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SHORT REPORT

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Topographic processing in developmental prosopagnosia: Preserved perception but impaired memory of scenes

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ABSTRACT

Anecdotal evidence suggests a relation between impaired spatial (navigational) processing and developmental prosopagnosia. To address this formally, we tested two aspects of topographic processing – that is, perception and memory of mountain landscapes shown from different viewpoints. Participants included nine individuals with developmental prosopagnosia and 18 matched controls. The group with developmental prosopagnosia had no difficulty with topographic perception, but was reliably poorer in the retention of topographic information. Additional testing revealed that this did not reflect a general deficit in visual processing or visual short-term memory. Interestingly, a classical dissociation could be demonstrated between impaired face memory and preserved topographic memory in two developmental prosopagnosics. We conclude that impairments in topographic memory tend to co-occur with developmental prosopagnosia, although the underlying functions are likely to be independent.

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Developmental prosopagnosia; face recognition; short-term memory; topographical orientation

Introduction

Developmental prosopagnosia (DP) is a lifelong disorder where the ability to recognize faces never fully develops. Reported problems range from not being able to recognize familiar colleagues, to not being able to recognize family members and, for some, their own face. Recent studies indicate that DP has a prevalence around 2.5% (Bowles et al., 2009; Kennerknecht et al., 2006; Kennerknecht, Ho, & Wong, 2008), and it is often found to run in families (Kennerknecht et al., 2006). Importantly, DP is dissociated from other developmental disorders that affect intellectual and social functioning (Duchaine, Murray, Turner, White, & Garrido, 2009), such as autism spectrum disorders (Dawson et al., 2002) and Turner's syndrome (Mazzola et al., 2006). However, more subtle impairments in perception and/or memory are often revealed (Behrmann & Avidan, 2005), and an ongoing controversy concerns the selectivity of the disorder (Gerlach, Klargaard, & Starrfelt, 2016).

Several case studies have provided anecdotal evidence of navigational problems in DP (De Haan & Campbell, 1991; Duchaine, Parker, & Nakayama, 2003; Grueter et al., 2007). Correspondingly, Iaria and Barton (2010) tested individuals with developmental topographical disorientation, and found that 6/9 also showed impaired performance with face stimuli. Corrow et al. (2016) recently reported a study of subjects with acquired ($N = 10$) and developmental prosopagnosia ($N = 7$) on a battery of topographic tests, including two landmark recognition tests (house and scene recognition) and two tests of route learning (the road map test and a test of cognitive map formation). They found that patients with acquired prosopagnosia with occipitotemporal lesions were frequently impaired in landmark recognition as well as route learning, while patients with more anterior lesions were only impaired in the former. Intriguingly, individuals with the developmental type of prosopagnosia showed normal outcome on these tests, with the exception of one DP who was impaired on the cognitive map formation. The authors rightfully claim that their results indicate that the developmental form of prosopagnosia is more face-selective than its acquired counterpart (Corrow et al., 2016). Thus, there seems to be a disparity between the anecdotal reports of navigational problems in DP and their normal performance on the different topographic tests used by Corrow et al. (2016). However, their test battery only contains measures from two categories of

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navigational strategies (Aguirre & D'Esposito, 1999) – that is, (a) landmark (or place) recognition, where specific locations are recognized, and (b) route learning, where sequential landmark-instruction paths are to be remembered. A third category, and the focus of this paper, is the recognition of metric representations of space, in which map-like representation preserve angle and distance relations between landmarks (Aguirre & D'Esposito, 1999). It can be argued that encoding and retrieval of surface configurations are important for recognition of both faces and landscapes, in that both entail the discrimination of invariant metric surface properties. In this study, we evaluate the ability in DP to encode and remember such surface configurations – that is, invariant metric representations of landscapes – using the *Four Mountains Test*, developed by Hartley et al. (2007). The Memory subtest of the Four Mountains Test has proved sensitive to hippocampal as well as extra-hippocampal volume in healthy adults (Hartley & Harlow, 2012).

