Reading in Developmental Prosopagnosia

Evidence for a Dissociation Between Word and Face Recognition

Starrfelt, Randi; Klargaard, Solja; Petersen, Anders; Gerlach, Christian

Published in:
Neuropsychology

DOI:
[10.1037/neu0000428](https://doi.org/10.1037/neu0000428)

Publication date:
2018

Document version
Peer reviewed version

Citation for published version (APA):
Starrfelt, R., Klargaard, S., Petersen, A., & Gerlach, C. (2018). Reading in Developmental Prosopagnosia: Evidence for a Dissociation Between Word and Face Recognition. *Neuropsychology*, 32(2), 138-147.
<https://doi.org/10.1037/neu0000428>

Paper in press *Neuropsychology*

©American Psychological Association, [2017]. This paper is not the copy of record and may not exactly replicate the authoritative document published in the APA journal. Please do not copy or cite without author's permission.

Reading in Developmental Prosopagnosia:

Evidence for a Dissociation Between Word and Face Recognition.

Randi Starrfelt ^{*,1}, Solja K. Klargaard ², Anders Petersen ¹, & Christian Gerlach ²

¹ Department of Psychology, University of Copenhagen, Copenhagen, Denmark

² Department of Psychology, University of Southern Denmark, Odense, Denmark

* Corresponding author: Randi Starrfelt
Dept. of Psychology
University of Copenhagen
O Farimagsgade 2a
DK-1351 Copenhagen K
Denmark
Email: Randi.Starrfelt@psy.ku.dk

Author note

Acknowledgements

This project was supported by a grant from the Danish Research Council for Independent Research | Humanities (DFF – 4001-00115) to CG. and RS. We wish to thank Brad Duchaine for sharing the Cambridge Face Memory test and the Cambridge Face Perception Test, Romina Palermo and Jon Brock for sharing the Faces and Emotions Questionnaire, and Simon Nielsen for providing the text-reading test. We are grateful to the participating subjects and thank Fakutsi for face value.

Abstract

Objective: Recent models suggest that face and word recognition may rely on overlapping cognitive processes and neural regions. In support of this notion, face recognition deficits have been demonstrated in developmental dyslexia. Here we test whether the opposite association can also be found, that is, impaired reading in developmental prosopagnosia.

Method: We tested 10 adults with developmental prosopagnosia and 20 matched controls. All participants completed the Cambridge Face Memory Test, the Cambridge Face Perception test and a Face recognition questionnaire used to quantify everyday face recognition experience. Reading was measured in four experimental tasks, testing different levels of letter, word, and text reading: a) single word reading with words of varying length, b) vocal response times in single letter and short word naming, c) recognition of single letters and short words at brief exposure durations (targeting the word superiority effect), and d) text reading.

Results: Participants with developmental prosopagnosia performed strikingly similar to controls across the four reading tasks. Formal analysis revealed a significant dissociation between word and face recognition, as the difference in performance with faces and words was significantly greater for participants with developmental prosopagnosia than for controls.

Conclusions: Adult developmental prosopagnosics read as quickly and fluently as controls, while they are seemingly unable to learn efficient strategies for recognizing faces. We suggest that this is due to the differing demands that face and word recognition put on the perceptual system.

Keywords: developmental prosopagnosia; reading; word recognition; face recognition

Introduction

Developmental prosopagnosia (DP) is a syndrome characterized by severely impaired face recognition, estimated to affect 2 - 2.5% of the population (Bowles et al., 2009; Kennerknecht et al., 2006). People suffering from DP fail to develop normal face recognition abilities and can have difficulties even in recognizing their own immediate family. While DP is still a relatively little known syndrome, an increase in studies has been seen over the last decade or so.

Neuropsychological studies have tried to delineate what perceptual or cognitive process(es) might be affected to selectively or disproportionally impair face recognition in DP, and there is growing consensus that a deficit in holistic or configural processing might be the core deficit (Liu & Behrmann, 2014; Palermo et al., 2011). A core question concerning DP is whether the syndrome affects face processing only, or if other visual recognition processes are also impaired (e.g., Duchaine & Nakayama, 2005; Gerlach, Klargaard, & Starrfelt, 2016). The relative selectivity of deficits is a general issue in developmental disorders, and is also debated in relation to more well-known syndromes like developmental dyslexia. Interestingly, a recent study has shown that participants with developmental dyslexia also show deficits in face and object recognition, and the authors conclude that these deficits did not result from problems with configural processing as in prosopagnosia (Sigurdardottir, Ivarsson, Kristinsdottir, & Kristjansson, 2015). Other studies of face recognition in developmental dyslexia have, however, indicated the presence of a dissociation between dyslexic / impaired reading and preserved face recognition (e.g., Smith-Sparke & Moore, 2009)

This debate about the selectivity of the deficit in developmental prosopagnosia and dyslexia, as well as the perceptual or cognitive processes involved, has clear parallels in the literature on acquired visual agnosia following brain injury (Behrmann & Plaut, 2014; Farah, 2004). In *pure alexia*, visual word processing is impaired, while identification of faces may be relatively

spared, and in acquired *prosopagnosia*, face recognition is impaired, while reading is commonly reported to be unaffected. This constitutes a double dissociation and suggests that words and faces are processed by different mechanisms and brain areas; or at least this is current textbook knowledge (Gazzaniga, Ivry, & Mangun, 2013). Processing of words and faces has been linked to specialized perceptual brain areas in ventral occipito-temporal cortex, lateralized to different hemispheres; the visual word form area (VWFA; Cohen et al., 2000) in the left fusiform gyrus, and the fusiform face area (FFA; Kanwisher, McDermott, & Chun, 1997) in the right hemisphere. The double dissociation between reading and face processing, and the modular theories of perception that often go with it, have, however, recently been challenged by studies suggesting a relationship between reading and face recognition (Behrmann & Plaut, 2014; Dundas, Plaut, & Behrmann, 2013, 2014; Roberts et al., 2015). The greatest challenge to traditional, modular views comes from a study showing that patients with acquired prosopagnosia show evidence of reading deficits, while patients with pure alexia show evidence of face processing deficits (Behrmann & Plaut, 2014), suggesting that the two functions rely on shared cerebral resources.

