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Review

A century of prosopometamorphopsia studies

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ABSTRACT

Prosopometamorphopsia is an extremely rare disorder of visual perception characterised by facial distortions. We here review 81 cases (eight new ones and 73 cases published over the past century) to shed light on the perception of face gestalts. Our analysis indicates that the brain systems underlying the perception of face gestalts have genuine network properties, in the sense that they are widely disseminated and built such that spatially normal perception of faces can be maintained even when large parts of the network are compromised. We found that bilateral facial distortions were primarily associated with right-sided and bilateral occipital lesions, and unilateral facial distortions with lesions ipsilateral to the distorted hemifield and with the splenium of the corpus callosum. We also found tentative evidence for the involvement of the left frontal regions in the fusing of vertical hemi-images of faces, and of right parietal regions in the fusing of horizontal hemi-images. Evidence supporting the remarkable adaptability of the network comes from the relatively high recovery rates that we found, from the ipsilateral hemifield predominance of hemi-prosopometamorphopsia, and from a phenomenon called cerebral asthenopia (heightened visual fatigability) which points to the dynamic nature of compensatory mechanisms maintaining normal face perception, even in chronic cases of prosopometamorphopsia. Finally, our analysis suggests that specialised networks for the representation of face gestalts in familiar-versus-unfamiliar faces and for own-versus-other face may be present, although this is in need of further study.

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1. Introduction

The term prosopometamorphopsia was introduced by Critchley (1953) to denote a visual distortion specific for faces. The condition famously features in Lewis Carroll's *Alice's Adventures in Wonderland*, in the scene where Alice carries a baby out of the Duchess's kitchen and sees it gradually grow 'a *very* turn-up nose', and then change into a pig (Blom, 2020; Carroll, 1865). In real life, Pablo Picasso and Francis Bacon may have experienced it themselves, judging by the split and otherwise distorted faces featuring in their art (Ferrari & Haan, 2000; Safran, Sanda, & Sahel, 2014). As these examples illustrate, prosopometamorphopsia may manifest as anything from distortions or even a scrambling of facial features to full transformations into animal faces, although the literature indicates that vertical splitting with ensuing hemidistortions may well be its most prevalent manifestation.

1.1. *Defining and demarcating the field*

Prosopometamorphopsia, or the phenomenon of perceiving facial distortions, affects the whole face or aspects thereof, and occurs during wakefulness in the absence of similar distortions in surrounding objects or scenes. As a distortion, it belongs to the 'third group of perceptual disorders', with the first two groups comprising hallucinations and illusions (Blom, 2010). It thus constitutes one of the 40-odd types of metamorphopsia (i.e., visual distortion) characteristic of Alice in Wonderland syndrome (which is the umbrella term for perceptual distortions in general; Todd, 1955; Blom, 2016). Clinically and scientifically, prosopometamorphopsia needs to be distinguished from facial hallucinations (seeing faces where there are none), hypnagogic face hallucinations (aptly dubbed 'faces in the dark'), facial pareidolia (seeing faces in amorphous or otherwise non-facial patterns such as a cloud or a clock), face illusions [such as the strange-face-in-the-mirror illusion (Caputo, 2010) and the more-or-less morphing face illusion (Van Lier & Koning, 2014)], and types of metamorphopsia that are not face-specific but may nonetheless affect the perception of facial features, such as hemimicropsia (seeing half of the visual field smaller; e.g., Cohen, Gray, Meyrignac, Dehaene, & Degos, 1994) and polyopia (seeing multiple copies of objects; e.g., Bekény & Péter, 1961). In the past prosopometamorphopsia was sometimes considered a mere feature or underlying cause of prosopagnosia (i.e., agnosia for faces; De Ajuriaguerra & Hécaen, 1964), but the one condition is not necessarily accompanied by the other, while underlying mechanisms appear to overlap only partially. Subserving two dissociable functions of the face-selective network, one is related to face recognition and, when dysfunctional, leads to prosopagnosia; whereas the other is linked to the perception of face gestalts and, when dysfunctional, leads to prosopometamorphopsia. Therefore, prosopagnosia and prosopometamorphopsia need to be carefully distinguished from each other. Neither should prosopometamorphopsia be confused with face adaptation (i.e., the finding that faces can be perceived slightly differently due to prior exposure to other faces; Webster & MacLeod, 2011). Finally, it should be distinguished from the flashed face distortion effect (also known as multiple-faces configuration), an experimentally induced illusion that involves a marked distortion of facial features when normal faces are presented in rapid succession to the peripheral visual field (Simas, Rocha, Sedycias, do Amaral, & de Menezes, 2008; Tangen, Murphy, & Tompson, 2011). Of note, patients with Alice in Wonderland syndrome may sometimes experience several types of metamorphopsia simultaneously but facial distortions only count as prosopometamorphopsia when non-facial distortions are of a different nature, such as plagiopsia (seeing things as being slanted), macrsomatognosia (experiencing one's own body as larger) or time distortions.

