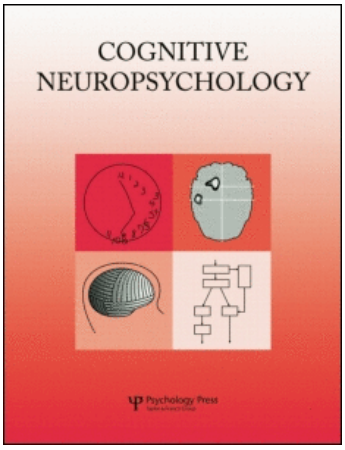

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Normal social cognition in developmental prosopagnosia

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Normal social cognition in developmental prosopagnosia

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Face perception provides information critical to cognitive computations about the social world. This raises the possibility that the development of mechanisms used for social cognition may depend on the presence of normal face perception mechanisms, and this notion partly motivates an aetiological model of autism spectrum disorder (ASD) that suggests that deficits in face perception lead to the social cognition impairments that characterize ASD. To investigate these issues, we examined social cognition in participants with developmental prosopagnosia (DP). A total of 2 male DPs with severe facial identity and facial expression deficits showed no signs of impaired social cognition on three measures. A total of 10 other DPs responded to an inventory measuring autistic traits, and all except one performed normally. These results indicate that social cognition mechanisms can develop normally in the context of developmental face-processing impairments.

Keywords: Social perception; Autism; Neuropsychology; Face perception; Emotion recognition.

Social cognition refers to the computations that underlie our ability to conceptualize and reason about the social world (Frith & Frith, 2007). One of the most important sources of information for these computations is faces, which provide information about identity, emotions, intentions, and other characteristics. Not surprisingly, impairments to face processing make navigation of the social world much more difficult (Yardley, McDermott, Pisarski, Duchaine, & Nakayama, 2008). The importance of facial information for social cognition raises the question of whether the development of social cognition depends on the presence of normally functioning face-processing mechanisms. To address this question, we assessed social cognition in participants who have face-processing deficits due to developmental problems.

Developmental prosopagnosia and social cognition

Developmental prosopagnosia (DP), sometimes referred to as congenital prosopagnosia (Behrmann & Avidan, 2005), is a neurodevelopmental condition defined by severe facial identity recognition deficits despite normal lower level visual processes (Behrmann, Avidan, Marotta, & Kimchi, 2005; Duchaine & Nakayama, 2006b; McConachie, 1976). DPs have difficulty recognizing faces in everyday encounters, particularly when contextual

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information is unavailable. In some cases, DP appears to result from deficits in mechanisms that are specialized for face processing (Duchaine, Yovel, Butterworth, & Nakayama, 2006; Nunn, Postma, & Pearson, 2001). Other cognitive deficits are sometimes seen in DP including problems with face detection (Garrido, Duchaine, & Nakayama, 2008), facial expressions (de Haan & Campbell, 1991; Duchaine, Yovel, Butterworth, & Nakayama, 2006), within-class object recognition (Behrmann et al., 2005; Duchaine, Germine, & Nakayama, 2007a; Duchaine & Nakayama, 2005), and navigation (McConachie, 1976). Several neural abnormalities associated with DP have been identified. Behrmann and colleagues (Behrmann, Avidan, Gao, & Black, 2007) found that the anterior fusiform gyrus was smaller in DPs than in controls and that its size correlated with one measure of face recognition performance, and Garrido et al. (2009) used voxel-based morphometry to show that a group of 17 DPs had less grey matter volume than controls in several temporal regions that respond more strongly to faces than to other stimuli. Studies using functional magnetic resonance imaging (fMRI) have found mixed results, with some DPs showing what appear to be normal fusiform face areas (FFA; Behrmann et al., 2005; Hasson, Avidan, Deouell, Bentin, & Malach, 2003) whereas others do not (Bentin, DeGutis, D'Esposito, & Robertson, 2007; Duchaine et al., 2006; Minnebusch, Suchan, Köster, & Daum, 2009; Van den Stock, van de Riet, Righart, & de Gelder, 2008) or do not show adaptation to repeated presentation of unfamiliar faces in FFA (Williams, Berberovic, & Mattingley, 2007). Genetic factors play a role in some cases of DP (Duchaine et al., 2007a; Grueter et al., 2007; Schmalzl, Palermo, & Coltheart, 2008), but little is known about the developmental trajectory of DP. Note that although DP is often referred to as congenital prosopagnosia, no evidence has been collected that demonstrates that face-processing impairments are present at birth or very early in life in DP so face processing may function normally for a period of time. However it seems likely that face-processing problems are present at birth and predate any experience with faces in a substantial proportion of DP cases.

