



# Are Social Cognition Deficits Present in Developmental Prosopagnosics?

Heidi Murray, BSc  
*2007 MSc Clinical Neuroscience Thesis*  
*Institute of Neurology, UCL*  
*Queen Square, London*



Supervisor: Dr. Brad Duchaine  
*Institute of Cognitive Neuroscience, UCL*  
*Queen Square, London*

*Word Count: 10,074*

Submitted as partial fulfillment of the requirements for the MSc in Clinical Neuroscience.  
University of London, 2007

**MSc Clinical Neuroscience**  
**2006/07**

UMI Number: U593940

All rights reserved

INFORMATION TO ALL USERS

The quality of this reproduction is dependent upon the quality of the copy submitted.

In the unlikely event that the author did not send a complete manuscript and there are missing pages, these will be noted. Also, if material had to be removed, a note will indicate the deletion.



UMI U593940

Published by ProQuest LLC 2013. Copyright in the Dissertation held by the Author.  
Microform Edition © ProQuest LLC.

All rights reserved. This work is protected against  
unauthorized copying under Title 17, United States Code.




ProQuest LLC  
789 East Eisenhower Parkway  
P.O. Box 1346  
Ann Arbor, MI 48106-1346

INST NEUROLOGY



19 0418640 7

INSTITUTE OF NEUROLOGY  
  
*The*  
ROCKEFELLER  
MEDICAL LIBRARY

ROCKEFELLER INSTITUTE LIBRARY  
UCL INSTITUTE OF NEUROLOGY  
THE NATIONAL HOSPITAL  
QUEEN SQUARE  
LONDON  
WC1N 3BG

## ACKNOWLEDGEMENTS

I would like to thank a number of people for their help throughout this project.

Thank you to Sarah White for advice on social cognition tests, for some of the control data used, and for her general guidance.

Thank you to Martha Turner for administering the ADOS and for taking time to explain the test to me.

Thank you to Sarah Blakemore, who was kind enough to let me borrow the WASI for testing.

Thank you to Laura Germine for putting the AQ questionnaire and Strange Stories test online for me, for answering technical questions regarding the website, and for taking the time to email the data from the tests to me.

Thank you to Lucia Garrido was helpful with my many questions, especially my questions involving statistics.

Thank you to Jamie Smith for his patience and support during my entire master's degree process.

Thank you to the IoN education unit for their assistance throughout the year.

And last but not least, thank you to Brad Duchaine for his supervisory guidance and for giving me the opportunity to work on this project.

<b>ABBREVIATIONS</b> .....	4
<b>1.0 ABSTRACT</b> .....	5
<b>2.0 INTRODUCTION</b> .....	6
2.1 <i>What are Autism Spectrum Disorders?</i> .....	6
2.2 <i>The association between facial processing and ASD</i> .....	7
2.3 <i>Models for the etiology of autism</i> .....	11
2.4 <i>Developmental prosopagnosia as a way to investigate the link between facial processing and social cognition</i> .....	14
2.5 <i>Social Cognition and Prosopagnosia</i> .....	18
2.6 <i>Tests of Social Cognition</i> .....	18
<b>3.0 METHODS 1</b> .....	22
3.1 <i>Subjects</i> .....	22
3.2 <i>Tests</i> .....	25
3.2.1 <i>WESCHLER ABBREVIATED SCALE OF INTELLIGENCE (WASI)</i> .....	25
3.2.2 <i>AUTISM DIAGNOSTIC OBSERVATION SCHEDULE (ADOS), MODULE 4</i> .....	26
3.2.3 <i>AUTISM-SPECTRUM QUOTIENT QUESTIONNAIRE (AQ)</i> .....	26
3.2.4 <i>STRANGE STORIES TEST</i> .....	27
3.2.5 <i>ANIMATIONS TEST</i> .....	28
<b>4.0 RESULTS 1</b> .....	30
4.1 <i>ADOS Results</i> .....	30
4.2 <i>AQ Results</i> .....	30
4.3 <i>Strange Stories Results</i> .....	33
4.4 <i>Animations Results</i> .....	36
<b>5.0 METHODS 2</b> .....	40
5.1 <i>Subjects</i> .....	40
5.2 <i>Tests</i> .....	41
5.2.1 <i>AUTISM-SPECTRUM QUOTIENT QUESTIONNAIRE (AQ)</i> .....	41
5.2.2 <i>STRANGE STORIES TEST</i> .....	41
<b>6.0 RESULTS 2</b> .....	42
6.1 <i>AQ Results</i> .....	42
6.2 <i>Strange Stories Results</i> .....	43
<b>7.0 DISCUSSION</b> .....	47
<b>8.0 CONCLUSIONS</b> .....	52
<b>9.0 REFERENCES</b> .....	53
<b>APPENDIX 1 – Autism-Quotient Questionnaire</b> .....	58
<b>APPENDIX 2 – Strange Stories Test</b> .....	61
<b>APPENDIX 3 – Animations Test</b> .....	68

**ABBREVIATIONS**

ADOS.....	Autism Diagnostic Observation
AQ.....	Autism-Spectrum Quotient
AS.....	Asperger's Syndrome
ASD.....	Autism Spectrum Disorder
CFMT.....	Cambridge Face Memory Test
CFPT.....	Cambridge Facial Perception Task
DSM – IV.....	Diagnostic and Statistical Manual of Mental Disorders
FFA.....	Fusiform face area
HFA.....	High-functioning Autism
IFC.....	Inferior Frontal Cortex
IQ.....	Intelligence Quotient
ITG.....	Inferior Temporal Gyri
PDD.....	Pervasive Developmental Disorders
PDD – NOS.....	Pervasive Developmental Disorders Not Otherwise Specified
SDD.....	Social Developmental Disorders
STS.....	Superior Temporal Sulcus
ToM.....	Theory of Mind
UCL.....	University College London
WASI.....	Wechsler Abbreviated Scale of Intelligence

## 1.0 ABSTRACT

Autism spectrum disorders (ASD) are characterized by severe and pervasive impairment of reciprocal social interaction and communication as well as a restricted range of behaviors and interests. A theory by Robert Schultz (Schultz 2005) proposes that the social cognition deficits of these disorders are caused by facial recognition areas that do not develop properly, resulting in the developing child not acquiring an interest in faces and social interactions, and eventually resulting in the impaired social cognitive traits characteristic of ASD subjects. To test this, developmental prosopagnosics were studied in order to investigate whether or not they presented with any social cognition problems, either supporting or rejecting facial processing problems as the cause of social cognition problems in autism spectrum disorders. Two methods were utilized: the first being case studies of two developmental prosopagnosics using autism diagnostic tools and theory of mind tests, and the second being a study of twelve developmental prosopagnosics using an online autism questionnaire and an online theory of mind test. The results show that all twelve subjects did well on the theory of mind tests and only one prosopagnosic scored above the ASD cut-off point on the autism-quotient questionnaire. This demonstrates that impaired face processing does not necessarily lead to social cognition deficits and so suggests that social cognition deficits in ASD are not caused by pre-existing face processing problems.