1.2. *Prosopometamorphopsia and the face-selective network*

Clinical descriptions of prosopometamorphopsia are extremely rare, although the condition is probably underreported (Podoll & Robinson, 2008). The first known case report stems from well over a century ago, when Lachmund (1904) reported on a 37-year-old woman who looked at herself in the mirror during a postictal twilight state and saw her face as '*changed*' and '*bigger*', with '*large, contorted eyes*'. Five years later Lenz (1909) described a 73-year-old woman who suddenly went blind, possibly due to bilateral occipital stroke, and, after partial recovery, had to cope with left-sided hemianopia with macular sparing while seeing familiar faces as '*so large, strange, and grimacing*'. During the decades that followed, Curschmann (1916) described a 35-year-old woman with migraine who saw grotesquely disfigured faces in hallucinated as well as real persons, and Zádor (1930) two cases in the context of experiments with the hallucinogenic substance, mescaline, derived from the peyote cactus. It took until World War II before prosopometamorphopsia was documented more frequently, notably in soldiers who had survived occipital gunshot or shrapnel wounds (e.g., Bodamer, 1947; Faust, 1951; Pichler, 1943). Since then it has also been described in other conditions, ranging from tumour, infarction, epilepsy, and multiple sclerosis to psychosis. Because of this, the etiology of prosopometamorphopsia is considered multifactorial in nature (ffytche & Howard, 1999). Regarding its pathophysiology, most lesion studies suggest the involvement of the occipital face area, fusiform face area, and splenium of the corpus callosum (Miwa & Kondo, 2007; Schroeder et al., 2017). In healthy volunteers, the condition has also been induced experimentally with the aid of electrical stimulation (Jonas et al, 2012, 2018; Parvizi et al., 2012; Rangarajan et al., 2014). On the basis of neurocognitive, neuroimaging, and electrophysiological studies, notably in the field of prosopagnosia, over the past 50 years a tentative model has been developed of a face-selective network, with the model by Duchaine and Yovel (2015) being the most comprehensive one to date. While many types of evidence have contributed to these models of face perception (Ahmed & Hodges, 2020), prosopometamorphopsia has hitherto played no substantial role in this. To amend this, we here present eight cases of this rare condition from our own clinical practice, supplemented by an overview of clinical cases lifted from the literature, which we analyse with the aim of refining the model devised by Duchaine and Yovel to accommodate evidence from prosopometamorphopsia.

2. Material and methods

The original reports presented below concern patients receiving treatment in two outpatient clinics in the Netherlands. We obtained written consent to publish from all. For the purpose of the literature review we performed a systematic search in PubMed, Embase, PsycINFO, Google Scholar, and the historical literature, up until January 15, 2021, using the search terms 'prosopometamorphopsia', 'facial metamorphopsia', 'metamorphopsia for faces', and 'hemi-prosopometamorphopsia'. The digital search was supplemented by backward searches. Papers were excluded when they contained no original case descriptions. From all eligible reports, the following data were extracted: i) year of publication, ii) sex and age of the patient(s), iii) phenomenological characteristics, iv) clinical diagnoses, v) test results, vi) type(s) of treatment, and vii) outcome. To allow for a proper comparison of cases we adopted the convention that 'distortions to the right side of the face' are those in the right hemifield when the point of fixation is at the facial midline, not the anatomical right side of the face of the person perceived, and adjusted this when necessary (e.g., Phua, Bhaskar, & Calic, 2019). Moreover, we used a tailor-made classification to distinguish different types of prosopometamorphopsia, comprising hetero-versus auto-prosopometamorphopsia (for distortions perceived in other people's faces versus those perceived in one's own face), hemi-prosopometamorphopsia (for unilateral distortions across the midline), and zoanthro-prosopometamorphopsia (for transformations of human faces into animal faces) (Table 1). Aided by that classification, we used the conceptual framework of lesion network mapping (Boes et al., 2015) to relate the evidence to Duchaine and Yovel's (2015) face-selective network.

3. Results

3.1. Novel case reports

Over the course of 2019 and 2020 we saw eight patients with prosopometamorphopsia, five of whom at our neurology outpatient clinic in Amsterdam and three at our outpatient clinic for uncommon psychiatric syndromes in The Hague. There were four female and four male patients, and their combined mean age was 49 years (range 19–74 years). All but two of the patients (75%) experienced hemi-hetero-prosopometamorphopsia (i.e., seeing half of other people's faces distorted) with vertical splitting across the midline. All underwent psychiatric and neurological examination, blood tests, neuroimaging (CT and/or MRI head), and (when indicated) EEG. Clinical presentations, test results, diagnoses, and treatment outcomes are summarised in Table 2. Further analyses are provided below, along with the cases extracted from the literature.

Table 1 – Classification of types of prosopometamorphopsia.

Autoprosopometamorphopsia	Seeing one's own face distorted (e.g., in a mirror)
Heteroprosopometamorphopsia	Seeing other people's faces distorted
Hemiprosopometamorphopsia	Seeing only the right or left half of faces distorted
Zoanthroprosopometamorphopsia	Seeing human faces morph into animal faces

3.2. Systematic review

Our literature search initially yielded 17 articles on prosopometamorphopsia. Cross-references and a search in the historical literature yielded another 46 papers and book chapters, plus one master thesis. Together these 63 texts described 76 patients diagnosed with the condition. Since three case descriptions were very similar and probably based on the same patient, we collapsed these into a single case (Hécaen, de Ajuriaguerra, Magis, & Angelergues, 1952; Hécaen & Angelergues, 1962; De Ajuriaguerra & Hécaen, 1964); the same held true for the case descriptions by Freixo (2016) and Almeida et al. (2020), which we likewise collapsed into a single case. The remaining 73 unique cases were added to the eight we had seen in our treatment centres and evaluated.

