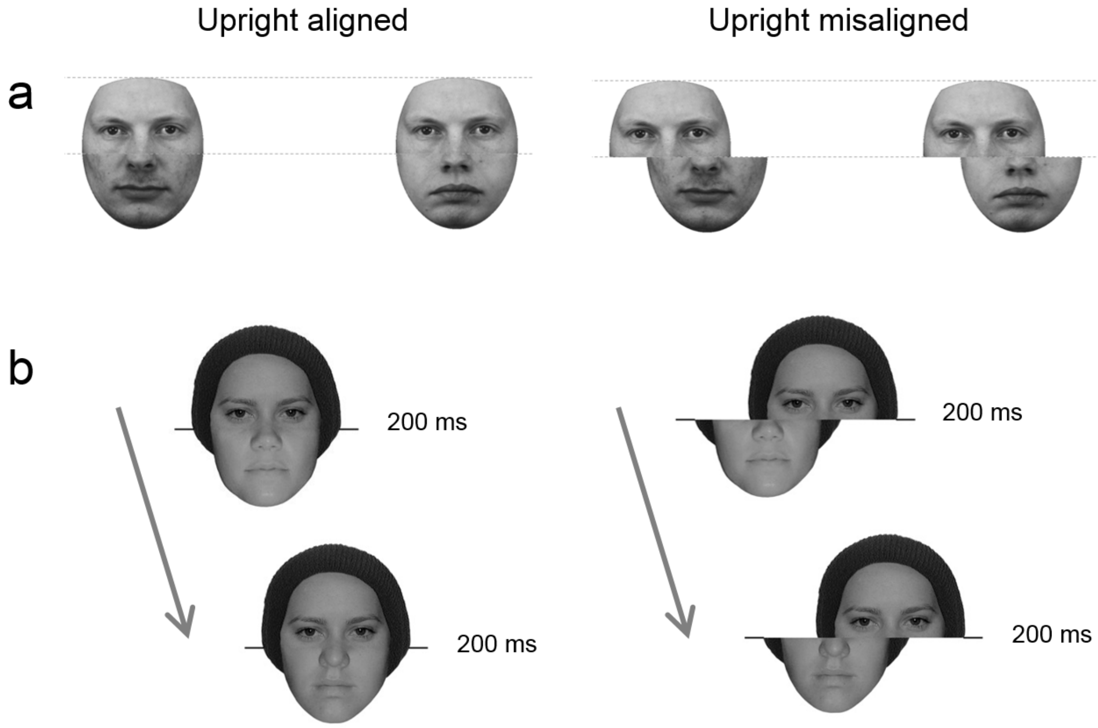

Figure 4.1. (a) In our first experiment, trials presented pairs of composite arrangements simultaneously. Composites were visible until a response was registered. (b) In our second experiment, trials presented pairs of face composites sequentially. Composites were presented for 200 msec each, with an inter-stimulus-interval of 400 msec during which a black display was presented.

Testing took place at City, University of London. Participants judged whether the regions shown within the guidelines were identical or not. Composite displays were presented until a response was registered. Participants were asked to respond with both speed and accuracy. Each pair was presented twice in each alignment condition with side (left or right) counterbalanced, yielding 120 'same' trials and 120 'different' trials (10 pairs × 2 presentations × 2 levels of alignment × 3 composite types). Composite type (upright faces, inverted faces, pseudo-words) was interleaved randomly within blocks of 60 trials. Six practice trials were provided. The experiment was programmed in MATLAB (The MathWorks, Natick, MA) using the Psychophysics Toolbox (Brainard, 1997; Pelli, 1997).

Prior to testing the DPs and age-matched controls, we piloted our novel procedure on a group of 25 young neurotypical adults (M_{age} = 18.92 years, SD_{age} = 1.42 years, 3 males) to ensure the tasks yielded the expected results. These data are provided in the supplementary material. The sample exhibited a clear composite effect for upright faces that accords closely with the existing literature. Reassuringly, we found disproportionate effects of Alignment on ‘same’ trials, where the presence of the illusion makes it harder to detect that target regions are identical, consistent with previous reports (e.g., Le Grand, Mondloch, Maurer, & Brent, 2004). As expected, composite effects were greatly diminished when arrangements were constructed from inverted faces.

4.3.2 Results

Where stimulus displays are visible until participants respond, there is a trade-off between response speed and response accuracy; slower responding allows observers to collect more perceptual evidence, and thereby reduce errors. Under these conditions, many observers approach ceiling on accuracy measures (e.g., Calder, Young, Keane, & Dean, 2000; Palermo et al., 2011). To facilitate clear interpretation, we therefore present both the response speed and accuracy data (Table 4.2).

			Aligned same	Misaligned same	Aligned different	Misaligned different
Upright faces	Accuracy (%)	NT	73.8 (20.0)	95.3 (7.2)	95.9 (6.6)	92.2 (9.3)
		DP	80.6 (15.2)	92.8 (8.0)	90.9 (9.5)	89.1 (14.3)
	RT (ms)	NT	2840 (1228)	1964 (711)	1937 (992)	1925 (1006)
		DP	3257 (1241)	2430 (981)	2383 (945)	2243 (974)
Inverted faces	Accuracy (%)	NT	94.1 (6.4)	95.3 (5.9)	86.9 (14.6)	95.3 (14.8)
		DP	96.9 (4.0)	94.1 (12.3)	84.7 (12.3)	82.2 (10.6)
	RT (ms)	NT	2028 (741)	1882 (661)	2095 (990)	1882 (909)
		DP	2323 (969)	2120 (753)	2438 (787)	2522 (1068)

Table 4.2. Mean accuracy and response time measures from Experiment 1. Standard deviations are shown in parentheses.

4.3.2.1 Accuracy

First, we compared the composite face effects exhibited by the groups in their accuracy data (Figure 4.2). Our analyses revealed evidence of clear composite effects for upright faces. As expected, we observed a significant main effect of Alignment

[F(1,30) = 19.04, p < .001, η^2 = .388], a main effect of Trial Type [F(1,30) = 5.91, p = .021, η^2 = .165], and an Alignment × Trial Type interaction [F(1,30) = 36.72, p < .001, η^2 = .550]. The analysis indicated that the composite effects exhibited by the controls and DPs did not differ. We observed no main effect of Group [F(1,30) = .145, p = .706, η^2 = .005], and the effects of Alignment [F(1,30) = .1.35, p = .254, η^2 = .043], Trial Type [F(1,30) = 1.41, p = .245, η^2 = .045], and the Alignment × Trial Type interaction [F(1,30) = 2.99, p = .094, η^2 = .091], did not interact with Group. We also note that the Alignment × Group interaction failed to reach significance when the analysis was restricted to 'same' trials [F(1,30) = 2.61, p = .117]. When considered separately, the neurotypical controls showed effects of Alignment [F(1,15) = 12.187, p = .003, η^2 = .448] and an Alignment × Trial Type interaction [F(1,15) = 35.161, p < .001, η^2 = .701]. Clear effects of Alignment [F(1,15) = 6.855, p = .019, η^2 = .314] and an Alignment × Trial Type interaction [F(1,15) = 8.238, p = .012, η^2 = .355] were also seen in the DP group.

Neither group showed evidence of composite effects for inverted faces. The analysis revealed a significant effect of Trial Type [F(1,30) = 23.43, p < .001, η^2 = .439], but the effects of Alignment [F(1,30) = 1.29, p = .264, η^2 = .041], and the Alignment × Trial Type interaction [F(1,30) = .41, p = .527, η^2 = .013] failed to reach significance. As expected, the main effects of Trial Type [F(1,30) = 60.96, p = .000, η^2 = .670] and Alignment [F(1,30) = 16.71, p = .000, η^2 = .358] both varied significantly as a function of Composite Type (upright face, inverted face). We observed no main effect of Group [F(1,30) = .09, p = .763, η^2 = .003], and none of the other main effects or interactions varied as a function of group [all F's < 0.9, p's > .35].

4.3.2.2 Response times

Next, we compared the composite face effects exhibited by the groups in their response time data (Figure 4.2). Analysis of response latencies for the upright faces revealed main effects of Alignment [F(1,30) = 56.339, p < .001, η^2 = .653], and Trial Type [F(1,30) = 28.80, p < .001, η^2 = .490], and an Alignment × Trial Type interaction [F(1,30) = 32.219, p < .001, η^2 = .518]. The analysis indicated that similar composite face effects were seen for controls and DPs. No effect of Group was observed [F(1,30) = 1.496, p = .231, η^2 = .048], and the effects of Alignment [F(1,30) = .101, p = .753, η^2 = .003], Trial Type [F(1,30) = .101, p = .753, η^2 = .003], and the Alignment × Trial

Type interaction [$F(1,30) = .424$, $p = .520$, $\eta^2 = .014$], did not vary as a function of Group. Once again, the Alignment × Group interaction failed to reach significance when the analysis was restricted to 'same' trials [$F(1,30) = .043$, $p = .838$]. The neurotypical controls showed effects of Alignment [$F(1,15) = 25.108$, $p < .001$, $\eta^2 = .626$] and an Alignment × Trial Type interaction [$F(1,15) = 14.720$, $p = .002$, $\eta^2 = .495$]. Highly significant effects of Alignment [$F(1,15) = 31.517$, $p < .001$, $\eta^2 = .678$] and an Alignment × Trial Type interaction [$F(1,15) = 19.722$, $p < .001$, $\eta^2 = .568$] were also seen in the DP group.

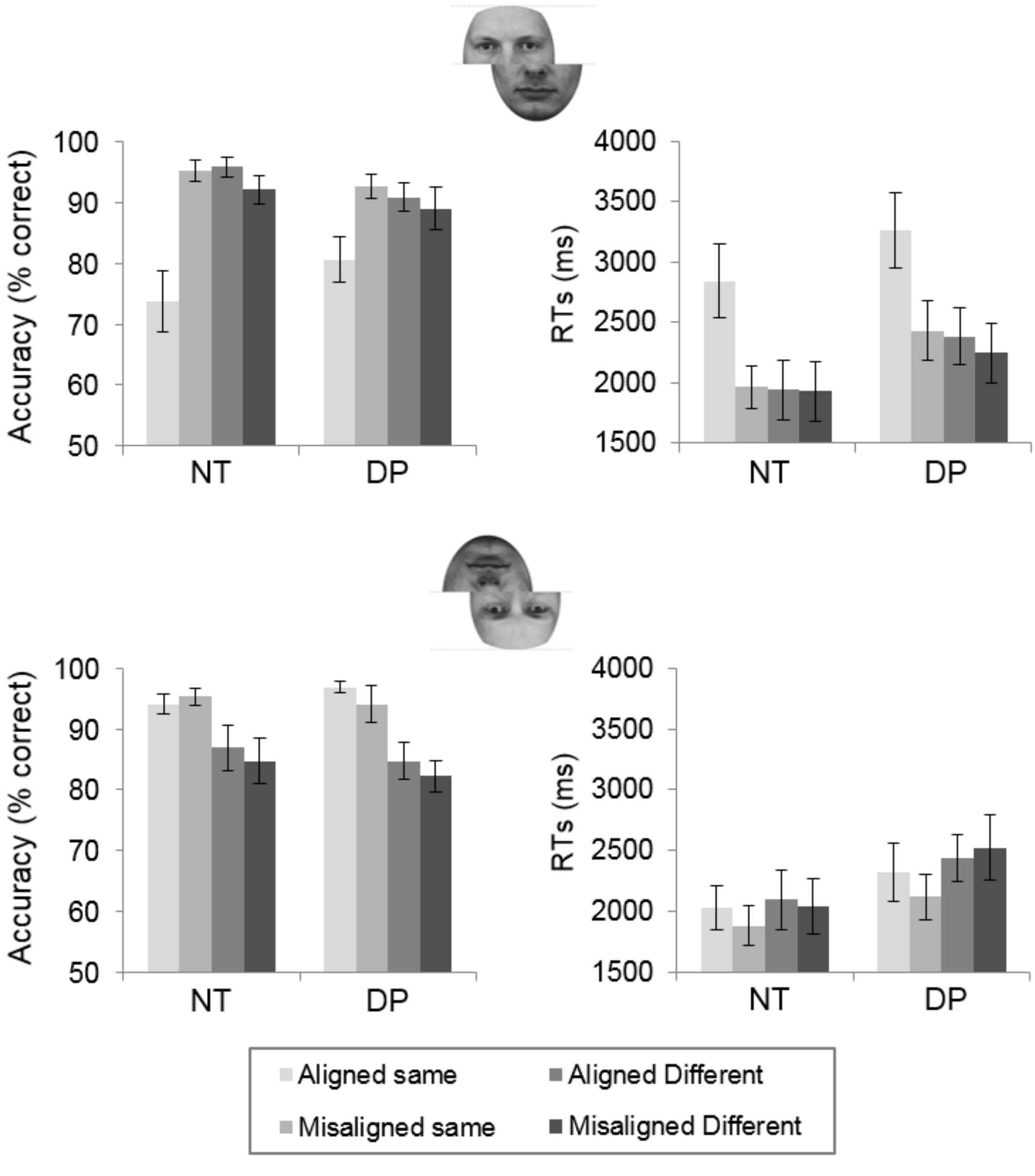

Figure 4.2. Results from Experiment 1 for composite arrangements constructed from upright faces (top) and inverted faces (bottom). Error bars represent ±1 standard error of the mean.

Neither group showed evidence of a composite face effect for inverted faces in their response time data. The main effects of Trial Type [$F(1,30) = 3.421$, $p = .075$, $\eta^2 = .102$] and Alignment [$F(1,30) = 2.831$, $p = .103$, $\eta^2 = .086$], and the Alignment × Trial Type interaction [$F(1,30) = 2.808$, $p = .104$, $\eta^2 = .086$], all failed to reach significance. The main effect of Alignment [$F(1,30) = 20.646$, $p < .001$, $\eta^2 = .408$] and the Alignment × Trial Type interaction [$F(1,30) = 10.638$, $p = .003$, $\eta^2 = .262$] varied significantly as a function of Composite Type (upright faces, inverted faces). No main effect of Group was observed [$F(1,30) = 1.459$, $p = .236$, $\eta^2 = .046$] and none of the effects or interactions varied as a function of Group [all F's < 0.8, p's > .38].

4.3.2.3 Individual differences

Next we sought to determine how susceptibility to the composite face effect related to individual differences in face processing ability in our sample of 16 DPs. Scores on the CFMT ($r = -.186$, $p = .491$) and the upright CFPT ($r = .219$, $p = .416$) failed to correlate with a measure of the composite effect based on accuracy (Δaccuracy = $\%Correct_{aligned} - \%Correct_{misaligned}$).

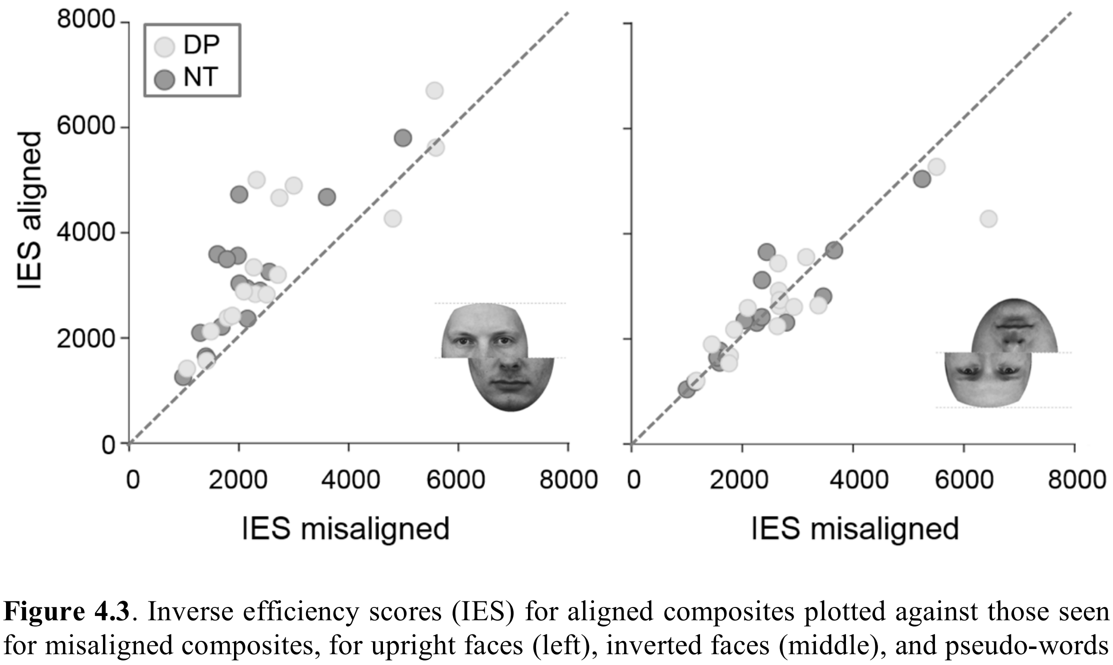

Figure 4.3. Inverse efficiency scores (IES) for aligned composites plotted against those seen for misaligned composites, for upright faces (left), inverted faces (middle), and pseudo-words (right). Points lying to the left of the dashed line are indicative of typical composite effects (performance misaligned > performance aligned).

Similarly, composite effects based on response time (Δlatency = $RT_{aligned}$ - $RT_{misaligned}$), failed to correlate with performance on the CFMT (r = .194, p =.471) or the upright CFPT (r = -.072, p =.792). Finally, we sought to derive a single measure of performance that combined response times and accuracy. We therefore computed Inverse Efficiency Scores (IES; Figure 4.3) by adjusting participants' response times (RTs) upwards in proportion to their error rate [IES = RT / % correct] (Townsend & Ashby, 1978). No correlation was observed between composite face effects (ΔIES = $IES_{aligned}$ - $IES_{misaligned}$) and their performance on the CFMT (r = .216, p =.422) or their CFPT scores (r = -.176, p = .514).

4.4 Experiment 2

In our first experiment, we examined whether 16 individuals with DP exhibited diminished composite face effects using a simultaneous matching paradigm. Contrary to previous reports (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011), we found that the DPs and controls exhibited comparable composite face effects. However, DP is known to be a heterogeneous condition (Eimer, Gosling, & Duchaine, 2012; Stollhoff, Jost, Elze, & Kennerknecht, 2011; Susilo & Duchaine, 2013). For example, some individuals appear to perceive facial expressions normally, whereas others exhibit impaired expression recognition (Biotti & Cook, 2016; Duchaine, Parker, & Nakayama, 2003; Duchaine, Yovel, Butterworth, & Nakayama, 2006; Humphreys, Avidan, & Behrmann, 2007). Similarly, some individuals with DP recognize objects normally, while others exhibit broader object recognition deficits (Behrmann, Avidan, Marotta, & Kimchi, 2005; Biotti, Gray, & Cook, 2017; Dalrymple, Elison, & Duchaine, 2017; Duchaine, Germine et al., 2007). In light of this heterogeneity, it is possible that a subgroup of the DP population exhibits diminished composite effects, but is under-represented in our first sample.

Moreover, the use of simultaneous matching in Experiment 1 differs from the sequential matching tasks employed in the previous studies that have reported group differences (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011). In our second experiment, we therefore tested a different group of DPs with a sequential matching composite task.

4.4.1 Methods

4.4.1.1 Participants

Twenty-four individuals with DP (M_{age} = 40.1 years, SD_{age} = 13.2 years, 6 males) participated in the study. The performance of the DPs was compared to a control group comprising 22 neurotypical adults (M_{age} = 45.8 years, SD_{age} = 13.9 years, 5 males). All observers were US residents. Ethical approval was granted by the local ethics committee and the study was conducted in line with the Declaration of Helsinki. All participants provided informed consent prior to testing.

4.4.1.2 Diagnostic testing

DP participants were recruited through the Dartmouth/Harvard/UCL Prosopagnosia Research Center website (www.faceblind.org). All complained of lifelong face recognition difficulties that affected their daily lives.

Participant	Age	FFT_{US} %	ONFT A′	CFMT %	CFPT upright [errors]
F1	23	8 (−7.2)	.87 (−4.5)	45.83 (−3.1)	54 (−1.4)
F2	26	23 (−5.9)	–	58.33 (−2.0)	–
F3	27	27 (−5.5)	.81 (−7.5)	51.39 (−2.6)	54 (−1.4)
F4	27	–	.83 (−6.5)	55.56 (−2.3)	66 (−2.4)
F5	29	63 (−2.2)	.69 (−13.5)	50.00 (−2.8)	54 (−1.4)
F6	31	61 (−2.4)	.98 (1.0)	56.94 (−2.1)	52 (−1.3)
F7	32	51 (−3.3)	.89 (−3.5)	54.17 (−2.4)	78 (−3.4)
F8	34	–	.77 (−9.5)	56.94 (−2.1)	48 (−.9)
F9	38	58 (−2.7)	.87 (−4.5)	61.11 (−1.8)	62 (−2.1)
F10	38	36 (−4.7)	.77 (−9.5)	47.22 (−3.0)	56 (−1.6)
F11	41	28 (−5.4)	.87 (−4.5)	38.89 (−3.8)	92 (−4.5)
F12	41	40 (−4.3)	.91 (−2.5)	47.22 (−3.0)	42 (−.4)
F13	44	61 (−2.4)	.82 (−7.0)	52.78 (−2.5)	–
F14	44	40 (−4.3)	.90 (−3.0)	51.39 (−2.6)	34 (+.2)
F15	46	50 (−3.4)	.81 (−7.5)	58.33 (−2.0)	62 (−2.1)
F16	51	45 (−3.9)	.91 (−2.5)	61.11 (−1.8)	42 (−.4)
F17	60	33 (−5.0)	.75 (−10.5)	51.39 (−2.6)	70 (−2.7)
F18	62	48 (−3.6)	.81 (−7.5)	50.00 (−2.8)	70 (−2.7)
M1	23	26 (−5.6)	.90 (−3.0)	47.22 (−3.0)	92 (−4.5)
M2	28	24 (−5.8)	.94 (−1.0)	51.39 (−2.6)	62 (−2.1)
M3	34	56 (−2.9)	–	45.83 (−3.1)	80 (−3.5)
M4	58	33 (−5.0)	.81 (−7.5)	50.00 (−2.8)	78 (−3.4)
M5	62	43 (−4.0)	.93 (−1.5)	56.94 (−2.1)	62 (−2.1)
M6	63	40 (−4.3)	.87 (−4.5)	56.94 (−2.1)	50 (−1.1)
DP mean		40.64	.85	52.37	61.82
DP SD		14.63	.07	5.47	15.48
Comparison mean		87.5	.96	80.4	36.7
Comparison SD		11.0	.02	11.0	12.2

Table 4.3. Scores for each developmental prosopagnosic in Experiment 2 on the Cambridge Face Memory Test (CFMT), The Famous Faces Test (FFT_{US}), and the Old-New Faces Test (ONFT). *Z*-scores are shown in parentheses. Negative *z*-scores denote performance worse than the typical mean. The mean and standard deviation of the comparison samples are provided below.