## 2.0 INTRODUCTION

### 2.1 *What are Autism Spectrum Disorders?*

Human beings are naturally a very social species. We rely on communication and social interaction for a majority of our everyday activities, which is why Autism Spectrum Disorders (ASD) can make life more challenging for those diagnosed with an ASD as well as surrounding friends and family. ASD is a category of neurodevelopmental disorders characterised by severe and pervasive impairment of reciprocal social interaction and communication, as well as a restricted range of behaviors and interests. This category is classified by the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) (Stark & Ellis 1981; American Psychiatric Association Task Force on DSM-IV 1995; Schwarzer *et al.* 2006) under Pervasive Developmental Disorders (PDD) and includes Autism, Pervasive Developmental Disorder Not Otherwise Specified (PDD-NOS), Asperger's Syndrome (AS), Rett's Disorder, and Child Disintegrative Disorder. The rate of PDD within the general population is thought to range from 27.5 to 70 per 10,000 individuals (Fombonne 2003), with autism, in particular, affecting 2 to 11 per 10,000 individuals (American Psychiatric Association Task Force on DSM-IV 1995; Fombonne 2003; Centers for Disease Control and Prevention 2007).

The most common standardized tool for the assessment of an ASD is the Autism Diagnostic Observation Schedule (ADOS) (Lord *et al.* 2001; Lord *et al.* 2002). It involves a series of structured and semi-structured tasks based on the diagnostic criteria found in the DSM-IV and it allows the experienced examiner to observe the social and communication skills of the examinee. The aforementioned diagnostic criteria are divided into three main categories: (1) qualitative impairment in social interaction, (2) qualitative impairment in



communication, and (3) restricted and repetitive and stereotyped patterns of behavior, interests, and activities (American Psychiatric Association Task Force on DSM-IV 1995). One of the subcategories under the “impairment of social interaction” category includes the inability to communicate nonverbally, such as through eye-to-eye contact and through facial expressions, during social situations (American Psychiatric Association Task Force on DSM-IV 1995; Lord *et al.* 2002). This puts an emphasis on the importance of recognizing, understanding, and interpreting the face for communication and how this is lacking in ASD subjects.

## 2.2 *The association between facial processing and ASD*

The level of performance on facial and emotional recognition tasks in autistic subjects is conflicting and can range from severely impaired to levels that, on the surface, appear similar to control subjects. For example, a pioneering study by Langdell (Langdell 1978) showed that not only did autistic children not show the inversion effect, which is the decrement in recognition seen in control subjects when viewing upright faces compared to inverted faces, but they actually performed better than controls in recognizing inverted faces. Hobson *et al.* (Hobson *et al.* 1988) also showed that adolescents with autism performed better in both expression and identity matching of inverted faces compared to controls. In contrast, Tuenisse and deGelder (Teunisse & deGelder 2003) found that that not only was the inversion effect seen in a group of adolescents with autism, but these subjects also responded faster and had a better recognition rate when faces were presented upright compared to inverted. A more recent study by Lahaie *et al.* (Lahaie *et al.* 2006) also demonstrated similar results. They found the inversion effect present in a group of sixteen adolescent and adult autistics, with a

significantly higher error rate in the autistic group for inverted faces compared to upright faces, which was not seen in controls. However, significantly different reaction times were seen in both groups, with a significantly longer reaction time present when viewing inverted faces compared to controls. Lahaie suggests that their conflicting results in comparison to Langdell and Hobson *et al.* (Langdell 1978; Hobson *et al.* 1988) could be because these studies only looked at error rates, which were weak, rather than also investigating reaction time, and also because these studies used a small number of trials in comparison to the 8 blocks of 26 trials used by Lahaie *et al.*

In addition to these findings, another study found that children with autism have a significant deficit in facial recognition compared to PDD-NOS and other non-PDD developmental disorders based on the results of the K-ABC Face Recognition Task (Klin *et al.* 1999). These children were matched for chronological age, non-verbal mental age, and verbal mental age, so the deficit could not be attributed to verbal or nonverbal impairments. Also, two visual perceptual control tasks were administered to ensure that there were not deficits due to general task demands or general visual perceptual deficits. The first test was the Gestalt Closure task, which required the subject to identify incomplete inkblot pictures by mentally filling in missing gaps. The second was a special memory task, in which the subject was shown a page with an arrangement of random pictures and was allowed to look at the page for five seconds and was then required to recall where on the page the pictures were. There were no differences on either of these tasks between the autism and control groups, so the deficit seen could not be due to visual memory deficits.

Barton *et al.* (Barton *et al.* 2004) found a mix of results in a group of 24 adults with a variety of social developmental disorders (SDD). One group of 8 performed at the

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

same level as normal controls on the facial recognition task. The remaining subjects were impaired in facial recognition, but still performed better than a comparison group of prosopagnosia subjects (subjects identified as having an impairment in facial perception and facial memory). These conflicting results could possibly be due to the heterogeneity of symptoms and symptom severity as well as the methods of compensation in some ASD subjects in an effort to overcome their facial recognition deficits. However, despite these conflicting results, the large number of autistic subjects who have been found to have facial recognition problems has suggested to some researchers that the process of recognizing faces has an important relationship with the pathology of the disorder.

The ability of persons with ASD to compensate for their impairment can be revealed in some tasks by increasing the sensitivity of the task through reaction times. An example of the importance of reaction time was noted in facial and emotional recognition tasks in three boys diagnosed with AS (Njokiktjien *et al.* 2001). Two of the boys were able to accurately identify facial emotions, however their reaction time was very slow in most identifications. One of these boys appeared to choose very salient features, such as an open mouth with teeth showing for a happy face, to identify the emotion being depicted, which assisted in increasing his reaction time in some identifications. The third boy was not able to compensate for his deficits and was not only slow, but very inaccurate in the identification of emotion.

Because of the pattern of familial inheritance and a focus on twin studies in ASD, as well as an early onset of impairment, there appear to be strong genetic mechanisms involved (Pickles *et al.* 1995), however a concrete neural basis of origin has not yet been identified. Large strides have been made regarding the pathology of autism by gathering information through human lesion, postmortem, and functional imaging studies, many of

which implicate activation in temporal lobe structures (Sweeten *et al.* 2002). The limbic system and the amygdalae, in particular, have been implicated as playing an important role in the recognition of facial expressions as well as applying emotional significance to important environmental stimuli, with particular emphasis on socially-related stimuli (Morris *et al.* 1998; Anderson & Phelps 2001; Adolphs *et al.* 2002; Gaigg & Bowler 2007). The processing of negative expressions has been linked to the left amygdala while general face processing, regardless of emotion, has been linked to the right amygdala (Iidaka *et al.* 2001).

The fusiform face area (FFA) has also been explored using both emotional faces and non-salient faces. Healthy subjects have shown that there is a stronger activation of the FFA when looking at facial expressions compared to neutral faces (Pessoa *et al.* 2002). In comparison, ASD subjects show varying patterns of activation. One study compared ASD subjects to controls and found significantly less activation in the middle aspect of the right fusiform gyrus, but significantly greater activation of the inferior temporal gyri (ITG). The author hypothesizes that this abnormal pattern utilizes a feature-based approach for the recognition and analysis of faces rather than the holistic approach that most people use (Schultz *et al.* 2000). In comparison, a different study of ASD subjects found significant activation of the FFA and inferior occipital gyrus (Hadjikhani *et al.* 2004; Hadjikhani *et al.* 2007). However, in addition, a pattern of hypoactivation was seen in other areas involving face processing as well as areas identified in emotional identification and processing, including: the right amygdala, inferior frontal cortex (IFC), superior temporal sulcus (STS), and face-related somatosensory and premotor cortex (Hadjikhani *et al.* 2007). These results would support an ASD pathology that involves the

limbic system, but where FFA function may or may not be an additional symptom of the syndrome rather than a part of the cause.