3.3. Main findings

3.3.1. Demographics. Of the 81 cases of prosopometamorphopsia, 64% were women and 35% men; in 1% the sex was not reported (Table 2 and Supplementary Material). Age was reported in 98% of the cases, while one patient was variously designated as 60 and 68 years of age in the original paper (Ebata, Ogawa, Tanaka, Mizuno, & Yoshida, 1991). As was the case for our own patients, the mean age of the whole group was 49 years (range 14–94 years).

3.3.2. Phenomenology. All but seven patients (92%) reported visual distortions exclusive to faces. A patient described by Wagner (1943) perceived 'broad faces' as well as 'strangely distorted hands'. The two patients described by Safran, Achard, Duret, and Landis (1999) perceived 'thin faces' and similar distortions to the left shoulder. Two patients described by Blom (2020) recounted of several additional types of metamorphopsia besides prosopometamorphopsia. A patient reported on by Lachmund (1904) experienced prosopometamorphopsia and coloropsia (i.e., seeing hues of gold and silver) and one of Bodamer's (1947) patients prosopometamorphopsia and achromatopsia (seeing no colors). The distortions the majority of patients (69%) reported were indicative of pure hetero-prosopometamorphopsia, with 5% reporting signs of pure auto-prosopometamorphopsia, and 25% symptoms of both types (while 1% was unspecified). Two patients (3%) reported distortions characteristic of zoanthro-prosopometamorphopsia, one of whom saw other people's faces morph into dragon faces, while the other one perceived them '*as fish heads*' (Blom, Sommer, Koops, & Sacks, 2014; Whiteley & Warrington, 1977). There were no such transformations in the auto-prosopometamorphopsia group. Hemi-prosopometamorphopsia with vertical splitting was documented for 51% of the patients, of whom 66% reported distortions indicative of hemi-hetero-prosopometamorphopsia, 4% of hemi-auto-prosopometamorphopsia, and 30% of a combination of both (i.e., perceiving hemi-distortions in other people's faces as well as in their own face). Of all the hemi-cases, 59% exclusively perceived distortions in the right field of vision, versus 41% in the left. Nine patients (i.e., 23% of the

Table 2 – Overview of eight cases of prosopometamorphopsia from our own clinical practice.

Case nr.	Sex, age (years)	Type of prosopometamorphopsia (characterisation)	Clinical diagnosis	Test results	Treatment	Outcome
1	F, 63	Hetero-prosopometamorphopsia since 1 day (seeing the left half of faces as vertically elongated, with a large eye, and the right eye replaced by a hole) + autoprosopometamorphopsia (one day later, unspecified)	Brain hemorrhage; migraine (preexistent)	CT and MRI: left parieto-occipital bleed; no underlying causes	–	Symptom-free in 2 days (and at follow-up 4 months later)
2	M, 74	Hemi-hetero-prosopometamorphopsia since 1 day, 15 min each time (seeing the left side of faces move away from the right side, leaving the 2 halves detached)	Brain infarction, probably with secondary epilepsy	CT: mild global atrophy; MRI: diffusion restriction in the left medial occipital horn	–	Symptom-free in several days
3	F, 54	Hemi-hetero-prosopometamorphopsia since 1 day (seeing the right side of faces deformed, including the eye)	Brain hemorrhage; migraine (preexistent)	CT and MRI: right parieto-occipital lobar hemorrhage; no underlying causes	–	Symptom-free in 6 months
4	F, 19	Hemi-hetero-prosopometamorphopsia since 3 weeks, 6 times, 15 min each time (seeing the left side of faces 'melt', with the eye moving downwards, protruding over the cheekbone, and the right and left halves of the face ending up detached)	Frontal-lobe epilepsy; cannabis use (preexistent, and prior to first attack)	EEG: epileptic focus in the left frontal lobe; MRI: normal	–	Symptom-free after consultation
5	M, 38	Hetero-prosopometamorphopsia (seeing a third eye on the middle of people's forehead and a fourth eye at their right temple) + autoprosopometamorphopsia (once, seeing the left corner of his own mouth as drooped)	Brief psychotic disorder due to stress, sleep deprivation, and possible substance abuse (unconfirmed)	EEG, MRI and CSF normal	lorazepam	Symptom-free in 2 weeks
6	F, 45	Hetero-prosopometamorphopsia since 2 years, several times a day, 3–5 min each time (seeing a vertical zigzag line across faces, leaving the 2 halves detached; once also a diagonal elongation of faces)	Alice in Wonderland syndrome, posttraumatic stress disorder, chronic pain after 2 traffic accidents with head injury	EEG and MRI normal	gabapentine, carbamazepine, levetiracetam	Unaltered after 4 months
7	M, 49	Hemi-hetero-prosopometamorphopsia (seeing people's right eye as a large black dot, reminiscent of a large pupil)	Brain hemorrhage	CT and MRI: right occipital bleed	–	Symptom-free in 2 days
8	M, 48	Hemi-hetero-prosopometamorphopsia (seeing people's right eye replaced by skin)	Brain infarction	MRI: left parieto-occipital infarction	–	Improved in 3 days