A related line of studies have looked at the impact of reading development on brain organization. Perhaps the most intriguing results come from studies showing that learning to read affects the cerebral substrate for face processing in a systematic way, and may even drive the lateralization of face processing to the right hemisphere (Dehaene & Cohen, 2007; Dehaene et al., 2010; Dundas et al., 2013). Similarly, with increasing reading proficiency, right hemisphere activation for words decreases (Turlington, Gareau, Flowers, Zeffiro, & Eden, 2003). Of course, the direct effect of learning to read on brain structure and function is hard to disentangle from cerebral maturation, but studies of people learning to read as adults indicate that the hemispheric shift in face selective areas is a result of reading acquisition, as it also occurs in adults learning to read (Dehaene et al., 2010)

On this basis, it becomes interesting to investigate whether a developmental deficit in face recognition also affects reading development. We test this on a behavioural level by assessing reading of letters, words, and text in 10 subjects with DP and a group of matched controls. If face processing and reading are dissociable processes, we would expect the group of developmental prosopagnosics – or at least some of them – to perform normally on reading tests. If, on the other hand, a normal development of the cognitive and cerebral processes involved in face recognition is necessary for acquisition of fluent reading, then reading may be impaired in DP. To ensure that reading skills were measured sensitively, we included both experimental and psychophysical tests tapping different levels of visual and lexical processing. In three experiments, we test 1) The word length effect and naming latency for single words of different lengths; 2) The word superiority effect; 3) Text reading speed and comprehension.

While the reading latency of normal readers is little affected by the number of letters in a word, a *word length effect* (WLE) is a common symptom of both developmental and acquired reading disorders (Barton, Hanif, Björnström, & Hills, 2014). Word length effects are also characteristic in beginning readers, and a drastic reduction of this effect characterizes successful reading acquisition. In pure alexia, RTs may increase with hundreds of milliseconds per additional letter in a word, and indeed the word length effect is often considered a defining feature of this syndrome (Starrfelt & Shallice, 2014). Smaller, but abnormal word length effects have also been reported in some patients with acquired prosopagnosia (Behrmann & Plaut, 2013; Petersen et al., 2016), but normal reading latency and word length effects have been demonstrated in other such patients (Hills, Pancaroglu, Duchaine, & Barton, 2015; Susilo, Wright, Tree, & Duchaine, 2015). A recent study (likely conducted simultaneously with the present one) (Rubino, Corrow, Corrow, Duchaine, & Barton, 2016) showed that a group of DPs did not show abnormal word length effects

or reading RTs. Because an abnormal WLE is a strong indication of the presence of a reading disorder, we test this effect, as well as single word reading RTs in Experiment 1.

Fluent reading is characterised by fast and parallel processing of letters in words; a likely explanation for the minimal WLE in proficient readers. Indeed, normal readers typically identify letters in words faster than they identify single letters, a phenomenon known as the *word superiority effect* (WSE) (Reicher, 1969; Wheeler, 1970), which is what we investigate in Experiment 2. We have recently developed a psychophysical paradigm for measuring the WSE comparing short words and single letters, and found that the effect is robust over a range of exposure durations in healthy participants (Sand, Habekost, Petersen, & Starrfelt, 2016; Starrfelt, Petersen, & Vangkilde, 2013). Patients with pure alexia, which is thought to be caused by a breakdown in parallel letter processing, did not show a WSE in this paradigm (Habekost et al., 2014). In typical experiments testing the WSE, stimuli are presented very briefly and then masked, and the classical explanation for the effect is that there is more top-down support from lexical representations for word processing than for single letters or strings of unrelated letters (McClelland & Rumelhart, 1981). Because the experiment uses very brief exposure durations, it is visually a very challenging task that should be sensitive even to subtle deficits in visual word processing in the DP group.

The last experiment (Experiment 3) is more straightforward, and simply tests text reading speed and comprehension. Here, we are interested in measuring whether the DPs can read a text as quickly as controls, and if they understand what they read at the same level. In sum, we measure reading performance in a variety of ways designed to tap both visual recognition of letters and words, reading aloud, and text reading speed and comprehension.

General Method

Participants.

All participants provided written informed consent according to the Helsinki declaration to participate in the project. The Regional Committee for Health Research Ethics in Southern Denmark has assessed the project, and ruled that it did not need formal registration.

Participants with developmental prosopagnosia (DPs).

Following coverage of DP in Danish media, we have been contacted by a number of people complaining of face recognition problems. They all report life long difficulties recognizing friends, colleagues, and sometimes even close family members and themselves by their faces.

10 of these subjects are included in the current study. Inclusion was based on abnormal performance, defined as 2SD's below the mean of a matched control group (see below,) on the Cambridge Face Memory Test (CFMT; Duchaine & Nakayama, 2006), the most commonly used diagnostic test for prosopagnosia. In addition, all included DP's also report severe difficulties with face recognition in their everyday life, as evaluated by the face recognition part (29 items) of the Faces and Emotion Questionnaire (FEQ; Freeman, Palermo, & Brock, 2015). Three DPs were left handed and all performed within the normal range on The Autism-Spectrum Quotient (AQ) questionnaire (Baron-Cohen, Wheelwright, Skinner, Martin, & Clubley, 2001). 7/10 DPs had a structural MRI, which showed no structural abnormalities. See Table 1 for an overview of age, gender, and basic test scores. The DPs did not receive remuneration for their participation in this study. The DPs (and controls) included in this study have also participated in other studies, and their performance in object recognition (Gerlach et al., 2016) and topographic memory tasks (Klargaard, Starrfelt, Petersen, & Gerlach, 2016), have previously been reported. We report here all

tests where reading is measured. For the DPs to be anonymous, and yet recognizable across publications, their original project subject-numbers are kept in the text and tables.

