Article type: Focus Article

Article title: Guidelines for studying developmental prosopagnosia in adults and children

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Abstract

Developmental prosopagnosia (DP) is a neurodevelopmental condition characterized by severe face identity recognition problems that results from a failure to develop the mechanisms necessary for adequate face processing¹. It occurs in children and adults with normal visual acuity, and without intellectual impairments or known brain injuries. Given the importance of face recognition in daily life, and the detrimental effects of impaired face recognition, DP is an important area of study. Yet conventions for classifying individuals as DP for research purposes are poorly defined. In this focus paper we discuss: 1) criteria for an operational definition of DP; 2) tests of face recognition, and conventions for classifying individuals as DP; and 3) important considerations regarding common associations and dissociations, and cognitive heterogeneity in DP. We also highlight issues unique to studying DP in children, a relatively new endeavour that is proving to be an important complement to the work with adults. Ultimately we hope to identify challenges researchers face when studying DP, and offer guidelines for others to consider when embarking on their own research pursuits on the topic.

Developmental prosopagnosia (DP) is a neurodevelopmental condition characterized by severe face identity recognition difficulties that occur when face processing mechanisms fail to adequately develop¹. DP occurs in the absence of brain injury, in people without intellectual impairment and with otherwise typical vision. Sometimes referred to as congenital prosopagnosia^[1]³, DP has been estimated to affect 2-2.9% of the population^{4,5}. Face recognition deficits in DP can be as severe as those reported in acquired prosopagnosia, which results from brain damage¹. Studies on the psychosocial consequences of DP in adults and children indicate that DP can have a profound impact on social and psychological functioning, creating difficulties making and maintaining relationships, participating in social activities, as well as increased levels of anxiety⁶⁻⁸. Given the importance of face recognition in daily life and the detrimental effects of DP, it is particularly important to discover what underlies face recognition deficits and, in doing so, inform potential treatment strategies. Studying DP also has the potential to provide information about the development of high-level vision, and other developmental disorders.

In recent years there has been a surge of interest in developmental prosopagnosia. A Scopus search for articles or reviews that make reference to developmental (or congenital) prosopagnosia reveals 216 publications since the year 2000, a special issue in *Cognitive Neuropsychology* in 2012, and a forthcoming special issue in the *Quarterly Journal of Experimental Psychology*. This compares to only 13 publications between 1960 and 1999. Despite this interest, conventions for classifying individuals as DP for research purposes are still poorly defined, potentially limiting progress in understanding DP. In this focus paper we will discuss how to best define DP and how to determine whether an adult or child should be classified as DP in research studies. We examine commonly used tests that tap different aspects of the face recognition process, and discuss which of these seem most suitable for the measurement of abilities in adults, and also importantly, in children (see Table 1). We also discuss tests and questionnaires that measure associated abilities and disabilities (e.g., object processing, autism) which are useful in determining whether the child or adult is likely to have a broader developmental condition rather than a more “pure” case of DP (Table 1). We argue that it is important for the progress of the field to study those with relatively circumscribed face deficits, and also those with broader deficits, but to be transparent as to which individuals and/or groups are included in the sample.

[¹] “Congenital” prosopagnosia implies that face recognition difficulties were present at birth, while “developmental” prosopagnosia is agnostic as to when in the developmental process the difficulties emerge. As such, some cases where brain injury occurred early in development and resulted in face recognition difficulties have been referred to as developmental prosopagnosia (e.g., 2). However, these are excluded from our definition, which focuses on those with no known brain injury.

OPERATIONAL DEFINITION OF DP

To develop a coherent body of research on DP, it would be useful to have relatively standard diagnostic criteria across studies. One critical aspect of defining a disorder of face recognition is defining face recognition itself. Face recognition involves perceiving a face, encoding it into memory, and then subsequently retrieving that memory to determine whether the face is familiar and who it might be⁹. An impairment of face recognition could result from a failure of any one of these processes. Therefore, like other developmental disorders (e.g. autism¹⁰), there are likely to be subtypes of DP, rather than one single form. In adults, some individuals with DP have normal face *perception* despite their face *memory* deficits¹¹⁻¹⁵. This may also be the case for children, although a recent study suggests that deficits in both face perception and memory are more common in children than adults¹². To accommodate this heterogeneity, DP is best defined as a deficit of face memory, with normal *or* abnormal face perception. The reverse, impaired face perception with preserved face memory, has been reported in the literature on acquired prosopagnosia for faces that were seen prior to brain damage¹⁶. However, in DP, it seems unlikely that an individual whose face perception has always been impaired would have the opportunity to acquire accurate face memories, making face memory and face perception difficult to disentangle in DP.