Convergent diagnostic evidence for the presence of DP was collected using the CFMT, the Old-New Face Recognition Test (ONFRT; Duchaine & Nakayama, 2005), and a Famous Faces Test suitable for use with US residents (FFT_{US}; Duchaine & Nakayama, 2005).

When tested on the CFMT, all DPs scored at least 1.7 standard deviations below the mean performance of the comparison sample described by Duchaine and Nakayama (2006a). All DPs tested[2] also scored at least 2 standard deviations below the mean of the controls on the FFT_{US} and the ONFRT (comparison data taken from Duchaine, Yovel, & Nakayama, 2007; Susilo, Wright, Tree, & Duchaine, 2015). DPs also completed the CFPT and the Leuven Perceptual Organization Screening Test (L-POST; Torfs, Vancleef, Lafosse, Wagemans, & de-Wit, 2014). All DPs scored within the normal range on the L-POST, suggesting typical mid-level vision. Detailed diagnostic results are provided in Table 4.3.

4.4.1.3 The composite task

The stimuli and procedure were adapted from the composite task employed by Susilo et al. (2013; Experiment 3). Face composites were constructed from greyscale photographs of Caucasian male children posing neutral expressions (Figure 4.1b). The children were photographed wearing a black ski-cap to occlude their hairline. When viewed from 40 cm, aligned faces subtended 10° vertically and 6.5° horizontally, and misaligned faces 10° × 9°. All subjects were tested remotely via www.testable.org, a platform that enables precise control of experiments conducted online[11]. Participants were asked to do the task in an environment in which they would not be disturbed and to employ a viewing distance of around 40 cm.

Experimental trials presented two face composites sequentially for 200 ms each, with an inter-stimulus interval of 400 ms during which a black display was presented. Composites were either both aligned or both misaligned, both upright or both inverted (Figure 4.1b). Participants were asked to indicate with a keypress whether the target regions (the face halves containing the eyes) were the “same” (identical) or “different” (not identical) while ignoring the distractor regions, which were always different. There

[11] One DP had technical difficulties, but a switch to another browser resolved the issue. This individual completed approximately one third of the trials before the task crashed, at which point the individual switched browsers and did the full task on the new browser.

were 90 trials per orientation; 60 in which the target regions were the same (30 aligned, 30 misaligned) and 30 where the target regions were different (15 aligned, 15 misaligned), making 180 trials in total. Orientation (upright, inverted), Alignment (aligned, misaligned), and Trial Type (same, different) were randomly interleaved. Six practice trials were provided.

4.4.2 Results

Matching procedures that present composites sequentially for pre-determined intervals (in this case 200 ms) afford less opportunity for a trade-off between speed and accuracy, because participants cannot accumulate more perceptual evidence by responding slowly. In Experiment 2, our primary analyses focus on accuracy (% correct). Descriptive statistics for accuracy scores and RTs achieved by the two groups are presented in Table 4.4.

			Aligned	Misaligned
Upright faces	Accuracy (%)	NT	65.9 (16.5)	92.0 (9.4)
		DP	67.6 (16.9)	91.2 (8.0)
	RT (ms)	NT	1105 (362)	874 (234)
		DP	1090 (277)	978 (255)
Inverted faces	Accuracy (%)	NT	88.0 (12.9)	88.8 (13.8)
		DP	86.7 (11.2)	91.7 (8.6)
	RT (ms)	NT	928 (247)	889 (215)
		DP	991 (260)	999 (303)

Table 4.4. Mean accuracy and response time measures from Experiment 2. Standard

4.4.2.1 Accuracy

The combined dataset was subjected to ANOVA with Alignment (misaligned, aligned) and Orientation (upright, inverted) as within-subjects factors, and Group (DP, NT) as a between-subjects factor (Figure 4.4). The analysis revealed main effects of Orientation [$F(1,44) = 30.96$, $p < .001$, $\eta^2 = .413$] and Alignment [$F(1,44) = 84.33$, $p < .001$, $\eta^2 = .65$], as well as a highly significant Alignment × Orientation interaction [$F(1,44) = 75.21$, $p < .001$, $\eta^2 = .63$], reflecting a larger difference between aligned and misaligned trials when composites were shown upright. The main effect of Group was not significant [$F(1,44) = 0.20$, $p = .65$], and neither the Group × Orientation interaction [$F(1,44) = 0.07$, $p = 0.79$], nor the Group × Alignment interaction [$F(1,44) = 0.61$, $p = .44$] reached significance. Most critically, however, the Orientation × Alignment interaction did not vary as a function of Group [$F(1,44) = 0.75$, $p = .39$]. As expected, controls' ability to discriminate the misaligned target regions exceeded their discrimination of the aligned targets when the faces were upright [$t(21) = 6.95$, $p < .001$, Cohen's $d = 1.48$], but not when arrangements were inverted [$t(21) = .33$, $p = .75$]. The DPs exhibited a similar pattern, but their ability to discriminate the misaligned target regions exceeded their discrimination of the aligned targets in both the upright [$t(23) = 7.78$, $p < .001$, Cohen's $d = 1.59$] and inverted [$t(23) = 2.70$, $p = .013$, Cohen's $d = .55$] conditions.

Unlike controls, DPs showed an effect of alignment for inverted trials. Nevertheless, we do not believe this difference is indicative of qualitatively differently face processing. First, the Alignment × Orientation interaction did not vary as a function of Group; both the DP and NT controls showed much larger alignment effects for upright faces than for inverted faces. Second, it is not uncommon for typical observers to show small but significant composite effects for inverted faces[12]. For example, Susilo and colleagues (2013) used the same inverted composite task used here and found a significant alignment effect in a large sample of typical observers ($N = 242$) with a magnitude similar to that exhibited by the DPs in this experiment (Typical observers: 4.0%, DPs: 5.0% respectively).

4.4.2.2 Response times

The response latency data was analysed using a mixed-model ANOVA with Orientation (upright, inverted) and Alignment (aligned, misaligned) as within-subjects factors, and Group (DP, NT) as a between-subjects factor. Main effects of Orientation [$F(1,44) = 12.71$, $p = .001$, $\eta^2 = .22$] and Alignment [$F(1,44) = 22.04$, $p < .001$, $\eta^2 = .32$] were observed, as well as a significant Orientation × Alignment interaction

[12] Composite face stimuli that include a gap of a few pixels between the target and distractor regions may be less likely to produce composite effects when arrangements are inverted (Rossion & Retter, 2015). It remains unknown how the presence or absence of this feature affects composite face processing in observers with DP. Addressing this issue in future studies of the composite effect in DP may prove worthwhile.

[F(1,44) = 21.80, p < .001, η^2 = .32]. However, no main effect of Group was observed [F(1,44) = .46 p = .50]. The effects of Orientation [F(1,44) = .60, p =.44], Alignment [F(1,44) = 2.58, p = .12], and the Orientation × Alignment interaction failed to interact with Group [F(1,44) = .88, p = .35].

4.4.2.3 Individual differences

Once again, no correlation was observed between the DPs' composite face effects (Δaccuracy = %Correct$_{aligned}$ - %Correct$_{misaligned}$) seen in the upright condition and their scores on the CFMT (r = -.05, p = .81) or CFPT (r = -.07, p = .77).

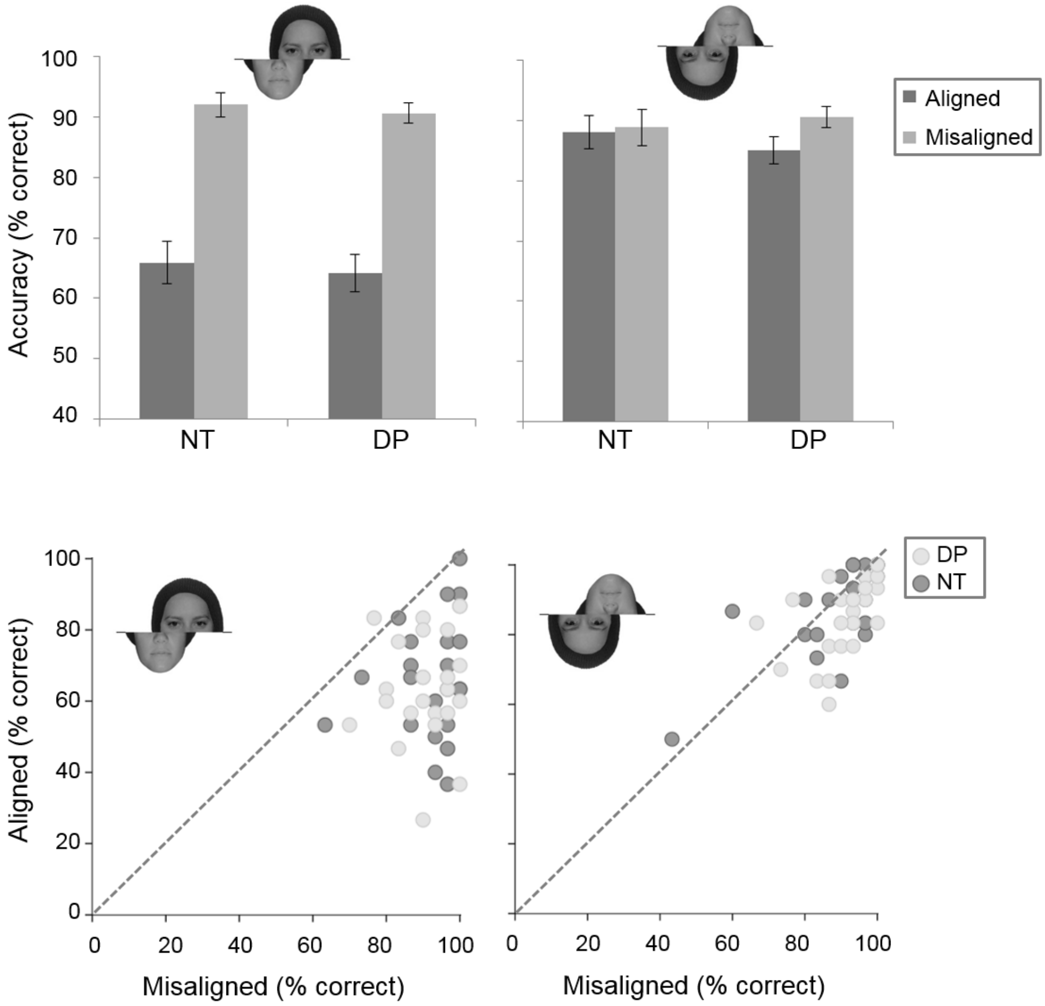

Figure 4.4. Results of Experiment 2. Top panels present accuracy scores for the two groups on the upright (left) and inverted composites (right). Error bars represent ±1 standard error of the mean. Bottom panels show accuracy scores seen for aligned composites plotted against those seen for misaligned composites, for upright faces (left) and inverted faces (right). Points lying to the right of the dashed line are indicative of typical composite effects (performance misaligned > performance aligned).

We present the individual effects seen for the DPs and age-matched controls (Figure 4.4) to illustrate that the failure to find a group difference is not due to the presence of outliers.

Some cases of developmental prosopagnosia appear to have an apperceptive profile – whereby individuals have problems forming perceptual descriptions of faces – while other cases may have selective problems with face learning or face memory (De Renzi et al., 1991). Insofar as the whole-face binding revealed by composite face effect has been characterised as a face encoding process (Murphy et al., 2017; Rossion, 2013), it is possible that susceptibility to the composite face effect is reduced only in apperceptive cases of DP. We took advantage of the large sample size employed in Experiment 2 to examine this possibility in more detail. The DPs were split into apperceptive (N = 12) and non-apperceptive (N = 12) subgroups. Members of the apperceptive subgroup performed at least 2 SDs below the mean of the comparison sample on the CFPT. Contrary to the foregoing speculation, however, we found no difference in the size of the composite effects (Δaccuracy) exhibited by the subgroups in the upright [$t(22) = .324$, $p = .749$] or inverted [$t(22) = .273$, $p = .787$] conditions. The lack of relationship between scores on the CFPT and composite effect sizes accords with previous findings with typical observers (Rezlescu et al., 2017) and DPs (Palermo et al., 2011).

4.5 Discussion

The present study assessed whether individuals with DP exhibit diminished composite face effects at the group level. Across two experiments conducted on separate samples and using different paradigms, we find no evidence for diminished composite-face effects in this population. In our first experiment, a group of 16 DPs showed typical composite face effects when tested on a simultaneous matching procedure. In our second experiment, a separate group of 24 DPs also showed typical composite face effects when tested on a sequential matching procedure. Contrary to previous reports (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011), these findings indicate that diminished composite face effects are not a characteristic feature of DP. These results have important implications, both for our understanding of DP and for our interpretation of the composite face effect.

4.5.1 Composite face effects in developmental prosopagnosia

Our results accord with findings from previous case studies that have described typical composite face effects in individual DPs (Le Grand et al., 2006; Schmalzl et al., 2008; Susilo et al., 2010). In particular, Le Grand and colleagues (2006) described typical composite effects in seven out of eight DPs tested. Similarly, having tested seven family members with DP, Schmalzl et al. (2008) found typical composite effects in the four youngest cases (aged 4-40 years) and atypical composite effects only in the three oldest cases (aged 66-87 years). Interestingly, we note recent findings from typical observers suggesting that composite face effects may behave differently in samples of older adults; for example, the composite processing of older observers may be less efficient (Wiese, Kachel, & Schweinberger, 2013) and be more susceptible to general factors (Meinhardt, Persike, & Meinhardt-Injac, 2016). In contrast, our results are inconsistent with previous reports of reduced composite face effects in DP at the group level (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011). Having examined the processing of upright and inverted face composites in 40 individuals with DP (aged 21-63 years), our results suggest most members of this population exhibit normal composite face effects. On the other hand, close examination of the previous group studies calls their conclusions into doubt.

In their first experiment, Palermo and colleagues (2011) found that a sample of 12 DPs were slower to name the emotion of a target region when aligned with a distractor region expressing an incongruous emotion. However, inspection of the distribution suggests this difference was strongly influenced by the results from a single DP whose aligned RTs were considerably faster than their misaligned RTs - a reversed composite effect (see Palermo et al., 2011, Figure 5). Further complicating interpretation, neither the DPs nor the controls showed composite effects in their error rates. In their second experiment, controls and nine DPs were required to match the top halves of face composites presented sequentially for 200 ms each. Given the short presentations, accuracy is the most critical measure of composite effects, and the DPs and controls showed clear and nearly identical composite effects in their accuracy data. The evidence for atypical composite effects cited by the authors is derived from RTs. However, the Alignment × Group interaction seen in the RT data failed to reach significance when analysed in the standard manner ($p > .3$). The group difference was only significant when adjusted for performance in the baseline misaligned condition, a point we discuss further below.

Avidan and colleagues (2011) reported that a sample of 14 individuals with DP showed diminished effects of alignment both in their RTs and error rates, when matching upright composites presented sequentially. The age of the DP sample is older than is typical in this literature; half the DP participants were aged 60 years or older (mean age = 52.5 years; range 31-79 years). Inspection of the single-case data is further complicated by the fact that aligned and misaligned trials were blocked, and completed in a different order by different DPs. Whilst this treatment may have little effect on the performance of typical observers (e.g., Le Grand et al., 2004), DPs may be prone to order effects resulting from practice, fatigue, or test anxiety. Within their DP sample, those individuals who showed weaker composite face effects showed greater local bias (r = .52) on a compound letter task (Navon, 1977). Where observed, weaker composite face effects therefore seem to be related to wider global processing difficulties. It is possible that a subgroup exists within the DP population characterized by a global processing deficit affecting performance on composite face and compound letter tasks. However, the present results together with previous reports, suggest that this profile is relatively uncommon. For example, many DPs exhibit typical perception of global motion and Glass patterns (Le Grand et al., 2006), typical Gestalt completion (Duchaine, 2000; Duchaine et al., 2006), and process compound 'Navon' stimuli typically (Duchaine, Germine et al., 2007; Duchaine, Yovel et al., 2007; Schmalzl et al., 2008).

Lastly, Liu & Behrmann (2014) reported that eight DPs showed reduced composite effects for left and right face halves when tested using the complete design. However, several factors undermine our confidence in these findings. First, the three DPs with the lowest holistic processing index, exhibited surprisingly normal performance on the diagnostic tests (e.g. MN and SH had CFMT scores of 73.6% and 79.2%, and WA exhibited above average famous face recognition). Second, inspection of the composite results indicates that the DPs performed much worse in the baseline misaligned condition than the matched controls. This feature of the data suggests that the reduced composite effects described reflect problems encoding local regions rather than aberrant integration processes. Distractor halves perceived as homogenous or nondescript by prosopagnosics may afford weaker perceptual prediction, and thereby exert less illusory bias in the aligned condition, than distractor halves perceived as distinctive. In an attempt to factor in baseline differences, the authors computed a holistic processing index, where modulation in the aligned condition is expressed relative to misaligned performance. Crucially, this measure and similar indices (see Avidan et al., 2011; Palermo et al., 2011) make unfounded assumptions about the relationship between performance in misaligned conditions and susceptibility to the composite effect; it is not clear what constitutes a "typical" composite effect where observers exhibit atypical misaligned performance.

Traditionally, it has been assumed that the face inversion (Yin, 1969), composite face (Young et al., 1987), and part-whole effects (Tanaka & Farah, 1993), reflect the operation of a single process or mechanism (Farah et al., 1998; Maurer et al., 2002; McKone & Yovel, 2009; Piepers & Robbins, 2013). However, mounting evidence suggests that individuals' susceptibility to the composite face effect not only fails to correlate with their face recognition ability, but also appears weakly related to other putative measures of holistic face processing (Rezlescu et al., 2017; Wang et al., 2012; but see DeGutis, Wilmer et al., 2013). As a result, we do not wish to claim that every facet of holistic face processing is typical in DP. Given that different measures of holistic processing are unrelated or weakly correlated in the typical population, neuropsychological dissociations might also be seen in the DP population. While DPs may show typical susceptibility to the composite face effect, other effects attributed to holistic face processing may be aberrant; for example, many DPs may show diminished face inversion effects (Duchaine et al., 2006; Shah, Gaule, Gaigg, Bird, & Cook, 2015; Tree & Wilkie, 2010), absent part-whole effects for the eye region (DeGutis et al., 2012), and commonly report excessive reliance on local features for identity recognition (DeGutis et al., 2012; Shah, Gaule, Sowden et al., 2015).

It is worth noting an interesting inconsistency in the DP literature highlighted by our findings. In both experiments, our DPs showed large composite effects with upright faces yet little or no composite effects with inverted faces (see also Susilo et al., 2010). Most DPs also show better performance with upright faces than inverted faces when tasks are sensitive and performance is not affected by restrictions of range (Duchaine, Germine et al., 2007; Duchaine, Yovel et al., 2007; Garrido, Duchaine, & Nakayama, 2008). Similarly, a study comparing event-related potentials (ERPs) indicated upright and inverted Mooney faces were processed differently by DPs (Towler, Gosling, Duchaine, & Eimer, 2016). These results indicate that DPs process upright and inverted faces differently, however they are inconsistent with findings from an ERP study of face processing in DP (Towler, Gosling, Duchaine, & Eimer, 2012). In typical observers, inverted faces reliably elicit larger N170 potentials than upright faces (Bentin, Allison, Puce, Perez, & McCarthy, 1996; Eimer, 2000; Rossion et al., 1999). A group of 16 DPs, however, showed no difference in their N170s to upright and inverted faces at the group level (Towler, Fisher, & Eimer, 2017; Towler et al., 2012). While the reason for the discrepancy between these findings is unclear, it appears that behavioural inversion effects and the N170 inversion effect are measuring different aspects of face processing.

4.5.2 Composite face effect and face recognition ability

The view that individual differences in holistic face processing, inferred from susceptibility to the composite face effect, predict face recognition ability is widespread (DeGutis et al., 2013; Farah et al., 1998; Maurer et al., 2002; Piepers & Robbins, 2013; Richler et al., 2011). This interpretation owes much to the correlated observations that orientation inversion renders faces harder to recognise (Yin, 1969) and greatly reduces the composite face effect (Young et al., 1987). Consistent with this view, composite studies employing the congruency design have found a positive correlation between composite effects and face recognition ability (DeGutis et al., 2013; Richler et al., 2011). However, the functional significance of the composite face effect has been called into question by other studies that have found little or no correlation between typical observers' composite face effects – measured using the standard design – and their face recognition ability (Konar et al., 2010; Rezlescu et al., 2017; Wang et al., 2012). Reports of diminished composite face effects in DP (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011) have been cited as evidence that the process responsible for the composite face effect makes a necessary contribution to face recognition ability (Murphy et al., 2017). Our findings suggest this inference is potentially misleading.