Table 1. Age, gender and performance (raw scores) on the Cambridge Face Memory Test (CFMT), the Cambridge Face Perception Test (CFPT), and the Face recognition questionnaire (FEQ) for the 10 participants with developmental prosopagnosia, and the mean and SD for the controls' scores on these tests. Values in boldface designate performance deviating more than 2 SDs from the mean of the matched control group. In the CFMT, a low score indicates a deficit, while in the CFPT and in the FEQ a high score indicates a deficit. The maximum score on the FEQ is 87. The MRI column indicates if the participant has been scanned.

Subject	Age	Gender	Handedness	CFMT	CFPT	FEQ	MRI
PP04	57	M	Right	37	86	71	no
PP07	40	F	Right	41	60	66	yes
PP09	40	F	Left	43	70	52	no
PP10	34	F	Right	33	58	62	yes
PP13	51	M	Right	35	42	64	yes
PP16	23	F	Left	39	64	54	yes
PP17	49	F	Right	35	88	56	yes
PP18	38	F	Left	30	78	69	yes
PP19	16	M	Right	33	48	53	yes
PP27	25	M	Right	42	66	59	no
Control mean (SD)				59.1 (7.9)	41.3 (11.4)	22.4 (11.4)	

Control subjects.

We compared the 10 DPs with 20 control subjects; two matched on age, gender, and educational level to each individual with DP (three left handed). Thus the groups were comparable in terms of age (DP $M = 37$, range = 16-57; Control $M = 37$, range = 16-56) and years of education (DP $M = 15.5$, range = 11-17; Control $M = 15.2$, range = 10-17). All controls performed within the normal range on the CFPT and the CFMT, evaluated by the Bowles (2009) norms. Controls received gift certificates of ~120 DKK (~20 USD) per hour for their participation.

The primary group statistical analyses below are based on confidence intervals (CI) and their degree of overlap (Cumming, 2014) but null-hypothesis testing significance values (p) are also provided. Individual data for the DPs and summary data for controls in all experiments are presented in the Supplementary table S1, where individual test scores significantly different from controls based on single case statistics are also highlighted.

Experimental Investigation

Experiment 1. Reading latency and word length effects.

Stimuli and procedure.

In this experiment, we tested whether our sample with DP showed a word length effect (WLE) in single word reading, and whether RTs in single word reading were elevated. The exact same paradigm has previously revealed elevated RTs and WLEs in patients with pure alexia (Habekost, Petersen, Behrmann, & Starrfelt, 2014; Starrfelt, Nielsen, Habekost, & Andersen, 2013). Stimuli were 150 words of 5–7 letters (50 of each length matched for word frequency and neighbourhood-size). Reading RTs were measured by a voice key (a microphone connected to a response box). The WLE is calculated using linear regression, where the slope represents the

additional time needed per additional letter in a word. Mean overall RT was also calculated for each subject.

Results.

Voice key errors (setting of the microphone too early /late) were excluded from the analysis. RTs were analysed for correct trials only. RTs were trimmed by excluding RTs from trials deviating more than 2.5 SD for each individual at each word length. On average 4.9% (range: 1.3 - 7.3) of the trials for each DP was removed due to voice key errors or trimming. For the control participants it was 3.3% (range: 0.7 - 6).

The DP-group made on average 1% errors (range: 0 - 5%) whereas the control group made on average 0.5% errors (range: 0 - 3%). The mean WLE for the DP group was 10 ms (95% CI = [2, 19]) and 11 ms (95% CI = [6, 16]) for controls. The mean correct reading RT for the DP group was 569 ms (95% CI = [519, 619]) and 521ms (95% CI = [489, 553]) for controls. The overlap in the CIs of the two groups indicate they do not differ reliably in WLE ($t_{28} = 0.11, p = .91$) or in latency ($t_{28} = -1.81, p = .08$). See also Figure 1.

Figure 1. The individual word length effects for each control and DP subject (grey circles), and the grand mean (black symbols) and 95% CI of the grand mean for each group (panel a). Panel b shows the mean reading RT for each participant (grey circles) and the grand mean and CIs for developmental prosopagnosics ($N = 10$) and control participants ($N = 20$).

a)

b)

Experiment 2. Word superiority effect.

Stimuli and procedure.

This paradigm consists of two experiments, an RT task for practising the stimuli (Experiment 2a) and a psychophysical task using limited exposure durations (Experiment 2b), both described in detail in Starrfelt, Petersen, et al. (2013; Experiments 1 and 2). The stimuli are the same in both the RT and the psychophysical task; 25 letters ('w' excluded), and 25 three letter words. The words are confusable in the sense that none can be identified uniquely by seeing one letter only, and for most of the words all three letters must be processed for the word to be correctly identified (see Appendix in Starrfelt, Petersen, et al., 2013) .

Experiment 2a. Letter and word naming.

This is a computerised naming task and the procedure is similar to Experiment 1. The purpose of this RT-task is to familiarise subjects with the word stimuli employed in the psychophysical paradigm (Experiment 2b). Everyone knows the letters of the alphabet, and we wanted to ensure that the subjects also knew which words were used as stimuli. The letter and word conditions included 50 trials each, and were run separately, the letter task first. The task was to name the word or letter presented on the screen. There were ten practice trials in each condition.

Results.

Voice key and naming errors were recorded, and excluded from the RT analysis. RTs were trimmed for each participant by excluding RTs from trials deviating more than 2.5 SD from the individual mean. 2.9% (range: 0 - 8) and 1.3% (range: 0 - 4) of the trials for each DP were removed due to voice key errors or trimming for letters and words respectively. For the control

participants it was 1.2% (range: 0 - 4) and 0.6% (range: 0 - 4) respectively. 9 DPs participated (PP16, who did not complete the test, and the two controls matched to PP16 were excluded from the analyses).

The DP-group made no errors. The controls made on average 0.9% errors with letters (range: 0 - 6%) and 0.4% errors with words (range: 0 - 4%). For the DPs the mean RT to letters was 488 ms (95% CI = [450, 526]) and 451 ms (95% CI = [417, 484]) to words. For controls the mean RT to letters was 473 ms (95% CI = [452, 495]) and 466 ms (95% CI = [446, 486]) to words. The overlap in the CIs of the two groups indicate they do not differ reliably in latency for the letters ($t_{25} = -0.79, p = .44$) or words ($t_{25} = 0.90, p = .37$). See also Figure 2.