Although some individuals with DP also have autism spectrum disorder (ASD) (Duchaine, Nieminen-von Wendt, New, & Kulomaki, 2003; Kracke, 1994), most DPs seem to have normal social cognition. Many are employed in jobs that appear to require good social cognitive abilities (Behrmann et al., 2005; Duchaine et al., 2007a; Garrido et al., 2008), and their behaviour during laboratory visits suggests normal social abilities. However, no formal data on social cognition in DP have been published. Such data would be relevant to three issues. First, if social cognition is normal in DP, it would demonstrate that social cognition mechanisms can develop normally even when face-processing mechanisms have not done so. Secondly, measurement of social cognition in DP would examine implications of an aetiological theory of ASD that we elaborate on in the next section (Schultz, 2005). Finally because face-processing problems are common in ASD, the definition for DP mentioned above does not clearly differentiate it from ASD. Both are developmental disorders with face-processing deficits, and it has been suggested that ASD and prosopagnosia are closely linked (Schultz, 2005). To examine whether DP and ASD are separate conditions, it is necessary to demonstrate that individuals with DP do not meet some of the criteria for ASD, and this could be done by showing that DPs have normal social cognition.

Face processing and the aetiology of autism spectrum disorder

Many studies have shown that individuals with ASD exhibit cognitive and neural face-processing abnormalities (for reviews see Behrmann, Thomas, & Humphreys, 2006; Dawson, Webb, & McPartland, 2005; Schultz, 2005; but see Jemel, Mottron, & Dawson, 2006), and several developmental possibilities have been suggested to explain face-processing deficits in ASD (Baron-Cohen et al., 2000; Dawson et al., 2005; Gauthier & Nelson, 2001). One model connects a number of findings to propose a causal link between impaired face processing and impaired social cognition (Schultz, 2005). Specifically, this

hypothesis suggests that an important factor contributing to the development of ASD involves a cascade of neurobiological events—namely, (a) abnormal functioning of the amygdala, a structure that plays a key role in alerting other brain systems to the emotional salience of perceptual events; (b) reduced attention to emotional and socially relevant stimuli, including faces; (c) reduced facial input to cortical areas involved in face perception, in particular fusiform gyrus; (d) reduced development of perceptual skills allowing advanced computations necessary for facial identity and facial expression recognition; and (e) impaired development of social cognition.

This model draws support from the common co-occurrence of face-processing impairments in ASD and from neuroimaging studies that show hypoactivation of the amygdala and the FFA in individuals with ASD (Schultz et al., 2003). In addition, a relationship between face processing and social cognition was suggested by Schultz et al.'s (2003) demonstration that the FFA is strongly activated by animations depicting faceless geometric shapes moving in ways that lead observers to make mental-state attributions about the shapes. Together these findings have raised the possibility that dysfunction in the fusiform gyrus could cause both the face-processing problems and the social cognition problems in ASD (Schultz, 2005). However, ASD and face recognition impairments are not always observed in conjunction, and a causal relationship is challenged by reports of individuals with ASD who show normal face perception (Barton et al., 2004; Hefter, Manoach, & Barton, 2005). Because the model proposes that developmental deficits of face processing contribute to the social cognition impairments characteristic of ASD, evidence showing that DP is accompanied by social cognition deficits would provide support for the model. In contrast if social cognition in DPs is normal, the dissociation between face processing and social cognition will indicate that the development of social cognition does not require face processing to develop normally. This inference though assumes that face processing is impaired in DP from birth, but if future work shows that many or all cases of DP have normal face processing at birth, and deficits only emerge later, then the interpretation of normal social cognition in DP will need to be reconsidered.

We assessed social cognition in DP in two ways. We carried out in-depth assessments of two DPs, which consisted of an interview asking about their social lives and three measures of social cognition and social interest: the Autism Diagnostic Observation Schedule (ADOS; Lord, Rutter, DiLavore, & Risi, 2002), the Autism-Spectrum Quotient Questionnaire (AQ; Baron-Cohen, Wheelwright, Skinner, Martin, & Clubley, 2001b), and the Animations task (Abell, Happé, & Frith, 2000). These tasks were chosen because, given the putative link between the development of face processing and the development of social cognition, we wanted to use tasks that have revealed deficits in ASD participants. The ADOS is commonly used to assess ASD (Bertrand et al., 2001; Gotham, Risi, Pickles, & Lord, 2007), the animations task has revealed impairments in people with ASD (Abell et al., 2000; Castelli, Frith, Happé, & Frith, 2002), and the AQ is sensitive to ASD traits (Baron-Cohen, Wheelwright, Hill, Raste, & Plumb, 2001a; Baron-Cohen et al., 2001b; Hoekstra, Bartels, Cath, & Boomsma, 2008; Woodbury-Smith, Robinson, Wheelwright, & Baron-Cohen 2005). Secondly, to examine whether our results from the in-depth assessments are typical of DP, 10 DPs were assessed with the AQ.

EXPERIMENT 1: IN-DEPTH ASSESSMENT OF SOCIAL COGNITION IN TWO DPs

Method and results

Participants

D.H. is a right-handed male architect and designer who was 31 years old at the time of testing. Like all prosopagnosic participants in this study, D.H. contacted the Prosopagnosia Research Center at Harvard University/University College London (UCL) because he

experienced significant problems with face recognition in everyday life. His full-scale IQ, measured with the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999), was 126 (verbal IQ, VIQ, 117; performance IQ, PIQ, 129). His visual acuity and low-level vision were normal. D.H. believes that his grandmother shares his face recognition difficulties, but she has not been tested. D.H. did suffer from birth complications, but a radiologist could not find any evidence in a structural magnetic resonance imaging (MRI) scan of anoxia or other effects from that incident. If birth complications caused D.H.'s prosopagnosia, he would not be considered a DP, but he would be ideal for the purposes of this study in that his face processing would have been impaired from birth. D.H. recalls difficulty recognizing faces as a child and has many stories about failures to recognize people he knew well. He tends to rely on the voice for emotional information and makes use of alternative strategies to recognize individuals. For example, D.H. stated: "In the course of an office meeting with new people, I will make a diagram of where people are sitting with notes on what shirt they are wearing."