Typical composite effects in the DPs tested here, and in other cases described previously (Le Grand et al., 2006; Schmalzl et al., 2008; Susilo et al., 2010), as well as evidence that some acquired prosopagnosics exhibit normal face composite effects (Finzi et al., 2016), suggest a complex relationship between susceptibility to the composite face effect and face recognition ability. Face recognition is thought to depend on a processing stream that can be fractionated at several stages (Bruce & Young, 1986). The whole-face binding indexed by the composite effect appears to be intact in individuals with DP suggesting that the locus of their impairment lies elsewhere in the face processing stream. However, the binding process revealed by the composite effect may still make a causal contribution to face recognition ability; i.e., the composite process may be necessary, but not sufficient, for typical face perception. Cases of acquired prosopagnosia have been described where face recognition deficits are associated with aberrant composite effects (e.g., Busigny et al., 2010; Busigny et al., 2014; Ramon et al., 2009), and no neuropsychological cases have been described who show no evidence of a composite effect but normal performance on tests of face perception and face recognition.

Typical composite face effects in DP and in some cases of acquired prosopagnosia (Finzi et al., 2016; Rezlescu et al., 2012), accord with other evidence that the processes underlying the composite effect are difficult to disrupt. Photographic negation disrupts observers' ability to encode 3D face shape (Kemp, Pike, White, & Musselman, 1996), but has little effect on the strength of the composite face effect (Hole, George, & Dunsmore, 1999; Taubert & Alais, 2011). Similarly, composite effects can be seen with abstract cartoon faces that contain only schematic facial features, but bear little resemblance to naturalistic faces (Murphy et al., 2017). Moreover, several markers of face processing, notably the ability to use the internal features (Ellis, Shepherd, & Davies, 1979; Osborne & Stevenage, 2008; Young, Hay, McWeeny, Flude, & Ellis, 1985) and achieve view-point invariance (Longmore, Liu, & Young, 2008), are strongly modulated by facial familiarity. In contrast, compelling composite effects can be seen with entirely unfamiliar faces (Hole, 1994). Together with the findings from prosopagnosia, insensitivity to negation, abstraction, and familiarity, suggest that the composite face effect is resilient and disrupted only by gross changes to the faciotopy (e.g., misalignment, inversion) or catastrophic damage to the face processing stream.

4.5.3 Face composite designs

Like most previous studies of composite effects in DP (e.g., Avidan et al., 2011; Le Grand et al., 2006; Palermo et al., 2011; Schmalzl et al., 2008), we employed the standard design in both experiments, where the distractor regions always differ. There has been considerable debate about the merits of an alternate congruency design, employing a full factorial combination of target regions (same, different) and distractor regions (same, different) (Richler & Gauthier, 2014; Rossion, 2013). Some authors have suggested that congruency designs mitigate the effects of response bias (for discussion see Richler & Gauthier, 2014). However, congruency designs have been criticized because the predicted effect on congruent-different trials – where different distractor halves are paired with different target halves – is unclear (Robbins & McKone, 2007), and because the congruency design produces composite effects for stimuli that do not yield demonstrable composite illusions (Rossion, 2013). The additional trials may induce domain-general facilitation / interference effects that differ from the illusory interference seen for upright-aligned face composites (Murphy et al., 2017; Rossion, 2013). Crucially, because the standard design is thought to limit the domain-general effects of congruency, the present findings represent a conservative test of the hypothesis that composite face effects are diminished in DP. Where observed, domain-general congruency effects may be expected to attenuate a group difference arising from a face-specific deficit.

4.5.4 Conclusion

In summary, we have described two experiments that sought to compare the composite face effects seen in typical observers and those with DP. Having employed complementary procedures and independent samples we find convergent results: evidence of highly significant composite effects in typical controls and DP groups that were indistinguishable. Contrary to previous reports, these results suggest that the whole-face binding process indexed by the composite face effect is intact in DP, indicating that the locus of this condition lies elsewhere in the face processing stream.

4.6 Supplementary material

4.6.1 Pseudo-word composite task

In addition to the upright and inverted composite face conditions employed in Experiment 1, we also examined composite effects for pseudo-words, because they resemble the effects found for upright faces (Anstis, 2005). By employing an additional comparison with a non-face composite effect we hoped to determine whether any diminished composite effects result from a face-specific deficit or from a non-specific problem affecting global processing of configurations. We elected to use pseudo-words in light of recent suggestions that the visual processing of words and faces may recruit similar neurocognitive mechanisms (Behrmann & Plaut, 2013; Hills, Pancaroglu, Duchaine, & Barton, 2015; Ipser, Ring, Murphy, Gaigg, & Cook, 2016).

Figure 4.5. Trials presented pairs of composite arrangements simultaneously. Composites were visible until a response was registered.

Four-letter pseudo-words written in lower-case Juice ITC font were used to create the composites following the procedure described by Anstis (2005). Pseudo-word composites subtended 8° of visual angle, vertically. The to-be-judged regions subtended 4°. In the misaligned conditions, the horizontal offset corresponded to approximately 25% the width of pseudo-word. 40 pseudo-word composites were employed. Each composite was allocated a partner arrangement of the same type with which it would be presented simultaneously. For half the composites pairs, the target regions were identical, for half the pairs the target regions differed. The distractor regions within each pair were always different. The two target regions appeared at the same vertical position in the display (the lower edge of each target region was aligned to the vertical midpoint of the display). Two dashed guidelines were imposed over the arrangements to clearly delineate the stimulus regions to be judged. Example displays are presented in Figure 4.5. Participants judged whether the regions shown within the guidelines were or were not identical. Composite displays were presented until a response was registered. Participants were asked to respond with both speed and accuracy. Arrangements were shown until a response was registered. Each pair was presented twice in each alignment condition with side (left or right) counterbalanced. Composite type (upright faces, inverted faces, pseudowords) was interleaved randomly within blocks of 80 trials.

4.6.2 Pilot testing of composite tasks for upright faces, inverted faces, and pseudo-words

Before testing the simultaneous matching task on the sample of DPs and age-matched controls, we piloted the task on a sample of 25 young neurotypical adults (M_{age} = 18.92 years, SD_{age} = 1.42 years, 3 males). We describe the results here (see Table 4.5 and Figure 4.6).

		Aligned Same		Misaligned same		Aligned different		Misaligned different	
Accuracy (% correct)									
	Upright faces	68.2	(22.5)	94.2	(8.5)	97.8	(3.3)	95.4	(7.2)
	Inverted faces	93.2	(6.8)	95.0	(6.1)	87.8	(8.8)	86.2	(9.9)
	Pseudo-words	84.8	(21.2)	93.8	(9.2)	84.6	(14.2)	80.0	(18.7)
RT (ms)									
	Upright faces	2148	(1080)	1402	(475)	1348	(427)	1280	(356)
	Inverted faces	1555	(538)	1427	(423)	1481	(471)	1538	(508)
	Pseudo-words	3014	(1134)	2335	(999)	2574	(868)	2181	(675)

Table 4.5. Descriptive statistics for the piloting conducted with young neurotypical controls. Standard deviations are shown in parentheses.

4.6.2.1 Accuracy

Analysis of the accuracy data (% correct) for the upright face composites revealed a main effect of Alignment [$F(1,24) = 29.12$, $p = .000$, $\eta^2 = .548$]. Target regions were harder to discriminate in the aligned than in the misaligned condition. We also observed a main effect of Trial Type [$F(1,24) = 28.62$, $p = .000$, $\eta^2 = .544$] and a significant Trial Type × Alignment interaction [$F(1,24) = 45.93$, $p = .000$, $\eta^2 = .657$], whereby aligned distractors were particularly detrimental when targets were the same.

No composite effect was observed for the inverted face arrangements. We did not see a main effect of Alignment [$F(1,24) = .01$, $p = .922$, $\eta^2 = .000$], nor an Alignment × Trial Type interaction [$F(1,24) = 2.36$, $p = .137$, $\eta^2 = .09$]. We observed a main effect of Trial Type [$F(1,24) = 12.12$, $p = .002$, $\eta^2 = .336$], whereby participants made more errors when the target regions differed than when they were identical.

Analyses suggested only a weak pseudo-word composite effect in the accuracy data of the young adults. While we found a main effect of Trial Type [$F(1,24) = 9.15$, $p = .006$, $\eta^2 = .276$] and an Alignment × Trial Type interaction [$F(1,24) = 5.74$, $p = .025$, $\eta^2 = .193$], the critical main effect of Alignment failed to reach significance [$F(1,24) = 1.25$, $p = .274$, $\eta^2 = .05$].

4.6.2.2 Response times

Analysis of response latencies (ms) revealed a main effect of Alignment for upright face composites [$F(1,24) = 21.41$, $p = .000$, $\eta^2 = .471$]. Participants were slower to discriminate target regions when the distractors were aligned than when distractors were misaligned. We also found a main effect of Trial Type [$F(1,24) = 31.08$, $p = .000$, $\eta^2 = .564$], which interacted significantly with Alignment [$F(1,24) = 16.04$, $p = .001$, $\eta^2 = .401$]. When distractor and target regions were aligned, we observed a disproportionate interference effect on same trials.

The response latency analysis revealed little evidence of a composite effect for inverted faces. While we observed a significant Alignment × Trial Type interaction [$F(1,24) = 5.42$, $p = .029$, $\eta^2 = .184$], we found no main effects for either Alignment [$F(1,24) = .83$, $p = .371$, $\eta^2 = .034$], nor Trial Type [$F(1,24) = .14$, $p = .707$, $\eta^2 = .006$].

The response latency analysis revealed a strong composite effect for pseudo-words. We observed a significant main effect of Alignment [$F(1,24) = 71.30$, $p = .000$, $\eta^2 = .748$], whereby participants took longer to discriminate target regions in the aligned condition. We also observed a significant main effect of Trial Type [$F(1,24) = 8.97$, $p = .006$, $\eta^2 = .272$] and a significant Alignment × Trial Type interaction [$F(1,24) = 15.68$, $p = .001$, $\eta^2 = .395$]. Overall participants responded slower on same trials, but this effect was particularly pronounced in the aligned condition.

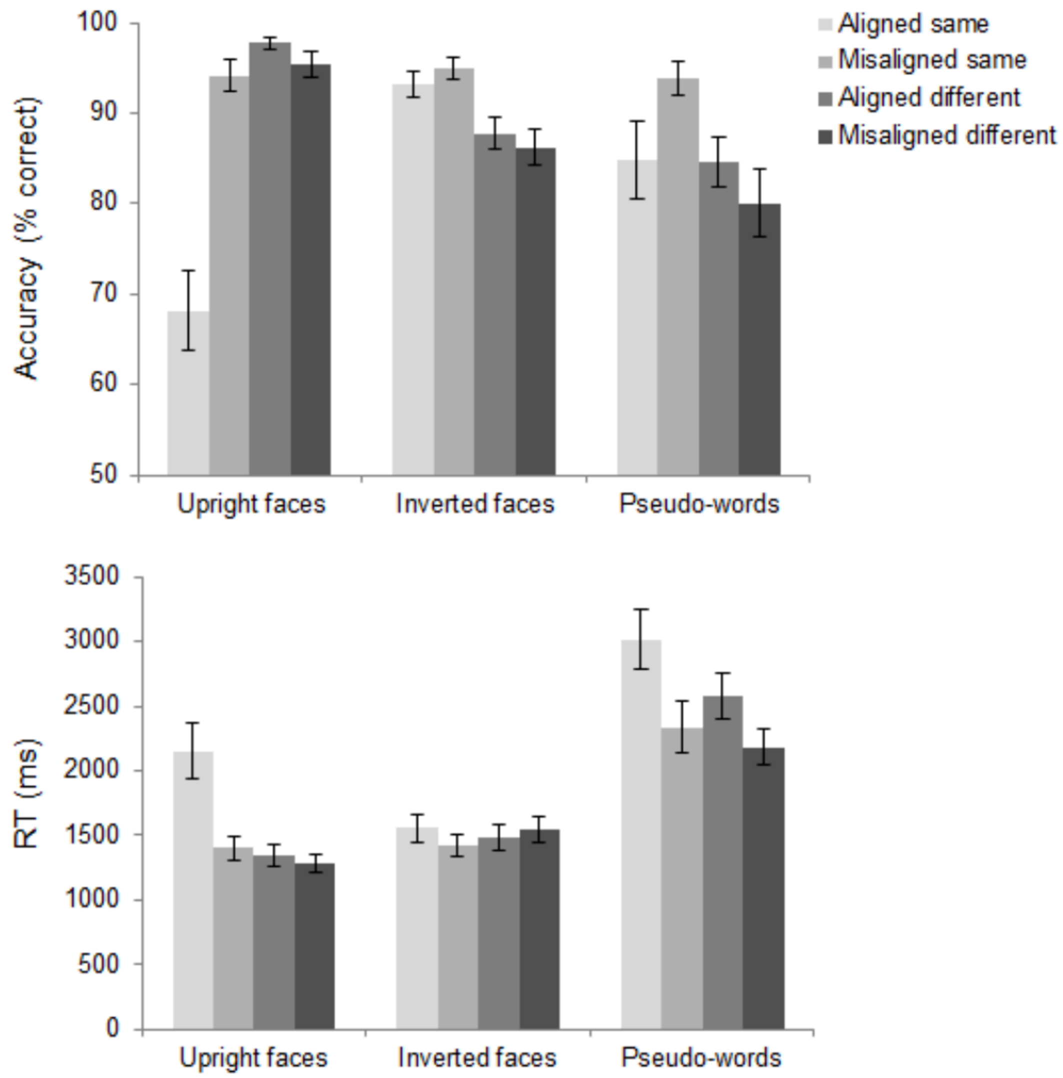

Figure 4.6. Mean accuracy and RTs exhibited by young NT controls during the piloting procedure. Error bars represent ± standard error of the mean.

4.6.3 Group comparison: pseudo-words

Group analyses for the upright and inverted face composites are reported in the main text of the paper. Here we describe additional comparison of the pseudo-word composite effects exhibited by the two groups (see Table 4.6 and Figure 4.7).

		Aligned Same		Misaligned same		Aligned different		Misaligned different	
Accuracy (%)	NT	93.1	(10.6)	95.3	(8.7)	85.0	(15.1)	88.4	(12.2)
	DP	94.7	(5.3)	95.9	(6.6)	90.3	(14.0)	95.6	(4.4)
RT (ms)	NT	3954	(1314)	2891	(962)	3107	(971)	2684	(777)
	DP	4136	(1369)	3342	(1109)	3511	(1217)	2739	(808)

Table 4.6. Mean accuracy and RTs exhibited by the NT and DP groups in the pseudo-word condition.

4.6.3.1 Accuracy

We observed a significant effect of Alignment [$F(1,30) = 8.33$, $p = .007$, $\eta^2 = .217$], whereby participants made more errors in the aligned condition. The main effect of Trial Type was also significant [$F(1,30) = 4.446$, $p = .043$, $\eta^2 = .129$], but the Alignment × Trial Type interaction did not reach significance [$F(1,30) = 1.078$, $p = .308$, $\eta^2 = .035$]. The pseudo-word composite effects were comparable for the two groups: No main effect of Group was observed [$F(1,30) = 2.651$, $p = .114$, $\eta^2 = .081$], and neither the main effect of Alignment [$F(1,30) = .049$, $p = .826$, $\eta^2 = .002$], the main effect of Trial Type [$F(1,30) = 1.220$, $p = .278$, $\eta^2 = .039$], nor Alignment × Trial Type interaction [$F(1,30) = .302$, $p = .587$, $\eta^2 = .010$], varied as a function of Group.

4.6.3.2 Response times

Both groups showed evidence of pseudo-word composite effects in their response latency data. Main effects of Trial Type [$F(1,30) = 51.765$, $p = .000$, $\eta^2 = .633$] and Alignment [$F(1,30) = 95.193$, $p = .000$, $\eta^2 = .760$] were observed, as well as a significant Alignment × Trial Type interaction [$F(1,30) = 15.495$, $p = .000$, $\eta^2 = .341$]. No main effect of Group was observed [$F(1,30) = .560$, $p = .460$, $\eta^2 = .018$]. Neither the effects of Alignment [$F(1,30) = .065$, $p = .801$, $\eta^2 = .002$], nor Trial Type [$F(1,30) = .297$, $p = .590$, $\eta^2 = .010$], varied as a function of Group. However, a significant Alignment × Trial Type × Group interaction was observed [$F(1,30) = 1.220$, $p = .001$, $\eta^2 = .310$]. Whereas the NTs showed disproportionate effects of Alignment on same trails, the DPs showed significant effects of Alignment on both same and different trials.

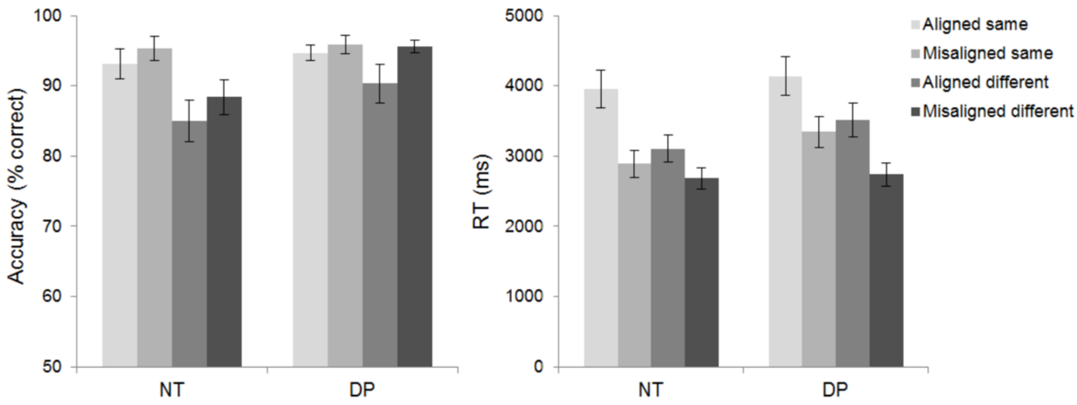

Figure 4.7. Mean accuracy and RTs exhibited by the DPs and aged-matched NT controls during the piloting procedure. Error bars represent ± standard error of the mean.

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Chapter 5: Is developmental prosopagnosia best characterised as an apperceptive or mnemonic disorder?

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Chapter 5: Is developmental prosopagnosia best characterised as an apperceptive or mnemonic disorder?

5.1 Abstract

Traditionally, developmental prosopagnosia (DP) has been thought of as an apperceptive condition that hinders individuals' ability to encode face structure. However, several authors have recently raised the possibility that some DPs, perhaps the majority, may be able to form accurate percepts, but are unable to maintain those percepts over time. The present study sought to distinguish these possibilities. In Experiment 1, 72 DPs and 54 typical controls completed the Cambridge Face Perception Test, a task that measures face perception ability in a way that minimises the memory demands. Not only were the DPs impaired at the group level, but closer analysis suggested that the entire DP distribution was shifted relative to the scores of controls. In Experiment 2, a subset of these participants (16 DPs; 22 controls) completed a delayed match-to-sample task with face and car stimuli, with a retention interval of 1-second (low demand) or 6-seconds (high demand). As expected, participants with DP were worse than typical observers at matching faces, and were disproportionately impaired at matching faces relative to cars. However, the relative degree of impairment seen in the DPs did not interact with retention interval; they exhibited similar levels of impairment when matching faces with 1- and 6-second delays. Some heterogeneity is likely in any neurodevelopmental population, and DP is no different. Generally, however, these findings suggest i) that in the majority of cases, DP is associated with some degree of apperceptive impairment, and ii) STFM impairment may be relatively uncommon in this population.

5.2 Introduction

Developmental prosopagnosia[13] (DP) is a neurodevelopmental condition associated with difficulties recognising familiar faces and distinguishing unfamiliar faces, that occurs in people with normal intelligence and typical visual acuity, and in the absence of manifest brain injury (Behrmann & Avidan, 2005; Duchaine & Nakayama, 2006b;

[13] We use the term *developmental prosopagnosia* instead of *congenital prosopagnosia* to indicate the possibility that in some cases the disorder may appear during development and not necessarily from birth.

Susilo & Duchaine, 2013). Historically, the condition was thought to be rare (McConachie, 1976), but current estimates suggest that 2% of the general population may experience face recognition difficulties severe enough to disrupt their daily lives (Kennerknecht et al., 2006; Kennerknecht, Ho, & Wong, 2008). The fact that DP often runs in families suggests the condition has a genetic component (Duchaine, Germine, & Nakayama, 2007; Johnen et al., 2014; Schmalzl, Palermo, & Coltheart, 2008), a finding that accords with the broader view that face recognition ability is a heritable trait (Shakeshaft & Plomin, 2015; Wilmer et al., 2010; Zhu et al., 2010). At the neural level, studies suggest that DP is associated with reduced structural (Gomez et al., 2015; Song et al., 2015; Thomas et al., 2009) and functional (Avidan & Behrmann, 2009; Lohse et al., 2016; Rosenthal et al., 2017) connectivity within the occipito-temporal face processing network. Due to their characteristic deficits, DPs often rely on non-facial cues like voice, hairstyle, and walking gait to recognise familiar others (Cook & Biotti, 2016; Shah, Gaule, Sowden, Bird, & Cook, 2015).

5.2.1 Apperceptive characterisation

Traditionally DP has been thought of as an apperceptive form of prosopagnosia (De Renzi, Faglioni, Grossi, & Nichelli, 1991); a condition with a perceptual origin that hinders individuals' ability to encode the structure of faces (Behrmann & Avidan, 2005; Duchaine & Nakayama, 2006b; Susilo & Duchaine, 2013). Consistent with this view, many DPs exhibit difficulties distinguishing unfamiliar faces presented simultaneously (Avidan, Tanzer, & Behrmann, 2011; Biotti & Cook, 2016; Biotti, Gray, & Cook, 2017; Duchaine et al., 2007; Shah, Gaule, Gaigg, Bird, & Cook, 2015; White, Rivolta, Burton, Al-Janabi, & Palermo, 2017) or sequentially, either side of sub-second interval (Duchaine, Yovel, Butterworth, & Nakayama, 2006; Fisher, Towler, & Eimer, 2017; Le Grand et al., 2006; Yovel & Duchaine, 2006). In addition to problems matching or recognising facial identities, many DPs appear to have problems recognising facial emotion (Biotti & Cook, 2016; Burns, Martin, Chan, & Xu, 2017; Duchaine et al., 2006), facial age (Ariel & Sadeh, 1996), and facial gender (Ariel & Sadeh, 1996; Esins, Schultz, Stemper, Kennerknecht, & Bulthoff, 2016). Moreover, electrophysiological markers thought to index early face encoding (e.g. the N170 ERP component) are often atypical in cases of DP (Fisher, Towler, & Eimer, 2016; Towler, Fisher, & Eimer, 2017; Towler, Gosling, Duchaine, & Eimer, 2012; Towler, Parketny, & Eimer, 2016). This profile of deficits is consistent with a locus of impairment early in the face processing stream, before the processing of identity and other facial attributes bifurcates (Bruce & Young, 1986; De Renzi et al., 1991; Haxby, Hoffman, & Gobbini, 2000).