### *2.3 Models for the etiology of autism*

There have been a number of hypotheses that attempt to explain the etiology of ASD and, in particular, autism. Schultz (Schultz 2005), for example, hypothesizes that the amygdala and other related structures causes an infant to become particularly interested in faces, leading to an “enhanced salience of faces”. This interest, experience, and increased skill in perception enhances connections in the FFA and other face processing areas, resulting in a person becoming an expert with faces. Facial expertise and the understanding of emotion which comes from facial expressions provides a “scaffolding” upon which the understanding of social skills and social interaction is formed. A diagram of this model can be seen in figure 2.1. This hypothesis goes on to suggest that if there is a disruption in the initial development of face processing areas, then this causes a disruption in the ability to acquire proper social skills, resulting in social developmental disorders such as autism. Other evidence suggests that abnormalities in the amygdala and its associated regions may lead directly to an autistic subject’s impaired emotional and social cognition, for example the “amygdala theory of autism” (Baron-Cohen *et al.* 2000). As the name would imply, this theory suggests that amygdala, dysfunction could be a possible cause of social cognition deficits seen in autistics. These deficits include the inability to understand others’ intentions, feelings, and desires, an inability to interact in complex social situations, and the lack of empathy of others’ state of mind. In addition, a review of lesion studies in both humans and non-human primates suggests lesions to the temporal lobe, and more specifically areas

surrounding the amygdala, results in impairments in social cognition that mirror the symptoms of autism (Sweeten *et al.* 2002). The difference between the hypotheses presented by Baron-Cohen et al. and Sweeten at al. compared to Schultz is that the former propose that amygdala dysfunction leads directly to the social and emotional problems of autistic subjects, whereas Schultz suggests that an autistic's social problems stem from the influence of the amygdala causing abnormal development of the FFA and other facial processing regions, and that the inability to process faces properly leads to the social cognition problems of autism.

POSSIBLE THEORY OF THE ETIOLOGY OF AUTISM

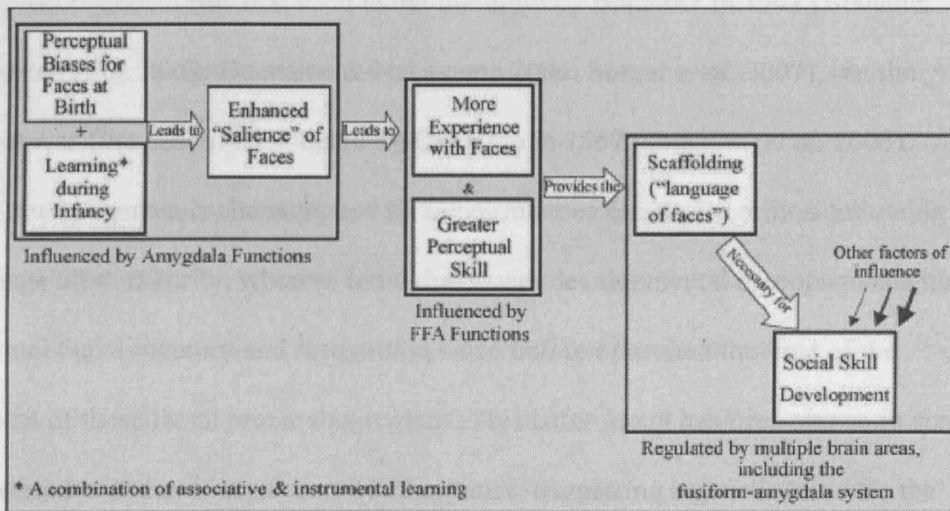


Figure 2.1: A model suggested by Schultz (Schultz 2005) associating the amygdala, FFA, and other brain areas with the social cognition deficits present in autistic subjects.

#### *2.4 Developmental prosopagnosia as a way to investigate the link between facial processing and social cognition*

As previously mentioned, prosopagnosia is an impairment in facial perception and facial memory despite intact intellectual and cognitive function (Sorger *et al.* 2007) and can be the result of abnormalities in perception and / or memory (Harris & Aguirre 2007). The term prosopagnosia was first used in the literature by Bodamer in 1947 (Bodamer 1947; Sweeten *et al.* 2002; Duchaine & Nakayama 2006; Sorger *et al.* 2007), but the impairment was first described in detail by Quaglino in 1867 (Quaglino *et al.* 2003). Acquired prosopagnosia is characterized by impairment of facial recognition following brain damage after maturity, whereas individuals with developmental prosopagnosia have had abnormal facial memory and recognition since before or around the time of the development of these facial processing regions. This latter group has been shown in some cases to present with familial patterns of inheritance, suggesting a genetic basis for the impairment (Behrmann & Avidan 2005). For the purposes of comparing social cognition in autistics to that in prosopagnosics, the subjects used for testing are developmental rather than acquired prosopagnosics. This is because both autistics and developmental prosopagnosics have deficits of the developing brain rather than deficits caused by brain damage after these regions have completely formed.

Similarities have been seen between prosopagnosia and some autistic subjects in regards to the way that both groups have been shown in some cases to process faces at a local level rather than utilizing configural processing at a global level (Boutsen & Humphreys 2002; Behrmann & Avidan 2005; Behrmann *et al.* 2006; Rondan & Deruelle 2007). A long standing theory has been that it is this loss of configural processing that results in an impairment in facial processing because the face is seen as pieces rather than



## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

as a whole (Behrmann & Avidan 2005). The inability to view faces in a global manner is supported by the inversion effect (Leder & Bruce 2000). Some individuals with autism (Langdell 1978; Hobson *et al.* 1988), as previously mentioned, as well as some individuals with prosopagnosia (Nunn *et al.* 2001; Boutsen & Humphreys 2002) do not show the decrement in performance that is seen in normal subjects and some even show inversion superiority (Farah *et al.* 1995; de Gelder *et al.* 1998). Because they are identifying the face based on its parts rather than on the face as a whole, changing the orientation of the face from an upright to an upside-down position does not cause the normal decrement in performance, and can even increase proficiency in face perception. Since normal subjects are used to processing faces in an upright position, they must switch to a piece-meal form of processing when the face is turned upside-down, costing performance accuracy and reaction time.

In contrast to this configural processing hypothesis is evidence that not all prosopagnosics suffer from this type of processing impairment. Duchaine (Duchaine 2000) tested a developmental prosopagnosia subject using the three tests from the Kit of Factor-References Cognitive Tests, which were tests used by Levine and Calvanio (Levine & Calvanio 1989) to show configural processing deficits in a different prosopagnosia subject. He found that his subject scored well on all three tests, and therefore a general purpose configural processing impairment could not be the cause of all prosopagnosias. This shows that there could possibly be a number of processing pathways that are abnormal during development, resulting in prosopagnosias that seem to have similar facial processing deficits on the surface, but function differently when tested.