General method

Participants

Nine subjects with DP and 18 typically developing controls were included in this study. All subjects had normal (or corrected-to-normal) vision, no learning disability, and no known history of neurological damage or psychiatric illness. All participants provided written informed consent according to the Helsinki declaration. The Regional Committee for Health Research Ethics of Southern Denmark evaluated the project as not requiring formal registration.

Participants with developmental prosopagnosia (DPs)

All DPs independently contacted us with subjective concerns about their ability to recognize faces. They completed structured interviews regarding their everyday difficulty with facial recognition and family history. All reported difficulties recognizing friends, colleagues, and sometimes close family members and themselves by their faces, and these problems had been present throughout their life.

As a first screening for DP, we used the *Cambridge Face Memory Test* (CFMT; Duchaine & Nakayama, 2006) and the *Cambridge Face Perception Test* (CFPT; Duchaine, Germine, & Nakayama, 2007), kindly provided by Brad Duchaine and translated into Danish. The inclusion criterion was initially performance below 2 standard deviations on CFMT or CFPT compared to the age- and gender-adjusted norms provided by Bowles et al. (2009).

All included participants also completed the first part of the Faces and Emotion Questionnaire (FEQ; 29 items; Freeman, Palermo, & Brock, 2015), translated into Danish by the last author. The final inclusion criterion for DP was a performance on CFMT and the FEQ (indexing everyday difficulties with face recognition) below 2 standard deviations of that of the matched control group. As seen in Table 1, the performance of 5/9 DPs was also 2 standard deviations below the control mean on the CFPT. All DPs performed within the normal range (score of 32 or more) on the *Autism-Spectrum Quotient* (AQ; Baron-Cohen, Wheelwright, Skinner, Martin, & Clubley, 2001). DPs did not receive remuneration for their participation in this study. We have kept their project subject-numbers in text and tables in order to enable comparisons across publications.

Control subjects

Two controls were matched for each DP on age, gender and educational level, making the groups comparable in terms of age (DP: $M = 38.9$ years, range = 16–57; control: $M = 37.8$ years, range = 16–56) and years of education (DP: $M = 16.1$, range = 13–17; control: $M = 15.3$, range = 10–17). All controls performed within the normal range on the CFPT and the CFMT, evaluated by the Bowles et al. (2009) norms. Controls received gift certificates of ~120 DKK (~20 USD) per hour for their participation.

Statistical analyses

Comparisons are primarily based on "new statistics" where the interpretation of data is based on confidence-intervals (Cumming, 2013), although significance tests are also reported.

In single-case statistics, when the performance of an individual with DP is compared with that of a small control sample, we used the Bayesian test for a deficit developed by Crawford, Garthwaite, and Porter (2010) (implemented in the program SingleBayes_ES).

When age and education were found to correlate reasonably ($r > .3$) with performance in the control

Table 1. Age, gender and performance scores of 9 DPs on the Cambridge Face Memory Test upright faces, the Cambridge Face Perception Test upright faces, the Face Recognition Questionnaire, and accuracy and median RT for topographical perception and topographical memory.

						TP (perception)		TM (memory)	
	Age (years)	Gender	CFMT (max 72)	CFPT deviation	FQ score	Accuracy (max 12)	RT (s) Mdn	Accuracy (max 12)	RT (s) Mdn
Individual DPs									
PP04	57	M	37	86	61	8	6.8	8	9.5
PP07	40	F	41	60	57	9	11.5	9	7.5
PP09	40	F	43	70	45	11	15.7	9	8.8
PP10	34	F	33	58	53	10	25.4	6	15.3
PP13	51	M	35	42	55	10	15.7	4	6.4
PP17	49	F	35	88	48	12	8.8	10[a]	6.3
PP18	38	F	30	78	59	7	6.4	6	9.1
PP19	16	M	33	48	46	10	8.9	9[a]	9.0
PP27	25	M	42	66	51	6	12.6	8	6.6
Group means (SD)									
DP	38.9 (12.9)		36.6 (4.5)	66.2 (16.0)	52.8 (5.7)	9.2 (1.9)	11.5 (7.9)[b]	7.7 (1.9)	8.8 (2.8)[b]
Control	37.8 (12.1)		61.1 (6.7)	41.3 (11.9)	18.4 (9.4)	9.3 (1.7)	12.5 (6.8)[b]	10.1 (1.6)	7.1 (3.4)[b]