Face memory deficits can be measured using tests of face recognition (see below; Table 1), but DP should also be characterised by the experience of face recognition difficulties in daily life. For some individuals with DP these experiences include failures identifying very familiar people, such as family members, whereas other individuals with DP primarily report failures recognising familiar but less often seen people, such as neighbours, when seen out of context (e.g., at the mall). Individuals with DP are also less likely to report a sense of familiarity for unrecognised faces¹⁷. However, self-report is subjective, both in terms of the report and the interpretation of the report, and individuals may have only minimal insight into their face recognition abilities¹⁸ (but see ¹⁹). For example, failing to recognize a familiar person out of context is a relatively ubiquitous experience and there is no standard for when an anecdotal report crosses into evidence of pathology in face recognition. Furthermore, some individuals with DP were unaware that the face recognition difficulties they experienced were not shared by others until they participated in testing sessions or learned about DP and realized that it explained many of their life experiences^{4, 20}. Unlike acquired prosopagnosia, in which an individual is typically aware that they have lost an ability they formerly possessed, individuals with DP do not have experience with normal face recognition to provide a point of comparison¹. Children, in particular, may lack awareness of their difficulties hence it is often parents who provide the key anecdotal evidence of their child's everyday face difficulties based on observations they have made⁶. Thus in children, identifying DP

requires keen observation by parents or teachers, combined with the knowledge of the disorder, and/or the time, resources, and skill set to research the signs and symptoms. Recently, several researchers have attempted to improve the assessment of everyday face difficulties by designing self-report questionnaires. A 15-item questionnaire¹⁹ contains questions on face identity recognition, as well others on other aspects of faces such as attractiveness and emotions. The more recent Prosopagnosia Index (PI20) consists of 20 items specific to face identity recognition, and may prove useful in screening and standardising the assessment of everyday face recognition problems²¹. No such scales are yet available for children.

We recommend that a definition of DP include objectively poor performance on tests of face memory (just ‘how poor’ is discussed below), along with a subjective feeling of repeated face recognition failures in daily life. Tests are needed because even if people know that they are poorer than average at face recognition, they may not have insight into the degree of their deficit¹⁸ (i.e., whether the classification would be as a “poor recogniser” rather than the more severe DP). Alternatively, if a person performs at DP levels on multiples tests of face recognition but reports no everyday difficulties (via interview and/or questionnaire) then they would seem qualitatively different to people who are classified as DP. As such, people who are identified via large-scale screening exercises that only include face recognition tests and no measure of subjective difficulty should only be classified as “potential” DPs. There is also the interesting case of people who report everyday difficulties but perform at typical levels on multiple tests of face recognition. These individuals are typically excluded from DP samples but it would be informative to determine why this occurs.

An additional definitional factor that is often raised regarding DP is whether the recognition deficit is specific to faces or also affects the processing of other objects. We expect that individuals with DP would be able to recognise objects at the “basic” level (e.g., bicycle, car, etc.) and this is typically the case²² when they are asked to name pictures of line drawn objects in the Birmingham Object Recognition Battery (BORB)²³ (although note that these types of tests can suffer from ceiling effects). However, it is less clear whether they should be proficient at distinguishing similar non-face objects (e.g., different bicycles). There are cases of “pure” DP where the individual does not appear to have difficulty differentiating between non-face similar objects²⁴⁻²⁶, and the double dissociation of an individual able to recognise faces but not objects²⁷. However, some people have difficulty with faces and objects^{24, 28, 29}. We argue that these people should still be considered DP. However, in these cases it is important to demonstrate that the individual can perform normally on some cognitive tasks to ensure that poor scores cannot be explained by general deficits of cognitive functioning or a failure to

understand task instructions, and that they have adequate visual acuity (see Table 1 for example tasks). This is particularly important when working with children, with whom attention and effort levels should also be carefully monitored. Moreover, it would be useful for studies to try and differentiate those with more face-specific problems from those with more general visual memory impairments, as the aetiologies of these may vary.

TESTS OF FACE RECOGNITION

One difficulty with studying DP is the lack of well-designed clinical tests that can be used to reliably evaluate face recognition^{4, 20}. To get a complete picture of facial identity recognition abilities, it is important to use multiple measures of face memory. In addition, face perception tasks are useful to help qualify the nature of the identity recognition deficits.

Face memory

Assessments of familiar face memory are an obvious choice of task because of their ecological validity. Photos of personally familiar individuals provided by a family member or taken for the purpose of the task are ideal, particularly when controlled for extra facial cues like hair, jewelry, glasses, and clothing. Drawbacks of this type of test are that it can be difficult to acquire a large number of carefully controlled photographs, the individual can often deduce who will be included in the test, and the time and effort required to make customized tests for each individual. Additionally, it is difficult to control the level of familiarity; some images may be of close friends and family, while others may be of co-workers, teachers, or acquaintances. Familiar face tasks also require a special control group of individuals who are similarly familiar with the individuals featured in the test, making it difficult to get an adequate control sample to compare to the DP's performance.