hemi-group) saw half of the face 'drooping downward' or 'melting'. In addition to these hemicases, Young, Heros, Ehrenberg, and Hedges (1989) described a unique case of horizontal splitting. In the remainder of the patients, faces were perceived as being either wider or narrower, vertically elongated or flattened, and/or distorted in numerous other ways, with parts appearing blurry or wavy, noses typically being bent downwards or sideways, eyes and/or mouths appearing larger or smaller, eyebrows being located in different positions, and colours looking unnatural or being absent. Typically, each individual type of distortion was seen in every person the patient looked at, although some patients experienced other distortions when they looked at themselves in the mirror. Two patients (3%) said that faces looked '*like a Picasso painting*', thus aptly indicating that they had lost the ability to perceive faces in their proper spatial gestalt, with three more patients (4%) reporting them to be 'kaleidoscopically changing', and one patient describing seeing an ear on top of people's heads and a shortened arm attached to faces. Several other remarkable cases featured eyes that 'behaved' extraordinarily. Apart from seeing them growing smaller or larger, some patients described them as contorting, changing colour, disappearing altogether, or moving towards an area outside the face, while one of our own patients saw a third eye in the middle of people's foreheads and a fourth at their right temples. Two others (3%), while standing in front of the mirror, saw one eye popping out of its socket and slithering down the cheek. Another patient saw people's eyes leaving the skull and rotating in front of it, and yet another reported seeing the left lens of people's spectacles rotating and floating through the room. Comorbid prosopagnosia was noted in seven cases (9%) only.

3.3.3. Neuroanatomy. In 78% of the cases, the original reports allowed for localisation of the underlying lesion or dysfunction (including one fMRI study by Dalrymple et al., 2014), with right-sided lesions being observed in 47%, left-sided lesions in 41%, and bilateral lesions in 12%. In 70% of the patients with hemi-prosopometamorphopsia, sidedness corresponded with demonstrable pathology in the ipsilateral hemisphere and in 30% with abnormalities in the contralateral hemisphere. There were no hemicases with bilateral cerebral involvement. The occipital lobe was involved in 53% of the cases, the corpus callosum in 28% (splenium 27%, genu 1%), the temporal lobe in 27%, the parietal lobe in 15%, and the frontal lobe in 6% [with only two cases (3%) showing exclusive frontal pathology]. There were no cases with exclusive left-temporal involvement. Bilateral and unspecified facial distortions were primarily associated with right-sided (61%) and bilateral lesions (30%), and with left-sided lesions in 9% only. All lesions to the left occipital lobe resulted in hemi-prosopometamorphopsia, of which 79% was ipsilateral. Likewise, of all the lesions to the corpus callosum, 94% resulted in hemi-prosopometamorphopsia, of which 57% was ipsilateral when lesions were located to the left side of the splenium/left forceps major and 75% when the right side of the splenium/right forceps major was compromised. Colour changes in faces or parts thereof (reported by 16% of the patients) were seen in connection with occipital lesions (in five patients, of whom one also suffered from achromatopsia), right temporo-occipital lesions (three patients), and lesions to the right side of the splenium/right forceps major (one patient). Four other patients reporting (parts of) faces changing colours did so in the context of epilepsy, migraine, mescaline intoxication, and Alice in Wonderland syndrome. Of the two people (3%) who perceived faces as smoother and younger-looking, one had right-temporal involvement and one involvement of the right fusiform face area, while a third patient who showed a lesion to the right parieto-temporo-occipital area perceived faces as older-looking. No such age-related alterations were reported by patients with left-sided lesions. In the seven cases complicated by prosopagnosia, right-sidedness and involvement of the occipital lobe were the most salient neural features (two bilateral occipital, two right parieto-occipital, two right temporo-occipital, one right temporal).

3.3.4. Etiology. A clinical diagnosis was made in 93% of the cases, while etiopathophysiology was established in 84% of the cases, mostly with the aid of EEG, neuroimaging, biopsy, and/or surgery. In the whole case series, the largest group (33%) had suffered a brain infarction, with epilepsy being involved in 10% (plus several other possible cases, often secondary to structural lesions), haemorrhagic stroke in 9%, complications of surgery also in 9% (of which almost half were due to prophylactic surgical procedures), migraine in 5% (plus one possible case), head injury in 5%, brain tumour in 5%, and mescaline intoxication in 3%. For the remaining 5%, miscellaneous causes were described, comprising 'parietal softening' [sic], brain haematoma, multiple sclerosis, and subarachnoid haemorrhage (all single cases). In 24% no *structural* abnormalities were demonstrated. These patients received diagnoses such as epilepsy, migraine, schizophrenia, brief psychotic disorder, Alice in Wonderland syndrome, transient ischemic attack, and Charles Bonnet syndrome.

3.3.5. Treatment outcomes. Treatment was detailed in 61% of the cases, with interventions mostly addressing the underlying cause (e.g., antiepileptics for epilepsy, migraine prophylaxis for migraine, etc.). In the whole group, 57% of the patients attained full or virtually full recovery, 10% partial recovery, and 13% no recovery (with 19% of the outcomes going unmentioned). Roughly half of the patients showing full recovery had received active treatment (e.g., surgery to remove brain tumours, antiepileptics to treat epilepsy) whereas the other half had undergone no treatment, unknown treatment, conservative treatment, or secondary prevention (e.g., aspirin, mannitol, clopidogrel) while recovering nonetheless. The time to recovery ranged from hours to years, with days to weeks being the most frequent interval, while in two cases recovery did not occur until after decades of frequently recurring symptoms (Blom et al., 2014; Blom, 2020). Of note, we operationalised 'full recovery' as recovery from prosopometamorphopsia, not necessarily of adjuvant symptoms or underlying disorders. In our own case series, all eight patients recovered quickly, except for the one patient diagnosed with Alice in Wonderland syndrome (Table 2). The cases from the literature showed considerable variation in outcome. In one patient, for instance, his prosopometamorphopsia was replaced by partial blindness (Pichler, 1943), while another patient with a massive glioblastoma infiltrating

almost the entire right hemisphere was relieved of his facial distortions within two days post-surgery - and had even resumed work - but died four months later, probably due to complications (Geyer, 1963). Except for these two extreme examples, the other patients who were reported to have attained (full) recovery appear to have done well. Remarkably, there were no significant differences in the recovery rates as a function of underlying cause. What is more, even the cases resulting from brain infarction, tumour, and surgery showed good recovery rates, with six out of seven instances of brain surgery (mostly for tumour resection) ending in full or virtually full recovery.

