REVIEW

Progress in perceptual research: the case of prosopagnosia
[version 1; peer review: 2 approved]

Andrea Albonico, Jason Barton

Human Vision and Eye Movement Laboratory, Departments of Medicine (Neurology), Ophthalmology and Visual Sciences, Psychology, University of British Columbia, Vancouver, Canada

Abstract
Prosopagnosia is an impairment in the ability to recognize faces and can be acquired after a brain lesion or occur as a developmental variant. Studies of prosopagnosia make important contributions to our understanding of face processing and object recognition in the human visual system. We review four areas of advances in the study of this condition in recent years. First are issues surrounding the diagnosis of prosopagnosia, including the development and evaluation of newer tests and proposals for diagnostic criteria, especially for the developmental variant. Second are studies of the structural basis of prosopagnosia, including the application of more advanced neuroimaging techniques in studies of the developmental variant. Third are issues concerning the face specificity of the defect in prosopagnosia, namely whether other object processing is affected to some degree and in particular the status of visual word processing in light of recent predictions from the “many-to-many hypothesis”. Finally, there have been recent rehabilitative trials of perceptual learning applied to larger groups of prosopagnosic subjects that show that face impairments are not immutable in this condition.

Keywords
face recognition, neuroimaging, diagnosis, rehabilitation, object recognition
The face is a complex structure. It has a complicated three-dimensional shape, a substantial degree of mobility, and structural constraints that make all faces fairly similar; all of these issues present challenges to a perceptual system. Nevertheless, perhaps because of the social importance of faces, humans have developed the ability to recognize faces rapidly and accurately and with seemingly little effort. Indeed, recent estimates are that the typical person can remember and recognize about 5000 faces¹.

However, for some people, face recognition is not so easy. Prosopagnosia is a condition marked by the loss of familiarity for faces and the consequent inability to identify people by their faces². Although prosopagnosic subjects frequently turn to other cues such as voice, hairstyle, or anomalous facial features, these strategies have their limitations; as a result, prosopagnosic subjects still often find social situations stressful, and recent work has shown that they can suffer from anxiety, depression, and social withdrawal³,⁴.

Studies of prosopagnosia have a time-honoured place in research on face recognition. Neuropsychological observations have played key roles in the development of cognitive models of face processing⁵ and pointed to the cerebral substrates of face recognition⁶. Even in an era when advances in face research are coming from psychophysics, functional neuroimaging, and primate neurophysiology, there are still important contributions from work on prosopagnosia. This has been spurred particularly by the recognition of a developmental variant⁷. Although acquired prosopagnosia is rare, developmental prosopagnosia appears to be more common but debate on its exact prevalence continues⁸. Nevertheless, the greater availability of developmental subjects has led to an increase in the number of prosopagnosic studies. In this review, we focus on four areas of recent progress in the fields of acquired and developmental prosopagnosia.

The diagnosis of prosopagnosia
Uniform definitions are a starting point for research into a condition. The core deficits in prosopagnosia are the loss of familiarity for previously known faces and the inability to learn to recognize new faces. In the past, this was often shown by tests using famous faces or in case studies by demonstrations that the subject could not recognize friends or family members. However, it is difficult to derive uniform diagnostic criteria from such tests. Familiarity for famous faces is affected by the subject’s age, culture, education, and interests, for example, and carefully matched controls are essential for interpreting the results of such tests. This has led to supplementation of famous face tests by the increasing use of tests that assess short-term familiarity. These show faces in a learning phase and then present these “target” faces along with new “distractor” faces in a test phase in which subjects are asked to indicate which were the faces they had learned. The most well-known examples are the Warrington Recognition Memory Test⁹ and the Cambridge Face Memory Test¹⁰, the latter of which has the desirable feature of testing recognition across changes in pose or lighting. Compared with tests that use famous or personally known faces, tests of short-term familiarity provide limited exposure and lack the semantic and perceptual richness of long experience but have the advantage of uniformity in the degree of learning and testing. For the Cambridge Face Memory Test, there has also been substantial normative work showing good internal consistency (Cronbach’s alpha ranges from 0.83 to 0.89) and no effects of intelligence or the ethnic mix of faces in the subject’s life experience. There is a very modest advantage for women but a more significant effect of age in that accuracy declines for those over the age of 50¹¹⁻¹³. Also, versions of this test have been developed for use in children¹⁴.