Similarities have also been seen in the gaze pattern of both autistic and developmental prosopagnosia subjects. Both tend to look significantly more at outlying parts of the face when identifying faces, as shown in figure 2.2 and 2.3, whereas normal controls would focus more on the central part of the face. Schwarzer et al. (Schwarzer *et al.* 2006) showed this in a group of hereditary prosopagnosia subjects (he uses the term hereditary to describe the familial transmission of impairment in facial recognition). The differing gaze patterns were consistent for both controls and prosopagnosics whether they were viewing familiar or unfamiliar faces. With regards to autistics, their dispersed gaze patterns differ slightly from prosopagnosics because their gaze tends to try to avoid looking at central features of the face whereas prosopagnosics still look at central parts of the face, but rely more on surrounding features in comparison to normal subjects (Pelphrey *et al.* 2002; Schwarzer *et al.* 2006). A differing gaze pattern in comparison to controls may reflect the way that the two groups try to recognize the person at whom they are looking. A possible theory is that people who do not have an impairment look at the main features of the face and how those features are spaced. They take in the face as a whole, processing it in a global fashion. Prosopagnosics and autistics, on the other hand, search the face for recognition and use other features such as overall face shape and hairstyle to piece together who the person is that they are seeing.

Gaze Patterns in Prosopagnosia versus Control Subjects

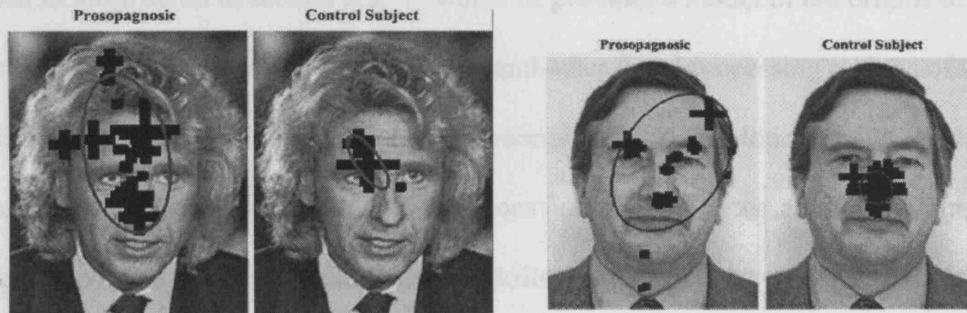


Figure 2.2 - These images show the gaze pattern of a prosopagnosia subject compared to a control subject. As you can see, the prosopagnosia subjects tend to look at more outlying regions of the face, whereas the control subjects tend to look towards a central region of the face. Images are from Schwarzer et al. (Schwarzer *et al.* 2006), but were originally found in Stark and Ellis (Stark & Ellis 1981).

Gaze Patterns in Autistic versus Control Subjects

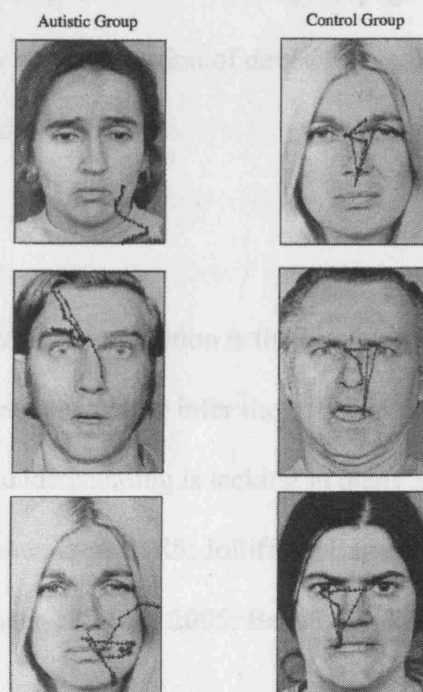


Figure 2.3 - This figure shows samples of scanpaths from autistic and control subjects who were given the task of identifying the emotion of the person in the picture. Images are from Pelphrey et al. (Pelphrey *et al.* 2002).

### *2.5 Social Cognition and Prosopagnosia*

In this paper, we explore the theory put forward by Schultz (Schultz 2005), as described in more detail in section 2.3, in which he provides a model of the origins of autistic behavior. This theory requires the FFA and other facial processing regions of the brain to function normally in order for a child's social skills to develop normally. In the absence of normal facial processing, the child does not have the necessary "scaffolding" upon which to develop higher social cognitive skills, leading to the symptoms that define autism, especially the inability to understand non-verbal communication during social interactions. In this theory, normal face processing would be a prerequisite for normal social cognition, and this, therefore, would predict that individuals with severely impaired face processing would also show social cognition deficits. Schultz even states that "autism has close parallels to associative agnosia, and in particular may represent a naturally occurring form of developmental associative prosopagnosia (Schultz 2005). To test this, we will examine the social cognition of developmental prosopagnosics to see if they have normal social cognition skills.

### *2.6 Tests of Social Cognition*

An important aspect of social cognition is the idea of "theory of mind" (ToM). This is a person's ability to understand and infer the beliefs, intentions, and mental states of others. This inference and understanding is lacking in many ASD subjects, in particular autistics (Baron-Cohen *et al.* 1985; Jolliffe & Baron-Cohen 1999; Baron-Cohen & Belmonte 2005; Hale & Tager-Flusberg 2005; Beaumont & Newcombe 2006). Two

tests that were designed to require ToM processing are a stories recall test and an animations test.

The Strange Stories task was devised by Happé (Happe 1994) to look at the ToM in autistic individuals. The original version had 24 short stories in total and included a comprehension question and a justification question. They were created using ToM concepts that were more “contextually embedded” than previous ToM tests on which some subgroups of autistics did as well as controls. The twelve story types were based on the following concepts: lie, white lie, joke, pretend, misunderstanding, persuade, appearance / reality, figure of speech, sarcasm, forget, double bluff, and contrary emotions. Happé found that the autistic subjects, even those with a normal IQ, had difficulties with mental state attribution compared to both controls and mentally handicapped subjects. Jolliffe and Baron-Cohen (Jolliffe & Baron-Cohen 1999) replicated these results in adults: 17 autistic subjects, 17 AS subjects, and 17 controls. They found that the autistic and Asperger subjects used mental terms, but failed to use terms that were appropriate to the context of the story, with the autism performing slightly below that of the Asperger’s group. The Happé Strange Stories were revised by Fletcher et al. (Fletcher *et al.* 1995), who divided the stories into one test group, “theory of mind stories”, and two control groups, “physical stories” and “unlinked sentences”. The subjects were required to read each story and then answer one question on that story. The physical stories required inference beyond information stated, which mimicked the ToM stories, however they did not require any insight into the mental state of characters involved. The unlinked sentences were composed of random sentences, which did not require the inference or integration of information, but as in the other two groups of

stories, it required the reader to utilize short-term memory and to pay attention to the meaning of each sentence.

Another test that has been used to investigate ToM is an animations task based on a paradigm created by Heider and Simmel (Heider & Simmel 1944). They created a two and a half minute film, in which a rectangle, triangle, and star were shown moving around at various speeds and directions. At the end of each segment, the subject was asked to describe what he or she saw. They found that most of the 34 subjects would apply human attributions to the simple movements of the inanimate objects. Exceptions were two subjects who still described the movement of the geometric shapes in animate terms, but described the activity as birds rather than humans, and one subjects used mostly geometric terminology, describing the objects as purely inanimate. Castelli et al. (Castelli *et al.* 2000; Castelli *et al.* 2002) used a modified version of this task for the computer consisting of twelve short clips, each approximately 40 seconds long. The twelve clips were composed of four ToM, four goal-directed, and four completely random sequences. Each clip involved a large red triangle and a smaller blue triangle, and half of the sequences utilized an enclosure that the triangles could move in and out of. 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 with little to no use of mental state inference, such as dancing or fighting. In the random sequences, the triangles moved in a completely random manner around the screen. After each clip, the subjects were asked to describe what they saw the triangles doing. Castelli et al. (Castelli *et al.* 2002) showed that there was not a significant difference between descriptions given for

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

the goal-directed or random sequences between the autistic group and the control group, however there was a significant difference between the autistic and control groups for the ToM animations, with the autistics giving “fewer and less appropriate” descriptions.