Famous faces provide an alternative method that circumvents many of these issues and can be used for multiple participants to enable comparison across DPs and with a control group. However, the issue of degree of familiarity must still be taken into account and participants should be tested later with the names of the celebrities in the task to determine whether they have had exposure to the face of the individual and could reasonably be expected to recognize them. Many famous people (especially politicians and sportspeople) are typically only famous in one or two countries, which means that these faces cannot be used across countries (although the test formats can be the same). It is also important to note that some DPs report that they have little interest in celebrities or watching TV because they find it difficult to recognize faces. These issues are exacerbated when designing tasks for children:

finding famous individuals who are familiar to children of a wide range of ages is challenging because different cohorts are often exposed to different celebrities. Another issue with famous face tests is that participants may perform well by using compensatory strategies, such as identifying distinctive features (e.g., a beauty spot on Marilyn Monroe), which may be more common on very famous faces. As a result, a poor score is often more informative than a high score: if an individual performs poorly at identifying the faces in a familiar face task despite demonstrating familiarity with the identities in the test based on names, this allows confidence that the person is impaired. However, a high score on the task could be explained by alternative recognition strategies (e.g. memory for distinctive traits of familiar faces).

Unfamiliar face memory tests can supplement tests of familiar face memory, and provide a common starting point for all participants because level of familiarity is controlled⁴. Some existing clinical tasks for adults such as the Warrington Recognition Memory for Faces Test (RMF)³⁰ and the Benton Facial Recognition Test (BFRT)³¹ have been formally evaluated and deemed to be poor measures of true face recognition^{32, 33}. Individuals with impaired face recognition can do well on these tasks by using extra-face cues (e.g. the RMF includes hair and clothing), or by matching images (e.g. the BFRT includes distinctive hairlines and uses simultaneous presentation with unlimited response times). Newer tasks, such as the Cambridge Face Memory Test (CFMT)³⁴ were carefully designed to reduce extra-face cues as well as other image cues such as lighting and low-level visual properties. In the original CFMT, participants learn six faces of adult males and are tested with a three-alternative forced choice format. There are three parts that increase in difficulty as the test progresses: the introduction involves immediate recall of individual faces while later sections involve recall of any of the six target faces and introduce changes in lighting and viewpoint and finally added visual noise (Figure 1a). Bowles and colleagues⁴ found a significant decline in performance on the CFMT after the age of 50, so performance on these tasks should be compared to age-matched controls. Alternatively, age-adjusted z-scores can be calculated from the formulas contained in Bowles et al.⁴ The CFMT is freely available to researchers and is commonly used due to its validity and reliability, which makes it useful in screening for DP^{14, 34, 35}. In addition, several versions exist to allow follow-up testing and testing of different races (CFMT-Chinese³⁶) and ethnicities (CFMT-Australian¹⁴). Another advantage of using the CFMT is that there are matched tests for within-category object recognition – the Cambridge Car Memory Test (CCMT)³⁷ and the Cambridge Bicycle Memory Test (CBMT)³⁸ (Figure 1b) although note that other tests of object recognition are available e.g., The Vanderbilt Expertise Test (VET)³⁹.

Like all tests, the CFMT does have limitations. Some people perform poorly on tests of famous face recognition, but not on the CFMT, perhaps because they have used effective compensatory

strategies, leading to an ambiguous classification^{14, 40}. In these cases, administering two different versions of the CFMT can help reach a consensus¹³. Some people will perform poorly on one of the two versions, increasing the likelihood of a correct diagnosis of DP (e.g., some Australians perform poorly on the CFMT-Australian but not the original CFMT, possibly because the faces in the former test match the ethnicity of the faces most commonly experienced in Australia and make it more difficult to match distinctive features)¹⁴. Alternatively, others will perform at “poor”, but not DP levels on either test, excluding a diagnosis of DP¹⁴. For some of these cases, the short delays between memorization and recognition in the CFMT may mask difficulties retaining faces over long delays (i.e., they should have difficulty with famous faces but not other tasks)^{14, 41}. The CFMT-Australian includes a 20 min delay section that may prove useful in disambiguating these cases¹³. As noted earlier, the faces used in CFMT tasks are carefully controlled, and it has been argued that these kinds of experimental manipulations are likely to alter the processing of *unfamiliar* faces and that tests should also be designed with more “real” faces (e.g., that include hair)⁴². It is of interest here to note that we designed a version of the CFMT (*CFMT-Films*) where the study stage involves watching short film clips of people interacting, and this correlates highly with the original CFMT¹⁷.