T.U. is a right-handed male graphic designer tested when he was 32 years old. His full-scale IQ score was 128 (VIQ, 119; PIQ, 132). His visual acuity and low-level vision were normal. T.U. has not experienced any brain damage and reports lifelong difficulties with face recognition. He often uses voices, gait, clothing, and hair to recognize people. T.U. sometimes makes sketches of people in meetings to help him recall the participants. These sketches depict individuals with distinctive hairstyles and clothing but without facial features.

Control data for several tests discussed below were obtained from published papers, but controls for the famous faces task and the facial expression task were 18 Londoners, 7 men and 11 women, with an average of age of 28.9 years ($SD = 5.7$). Their average full-scale IQ was 118.9 ($SD = 8.8$; VIQ, 112.8; PIQ, 122.0) so they are well matched to D.H. and T.U.

We used Crawford and Howell's (1998) modified t test to calculate significance for the difference between each DP score and the control scores. This test was designed to compare data from single cases to small control groups.

Facial identity tests

Famous Faces. A total of 60 celebrity faces were presented for 3 s each (Duchaine, Yovel, & Nakayama, 2007b). Participants were asked to name them or provide uniquely identifying information about them. Table 1 shows that D.H. and T.U. both performed very poorly on this task relative to controls despite being familiar with the celebrities, and their scores were significantly different from those of controls: D.H., $t(17) = -3.08$, $p = .003$; T.U., $t(17) = -2.17$, $p = .022$.

Cambridge Face Memory Test (CFMT). The CFMT requires recognition of a set of six target faces from novel views in which the pose and

Table 1. *D.H.'s and T.U.'s scores on face tests relative to controls*

	n	*Control mean*	*Control SD*	*D.H.*	*T.U.*
Famous Faces Identified (total = 60)	18	46.9	8.5	20**	28*
Famous Faces Exposed (total = 60)	18	54.1	5.9	60	60
Cambridge Face Memory Test (total = 72)	20	59.6	7.6	28**	42*
Face Old-New Discrimination - A'	21	0.96	0.02	0.79**	0.83**
Cambridge Face Perception Test Upright (Errors)	21	36.7	12.2	76**	64*
Cambridge Face Perception Test Inverted (Errors)	21	65	9.8	78	44
Films Facial Expression Task (%)	18	89.5	5.7	75.9*	70.7**

Note. Asterisks indicate significant effects computed using Crawford and Howell's modified t test (1998).
*$p < .05$. **$p < .01$.

lighting are different from those of the study images (for details, see Duchaine & Nakayama, 2006a). The CFMT has 72 items (see Figure 1a). Each item has three choices so chance performance is 24. Table 1 shows that both D.H. and T.U. had difficulty with this test relative to controls (Duchaine et al., 2007b). D.H.'s score was more than four standard deviations below the mean whereas T.U.'s score was more than two, and both scores were significantly different from the control scores: D.H., $t(19) = -4.05$, $p < .001$; T.U., $t(19) = -2.25$, $p = .018$.

Face Old–New Test. In the study phase, participants are shown 10 target faces for 3 s each, and this cycle is repeated (Duchaine & Nakayama, 2005). Participants are then asked to decide whether 50 faces shown one at a time are targets or nontargets (20 targets, i.e., 10 targets repeated twice; 30 unique nontargets). We computed A', an unbiased measure of discrimination (Macmillan & Creelman, 1991), from the participants' hits and false alarms. Table 1 shows that D.H. and T.U. both did very poorly with this test relative to controls—D.H., $t(20) = -8.31$, $p < .001$; T.U., $t(20) = -6.35$, $p < .001$—with scores 7 and 5 standard deviations below the control mean, respectively.

Cambridge Face Perception Test. This test was designed to measure facial identity perception so memory demands are minimal (Duchaine et al., 2007b). For each item, a three-quarter profile target face was presented above six test faces in frontal view. The six faces were created by morphing the target face with one of six faces, so each test face came from a different morph continuum. Each test face contained a different proportion of the target face in it, ranging from 88% to 28% in jumps of 12% (see Figure 1b). There were eight items, and each was presented upright once and inverted once. Participants had one minute to sort the test faces in each item based on their similarity to the target face. Errors were computed by summing the deviations from the proper position of each face in an item and computing the total number of upright errors and total inverted errors. Table 1 shows that both D.H. and T.U. had scores more than two standard deviations above the mean for the upright faces: D.H., $t(20) = 3.15$, $p = .003$; T.U., $t(20) = 2.19$, $p = .02$. Neither of their inverted scores was significantly worse than those of controls, indicating that low-level vision problems were not causing their poor upright scores. Interestingly, T.U. made far fewer errors with inverted faces than with upright faces. He repeated the test at a later date and showed the same pattern though with a smaller difference (5 more upright errors rather than 20).