Note: DPs = participants with developmental prosopagnosia; CFMT = Cambridge Face Memory Test; CFPT = Cambridge Face Perception Test; FQ = Face Recognition Questionnaire; TP = topographical perception; TM = topographical memory; RT = reaction time; M = male; F = female. On the CFMT lower values indicate a deficit (scores 0–72), while on the CFPT and the FQ higher values indicate a deficit. Values in bold face designate abnormal performance applying Bayesian test for deficit with single-case statistics (Crawford et al., 2010), with age as a covariate in TM (Crawford et al., 2011). RT (correct trials only) in seconds.
[a]Cases showing a classical dissociation in outcome on TM versus CFMT. [b]Median and interquartile range for RTs.

sample we controlled for these variables by means of the Bayesian test for a deficit with covariates developed by Crawford, Garthwaite, and Ryan (2011; implemented in the program BTD_Cov). A score was considered abnormal if the one-tailed probability that the score could be an observation from the control population was less than .05.

Experiment 1. Topographic processing

Method

The Four Mountains Test (FMT) includes two subtests: The first measures topographic perception (TP) in a concurrent match-to-sample paradigm. The second measures topographic short-term memory (TM) in a delayed match to sample test. For the purpose of this study, the original A4 booklet version of the test was computerized using E-prime 2 while keeping all original features intact. A short summary of the FMT will suffice here, as it is described in detail in Hartley et al. (2007).

Stimuli

The perception and memory subtests both use landscape stimuli of a semi-circular mountain range containing four hills (or mountains) within the centre of view (see Figure 1). Topographical parameters are manipulated by altering the shape, size and relative location of the four hills. The perspective of the perceiver is from one of seven locations. Both subtests involve the same rotation/transformation processes. Non-topographical conditions, such as the time of day and the season, also vary. Each subtest comprises 15 items (12 test items and 3 practice items), with each test item being composed of one sample image, one target image (correct response, with all topographical information preserved from the sample image) and three categorical foils (one spatial, one configural and one elemental foil; see Hartley et al., 2007). Within each trial, each of the four alternatives was shown from a different perspective to avoid local matching based on small-scale features. No images were repeated, and the location of the target image changes both within and between subtests (Hartley et al., 2007).

Procedure

Instructions were presented on screen as well as given orally by the experimenter. The participant was instructed to identify a target image among four alternative test images. Responses were made using a keyboard, and the participant was seated approximately 70 cm from the screen. Subtests were completed in the order TP → TM, each starting with three practice items with feedback. In the TP part, sample and test (one target and three foils) images were displayed concurrently until a response was made or the time had run out at 60 s. The participant reported which of four images matched the

