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Research report

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developmental prosopagnosia

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ABSTRACT

Face identity matching is selectively impaired in

Individuals with developmental prosopagnosia (DP) have severe face recognition deficits, but the mechanisms that are responsible for these deficits have not yet been fully identified. We assessed whether the activation of visual working memory for individual faces is selectively impaired in DP. Twelve DPs and twelve age-matched control participants were tested in a task where they reported whether successively presented faces showed the same or two different individuals, and another task where they judged whether the faces showed the same or different facial expressions. Repetitions versus changes of the other currently irrelevant attribute were varied independently. DPs showed impaired performance in the identity task, but performed at the same level as controls in the expression task. An electrophysiological marker for the activation of visual face memory by identity matches (N250r component) was strongly attenuated in the DP group, and the size of this attenuation was correlated with poor performance in a standardized face recognition test. Results demonstrate an identity-specific deficit of visual face memory in DPs. Their reduced sensitivity to identity matches in the presence of other image changes could result from earlier deficits in the perceptual extraction of image-invariant visual identity cues from face images.

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1. Introduction

Developmental prosopagnosia (DP) is a lifelong impairment in the ability to recognise faces in the apparent absence of brain damage or other cognitive impairments (for recent reviews see Susilo & Duchaine, 2013; Towler & Eimer, 2012). DP affects approximately 2% of the population (Kennerknecht et al., 2006; Kennerknecht, Pluempe, & Welling, 2008), and evidence from family and twin studies suggests that there may be a genetic component to this disorder (Duchaine, Germine, & Nakayama, 2007; Lee, Duchaine, Wilson, & Nakayama, 2010). The exact nature of the face processing deficits in DP is still largely unknown. Successful face recognition is based on a number of successive processing stages. These stages include the part-based and holistic perceptual processing of face images, constructing representations of identity-related visual information and retaining them in memory, and matching this information with the visual properties of a currently seen face (for a cognitive model of the stages involved in face recognition, see Bruce & Young, 1986).

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Impairments of any of these processes can result in impairments of face recognition, as experienced by individuals with DP.

Neuroimaging studies of DP have shown that in contrast to face recognition disorders caused by brain injury (acquired prosopagnosia; Bodamer, 1947), the occipito-temporal "core" face processing network (e.g., Haxby, Hoffman, & Gobbini, 2000, 2002; Kanwisher, McDermott, & Chun, 1997) appears to be largely intact in DP (Avidan & Behrmann, 2009; Avidan, Hasson, Malach, & Behrmann, 2005; Avidan et al., 2014; Furl, Garrido, Dolan, Driver, & Duchaine, 2011; Hasson, Avidan, Deouell, Bentin, & Malach, 2003; but see also Berhmann, Avidan, Gao, & Block, 2007; Garrido et al., 2009). However, investigations of face-specific event-related potential (ERPs) in DP are now beginning to reveal systematic differences between DPs and control participants, both at early visualperceptual stages of face processing, and at later memoryrelated stages associated with the recognition of facial identity (see Towler, Fisher, & Eimer, 2017, for review). Most ERP studies of DP have focused on the N170 component, which is the earliest face-sensitive ERP component that emerges at occipital-temporal electrode sites approximately 170 msec after stimulus onset. The N170 reflects an enhanced negativity for faces as compared to non-face objects, and is assumed to be generated during the structural encoding of faces and face parts in face-selective occipitotemporal visual areas (e.g., Bentin, Allison, Puce, Perez, & McCarthy, 1996; Eimer, 2000a, 2000b). Most individuals with DP show normal N170 components to faces versus non-face objects (Towler, Gosling, Duchaine, & Eimer, 2012), suggesting that the ability to perceptually discriminate between faces and non-faces is largely intact. However, changes to the prototypical spatial configuration and contrast properties of face images (such as presenting face images upside-down, spatially scrambling internal facial features, or contrast-inverting the eye region) produce atypical N170 amplitude modulations in individuals with DP (Fisher, Towler, & Eimer, 2016b; Towler et al., 2012; Towler, Kelly, & Eimer, 2016; Towler, Parketny, & Eimer, 2016). This suggests that perceptual face processing mechanisms in DP may be less well tuned to the spatial configuration and contrast properties that are the defining features of a typical upright face, and are therefore less sensitive to deviations from a prototypical visual face template.