4. Discussion

4.1. Network properties

Our analysis of 81 cases of prosopometamorphopsia confirms that, physiologically, spatial gestalt aspects of face processing are largely dissociated from spatial gestalt aspects of object processing. The relationship between face and object processing more generally has long been debated, but findings over the past 30 years are chiefly in favour of the specificity hypothesis (although controversies remain; see Duchaine & Yovel, 2015). Especially since the work by Haxby, Hoffman, and Gobbini (2000), it has become customary to divide the face-selective network into a core system and an extended system, the latter comprising non-visual structures such as the insula, limbic system, and auditory cortex, which play a role in adding emotional context, salience, multimodal experience, and other features. Although the core system is often referred to as a network, it is mostly conceptualised as a system comprising a ventral and a dorsal stream, running from occipital towards more rostral areas (e.g., Duchaine & Yovel, 2015). The present study indicates that the perception of spatial aspects of face features and spatially normal face gestalts is also carried out in a widely disseminated network with involvement of occipital-to-frontal parts, left-to right-hemispheric parts, and interhemispheric transfer through the corpus callosum. The relationship of the face-gestalt network to the core face system is unclear from the evidence reviewed, but it seems to be built in such a way that faces can still be perceived as spatially normal when relatively large parts of the network are missing or malfunctioning. Perceptual distortions seem to relate to an imbalance of activity across the network, or diaschisis, that resolves with recalibration of network activity over time. Evidence for this notably comes from the cases of head trauma, tumour, and surgery, which show that even in the presence of dramatic tissue loss, faces are typically only temporarily perceived in a distorted fashion. Particularly some of the older cases of occipital war wounds are testimony to the brain's impressive plasticity, where initial states of cortical blindness were followed by a return of vision within days to weeks, with prosopometamorphopsia lingering for relatively short periods of time only (e.g., Bodamer, 1947; Geyer, 1963). The dependency of normal spatial configurations of facial gestalts on network properties is also confirmed by our finding that left-sided lesions do not always lead to right-sided hemicases and vice versa, as one would expect on the basis of retinotopic representation in extrastriate cortex. This is especially true in connection with lesions to the splenium of the corpus callosum and their extension to the left and right forceps major, where the left side produces almost as many right-sided as left-sided distortions, while the right side mostly produces right-sided distortions. Given the function of the corpus callosum as the communication highway connecting the two hemispheres, this may not be surprising, but even lesions to occipital, temporal, and parietal areas do not always generate contralateral distortions.

4.2. Cerebral asthenopia

Further evidence for the face-gestalt network's dynamic network properties comes from a phenomenon that in the older literature is called 'cerebral asthenopia' (i.e., heightened fatigability; Willanger & Klee, 1966). Thus, Ganssauge, Papageorgiou, and Schiefer (2012) reported three patients whose perception of facial distortions only kicked in after having fixated on their interlocutor's nose for several seconds. Nijboer, Ruis, van der Worp, and de Haan (2008) studied two adults who started to experience facial distortions 'within seconds', and, describing a female patient, Dalrymple et al. (2014) explained that '*faces appear normal when she first sees them and then become progressively more distorted*'. In the latter case, the distortions built up over a time span of 5–10 sec. Upon looking away, faces would become normal again, and then started to distort once more after 5–10 sec. Likewise, Bala et al. (2015) described a patient whose '*symptom intensity not only fluctuated during her hospitalization but also was changing dynamically while she was watching a face for a longer time*,' quoting her as saying that, '*The longer I watch, the more the face changes, some elements change and the overall face is different*'. Another patient described earlier by our group, typically saw faces change after several minutes (Blom, 2020; Blom et al., 2014). In all, these observations indicate that the face gestalt network is capable of effectively rerouting information for the purpose of maintaining normal face perception - even in the event of massive tissue loss, as indicated by the aforementioned lesion studies. The asthenopic descriptions further hint at the importance of functional properties of the network. The implication is that the normal perception of face gestalts is subserved by a densely interconnected network with considerable adaptability of local functions, although the exact nature of this adaptability is in need of further elucidation. It may also be seen as evidence in favour of the existence of 'face templates' in that the face-selective network generates closed, intrinsic functional states that are not actively *created* by sensory input but merely *modulated* by it. This will be worked out below.