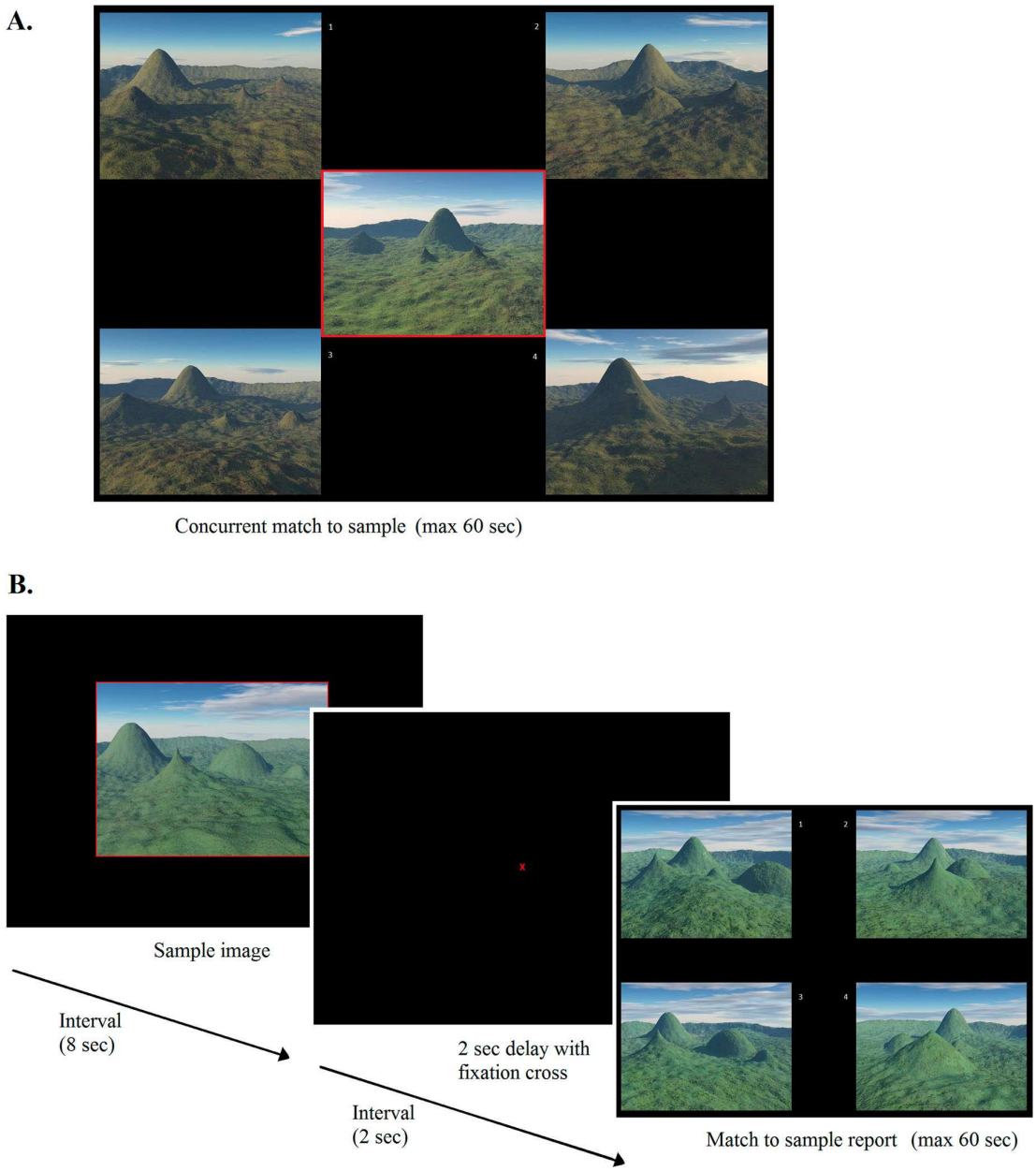

Figure 1. Stimuli presentation in computerized version of Four Mountains Test. (A) Concurrent match to sample, measuring topographical perception. (B) Delayed match to sample, measuring topographic memory. For more detailed description of paradigm see Hartley et al. (2007). [To view this figure in colour, please see the online version of this Journal.]

sample image by pressing 1, 2, 3 or 4 on the keyboard. In the TM part, the sample image was presented in isolation for 8 s; then a black screen with a fixation cross was shown for 2 s, after which the four test images were displayed. Apart from the change from concurrent to delayed match to sample, the procedures in the TP and TM subtests were the same. Accuracy and reaction time (RT) were recorded (and listed in Table 1). However, since the RT measure is not a part of the original test our focus is on the accuracy measure.