There are many other tests of face processing and these were recently reviewed in detail and categorized¹⁵. Diagnostic tests can be divided into three main types: (a) tests of face perception, which can include detecting faces in arrays or discriminating or matching simultaneously seen faces; (b) tests of face recognition, such as the tests for short- and long-term familiarity which were discussed above; and (c) tests of face identification, which involve naming or providing other information learned about the person whose face is shown. Prosopagnosic subjects are impaired on both recognition and identification. Performance on tests of face perception can be used to differentiate between prosopagnosic subjects who have an apperceptive variant, in which there is an under-specification of facial structure by perceptual processing, or an associative or amnestic variant, in which the problem is not perception but the ability of perceptual information to access facial memories¹⁶. Examples of tests assessing face perception are the Benton Facial Recognition Test¹⁷, the Cambridge Face Perception Test¹⁸, the Glasgow Face Matching Test¹⁹, and the Caledonian Face Test²⁰. Tests of face imagery have also been used to clarify the status of facial memories and diagnose the amnestic variant²¹.

Self-report questionnaires are becoming more common tools in diagnosing prosopagnosia. They are quick and easy, do not require equipment, do not need to be done in person and hence can be used to screen a large number of subjects, even at a distance. Among those are the Kennerknecht 15-item questionnaire²², the 20-item Prosopagnosia Index²³, and the Cambridge Face Memory Questionnaire²⁴. A potential concern is that individuals may have only modest insight into their face recognition abilities²⁵,²⁶, particularly children²⁷, although some studies suggest that this might not be the case for adults using the Prosopagnosia Index²⁸. This concern might account for the fact that questionnaires may have high reliability but only modest sensitivity and specificity for diagnosing prosopagnosia²⁵. Because of these concerns, some have advocated that questionnaires always be supplemented by objective tests for diagnosis²⁹,³⁰.

Recent reviews have discussed how to incorporate these various instruments into a diagnostic approach. This may be less of an issue for acquired prosopagnosia, in which the combination of an appropriate lesion on imaging, the subject’s awareness of a change in face recognition after lesion onset, and poor performance on an objective test of face recognition makes the diagnosis plausible. For developmental prosopagnosia, there are no definite structural or genetic markers at present and so its diagnosis still rests solely on behavioural tests. One review
pointed out the wide variations between studies in the types of
tests, the number of tests, and the statistical cutoffs used. This
creates variable confidence in the diagnosis and introduces
heterogeneity that can confound comparisons across groups
and studies, an obstacle to scientific progress. As a result,
there have been proposals for more uniform diagnostic criteria.2,3,4
These include (i) subjective difficulty recognizing faces in daily
life; (ii) objectively impaired face recognition on at least two
tests of face recognition and criteria of at least 2 standard deviations
below control means; (iii) intact general perceptual and
memory function; and (iv) exclusion of other disorders associated
with impaired face recognition, such as autism spectrum
disorders.

Although reaching a firm diagnosis of developmental prosop-
agnosia has its hurdles, a recent study using qualitative methods
suggested that screening for it may be possible with a simple
list of 16 “hallmark symptoms” from experiences in daily life,
which anyone can review.27 The utility and sensitivity of this
approach need to be explored.

The neural basis of prosopagnosia

The older literature has shown that lesions of acquired prosop-
agnosia are bilateral2–4 or limited to the right hemisphere5,6,7,8,9
and reports of left-sided lesions alone are rare10–12. This is
consistent with evidence from functional neuroimaging that face
processing induces greater activation in the right hemisphere13.
The areas involved are the ventral occipito-temporal and fusiform
cortex or anterior temporal cortex or both. These anatomic
variants may correspond to functional variants8,10. Individuals
with occipito-temporal or fusiform lesions are more likely to have
an apperceptive variant11, whereas those with anterior tempo-
ral lesions have an amnestic variant along with better perceptual
function and more difficulty with face imagery12.

Although by definition subjects with developmental prosopagnosia
do not have large visible lesions, the status of their face process-
ing networks can be studied with more subtle neuroimaging
methods, including measures of cortical thickness, the degree
of functional activation, and connectivity within the network.
The results as they currently stand are not conclusive. There are
two main views. One proposes that developmental prosopagnosia
is marked by alterations in various regions of the face
network, particularly the fusiform gyrus, changes such as
reduced cortical thickness or density14–16, reduced face selectivity
of their activation17–21, local white matter abnormalities on
diffusion imaging14–16, or reduced feedforward connectivity from
early visual to occipito-temporal cortex22. The second proposes
a disconnection between posterior and anterior regions within
the face network23–25, on the basis of observations of preserved
activation of the fusiform and ventral occipito-temporal
cortex by faces26–28 and abnormalities in long white matter tracts
that link posterior and anterior temporal cortex29–31.