In support of these two ToM tests, an Autism-spectrum Questionnaire (Baron-Cohen *et al.* 2001) and the ADOS (2001) were administered to examine whether or not the subjects had a significant number of autistic characteristics. Combined, the AQ, ADOS, animations tests, and strange stories test were used to investigate how developmental prosopagnosics compare to the results seen in autistic subjects, particularly the poor ToM results seen in autistic subjects. If the developmental prosopagnosics’ social cognition is similar to scores seen in autistic subjects, this would be consistent with Schultz’s hypothesis that a failure to develop face processing leads to impaired social cognition. However, if the developmental prosopagnosic's social cognition is normal, this would demonstrate that developmental facial recognition deficits do not invariably lead to social cognition problems and would suggest that social cognition problems in ASD are not the result of face processing impairments.

### 3.0 METHODS 1

The first part of our methods focused on a more in-depth study of social cognition in two prosopagnosia subjects, TU and DH. The reasoning behind this is to first describe the nature of their prosopagnosia and then to look at their personal lives and social interactions in a more thorough manner. We would like to search for any social cognitive characteristics that might be similar to those seen in autistic subjects. Due to time constraints, this in-depth look into the social cognition was not possible for the large group of subjects.

#### 3.1 Subjects

TU is a 31 year old male who has had no known brain injuries throughout his life. He has been involved in prosopagnosia testing since 2005 and some of his have been published in a recent paper by Duchaine (Duchaine *et al.* 2007), results of which can be found under subject M30. DH is a 30 year old male who had a severe seizure shortly after birth, but has sustained no other known injuries that could have caused brain damage since then. In addition, DH has described facial recognition problems in his father and believes that there is a possibility that he could also have prosopagnosia. Both TU and DH have been tested with a number of standardized as well as more recently created batteries of facial recognition and memory tests to investigate their impairment in facial processing. Some of these main tests include the Cambridge Face Memory Test (CFMT), Cambridge Facial Perception Task (CFPT), and a famous faces task.

The CFMT uses six novel faces and four stages of testing: a practice stage that uses cartoon faces to mimic the stages to follow; an introduction / same images stage



## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

where the six novel faces are presented at a left 1/3 profile, a frontal view, and a right 1/3 view; a novel images stage in which the subject is presented with three faces and is forced to choose which face is one of the target faces previously seen; and finally a novel images with noise scenario, which is identical to the previous scenario, but with the addition of Gaussian noise disturbing the facial images. This final part of the test is thought to increase the dependence of facial recognition on the special mechanisms that are normally involved in every-day facial recognition and also keep performance from the ceiling.

CFPT uses one target face at a  $\frac{3}{4}$  profile position and six frontal view faces, which were created using a continuum of morphed faces. The first of the six faces was morphed to an 88% likeness of the target face. The remaining five faces are morphed at 76%, 64%, 52%, 40%, and 28%, decreasing the percentage of similarity to the target face. The six morphed faces and the one target face are then presented together and the subject is asked to sort the faces based on their similarity to the target face. Each trial is presented upright once and inverted once.

The famous faces test consists of 60 images of well-known people that are cropped so that little hair or clothing could be seen. The subjects are presented with each image for three seconds and then asked to name the person presented and provide identifying information about that person, for example if they were in a movie or if they had a particular political role. The test was a UK version since both TU and DH are from the UK.

Before testing, TU and DH were interviewed in order to understand more about their daily lives and social habits, to find out how prosopagnosia affects the way that they

interact with others, and to see if these two subjects have social cognition impairments characteristic of autistic subjects.

TU is a graphic designer who enjoys drawing and web design work. He has said that he enjoys social situations, but finds them very exhausting. He prefers to stay at home on the weekends, but will join his girlfriend to go to parties, galleries, or other social events. Before larger social events, his girlfriend will give him a reminder briefing on which friends are which and he usually works on the basis of a person recognizing him first. He says "if someone I know quite well walks straight past me and doesn't see me or look at me, I'm not going to trust myself because you can fool yourself...I keep thinking I see people who I think I know because of their mouths or their face shapes or whatever it is". He also mentioned that finally being diagnosed with prosopagnosia was a huge relief and that it actually made social situations easier because the people around him were much more understanding if he did not immediately recognize them. TU is able and willing to be a part of larger social situations, such as a larger party, but prefers interacting with smaller gatherings of friends. Overall, the interview with TU was very comfortable. He was soft-spoken at times, but the conversation was very smooth and easy; there were reciprocal questions being asked, jokes were made, and eye contact was maintained throughout the conversation.

DH, an architect, has learned to cope with prosopagnosia by expending extra effort in trying to remember people he meets, with meeting new friends in smaller groups tending to be easier than meeting people in larger groups (e.g. a wedding). He tends to be shy in these larger groups, but more outgoing when he is with close friends. While talking with him before testing he mentioned that he overcompensate at times for his

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

facial recognition problems, for example on his walk from the tube station to work in the mornings he is quite friendly and will say “hello” to more people than he probably should, just in case he knows the person walking next to him from the office but just doesn’t remember him outside of his work environment. Prosopagnosia makes it more difficult for him in social situations, however he is very willing to make the additional effort to try to recognize a person who he has already met but cannot immediately remember. On the weekends, he is more or less social depending on his mood. For example, if his wife is away for the weekend, he spends much of his weekend at home, or if she is around, they will watch a film or visit a gallery, meet his friends out, or go for a walk. Like TU, the interview with DH was very relaxed. He answered all of my questions and was very curious after the testing was completed as to the reasoning behind it all. He seemed kind, kept eye-contact, and although he didn’t answer my questions in as much detail as TU, he kept the interview conversation going effortlessly.

### *3.2 Tests*

#### 3.2.1 WESCHLER ABBREVIATED SCALE OF INTELLIGENCE (WASI) (1999)

All four subsections of the WASI (vocabulary, block design, similarities, and matrix reasoning) were used to investigate the intellectual abilities in DH and TU. This was done to show that if any deficits were observed in the strange stories or animations tasks, they could not be attributable to verbal or non-verbal impairments.

### 3.2.2 AUTISM DIAGNOSTIC OBSERVATION SCHEDULE (ADOS), MODULE 4 ( 2001)

The ADOS was administered by a trained and qualified researcher to see if subjects DH and TU presented with any traits that were similar to those seen in ASD subjects. Module 4, in particular, was used because it is specific for fluent adults. As previously mentioned, the ADOS is a series of structured and semi-structured tasks and questions which allow the interviewer to gain insight into any possible ASD characteristics of the interviewee. The five main sections include: language and communication, reciprocal social interaction, imagination, stereotyped behaviors and restricted interests, and other abnormal behaviors. A score of 7 is a useful cut-off point in the Communication plus reciprocal social interaction sections for an ASD, while a score of 10 or more is the cut-off point for autism.