4.3. Face recognition

Remarkably, in the cases we reviewed, even profound facial distortions rarely affected the patients' ability to recognise faces. This might well indicate that the networks for face feature and gestalt perception, and for face recognition do indeed hardly overlap. Alternatively, it may suggest that prosopagnosia may have gone unexplored, undetected, or unreported since its presence or absence was not always mentioned in the original texts. Moreover, several patients divulged that, even though faces did become distorted beyond recognition, they could still recognise people by the

sound of their voice, their clothes, or typical attributes such as glasses or a pony tail. This is a common strategy of patients with prosopagnosia and strongly suggests that their recognition of faces must have been impaired (Ahmed & Hodges, 2020). Other patients may have benefitted from the seconds to minutes they had before faces started to distort, giving them sufficient time to identify people before they were unable to do so any longer. A related finding is that some patients recounted they were unable to recognise faces except in photographs. Since one patient Hécaen et al. (1952) described reported that she had no difficulty in recognising faces in photographs '*because of the immobility of the features*', we may ask ourselves whether such cases may perhaps be due to adjuvant malfunctioning of the motion centre, V5, or the V3 areas involved in dynamic form perception. As a case in point, we ourselves once saw a patient (not included in the present case series) who suffered from a trailing phenomenon, accordingly seeing people's lips move discontinuously while hearing their speech without any interruptions. This experience had a profoundly alienating effect on her, comparable to watching actors in a dubbed motion picture. It is not unthinkable that variations of this mechanism may prevent patients from recognising actual faces with all their moving features, while leaving intact the ability to do so in stills. As we shall see below, another explanation may be damage to parts of the face-selective network that have a function in processing dynamic aspects of faces, notably those in the so-called dorsal stream.

4.4. *Familiar versus unfamiliar faces*

In the early case description Lenz (1909) recorded, he detailed that his patient curiously experienced distortions '*when looking at familiar faces*'. Well over a century later, Dalrymple et al. (2014) likewise described a patient who saw familiar faces distort more than unfamiliar ones. Jiang, Sasikumar, and Jin (2017), on the other hand, reported patients seeing facial distortions that were worse in strangers. An explanation for people noticing distortions more readily in familiar faces may be that especially subtler types of distortion (e.g., looking thinner or older) may go unnoticed in unfamiliar faces (Landi & Freiwald, 2017). However, in the cases cited above, distortions were typically far from subtle. An alternative explanation may therefore be that familiar and unfamiliar faces are processed by different parts of the face-gestalt perception network. Indirect evidence for this comes from an fMRI study in macaques, which not only showed that familiar faces engage the face-selective network to a larger extent, but also that two temporal areas outside the core system are recruited, tentatively called the 'familiar face recognition system' (comprising the perirhinal cortex and temporal pole; Landi & Freiwald, 2017). Whether a similar neural system exists in humans is as yet unknown, but, judging by the examples mentioned above, this may well be possible.

4.5. *Own versus others' faces*

The existence of neural structures dedicated to the recognition of either familiar or unfamiliar faces would also make sense given that some patients suffer solely from auto-prosopometamorphopsia, whereas others struggle with hetero-prosopometamorphopsia, and some with both types of distortion. After all, one's own face is probably the best known face to anyone, at least since the 19th century when mirrors became a household product. However, the four cases of pure auto-prosopometamorphopsia in our series hardly give us any clues as to the neuroanatomical correlates of this specialisation. One case of hemi-auto-prosopometamorphopsia described by Imai, Nohira, Miyata, Okabe, and Hamaguchi (1995) was attributed to a very localised spotty infarct between the left retrosplenium and the cingulate gyrus. Due to its specificity, it is in fact the only clue we get from the present case series for a possible structure devoted to 'own face'. A similar case described by Geyer (1963) and two by Blom (2020) were inconclusive regarding localisation. A possible reason for the lack of auto-prosopometamorphopsia cases in our series may be that, in an evolutionary sense, areas devoted to own (versus other's) face may be a relatively late addition to the face-selective network, possibly branched from areas devoted to familiar faces. Alternatively, auto-prosopometamorphopsia may simply have been under-reported in the cases we collected, since not all papers mentioned explicitly whether this symptom was present or not. In either case, the neural correlates of own-versus-other face perception remain to be elucidated, especially since a prior review by Devue and Brédart (2011) found evidence for the involvement of a complex bilateral network comprising frontal, parietal, and occipital areas in own-face recognition that did not allow for easy determination of the contribution of each recruited brain area.

4.6. *Role of the fusiform gyrus*

Another remarkable finding from our investigations is that the role of the fusiform face area in mediating prosopometamorphopsia would seem to be relatively small. In their groundbreaking work, Sergent, Ohta, and MacDonald (1992) were the first to chart object- versus face-specific brain areas with the aid of PET, identifying a circumscript area located in the right inferior temporal lobe (later termed the 'fusiform face area' by Kanwisher, McDermott, & Chun, 1997) which they found to be central to face perception and recognition. In their study they found that specifically the structure located in the right middle fusiform gyrus consistently showed activity when healthy volunteers viewed images of faces and face-like configurations, whereas it did not when the participants were looking at images of other objects such as houses or shoes. Since then, their finding has been replicated by numerous other imaging studies with varying study designs. However, in the patients analysed here, prosopometamorphopsia was attributed to pathology of the right fusiform gyrus in three cases only (Seron et al., 1995; Miwa & Kondo, 2007; Dalrymple et al., 2014), and to pathology of the *right temporal lobe* in two patients (Jung et al., 2018; Quevedo-Diaz et al., 2018), as well as to pathology of *right temporo-occipital or temporo-parietal regions* in eight others. While the latter two groups may also have contained cases of pathology to the right fusiform gyrus, its involvement is merely certain in as few as three of the 81 cases (4%) we reviewed, and, if we would allow all 13 cases to count, still only in 17%, which is extremely low for a region that is