Figure 2. Naming time. Mean RTs to single letters and words in Experiment 2a for DPs ($n = 9$) and controls ($n = 18$). Grey circles represent individual mean RTs, black symbols and interval show grand mean and 95% CI for each group.

Experiment 2b. A psychophysical test of the word superiority effect.

This experiment tested identification of briefly presented single stimuli (letters or words in different blocks) flashed at the centre of the screen and followed by a pattern mask. There were 200 trials in each experimental block. In total, subjects ran 400 trials per stimulus type in an ABBA-design (letters first), and the first and second blocks for each stimulus type were preceded by 30 and 15 practice trials, respectively. In each trial, a single stimulus (word or letter) was chosen randomly and presented for one of ten exposure durations (10-100 ms, randomly intermixed). The stimulus was terminated by a pattern mask shown for 500 ms. Participants were instructed to make an unspeeded report of the stimulus, if they were “fairly certain” of its identity. Responses were recorded by the experimenter. To ensure foveal presentation, participants were required to focus on a centrally placed cross and then initiate the trial by pressing the left mouse button.

Results.

We first compared the proportion of correct responses averaged across the ten exposure durations for the two groups for letters and words respectively.

The average proportion correct responses across all exposure durations was 0.74 (95% CI = [0.66, 0.82]) for letters, and 0.85 (95% CI = [0.82, 0.88]) for words in the DP-group. Hence, the WSE was large for the DPs ($d_z = 1.3$; 95% CI $M_{dif} = [0.04, 0.17]$). For the controls, the average proportion correct responses across all exposure durations were 0.74 (95% CI = [0.69, 0.78]) for letters, and 0.82 (95% CI = [0.80, 0.85]) for words. Accordingly, the WSE was also large for the control participants ($d_z = 1.3$; 95% CI $M_{dif} = 0.05, 0.12$). As can be seen from Figure 3 there is considerable overlap in the CIs for the DP-group and the control participants for both letters and words, and there is no reliable difference between their performance in this experiment (Letters: $t_{25} = -1.21, p = .24$; Words: $t_{25} = -0.11, p = .91$).

Figure 3. Word superiority. Shows the overall proportion correct responses for briefly presented words and single letters in Experiment 2b for DPs ($n = 9$) and matched controls ($n = 18$). Grey circles represent individual accuracy scores, black symbols and interval show grand mean and 95% CI for each group. The mean exposure duration for both groups was 55 ms.

We then compared the mean proportion of correct responses for letters and words for each of the ten exposure durations. Because four DPs received a slightly different set of exposure durations (caused by a computer problem resulting in a lower screen refresh rate), only five DPs (PP04, PP07, PP09, PP17 & PP27) and their matched controls ($n = 10$) were included in this analysis. As can be seen from Figure 4 the two groups performed quite similarly across the ten exposure durations. Performance generally increases with increased exposure duration, and the WSE is mainly evident at exposure durations between 20 and 60 ms for both groups.

Figure 4. Mean proportion correct responses at the ten exposure durations for words and letters, in the DP-group (panel a) and controls (panel c) in Experiment 2b. Also shown is the mean difference in accuracy between words and letters (reflecting the word superiority effect) at the ten exposure durations for DPs (panel b) and control (panel d).

Experiment 3. Text reading.

Stimuli and procedure.

The test was adapted from the standard 9th grade Danish reading tests (see Nielsen & Wilms, 2015 for a full description), and the main measure was reading time (in seconds). A text of 637 words was presented on the screen of a laptop computer. Instructions were to read the text carefully, as one would be required to answer questions about the text immediately after reading it.

The text was a popular scientific text from a biology book¹. Due to the length of the text, the reading time measure in this test includes a click on the mouse to move the text forward. Subjects also clicked the mouse to indicate that they had finished reading the text, and this was recorded as their RT. Immediately after this, four multiple choice questions about the content of the text were presented.

Results.

Two of the DPs (PP04 and PP16) and two control participants (one for PP09 and one for PP10) did not perform the test. Hence, the DP group comprised 8 subjects and the control group 14 subjects.

The mean reading speed of the DPs was 213 s (95% CI = [178, 247]) and 221 s (95% CI = [191, 251]) for the control participants, which is not reliably different ($t_{20}=0.37, p = .71$). In terms of accuracy on the text comprehension questions (four multiple choice questions), the DPs obtained a median score of 2 (range: 2-4) and the control participants a median score of 3 (range: 1-4), which is not reliably different (Mann-Whitney, $Z = -0.55, p = .62$). Hence, the performance of DPs and the control participants was quite alike in terms of both speed and comprehension.

Null finding or dissociation?

The overall results from this investigation shows that participants with DP, who are severely impaired in face recognition, perform on level with controls in four different reading tasks. Thus, there seems to be a dissociation between impaired face recognition and preserved reading. Such a dissociation has traditionally been considered strong evidence for independence of the underlying cognitive processes within cognitive neuropsychology (Shallice, 1988). It is also clear,

¹ One DP-participant was presented with a different text from the same book, matched in length and difficulty, by mistake.

however, that proving “normal performance” is more difficult than proving a deficit. The individual test-scores (see figures 1-3 and Supplementary table S1) show that none of the DPs had a general deficit in reading, and one DP even performed significantly superior to controls on a measure of visual word recognition. However, in order to provide positive evidence for a dissociation, a formal analysis is needed. To test whether there is a significant dissociation between face recognition and reading in the DP group, we used a method suggested by Crawford and colleagues (Crawford, Blackmore, Lamb, & Simpson, 2000). This analysis (implemented in the program *DiffDef.exe*) specifically tests whether there is evidence of a differential deficit (a dissociation) in a clinical sample compared to controls. As the core test of face recognition, the CFMT, is based on accuracy rather than RTs, we thought it most appropriate to compare face recognition performance on this test with a reading measure based on accuracy. We thus chose to test if there was evidence of a dissociation between accuracy in visual word recognition (overall correct score in Experiment 2b) and accuracy in the CFMT in the group of DPs.