### 3.2.3 AUTISM-SPECTRUM QUOTIENT QUESTIONNAIRE (AQ) (BARON-COHEN ET AL. 2001)

The AQ is a short, self-administered questionnaire with 50 items used to measure “the degree to which an adult with normal intelligence has the traits associated with the autistic spectrum” (Baron-Cohen *et al.* 2001). The two subjects were sent the questionnaire by mail, asked to circle the answers that were best associated to their own personal traits, and asked to return it in the accompanying envelope. Each question was scored either 0 or 1 based on the answer circled (see appendix 1 for the full questionnaire and scoring). In order to elucidate a useful cut-off score between an individual who shows a clinically significant number of ASD traits and an individual who does not, Baron-Cohen *et al.* (Baron-Cohen *et al.* 2001) analyzed questionnaires from a group of

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

58 adults with Asperger's syndrome or high-functioning autism (HFA) (all of whom had been diagnosed by psychiatrists using appropriate ASD diagnostic criteria) and a group of 174 adult controls selected at random living in East Anglia. An analysis of these two groups suggests that a total score of 32 or more on the questionnaire is a good cut-off point for individuals who have a clinically significant number of autistic traits and those who do not. At this point, 79.3% of AS / HFA subjects scored at this level, while only 2.3% of controls scored at this level.

### 3.2.4 STRANGE STORIES TEST

The twelve stories used in this experiment were those created by Fletcher et al. (Fletcher *et al.* 1995) based on Happé's Strange Stories (Happé 1994) and can be found in appendix 2. The questions to the unlinked stories were modified by Dr. Sarah White, Institute of Cognitive Neuroscience, UCL, in 2004 to increase difficulty by changing the questions from closed (e.g. "Was the mother brave?") to open-ended (e.g. "Who was brave?"). Dr. Sarah White also contributed the control data for this task. She and her colleagues collected the data through a number of different means, including local ads in the community and UCL, friends and acquaintances of the laboratory, UCL Institute of Cognitive Neuroscience subject database, et cetera. The original size of the control group was 46, but was reduced to 24 by eliminating those whose IQ's were below 110. This was done in order to better match the mean IQ of the controls to the IQ of the case studies. The mean full-scale IQ of the controls is 120 (sd = 7.1), the mean age is 36.1 (sd = 12.3), the gender ratio of males to females is 1.4:1.

Our two subjects, TU and DH, were given the stories face down. One story was written on each page and one question pertaining to that story was written on the following page. Each subject was given the same instructions, which can be found in appendix 2. When the subject was ready, he turned over the page with the story. Reaction time for how long he spent reading the story was recorded. He then turned over the sheet with the question on it, answered the question aloud, and his answer was recorded. Scoring was done on a zero to two point scale based on the accuracy of the answer given. See appendix 2 for stories, questions asked, and scoring.

### 3.2.5 ANIMATIONS TEST

The animations task, as previously described, was that used by Castelli et al (Castelli *et al.* 2000). The subjects were given identical instructions, which can be found in appendix 3, along with details regarding scoring. The scoring of each answer was based on three criteria: intentionality, appropriateness, and length. The intentionality score was on a scale of zero to five points and reflected the use of mental state terms. The appropriateness score used a point scale of zero to three and evaluated how well the subject understood the animations, as intended by the designer. The length score ranged from zero to four and was based on the number of clauses used to describe the animations.

The controls for this task were recruited by an email sent to UCL Institute of Neurology MSc students requesting volunteers, as well as through friends, colleagues, and acquaintances of those who received this initial recruiting email. Twelve controls were tested in total. The mean age of the controls is 29.6 (sd = 9.86) and the gender ratio

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

of males to females is 1:1. Due to time constraints, IQ tests were not performed on the control subjects, however every subject was either pursuing or had completed an undergraduate degree, with some having further education such as a master's degree or medical degree. For this reason, any differences that may arise are most likely not attributable to an intellectual deficit in the control group. None of the tested controls have been diagnosed with an ASD and none reported any sort of facial memory or facial processing deficits.

## 4.0 RESULTS 1

To begin, both TU and DH scored similarly on the verbal and performance sections of the WASI, as seen in table 4.1. TU has a full scale IQ of 128 while DH's full scale IQ is 126. Poor performance on any of the social cognition tests, therefore, cannot be attributed to low intelligence or an inability to understand the directions or the written material.

### 4.1 ADOS Results

Additionally, the results from the ADOS, module 4 showed that TU and DH do not have any social or communication problems that are characteristic of ASD subjects. TU scored zero points under the "communication" category and one point under the "reciprocal social interaction" category. This one point was because he slightly interrupted the examiner on a couple of occasions, for example by asking a question and then talking over the response. DH had zero points on both the "communication" and "reciprocal social interaction" categories. Both of these scores are far from the autism spectrum cut-off of 7 points, and even farther from the autism cut-off of 10 points, which are obtained when the "communication" and "reciprocal social interaction" scores are added together. Based on the ADOS, module 4, neither subject is diagnosable as having an autism spectrum disorder.

### 4.2 AQ Results

TU scored 15 on the AQ while DH score 14, which are both well below the cut-off score of 32. Both TU and DH's scores are significantly lower ( $p = 0.002$  and  $0.001$ ,



## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

respectively) than the mean AS / HFA mean score (n = 58, mean = 35.8, sd = 6.5) and neither score is significantly (p = 0.82 and 0.70, respectively) different from the control mean n = 174, mean = 16.4, sd = 6.3).

## WESCHLER ABBREVIATED SCALE OF INTELLIGENCE SCORES

<b>TU (age 31 years old)</b>				
	<b>Sum of T scores</b>	<b>IQ</b>	<b>Percentile</b>	<b>95% confidence interval</b>
<b>Verb.</b>	123	119	90	113 - 124
<b>Perf.</b>	135	132	98	125 - 136
<b>Full-4</b>	258	128	97	123 - 132
<b>DH (age 30 years old)</b>				
	<b>Sum of T scores</b>	<b>IQ</b>	<b>Percentile</b>	<b>95% confidence interval</b>
<b>Verb.</b>	121	117	87	111 - 122
<b>Perf.</b>	133	129	97	122 - 133
<b>Full-4</b>	254	126	96	121 - 130

Table 4.1 – Verbal IQ, performance IQ, and full scale IQ of TU and DH using the Wechsler Abbreviated Scale of Intelligence.

### 4.3 *Strange Stories Results*

With regards to the raw data from the strange stories tests, table 4.2 shows that the total scores for the physical stories and the unlinked sentence passages were much more variable between TU (phys = 12, unlink = 11) and DH (phys = 9, unlink = 2) compared to the similar score they obtained for the ToM stories (15 and 14, respectively). Graphs 4.1 and 4.2 elucidates this variability even more.