Facial expression

The previous tests demonstrate that D.H. and T.U. have severe facial identity perception and memory deficits. Next we examine whether they also have deficits with facial expression. DPs show mixed performance with facial expression, with some showing normal performance (Bentin et al., 2007; Duchaine, Parker, & Nakayama, 2003; Humphreys, Avidan, & Behrmann, 2007) whereas others have severe problems (de Haan & Campbell, 1991; Duchaine et al., 2006).

Films Facial Expression Task. Images of facial expressions were taken from movies, and they are more realistic and subtle than expressions used in most experiments. On each trial, an emotion state word was presented, and then participants were presented with three images of the same actor or actress making different facial expressions (see Figure 1c). Each image was presented for 500 ms. Participants chose which of the three expressions depicted the emotion state word.

Control averaged 89.5% correct ($SD = 5.7$). D.H. scored 75.9% correct, and T.U. scored 70.7%. D.H.'s score was more than two standard deviations below the control mean, and T.U.'s was more than three: D.H., $t(17) = -2.30$, $p = .017$; T.U., $t(17) = -3.19$, $p = .003$.

Social cognition

The face tests demonstrate that D.H. and T.U. both have deficits with facial identity and facial expressions. Next we assess their social cognition

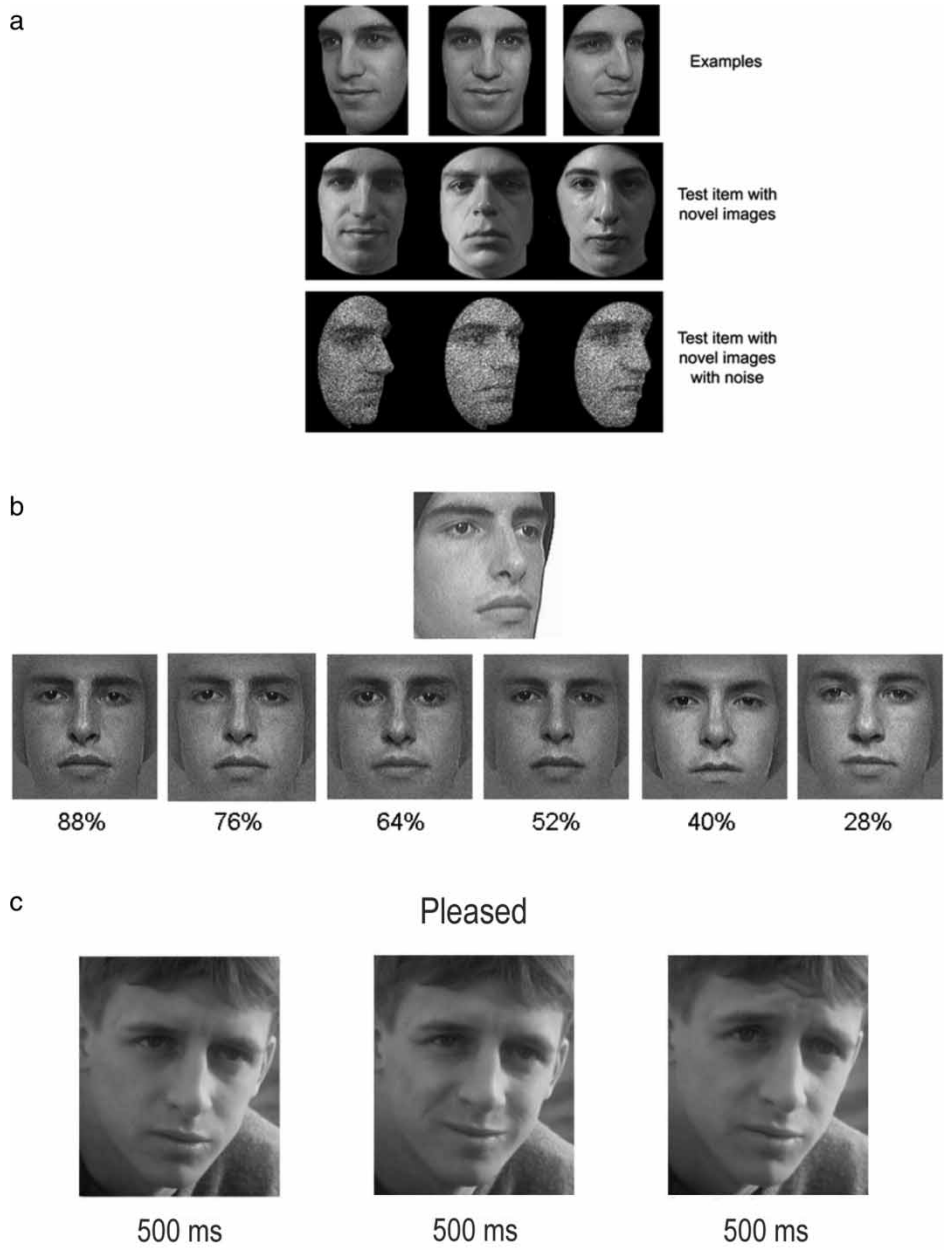

Figure 1. *Examples from the Cambridge Face Memory Test (CFMT), the Cambridge Face Perception Test (CFPT), and the Films Facial Expression Test (FFET). (a) This panel displays three study items from the CFMT, a test item showing the target face from the examples with different lighting, and a test item with a novel pose and added noise. (b) Panel b displays an example item from the CFPT. Each face is a morph created by morphing a frontal view of the target face with another face. The target face was morphed with six different faces to create the test faces. The percentages show the proportion of the morph that comes from the target face. The six faces are presented in a random order, and the participant has one minute to sort them according to their similarity with the target face. (c) Participants in the FFET are presented with an emotion word and must choose which one of three briefly presented faces best depicts that emotion. FFET images, unlike those shown here, are in colour.*

to see whether face-processing deficits due to developmental problems are associated with social cognition deficits.

Interview

A semistructured interview was carried out to learn more about D.H.'s and T.U.'s social lives to examine whether these suggest that they have problems with social cognition (see Appendix for questions).

D.H. was relaxed and kept the conversation flowing effortlessly. He is married, and although he enjoys socializing, he prefers small gatherings where it is easier to keep track of identity. D.H. tends to be shy in larger groups but more outgoing when he is with close friends. On his walk from the Underground station to work he reports that he says hello to more people than he probably should in case he knows the person walking next to him from the office but doesn't recognize him or her.

The interview with T.U. was very comfortable. He was soft spoken at times, but the conversation was smooth and easy. During the interview, T.U. asked reciprocal questions, made jokes, and maintained eye contact throughout the conversation. T.U. is not married but has had a number of girlfriends over the years. T.U. reports that he enjoys social situations but finds them exhausting. He prefers to stay at home on weekends but will go with his girlfriend to parties, galleries, and other social events. T.U. is able and willing to be part of larger social situations, but prefers interacting with small groups of friends.

Autism Diagnostic Observation Schedule