According to one influential apperceptive account, a failure to process faces holistically – whereby facial features are integrated into a non-decomposable whole (Farah, Wilson, Drain, & Tanaka, 1998; McKone & Yovel, 2009b; Piepers & Robbins, 2013) – may underlie the face recognition difficulties seen in DP (Avidan et al., 2011; DeGutis, Cohan, Mercado, Wilmer, & Nakayama, 2012; DeGutis, Cohan, & Nakayama, 2014; Liu & Behrmann, 2014; Palermo et al., 2011). Consistent with this view, individuals with DP are thought to be less sensitive to facial orientation (Duchaine et al., 2006; Shah, Gaule, Gaigg et al., 2015; Tree & Wilkie, 2010), and sometimes have problems distinguishing faces using feature configurations (Le Grand et al., 2006; Yovel & Duchaine, 2006). It has also been argued that some DPs show reduced susceptibility to visual illusions thought to index holistic face processing, including the part-whole (DeGutis et al., 2012) and composite face effects (Avidan et al., 2011; Liu & Behrmann, 2014; Palermo et al., 2011). Where observed, aberrant processing of configurations may extend to non-face stimuli (Avidan et al., 2011).

5.2.2 A deficit of perceptual encoding or perceptual maintenance?

The case for an apperceptive characterisation of DP is not as strong as it first appears. Several findings suggest that some DPs may encode face structure typically; for example, some individuals with DP exhibit broadly typical discrimination of unfamiliar faces presented simultaneously (Bowles et al., 2009; Dalrymple, Garrido, & Duchaine, 2014; McKone et al., 2011; Ulrich et al., 2017), and apparently normal recognition of facial emotion (Dobel, Bölte, Aicher, & Schweinberger, 2007; Humphreys, Avidan, & Behrmann, 2007; Lee, Duchaine, Wilson, & Nakayama, 2010; Palermo et al., 2011), facial age and facial gender (Chatterjee & Nakayama, 2013; DeGutis, Chatterjee, Mercado, & Nakayama, 2014). Many DPs also exhibit typical susceptibility to visual illusions thought to arise from the holistic encoding of facial structure, in particular the composite face effect (Biotti, Wu et al., 2017; Le Grand et al., 2006; Susilo et al., 2010; Ulrich et al., 2017). Notably, Biotti et al. (2017) recently described two group studies – using independent samples of 16 and 24 DPs – neither of which found evidence of reduced composite effects. These behavioural results indicate that early structural encoding may be intact in many cases of DP.

Rather than characterise DP as an apperceptive condition, several authors have raised the possibility that many cases of DP – perhaps even the majority – may be caused by impaired short-term face memory (STFM); that DPs may be able to form accurate percepts, but are unable to maintain those percepts over time (Dalrymple et al., 2014; Dalrymple & Palermo, 2016; Jackson, Counter, & Tree, 2017; Stollhoff, Jost, Elze, & Kennerknecht, 2011; Ulrich et al., 2017). A similar possibility has been suggested in the literature on autism spectrum disorder (ASD), where a systematic review concluded that a delay of a few seconds between the presentation of the target and test faces disproportionately impairs matching or recognition performance in this population (Weigelt, Koldewyn, & Kanwisher, 2012). While the suggestion that faces may benefit from domain-specific memory processing is relatively new, the implied dissociation between perceptual processes responsible for face encoding, and memory processes responsible for maintaining face representations, is consistent with evidence that face memory follows a different developmental trajectory relative to perceptual memory for other objects (Weigelt et al., 2013).

Consistent with the possibility that DP may be caused by aberrant STFM, many cases of DP have been described (Bowles et al., 2009; Dalrymple et al., 2014; McKone et al., 2011; Ulrich et al., 2017) who exhibit impaired performance on diagnostic tests with a memory component such as the Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006a), but perform within the typical range on tests with a minimal memory component such as the Cambridge Face Perception Test (CFPT; Duchaine et al., 2007). When DPs are required to retain faces in memory for brief periods, functional magnetic resonance imaging (fMRI) reveals wider activation in prefrontal regions implicated in working memory, relative to controls (Avidan, Hasson, Malach, & Behrmann, 2005), suggesting that percept retention may be effortful. Similarly, where observed, neural differences in DP are sometimes more pronounced in anterior (extended) regions of the face processing network, than in posterior (core) areas thought to be responsible for early structural encoding (Avidan et al., 2014).

5.2.3 Present study

The present study sought to examine whether DP is best characterised as i) a disorder of STFM, where these individuals initially form accurate perceptual descriptions of faces, but struggle to maintain these representations over time; or ii) as an apperceptive condition, where face recognition difficulties arise from poor encoding of face structure. In Experiment 1, we examined the performance of a large sample of DPs (N = 72) on the CFPT. In Experiment 2, we compared the face-matching ability of a subset of these DPs (N = 16) following 1- and 6-second retention intervals. Consistent with an apperceptive characterisation, we find that DPs not only perform poorly on the CFPT at the group-level, but show signs of a shifted distribution (Experiment 1) and show similar levels of matching impairment relative to controls at short and long retention intervals (Experiment 2).

5.3 Can DPs discriminate simultaneously presented faces?

Several studies have found that small samples of DPs make more errors on the CFPT than groups of matched TD controls (e.g., Shah, Gaule, Gaigg et al., 2015). As has been noted elsewhere, however, individual DPs sometimes fail to exhibit significant impairment at the single-case level – i.e., they score within 2 SDs of mean typical performance on this task (e.g., Bowles et al., 2009; Ulrich et al., 2017). Consequently, it is possible that group differences in CFPT performance, where observed, are driven by a handful of DPs with apperceptive impairments who produce outlying error scores.

A second possibility is that apperceptive deficits are widespread in the DP population, but that the CFPT does not always reveal clear evidence of impairment. The distribution of CFPT scores produced by DPs and controls might be expected to overlap to some degree given that the CFPT is known to yield noisy estimates of perceptual ability (Bowles et al., 2009). The sequential sorting task employed by the CFPT may also render it more susceptible to compensatory strategies such as moving closer to the display or seeking trivial details that distinguish faces. By adopting these strategies, DPs with apperceptive problems may sometimes achieve CFPT scores within the normal range.

It is difficult to distinguish these rival views by examining the scores from single cases of DP. However, these accounts make different predictions about the distributions of CFPT scores that should be seen in DP samples. According to the apperceptive subset view, the distribution of CFPT scores produced by TDs and DPs should differ only in terms of the lower tail of their distributions; i.e. the DP distribution should be identical to that of controls, with the exception of some outlying individuals at the lower tail who make a disproportionate number of errors. According to the shifted distribution view, however, evidence of impairment should be seen in both the upper and lower tail of the DP distribution – not only should the worst DPs make more errors than the worst controls, but the best DPs should be unable to achieve scores comparable with the best controls. We sought to test these rival predictions by examining the distribution of CFPT scores produced by a large sample of DPs and controls.

5.3.1 Methods

5.3.1.1 Participants

In total, 126 adults participated in Experiment 1, 72 with DP (30 males; $M_{age} = 42.34$ years, $SD_{age} = 11.77$ years) and 54 typically developed (TD) controls (23 males; $M_{age} = 39.20$ years, $SD_{age} = 13.36$ years). Neither participant age [$t(124) = 1.400$, $p = .164$] nor proportion of males [$X^2(1) = .01$, $p = .920$] differed significantly between the two groups. Ethical approval was granted by the local ethics committee. The study was conducted in line with the ethical guidelines provided by the 6th (2008) Declaration of Helsinki. All participants provided informed consent and were debriefed after the experimental procedure (i.e., the aims and rationale of the study were explained).

5.3.1.2 Diagnostic testing

DP participants were recruited through www.troublewithfaces.org and reported lifelong face recognition difficulties in the absence of brain injury or psychiatric disorder (e.g., ASD, schizophrenia). Diagnostic decisions were based primarily on participants' scores on the Twenty-Item Prosopagnosia Index (PI20; Gray, Bird, & Cook, 2017; Shah, Gaule, Sowden et al., 2015) and the CFMT (Duchaine & Nakayama, 2006a). As expected, the groups differed significantly in their PI20 [$t(124) = 29.156$, $p < .001$] and CFMT scores [$t(124) = 19.357$, $p < .001$]. Summary statistics for both groups are provided in Table 5.1 and detailed diagnostic information for each DP is provided as supplementary material. The development of standardised diagnostic criteria for DP still appears some way off (Barton & Corrow, 2016; Dalrymple & Palermo, 2016; Shah, Gaule, Sowden et al., 2015). However, the use of convergent self-report evidence and scores on objective, computer-based tasks may be a particularly effective approach to the identification and classification of DP; for example, less than 1.5% of the general population score below 65% on the CFMT *and* more than 65 on the PI20 (see Gray et al., 2017).

	PI20				CFMT (%)			
	M	SD	Min	Max	M	SD	Min	Max
Typical controls (N = 54)	37.96	9.09	20	56	84.98	8.92	65.28	100.00
Prosopagnosics (N = 72)	80.94	7.45	59	97	56.00	7.83	34.72	68.02

Table 5.1. Diagnostic information for the DP and TD samples employed in Experiment 1.

5.3.1.3 Stimuli and procedure

The CFPT assesses face perception ability in such a way as to minimize the memory demand on participants. Trials present a target face and a series of six faces that resemble the target to varying degrees (Figure 5.1a). Participants have 60 seconds to sort the six faces in order of target-face similarity. Eight trials present the target and test faces upright, eight present the faces inverted. Trials are scored by calculating deviations from the correct order. Participants were given the option of completing the CFPT using a trackpad or mouse – whichever they found easier to use. All participants were tested individually at the *troublewithfaces.org* lab, under tightly controlled conditions, in return for a small honorarium.

5.3.2 Results and discussion

Participants' scores on the CFPT were analysed using ANOVA with Orientation (upright, inverted) as a within-subjects factor, and Group (DP, TD) as a between-subjects factor (Figure 5.1b). The analysis revealed main effects of Orientation [$F(1,124) = 370.862$, $p < .001$, $\eta_p^2 = .749$] and Group [$F(1,124) = 10.650$, $p < .001$, $\eta_p^2 = .079$] with more errors seen when faces were inverted and less precise sorting exhibited by the DP group. However, it also yielded a significant Orientation × Group interaction [$F(1,124) = 251.784$, $p < .001$, $\eta_p^2 = .670$]. The DPs ($M = 50.64$, $SD = 15.35$) made disproportionately more errors than the controls ($M = 29.41$, $SD = 9.35$) on the upright trials of the CFPT [$t(124) = 9.601$, $p < .001$]. However, the DPs ($M = 69.86$, $SD = 13.11$) also made more errors than controls ($M = 63.37$, $SD = 15.74$) on the inverted trials [$t(124) = 2.522$, $p = .013$].

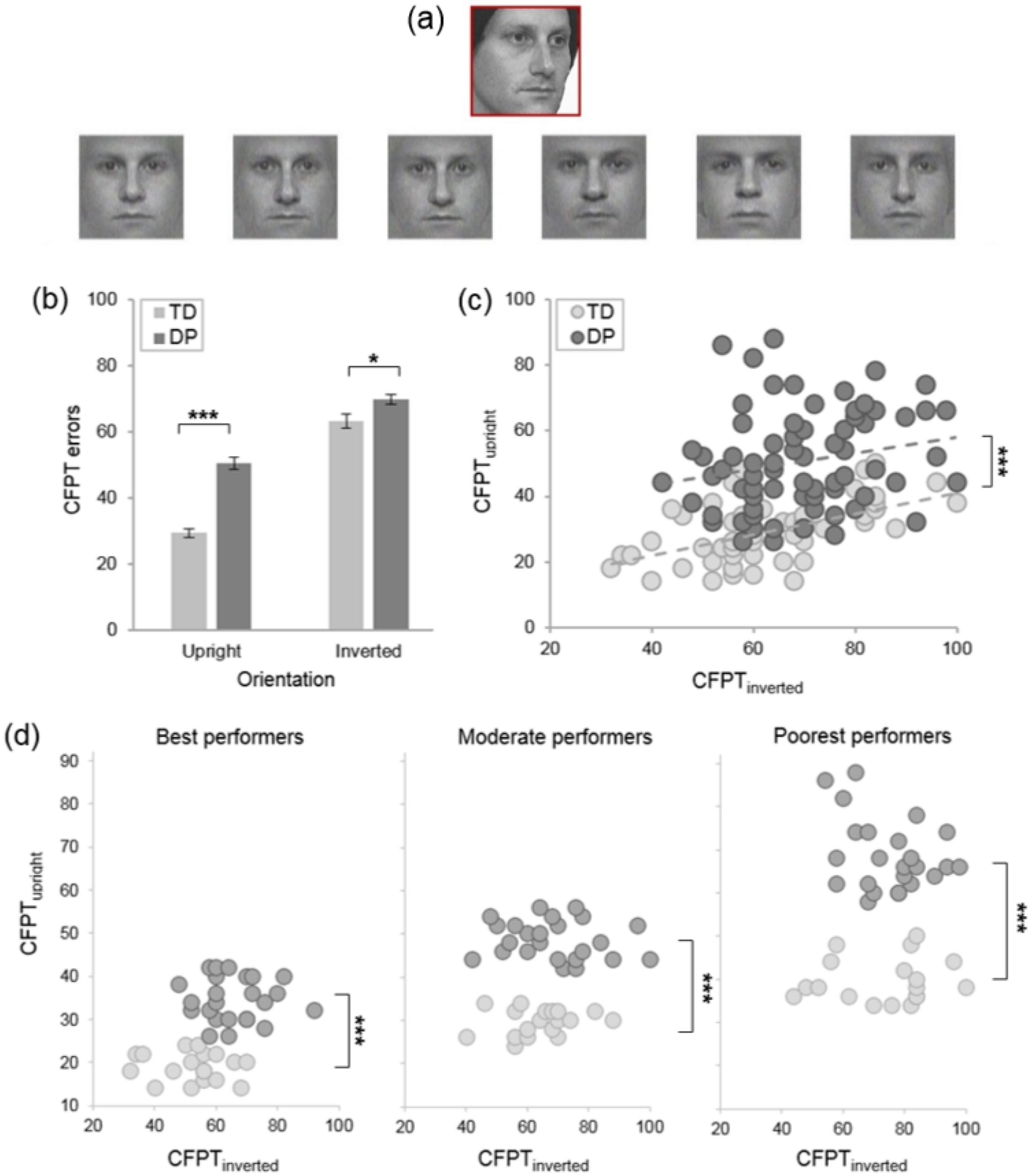

Figure 5.1. (a) Each trial of the Cambridge Face Perception Test presents simultaneously a target face and a series of six faces that resemble the target to varying degrees. Participants have 60 secs to sort the six items in order of target-face similarity. (b) Mean performance of the TD (N = 54) and DP (N = 72) groups in the upright and inverted conditions of the CFPT. (c) Each participant's performance on the upright trials plotted against their inverted performance. (d) Analysis of the best, moderate, and worst performers from the sample indicated that the entire distribution of DP scores was shifted relative to the distribution of TD scores. *denotes $p < .05$; ** denotes $p < .01$; *** denotes $p <.001$. Error bars denote ±1SEM.

While the scores of the TD observers were more sensitive to the orientation manipulation (upright vs. inverted presentation), this may simply reflect the fact that the DPs are closer to floor performance in the upright condition (also see Klargaard, Starrfelt, & Gerlach, 2018). In addition to the group difference (DPs < TDs) seen for the inverted trials of the CFPT, we found evidence of correlation between observers' scores on the upright and inverted trials ($N = 126$, $r = .370$, $p <.001$; Figure 5.1c). These findings accord with the view that the visual processing of upright and inverted faces may differ quantitatively (Gold, Mundy, & Tjan, 2012; Murphy & Cook, 2017; Sekuler, Gaspar, Gold, & Bennett, 2004; Susilo, Rezlescu, & Duchaine, 2013), not qualitatively (McKone & Yovel, 2009a; Rossion, 2008).

Next, we ranked the TD ($N = 54$) and DP ($N = 72$) samples based on individuals' performance on the upright trials of the CFPT and split each distribution into thirds: best performing TDs ($N = 18$, $M_{age} = 37.83$) and DPs ($N = 24$, $M_{age} = 43.08$), intermediate TDs ($N = 18$, $M_{age} = 37.94$) and DPs ($N = 24$, $M_{age} = 41.38$), and poorest performing TDs ($N = 18$, $M_{age} = 41.83$) and DPs ($N = 24$, $M_{age} = 42.58$). Strikingly, the TD controls outperformed the DPs at each level of their respective distributions: best performers [$t(40) = 11.304$, $p < .001$], intermediate performers [$t(40) = 15.596$, $p < .001$], poorest performers [$t(40) = 13.051$, $p < .001$] (Figure 5.1d). This pattern argues against the view that group differences on the CFPT reflect the presence of a few individual DPs with an apperceptive deficit. Instead, these results favour the view that the entire distribution of CFPT scores produced by the DPs is shifted relative to that of TD controls.

To illustrate how apperceptive impairment in DP might produce a shifted distribution of CFPT scores similar to that observed, we have shown the effects of inflating each typical observer's CFPT error score by 90% (Figure 5.2). This inflation coefficient is akin to the application of a hypothetical apperceptive deficit that increases the number of sorting errors made. As can be seen, this simple model provides a reasonable approximation of the distribution of scores seen in the DP sample. To be clear, we are not claiming that DP always impairs perceptual encoding of faces by 90%; rather, we present this demonstration as a proof-of-principle. We merely seek to illustrate that an apperceptive deficit might plausibly produce the distribution of CFPT scores seen in our DP sample.

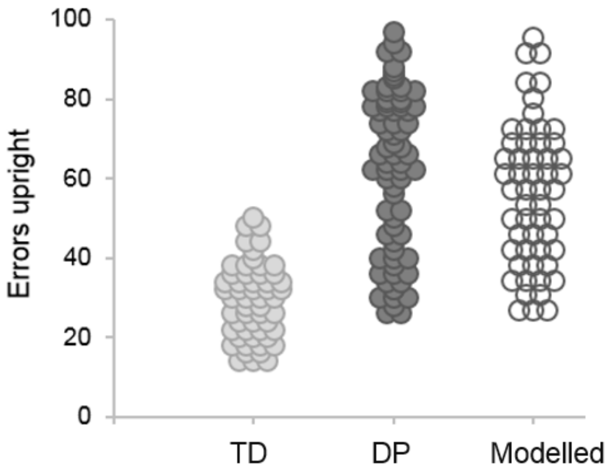

Figure 5.2. To illustrate the shifted distribution account, we modelled the effects of inflating each typical observer's error score by 90%, akin to the application of a hypothetic apperceptive deficit. This simple model provides a reasonable approximation of the range of scores seen in the DP population.

In light of its poor psychometric properties, researchers are discouraged from using the CFPT for diagnostic purposes (Bowles et al., 2009). In the present study, decisions to classify people as DP were therefore based principally on individuals' PI20 and CFMT scores. We note, however, that observers' CFPT scores correlated closely both with their CFMT (N = 126, r = -.665, p <.001) and PI20 scores (N = 126, r = .662, p <.001). Contrary to the prevailing view that the CFMT is a test of 'face memory', this finding suggests that the individual differences revealed by the PI20 and CFMT may in large part reflect individuals' ability to encode face structure.

5.4 Do face matching deficits seen in DP increase as a function of retention interval?

It has been proposed that many DPs – possibly the majority – experience deficits of STFM but exhibit intact encoding of face structure (Dalrymple et al., 2014; Dalrymple & Palermo, 2016; Jackson, Counter, & Tree, 2017; Stollhoff, Jost, Elze, & Kennerknecht, 2011; Ulrich et al., 2017). Experiments that directly test whether the face matching and face recognition deficits seen in DP are sensitive to memory load are therefore particularly important. For this reason, we sought to revisit a finding described by Shah, Gaule, Gaigg, Bird, and Cook (2015). This previous study utilised a delayed match-to-sample task whereby participants were required to identify a target stimulus from a test display of four items (target plus three lures). Memory demands were manipulated by varying the delay between the presentation of the target and the test array. This approach is useful as it allows systematic manipulation of the memory component of the task, but ensures the perceptual demands – associated with the encoding of target and test items – are held constant (Shah, Gaule, Gaigg et al., 2015). If DP is associated with impaired STFM, disproportionate impairment should be seen after longer retention intervals, relative to shorter retention intervals. Contrary to this prediction, however, Shah and colleagues found that their DP sample ($N = 15$) exhibited comparable deficits at short (2-second) and long (8-second) intervals.

In the original study described by Shah et al. (2015) the same images were used to present items in the study and test phases. Consequently, targets were always seen from the same frontal viewpoint. In the present study, we examined observers' ability to match items viewed from the same frontal perspective (constant-viewpoint matching), and across a viewpoint disparity of 45° (different-viewpoint matching). While constant- and different-viewpoint matching appear similar, they may differ substantially in their perceptual and mnemonic demands. First, observers sometimes match unfamiliar faces using superficial pictorial cues (Hancock, Bruce, & Burton, 2000; Megreya & Burton, 2006). Because rotation introduces substantial disparity between target and test images, different-viewpoint matching is less susceptible to this strategy than constant-viewpoint matching (Longmore, Liu, & Young, 2008). Instead, different-viewpoint matching is thought to tax observers' ability to form and maintain a view-invariant structural description (Bruce & Young, 1986; Marr & Nishihara, 1978). Second, a particular type of short-term memory – visual working memory (Baddeley, 1992, 1993, 2010) – has been hypothesised that supports the rotation and manipulation of percepts. While constant- and different-viewpoint face matching both tap some short-term memory processes, different-viewpoint matching places greater demands on visual working memory. In light of their different mnemonic demands, these two tasks may behave differently as a function of retention interval, and be differentially affected in DP.