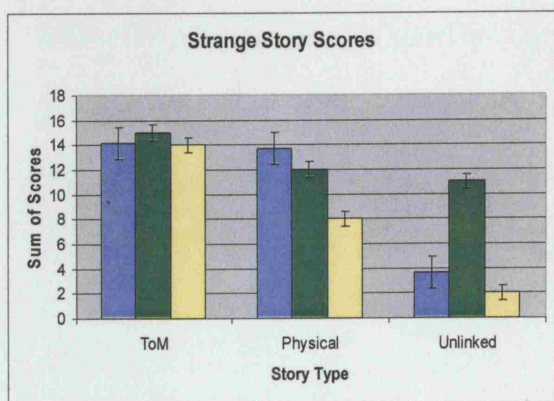
The statistical program used to compare the case study scores to the control data was created by Crawford and Garthwaite (Crawford & Garthwaite 2007). It computes a p value based on the mean, standard deviation, and sample size of the control group in comparison to the case study score. The p values for the case studies can be found below in table 4.3. There was not a significant difference between the control sample mean or the two case studies for either full-scale IQ (TU,  $p = 0.34$ ; DH,  $p = 0.5$ ) or age (TU,  $p = 0.69$ ; DH,  $p = 0.63$ ). The control sample, albeit small, is a good match to the age and intellectual abilities of TU and DH. The ToM scores (TU,  $p = 0.69$ ; DH,  $p = 0.63$ ) and reaction times (TU,  $p = 0.98$ ; DH,  $p = 0.38$ ) also showed no significant difference from the control means. This provides support that these two developmental prosopagnosics do not have a deficit in ToM. DH showed a significant difference in his physical score (TU,  $p = 0.26$ ; DH,  $p = 0.001$ ), but no significant difference was seen in either subject's reaction time (TU,  $p = 0.23$ ; DH,  $p = 0.45$ ) for this story type. The unlinked sentence passages showed no significant difference for reaction time (TU,  $p = 0.77$ ; DH,  $p = 0.52$ ), but showed a very significant different between scores (TU,  $p = 0.0005$ ; DH,  $p = 0.0002$ ), with TU scoring significantly higher than the controls and DH scoring significantly lower.

STRANGE STORY SCORES AND REACTION TIME

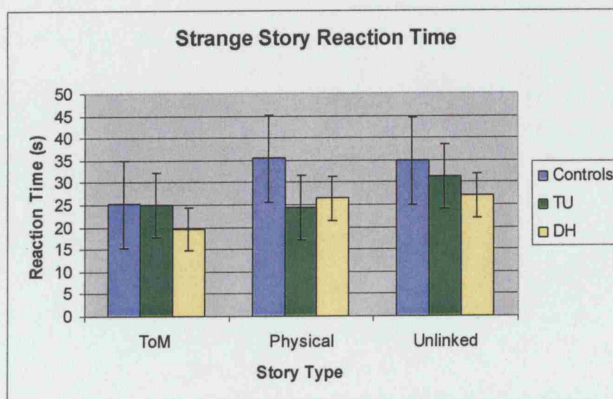
	fs IQ	Age	ToM score	ToM rt (s)	Phys score	Phys rt (s)	Unlinked score	Unlinked rt (S)
<b>Controls n = 24</b>	120.5 (7.1)	36.08 (12.3)	14.13 (1.5)	25.23 (6.2)	13.75 (1.5)	35.45 (11.6)	6.67 (1.0)	34.81 (11.7)
<b>TU</b>	128	31	15.0	25.1	12.0	24.5	11	31.3
<b>DH</b>	126	30	14.0	19.6	8.0	26.4	2	27.0

Table 4.2: The mean (s.d) full scale IQ and age for the control sample is shown, in addition to the mean (s.d) of the summed scores for each story group (theory of mind, physical, and unlinked sentences) and the mean (s.d) reaction time for each story group (theory of mind, physical, and unlinked sentences). The actual IQ, age, summed score, and mean reaction time are listed for TU and SH.

GRAPHIC COMPARISON OF MEAN STRANGE STORY SCORES AND REACTION TIMES



Graph 4.1: Compares the mean sum of scores for each story type for controls, TU, and DH.



Graph 4.2: Compares the mean reaction time to read each story type for controls, TU, and DH.

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

### ANALYSIS OF STRANGE STORY SCORES AND REACTION TIME

	fs IQ	age	ToM score	ToM rt	Phys score	Phys rt	Unlinked score	Unlinked rt
<b>TU</b>	0.34	0.69	0.56	0.98	0.26	0.23	0.0005*	0.77
<b>DH</b>	0.50	0.63	0.93	0.38	0.001*	0.45	0.0002*	0.52

Table 4.3: p values comparing the full scale IQs, ages, scores and reaction times of TU and DH to the control sample means. A significant difference is found for the physical score obtained by DH, as well as for the unlinked sentence passage scores obtained by both TU and DH.

#### 4.4 Animations Results

The final test results to be considered are for the animations tasks. Twelve control subjects with a mean age of 29.6 (sd = 9.86) were tested. As previously described, the animations task is rated based on three scoring criteria: intentionality (0-5), appropriateness (0-3), and length (0-4). Table 4.4 shows the mean scores of the control sample for each of these categories in comparison to the scores achieved by TU and DH. The scores are very similar between the control group and the two prosopagnosia, and the similarities can be seen in graphs 4.3, 4.4, and 4.5 below.

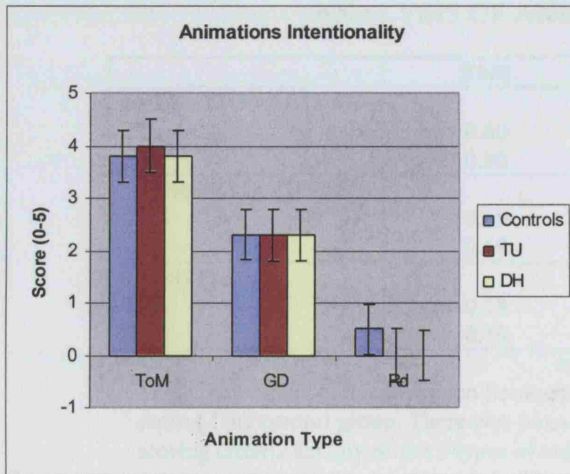
The Crawford and Garthwaite (Crawford & Garthwaite 2007) statistical software was also used to analyze the animations scores. The ages TU and DH were compared to the mean age of the control group (29.6, sd = 9.86), in addition to a comparison of the mean scores for all three animation types (refer to table 4.4 and graph 4.4). There was not a significant difference between the ages of TU and the controls ( $p = 0.89$ ) or DH and the controls ( $p = 0.97$ ). In addition, there were not any significant differences ( $p < 0.5$ ) for the intentionality scores appropriateness scores, or response length for either the ToM, goal directed, or random animation scenarios (refer to table 4.5 for exact p values). Neither TU or DH have a deficit in attributing human attributes to inanimate objects, which continues to support the previous evidence that these subjects do not have a ToM deficit.

ANIMATION TEST MEAN SCORES

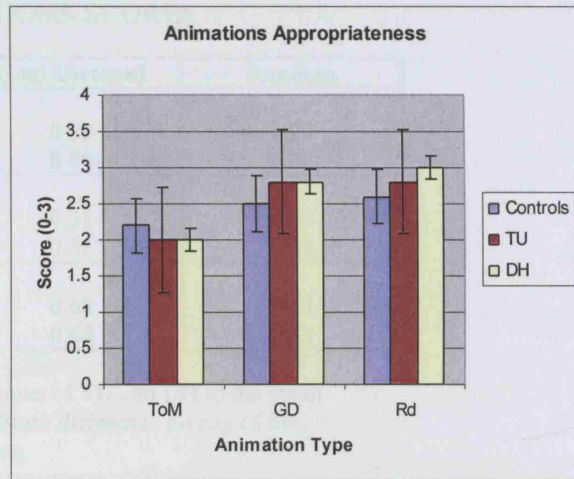
	ToM	Goal Directed	Random
<b>INTENTIONALITY (0-5)</b>			
Control (n = 12)	3.8 (0.6)	2.3 (0.4)	0.5 (0.4)
TU	4.0	2.3	0.0
DH	3.8	2.3	0.0
<b>APPROPRIATENESS (0-3)</b>			
Control (n = 12)	2.2 (0.5)	2.5 (0.3)	2.6 (0.4)
TU	2.0	2.8	2.8
DH	2.0	2.8	3.0
<b>LENGTH (0-4)</b>			
Control (n = 12)	2.8 (0.5)	1.8 (0.7)	1.6 (0.5)
TU	2.0	1.5	1.3
DH	2.5	1.5	1.3

Table 4.4: Each group of animations ToM, goal directed, and random, were scored using three criteria: intentionality (score range 0-5), appropriateness (score range 0-3), and length (score range of 0-4). The mean scores of each scoring criteria are presented for controls, TU, and DH.