The ADOS is a standardized observation designed to assess behaviours related to ASD (Lord et al., 2002). D.H. and T.U. were assessed with Module 4 because it was designed for fluent adults. The assessment gives summary scores in three categories: communication, reciprocal social interaction, and stereotyped behaviours and restricted interests.

The results from the ADOS showed that T.U. and D.H. do not have any social or communication problems characteristic of ASD. T.U. received 0 points under the "communication" and "stereotyped behaviours" categories, and 1 point under the "reciprocal social interaction" category because he slightly interrupted the examiner on a couple of occasions. D.H. had 0 points on all categories. Both scores are far from the autism spectrum cut-off of 7 points and even farther from the autism cut-off of 10 points.

Autism-Spectrum Quotient Questionnaire

The AQ is a self-administered questionnaire with 50 items that measures the degree to which an adult with normal intelligence has traits associated with the ASD (Baron-Cohen et al., 2001b). For each item, participants choose whether they "definitely agree", "slightly agree", "slightly disagree", or "definitely disagree" with the statement. The following are two example statements: "I prefer to do things the same way over and over again", and "Other people frequently tell me that what I've said is impolite, even though I think it is polite." The ASD-typical answer for each item in the questionnaire was either both "agree" choices or both "disagree" choices. ASD-typical responses received a point.

Baron-Cohen et al. (2001b) analysed questionnaires from a group of 58 adults with Asperger syndrome or high-functioning autism (HFA; all of whom had been diagnosed by psychiatrists using established ASD diagnostic criteria) and a group of 174 adult controls selected at random from East Anglia. This analysis suggested a total score of 32 or more on the questionnaire is an effective cut-off point. In their sample, approximately 79% of AS/HFA participants scored at or above this level while only 2.3% of controls scored at or above this level.

D.H. scored 14, and T.U. scored 15, which are slightly lower than the control mean of 16.4 ($SD = 6.3$). The AQ indicates that D.H. and T.U. have normal social cognition.

Animations task

The final task used to investigate D.H.'s and T.U.'s social cognition is the animations task, which assesses theory of mind (Abell et al., 2000; Castelli et al., 2002; Castelli, Happé, Frith, &

Frith, 2000). It was inspired by Heider and Simmel's cartoon (1944) in which a circle, a small triangle, and a larger triangle "interact". Observers usually describe the shapes' movements with mental-state terms. To systematically investigate this phenomenon, 12 clips were created, which were each approximately 40 s long (Abell et al., 2000). Each clip involved a large red triangle and a smaller blue triangle, and half of the sequences utilized an enclosure that the triangles moved in and out of. The sets consisted of four theory of mind (ToM) clips, four goal-directed clips, and four random sequences. The ToM animations involved the triangles moving in a way that elicited mental state attributions, such as one triangle seducing or coaxing the other. The goal-directed animations consisted of the triangles moving in a physical way, such as dancing or fighting. In the random sequences, the triangles moved in a completely random manner around the screen. After each clip, the participants were asked to describe what they saw the triangles doing. Castelli et al. (2002) found no difference between the descriptions provided by the autistic group and controls for the goal-directed or the random sequences. There was, however, a significant difference between the autistic and control groups for the ToM animations, with ASD participants giving shorter descriptions that referred to mental states less frequently. Similar results were found when high-functioning children with autism were tested with the animations task (Abell et al., 2000).