On half the trials, we used a retention interval of 1-second (low demand); on half the trials, we used a retention interval of 6-seconds (high demand). The short interval used in this study (1-second) is shorter than that employed previously (2-seconds; Shah, Gaule, Gaigg et al., 2015), thereby reducing further the memory demands in the low demand condition. We recognize, however, that the retention of percepts for 1 second still represents a memory demand. Crucially, our aim in the short interval condition was to minimise, not to eliminate, the memory demands of the matching task[14]. Participants' face matching ability was compared to that seen with cars to determine if deficits, where observed, were face-specific, or whether they extended to a non-face object category.

5.4.1 Methods

5.4.1.1 Participants

A subset of the DPs (N = 16, 6 males; M_{age} = 41.50 years, SD_{age} = 12.58 years) and TDs (N = 22, 9 males; M_{age} = 38.23 years, SD_{age} = 13.39 years) from Experiment 1 completed Experiment 2 (Table 5.2). None of the DPs were included in the sample described by Shah et al. (2015). Neither participant age [$t(36) = .763$, $p = .451$] nor proportion of males [$X^2(1) = .045$, $p = .551$] differed significantly between the two groups. Ethical approval was granted by the local ethics committee. The study was conducted in line with the ethical guidelines provided by the 6th (2008) Declaration of Helsinki. All participants provided informed consent and were fully debriefed after the experimental procedure.

As expected, the TD controls (M_{CFMT} = 85.1%, SD_{CFMT} = 10.2%; M_{PI20} = 39.0, SD_{PI20} = 9.0) differed significantly from the DPs (M_{CFMT} = 55.9%, SD_{CFMT} = 7.9%; M_{PI20} = 79.9, SD_{PI20} = 8.1) in their PI20 [$t(36) = 14.390$, $p < .001$] and CFMT [$t(36) = 9.574$, $p < .001$] scores. In addition to the CFMT, CFPT and the PI20, all participants in Experiment 2 (DPs and controls) also completed the Cambridge Car Memory Test (CCMT; Dennett et al., 2011) to measure their non-face object recognition ability. The TD controls (M = 73.9%, SD = 12.8%) and the DPs (M = 63.5%, SD = 8.4%) differed significantly in terms of their performance on the CCMT [$t(35) = 2.837$, $p = .008$]. All participants were also screened for colour blindness using Ishihara's Tests for Colour-Blindness (Ishihara, 1993).

[14] The key strength of this paradigm is that it allows the manipulation of memory demands in a way that leaves the perceptual demands of the task unaltered. Having a no interval condition (i.e., where the target is presented alongside the array of 4 test items) would have violated this logic. Although presenting the 5 faces simultaneously would further reduce the memory demands, this reduction would be confounded with an increase in perceptual and attentional demands.

Participant	Age	PI20	CFMT %	CFPT Upright (Errors)	CFPT inverted (Errors)	PI20 z-scores	CFMT z-scores	CFPT z-scores
F1	21	59	63.89	30	64	-2.31	-2.36	-0.06
F2	48	85	63.89	60	70	-5.17	-2.36	-3.27
F3	22	80	40.28	66	98	-4.62	-5.01	-3.91
F4	66	79	61.11	40	70	-4.51	-2.68	-1.13
F5	48	78	58.33	26	64	-4.40	-2.99	0.36
F6	25	70	44.44	74	68	-3.52	-4.54	-4.77
F7	38	73	59.72	48	64	-3.85	-2.83	-1.99
F8	40	85	59.72	34	75	-5.17	-2.83	-0.49
F9	53	84	52.78	62	58	-5.06	-3.61	-3.49
F10	38	90	50.00	66	80	-5.72	-3.92	-3.91
M1	30	85	59.72	48	84	-5.17	-2.83	-1.99
M2	47	77	68.06	42	72	-4.29	-1.90	-1.35
M3	52	94	55.56	88	64	-6.17	-3.30	-6.27
M4	55	80	50.00	44	84	-4.62	-3.92	-1.56
M5	41	80	61.11	44	70	-4.62	-2.68	-1.56
M6	40	79	45.83	34	76	-4.51	-4.39	-0.49
DP mean	41.5	79.87	55.90	50.37	72.56			
DP SD	12.6	8.13	7.89	17.33	0.09			
TD mean	39.2	37.96	84.98	29.41	63.37			
TD SD	13.4	9.09	8.92	9.35	15.74			

Table 5.2. Scores of each developmental prosopagnosic on the 20 Item Prosopagnosia Index (PI20), the Cambridge Face Memory Test (CFMT), The Cambridge Face Perception Test (CFPT). The z-scores provided for the CFPT are based on performance in the upright condition. *Note*. The prosopagnosics' scores on the diagnostic procedures were compared with the group of 54 controls described in Experiment 1 (23 males). All but one of the DPs scored at least two standard deviations below the comparison average on the PI20 and the CFMT. The case for including this individual (M2) in our DP sample was bolstered by his poor score (< 3 SDs below the mean) on a UK variant of the Famous Face Recognition Task.

5.4.1.2 Stimuli and procedure

Each category (faces, cars) comprised 50 exemplars. Both categories were further organised into 5 subsets of 10 exemplars based on approximate similarity. Cars were sorted into subsets based on their size and class (e.g. Saloons / Sedans / SUVs). Faces were sorted based on aspect-ratio, pigmentation, and eye-brow colour. Each exemplar was depicted twice: once in frontal view, once in 3/4 view. When viewed at 57 cm, the faces subtended 6° of visual angle vertically; the cars subtended 3.5° vertically. Face stimuli (male Caucasian faces) were created using FaceGen Modeller Version 3.3 (Singular Inversions Inc.). Car stimuli were generated through www.3dtuning.com.

The structure of the delayed matching task is shown in Figure 5.3. Each trial started with a fixation point (750 ms) on a blank screen. A single target stimulus was then presented centrally for 400 ms. Targets were always shown in frontal view. A given facial identity or car model could appear as a target only once in each viewing condition.

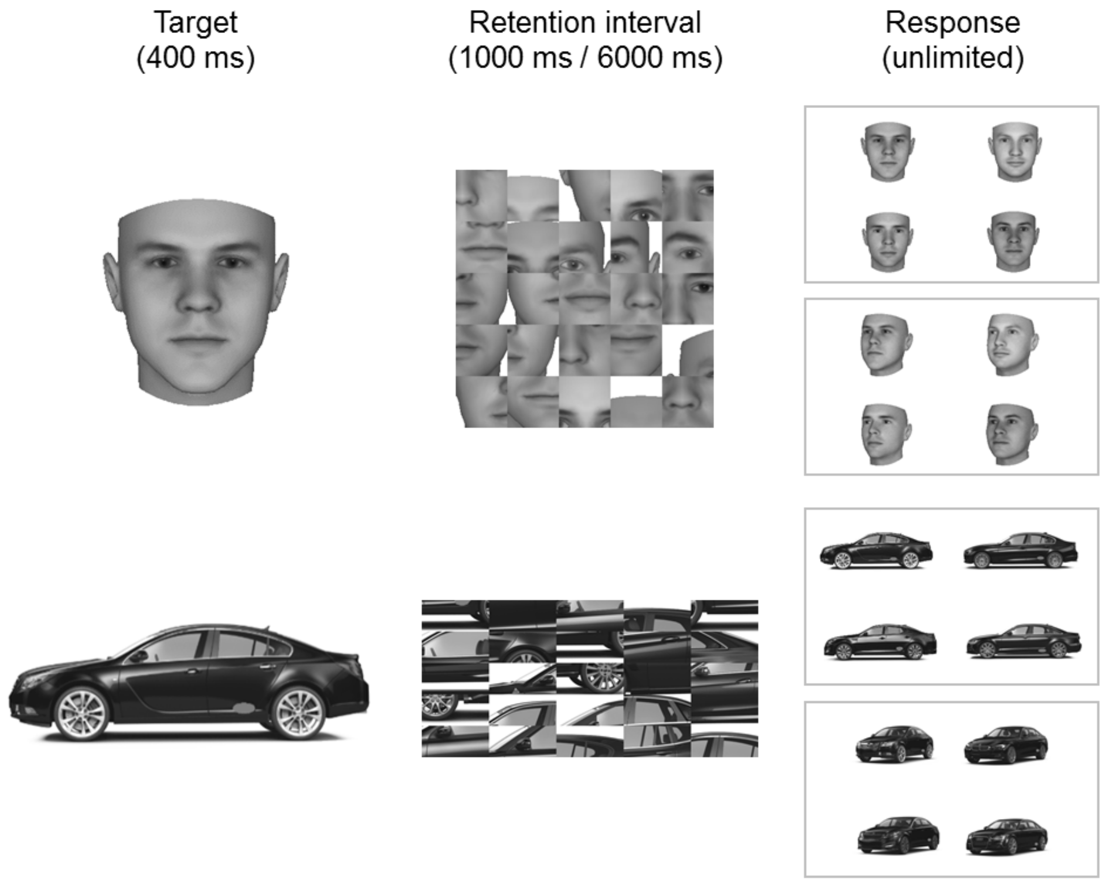

Figure 5.3. Illustration of the stimuli and procedure employed in our delayed matching task.

In all other respects, the choice of target was randomly determined by the experimental program. The offset of the target was followed by a retention interval during which a mask image was presented. The mask was constructed by recombining regions cropped from other target images from the same category. An array of four test items followed the retention interval. The array comprised the target and three lures selected at random from the same within-category subset. On half of the trials, test stimuli were presented in frontal view (here, the target and test stimuli were shown from the same viewpoint). On the remaining trials, test stimuli were presented in 3/4 view (here, the target and test stimuli were shown from different viewpoints). Test arrays were visible until a keypress response was registered. Participants were asked to respond with speed and accuracy.

The factorial combination of stimulus type (faces, cars), retention interval (short, long), viewpoint (frontal, 3/4) yielded eight types of trial, which were randomly interleaved. There were 20 trials of each type, yielding 160 trials in total. Given the large number of face and car stimuli required by the procedure it was necessary to recycle stimuli from each pool of 50 items. Some stimulus items therefore appeared multiple times across the procedure, either as targets or lures. Six practice trials preceded the experiment. No feedback was provided during the procedure. The task lasted approximately 45 minutes and included three short breaks. The task was programmed in MATLAB using the Psychophysics Toolbox (Brainard, 1997; Pelli, 1997).

5.4.2 Results and discussion

5.4.2.1 Group analyses

Matching accuracy (Figure 5.4a) was analysed using ANOVA with Stimulus Type (faces, cars), Retention Interval (short, long), and Viewpoint (frontal, 3/4) as within-subjects factors, and Group (DP, TD) as a between-subjects factor. The analysis revealed significant main effects of Viewpoint [$F(1,36) = 52.44$, $p < .001$, $\eta_p^2 = .59$] and Retention Interval [$F(1,36) = 48.41$, $p < .001$, $\eta_p^2 = .57$], whereby a change of viewpoint and a longer retention interval were associated with poorer matching accuracy, respectively. However, there was no main effect of Stimulus Type [$F(1,36) = 3.16$, $p = .084$, $\eta_p^2 = .081$], nor did we see a Retention Interval × Viewpoint interaction [$F(1,36) = .001$, $p = .982$, $\eta_p^2 = .000$]. As expected, we observed a significant effect of Group [$F(1,36) = 10.35$, $p = .003$, $\eta_p^2 = .22$], as well as a significant Group × Stimulus Type interaction [$F(1,36) = 6.11$, $p = .018$, $\eta_p^2 = .145$], indicating that the DPs were worse overall, but disproportionately impaired at face matching. Crucially, however, no further interactions with Group were seen on the face (all Fs < .45, ps > .50) or car trials (all Fs < .60, ps > .45). When matching accuracy for cars and faces was analysed in separate ANOVAs, we observed a significant effect of Group for face trials [$F(1,36) = 15.072$, $p < .001$, $\eta_p^2 = .295$], but not for car trials [$F(1,36) = 1.869$, $p = .180$, $\eta_p^2 = .049$].

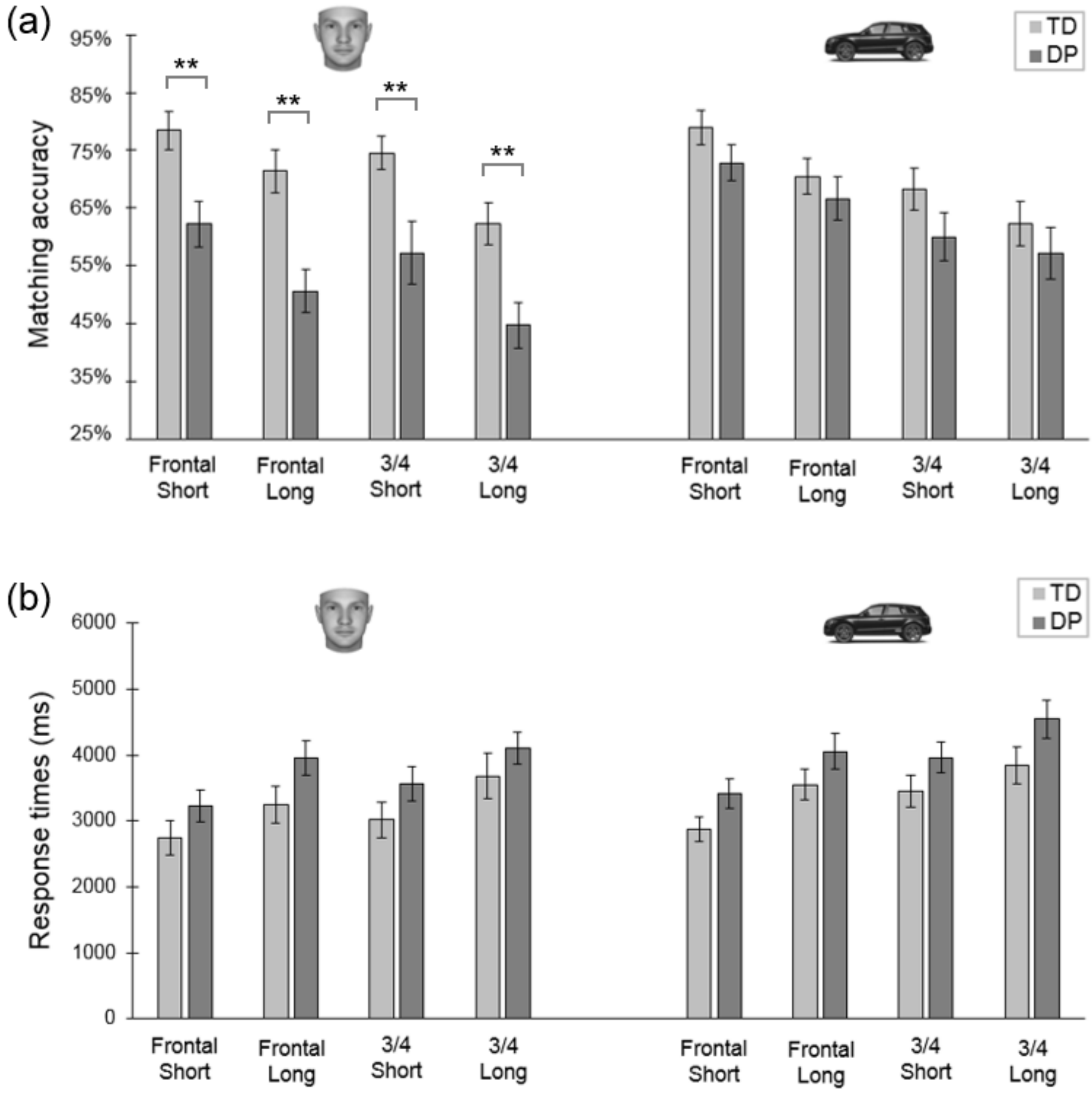

Figure 5.4. Mean (a) accuracy and (b) response times for the two groups on the delayed-matching task. Performance is broken down by Viewpoint (frontal, 3/4) and Retention Interval (short, long). Simple contrasts were non-significant unless otherwise indicated. *denotes $p < .05$; ** denotes $p < .01$; *** denotes $p < .001$. Error bars denote ±1SEM.

To evaluate the effects of the two within-subjects manipulations we computed measures expressing each observer's viewpoint effect (same-viewpoint matching accuracy – different-viewpoint matching accuracy) and their retention interval effect (short-interval matching accuracy – long-interval matching accuracy). The retention interval effects of the TDs (M = 9.7%, SD = 12.1%) and the DPs (M = 12.0%, SD = 8.7%) did not differ [$t(36) = .689$, $p = .495$] and all DPs exhibited retention interval effects within 2 SDs of the typical mean. Similarly, the viewpoint effects of the TDs (M = 6.5%, SD = 8.5%) and DPs (M = 5.5%, SD = 11.1%) did not differ [$t(36) = .319$, $p = .752$] and all DPs exhibited viewpoint effects within 2 SDs of the typical mean.

We also analysed participants' response times (Figure 5.4b) using ANOVA with Stimulus Type (faces, cars), Retention Interval (short, long), and Viewpoint (frontal, 3/4) as within-subjects factors, and Group (DP, TD) as a between-subjects factor. The analysis revealed main effects of Stimulus Type [$F(1,36) = 5.33$, $p = .027$, $\eta_p^2 = .129$], Viewpoint [$F(1,36) = 52.48$, $p < .001$, $\eta_p^2 = .593$], and Retention Interval [$F(1,36) = 98.94$, $p < .001$, $\eta_p^2 = .733$]. Overall, participants responded faster on face trials than on car trials, were faster when identifying frontal views of targets than 3/4 views, and were faster following short retention intervals than long retention intervals. The analysis revealed no main effect of Group [$F(1,36) = 2.65$, $p = .112$, $\eta_p^2 = .069$], nor a Group × Stimulus Type interaction [$F(1,36) = .01$, $p = .931$, $\eta_p^2 = .000$]. No further interactions with Group were seen on the face (all Fs < .75, ps > .39) or car trials (all Fs < .90, ps > .35). When analysed in separate ANOVAs, the response times of the DPs and the TD controls did not differ significantly on either face [$F(1,36) = 2.012$, $p = .165$, $\eta_p^2 = .053$] or car trials [$F(1,36) = 2.845$, $p = .100$, $\eta_p^2 = .073$].

In both the accuracy and response time analyses, Group failed to interact significantly with either Retention Interval or Viewpoint. In order to evaluate the strength of evidence provided by these null results, we subjected these interaction effects to Bayesian analysis in JASP (JASP-Team, 2018) with default prior width. Analysis of the Group × Retention Interval interaction seen in the accuracy data indicated that the observed data were 2.64 times more likely to occur under the null model, than under an alternative. The observed Group × Viewpoint interaction was 3.02 times more likely to occur under the null model, than under an alternative. Analysis of the Group × Retention Interval interaction seen in the response time data indicated that the observed data were 3.05 times more likely to occur under the null model, than under an alternative. The observed Group × Viewpoint interaction was 2.90 times more likely to occur under the null model, than under an alternative.

5.4.2.2 Correlational analyses

The group analyses described above reveal comparable deficits at short and long retention intervals, replicating the findings of Shah and colleagues (2015). The insensitivity of the DP deficit to retention interval suggests that poor perceptual encoding – not aberrant STFM – may be responsible for the face recognition problems seen in this population. If this view is correct, performance in our matching task should correlate with participants' scores on the CFPT – a measure of face encoding ability (Experiment 1). Consistent with this prediction, overall matching accuracy (i.e., collapsing across viewpoint and interval conditions) correlated closely with performance on the CFPT (r = -.743, p < .001; Figure 5.5a). Highly significant correlations were seen between CFPT scores and face matching accuracy in all conditions (Table 5.3). In the combined sample, CFPT scores were also correlated with overall car matching accuracy (r = -.326, p = .046), however this correlation was significantly weaker than that seen between the CFPT and face matching [z = 2.59, p < .001]. As expected, observers' matching ability at short intervals correlated closely with their performance at longer intervals for both faces (r = .810, p < .001) and cars (r = .689, p < .001; Figure 5.5b).

	Same view Short interval	**Same view Long interval**	**Different view Short interval**	**Different view Long interval**
Face accuracy	-0.696 (.000)	-0.703 (.000)	-0.613 (.000)	-0.621 (.000)
Face RTs	0.347 (.033)	0.411 (.010)	0.352 (.030)	0.273 (.097)
Car accuracy	-0.337 (.038)	-0.286 (.082)	-0.334 (.040)	-0.143 (.391)
Car RTs	0.280 (.089)	0.222 (.180)	0.226 (.173)	0.173 (.298)

Table 5.3. Correlations seen between participants' scores on the $\text{CFPT}_{\text{upright}}$ and their accuracy and response time (RT) performance in each of the matching conditions. Associated p values are reported in brackets.

