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Facing up to a change of concepts

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Abstract—**The term 'Prosopagnosia' refers to the inability to identify people using visual facial cues as a result of brain injury [1]. A further term, 'Developmental Prosopagnosia', has been proposed to refer to a condition with a similar dysfunction, but here there is an absence of any external brain injury. In recent years, these terms have been used increasingly, despite the lack of evidence about which specific functions are involved, and how they are affected during the development of human facial feature processing. Furthermore, most of these studies seem to address a dysfunction, rather than a function that never developed or has been lost altogether.**

We propose clarifying the terminology by distinguishing between whether the function is absent (prosopagnosia) or functioning at a suboptimal level (prosopdysgnosia). This distinction is particularly important as an increasing number of studies indicate varying degrees of function, rather than a function that is either present or not. Focusing on the variations of dysfunctions may help to form a better understanding of how facial features are processed and used by different observers. Moreover, studies describing functions that are lost can help us to understand which parts of a network are critical for face perception. One model that may prove particularly useful, both in the general understanding of the dysfunction and in the specific possibilities regarding rehabilitation is the Reorganization of Elementary Functions model (REF) [2]. This model has recently been applied both within consciousness studies [3] and blindsight [4].

Keywords-component; Prosopagnosia; Individual differences; Prosopdysgnosia; Visual Perception; Agnosia

I. INTRODUCTION

Visual Agnosia is a rare deficit where the observer is no longer able to recognize objects based on visual information [5]. This condition has even been reported to affect very specific domains, such as facial identification, referred to as Prosopagnosia [1]. People with prosopagnosia can often use cues other than visual, in the task of identifying other people (e.g. the sound of a person's voice). Nevertheless, they seem to have a profound "[…] inability to identify known people by the face" [6, p. 489]. The term 'Prosopagnosia' is often used not only to refer to a lost function, but also to dysfunctional facial recognition; here we argue that there is a need to distinguish between whether a function is missing (prosopagnosia), or merely operating at a suboptimal level (prosopdysgnosia). This clarification may ultimately yield a better understanding of the substrate underlying these cognitive functions and variations between observers, whereby we may progress towards a gradual understanding, where particular functions at one level of description may be achieved in different ways at the neuronal level [2,7].

Originally Bodamer [1] described patients who were able to identify individual facial features, while unable to bind these features together into a coherent whole; effectively rendering the patients unable to recognize people from facial information alone. A variety of prosopagnosia, seemingly not caused by injury or trauma, was later coined 'Developmental Prosopagnosia' [8]. Despite little initial attention, this dysfunction has since been reported with increasing frequency [9]. McConachie [8] described a 12-year old girl (AB) who was suffering from a severe deficit in facial identification and recognition. This became apparent when AB changed to a different school where the use of school uniforms prevented AB from using some of the feature cues she would normally rely on to identify people (i.e. clothing). Compensatory strategies potentially disguise cognitive deficits, as in the case of AB. Contrary to prior cases of prosopagnosia, AB did not seem to have any history or indication of traumatic injury, suggesting to McConachie [8] a developmental disorder. Also, AB's mother reported that she herself had difficulties identifying faces, however, this deficiency was not as severe as in the case of AB [8]. Such findings, naturally give rise to speculations as to whether there may be a hereditary component in the deficit [10,11]. For our present purposes, Developmental Prosopagnosia could be considered as one end of the spectrum of in a function that can have varying degrees [12]. This is further substantiated by reports of observers who populate the opposite end of the spectrum, the so-called 'super recognizers' [13, see also 34].

In a follow-up study of AB, De Haan and Campbell [6] reported that her deficit may not be limited to a severe dysfunction of facial recognition. She also has milder deficits, both in general object naming and in object recognition. It also seemed that AB had either retained or had developed some functions of face perception, since her performance was higher on a face-matching task [6]. Most studies of Developmental

Prosopagnosia rarely describe a pure inability to process face-specific information, as otherwise seemingly suggested by the label 'agnosia'. Studies that report on diagnostic tools for prosopagnosia, typically demonstrate variations in performance between individual observers [e.g. 14,15, see also 35]. Often, a performance criterion of two standard deviations from the mean is used as a cut-off, which is implicitly indicative of a dysfunction, rather than a lacking function [16].