For children, some general neuropsychological assessment batteries do include face recognition subtests^{43, 44}, however, like the RMT and the BFRT, they include superficial cues for recognition (e.g., hair and faces of multiple races) allowing children to infer the correct answers using extra-facial information rather than normal face recognition mechanisms. New tasks of face recognition such as the Cambridge Face Memory Test for Children (CFMT for Children)⁴⁵ and the Cambridge Face Memory Test – Kids (CFMT-Kids)²⁰ (Figure 1c) have been designed. Although they both follow the same general procedures as the original CFMT for adults, the CFMT for Children uses adult faces, five targets, and two-alternative forced choice format, while the CFMT-Kids uses children’s faces (mean age = 9.3 years), six targets, and three-alternative forced choice format. The existence of own-age biases in recognition⁴⁶ suggests that it may be preferable to use children’s faces in tests for children. However, given that these biases appear to be driven by experience⁴⁷, the faces of young adults would also be suitable. The advantage of the former is that it is easier for children, but the advantage of the latter is that chance level performance is 33%, allowing a larger range of scores above floor (i.e. scores two standard deviations below the control mean yet above chance are possible, see Figure 2). Other simple tests of unfamiliar face memory exist, such as Old/New Faces⁴⁸. This task involves memorizing ten target faces and then determining which from a set of two is a target face (i.e. the “old” face). The task is easy to create and quick to administer. It is also easy to create matched tasks using different classes of objects²⁶ (e.g., horses, cars, guns, tools,

houses, glasses, etc.) though one weakness of these tests is that they currently use the same images for study and test.

Face perception

The Cambridge Face Perception Test (CFPT)⁴⁹ is a useful tool for assessing face perception ability in adult participants (Figure 1d). It involves sorting adult morph faces on a continuum from most like to least like a target. The morph faces help avoid direct comparison or matching of features from the target face to the sorting faces. Additionally, the target face is presented at 3/4-profile view while the sorting faces are presented in frontal views. A limitation of the CFPT is that it requires using the mouse to re-order the continuum, so poor scores could reflect poor motivation to shift the faces in a time-limited (one minute per trial) task. Performance on the CFPT declines by middle age, and past young adulthood females perform better than males⁴. Therefore, both sex and age should be taken into account when administering the CFPT (age- and sex-adjusted z-score calculations for the CFPT are available⁴). Even when using age-matched controls, standard tasks such as the CFPT should not be used to evaluate face perception in individuals over 80 because of floor effects⁴. For children, the Dartmouth Face Perception Test (DFPT)¹² involves deciding which of three child faces looks the most like a target face that remains at the top of the screen (Figure 1e). Loosely based on the CFPT, the target face is presented at a 3/4-profile view and the choice faces are frontal views taken from a morph continuum between the target face and a distractor face. The morph strengths of the choice faces were titrated to systematically vary task difficulty.

One concern with face perception tests that use morphs is that the faces are unnaturally similar and contain elements of the target face, reducing the ecological validity of the task. In contrast to the CFPT and DFPT, the Glasgow Face Matching Test (GFMT)⁵⁰ does not use morphs and thus avoids this issue. In this task, participants are asked to determine whether face pairs are the same individual (match), or different individuals (mismatch), with no time limit. Faces are frontal views with neutral expressions. Matched faces are images of the same individual, taken with different cameras several minutes apart. Mismatched face pairs were chosen based on similarity measures generated through pilot work. In addition to retaining ecological validity that may be absent in tasks that use morphs, one advantage of the GFMT is that it provides a format that could be easily matched to an object-matching task (tasks involving morphs are more difficult to construct using objects, though it has been done⁵¹).

Special considerations for the design of tasks for children

One major challenge for designing face recognition tasks for children is achieving a level of difficulty that will be suitable for all ages: above floor for young children, but below ceiling for older children. These issues are more pronounced for tests of face memory because unlike face perception, which develops at the same rate as perception for other objects, face memory appears to develop more slowly than memory for other objects⁵¹. This difference makes it difficult to design matched tests of face and object memory that will help evaluate face-specificity: an object memory task that is matched in difficulty to a face memory task for older children may be poorly matched for younger children and vice versa. However, creating multiple versions of a task, for example an easier version of the face memory task for younger children, leads to other difficulties: 1) determining which task should be administered to an individual who is at an intermediate age, or determining a fixed age range for each test; 2) trying to establish a developmental trajectory for the task (i.e. comparing kids across a wide range of ages); and 3) comparing an individual child's score over time: if the individual was originally evaluated on a test for younger children, which test should be administered when the child has aged outside the cut-off for the original test? A move to a more difficult test could mask improvement, yet re-administering the original test for young children could lead to inflated scores.