The group analyses also indicate that, relative to controls, DPs showed similar levels of impairment in both the constant-viewpoint and different-viewpoint matching conditions. This finding suggests that observers may be using the same perceptual strategy to achieve both types of matching. Consistent with this possibility, we found that observers' (N = 38) different-viewpoint face matching ability correlated closely with their constant-viewpoint face matching ability (r = .846, p <.001). A similar relationship was seen for cars (r = .743, p < .001), as shown in Figure 5.5c. Some correlation was also seen between same-viewpoint face matching and same-viewpoint car matching (r = .376, p = .02), and between different-viewpoint face matching and different-viewpoint car matching (r = .387, p = .016). However, both between-class correlations were significantly lower than the within-class correlations seen for faces (z = 3.54, p <.001; z = 3.49, p < .001) and cars (z = 2.35, p = .019; z = 2.30, p = .021). Having collapsed across viewing angle and retention interval, a moderate correlation

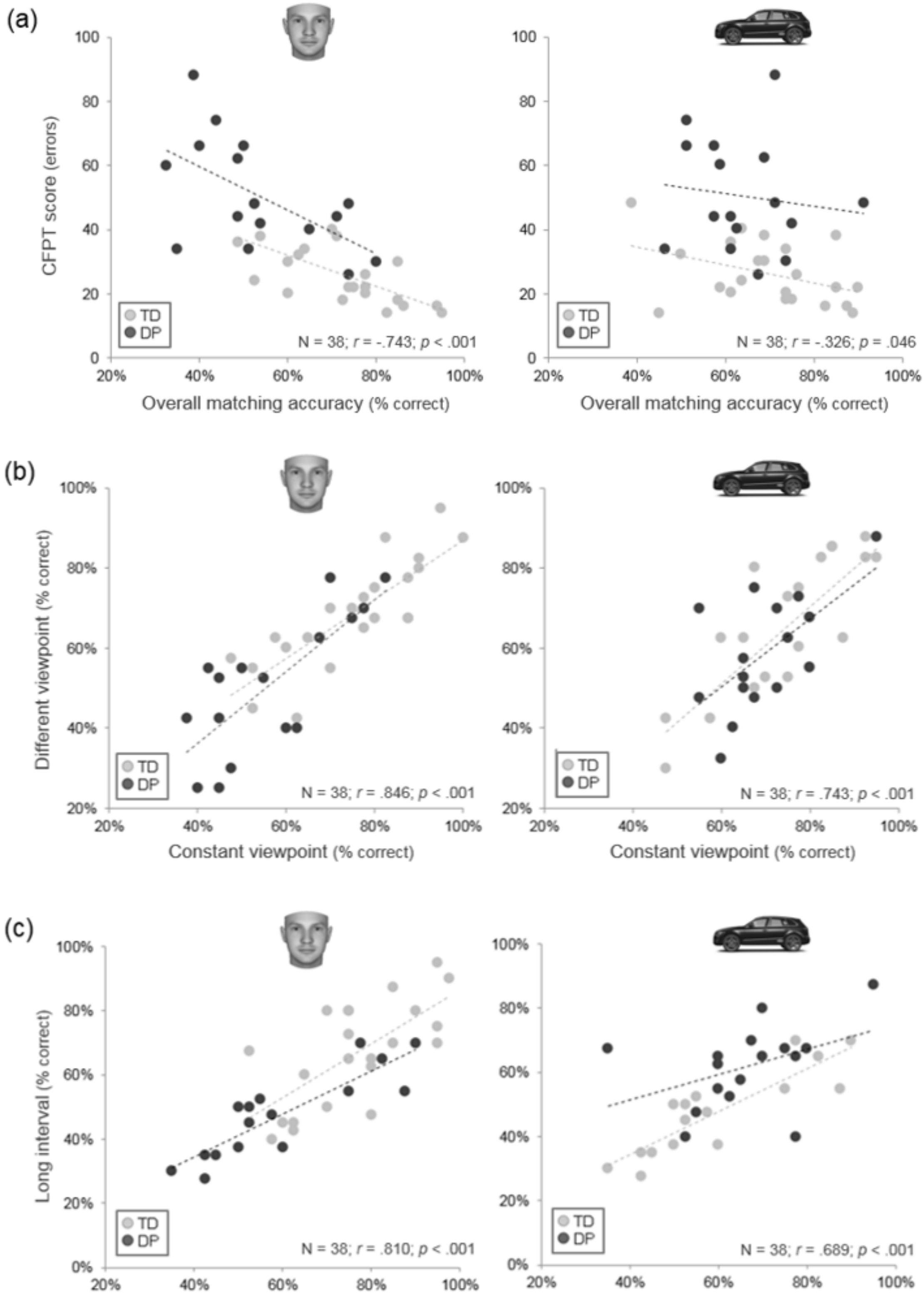

Figure 5.5. (a) The relationship between observers' CFPT scores and their face (left) and car (right) matching ability. (b) Scatterplots depicting the relationship between constant- and different-viewpoint matching for faces (left) and cars (right). (c) Scatterplots depicting the relationship between long- and short-interval matching accuracy for faces (left) and cars (right).

was seen between observers' face and car matching in the combined sample (r = .437, p = .006).

The correlations seen between observers' short- and long-interval matching accuracy (faces: r = .810; cars: r = .689), and between their constant- and different-viewpoint matching accuracy (faces: r = .846; cars: r = .743) indicate that the task has good reliability. Reassuringly, matching accuracy for faces and cars also correlated with our other measures of face and car processing (Table 5.4). In particular, strong correlations were observed in the combined sample between participants' face matching accuracy and their scores on the CFMT ($r = .67$, $p < .001$), but not the CCMT ($r = .25$, $p = .14$). Conversely, car matching accuracy correlated with scores on the CCMT ($r = .55$, $p < .001$), but not the CFMT ($r = .29$, $p = .082$).

	Faces		**Cars**	
	Accuracy	*RTs*	*Accuracy*	*RTs*
CFMT	0.671 (.000)	-0.192 (.247)	0.286 (.082)	-0.120 (.474)
CFPT	-0.743 (.000)	0.359 (.027)	-0.326 (.046)	0.234 (.158)
PI20	-0.628 (.000)	0.345 (.034)	-0.283 (.085)	0.384 (.017)
CCMT	0.246 (.141)	-0.319 (.055)	0.546 (.000)	-0.199 (.239)

Table 5.4. Correlations between participants' scores on the Cambridge Face Memory Test (CFMT), the upright condition of the Cambridge Face Perception Test (CFPT), the 20-item Prosopagnosia Index (PI20), the Cambridge Car Memory test (CCMT), and face and car matching performance. Associated p-values are shown in parentheses. Accuracy and response time (RT) measures have been collapsed across viewing conditions.

5.5 General discussion

The present study sought to examine whether DP is best characterised as i) a disorder of STFM, where these individuals initially form accurate perceptual descriptions of faces, but struggle to maintain these representations over time; or ii) as an apperceptive condition, where face recognition difficulties arise from poor encoding of face structure. First, we had 72 DPs and 54 TD controls complete the CFPT, a task that measures face perception in a way that minimises participants' memory load and is therefore thought to index structural encoding ability. We found that the DPs were clearly impaired at the group level, and showed evidence of a shifted distribution. Next, a subset of these participants (16 DPs and 22 TD controls) completed a delayed match-to-sample task for faces and cars, with a retention interval of 1-second (low demand) or 6-seconds (high demand). As expected, participants with DP were worse than TD controls at face matching. Interestingly, however, the relative degree of impairment exhibited by the DPs did not interact with retention interval. Observers' scores on the CFMT (Experiment 1) and the delayed matching task (Experiment 2) – both ostensibly measures of 'face memory' – were found to correlate strongly with their performance scores on the CFPT.

5.5.1 Evidence for an apperceptive characterisation

Some heterogeneity is likely in any neurodevelopmental population, and DP is no different. Generally, however, our results support an apperceptive characterisation of this condition. In our first experiment, we found that a large sample of DPs exhibited clear impairment on the CFPT when analysed at the group level. Despite the limitations of the CFPT – in particular, its psychometric properties make it poorly suited for quantifying individual differences (Bowles et al., 2009) – we found evidence that the distribution of CFPT scores seen in the DP sample was shifted relative to that of typical controls. Given that the CFPT is thought to measure individuals' ability to encode accurately the structure of faces, these results suggest i) that in the majority of cases, DP is associated with some degree of apperceptive impairment; and ii) that cases of DP arising solely from STFM impairment may be relatively uncommon in this population.

This conclusion is further suggested by the results of our second experiment where we found that the face matching deficits seen in DP were insensitive to retention interval; i.e., that very similar levels of impairment were seen at the short and long intervals. To date, only one other study has used a delayed match-to-sample task to explore the perceptual and mnemonic contributions to DP (Shah, Gaule, Gaigg et al., 2015). In this study, the authors found that 15 DPs exhibited comparable face matching deficits at short (2 seconds) and longer (8 seconds) intervals. We replicated this result in a sample of 16 different DPs. In addition, the present results show that DPs still exhibit similar impairments at short and long retention intervals when a 45° viewpoint disparity exists between target and test items. Importantly, the results of Experiment 2 exclude the possibility that DPs have a particular problem retaining percepts in a way that supports rotation and manipulation (working memory; Baddeley, 1992, 1993, 2010).

The view that the face matching deficits seen in DP are relatively insensitive to memory load is also suggested by a finding recently described by Jackson, Counter, and Tree (2017; Experiment 1). Rather than vary retention interval, the authors manipulated memory load by increasing the number of target faces observers had to memorise (one, two, three, or four). Participants were asked whether a single test image presented a second later was one of the targets. As expected, the authors found that matching accuracy decreased as a function of the number of target faces held in memory (a main effect of Memory Load), and that relative to controls, DPs performed poorly in all conditions (a main effect of Group). Crucially, however, the relative impairment of the DPs did not increase with memory load[15]. The insensitivity of the DPs' deficits to the memory load manipulation mirrors the findings of the present study. Once again, this result suggests that the matching deficits observed have a perceptual origin; for example, the DPs in the experiment described by Jackson and colleagues (2017) may have had problems forming a perceptual description of the test face, and thus exhibited poor matching at all levels of the memory load manipulation.

5.5.2 Reconsidering the case against apperceptive accounts

Many DP samples – including ours – include individual DPs who show marked impairment on the CFMT but who exhibit only marginal impairment on the CFPT. The fact that the CFMT (a task with both perceptual and memory components) is more likely to reveal clear deficits at the single-case level than the CFPT (a task that assesses face perception with minimal memory demands) has led many to speculate that DP may often be caused by aberrant STFM, and not impaired perceptual encoding (Bowles et al., 2009; Dalrymple et al., 2014; McKone et al., 2011; Ulrich et al., 2017). Where observed, however, we recommend authors to treat this apparent dissociation with caution. First, the CFPT is simply a less reliable measure than the CFMT (Bowles et al., 2009). The fact that the CFPT has relatively few trials, and the way sorting performance is scored, may make it less likely to reveal single-case differences than the

[15] In the second experiment described by Jackson et al (2017), trials presented four faces sequentially for 500 ms each, followed by a maintenance interval of one second. Participants were asked whether a single test image presented a second later was one of the targets. In their analysis, the authors examined how observers' discrimination varied as a function of the position of the target in the sequence (first, second, third, fourth). Although the DPs performed relatively poorly in all categories, their serial-position functions closely resembled those of the controls; for example, both the DPs and controls were more accurate when matching recently presented targets.

CFMT. Second, the CFPT and CFMT differ not only in terms of their respective memory components, but also in terms of their fundamental perceptual demands. It is unclear whether the basic structure of the sorting task employed in the CFPT renders it more susceptible to compensatory strategies (e.g., moving closer to the display, looking for trivial details that distinguish faces). Third, having analysed a large sample of DPs and controls we found evidence that observers' CFPT scores correlate closely with their CFMT scores (r = .665). This finding suggests that, despite its name, CFMT performance may be heavily influenced by individuals' perceptual encoding ability.

Individuals with an apperceptive face processing deficit would be expected to exhibit aberrant perception and recognition of facial expression (Biotti & Cook, 2016, 2017; De Renzi et al., 1991; Duchaine, Parker, & Nakayama, 2003). Studies describing (seemingly) typical recognition of facial emotion in DP (e.g., Humphreys et al., 2007; Ulrich et al., 2017) therefore appear to challenge the view that the majority of individuals with DP exhibit some degree of apperceptive impairment. We note, however, that sensitive psychophysical tasks – *and appropriate analyses* – may be required to detect expression recognition difficulties arising from impoverished structural description. Having employed expression morphing and the estimation of psychometric functions, Biotti and Cook (2016) found that subtle expression recognition deficits were relatively common in a sample of 17 DPs (see also Burns et al., 2017). In contrast, tasks that simply require participants to label prototypical expressions ('basic emotions') may be prone to ceiling effects and lack the sensitivity necessary to detect subtle deficits (for related discussion, see Ipser & Cook, 2015).

Recent evidence suggests that most individuals with DP show typical susceptibility to the composite face effect (Biotti, Wu et al., 2017; Esins et al., 2016; Le Grand et al., 2006; Susilo et al., 2010; Ulrich et al., 2017), a visual illusion thought to index holistic face processing (Murphy, Gray, & Cook, 2017; Rossion, 2013). While these results suggest that holistic face processing may be intact in DP, they by no means exclude all apperceptive accounts of the condition. For example, DPs may have an apperceptive problem that affects local feature descriptions. Consistent with this possibility, many DPs struggle to make judgements about local regions shown in isolation (Biotti & Cook, 2016; Duchaine et al., 2006; Liu & Behrmann, 2014). We also note recent evidence from aperture viewing paradigms suggesting that the ability to process local regions may be a key determinant of face recognition performance (Murphy & Cook, 2017).

5.5.3 Insensitivity of face matching deficits to viewpoint disparity

Different-viewpoint matching is thought to be a better test of face perception ability than constant-viewpoint matching (e.g., Duchaine & Nakayama, 2006a). To match unfamiliar faces across different viewpoints, observers must infer the 3D structure of a target face from an ambiguous 2D image depicting a single view. This represents a substantial computational challenge (Todd, 2004). In the absence of an image-change, constant-viewpoint matching can in principle be accomplished using superficial pictorial cues (Hancock et al., 2000; Megreya & Burton, 2006). One might therefore expect DPs to show greater impairment, relative to controls, when matching across different viewpoints. The fact that our DPs exhibited similar deficits when matching faces shown from the same viewing angle, and faces shown from different viewing angles (Experiment 2), is therefore striking. Rather than dissociation between constant-viewpoint and different-viewpoint face-matching, our results suggest association: our participants appear to have used a similar process in both conditions. This is further suggested by the fact that participants' constant-viewpoint matching ability was closely related to their different-viewpoint matching ability.

One possibility is that observers accomplished both types of face matching through superficial pictorial cues, and that DPs experience difficulties using this strategy. This seems unlikely for two reasons. First, image matching is by definition a domain-general process (Hancock et al., 2000; Megreya & Burton, 2006). Crucially, however, our DPs were unimpaired at car matching in our second experiment. Similarly, the DPs tested by Shah et al. (2015) showed typical matching of chairs, butterflies, and hands. These convergent findings argue against a simple picture matching deficit. Second, face matching accuracy – but not car matching accuracy – correlated with the face-recognition problems encountered by observers outside the lab, as measured by the PI20 (e.g., mistaking familiar people for strangers, failing to recognise people in the absence of vocal cues, problems recognising people wearing hats or different hairstyles). These difficulties seem unlikely to reflect aberrant processing of trivial pictorial cues. Instead, this correlation underscores the fact that the processes measured by our matching task have meaningful consequences for the day-to-day social interactions of our participants.

Instead, we favour the view that observers use 'face-centred' (Bruce & Young, 1986; Marr & Nishihara, 1978) structural descriptions to achieve both constant-viewpoint and different-viewpoint face matching[16]. We speculate that i) these structural descriptions augmented the matching performance of typical observers in both the constant-viewpoint and different-viewpoint matching conditions; and ii) the DPs were outperformed in all viewing conditions because they were hampered by imprecise structural descriptions. There is little doubt that seeing to-be-learned individuals in different poses, with different expressions, from different viewing angles (so-called exemplar variation) aids face learning (e.g., Ipser, Ring, Murphy, Gaigg, & Cook, 2016; Murphy, Ipser, Gaigg, & Cook, 2015). The suggestion that observers form face-centred descriptions of unfamiliar faces from a single 2D image may therefore seem counter-intuitive. Consider, however, that computer programs have been described that do precisely this: i.e., extrapolate a morphable, posable 3D model of a human face from a single image of a novel face, using the covariation present in a set of training images (e.g., FaceGen Modeller). Once derived, these morphable posable models can be used to estimate how the target face will appear from different viewing angles (e.g., Jones, Dwyer, & Lewis, 2017). In a similar way, the human visual system may use the statistical regularities present in the faces it has encountered in the past to estimate the likely 3D structure of novel faces.

5.5.4 Is DP associated with a face-specific or domain-general deficit?

It remains unclear whether the deficit seen in DP is face-specific or indicative of a domain-general impairment (Gerlach, Klargaard, & Starrfelt, 2016; Geskin & Behrmann, 2017). On the one hand, we observed a significant group difference on the CCMT and evidence of a modest correlation (r = .437) between face and car matching accuracy. In our first experiment, we also found that the DP group made more errors than the typical controls when sorting inverted faces, regarded by some as a measure of domain-general perceptual ability (e.g., Rossion, 2008, 2013). On the other hand, our DPs were unimpaired in the car matching condition of Experiment 2, and other authors,

[16] We use the term face-centred rather than view-invariant to reflect the fact that these representations do not exhibit perfect view invariance. We note, however, that observers' matching performance – in the present study and elsewhere – typically far exceeds chance even when pairs of unfamiliar faces are presented with large viewpoint disparities. Given the highly complex 3D shape of the human face, and the fact 3D structure must be recovered from a highly ambiguous 2D image, this is a remarkable achievement of the human visual system.

for example, Shah et al. (2015; $N = 15$ DPs) and Esins et al. (2016; $N = 16$ DPs), have found that DPs' performance on the CCMT is comparable with matched controls.

Evidence of idiosyncratic, inconsistent object recognition deficits in DP accords well with the independent disorders hypothesis - the view that forms of developmental agnosia affecting faces and objects are best thought of as independent neurodevelopmental conditions (Gray & Cook, 2018). This account predicts the existence of 'pure' cases of DP and developmental object agnosia (DOA), individuals who experience impaired face recognition but typical object recognition (Duchaine et al., 2006), and vice versa (Germine, Cashdollar, Düzel, & Duchaine, 2011). However, the independent disorders hypothesis also predicts that the incidence of DOA will be higher in DP than in the wider population due to common genetic or environmental risk factors. For example, susceptibility to aberrant structural development of occipito-temporal cortex (e.g., reduced density and coherence of white matter tracts or atypical neural migration) may be a common risk factor for DP and DOA (see also Susilo & Duchaine, 2013).

5.6 References

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Chapter 6: Overview and discussion

The present Chapter will provide a summary of the findings presented in the empirical Chapters (i.e. 2-5) and their significance within the wider context of the cognitive mechanisms that may play a role in the nature and aetiology of DP. For each study the limitations and pending issues will be examined, highlighting those questions that have not been answered and constitute the basis for future research. In Section 6.1 I will examine the experiments described in Chapter 2, which investigated facial emotion recognition in individuals with DP using sensitive psychophysical tests. In Section 6.2 I will review the experiment described in Chapter 3, which investigated body recognition in DP. In Section 6.3 I will examine the study described in Chapter 4, which tested the susceptibility of individuals with DP to the composite face illusion. In Section 6.4 I will review the experiments reported in Chapter 5, which measured the contribution of perceptual encoding and short term face memory in DP's recognition impairments. Finally, in Section 6.5 a general conclusion will summarise the key findings of this thesis and suggest future directions for the study of DP.

6.1 Impaired perception of facial emotion in Developmental Prosopagnosia (Chapter 2)

6.1.1 Summary and interpretation

This study sought to systematically examine expression recognition in DP, testing a sizable sample of individuals with DP on sensitive psychophysical tasks. Previous literature had reported mixed results: some authors failed to find emotion recognition deficits in DP (e.g., Palermo et al., 2011; Humphreys et al., 2007; Dobel, Bölte, Aicher, & Schweinberger, 2007), whereas others reported widespread difficulties (e.g., Burns, Martin, Chan, & Xu, 2017; Duchaine et al., 2006; Ariel & Sadeh, 1996; de Haan & Campbell, 1991; Duchaine, Murray, Turner, White, & Garrido, 2009; Minnebusch, Suchan, Ramon, & Daum, 2007; Schmalzl et al., 2008). In Experiments 1 and 2 participants had to recognise the emotion depicted by faces drawn from morph continua, wherby expressions varied in ambiguity (Experiment 1) or intensity (Experiment 2). We sought to determine whether the use of sensitive methods could reveal more widspread deficits in the DP population. Morphing technique allowed us to generate more sensitive tests by creating ambiguous stimuli that placed greater demands on perceptual and decision processes.

In Experiment 1 we investigated the ability of DPs to make binary categorisations of whole-face emotional expression stimuli created by morphing two distinct facial expressions. Three morph continua (i.e., anger-happiness, disgust-sadness, fear-surprise) were produced by blending together two faces of the same actor expressing different emotions. Each continuum contained seven stimuli which varied in emotion amount from 20% to 80% in equidistant 10% increments. Participants were presented with each stimulus individually and had to make binary decisions on which emotion was depicted. Data were modelled by fitting psychometric functions and revealed that relative to controls DPs were significantly less accurate when discriminating between fearful and surprised facial expressions. Crucially, the performance at this task highly correlated (r = .78) with the ability of participants to detect physical differences between faces, measured by the CFPT. Moreover, single-case analyses showed that only the apperceptive subgroup of DPs were impaired, whereas the non-apperceptive individuals did not differ from typically developed participants.

Experiment 2 was designed to investigate two possible accounts for the findings emerged in Experiment 1, given that impairments occurred only in the fear-surprise morph continuum. On one hand, participants with DP may struggle at integrating information from different facial regions into a perceptual whole, finding more difficult to form a unified perceptual description of facial expressions. On the other hand, DPs - particularly those with an apperceptive profile - may find it problematic to encode the local information in the eye region, resulting in difficulties discriminating between fearful and surprised faces. Therefore, Experiment 2 sought to examine participants' ability to recognise facial emotion from the eyes. Stimuli consisted of eye regions drawn from four different morph continua (i.e., happiness, sadness, anger, fear) in which we merged an actor's facial expression with their neutral expression, creating three levels of intensity for each emotion continuum (high-70%, moderate-50%, low-30%). Results showed that relative to controls prosopagnosics were impaired at categorising facial emotion in the 70% and 50% conditions. As for Experiment 1, when DPs were divided into two sub-groups only the apperceptive prosopagnosics showed impairment at this task.