Comparisons with other developmental disorders might be
informative. Researchers on dyslexia have suggested a model
in which a general risk for cortical anomalies is modulated by
other genetic and/or environmental factors that determine
the location and extent of such anomalies32. The latter deter-
mines the specific syndrome and can explain the frequent
co-association of developmental disorders. In this regard, we
note recent observations of associations between congenital
amusia and developmental prosopagnosia33,34. Along these lines,
others have speculated that abnormal neural migration may be
responsible for developmental prosopagnosia3.

Does developmental prosopagnosia have a genetic cause? Face
recognition abilities show a high degree of heritability in the
general population35,36, and early observations were that devel-
opmental prosopagnosia tended to run in families37,38, possibly
with an autosomal dominant pattern of inheritance39,40. However,
most neurodevelopmental disorders are polygenic combi-
nations of allelic variants present in the normal population.
Along these lines, a recent study of 24 subjects reported that
common single-nucleotide polymorphisms in the oxytocin recep-
tor gene are associated with developmental prosopagnosia41.
These preliminary results require replication in larger samples.

Is prosopagnosia only about faces?

A long-standing controversy is whether the impaired recogni-
tion in prosopagnosia is face-specific or affects other object
types. This has important theoretical implications for how object
recognition is organized in the visual system. The distributed
view suggests that object processing is performed by networks
of visual regions, and that some of these regions are involved
in the perception of several types of stimuli42–44. The modular
view claims that different categories of objects—particularly
faces—are processed by distinct dedicated cortical regions45–47.

Case studies of acquired prosopagnosia have produced mixed
results; some reported normal recognition of exemplars of other
objects48–50, and others showed impairments45,51,52. A recent
major review53 examined 238 cases of developmental prosop-
agnosia in the literature. The majority of subjects had evidence
of impaired object recognition, although a smaller number
had reasonable evidence that object recognition was intact,
given that they had both good accuracy and normal reaction
times on tests. Although the authors concluded that the frequent
association of face and object impairments supported a shared
mechanism for recognizing faces and other objects54,55, the
challenge for any comprehensive explanation is to account for
both frequent associations and occasional dissociations. One of
the most useful aspects of this review was the collection of
accompanying commentaries56–104, which suggested both various
hypotheses to explain this fact and methodologic limitations in
the currently available data that need to be addressed in future
work to allow a more definitive set of conclusions to be drawn.

A particular object type deserves comment—namely, words. One
of the difficulties in comparing faces and objects is that humans
have a great deal of experience and expertise with faces but such
expertise cannot be assumed for other object types. Take cars,
for example. A recent study found that, as a group, subjects
with developmental prosopagnosia tended to score low on the
Cambridge Car Recognition Test but that individual scores ranged quite widely, from excellent to poor. However, not everyone is a car expert and variable expertise could affect recognition performance. In another group of studies, when visual car recognition scores were adjusted for car expertise, as reflected by a subject’s semantic knowledge about cars, subjects with both acquired and developmental prosopagnosia tended to perform worse than expected.

In literate societies, visual words, in contrast to cars, are a category for which almost all subjects have considerable perceptual expertise. The “many-to-many hypothesis” proposes that face and visual word processing share and compete for neural resources in regions like the fusiform gyrus and that structural constraints cause visual words to be processed more on the left, in proximity to language processing, and faces secondarily to lateralize to the right. Lateralization is incomplete, though, and functional imaging shows overlap between face- and word-activated voxels. As a consequence, the hypothesis predicts that prosopagnosia from right lesions should be accompanied by mild reading deficits in the processing of words and that alexia from left lesions should be accompanied by mild face recognition problems. Whereas one study of three subjects with acquired prosopagnosia did show mild word recognition deficits, other studies of visual word processing in acquired prosopagnosia from right-sided lesions alone have not found impaired reading and the same is true for developmental prosopagnosia. On the other hand, the type of processing that is performed on words and faces may differ by hemisphere. Although subjects with acquired prosopagnosia from right-sided lesions may read normally, they often have trouble recognizing handwriting or font, and subjects with alexia from left lesions should be accompanied by mild face recognition problems. Whereas one study of three subjects with acquired prosopagnosia did show mild word recognition deficits, other studies of visual word processing in acquired prosopagnosia from right-sided lesions alone have not found impaired reading and the same is true for developmental prosopagnosia. On the other hand, the type of processing that is performed on words and faces may differ by hemisphere. Although subjects with acquired prosopagnosia from right-sided lesions may read normally, they often have trouble recognizing handwriting or font, and subjects with alexia from left lesions should be accompanied by mild face recognition problems.