Additionally, recent studies have begun to use the slightly different term of 'Congenital Prosopagnosia'. This is treated synonymously with Developmental Prosopagnosia, and is defined as an "impairment in face processing that is apparent from birth in the absence of any brain damage" [17, p. 180]. Some authors may have preferred the term 'Congenital Prosopagnosia' over 'Developmental Prosopagnosia', as the latter term has also been used to refer to acquired Developmental Prosopagnosia [18]. Generally, both Developmental and Congenital Prosopagnosia are used more broadly, referring to both deficits and dysfunctions in facial recognition, that are not caused by head injury or trauma [9,19]. We believe both terms to be problematic when addressing cognitive functions that are not developed from birth. In particular, the latter term is highly questionable, as one may argue that all newborns have Congenital Prosopagnosia. In fact, studies have suggested that it is not possible to see normal perceptual processing biases until a few months after birth [e.g. 20]. Only after the visual system matures, may one see whether a child has a developmental disorder affecting the ability to recognize faces.

As well as Developmental Prosopagnosia and Congenital Prosopagnosia, a third term has emerged, namely, 'Hereditary Prosopagnosia' [e.g. 10,11]. This also seem to be used analogous to the two other types of prosopagnosia, nevertheless, here the causal emphasis is on genetics (or upbringing). Studies of families, for example [e.g. 11], only give indirect evidence for hereditary components. One may indeed, question the contribution of nature and nurture in these types of studies. Both could be confounded by a number of other factors, such as how different people relate to their experiential qualia. As an example, it has long been argued that studies of synaesthesia suggest a hereditary element based on a strong female gender bias [e.g. 21]. Recent studies have established that this gender bias is, in fact, due to diverging cultural practices between how males and females discuss how they experience their surrounding environment [22]. To the knowledge of the author's, explicit genetic studies of face recognition have yet to be performed. There are a number of studies of twins [23,24], however, where it has been questioned whether these findings relate specifically to face perception [25].

The decreased ability to recognize faces may reflect a function that is much more prone to individual variation than we usually assume [10,12]. Already in her original paper, McConachie [8] briefly speculated, as to whether the phenomenon may be more widespread in the general population. In related areas, where we see individual perceptual differences, such as synaesthesia mentioned above, observers discover the fact that they perceive the world differently from other people only rather late in their development [26]. Similarly, to the case of AB, where her difficulties went largely unnoticed, until an external event (the use of school uniforms) revealed that AB did not perceive the world similarly to her peers. If no such external pressure is experienced, then it is easy to imagine that many observers could be unaware of their synaesthesia, or in the present case, decreased performance in identification based on visual facial cues. Studies often suggest using a lifelong report of poor face recognition as an additional diagnostic characteristic in addition to the two standard deviation cut-off on more objective tasks; however, in the light of the example above, such subjective criterion does not seem valid.

Our aim is to shift the emphasis from cause to function. We propose the more accurate term 'Prosopdysgnosia' (or 'Prosopodysgnosia') in order to emphasize a decrease in ability compared to a neurotypical observer, rather than Developmental, Congenital, or Hereditary Prosopagnosia; as all of these latter seem to carry connotations of cause. Naturally, if the decrease in function can be linked to a specific cause, such as a trauma, we find it relevant to add 'acquired', and if other causal links can be established (e.g. developmental) for prosopdysgnosia (or prosopagnosia) then it would make sense to use developmental prosopdysgnosia. However, it still seems that more work is needed before one can ascribe the cases to developmental disorders with any certainty. Even in the absence of any evidence of external trauma, it is still difficult to fully rule out the possibility that a dysfunction is not actually due to an external cause. Hence one may argue that McConachie [8] was premature in coining a term for something in the absence of evidence. Imaging studies have sometimes had difficulties distinguishing observers with Developmental Prosopagnosia from neurotypical observers [see 12]. Moreover, if face perception is a varying spectrum in the normal population, then Developmental Prosopagnosia at times would also be much more similar to neurotypical observers than would the very severe cases originally reported by Bodamer [1].