Results

Topographical perception (TP)

TP accuracy scores ranged from 6–12 in both groups (controls: $M = 9.28$, 95% confidence interval, CI [8.50, 10.06]; DPs: $M = 9.22$, 95% CI [8.00, 10.43]; see

Table 2. Experiment 2 results.

	K	t_0	C	α	w_{index}
Individual DPs					
PP04	3.95	9.63	72.62	1.09	.57
PP07	3.60	9.71	75.93	0.95	.57
PP09	2.78	30.48	46.00	1.09	.83
PP10[a]	1.73	7.19	27.35	0.29	.78
PP13[a]	2.57	32.44	48.22	1.15	.38
PP17	3.27	18.80	62.87	1.20	.55
PP18[a]	2.72	43.32	44.57	0.89	.68
PP19	3.25	7.37	36.41	0.31	.61
PP27[a]	3.76	6.90	71.52	0.61	.61
Group mean (SD)					
DPs	3.07 (0.70)	18.43 (13.68)	53.94 (17.39)	0.84 (0.35)	.62 (.13)
Controls	2.96 (0.98)	19.64 (11.36)	67.24 (22.01)	0.80 (0.47)	.54 (.12)

Note: DPs = participants with developmental prosopagnosia. CombiTVA parameters: K = short-term memory capacity; t_0 = threshold of conscious perception; C = processing speed; α = selectivity; w_{index} = distribution of attentional weighting. Units for the individual parameters are: K (letters), t_0 (ms), C (letters/second) An α-value of 0 denotes perfect selectivity, while 1 represents non-selectivity. α-values >1 indicates more attentional weight to distractors than targets. w_{index} ranges from complete rightward bias at 0 to complete leftward bias at 1 with .5 indicating equal weighting between the two visual fields. Numbers in bold indicate performance significantly below controls ($p < .05$; Crawford & Garthwaite, 2002).

[a]Subjects with topographical memory (TM) scores significantly below controls, as evaluated by Crawford and Garthwaite's (2002) methods.

Figure 2). Not one DP had a score below 2 standard deviations of the control mean (Table 1). The considerable overlap in CIs across groups suggests quite similar performance with regards to perception of the topographical layout, $t(25) = -0.08$, $p = .94$.

Topographical memory (TM)

At a group level, the TM performance level of DPs ($M = 7.67$, 95% CI [6.20, 8.87]) was reliably poorer than that of controls ($M = 10.06$, 95% CI [9.29, 10.75]), $t(25) = -3.47$, $p = .002$, Cohen's d (pooled SD) = 1.36 (see Figure 2). Also, better performance was associated with faster reaction times in the control group, $r(18) = -.519$, $p = .027$, 95% CI [−.767, −.026].

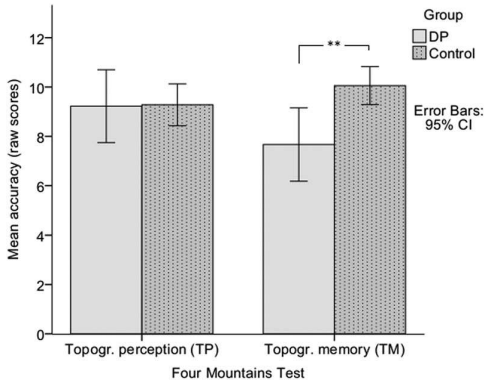

Figure 2. Performance (proportion correct) on subtests of topographic perception (TP) and topographic memory (TM) by group (developmental prosopagnosia, DP: N = 9; control: N = 18). Error bars show 95% confidence interval (CI) of the mean. **p < .01