Participants were told that they would see shapes moving around and were informed that the different animations would have different content. After each animation, participants were asked what the triangles were doing, whether they were randomly moving about, or whether they were doing something more specific. The scoring of each description was based on three criteria: intentionality, appropriateness, and length. The intentionality score was a scale from 0 to 5 points and reflected the use of mental-state terms. The appropriateness score used a scale from 0 to 3 and evaluated how well the participant understood the animation, as intended by the designer. The length score ranged from 0 to 4 and was based on the number of clauses used to describe the animation.

Descriptions of the animations provided by D.H. and T.U. were compared to those of 12 controls (average age = 29.6, $SD = 9.9$). The scores of these controls were similar to previous control scores (Castelli et al., 2002). Table 2 shows that D.H.'s and T.U.'s scores were very similar to the scores of the current controls. Like these controls, their intentionality scores were highest on the ToM animations and lowest on the random sequences. D.H. and T.U.'s scores for intentionality and appropriateness on the theory-of-mind clips were substantially higher than the mean scores of the ASD participants in Castelli et al.

Table 2. *Scores on the Animations task*

	Control mean	*Control SD*	*D.H.*	*T.U.*
Intentionality (0–5)				
Theory-of-mind clips	3.9	0.6	3.8	4.0
Goal-directed clips	2.3	0.4	2.2	2.3
Random sequence clips	0.5	0.4	0.0	0.0
Appropriateness (0–3)				
Theory-of-mind clips	2.2	0.5	2.0	2.0
Goal-directed clips	2.5	0.3	2.8	2.8
Random sequence clips	2.6	0.4	3.0	2.8
Length (0–4)				
Theory-of-mind clips	2.8	0.5	2.5	2.0
Goal-directed clips	1.8	0.7	1.5	1.6
Random sequence clips	1.6	0.5	1.3	1.4

(2002). Altogether, their scores indicate they conceptualized the animations in the same way the controls did.

EXPERIMENT 2: TESTING OTHER DPs WITH THE AUTISM-SPECTRUM QUOTIENT QUESTIONNAIRE

Method and results

The results from D.H. and T.U. indicate that their social cognition is normal, but considerable heterogeneity exists in developmental neuropsychological conditions, and cognitive and neural studies demonstrate that this is true for DP (Duchaine & Nakayama, 2005; Garrido et al., 2008; Harris, Duchaine, & Nakayama, 2005; Schmalzl et al., 2008). To investigate whether social cognition is normal in a larger sample of DPs, we examined how 10 other DPs did on the AQ.

Participants

These participants, 6 women and 4 men, contacted us because they regularly have difficulty recognizing faces in daily life. Their average age was 38.8 years ($SD = 10.3$). They were tested with the face tests discussed below in our laboratory, but they responded to the AQ using an online interface.

Facial identity tests

To establish that these participants were prosopagnosic, we compared their scores on the same three tests of face memory discussed above (Famous Faces, the CFMT, and the Face Old–New Discrimination) to those of the controls used above. The z scores for the 10 DPs are shown in Figure 2, with asterisks indicating significance levels computed using Crawford and Howell's (1998) method. Each DP had at least two scores significantly different from the control scores, and all scores were below average.

Films Facial Expression Test (FFET)

We have scores for 8 of the 10 DPs on the FFET (no scores for F30 and F49). All scores were below the control mean, but only 2 DPs had scores that were significantly different from those of controls: M54, $t(17) = -2.01$, $p = .03$; F45, $t(17) = -2.90$, $p = .005$.

AQ results

Figure 3 displays the AQ scores for the DPs. All but 1 of the DPs scored in the normal range. The average for the 10 DP participants was 19.7 ($SD = 8.0$), which is only slightly higher than the control mean of 16.4 ($SD = 6.3$; Baron-Cohen et al., 2001b). This difference was not significant, $t(182) = 1.3$, *ns*. F45's score of 37 was greater than the Asperger syndrome/high-functioning autism sample's average of 35.8 ($SD = 6.5$), indicating that F45 has autistic traits.

Discussion

To investigate social cognition in a group of DPs, we carried out an in-depth assessment of social cognition in two DPs and examined ASD traits using a questionnaire with 10 other DPs. The 2 DPs assessed had severe facial identity and facial expression deficits. Their social cognition was assessed with the ADOS, the AQ, the animations task, and an interview inquiring about their social lives. D.H. and T.U. showed no sign of social cognition impairments. To determine whether their results are typical of DP, the 10 DPs filled out the AQ. A total of 9 of the 10 DPs had scores indicating that they were not on the autistic spectrum, so normal social cognition appears to be common in DP.