The face perception deficits reflected by such atypical N170 responses in DPs are likely to adversely affect subsequent identity-related face processing stages, resulting in the severe face recognition problems experienced by individuals with DP. The processing of facial identity is associated with ERP components that emerge at post-stimulus latencies beyond 200 msec (N250 and N250r components). During the successful recognition of familiar faces and of learned target faces, an enhanced negativity at lateral posterior electrodes emerges at around 250 msec after stimulus onset (Gosling & Eimer, 2011; Tanaka, Curran, Porterfield, & Collins, 2006). This N250 component, which is assumed to reflect the activation of a stored representation of a particular individual face in longerterm visual memory, has also been observed for individuals with DP (Eimer, Gosling, & Duchaine, 2012; Parketny, Towler, & Eimer, 2015). However, the N250 in response to a learned target face was delayed in DPs as compared to age-matched

control participants (Parketny et al., 2015), suggesting that such identity matching processes are triggered less rapidly in DP. A similar N250 component has also been found in tasks where two face images are shown in quick succession. When the second face shows the same individual as the first face, an enhanced negativity is elicited bilaterally at occipitotemporal electrodes, relative to trials where faces of two different individuals are shown. This N250r ("r" for repetition) component is assumed to reflect the selective activation of a working memory representation of the first face that is triggered by an identity match with an on-line perceptual representation of the second face (Schweinberger & Burton, 2003; see also Begleiter, Porjesz, & Wang, 1995; Schweinberger, Pfütze, & Sommer, 1995; Schweinberger, Pickering, Burton, & Kaufmann, 2002; Schweinberger, Huddy, & Burton, 2004; Towler, Kelly, et al., 2016). In the face processing model proposed by Bruce and Young (1986), this process would correspond to the activation of a particular face recognition unit (FRU) in visual memory (see also Burton, Bruce, & Johnston, 1990). The fact that N250r components remain present when two different images of the same individual are shown (e.g., Bindemann, Burton, Leuthold, & Schweinberger, 2008; Kaufmann, Schweinberger, & Burton, 2009; Wirth, Fisher, Towler, & Eimer, 2015; Zimmermann & Eimer, 2013) shows that these components do not simply reflect a match between low-level visual image features, but are sensitive to higherlevel visual aspects of facial identity. N250r components to identity repetitions are not only elicited when face identity is task-relevant, but also when another face property has to be matched and identity can be ignored (Zimmermann & Eimer, 2014), indicating that the encoding of facial identity into working memory operates in an obligatory fashion for attended faces.

The goal of the present study was to use the N250r component to investigate the encoding and temporary working memory storage of identity-related face information in DP. Some behavioural studies have found that DPs are impaired in matching the identity of two successive unfamiliar face images (DeGutis, Cohan, & Nakayama, 2014; Shah, Gaule, Gaigg, Bird, & Cook, 2015), whereas other studies have shown no or little deficit (Ulrich et al., 2017). It is currently unknown whether individuals with DP have a particular deficit in detecting that dissimilar images of the same face belong to the same individual, or whether visual dissimilarity more generally impairs their ability to perceptually match other facial attributes, such as emotional expression. If there are any perceptual or working memory impairments in DP, these may be specific to representations of facial identity, and leave the representation of emotional expression unaffected. This has been suggested by studies showing that DPs are relatively normal in their ability to recognise categorically distinct basic emotions (Duchaine, Parker, & Nakayama, 2003; Humphreys, Avidan, & Behrmann, 2007; Palermo et al., 2011), more subtle and complex expressions (Duchaine, et al., 2007; Duchaine et al., 2003; Palermo et al., 2011) and are also able to successfully complete expression matching tasks (Bentin, DeGutis, D'Esposito, & Robertson, 2007; Garrido et al., 2009; Lee et al., 2010). DPs also show typical neural responses to emotional versus neutral faces (Avidan et al., 2014; Dinkelacker et al., 2011; Furl et al., 2011; Towler, Kelly, et al., 2016; Towler,

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