Results

The following measures were used in the analysis: The correlation between group (control vs. DP) and WordAccuracy (Exp 2b) $-.178$; the correlation between group and CFMT $= .868$; and the correlation between WordAccuracy and CFMT scores in the whole sample (DP and controls $N = 27$) $= .06$. This yields a $t_{24} = -7.0$, with p (two-tailed) < 0.00001 , indicating a very clear dissociation between results on the two measures for the DPs. As mentioned above, three DPs received slightly offset exposure durations in Experiment 2b. Although the mean (SD) of the exposure durations in this group were similar to the controls and other DPs (mean correct exposure 55 ms, $SD=0.01$; mean offset exposure 55.8 ms, $SD=0.06$), we found it appropriate to also perform this analysis including only the DPs receiving the exact same exposure durations as controls (i.e.

using the same data from Experiment 2 as reported in Figure 4). Here, the correlation between group and WordAccuracy (Exp 2b) was $-.062$; the correlation between group and CFMT = $.843$; and the correlation between WordAccuracy and CFMT scores in the whole sample (DP and controls $n = 15$) = $.06$. This yields a $t_{12} = -3.946$ with a p (two-tailed) < 0.002 , again indicating a clear dissociation between face and word recognition performance in the DP group.

Figure 5. Performance on Cambridge Face Memory Test and Word recognition accuracy (Experiment 2b) for individual subjects, showing the clear dissociation between face and word recognition in the DP group. The bold lines represent the mean score of the control group; the dotted lines show 2 SDs below the mean of the control group.

This finding strongly suggests functional independence between the two domains: Face recognition is impaired while word recognition is spared, and the DPs exhibit a difference between face and word recognition performance that far exceeds that found in the control sample. To further illustrate this dissociation, Figure 5 shows that while all DPs perform below 2 SDs of the control mean on the CFMT (this was a criterion for inclusion), none fall below 2 SDs of the control mean on the reading task. Rather, 7/9 DPs perform on level with the control mean or better in the word recognition task.

Discussion

The findings are clear; the participants with developmental prosopagnosia show no deficits in reading on any of the tests included. Their letter and word naming times are similar to controls, they show a normal word superiority effect, a normal (absence of a) word length effect in single word reading, and normal text reading speed and comprehension. As such, a clear dissociation between impaired face processing and preserved reading is evident in this group of DPs. This is also clearly demonstrated in a formal analysis that shows: (i) the DPs perform within the normal range on word recognition, (ii) they perform outside the normal range on face recognition, *and* (iii) the difference in the DPs performance with words and faces by far exceeds what can be expected in the normal population. This provides positive evidence for a dissociation between word reading and face recognition in this group. This lines up nicely with recent evidence from acquired prosopagnosia, where normal reading has also been demonstrated following unilateral lesions to right ventral temporo-occipital cortex (Hills et al., 2015; Susilo et al., 2015), as well as a recent demonstration of normal RTs and word length effects in DP (Rubino et al., 2016).

An important next question then, is whether this constitutes a single dissociation, or if a double dissociation can be demonstrated, where another group shows the opposite pattern of performance (impaired reading and intact face processing). This would indicate that reading and

face recognition, at least at some levels of processing, are independent. Looking at developmental reading disorders, a first glance at the literature indicates that face recognition and naming may be intact in developmental dyslexia (Russeler, Johannes, & Munte, 2003; Smith-Spark & Moore, 2009). This suggests a double dissociation between reading and face processing. However, there are also reports of deficits in various face tasks, as well as different brain activation patterns for faces in dyslexics (Monzalvo, Fluss, Billard, Dehaene, & Dehaene-Lambertz, 2012; Tarkiainen, Helenius, & Salmelin, 2003). A recent study has shown that adult dyslexics performed significantly below a matched control group on the Cambridge Face Memory Task (also used in our study), as well as other measures of face and object processing (Sigurdardottir et al., 2015), but suggest that different processes are impaired in dyslexic and prosopagnosic subjects. Specifically, they conclude that dyslexics' "holistic processing of faces appears to be intact, suggesting that dyslexics may instead be specifically impaired at part-based processing of visual objects" (Sigurdardottir et al., 2015, p. 739).

This explanation lines up nicely with the suggestion that a general deficit in global shape perception, affecting both faces and objects, is at the core of DP (Gerlach et al., 2016). There is ample evidence suggesting that face recognition is particularly dependent on some kind of holistic or configural processing, which requires not only the identification of the parts (eyes, nose etc.), but also the specific relations between these parts; the configuration (Tanaka & Gordon, 2011). The same is true for efficient object recognition, especially when fine grained discriminations are required (Gerlach, 2009). Words, on the other hand, are read by recognizing the letters in parallel (parts first) and not by identifying the outline or global shape of the word (Grainger, Dufau, & Ziegler, 2016; Grainger & Whitney, 2004; Pelli, Farell, & Moore, 2003). On this basis, it seems plausible that the face recognition problems in our sample of DPs could be caused (at least partly) by impaired processing of global shape. As global shape plays no part in

word recognition, reading is intact in these individuals despite the fact that word recognition is a perceptually demanding task.

Sigurdardottir et al. (2015) suggested that the face and object recognition deficits they observed in developmental dyslexia could be taken as evidence for ventral stream dysfunction. Their study, like ours, was a behavioural study, however, and does not speak directly to the cerebral underpinnings of the observed pattern of preserved and impaired functions. Behavioural results may, however, challenge or constrain such theories, however, if the proposed models cannot explain our findings. As mentioned in the introduction, deficits in word and face perception have been linked to specific areas in the visual ventral stream, and recent evidence has suggested that there may even be a developmental trajectory for hemispheric specialization that is at least partly caused by learning to read (Behrmann & Plaut, 2013; Cantlon, Pinel, Dehaene, & Pelphrey, 2011; Dundas et al., 2014; Dundas, Plaut, & Behrmann, 2015). The “neuronal recycling” hypothesis is so far the most specific regarding the relationship between the two processes (Dehaene et al., 2010). This hypothesis suggests that when we learn to read, some brain areas originally contributing to face processing are “recycled” to be used in visual word recognition, and results supporting this come from studies of both children and illiterates learning to read (Cantlon et al., 2011). Our results show that normal visual word recognition can be acquired in spite of a developmental impairment in face recognition. If it is correct that face recognition is bilaterally distributed from birth, but becomes more right lateralised with reading acquisition, then how might this work in DP to leave their reading functions unaffected? One possibility is that DP is a disorder characterised by cerebral dysfunction primarily in the posterior right hemisphere, perhaps parallel to the left hemisphere dysfunction seen in developmental dyslexia (Norton, Beach, & Gabrieli, 2015; Shaywitz, Lyon, & Shaywitz, 2006). This dysfunction in prosopagnosia thus affects the face processing network in the right hemisphere only or primarily, leaving the left hemisphere part of the network, where visual