Overall, the use of sensitive tasks, which avoided performance accuracy ceiling effects, allowed to reveal widespread difficulites recognising facial emotion in DP.

Specifically, prosopagnosics were less accurate at making binary classifications of whole-face expressions (Experiment 1), and even when the emotion had to be recognised locally using cues from the eye-region only (Experiment 2). These difficulties appeared to be face-specific, as performance on a comparable vocal affect recognition task was intact (Experiment 3). One of the most compelling findings of this study was the striking correlation between facial emotion recognition accuracy and performance on the CFPT. Individual differences observed within the DP population, which in this study was broadly divided into apperceptive and non-apperceptive subtypes, provided a strong evidence that different behavioural profiles may stem depending on where the deficit occurs in the face processing stream. Specifically, apperceptive DPs may have difficulties forming view-invariant structural descriptions of faces at early stages of encoding, with consequent problems in the processing of both identity and expression (Bruce & Young, 1986; Haxby et al., 2000).

6.1.2 Limitations and outstanding questions

The leading models of face perception contamplate a clear separation between the processing of identity and expression (e.g., Bruce & Young, 1986). However, there is evidence suggesting that these two processing streams may be less independent than currently thought (e.g., Johnson, 2005; de Gelder, Frissen, Barton, & Hadjikhani, 2003; Knight & Johnston, 1997). For example, emotion-identity interference has been shown in a task where observers had to judge either the identity or the expression of stimuli which kept varying orthogonally along these directions (Schweinberger, Burton, & Kelly, 1999). In his commentary on the study, Van den Stock (2017) suggested two explanations of our results. First, assuming an emotion-identity interaction, a reduced performance on emotion recognition tasks in DP would be predictable irrespective of the locus of impairment in the processing stream, as the main characteristic of DP consists of identity recognition deficits. Therefore, one of the claims of Van den Stock (2017) is that emotion recognition impairments in DP may originate later identity processing and not at the structural description phase as suggested by our findings. However, the fact that in our study expression recognition strongly correlated with performance on the CFPT hinted that deficits were associated with poor structural encoding. Moreover, many DPs have been reported to exhibit poor identity recognition but intact emotion recognition (e.g., Dobel, Bölte, Aicher, & Schweinberger, 2007;

Humphreys, Avidan, & Behrmann, 2007; Palermo et al., 2011), suggesting that emotion deficits are not an inevitable consequence of identity problems.

The second point raised by Van den Stock in his commentary (2017) concerns the specificity of the difficulties observed. Aberrant expression recognition in DP could reflect a domain-general deficit affecting general visual processing. The main argument against this view consists of the evidence of lack of relationship between expression recognition accuracy in both Experiment 1 and Experiment 2 and performance on tests of object recognition (i.e., the Cambridge Car Memory Test and the Cambridge Bike Memory Test). Nevertheless, the extent to which these tasks tap similar mechanisms remains unknown and needs additional examination.

Crucially, the experiments presented in Chapter 2 exposed the importance of task sensitivity in detecting emotion recognition impairments in DP. On one hand, when tasks are too easy participants with DP have sufficient ability to label prototypical 'basic' emotions, performing within typical ranges. Conversely, very demanding tasks may also fail to reveal clear differences because the performance of the controls can be variable. Accordingly, in Experiment 2 we found clear discrepancies when 50% of emotion information was provided to the observer, but failed to detect differences when only 30% of task-relevant information was shown. It is important that forthcoming studies use sensitive tasks where typical observers score within the 'dynamic range', avoiding both ceiling and floor effects.

A spontaneous consideration on these results may question the relevance of such deficits, given that they appear to be relatively subtle. As a matter of fact, the expression of emotions in real life is often subtle itself, and efficient social interactions rely greatly on the ability to perceive and accurately classify these subtle facial changes. A failure in detecting slight expressions may result in inadequate behavioural responses, which may be a cause of social disabilities. In the DP population, in particular, these deficits may exacerbate identity face recognition difficulties, affecting the development of social skills, and ultimately reducing the quality of social interactions.

A key open question that needs to be addressed relates to the frequency of expression recognition deficits in DP. Future research will have to clarify whether all DPs or only an apperceptive subset of them show signs of impaired facial emotion recognition.

6.2 Impaired body perception in Developmental Prosopagnosia (Chapter 3)

6.2.1 Summary and interpretation

The study presented in Chapter 3 aimed to investigate body recognition in DP. Previous evidence on the topic was limited, and existing behavioural studies reported typical body and body parts matching accuracy in prosopagnosics (Duchaine, Yovel, Butterworth, & Nakayama, 2006; Rivolta, Lawson, & Palermo, 2016; Shah, Gaule, Gaigg, Bird, & Cook, 2015). However, many parallels between body and face perception had been reported, including the recruitment of adjacent brain areas in the visual cortex (Peelen & Downing, 2007) and comparable inversion effects (e.g., Cook & Duchaine, 2011; Reed, Stone, Bozova, & Tanaka, 2003; Robbins & Coltheart, 2012). Our study aimed to explore body recognition in a sizable sample of individuals with DP and using a sensitive task.

Individuals with DP ($N = 20$) and typical observers ($N = 23$) completed a delayed match-to-sample task in which they had to match exemplars of faces, headless male torsos and cars. As expected, prosopagnosics were impaired at the face trials, but crucially they also showed evidence of impaired body and object recognition accuracy at the group level, and several individuals exhibited difficulties also at the single case level. However, we observed little or no relationship between observers' car perception ability and their body perception ability. The lack of relationship between observers' performance at cars and torsos trials suggested that body and object recognition impairments in DP may co-occur independently.

Taken together these results revealed that some individuals with DP tend to show impaired body perception. This finding accords with the evidence of many neuro-cognitive correspondences between faces and bodies. In fact, given the proximity of brain areas subtending the processing of faces (i.e., the occipital face area and the fusiform face area) and bodies [i.e, the extrastriate body area (EBA) and the fusiform body area (FBA)], perfectly typical body recognition would be unlikely. Instead, differences in the organisation of white matter tracts in occipitotemporal regions well-documented in DP (e.g., Gomez et al., 2015; Thomas et al., 2009) may explain co-occurring body perception deficits. Similarly, looking at cognitive similarities in the processing of faces and bodies, both possibly relying on configural information, it is expected that those individuals with DP experiencing aberrant holistic processing may exhibit problems with the perception of both categories of stimuli.

6.2.2 Limitations and outstanding questions

One of the aspects that may weaken the conclusions of this study is the use of torsos, instead of whole bodies. Some authors may argue that torsos would be better described as body parts, and previous studies on body recognition have used whole figures, headless or with masked faces. Accordingly, the inversion effect indicative of holistic processing has always been found using whole bodies. However, torsos unlike other body parts elicit strong responses both in EBA and FBA (Taylor, Wiggett, & Downing, 2007). The decision of using torsos was influenced by the possibility that observers, especially those with body perception deficits, would benefit from limb-matching strategies. Moreover, torsos allowed us to present body stimuli in a scale that emphasised 3D shape variation minimising image-matching strategies. It is likely that whole bodies may recruit different mechanisms than those involved in torso processing. However, the stimuli used in this study were sufficiently challenging for the DPs.

A crucial conclusion of the study was based on the lack of correlation between car and body matching accuracy. Given the null result, it is imperative to acknowledge that the combined sample size (N = 43) was not particularly big for making confident inferences and strong interpretations. However, the key result for the purpose of the study was the lack of association that emerged between the two non-face conditions. If the group differences observed for the three categories reflected a single domain-general deficit, body and car perception ability would have clearly correlated.

Critically, the two types of deficit do not seem to be associated and they may likely present as independent developmental agnosias. This idea would not be surprising as co-occurrence is a common phenomenon in developmental disorders. For example, it is well established that Autism Spectrum Disorder tends to present with an array of other developmental conditions such as alexithymia (Bird & Cook, 2013), ADHD (Leitner, 2014), and dyslexia (Jones et al., 2009). While the probability to develop one of these disorders is higher in the autistic population compared to the general population, all the listed conditions can occur independently from each other. Co-occurrence indicates that common genetic or environmental factors may play a role in the aetiology of different neurodevelopmental disorders. Since the publication of the present study, Gray and Cook (2018) proposed that DP may be due to an increased susceptibility to atypical neurostructural development of the occipitotemporal cortex, resulting in a greater predisposition to various forms of visual agnosia. Future research must explore whether DP, developmental object agnosia and developmental body agnosia can exist as independent conditions, rather than as manifestations of a common aberrant domain-general mechanism.

Finally, this study raised interesting considerations for a new framework of body recognition research. What can be defined as body in the context of social perception research remains unanswered. Even where people use whole bodies there are some disagreements about the inclusion of the head region, and the inclusion of feet and ankles, which often are cropped. For example, the presence of the face or the head may influence the inversion effects observed with body stimuli. In many cases, body stimuli are also clothed and it is not clear how this affects the underlying visual processing. In fact, the presence of clothing occludes surface reflectance information and often introduces sharp boundaries between different stimulus regions. Future studies will have to clarify how these aspects impact on observers' visual processing of body stimuli.

6.3 Normal composite face effects in developmental prosopagnosia (Chapter 4)

6.3.1 Summary and interpretations

The study presented in Chapter 4 sought to investigate whether DP is associated with reduced susceptibility to the composite face illusion. Faces are typically perceived using holistic processing, whereby local features are integrated into a unified whole. When the top half of one face is paired with the bottom half of a different face, observers tend to fuse them perceptually when presented upright and aligned. This 'composite effect' (CFE) results from an implicit tendency to integrate information from disparate regions when faces are presented canonically (i.e., the effect disappears when the two halves are misaligned or when faces are inverted). Some authors have proposed that susceptibility to this illusion may be associated to better face recognition skills (e.g., Rivest, Moscovitch, & Black, 2009; Rossion, 2013; Murphy, Gray, & Cook, 2017). Accordingly, previous studies showed that individuals with DP are less sensitive to the CFE, suggestive of reduced holistic face processing (e.g., Palermo et al., 2011; Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014).

Two independent samples of prosopagnosics took part in tasks investigating CFE using different paradigms. In Experiment 1 (N_{DPs} = 16) participants took part in a simultaneous composite matching task, while in Experiment 2 (N_{DPs} = 24) observers completed a sequential composite matching task. In both tasks participants had to match the eyes of composite displays presented either upright or inverted, aligned or misaligned. Results failed to show evidence of diminished CFEs in both samples of individuals with DP, who responded faster and more accurately when composites were inverted or misaligned.

These findings conflict with previous evidence reporting diminished CFE at the group level in participants with DP (Palermo et al., 2011; Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014). However, the interpretation of previous studies' results may be compromised by the composition of their samples. In the typical population age plays a critical role in observers' sensitivity to the composite face illusion and older participants have been reported to be less sensitive to configural processing (Meinhardt, Persike, & Meinhardt-Injac, 2016; Wiese, Kachel, & Schweinberger, 2013). Accordingly, aberrant CFE in DPs is likely to be evident only in older cases (e.g., Schmalzl, Palermo, & Coltheart, 2008; Avidan, Tanzer, & Behrmann, 2011).

Another factor that may lead to dissimilar results consists of the way performance is measured. Previous reports have mainly focussed their analyses on response times. For example, in Palermo and colleagues (2011) DPs and controls exhibited comparable CFEs in their accuracy data and the group difference only emerged in response times when these were adjusted for error rates in the misaligned condition. In particular, the authors failed to show a significant Alignment × Group interaction for RTs. Furthermore, the score distribution suggested that the group difference was highly influenced by the performance of a single DP who exhibited a reversed composite effect (i.e., their aligned RTs were much faster than their misaligned RTs).

Finally, specific problems encoding local face regions, which are often observed in DP, may influence the occurrence of atypical CFEs. For example, DP participants in Liu and Behrmann (2014) performed significantly worse than typical observers in the baseline misaligned condition, indicative of reduced local processing. Composite face tasks require observers to encode local regions and make perceptual decisions on their similarity or dissimilarity. Therefore, the execution of this task is based on the assumption that observers exhibit typical local processing of faces. Distractor halves perceived as non-distinctive by DPs may lead to a weaker perceptual prediction and exert less illusory bias in the aligned condition, compared to distractor halves which are perceived as distinctive. In fact, any attempts to account for baseline differences by modulating the holistic processing index based on the performance in the misaligned condition result misleading; it is not clear what is a 'typical' CFE when participants display an atypical misaligned performance. Hence, assuming impaired configural processing using the composite face task in a population exhibiting atypical local processing, becomes problematic.

6.3.2 Limitations and outsgtanding questions

Differences in the task designs employed by our study and previous studies may contribute to the mixed findings. The choice of using the original versus the congruent (or complete) design in composite tasks complicates the interpretation of results. While in original designs the distractor regions always differ, in congruent designs a full factorial combination of target regions (same and different) and distractor regions (same and different) is used (e.g., Richler & Gauthier, 2014; Rossion, 2013), and holistic processing can be measured as a Congruency × Alignment interaction (i.e., performance is better in congruent trials and this congruency effect is modulated by alignment). It has been suggested that congruent designs allow to control for the effects of response bias, which vary as a function of congruency. For example, in original designs participants may have a tendency to respond 'different' in aligned trials for two potential reasons: i) less perceptual discriminability due to holistic processing; ii) a response bias due to the incongruent bottom halves. Crucially, since in original designs same trials, whose analysis is critical for the CFE, are always incongruent, participants' responses can be affected by a congruency interference (see Richler & Gauthier, 2014).

Our choice of using the original paradigm stems from concerns that effects obtained with the congruent design may confound the CFE with other effects attributable to general cognitive factors such as response facilitation, generic interference, attentional and decisional biases (Rossion, 2013). These effects may partially explain why congruent designs often yield to composite effects with stimuli that generate no tangible visual illusion (e.g., unfamiliar Chinese symbols, Hsiao & Cottrell, 2009), while the original design produces effects only for faces, and bodies to a certain degree (for a review see Murphy, Gray, & Cook, 2017). It is vital that our findings are replicated using other paradigms, including the complete design. This would provide further support to our conclusions. Encouragingly, our findings have been recently replicated using a novel psychophysical variant of the composite face task that overcomes many of the problems associated with the different matching paradigms (Gray & Cook, 2017).

Some readers may object to the fact that different face stimuli were used in Experiments 1 and 2. Furthermore, how can we compare the results of simultaneous versus sequential matching tasks? Nevertheless, the fact the these experiments differ in their stimuli, procedures, and participants can be considered a strenght as the two datasets consist of independent tests of the same hypothesis, providing additional support to our conclusions.

Taken together these considerations show a complex picture where holistic processing seems to be impaired in many DPs, yet our participants did not show reduced sensitivity to the composite face illusion. The CFE is not the only measure known to reflect holistic face processing. In fact, other perceptual phenomena like the inversion and the part-whole effects have been shown to originate from perceiving faces holistically (section 1.3.3.1). Traditionally these three phenomena have been thought to emerge as by-products of a single underlying mechanism (e.g., Farah, Wilson, Drain, & Tanaka, 1998; Maurer, Le Grand, & Mondloch, 2002), but recent evidence suggests that people's susceptibility to the composite face illusion is rather weakly associated to other measures of holistic processing (see Rezlescu, Susilo, Wilmer, & Caramazza, 2017). Therefore, our conclusions do not deny the existence of aberrant holistic processing in DP. On the contrary, we believe that holistic processing has a multifacet nature and different aspects of it may be impared in the DP population. Future research is necessary in order to distinguish between types of holistic face processing deficit, and to show how different profiles of impairment contribute to face recognition difficulties.

Relative to this last point, our results directly inform the ongoing debate on whether sensitivity to the CFE is associated to face recognition ability. While some authors believe that holistic processing predicts face recognition in the general population, relying on evidence of positive correlations (e.g., DeGutis, Wilmer, Mercado, & Cohan, 2013; Richler, Cheung, & Gauthier, 2011), other studies have questioned these conclusions, finding little or no correlation (e.g., Rezlescu, Susilo, Wilmer, & Caramazza, 2017; Wang, Li, Fang, Tian, & Liu, 2012). In addition, our results indicate that inferring face recognition ability from the CFE's results can be misleading. Face recognition may be a complex skill which relies on multiple cognitive mechanisms -

the process reflected by the susceptibility to the CFE may make a necessary causal contribution to face recognition, but it may not be a key source of individual differences.

6.4 Is developmental prosopagnosia best characterised as an apperceptive or mnemonic disorder? (Chapter 5)

6.4.1 Summary and interpretations

The study presented in Chapter 5 sought to investigate whether DP is associated with aberrant structural encoding of faces (i.e. apperceptive deficit) or difficulties maintaining visual percepts over time (i.e. mnemonic deficit). First, apperceptive deficits were explored by testing a large sample of DPs on the Cambridge Face Perception Test (CFPT) (Experiment 1). Next, a subset of DPs completed a delayed match-to-sample task with face and car stimuli, where both retention interval and viewpoint were manipulated (Experiment 2).

The aim of Experiment 1 was to investigate apperceptive impairments using the CFPT in a large sample of individuals with DP (N = 72). Participants had to sort six faces based on similarity with a target face. Stimuli were presented either upright or inverted, for a total of 16 trials. Many studies had reported that groups of DPs make more errors on the CFPT compared to controls. However, these differences rarely are reflected at the single-case level. Two competing explanations guided Experiment 1. One possibility is that group differences in the CFPT performance, where observed, are driven by a few DPs with apperceptive impairments. The second possibility is that apperceptive deficits are widespread in the DP population, but the CFPT may not be sensitive enough to detect them, leading to distributions of DPs' and controls' scores that overlap to some degree. In addition, the sequential sorting task employed by the CFPT may render it more prone to compensatory strategies, allowing apperceptive DPs to achieve scores within the normal range. Results revealed that relative to controls, DPs were impaired at the group level. Having split the groups based on their performance on the upright trials, we found evidence that the distribution of DP scores was shifted. Not only the worst-performing but also the best-performing DPs performed worse than the worst-performing controls and the best-performing controls, respectively. We found that inflating the CFPT scores of the typical observers by 90% provided a reasonable model of the distribution of DP scores. These findings support our second hypothesis and suggest that some DPs may score typically due to the use of compensatory strategies, such as excellent attention to detail or visual acuity.

The aim of Experiment 2 was to determine whether the face matching deficits observed in DP increase as a function of retention interval. A subset of DPs ($N = 16$) completed a delayed match-to-sample task using face and car stimuli, whereby participants had to select a target stimulus from a test display of four items. Memory demands were manipulated by introducing either a short (1 second) or a long (6 seconds) retention interval between target presentation and test array. This manipulation allowed us to measure the contribution of perceptual versus mnemonic impairment by maintaining the perceptual demands constant. In particular, if DP is associated with an apperceptive impairment, similar deficits will emerge under conditions of high and low memory demand. On the other hand, if DPs exhibit a mnemonic profile, disproportionate impairment will emerge in the high memory demand condition. In addition, we sought to determine whether a change in viewpoint from target to test affected performance, particularly under different mnemonic demands. Results revealed that relative to controls, DPs were less accurate at matching faces. Crucially, the impairment was insensitive to both changes in retention interval and viewing angle, indicating that matching difficulties in DP may likely stem from an apperceptive deficit.

Overall, this study revealed that DPs are impaired at the group level at a simultaneous sorting task which minimises memory demand (the CFPT), indicative of an apperceptive impairment (Experiment 1). Moreover, DPs were impaired at face matching, but the relative degree of impairment did not interact with retention interval (i.e., the levels of impairment were similar when matching faces viewed with a 1-second delay and when matching faces viewed with a 6-second delay) (Experiment 2).

Taken together these results suggest an apperceptive account of many cases of individuals with DP. Although heterogeneity is likely in the DP population, the lack of positive evidence for the mnemonic accounts is problematic. Consistent with an apperceptive account, the DPs made more errors on the CFPT at the group level compared to typically developed individuals. Moreover, not only the lower end but the entire distribution of DPs' scores was shifted, indicating that also the best performing DPs performed worse, on average, than controls. This result is likely to be due to poor perceptual encoding rather than impaired short term face memory, as memory load is kept minimal in this test. In support of this account, we showed that face matching deficits in DPs are insensitive to retention interval, not only when matching across the same viewpoint, but also when the perspective of test items is rotated from the target. This finding suggests that prosopagnosics are able not only to retain percepts, but also mentally manipulate and rotate them, indicative of unimpaired working memory. The apperceptive account accords well with evidence of widespread expression recognition deficits in this population (Biotti & Cook, 2016; Duchaine, Parker, & Nakayama, 2003), and it may support recent suggestions of typical composite face effect in DP, whereby the lack of susceptibility to the perceptual illusion may originate from poor local processing rather than typical holistic processing.

Finally, a striking result of this study consists of the insensitivity of face matching deficits to changes in viewpoint in DPs. We speculate that participants used a similar process when matching faces shown from the same and from different angles. This conjecture is reinforced by the close relationship that emerged between constant-viewpoint and different-viewpoint matching abilities. Crucially, we exclude the possibility that observers may have used superficial pictorial cues to accomplish face matching in both conditions. In fact, DPs were unimpaired at car matching and it is well established that image matching relies on a domain-general process. Instead, it is likely that participants used view-invariant 'face-centred' structural descriptions to achieve both constant-viewpoint and different-viewpoint face matching. But while controls could benefit from these mechanisms, DPs were outperformed in both conditions because of less precise structural descriptions.

6.4.2 Limitations and outstanding questions

The fact that many DPs show typical performance on the CFPT has been taken as evidence of mnemonic impairment by previous reports (e.g., Bowles et al., 2009; Ulrich et al., 2017). Conversely, we showed that differences at the group level in the CFPT may rather indicate an apperceptive deficit, whereby, given the poor reliability of the CFPT, some apperceptive DPs may use compensatory strategies to perform typically. The CFPT is known to be a less reliable measure compared to other tests of face recognition ability due to its susceptibility to the use of compensatory strategies, reliance on low-level visual processing, and procedure requirements (e.g., use of trackpad, limited time). Therefore, the noisy nature of the CFPT limits the strength of our conclusions. Future research in the filed should prioritise the development of a sensitive and reliable test of perceptual encoding. Such a measure might reveal even clearer evidence of widespread apperceptive impairment in the DP population.