We have been confronted with the above difficulties when designing our tests of face and object processing for children. We have designed multiple tests of face memory (CFMT-Kids²⁰, Old/New Faces), matched object memory tasks (CBMT³⁸, Old/New Flowers¹²), and a test of face perception¹² (see Table 1). The CFMT-Kids²⁰ seems to be a useful tool for assessing face memory in children 10 and above, but suffers from floor effects in younger children. We designed a new version of the task for younger children by reducing the number of targets from six to four, but it is still close to floor in younger age groups. To address this difficulty, we designed a CFMT-style test that involves immediate recall only. Although this new test improves scores across the board, it leads to ceiling effects in older children and does not evaluate the ability to maintain multiple identities in memory at one time (Figure 2).

In contrast to the face memory tasks, our other tasks, such as the Dartmouth Face Perception Test (DFPT)¹², have been useful for testing children between the ages of 7 to 12 years. Control means are a reasonable distance from floor and ceiling, and standard deviations are small enough that the task may be useful for a larger age range. As a result, we have been placing particular emphasis on impaired face perception, combined with poor face memory scores and evidence of difficulties with face recognition in daily life, when identifying prosopagnosia in young children⁶.

STATISTICAL GUIDELINES

One popular criterion for neuropsychological impairment is a score that falls more than two standard deviations below the mean for age-matched controls³⁴. In terms of face recognition, scores on the CFMT and CFPT that are greater than two standard deviations below the control mean tend to correspond with clinically significant complaints of face recognition difficulties^{4, 52}. While two standard deviations below the mean is a useful guideline, statistical techniques have been developed to formally test the probability that a single individual's score is taken from the control group⁵³. If the individual's score is statistically unlikely to come from the group (i.e. $p < 0.05$), the individual is considered impaired for the ability that is being measured. Software to perform these calculations using the group size, mean, standard deviation, and individual's test score can be downloaded for free (SINGLIMS)^{54, 55}. To corroborate case-by-case diagnoses of DP, we suggest reporting whether scores fall more than two standard deviations below the mean and whether they meet statistical significance^{12, 56}. Similarly, we suggest using both statistical methods when reporting performance on other tasks (e.g. objects, facial expression, gender).

As discussed above, identifying cases of DP in children can be particularly challenging due to floor effects. Children produce variable results both between subjects, and even trial-to-trial within subjects^{57, 58}. If the control mean on a given task is low and the standard deviation is high, it becomes difficult to detect a score that is more than two standard deviations below the mean (see Figure 2). These factors similarly affect the outcome of statistical tests. It is difficult to advise how to classify children who are on the cusp or who have ambiguous results, but it is recommended that case descriptions provide as much information as possible to justify inclusion or exclusion of a particular case from the DP group. A combination of anecdotal reports of failures of face recognition in daily life, in addition to multiple test scores of face memory and face perception compared to a two standard deviation from the mean cut-off and formal statistical analyses such as the Crawford, Garthwaite, and Howell⁵³ modified statistics is appropriate. These statistical tests can be used to test for a deficit on one test relative to a control group (SINGLIMS)^{54, 55} or for dissociations in performance on two tests (DiffBayes_ES_CP.EXE software)⁵⁹⁻⁶¹. Ideally, when drawing conclusions with regards to a given hypothesis, some cases will provide clear results. However, cases that are more ambiguous should also be included to present a more complete picture of the behaviors being studied.

ASSOCIATIONS AND DISSOCIATIONS

For most research studies, the ideal participants would have pure prosopagnosia (i.e. difficulties with facial identity recognition alone). However, researchers do not appear to routinely report whether

the people in their DP sample show impairments with other aspects of face processing, such as expression or gender recognition. We suggest that this might be due to the time-consuming nature of administering a battery of tests to examine various aspects of face processing (e.g., expressions, eye gaze, gender, sex etc.), coupled with a paucity of brief, widely accepted, reliable tests to diagnose deficits in these processes.

Similarly, there are currently no formally established exclusion criteria for individuals who show impairments distinguishing between non-face objects. Moreover, some have suggested that purely face-specific deficits cannot occur and that all prosopagnosics must have corresponding, though often milder, recognition deficits, such as object agnosia or word agnosia, that can be revealed through the use of carefully designed tasks^{62, 63}. Although comorbid object processing deficits are common^{24, 28}, there are individuals who only have difficulty with faces^{24, 26}. Navigational difficulties may also be associated with DP, with approximately 15% of adults in the faceblind.org database reporting severe navigational difficulties⁶⁴, although it is not yet known whether this rate would be higher than that reported by the general population. These comorbidities may be explained by the proximity between face, place, and object processing areas in the brain⁶⁴. At least for objects, tests are available to enable comparisons between face and object processing abilities (see Table 1) and we recommend that researchers note whether those they classify as DP appear to have face-selective or more widespread deficits.