word processing comes to be located, unaffected. To date, little is known about the cognitive correlates of the neuronal recycling process, and whether face recognition may deteriorate as a result of learning to read, and to our knowledge, no developmental studies have specifically addressed this question. Ventura et al. (2013) have, however, reported *superior* performance of literate compared to illiterate adults on a holistic face perception task, and claim that so far no clear behavioural consequences of literacy on face-recognition abilities have been documented

Another, but related, account of the relationship between visual word recognition and face recognition is the “many-to-many” account suggested by Behrmann & Plaut (2013). They propose that many posterior regions are necessarily engaged in the representation of multiple visual stimulus classes including words and faces, and that these regions form distributed but integrated large-scale circuits. They review evidence suggesting that the cerebral areas affected in DP are localised more anteriorly in the face processing network, and do not implicate the fusiform areas affected by learning to read. If the fusiform regions involved in face processing are intact in DP, then the cerebral competition between reading and face processing in these regions might also proceed normally. This would result in the pattern of performance we report, impaired face recognition (due to cerebral dysfunction outside the posterior fusiform regions), and normal reading. The many-to-many account is partly based on studies of brain injury patients, showing that that reading is affected in acquired prosopagnosia, and face recognition affected in pure alexia (Behrmann & Plaut, 2014), which is taken as evidence for shared, distributed networks being involved in both reading and face recognition. A recent study of acquired prosopagnosia has, however, suggested a modification of this claim: Hills, et al. (2015) showed that patients with prosopagnosia due to unilateral right hemisphere damage did not have impaired word recognition, evidenced by normal RTs and WLEs (see Susilo et al., 2015 for similar findings). The acquired prosopagnosics did, however, show deficits in discriminating font or handwriting, which may

suggest a hemispheric division of labour somewhat different from that originally suggested by Behrmann & Plaut (2013). Interestingly, when the same test of font discrimination was presented to participants with DP, no consistent deficits were found (Rubino et al., 2016).

In conclusion, our behavioural findings clearly suggests that face recognition can be impaired while reading is unimpaired, supporting a model where face and word recognition rely on cognitive and cerebral processes that are independent at least at some levels of processing. This conclusion is supported by other findings in both acquired and developmental prosopagnosia. Evidence for the opposite dissociation (impaired reading and preserved face recognition) seems less conclusive, although there are studies showing preserved face recognition in both developmental dyslexia (Smith-Spark & Moore, 2009) and pure alexia (Turkeltaub et al., 2014). Regardless of the neural processes involved, it is intriguing from a learning perspective that the developmental prosopagnosics can learn to read as fluently as normal subjects, while they are seemingly unable to learn efficient strategies for recognizing faces.

References

- Baron-Cohen, S., Wheelwright, S., Skinner, R., Martin, J., & Clubley, E. (2001). The autism-spectrum quotient (AQ): evidence from Asperger syndrome/high-functioning autism, males and females, scientists and mathematicians. *Journal of autism and developmental disorders*, 31(1), 5-17.
- Barton, J. J., S., Hanif, H. M., Björnström, L. E., & Hills, C. (2014). The word length effect in reading: A review. *Cogn Neuropsychol*(31), 378-412.
- Behrmann, M., & Plaut, D. C. (2013). Distributed circuits, not circumscribed centers, mediate visual recognition. *Trends Cogn Sci*, 17(5), 210-219. doi: 10.1016/j.tics.2013.03.007
- Behrmann, M., & Plaut, D. C. (2014). Bilateral hemispheric processing of words and faces: evidence from word impairments in prosopagnosia and face impairments in pure alexia. *Cereb Cortex*, 24(4), 1102-1118. doi: 10.1093/cercor/bhs390
- Bowles, D. C., McKone, E., Dawel, A., Duchaine, B., Palermo, R., Schmalzl, L., . . . Yovel, G. (2009). Diagnosing prosopagnosia: effects of ageing, sex, and participant-stimulus ethnic match on the Cambridge Face Memory Test and Cambridge Face Perception Test. *Cogn Neuropsychol*, 26(5), 423-455. doi: 10.1080/02643290903343149
- Cantlon, J. F., Pinel, P., Dehaene, S., & Pelphrey, K. A. (2011). Cortical Representations of Symbols, Objects, and Faces Are Pruned Back during Early Childhood. *Cerebral Cortex (New York, NY)*, 21(1), 191-199. doi: 10.1093/cercor/bhq078
- Cohen, L., Dehaene, S., Naccache, L., Lehericy, S., Dehaene-Lambertz, G., Henaff, M. A., & Michel, F. (2000). The visual word form area: spatial and temporal characterization of an initial stage of reading in normal subjects and posterior split-brain patients. *Brain*, 123 (Pt 2), 291-307.
- Crawford, J., Blackmore, L., Lamb, A., & Simpson, S. (2000). Is there a differential deficit in fronto-executive functioning in Huntington's disease. *Clinical Neuropsychological Assessment*, 1(4), 20.
- Cumming, G. (2014). The new statistics: why and how. *Psychol Sci*, 25(1), 7-29. doi: 10.1177/0956797613504966
- Dehaene, S., & Cohen, L. (2007). Cultural recycling of cortical maps. *Neuron*, 56(2), 384-398. doi: 10.1016/j.neuron.2007.10.004
- Dehaene, S., Pegado, F., Braga, L. W., Ventura, P., Nunes Filho, G., Jobert, A., . . . Cohen, L. (2010). How learning to read changes the cortical networks for vision and language. *Science*, 330(6009), 1359-1364.
- Duchaine, B., & Nakayama, K. (2005). Dissociations of face and object recognition in developmental prosopagnosia. *Journal of cognitive neuroscience*, 17(2), 249-261. doi: 10.1162/0898929053124857
- Dundas, E. M., Plaut, D. C., & Behrmann, M. (2013). The joint development of hemispheric lateralization for words and faces. *Journal of Experimental Psychology General*, 142(2), 348-358. doi: 10.1037/a0029503
- Dundas, E. M., Plaut, D. C., & Behrmann, M. (2014). An ERP investigation of the co-development of hemispheric lateralization of face and word recognition. *Neuropsychologia*, 61, 315-323. doi: 10.1016/j.neuropsychologia.2014.05.006
- Dundas, E. M., Plaut, D. C., & Behrmann, M. (2015). Variable Left-hemisphere Language and Orthographic Lateralization Reduces Right-hemisphere Face Lateralization. *Journal of cognitive neuroscience*, 27(5), 913-925. doi: 10.1162/jocn_a_00757
- Farah, M. J. (2004). *Visual agnosia*: The MIT press.
- Gazzaniga, M. S., Ivry, R. B., & Mangun, G. R. (2013). *Cognitive neuroscience: the biology of the mind* (Fourth edition. ed.). New York, N.Y.: W. W. Norton & Company, Inc.
- Gerlach, C. (2009). Category-specificity in visual object recognition. *Cognition*, 111(3), 281-301. doi: 10.1016/j.cognition.2009.02.005
- Gerlach, C., Klargaard, S. K., & Starrfelt, R. (2016). On the Relation between Face and Object Recognition in Developmental Prosopagnosia: No Dissociation but a Systematic Association. *PLoS ONE*, 11(10), e0165561. doi: 10.1371/journal.pone.0165561