Single-case statistics, with age as a covariate, showed that four DPs (PP10, PP13, PP18 and PP27) had accuracy scores significantly below those of the controls (one-tailed $p < .05$), as shown in Table 1. A concern that could be raised, however, is how reliable these scores are, given that they are based on a test with 12 trials. To address this, we computed the Spearman–Brown-corrected split-half reliability of the TM task. This was .644, which yields a standard error of measurement of 1.19 [$2 \times \sqrt{(1 - .644)}$]. Using this standard error of measurement, we estimated the upper 95% CI bound for any true score by adding +1.96 [1.19×1.65 (the z-score representing the 95% percentile)] to the observed score. These upper bound estimates thus represent the best score the patient could potentially have received on the test if – based on the test's reliability – the measurement was as off as it can be within the 95% CI. Reassuringly, even when such upper bound estimates were used in the single-subject analyses, instead of the observed scores, it was estimated (Bayesian point estimate) that 7% of the control population would obtain the same or lower score than PP10, 2% would obtain the same or lower score than PP13, and 9% would obtain a the same or lower score than PP18. In comparison, 30% of the control population would obtain the same or lower score than PP27. Hence, even when based on the upper 95% CI bound of the observed score, PP13 still scored significantly outside the normal range, and PP10 and PP18 scored in the very low end of it, whereas PP27 scored comfortably within the normal range. We note, that even with these three subjects and their

controls removed, there is still a reliable group difference between the DP and the control group ($M_{diff} = -1.58$, 95% CI [−2.38, −0.74], $p = .008$, based on bias corrected bootstrap analysis with 1000 samples). Hence, the group-level association is not simply driven by the three individuals who perform worst on the TM task.

While all the DPs scored significantly below the mean of the control group on the CFMT when considered individually, only four did so on the TM test. Moreover, two DPs (PP17 and PP19) fulfilled the criteria suggested by Crawford, Garthwaite, and Gray (2003) for a putatively classical dissociation with impaired performance on the CFMT but with performance within the normal range on the TM (PP17: one-tailed $p < .01$, $Z_{DCC} = -3.2$, 95% CI [−4.70, −1.93]; PP19: one-tailed $p = .015$, $Z_{DCC} = -2.9$, 95% CI [−4.45, −1.62]).

Experiment 2. Whole and partial report

The findings indicate a group-level impairment in the retention of topographical information in DP. The DP impairment of topographic memory, however, is not consistently related to the observed face recognition impairment, as 2/9 DPs showed a classical dissociation. At least in some cases of DP, then, face recognition deficits can exist without topographic memory problems, even though topographic memory ability is found to be reliably poorer in DP at a population level. The latter observation of impairments in two unrelated domains, however, could be a "trivial" consequence of a deficit in a cognitive process necessary for normal performance in both domains. Common for the CFMT and the TM task is that information must be retained for a short interval. Hence, the group-level impairment observed in both domains could reflect a general impairment in visual short-term memory (VSTM).

To rule this out, we next report data from assessment based on the theory of visual attention (TVA; Bundesen, 1990, 1998), modelling the efficiency of the first stages of object-based attention and categorization. TVA-based assessment is a theoretically grounded method well documented as being specific and sensitive, as well as suitable for testing clinical groups (Habekost, 2015). A combined whole and partial report paradigm (CombiTVA) was applied to estimate the following parameters of general visual processing: VSTM capacity (K), visual processing speed (C), the threshold of conscious perception (t_0), the spatial distribution of attention (w_{index}), and the efficiency of top-down control of attention (alpha). For detailed descriptions of the paradigm, modelling procedure, and estimated parameters see Habekost (2015) and Vangkilde, Bundesen, and Coull (2011).