Face identity recognition deficits also exist in other developmental disorders, such as autism⁶⁵⁻⁶⁸ and Turner syndrome⁶⁹, but face recognition deficits can and do exist in individuals who have normal social functioning⁶⁷. It is unclear at this point whether the face recognition deficits that accompany other developmental disorders should be classified as DP, but given the much broader cognitive effects in disorders like autism spectrum disorders (ASD), it is possible that face recognition deficits outside DP may be qualitatively different (see ⁷⁰ for example). Thus it is advisable to exclude individuals with social developmental disorders from studies of DP, unless they are a particular population of interest.

When working with children with no formal diagnosis of autism, we recommend including measures of autistic tendencies to confirm that the child does not have autism. The Autism Diagnostic Observation Schedule (ADOS)⁷¹ is one component of the gold standard assessment battery for ASD, but trained personnel must administer it, and research reliability is difficult to achieve. It is also time consuming to administer (30 minutes to > 1 hour, plus scoring), and additional assessments are still required for diagnosis. Alternative, more practical, measures for researchers are parent report questionnaires such as the Social Responsiveness Scale – 2 (SRS-2)⁷² and the Repetitive Behavior Scale – Revised⁷³. While these questionnaires are not sufficient for diagnosing ASD, they can be used as

guidelines such that individuals with high scores (indicating autistic tendencies) can be further tested or excluded from the study as a precaution. The Autism Quotient (AQ)^{74, 75} is commonly used among researchers as a quick assessment of autistic tendencies, but parents complain that the items are antiquated (e.g. “S/he is not very good at remembering phone numbers.”) and may therefore be less current than the SRS-2 and the RBS-R. For adults, some previous studies¹⁵ have tended to exclude individuals who scored 32 or above on the AQ, which was a range indicative of a possible ASD⁷⁴. One drawback of using the AQ with DPs is that it includes questions related to face recognition ability and related social interaction and could therefore result in inflated scores (e.g., “When I’m reading a story, I can easily imagine what the characters might look like”, “I find social situations easy”, “I enjoy meeting new people”). Additionally, we note that the AQ is designed to be descriptive rather than diagnostic and that there are updated mean AQ scores for typical and ASD populations⁷⁶. Other questionnaires are also available to measure autistic traits in adults (see ⁷⁷ for a comparison).

In terms of strictly visual comorbidities, we have noticed a recent increase in the number of children requesting assessment for face recognition deficits who already have a diagnosis of Cortical Visual Impairment (CVI). CVI is defined as visual impairment affecting both visual fields despite normal oculomotor systems. It can be related to abnormal development of the visual cortex^{78, 79} due to congenital brain malformation⁷⁸, with the location and extent of the cortical abnormalities determining the functional outcomes⁸⁰. Some individuals with CVI have prosopagnosia⁸⁰ making it an open question whether a diagnosis of CVI, in some cases, would be better classified as DP, or whether some or all individuals with DP may have additional symptoms that would qualify for a diagnosis of CVI.

Another consideration when identifying DP is whether the individual had early visual disturbances such as congenital cataracts, amblyopia or strabismus. There is evidence that normal visual experience with faces from an early age is critical for the development of normal face recognition⁸¹⁻⁸⁵. For example, some individuals who had congenital cataracts have long-lasting face processing deficits, even if the cataracts were removed at an early age (e.g. within the first 8 months of life) and acuity is restored to normal^{81-83, 86}. This is especially true for individuals with bilateral or left eye cataracts⁸⁴, possibly because in infancy information from the left eye projects almost exclusively to the right hemisphere⁸⁷, which is thought to be particularly important for face processing⁸⁸⁻⁹¹. Thus one issue concerns whether an individual with face recognition deficits who had congenital cataracts should be excluded from research on DP.

One might predict that early amblyopia, a condition where the vision in one eye is reduced, could similarly affect face processing and could therefore raise similar flags for participant inclusion.

However, at present there is no empirical evidence that face recognition deficits are more common in individuals with amblyopia than the general public. While some researchers might advocate for excluding these cases, if low-level vision is normal or corrected-to-normal, and the intricacies of the case are described in detail, cases with suspected aetiologies may help elucidate the developmental processes leading to DP. These cases may also represent an interesting sample to compare to DPs with no history of issues with low-level vision.

COGNITIVE HETEROGENEITY

Different phenotypes of DP have been identified in adults: some individuals with DP are only impaired with facial identity recognition^{15, 56, 92-94}, while others may have problems with face detection^{56, 94}, expression recognition²⁶, or gender discrimination²⁶. These dissociations can provide information about which abilities are mediated by distinct neural mechanisms. Addressing heterogeneity, associations, and dissociations in children with DP is particularly important: dissociations between different types of face processing abilities in children with DP can provide clues as to when in development these abilities begin to rely on separate mechanisms. Identifying and understanding the different subtypes of DP in childhood and adulthood will help inform choices for therapeutic interventions.