Input from face processing is critical for social cognitive computations but our results suggest that social cognition is normal in DP and that the mechanisms carrying out these computations can develop normally even when face processing does not develop properly. This point was demonstrated especially clearly by D.H.'s and T.U.'s results. Both men have severe facial identity and expression recognition deficits that impact their daily lives but their assessment showed that they had no problems with social cognition. Their results contrast with those of people who have experienced a much greater absence of facial input—the congenitally blind. Although congenital blindness is

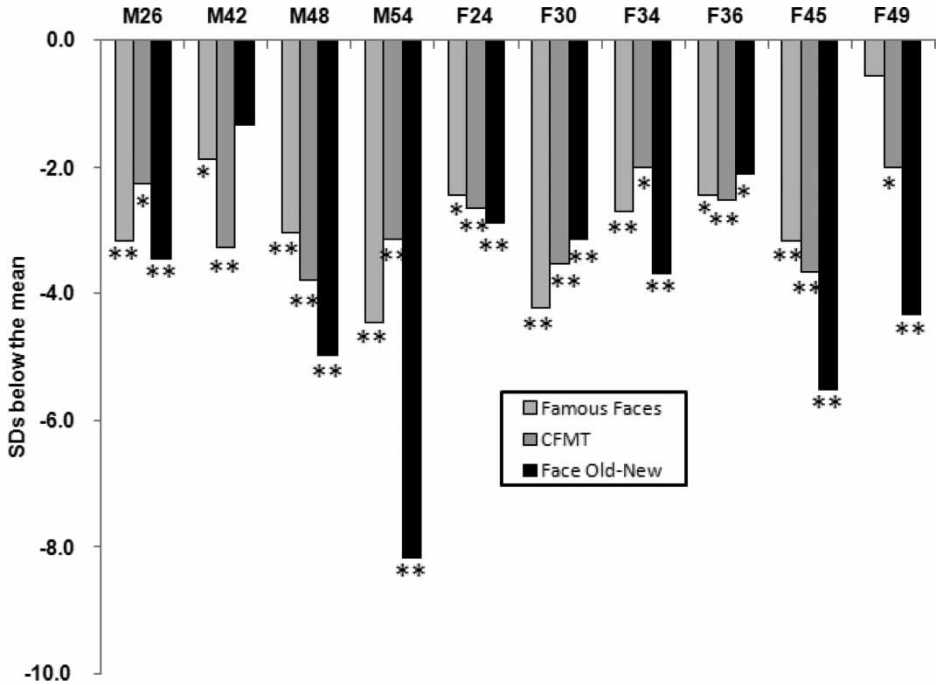

Figure 2. *z scores for the 10 participants with developmental prosopagnosia (DPs) on three face memory tests: Famous Faces, the Cambridge Face Memory Test, and the Face Old–New Discrimination. Each score was tested for significance using Crawford and Howell's (1998) modified* t *test.* * $p < .05$; ** $p < .01$.

certainly not sufficient to cause ASD, Hobson and Bishop (2003) reported that they do show increased autistic behaviours and some social impairment. The cause of the social difficulties in the blind are unknown, but Hobson and Bishop (2003) point out that vision plays a key role in linking children with others and so may contribute to the development of social cognitive processes. Presumably facial input would be a critical visual feature in such a scenario, and the results from the DPs suggest that some facial input, even if it is not normal, is sufficient for typical social cognitive development.

Our conclusion that social cognition can develop normally even in the absence of normal facial input assumes that face processing is dysfunctional in DPs from an early age. It seems likely that face-processing deficits in some or most DPs are present at birth, which is why DP is referred to as "congenital prosopagnosia" by many laboratories (Behrmann & Avidan, 2005; Schmalzl et al., 2008; Williams et al., 2007). However, at present there is no evidence to support this view (note, though, that there is also no evidence demonstrating very early dysfunctional face processing in ASD). A better understanding of the developmental course of face processing in DP should shed light on if and when facial input is necessary for the development of social cognition, and the current results will need to be reconsidered if face processing in DP is usually normal in the early years of life.

Although our results indicate that social cognition is normal in the DPs, social cognition is an umbrella term for what may be a number of mechanisms. The tasks used were chosen because they are sensitive to the social cognition deficits seen in ASD (Abell et al., 2000; Baron-Cohen et al., 2001b; Castelli et al., 2002; Castelli et al., 2000; Lord et al., 2002). Presumably these tasks assess some mechanisms important for social cognition but fail to measure others (Frith & Frith, 2007).