Method

Stimuli and procedure

Coloured letters (red = targets, blue = distractors) were presented in a circular display on a black background for one of six durations (i.e., 10, 20, 50, 80, 140, and 200 ms) and terminated by a pattern mask (red and blue letter fragments), which covered each possible stimulus position for 500 ms. The letters were drawn randomly without replacement from a set of 20 capital letters (ABDEFGHJKLMNOPRSTVXZ) written in Ariel (broad) font, point size corresponding to 2.7° × 2.3° of visual angle. Seated approximately 60 cm from the screen, and with central fixation continuously emphasized, participants were asked to report any target letters they were "fairly certain" of having seen, while refraining from guessing. Participants responded by typing the letters in any order on a standard keyboard. The trial outline is shown in Figure 3.

One session contained 24 practice trials and nine experimental blocks of 36 trials. Blocks were counterbalanced with regards to the number of target and distractor letters as well as the exposure durations.

Results

Individual DP scores and group-level scores are shown in Table 2. On average, the threshold for visual perception (t_0) was similar across groups (DPs: $M = 18$ ms, 95% CI [10, 28]; controls: $M = 19$ ms, 95% CI [15, 25]), $t(25) = -.24$, $p = .81$. The average processing speed (letters/second) did not differ between groups (DPs: $M = 54$, 95% CI [42, 65]; controls: $M = 67$, 95% CI [57, 77]), $t(25) = -1.58$, $p = .13$. Both groups show a maximum VSTM capacity of about three letters (DP: $M = 3.07$, 95% CI [2.59, 3.49]; controls: $M = 2.97$, 95% CI [2.49, 3.42]), $t(25) = 0.29$, $p = .78$. While there was no reliable group difference in VSTM capacity, we note that the three DPs with TM scores below 2 standard deviations of the control group also displayed the lowest VSTM capacity. Note, however, that none of these scores fall outside the normal range when tested using single-case statistics (one-tailed $p > .1$).

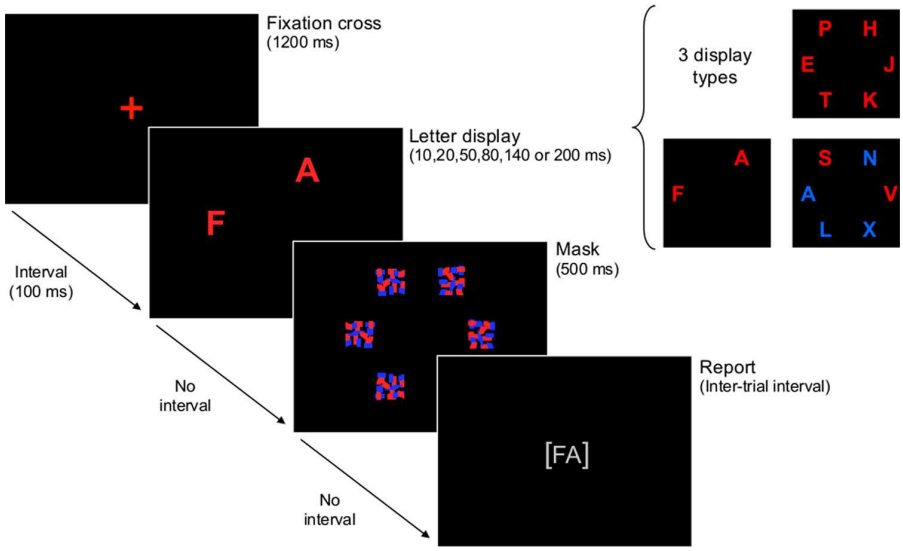

Figure 3. Trial outline of CombiTVA paradigm, where letter stimuli are displayed at brief exposure durations.

There was no reliable group difference in the spatial weighting of attention (w_{index}; DP, $M = .62$, 95% CI [.54, .71]; controls, $M = .55$, 95% CI [.49, .59], $t(25) = 1.51$, $p = .14$, or in the efficiency of top-down controlled selectivity (alpha; DP, $M = .84$, 95% CI [0.62, 1.05]; controls: $M = .80$ (95% CI [0.60, 1.00]), $t(25) = 0.265$, $p = .79$. In conclusion, no group-level differences were evident on any of the five parameters of visual attention including VSTM capacity. We note that failure to find a group difference in VSTM capacity is not simply due to lack of statistical power, as the DP group had higher mean VSTM capacity than the control group.