While individual differences can provide unique information about DP, distinct phenotypes of DP suggest the need for caution when planning group studies (c.f. ⁹⁵). There are no clear guidelines for which individuals can be grouped together, and how one determines inclusion criteria may depend on the primary research question. Thus, we recommend that when presenting group data on DP, individual data be included to provide a more complete picture of the distribution of scores. One difficulty here is that many of the measures suitable for assessing group differences in face processing mechanisms do not have the required psychometric properties to reliably determine whether an individual is impaired or not. As an example, faces are represented as an integrated whole more than most other types of objects, a process termed configural or holistic processing^{96, 97}. Holistic coding can be measured with the composite effect⁹⁶ and part-whole effect⁹⁸, and recent studies have typically shown impairments in groups of DPs^{15, 99, 100}. However, current versions of holistic processing tasks are not appropriate for evaluating holistic processing at an individual level. One way that researchers have attempted to compensate for the low to moderate reliability of each task has been the cognitive neuropsychological approach of using more than one version of a holistic coding task to try to obtain consensus for each individual¹⁰¹. Interestingly, individuals with DP can show typical holistic coding^{101, 102}, indicating that

while measuring holistic coding may be useful for the differentiation of potential subtypes, it is not a defining feature of DP.

With such a complex, heterogeneous disorder, it is likely that a portion of the population will produce borderline or ambiguous results. Especially with children, it is possible for an individual to score in the impaired range for one test of face memory, but then score in the normal range for a different test of the same ability. While it is perhaps simplest to disregard these individuals as uninterpretable, these variable profiles may provide a more complete account of the disorder and its relationship to the hypothesis. It is therefore recommended to include these ambiguous cases in reports, but to interpret them with caution, or to only draw firm conclusions from the clear cases. This again speaks to the drawbacks of group studies, where borderline or inconsistent cases may dilute group effects.

SUMMARY OF GUIDELINES AND FUTURE DIRECTIONS

Table 1 provides examples of tasks that are available for the assessment of DP in children and adults. It would be ideal to have valid, reliable, and brief tests to measure each of the processes involved in face recognition to determine the locus of the impairment (e.g., intact face perception but impaired face memory). In addition, it would be preferable to have more than one test available to measure each of these processes, though at present, only CFMT tasks are available in more than one version. It is also desirable to use a greater mixture of highly controlled and ecologically valid tasks. The tests need to be sensitive to diet of faces experienced by individuals and so will need to vary by race, and perhaps ethnicity¹⁴. Finally, each of the tests needs to be age-appropriate – easy enough for children and sensitive to declines in performance that begin from middle-age. The design and validation of additional tests could be guided by the process of test design for other conditions, such as developmental dyslexia, where a number of appropriate tests are available to distinguish between different subtypes of the disorder¹⁰³. We note that most recently devised tests of face recognition are freely available to researchers via contacting the authors of the test. However, the field may wish to consider establishing a central website for tests. We know of one that hosts numerous tests of language processing (<http://www.motif.org.au>), from which tests can be accessed by those with appropriate credentials and which provides updates to researchers.

Given the available tests, we propose the following certain minimum guidelines for evaluating face recognition in adults. First, the person should report repeated instances of difficulty recognizing faces (not names) in everyday life, either via questionnaire or interview, and not report any brain injury

or disorder that would be expected to result in face recognition impairments (e.g. temporal lobe epilepsy). Second, the person should demonstrate impaired face recognition memory (or perception), ideally on more than one test (e.g., famous faces and CFMT; CFMT and CFMT-Australian; CFPT and CFMT). To differentiate poor recognisers from DPs, we suggest examining z-scores and significance levels on statistical tests such as Crawford, Garthwaite, & Howell's (2009) modified t-tests. While a stringent criteria would advocate that performance should be significantly impaired and below two standard deviations on all tests of face memory, in practice it is typically more instructive to examine performance on the range of tests included and be clear about which cases may be ambiguous. These criteria would enable a diagnosis of DP, but would not be informative as to whether the impairments extended to other objects and if so, whether this is indicative of a general cognitive impairment. For the former, we advocate including a test of object memory; for the latter we advocate assessing cognitive functioning with brief measures. We also recommend asking whether the individual knows of any current or previous visual impairment, and to measure visual acuity, which if impaired could suggest a low-level origin for DP. Finally, a measure of social functioning and/or presence of autistic traits can help to differentiate a diagnosis of DP from a broader developmental disorder of social function.

Similar guidelines should be followed for children, but special considerations must be made. For instance, reports of real world face recognition problems are still important, but will likely come from parents rather than the child. Diagnosis of DP in children can be hindered by floor effects in tests of face memory, particularly in younger children. As such, it is recommended to evaluate face perception in children with suspected DP as a way of providing convergent evidence for face processing deficits. Given the variability in test taking in childhood (both within, and between individuals), ambiguous cases are likely to arise. These cases may be informative, and should be included in scientific reports, but appropriate caution should be used when offering interpretations of the data.