considered to play a key role in the face-selective network. This observation adds weight to the conclusion that face gestalt processing is dissociated from other aspects of face processing (such as face recognition) and that the role of the fusiform face area is limited to the processing of non-structural facial aspects such as face identity and facial expressions. Additionally or alternatively, we can postulate that the function of the right fusiform gyrus in face gestalt processing (contrary to the situation in face *recognition*) is easily taken over by other parts of the network. An obvious candidate structure would then be the left fusiform gyrus, even though in right-handed individuals that region is believed to exert predominantly linguistic rather than face-specific functions (Allison, Puce, & McCarthy, 2002; Dehaene et al., 2010; Dundas, Plaut, & Behrmann, 2013). A non-face-specific function of the left fusiform gyrus is consistent with our finding that the left temporal lobe was not implicated in any of the cases of prosopometamorphopsia that we evaluated. Moreover, it is in line with an experimental study by Rangarajan et al. (2014), who implanted subdural intracranial electrodes over the fusiform gyrus in 10 patients undergoing invasive intracranial monitoring for intractable epilepsy, and found a clear functional dissociation between the right and left fusiform gyrus, with the first region playing a part in the processing of facial information, and the latter in language.

4.7. *The face-selective network*

Duchaine and Yovel's (2015) face-selective network model, in itself an extension of the model proposed by Haxby et al. (2000), primarily suggests the involvement of the occipital face area, fusiform face area, and posterior superior temporal sulcus face area, all with bilateral involvement yet right-sided dominance (in right-handed individuals). It also suggests that face-specific information can enter the system via multiple pathways, starting from early visual cortex, from whence it is then thought to be relayed to the aforementioned face-specific regions in a serial fashion. Regarding the function of these regions, the authors propose that the right fusiform gyrus (either the middle part or the middle and posterior part) is involved in the processing of face identity. As noted above, this might account for why we ourselves found so little involvement of the fusiform face area in connection with prosopometamorphopsia. It might also be the reason why agnosia for faces was so rare in the case series presented here and why comments on facial expression, although not assessed systematically in the original studies, were far outnumbered by those on shape and gestalt. Citing studies by Fox, Moon, Iaria, and Barton (2009) and Said, Haxby, and Todorov (2011), Duchaine and Yovel (2015) further suggest that the posterior superior temporal sulcus is involved in the processing of dynamic facial information such as facial expression, while, on the other hand, they consider the occipital face area to be primarily involved in the processing of static features. In some other studies, the inferior frontal gyrus has been implicated in the processing of dynamic facial features, also and specifically in the processing of eyes, although evidence for the latter claim is weak and further studies are necessary (Chan & Downing, 2011). In their model, Duchaine and Yovel further mention the anterior temporal lobe and the anterior superior temporal sulcus but note that their roles in the processing of facial information is still far from clear. In all, they organise the face-selective network into a ventral and a dorsal stream, with the ventral stream (comprising occipital face area, fusiform face area, and anterior temporal lobe) playing a role in the processing of form information, i.e., invariant features such as sex, age, and identity, and also facial expression; and the dorsal stream (comprising posterior superior temporal sulcus, anterior superior temporal sulcus, and inferior frontal gyrus) serving the processing of information from dynamic faces, including expression, eye gaze, and mouth movements. Moreover, they posit that information in both streams is processed in a parallel, hierarchical fashion, running from occipital towards more frontal areas.

4.8. *Refining the model*

As noted above, the evidence reviewed is insufficient to determine the anatomical relationship of face-gestalt processing to current face-selective network models. However, we are able to make general statements about the face-gestalt network that help refine existing accounts. The findings presented here suggest that face gestalt processing is probably greatly facilitated by interhemispheric transfer, with the splenium of the corpus callosum playing a key role in connecting specialised components on either side in such a way that they function in a concerted manner to maintain the capacity for perceiving normal face gestalts under all but the most severe circumstances. After all, it is hard to explain why else facial distortions would be so rare (even in the likelihood of underreporting) and why the numerous brain disorders affecting people every day would not do any more damage to face perception. Elaborating on a hypothesis proposed by Llinás and Ribary (1994) about perceptions as closed, intrinsic functional states, we pose that the face-gestalt related components of the face-processing network are involved in the *active and independent creation of facial images*, irrespective of the presence or absence of any visual input. We conceptualise the images thus created as 'face templates', not composed of pieces of information from the outside world, but rather created internally, fully formed, and merely *modulated* by information relayed by the retina, the optic tract, and primary visual cortex towards the face-selective system as a whole. Thus, we hypothesise that information from the outside world has the function of *restricting the degrees of freedom* of the face-gestalt network, and thus of preventing it from creating just any random face configuration on the basis of its generic 'face template', which it indeed does in the presence of rudimentary input signals and under very poor lighting conditions (Caputo, 2010). This would go a long way to explain why facial distortions are so rare, why human faces - in even rarer cases - can be perceived as fully-formed animal faces, why low-level visual input has the capacity to evoke facial illusions and pareidolias, why we see 'faces in the dark' before falling asleep, why entry points into the network are probably indeed manifold, as suggested by Duchaine and Yovel (2015), and why even large-scale tissue loss would hardly seem to impair the network's function in creating fully formed, coherent faces - even though in some cases, such as in prosopagnosia and Capgras' syndrome, the recognition or identification of faces may be problematic.