General discussion

We tested nine individuals with developmental prosopagnosia (DP) and 18 matched controls on a test of topographic perception and topographic memory, the Four Mountains Test (Hartley et al., 2007). All DPs showed normal perception of topographical features (topographical perception). However, the short-term retention of information was reliably poorer in the DP group than in the control group (topographical memory: Cohen's $d = 1.36$), and four DPs were significantly impaired on a single-case level. These findings indicate that memory for spatial/navigational information is impaired in some subjects with DP.

Interestingly, impairments of topographic memory and face memory were not found to be consistently related, as two subjects with DP showed a classical dissociation with: (a) impaired face recognition performance, (b) normal performance on topographic memory, and (c) a difference between face recognition and topographic memory performance more extreme than what can be expected in the population with normal face and topographical recognition abilities (cf. Crawford et al., 2003). This, seen in connection with the report of normal performance on measures of landmark recognition and route learning in another group of DPs (Corrow et al., 2016), strongly suggests that face recognition problems are not necessarily associated with deficits in topographic orientation. The deficits may co-occur, but are likely to be independent.

A possible explanation for the co-occurring deficits of face and topographical memory in our study was examined using assessment methods based on theory of visual attention (Bundesen, 1990), which yields estimates of central parameters such as VSTM capacity, visual processing speed, and spatial distribution of attention. These analyses revealed no signs of deficits, neither at the group nor at the single-case level.

The co-occurrence of impaired topographic memory in DP resembles findings in acquired prosopagnosia (after brain lesion) where navigational problems frequently have been reported (Aguirre & D'Esposito, 1999). A possible explanation for these associations could lie in the close proximity of the cortical areas contributing to topographic memory (the right parahippocampal region; Burgess, Maguire, &

O'Keefe, 2002) and face recognition (the right "fusiform face area"; Kanwisher & Yovel, 2006), respectively. At least, an association between impaired face recognition and topographic memory would be expected if DP partly reflects abnormal connectivity in the occipitotemporal region (Avidan et al., 2014). The topographic memory subtest of the Four Mountains Test has proved sensitive to hippocampal as well as extra-hippocampal volume in healthy adults (Hartley & Harlow, 2012). Such an account could explain why problems with face recognition and topographic memory often are associated without being functionally related. This is further supported by the finding that different lesion topology in acquired prosopagnosia is related to different outcomes on tests of topographic orientation (Corrow et al., 2016).

The type of topographic processing assessed in this study can be characterized as the ability to encode and retain metric representations of naturalistic environments in the domain of allocentric space. We here emphasize the term "naturalistic", as the landmarks used (i.e., four hills of differing size and shape) are relatively undifferentiated in comparison to, for example, manmade landmarks, which are more readily classified by some distinctive property or function (i.e., a bus stop, a school, a playground). Importantly, the relatively undifferentiated nature of the landmarks in the mountain landscapes used puts more demand on the use of metric representations of space, where the relief features or surface configuration are encoded. The topographic task successfully avoids cognitive strategies solely based on local information or distinctive features of landmarks – that is, it does not measure the ability to recognize specific locations (landmark recognition), but rather the ability to recognize Euclidean relationships between rather undifferentiated landmarks. Furthermore, the task does not measure navigational ability dependent on route learning, but rather the ability to generate and apply metric representations of new environments.

Further investigations are warranted to assess whether the observed deficit in topographic memory has an impact on real-life topographical orientation and to what degree. Studies relating the subjective experience of navigational problems in everyday life with experimental tests of different aspects of topographic processing, including topographic allocentric memory, are needed to inform our understanding of these functions, as well as their co-occurrence with acquired and (to a lesser degree) developmental prosopagnosia.

Disclosure statement

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