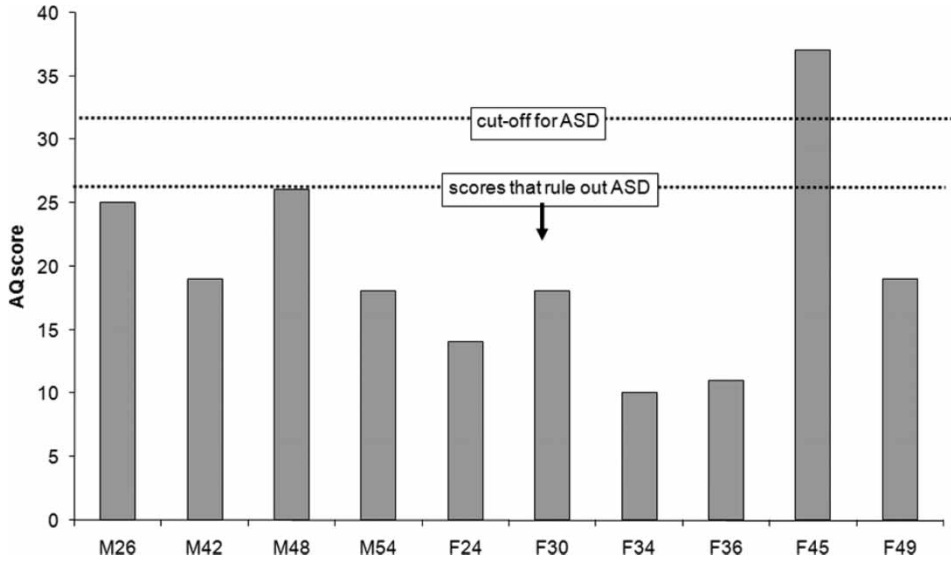

Figure 3. *Scores for the participants with developmental prosopagnosia (DPs) on the Autism-Spectrum Quotient Questionnaire (AQ). Of the 10 DPs, all but F45 scored in the normal range. Her score suggests that she has autistic traits.*

Given these considerations, it will be worthwhile to carry out further testing of DPs with tests that measure different aspects of social cognition. This testing might include advanced theory of mind tasks such as the faux pas task (Stone, Baron-Cohen, & Knight, 1998) and nonface emotion recognition tasks such as the Movie Stills task without faces (Adolphs & Tranel, 2003). Formal measures of DPs' ability to empathize with others' emotions would also be valuable (Dziobek et al., 2008; Spreng, McKinnon, Mar, & Levine, 2009). It would be expected that social cognition mechanisms that rely heavily on input from faces would be most likely to be impaired in DP. Note, however, that tasks assessing such mechanisms should not require face processing since failure on such tasks by DPs could result from face-processing problems rather than from deficits in social cognition mechanisms.

The demonstration that the development of social cognitive mechanisms is not dependent on normal face processing is also relevant to the aetiological theory of autism discussed above (Schultz, 2005). Several groups have demonstrated hypoactivation in the FFA when ASD participants view faces (Critchley et al., 2000; Dalton et al., 2005; Pierce & Courchesne, 2000; Schultz et al., 2000), and Schultz et al. (2003) found that normal participants showed activation of the FFA when shown animations similar to those in the animations task. This last result suggests that fusiform regions involved in face processing may also be critical for social cognition, which raised the possibility that the face-processing deficits and social cognition deficits in ASD result from the same problems in the fusiform gyrus. Our results, however, demonstrate that developmental face-processing problems do not invariably lead to social cognition deficits and so raise questions about this model of ASD. It is possible, though, that the events that lead to impaired facial processing in ASD are different from those that lead to DP. Moreover, the nature of the face-processing impairments in ASD and DP may be different. Future research could test both groups with similar tasks and characterize the impaired mechanisms that lead to difficulties in processing faces in each group. It may be that only certain face-processing computations or areas, which are impaired in ASD but not in most DPs, influence the development of social cognition.

Schultz (2005) acknowledged that the causal connection between face processing and social

cognition in ASD could point in the opposite direction. Preexisting social cognition deficits might lead to a lack of interest in faces, which could negatively impact face-processing development. Given that individuals with ASD have social cognition deficits by definition, this possibility is challenging to test in ASD. Our results, however, do suggest that preexisting problems with social cognition, at least for those aspects of social cognition tested here, do not lead to the face-processing deficits seen in DP. The frequent co-occurrence of object recognition problems in DP (Behrmann et al., 2005; Duchaine et al., 2007a; Duchaine & Nakayama, 2005) provides additional evidence that DP is not caused by social cognition deficits, because deficient attention to social interactions seems unlikely to lead to problems with object recognition.

A final motivation for this investigation was to demonstrate that DP and ASD are separate conditions. Schultz (2005) has pointed out that they have a number of parallels, but research into DP has implicitly assumed that DP is qualitatively different from ASD. The results demonstrate that DP is commonly accompanied by normal social cognition and so show that DP is, in fact, a separate condition from ASD. D.H. and T.U.'s failure to show perseverative behaviours also differentiates DP from ASD. Additional evidence that DP and ASD are separate conditions is provided by the opposite dissociation—ASD with normal face processing (Barton et al., 2004; Hefter et al., 2005). The score of F45 on the AQ, however, suggests that some individuals classified as DP may be on the autism spectrum, and this possibility should be explicitly assessed in DPs involved in research.

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APPENDIX

Questions asked during the interviews with D.H. and T.U.:

- Could you tell me a little bit about yourself?
- Do you consider yourself more of a shy person or an outgoing person?
- Do you feel that you have become more or less outgoing than when you were a child?
- When you were younger did you have a large group of friends or did you prefer one or two friends that you spent all of your time with?
- How old were you when you had your first boyfriend or girlfriend?
- And have you been in any serious relationships since then?
- Do you enjoy meeting new people? Do you find it difficult to meet new people?
- What do you like to do on the weekends?
- Do you enjoy being in a more social environment like a party, or would you rather stay at home and watch a movie or read a book?
- How has prosopagnosia affected the way you interact with others?