4.9. Roles of individual network components

We have insufficient anatomical data to allocate specific face-gestalt symptoms to specific regions of the face processing network. However, we can draw conclusions about the contribution of individual hemispheres and interhemispheric transfer. We found unilateral disruptions of the network on either side to produce unilateral facial distortions, with sidedness following no strict rules. The suggestion is that normal face gestalt processing requires balanced network activity across the two hemispheres and that it can be disrupted either by network dysfunction in one hemisphere or by impaired interhemispheric communication due to a splenial lesion. The ipsilateral predominance of hemi-prosopometamorphopsia suggests that spatial configuration information from the intact hemisphere dominates the lesioned hemisphere, leading to distortions in the hemifield ipsilateral to the lesion. On the basis of a single finding, we also cautiously suggest that the inferior frontal gyrus may play a role in fusing left and right hemifaces into a coherent whole. As noted, frontal involvement featured in only four of the cases we collected. If our own case (no. 4, Table 2) of isolated frontal pathology is anything to go by, the left frontal lobe may indeed be involved in fusing hemi-images of faces into a coherent whole, with malfunctioning resulting in hemi-prosopometamorphopsia with detachment of the two halves. That said, two other patients from our case series reported this kind of midline detachment while showing no demonstrable frontal pathology (one had mild global atrophy, the other was suspected of epilepsy despite a normal EEG). In all, the function of the frontal cortex in face-gestalt perception is in need of further elucidation. The same holds true for the role of right parietal areas, which were found to be compromised in a patient who experienced horizontal splitting while suffering from a tumour in the right parietal lobe (Young et al., 1989). Regarding the white-matter pathways connecting the various hubs in the face-selective network, Geschwind, Pourtois, Schwartz, van de Ville, and Vuilleumier (2012), using diffusion tractor imaging in 22 healthy participants, found evidence of strong connections between (especially) the right-sided occipital and fusiform face areas along the ventral visual pathway. However, they found no evidence of comparable connections between these areas and the remainder of the network, which instead appear to be connected via a dorsal system that includes the superior temporal sulcus and fronto-parietal areas. On the basis of the strong network properties that we found, as well as the central role of the corpus callosum, we suggest that the corpus callosum might well play a much larger role in the perception of normal face gestalts than hitherto suspected.

4.10. Limitations

Due to the modest number of published cases and the lack of uniformity in the reports, our synthesis of prosopometamorphopsia is somewhat limited. Most of the original studies only allowed us to obtain rather rough indications of the brain regions involved since most did not describe specific structures such as the fusiform or occipital face areas, while 25% of the original accounts (especially the older ones) lacked neuroimaging data altogether. As a consequence, we were unable to make anatomical claims for neural representations beyond inferences about gross hemispheric or lobar associations derived from the data reviewed. Secondly, since we also found case reports under titles such as 'hallucination', 'optical agnosia', and 'prosopagnosia', we suspect that our review, although the most comprehensive to date, is probably not exhaustive. Especially cases from the literature on hallucinogen persisting perception disorder (HPPD, the drug-induced variant of Alice in Wonderland syndrome) were conspicuously lacking. Finally, since handedness was not mentioned in many of the original reports and we consequently decided to leave this variable out of the equation, we were forced to allow for a 10% error margin in our calculations on the sidedness of the face-selective network (based on the fact that 10% of all people are left-handed).

5. Conclusions

On the basis of an analysis of eight new cases of prosopometamorphopsia and 73 cases described over the past century, we conclude that the face-gestalt network is widely disseminated and densely interconnected, probably due to the central role of the corpus callosum with its unsurpassed connecting properties. We hypothesise that the network is involved in creating internally mediated, generic 'face gestalt templates', which are modulated rather than created by visual input from the outside world. Our evaluation suggests that bilateral facial distortions are primarily associated with right-sided and bilateral occipital lesions while unilateral facial distortions, typically ipsilateral to the lesion, are associated with left-sided occipital lesions and lesions to the splenium of the corpus callosum. Colour changes in faces are also primarily seen in connection with occipital lesions. We found little involvement of the right fusiform gyrus in prosopometamorphopsia. This is in line with the well-established role of the fusiform face area in face recognition and points to a dissociation between spatial gestalt and recognition aspects of face processing that has not previously been recognised. That the left fusiform gyrus was not involved at all, is consistent with its purported role in linguistic rather than face-selective processes. We also found preliminary evidence for the involvement of left frontal cortex in the fusing of vertical hemi-images of faces, and for right parietal areas in fusing horizontal hemi-images. Further evidence for the interconnectedness of the face-selective network comes from the surprisingly good recovery rates reported, even in cases of gross anatomical damage irrespective of underling pathology, which demonstrates that the system as a whole, even in long-lasting cases of prosopometamorphopsia, is capable of fully restoring the perception of faces. The curious phenomenon of cerebral asthenopia and the ipsilateral hemifield predominance of hemi-prosopometamorphopsia are also consistent with this view. Finally, our analysis indicates that specialised networks for the representation of familiar-versus-unfamiliar faces, and for own-versus-other face, probably exist outside the core system, although the location of these networks and their connection with the core system are in need of further study.

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Credit author statement

Jan Dirk Blom: Conceptualisation, Methodology, Data acquisition, Formal analysis, Original draft preparation - Writing, Reviewing and Editing, Visualisation, Supervision; **Bastiaan C. ter Meulen:** Data acquisition, Writing - Reviewing and Editing; **Jitze Dool:** Data acquisition, Writing - Reviewing and Editing; **Dominic H. ffytche:** Writing - Review and Editing, Validation, Supervision.

Declaration of competing interest

The authors report no competing interests.

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Supplementary data

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