The basic finding of Experiment 2 was the lack of Retention Interval × Group interaction, whereby DPs did not show disproportionate impairments in the high memory demand condition. This results leaves open the possibility that a memory impairment may already occur within the 1 second interval, which corresponded to the low demand condition. The only way to address this point would have been adding a simultaneous matching condition, which did not require any memory retention. Critically, a no-interval condition would have changed the perceptual demands of the task, as five faces would appear in the test array rather than four. Therefore, any reduction in memory demand associated with the simultaneous presentation of five faces would be confounded with an increase in perceptual and attentional demands. Instead, we chose to consider performance at the CFPT as a proof that matching impairments are not associated to aberrant STFM. In fact, the performance distribution of a large sample of DPs was entirely shifted, suggestive of a clear apperceptive profile in most prosopagnosics. Moreover, even considering the occurrence of a mnemonic deficit at shorter retention intervals, it is rather challenging to explain the lack of any group difference increment as a function of retention interval. On the other hand, it is crucial to replicate these results using other experimental parameters, in particular longer retention intervals. In fact, our results do not exclude the possibility that DPs may have difficulties at face learning.

Overall, this study provides evidence against a mnemonic impairment in most cases of DP, but focussing only to a specific type of memory: The short-term face memory. These findings do no discount the existence of a memory impairment occurring after encoding, and involving the association of the percept to semantic information stored in long-term memory. Emerging evidence reveals several cases of people who report severe difficulties in everyday face recognition, despite typical performance at the CFMT and the CFPT. Specific impairments involving semantic memory may be only detectable by tests measuring familiar face recognition (e.g., the Famous Faces Test), which require the access to semantic information to be able to retrieve a person's identity. Future research needs to be done to investigate different profiles of DP, shifting the focus from unfamiliar to familiar face recognition, starting from unravelling the neuro-cognitive mechanisms that regulate face learning.

6.5 General conclusion

This thesis sought to investigate profiles of impairment and cognitive mechanisms of DP using sensitive behavioural tasks. The heterogeneous nature of DP often yielded to mixed results and contradictory evidence. Traditionally, DPs have been clustered into two subgroups (i.e., apperceptive vs mnemonic), depending on where the deficit occurred in the face processing stream. Evidence for an apperceptive account was supported by studies showing face perception difficulties at the group level on face matching tasks, facial emotion, age and gender recognition (section 1.3.3). In addition, a major debate reported in this thesis consisted of the nature of impairment, which for some authors is face specific, while for others originates from an aberrant domain-general mechanism (section 1.3.5.2). Inconsistent findings were also found in studies investigating cognitive mechanisms responsible for the condition. A pivotal account of DP suggested that impaired holistic processing of faces may be a crucial factor leading to poor face recognition (section 1.3.3.1). Several studies found reduced composite face effects in DPs, suggestive of diminished holistic processing in this population (section 1.3.3.1.2). Finally, previous literature tried to associate DP with aberrant short term face memory, assuming normal face perception from typical performance of many DPs on the CFPT, a test which minimises memory demands (section 1.3.4).

6.5.1 Towards an apperceptive account of DP

The findings described in Chapter 2 provide strong evidence of widespread facial emotion recognition deficits at the group level in DP. Furthermore, aberrant expression recognition seems to be associated to apperceptive types of DP, whereby perceptual deficits affect early stages of structural encoding, before the neuro-cognitive pathways that analyse changeable and invariant facial information bifurcate. Crucially, apperceptive problems appear to be relatively common within the DP population as shown by our CFPT findings collected in a large sample of prosopagnosics (Chapter 5). In this study, we showed that the entire distribution of DPs scores at the CFPT was shifted relative to controls, suggesting that difficulties were present at the group level despite some DPs scoring within the typical range (Experiment 1, Chapter 5). Accordingly, DPs impairments on a delayed match-to-sample task did not disproportionally increase from a short 1-second delay to a longer 6-second delay, excluding a mnemonic deficit and supporting the apperceptive account (Experiment 2, Chapter 5).

6.5.2 Local (rather than global) face processing may explain deficits in DP

Contrary to some leading accounts of DP our findings seem to support the idea of impaired local processing. For example, previous studies have attempted to describe DP as a failure of holistic processing, whereby DP individuals struggle to integrate local face features into a perceptual whole. This idea emerged largely from evidence of reduced susceptibility of DPs to the composite face illusion (e.g., Palermo et al., 2011; Avidan, Tanzer, & Behrmann, 2011; Liu & Behrmann, 2014). Instead, we showed comparable CFE in DPs and controls using two independent samples of prosopagnosics and different paradigms (Chapter 4). Our results suggest that although some DPs may show aberrant holistic processing, the processes responsible for the CFE appear to be unimpaired in this population, reinforcing the idea that holistic processing does not stem from a single underlying mechanism (see Rezlescu, Susilo, Wilmer, & Caramazza, 2017). One possibility emerging from some of the findings presented in this body of work is that processing of local features may be affected in many DPs. For example, we showed that the recognition of facial emotion was impaired also when judging expressions displayed by the eyes only (Experiment 2, Chapter 2). We advanced the possibility that some DPs, in particular those with apperceptive profiles, may struggle to encode facial shape from a very early stage of face processing, with consequent problems perceiving multiple facial attributes, such as identity or expression.

Interestingly, new evidence emerged that challenges holistic accounts of face inversion effect (Murphy & Cook, 2017). Using a dynamic aperture paradigm, the authors tested face recognition of upright and inverted faces shown either in their entirety or viewed through a dynamic aperture that moved incrementally across the face. Results showed that regardless of the orientation of the face, perceiving a whole-face is associated to better face perception than viewing the same face region-by-region. These findings suggest that perceptual decrements when viewing inverted faces may stem from poor descriptions of local regions rather than reduced configural processing.

6.5.3 The domain-specificity issue in DP and co-occurring developmental agnosias

The study described in Chapter 3 addressed the domain-specificity issue, exploring non-face recognition in DP. Findings revealed co-occurring deficits of body and car perception, which were not associated to each other, excluding a domain-general account. We speculated the existence of different forms of developmental agnosia affecting the perception of faces, bodies, and object, which may co-occur as independent neurodevelopmental conditions. These may account for the fact that many of our participants exhibit non-face recognition deficits. This aspect makes it pivotal to include non-face control conditions when testing DPs and employing object recognition measures, such as the CCMT, in the diagnostic assessment.

Co-occurrence is a typical phenomenon of many developmental disorders and common genetic, epigenetic and environmental factors may predispose one individual to a range of neurodevelopmental conditions. In the specific case of DP, it has been shown that abnormal development of the inferior longitudinal fasciculus contributes in face recognition impairments typical of this condition (e.g., Thomas et al., 2009; Song et al., 2015). Furthermore, depending on the specific locus of impairment along the tract, it is possible to predict perceptual deficits affecting non-face stimuli (Gomez et al., 2015). Therefore, co-occurring object recognition deficits seem to be by-products of aberrant neuro-substrates common to the neuro-cognitive processing of faces, objects, or bodies (see Gray & Cook, 2018). This idea goes against proposed accounts of DP resulting from an atypical single-mechanism responsible for the processing of faces as well as other classes of objects (e.g., Geskin & Behrmann, 2018). We rather propose that, where observed, object recognition impairments in DP present as independent developmental agnosias. Accordingly, we also predict the existence of dissociations, whereby the abnormal structural development of occipitotemporal areas can selectively impair the perception of object, or bodies, leaving face perception unaffected.

6.5.4 Toward an apperceptive characterisation of DP

Chapter 5 explored perceptual and mnemonic accounts of DP, finding that prosopagnosics are impaired at face matching both on a simultaneous task which minimises memory demands (Experiment 1, Chapter 5), and on a delayed match-to-sample task were retention interval and viewpoint are manipulated from target to test

(Experiment 2, Chapter 5). Crucially, DPs do not show signs of disproportionate impairment as a function of retention interval, indicating that their deficit has an apperceptive, rather than mnemonic, origin. Previous studies have excluded apperceptive deficits by looking at CFPT results, a test whose reliability has been questioned, and from evidence of typical recognition of basic facial expressions. However, we showed that normal performances on the CFPT by some prosopagnosics may indicate the use of excellent compensatory strategies rather than typical face perception. In support, we presented evidence of a distribution shift in a large sample of DPs, indicative of deficits at the group level. This finding was corroborated by results on a delayed match-to-sample task (Experiment 2), where an increment in retention interval from 1 second to 6 seconds did not affect the size of impairment in DPs. We concluded that most DPs appear to have an apperceptive impairment that prevents them from reaching a typical performance even on very simple face recognition tasks.

Taken together the findings reported in this thesis expand and enrich previous evidence. First, we show that apperceptive deficits seem to be relatively common in DP and difficulties deriving a structural description of face stimuli can be distributed in this population. Accordingly, prosopagnosics with severe perceptual deficits may also show co-occurring expression recognition impairments. Second, we illustrate that forms of developmental agnosia, especially those affecting face, body and object perception, can likely co-occur as independent neurodevelopmental conditions rather than as by-product of a common aberrant domain-general process. Finally, we demonstrate that DP is not associated with reduced susceptibility to the composite face illusion, suggesting that other mechanisms may give rise to face recognition difficulties in this population.

6.6 Limitations and future directions

6.6.1 Theoretical definitions and constructs

One of the main ongoing challenges in visual perception research consists of a lack of established and generally accepted theoretical definitions of key constructs. The studies presented in this thesis sought to investigate individual processes by using purposely-designed tasks in which we tried to measure the effect of the manipulation of specific variables on the process of interest (e.g. varying retention interval while maintaining the perceptual demand constant to tap short term face memory). However, we do not intend to claim that our tasks tap one single process eliminating completely the effects of other processes. On the contrary, our intention is that of maintaining stable the influence of other interacting processes while varying the contribution of the process of interest.

Existing theoretical positions tend to consider and refer to functions as independent constructs which can be potentially studied in isolation (e.g. Weigelt et al., 2013; Ulrich et al., 2017). This position posits a great empirical challenge. In fact, we believe that it is particularly difficult trying to distinguish between functions (e.g. local vs. global processing, face memory vs. face perception) without a clear definition of these constructs and without addressing their inevitable interplay. We may wonder, for example, where one starts and another one ends. A concrete example is provided by composite face tasks, which are used to study face holistic processing employing an ingenious paradigm that breaks down local versus global processing. Though, it is not clear what constitutes a 'pure' feature-based processing given that there is a degree of feature integration even in local processing – we may wonder to what extent the eye region is perceived using global versus local processing, for example. Therefore, when we observe normal composite face effects in DP not only are we not assuming normal holistic processing, but we are also avoiding strong claims relative to unimpaired local processing in our prosopagnosics.

Another example of the tendency to consider cognitive processes as independent and potentially dissociable constructs arises from the DP diagnostic practice. Among various tests, the most commonly employed diagnostic measures (i.e. the CFMT and the CFPT) have been designed to measure either face memory or face perception, as they were implicitly thought of as separable components. Nonetheless, these measures inevitably tap both processes and they cannot be referred to as pure measures of either face memory or face perception. A failure in acknowledging their interplay may lead to misleading interpretations of diagnostic results. For example, the fact that the CFMT (a task with both perceptual and memory components) is more likely to reveal clear deficits at the single-case level than the CFPT (a task that assesses face perception with minimal memory demands) has led many to speculate that DP may often be caused by aberrant short term face memory, and not impaired perceptual encoding (e.g. Bowles et al., 2009; Dalrymple et al., 2014; McKone et al., 2011; Ulrich et al., 2017). Crucially, the CFPT and CFMT differ not only in terms of their respective memory components, but also in their fundamental perceptual demands (i.e. they present different facial identities under different viewing conditions). Hence, not only do both these tasks have a perceptual demand, but they may even tax different types of perceptual processes. It is therefore misleading to equate the CFMT and CFPT to perceptual and mnemonic conditions in a controlled experimental manipulation. Although they differ in their respective memory demands, the differential memory load is confounded with numerous perceptual differences.

Inadequate definitions limit research, including the studies described in this thesis. Therefore, future research will need to nail down constructs in order to design tasks that better tap one process or another. For example, whenever researchers explore mnemonic accounts in DP, we encourage proponents to articulate more clearly what types of memory processes are thought to be impaired, whether perceptual encoding is preserved entirely, and what constitutes 'short-term' and 'long-term' face memory.

6.6.2 Heterogeneity in DP

DP is considered a heterogeneous condition – individuals with DP have been reported to vary greatly in terms of specific combinations of face-specific and co-occurring non-face deficits. Our prosopagnosics varied consistently regarding the specificity of their deficits. Overall, co-occurring object recognition impairments were inconsistent. In some studies, we were able to find clear group differences in object conditions, while in others non-face recognition was impaired only at the single case level. It appears that different studies may obtain different findings depending on the specific composition of the DP group (e.g., whereby samples are composed by more apperceptive DPs, researchers might find greater group differences at facial emotion recognition tasks, and vice versa).

Apparent heterogeneity in DP has also been inferred by putative dissociations between performance on the CFMT and CFPT. First, artefactual dissociations may arise from the fact that the CFMT plays a key role in the diagnosis of DP, while the CFPT does not (i.e., a clear deficit on the CFMT is widely seen as necessary for a DP diagnosis). Where individuals fail to reach this criterion, they are often excluded from DP research. In contrast, CFPT scores are free to vary and they are usually provided only as an indication of whether a DP is apperceptive or mnemonic (see Chapter 2 and Chapter 4). Therefore, considering this prevailing bias, it is unsurprising that the DP literature reports many individuals who exhibit a clear deficit on the CFMT but not on the CFPT. The practice of preselecting individuals based on extremely poor CFMT scores (i.e., <2 SDs below the mean), and then reporting single-case analyses to imply that an individual's CFMT deficit exceeds their CFPT deficit is known as 'double dipping' (Kriegeskorte, Simmons, Bellgowan, & Baker, 2009). Second, the CFPT is known to have poorer psychometric properties compared to the CFMT and it is therefore more likely to yield to noisy performance. For example, the format of the CFPT may render it more susceptible to compensatory strategies, than the CFMT (e.g. the side-by-side presentation of the faces, or the opportunity to study each trial display for a longer interval). Third, as previously mentioned, it is not clear whether meaningful theoretical inferences can be drawn from differential impairments on the CFPT and the CFMT, given that they differ not only in terms of their respective memory components, but also in their fundamental perceptual demands. Hence, the overarching point is that these putative dissociations between performance on the CFMT and the CFPT cannot be relied upon to make strong claims about DP heterogeneity.

A way to control for heterogeneity in future studies will require the employment of larger samples of DPs, tested on a variety of different (and more sensitive) measures. Although results from single cases can be very informative when using sensitive measures, the combination of poor tests and single case analyses can give the illusion of more heterogeneity than there actually is. We therefore encourage future research to test DPs on an array of reliable measures which tap several different processes. For example, as already suggested by other authors (Geskin & Behrmann, 2017), DPs could be tested on many different categories of non-face objects. This may yield to a variety of impairment profiles, possibly due to particular mid-level vision deficits rather than an aberrant general mechanism.

6.6.3 Methodological considerations

Some of the limitations of the individual chapters have been reported in the previous sections. However, some general methodological considerations may also apply. First, measures of reliability and validity of our tasks were not formally reported and neither stated explicitly. Overall, good convergent validity was implied as performance on the face trials in our tasks correlated with diagnostic tests (i.e., the CFMT, the CFPT, and the PI20). In particular, correlations with the PI20 suggested that what we measured with our tests was related to people's actual difficulties outside the laboratory. Validity of the tasks presented in Chapter 2 was assessed by additional correlational analyses conducted post hoc. Participants' performance on the emotion recognition task in Experiment 1 correlated with their performance on the emotion recognition task in Experiment 2 ($r = 0.43$, $p = .009$). The delayed matching task presented in Chapter 3 had good convergent validity as performance in our face and car conditions correlated with our diagnostic measures of face and car perception, respectively (Table 3.2). Finally, the validity of the delayed match-to-sample task presented in Chapter 5 was implied by strong correlations observed in the combined sample between participants' face matching accuracy and their scores on the CFMT ($r = .67$, $p < .001$), but not the CCMT ($r = .25$, $p = .14$). Conversely, car matching accuracy correlated with scores on the CCMT ($r = .55$, $p < .001$), but not the CFMT ($r = .29$, $p = .082$). We were also able to assess post hoc the internal reliability of the matching task in Chapter 5 by analysing the correlations between observers' short- and long-interval matching accuracy (faces: $r = .810$; cars: $r = .689$), and between their constant- and different-viewpoint matching accuracy (faces: $r = .846$; cars: $r = .743$), which are equivalent to split-half measures of reliability.

In the light of these considerations, it is crucial for future research to use reliable tasks. The employment of sensitive psychophysical measures could be a useful direction, especially when trying to avoid ceiling performance which may hinder more subtle impairments (not uncommon in DP). Assuring good reliability is especially important when interpreting null results. As discussed above, apparent dissociations between performance on the CFMT (a task with greater reliability) and the CFPT have often been taken as proof of a disproportionate impairment of face memory versus face perception, inflating the putative heterogeneity of the DP population.

Another interesting post hoc analysis would have been looking at individual profiles considering DP participants who took part in all studies presented in this thesis. Regrettably, none of the participants participated in all the four studies. Our research is constrained by the availability of our participants, who generously give their time to take part in the studies. Nevertheless, future research is encouraged to test many DP participants on a range of measures. Previous studies who assessed prosopagnosics on several different tests had a limited number of participants, but they have been highly influential for the DP literature (e.g. Le Grand et al., 2006; Duchaine, Yovel, Butterworth, & Nakayama, 2006).

A further consideration concerns the way we analysed some of the correlations presented in the chapters. All the correlations of principle interest were significant at $p < .001$ and comfortably survived correction for multiple comparisons. Moreover, many of the correlations reported were independent of one another and thus did not increase type-I error rate.

However, some may argue that our correlations were inflated as the range of abilities was broader than in the general population given that we collapsed the data obtained from controls with those obtained from individuals with DP. Therefore, the strength of the correlation would not have emerged if the same task was given to a group of people randomly sampled from the general population. Crucially, we are not trying to present results as something it would be naturally observed in the general population. Our aim is to use individual differences to examine whether a functional relationship exists between X and Y, rather than estimating the strength of the correlation seen between X and Y in the typical population.

Finally, some of our studies lacked control tasks. For example, it would have been useful to add a visual perception task in the emotion recognition study presented in Chapter 2, in which participants had to recognise non-face objects drawn from morph continua. Performance on this task may have been beneficial to exclude any potential domain-general visual perception deficit in our DP sample. Instead, we relied on DPs' performance on the Cambridge Car Memory Test (CCMT) and the Cambridge Bike Memory Test (CBMT) to assume typical general visual perception.

6.6.4 Considerations on diagnostic criteria

A standard diagnostic battery for the assessment of DP does not currently exist and different research laboratories are free to use different tests to recruit DP participants. In the studies reported here diagnostic decisions were typically based on the CFMT, the CFPT, and the PI20. However, while performance on the CFMT and the PI20 were taken as the main proof for a diagnosis, results on the CFPT were usually reported to provide an indication of whether a DP was apperceptive or mnemonic. While the use of the CFMT is conventional, results from the CFPT and the PI20 are controversial.

Some authors have queried the CFPT because of its poor reliability and likelihood to yield to noisy performance. These limitations are mainly due to its format which may render it more susceptible to the use of compensatory strategies, compared to the

CFMT. Therefore, if the CFPT lacks the reliability necessary to diagnose someone as DP or not DP, we should be cautious about making apperceptive versus non-apperceptive classifications on this basis. Future research needs to generate a new measure of face perception, with better psychometric properties and less space for the use of compensatory strategies.

The employment of the PI20 in the diagnostic practice of DP has also been debateable. The use of self-reports of face recognition difficulty has been discouraged based on evidence that people have little insight into their ability to recognise faces (e.g., Bowles et al., 2009; McGugin, Richler, Herzmann, Speegle, & Gauthier, 2012; but see Palermo et al., 2017). Moreover, some authors have erroneously implied that the correlation found in the original study between the PI20 and the CFMT (Shah et al., 2015) was an estimate of the relationship between PI20 scores and face recognition ability in the general population (e.g., Bate & Tree, 2017; Bobak, Pampoulov, & Bate, 2016; Palermo et al., 2017). However, the original study employed a sample with an incidence rate of DP (21%) which was clearly higher than the one observed in the general population, in order to test the relationship between PI20 scores and CFMT results across the entire range of abilities and hence validate the PI20 as a diagnostic instrument. This aspect contributed in increasing the strength of the correlation observed, as the sample used presented a distribution of abilities which is not typically found in the general population. However, a follow-up study using a narrower range of performances, confirmed the existence of a significant relationship between PI20 scores and performance on the CFMT (Gray, Bird, & Cook, 2017).

Crucially, we never included our DP participants based exclusively on self-report evidence. Instead, we used PI20 scores as a diagnostic complement to objective computer-based tests, which can sometime yield to imprecise evidence. For example, variables such as lack of motivation, boredom, test anxiety, and poor mouse control have been reported to affect the execution of these tests. Furthermore, the use of compensatory strategies may lead to typical execution of diagnostic tests in individuals with a lifelong history of face recognition difficulties.

Non-face recognition deficits in our participants have been assessed using the CCMT as this measure is matched in format and difficulty with the CFMT, providing a good comparison with face recognition difficulties. However, we recognise that the CCMT alone may be too restrictive to investigate the complexity of non-face object recognition. As recently suggested by some authors, future research should include tests which assess a wider range of different non-face objects (see Geskin & Behrmann, 2017). Moreover, including measures of mid-level vision may also be extremely interesting as it may unveil specific difficulties which would reflect in high-level vision (e.g., a specific deficit at perceiving curvature, but intact perception of straight lines, may affect the perception of objects such as flowers and cars, but not buildings).

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