- Grainger, J., Dufau, S., & Ziegler, J. C. (2016). A Vision of Reading. *Trends Cogn Sci*, 20(3), 171-179. doi: 10.1016/j.tics.2015.12.008
- Grainger, J., & Whitney, C. (2004). Does the human mind read words as a whole? *Trends in Cognitive Science*, 8(2), 58-59. doi: 10.1016/j.tics.2003.11.006
- Habekost, T., Petersen, A., Behrmann, M., & Starrfelt, R. (2014). From word superiority to word inferiority: visual processing of letters and words in pure alexia. *Cogn Neuropsychol*, 31(5-6), 413-436. doi: 10.1080/02643294.2014.906398
- Hills, C. S., Pancaroglu, R., Duchaine, B., & Barton, J. J. S. (2015). Word and text processing in acquired prosopagnosia. *Ann Neurol*, 78(2), 258-271. doi: 10.1002/ana.24437
- Kanwisher, N., McDermott, J., & Chun, M. M. (1997). The fusiform face area: a module in human extrastriate cortex specialized for face perception. *Journal of Neuroscience*, 17(11), 4302-4311.
- Kennerknecht, I., Grueter, T., Welling, B., Wentzek, S., Horst, J., Edwards, S., & Grueter, M. (2006). First report of prevalence of non-syndromic hereditary prosopagnosia (HPA). *American Journal of Medical Genetics Part A*, 140(15), 1617-1622. doi: 10.1002/ajmg.a.31343
- Klargaard, S. K., Starrfelt, R., Petersen, A., & Gerlach, C. (2016). Topographic processing in developmental prosopagnosia: Preserved perception but impaired memory of scenes. *Cogn Neuropsychol*, 1-9. doi: 10.1080/02643294.2016.1267000
- Liu, T. T., & Behrmann, M. (2014). Impaired holistic processing of left-right composite faces in congenital prosopagnosia. *Frontiers in Human Neuroscience*, 8, 750. doi: 10.3389/fnhum.2014.00750
- McClelland, J. L., & Rumelhart, D. E. (1981). An interactive activation model of context effects in letter perception: I. An account of basic findings. *Psychological Review*, 88(5), 375-407. doi: 10.1037/0033-295X.88.5.375
- Monzalvo, K., Fluss, J., Billard, C., Dehaene, S., & Dehaene-Lambertz, G. (2012). Cortical networks for vision and language in dyslexic and normal children of variable socio-economic status. *NeuroImage*, 61(1), 258-274. doi: 10.1016/j.neuroimage.2012.02.035
- Nielsen, S., & Wilms, I. L. (2015). Cognitive ageing on latent constructs for visual processing capacity: A novel Structural Equation Modelling framework with causal assumptions based on A Theory of Visual Attention. *Frontiers in Psychology*, 5. doi: 10.3389/fpsyg.2014.01596
- Norton, E. S., Beach, S. D., & Gabrieli, J. D. (2015). Neurobiology of dyslexia. *Curr Opin Neurobiol*, 30, 73-78. doi: 10.1016/j.conb.2014.09.007
- Palermo, R., Willis, M. L., Rivolta, D., McKone, E., Wilson, C. E., & Calder, A. J. (2011). Impaired holistic coding of facial expression and facial identity in congenital prosopagnosia. *Neuropsychologia*, 49(5), 1226-1235. doi: 10.1016/j.neuropsychologia.2011.02.021
- Pelli, D. G., Farell, B., & Moore, D. C. (2003). The remarkable inefficiency of word recognition. *Nature*, 423(6941), 752-756. doi: 10.1038/nature01516
- Petersen, A., Vangkilde, S., Fabricius, C., Iversen, H. K., Delfi, T. S., & Starrfelt, R. (2016). Visual attention in posterior stroke and relations to alexia. *Neuropsychologia*, 92, 79-89. doi: 10.1016/j.neuropsychologia.2016.02.029
- Reicher, G. M. (1969). Perceptual recognition as a function of meaningfulness of stimulus material. *J Exp Psychol*, 81(2), 275-280.
- Roberts, D. J., Lambon Ralph, M. A., Kim, E., Tainturier, M. J., Beeson, P. M., Rapcsak, S. Z., & Woollams, A. M. (2015). Processing deficits for familiar and novel faces in patients with left posterior fusiform lesions. *Cortex*, 72, 79-96. doi: 10.1016/j.cortex.2015.02.003
- Rubino, C., Corrow, S. L., Corrow, J. C., Duchaine, B., & Barton, J. J. (2016). Word and text processing in developmental prosopagnosia. *Cogn Neuropsychol*, 33(5-6), 315-328. doi: 10.1080/02643294.2016.1204281
- Russeler, J., Johannes, S., & Munte, T. F. (2003). Recognition memory for unfamiliar faces does not differ for adult normal and dyslexic readers: an event-related brain potential study. *Clin Neurophysiol*, 114(7), 1285-1291.
- Sand, K., Habekost, T., Petersen, A., & Starrfelt, R. (2016). The Word Superiority Effect in central and peripheral vision. *Visual Cognition*, 24(4), 293-303. doi: 10.1080/13506285.2016.1259192