Also, work linking face perception to general expertise seems to further support the proposed distinction [e.g. 25,27]. Investigations of face perception often reveal that observers have a processing advantage for upright compared to inverted faces [e.g. 27]. In a group of colour-grapheme synaesthetes we have previously shown a decreased upright advantage, but only a few of our observers had complaints related to their ability to recognize faces [28]. Moreover, Gauthier et al. [25] recently linked poor performance on face-recognition tasks to a general decrease in object recognition (as also later reported in AB [6]); this allows for some speculation into the degree of difference between face perception and object perception.

In addition, studies of the so-called 'own-race bias' in perception – a bias in the perceptual processing of faces – further demonstrate that this process is not present from birth and this bias is only established when the child is at least three months old [20]. Moreover, a study by Bar-Haim, Ziv, Lamy, and Hodes [29] showed that the own-race bias is established through an interaction with the surrounding environment, supporting the idea that face perception is acquired, and is fine-tuned through environmental exposure and expertise.

If, in fact, face and object perception are modulated by a general domain non-specific function, as suggested by Gauthier et al. [25], which develops through environmental exposure and interaction [20,29], then it is even more important to be able to locate the underlying domain-specific substrates in face perception. This may prove important in the development of intervention strategies for people with suboptimal facial recognition. These in turn, may not be an effective method in treating more severe cases of prosopagnosia.

With some caution, one might also consider the theoretical consequences of the argument above. The debate between strong localization and the equipotentiality of the brain has historically been discussed with varying emphasis, reaching a compromise between the two extreme positions today [30]. If we accept that cognitive functions such as perception are represented in varying degrees [12, see also 31], rather than as cognitive modules [32]. Cognitive functions, such as face perception, may be described as processes of interacting strategies rather than separated, individual modules, hereby relating to different neural networks. This would mean for instance that face perception may be achieved in a number of different ways encompassing varying strategies from observer to observer [e.g. 2,30]. The proposal of a distinction between prosopagnosia and prosopdysgnosia seem to support this view. Which is further supported by the number of varying sub-strategies that can be employed to identify other people, as an example, some people have been reported who have difficulties in identifying others from their voice alone (phonagnosia) similar to prosopagnosia in vision [36,37].

One model of functional reorganization following brain damage, does in fact predict that a neural injury may not result in the permanent abolishment of particular functions, but, rather, in the development of different strategies to replace them. In the recently proposed Reorganization of Elementary Functions (REF) model, the orchestration of 'elementary functions' (specific functions localized to a particular neural region) form 'algorithmic strategies' in larger brain networks [2]. According to the model, such algorithmic strategies subserve behavioural and cognitive functions, but not in a 1 to 1 fashion [7]. Rather, in the case of an injury to some region in the brain, following which some elementary functions are damaged, the associated strategies and behaviour or cognition are also (temporarily) lost. However, in many cases, new strategies consisting of different or partially different sets of elementary functions realize the relevant behaviour or cognition. Nevertheless, at the level of the algorithmic strategies, the same function will be realized in a different way. In this view, it is highly plausible that we may find intervention strategies that can alleviate milder problems like prosopdysgnosia, whereas the damage to elementary functions in prosopagnosia may be too severe to be rehabilitated.

Models, such as the REF-model unfold a more complex picture of different possible strategies with different degrees of efficiency. As such, they fit nicely into other developments in visual perception research, where conscious experience of objects contains several levels of representation [see 31,33]. Moreover, this corresponds to the different strategies used in identifying people, where we rely on a number of different cues (e.g. clothes as in the case of AB).

II. Conclusion

We believe that the emphasis on function should be explicit, similarly to the distinctions between alexia and dyslexia. Moreover, distinguishing pure cases of prosopagnosia from different kinds of prosopdysgnosia may lead to a better understanding of the neural mechanisms constituting the algorithmic strategies behind face perception. Furthermore, the difficulty in accurately gaging the cause of the developmental disorder, seem to warrant the more accurate term 'prosopdysgnosia', and therefore the term 'prosopagnosia' should be reserved for cases where the facial identification through visual cues is abolished, or severely limited.

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