CONCLUSIONS

Individuals with DP have severe face recognition deficits in the absence of brain damage and despite normal low-level vision and intellect. The heterogeneity of the disorder, and the variety of comorbidities that can accompany it, lead to challenges for researchers. We provided guidelines for studying DP based on our experiences. Although face memory impairment is critical for identifying DP in adults, face perception might be the most reliable way to identify DP in kids. Real life difficulties with face recognition also need to be considered. The use of single case modified t-tests can supplement the

traditional approach of using two standard deviations below the mean as a cut-off for impairment. Overall it is essential to be transparent and include as much information as possible for all cases.

Notes

The writing of this article was supported by the Australian Research Council (ARC) Centre of Excellence in Cognition and its Disorders (CCD) [grant number CE110001021] <http://www.ccd.edu.au>. KAD was supported by a Banting Postdoctoral Fellowship awarded by the Canadian Institutes of Health Research. RP was supported by the CCD and ARC Discovery Project grants (DP110100850 and DP1401743). KAD and RP would like to thank the many children and adults whose generous participation in our research projects has helped us to establish these guidelines. Special thanks to Brad Duchaine and Linda Jeffery for their insightful comments on previous drafts.

There are no conflicts of interest.

Table 1. Some available measures that can be used determine whether children and adults have significant face recognition difficulties commensurate with developmental prosopagnosia (DP), and whether they also present with other associated difficulties.

Abilities	Children (ages 6+)	Adults (ages 18+)
Face recognition tests		
Face perception	Dartmouth Face Perception Test (DFPT) ¹²	Cambridge Face Perception Test (CFPT) ⁴⁹ Glasgow Face Matching Test (GFMT) ⁵⁰
Face memory – unfamiliar faces	Cambridge Face Memory Test – Kids (CFMT-Kids) ²⁰ Cambridge Face Memory Test – Children (CFMT-Children) ⁴⁵	Cambridge Face Memory Test (CFMT) ³⁴ Old-New Face Recognition test ⁴⁸
Face memory – familiar faces	Familiar faces provided by families	Familiar faces provided by families Famous face Tests ¹⁷
Subjective reports of everyday face recognition difficulties	Parent and child reports of face recognition difficulties	Anecdotal reports of face recognition difficulties Prosopagnosia Index (PI-20) ²¹
Other tests and measures		
Within-category object memory (used to determine whether face <i>and</i> object memory impaired)	Cambridge Bicycle Memory Test (CBMT) ³⁸ Old-New Flowers ¹²	Cambridge Car Memory Test ³⁷ Vanderbilt Expertise Test ³⁹ Old-New Tests ²⁶
Selective social deficits (used to determine whether social deficits evident)	Social Responsiveness Scale - 2 ⁷² Repetitive Behavior Scale - Revised ⁷³	Autism Quotient ⁷⁴

Visual Acuity (used to determine adequate visual acuity)	Parental report of visual impairments HOTV Distance Pediatric Eye Chart	Self-report of visual impairments Visual acuity test using Sloan font
General functioning	Parent report of any brain injury Birmingham Object Recognition Battery ²³ Wechsler Abbreviated Scale of Intelligence - II ¹⁰⁴	Self-report of any brain injury, disorder, educational level and occupation. Birmingham Object Recognition Battery ²³ Digit Span (backwards) from the Wechsler Adult Intelligence Scale - IV ¹⁰⁵ Raven's Progressive Matrices ¹⁰⁶

Figures

Figure 1. a) Format of Cambridge Face Memory Test (CFMT, Duchaine & Nakayama, 2006a) for adults. The target face is for illustrative purposes and is not used in the actual test. b) Sample trial from Cambridge Bicycle Memory Test (CBMT, Dalrymple & Duchaine, 2013) c) Sample trial from Cambridge Face Memory Test – Kids (CFMT-Kids, Dalrymple, et al, 2012) d) Cambridge Face Perception Test (CFPT, Duchaine, Yovel, & Nakayama, 2007) e) Dartmouth Face Perception Test (for children) (DFPT, Dalrymple, Garrido, & Duchaine, 2014).

Figure 2. Mean scores by age for different versions of Cambridge Face Memory Test – Kids (CFMT-Kids, Dalrymple et al., 2012). Six-target version follows the format of the CFMT for adults, with 72 trials in total. Four-target version follows the same format as the CFMT for adults, but uses 4 targets instead of 6, with 48 trials in total. The Immediate recall version presents the Introductory section of the CFMT-Kids six-target version twice, for a total of 12 targets, with 36 trials in total. Specifically, children learn a target and then pick it out from a choice of three, but are never required to hold multiple targets in memory. Error bars represent standard deviation. Two standard deviations below the mean is one convention for calculating a cut-off for impairment. Dotted line represents chance level performance.

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