- Shaywitz, B. A., Lyon, G. R., & Shaywitz, S. E. (2006). The role of functional magnetic resonance imaging in understanding reading and dyslexia. *Dev Neuropsychol*, 30(1), 613-632. doi: 10.1207/s15326942dn3001_5
- Sigurdardottir, H. M., Ivarsson, E., Kristinsdottir, K., & Kristjansson, A. (2015). Impaired recognition of faces and objects in dyslexia: Evidence for ventral stream dysfunction? *Neuropsychology*, 29(5), 739-750. doi: 10.1037/neu0000188
- Smith-Spark, J. H., & Moore, V. (2009). The representation and processing of familiar faces in dyslexia: differences in age of acquisition effects. *Dyslexia*, 15(2), 129-146. doi: 10.1002/dys.365
- Starrfelt, R., Nielsen, S., Habekost, T., & Andersen, T. S. (2013). How low can you go: Spatial frequency sensitivity in a patient with pure alexia. *Brain Lang*, 126(2), 188-192. doi: <http://dx.doi.org/10.1016/j.bandl.2013.05.006>
- Starrfelt, R., Petersen, A., & Vangkilde, S. (2013). Don't words come easy? A psychophysical exploration of word superiority. *Frontiers in Human Neuroscience*, 7, 519. doi: 10.3389/fnhum.2013.00519
- Starrfelt, R., & Shallice, T. (2014). What's in a name? The characterization of pure alexia. *Cogn Neuropsychol*, 31(5-6), 367-377. doi: 10.1080/02643294.2014.924226
- Susilo, T., Wright, V., Tree, J. J., & Duchaine, B. (2015). Acquired prosopagnosia without word recognition deficits. *Cogn Neuropsychol*, 32(6), 321-339. doi: 10.1080/02643294.2015.1081882
- Tanaka, J. W., & Gordon, I. (2011). Features, Configuration, and Holistic Face Processing. In A. J. Calder, G. Rhodes, M. H. Johnson & J. V. Haxby (Eds.), *The Oxford handbook of face perception* (pp. 177 - 194). New York: Oxford University Press.
- Tarkiainen, A., Helenius, P., & Salmelin, R. (2003). Category-specific occipitotemporal activation during face perception in dyslexic individuals: an MEG study. *NeuroImage*, 19(3), 1194-1204.
- Turkeltaub, P. E., Gareau, L., Flowers, D. L., Zeffiro, T. A., & Eden, G. F. (2003). Development of neural mechanisms for reading. *Nat Neurosci*, 6(7), 767-773. doi: 10.1038/nn1065
- Turkeltaub, P. E., Goldberg, E. M., Postman-Caucheteux, W. A., Palovcak, M., Quinn, C., Cantor, C., & Coslett, H. B. (2014). Alexia due to ischemic stroke of the visual word form area. *Neurocase*, 20(2), 230-235. doi: 10.1080/13554794.2013.770873
- Ventura, P., Fernandes, T., Cohen, L., Morais, J., Kolinsky, R., & Dehaene, S. (2013). Literacy acquisition reduces the influence of automatic holistic processing of faces and houses. *Neuroscience Letters*, 554, 105-109.
- Wheeler, D. D. (1970). Processes in word recognition. *Cognitive Psychology*, 1(1), 59-85. doi: [http://dx.doi.org/10.1016/0010-0285\(70\)90005-8](http://dx.doi.org/10.1016/0010-0285(70)90005-8)

Supplementary Table S1.

Subject	Age	Gender	Hand	CFMT	CFPT	FEQ	Exp1 RT (ms)	Exp1 WLE (ms)	Exp2a LetterRT (ms)	Exp2a WordRT (ms)	Exp2b Letter acc	Exp2b Word acc	Exp3 RT	Exp 3 Acc of 4
PP04	57	M	Right	37	86	71	525	2	507	432	0,61	0,84	na	na
PP07	40	F	Right	41	60	66	455	8	431	<u>377</u>	0,69	0,82	201	2
PP09	40	F	Left	43	70	52	713	16	487	478	0,68	0,76	217	2
PP10	34	F	Right	33	58	62	576	7	453	401	0,84	0,85	191	3
PP13	51	M	Right	35	42	64	573	33	446	431	0,69	0,86	226	2
PP16	23	F	Left	39	64	54	553	13	na	na	na	na	na	na
PP17	49	F	Right	35	88	56	508	-10	523	469	0,81	0,87	291	4
PP18	38	F	Left	30	78	69	616	5	587	495	0,63	0,85	150	2
PP19	16	M	Right	33	48	53	616	22	506	505	0,89	0,91	191	3
PP27	25	M	Right	42	66	59	555	7	452	466	0,84	0,88	236	2
Ctrl mean (sd)	37 (12)			59.1 (7.9)	41.3 (11.4)	22.4 (11.4)	521 (68)	10.7 (10.9)	473 (44)	465 (40)	0.73 (.09)	0.82 (.05)	221 (52)	2.6 (.8)