Can prosopagnosia be treated?

Spontaneous resolution of acquired prosopagnosia is rare, and developmental prosopagnosia is a lifelong disorder. Hence, means of improving face recognition skills in these populations are of clinical interest. But can it be done? Neuroimaging shows that face processing activates a widely distributed network, including occipito-temporal, superior temporal, anterior temporal, and inferior frontal regions in both hemispheres, though more on the right. It is highly unlikely that acquired lesions will eliminate all components of this network; furthermore, some studies in developmental prosopagnosia continue to show activation of this network by faces. The open question is whether surviving components of the face network in a given prosopagnosic subject have any capacity for functional reorganization or modulation that could allow face recognition to improve through a rehabilitative approach.

Most work has focused on behavioural interventions, although there is one intriguing report of transient improvement of developmental prosopagnosia after intranasal inhalation of oxytocin. These rehabilitative attempts have been reviewed in detail. Approaches can be divided into compensatory strategies, which aim to achieve person recognition by circumventing the face processing impairment, and remediation, which aims to improve that impairment. In terms of the process targeted, they can also be divided into those that focus on enhancing mnemonic function, which has been used in a few case studies, and those that target perceptual function. As examples of the latter, a few older case studies attempted to enhance attention to facial features, though results on face recognition were variable.

The most significant recent advances have been trials of perceptual learning in groups rather than single cases of prosopagnosia. In one study of 24 subjects with developmental prosopagnosia, subjects learned over the course of 2 weeks to discriminate distances between facial features, namely the distance between the eyes and eyebrows or between the nose and the mouth. These “spatial relations” can be thought of as indices of the complex geometry of faces, and studies show that some people with prosopagnosia are impaired in perceiving them. This trial found improved face perception (but only if the test faces had a similar frontal view) and some modest improvements in subjective reports of daily experience with faces. A second study of 10 subjects with acquired prosopagnosia used morphed faces to train subjects over the course of 11 weeks to perceive finer and finer differences in facial shape; at the same time, the study introduced irrelevant variations in the expression and viewpoint of the face. In these subjects, compared with a control condition, there was a 21% absolute increase in perceptual sensitivity to facial shape after training, which generalized over new views and expressions. Importantly, there was also a 10% increase for new faces on which subjects had not trained, indicating that subjects were acquiring new skills rather than just learning a set of faces. The effects of training were still evident 3 months later. Although some but not all subjects related anecdotes pointing to improved face recognition in daily life, future studies will require formal evaluation of real-life benefit before such methods are translated to the clinic.

These rehabilitative studies represent a starting point. Although neither training method represents a “cure”, they provide evidence that face processing can be changed in prosopagnosia. They also suggest that there may be individual differences in training potential. Further work is required to determine whether the perceptual gains from learning can be augmented further by better training design or the use of adjunctive methods to promote plasticity during learning.

Grant information

This work was supported by the Natural Sciences and Engineering Research Council of Canada (RGPIN 319129) and Canada Research Chairs (950-228984).

The funders had no role in study design, data collection and analysis, decision to publish, or preparation of the manuscript.


Page 8 of 9
Open Peer Review

Current Peer Review Status: ✔ ✔

Editorial Note on the Review Process

F1000 Faculty Reviews are written by members of the prestigious F1000 Faculty. They are commissioned and are peer reviewed before publication to ensure that the final, published version is comprehensive and accessible. The reviewers who approved the final version are listed with their names and affiliations.

The reviewers who approved this article are:

1. Richard Cook
   Department of Psychological Sciences, Birkbeck, University of London, London, UK
   **Competing Interests:** No competing interests were disclosed.

2. Galia Avidan
   Department Psychology, Ben-Gurion University of the Negev, Beer-Sheva, Israel
   **Competing Interests:** No competing interests were disclosed.

The benefits of publishing with F1000Research:

- Your article is published within days, with no editorial bias
- You can publish traditional articles, null/negative results, case reports, data notes and more
- The peer review process is transparent and collaborative
- Your article is indexed in PubMed after passing peer review
- Dedicated customer support at every stage

For pre-submission enquiries, contact research@f1000.com