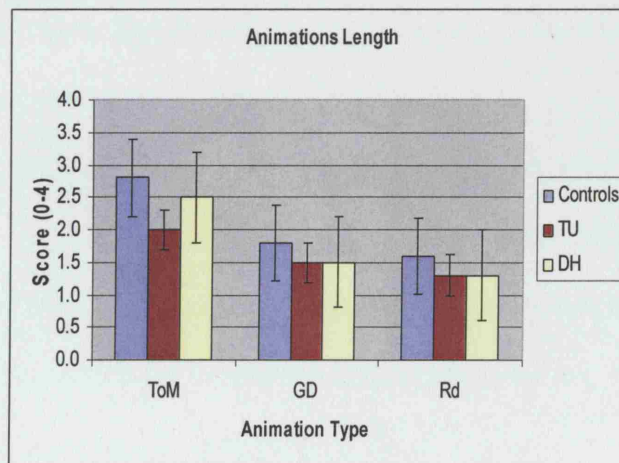
GRAPHIC COMPARISON OF ANIMATION TEST MEAN SCORES



Graph 4.3: Compares the mean intentionality scores of the control group, TU, and DH.



Graph 4.4: Compares the mean appropriateness scores of the control group, TU, and DH.



Graph 4.4: Compares the mean response length of the control group, TU, and DH.



## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

### ANALYSIS OF ANIMATIONS SCORES

	<b>ToM</b>	<b>Goal Directed</b>	<b>Random</b>
<b>INTENTIONALITY</b>			
TU	0.60	0.99	0.30
DH	0.90	0.99	0.30
<b>APPROPTIATENESS</b>			
TU	0.69	0.31	0.63
DH	0.69	0.31	0.35
<b>LENGTH</b>			
TU	0.18	0.68	0.61
DH	0.39	0.68	0.61

Table 5: p-values of a comparison between the scores of TU and DH to the mean score of the control group. There was not a significant difference on any of the scoring criteria for any of the 3 types of animations.

## 5.0 METHODS 2

The second part of this experiment involved a group of ten prosopagnosia subjects plus TU and DH, making a total of twelve subjects. The reasoning behind this larger group was to test whether or not the case studies were an exception to a general population of prosopagnosics. In contrast to method 1, the tests given to the ten prosopagnosics in method 2 were taken online due to time constraints of the project.

### 5.1 Subjects

The ten prosopagnosic subjects were chosen from a larger database of prosopagnosics who have contacted either [www.faceblind.org](http://www.faceblind.org) or Dr. Brad Duchaine directly because of their facial recognition problems. All have reported these facial recognition problems for as long as they can remember and all have undergone similar batteries of facial memory and facial recognition tests as TU and DH, specifically CFMT, CFPT, and famous faces test. These tests were described in greater detail in the aforementioned methods section. The ten subjects, in addition to TU and DH, performed very poorly on these tests of facial memory and recognition.

The mean age of the complete group of twelve subjects is 38.3 and the gender ratio of males to females is 1:1. The comparative control group for this strange stories task is the same group of 24 subjects described for the strange stories task in method 1. The mean age of this group is 36.1 and the male to female ratio is 1.4 to 1.

## 5.2 Tests

### 5.2.1 AUTISM-SPECTRUM QUOTIENT QUESTIONNAIRE (AQ)

The AQ contained the same questions and scoring as previously described, however it was given online rather than in a paper form. Each subject clicked next to the answer that best described their personal traits in relation to the question being asked: “definitely agree”, “slightly agree”, “slightly disagree”, “definitely disagree”.

### 5.2.2 STRANGE STORIES TEST

The strange stories test used the same instructions, stories, and questions as aforementioned, however it was also given online. Each subject read a story and, when finished, he or she would click a button to move on to the question. Reaction time was recorded for the time that it would take the subject to read the story, however, as in the paper version, he or she would have unlimited time to answer the question. The subject would type the answer to the question and then push a button to move on to the next story.

## 6.0 RESULTS 2

### 6.1 AQ Results

Each of the twelve developmental prosopagnosics completed the AQ. The raw data can be seen in table 5.1 and a plot of these scores is presented in graph 5.1. The mean score of the group is 18.0 (sd = 8.15), which is well below the cut-off score of 32 suggested by Baron-Cohen et al. (Baron-Cohen *et al.* 2001). All of the subjects performed below the ASD cut-off point with the exception of RS, who obtained a score of 37 on the questionnaire.

An independent t-test was performed to see if there was a significant difference between Baron-Cohen et al.'s AS / HFA group and control group compared to our developmental prosopagnosics. Table 5.2 shows that there is a significant difference ( $p < 0.001$ ) between the mean score of the prosopagnosics compared to AS / HFA group, however there was not a significant difference between the prosopagnosics and the control group ( $p = 0.405$ ). This provides further support that the average developmental prosopagnosic does not have enough autistic character traits to be categorized as having an ASD, however, there are exceptions. For example, as previously noted, RS had a score of 37 that surpassed the cut-off point of 32. According to the Baron-Cohen data, 50% of AS / HFA subjects (61.5% of AS / HFA females) scored at or above 37, while only 0.6% of controls (0.0% of control females) scored at or above this score. We, therefore, looked to see if there was a significant difference between her score and the mean scores obtained for the AS / HFA and control groups. Using Crawford and Garthwaite's single case analysis software (Crawford & Garthwaite 2007), a significant difference was seen between RS and controls as a whole ( $n = 174$ , mean = 16.4, sd = 6.3,  $p = 0.001$ ) and RS

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

and control females ( $n = 98$ , mean = 15.4,  $sd = 5.7$ ,  $p = 0.0003$ ). A significant difference was not seen, however, between RS and the AS / HFA group as a whole ( $n = 58$ , mean = 35.8,  $sd = 6.5$ ,  $p = 0.856$ ) or between RS and AS / HFA females ( $n = 13$ , mean = 38.1,  $sd = 6.9$ ,  $p = 0.814$ ).

RS's scores prompted a closer look at individual scores of the remaining subjects, so their scores were compared to both the AS / HFA and control groups as a whole, as well as divided by gender. The eleven remaining prosopagnosics' individual scores were not significantly different from the control group as a whole or when compared by gender. Nine prosopagnosics's scores were significantly different ( $p < 0.01$ ) from the AS / HFA group as a whole and by gender, however MT and DG did not show this significant difference ( $p = 0.204$  and  $0.518$  respectively when compared to the group as a whole and  $p = 0.141$  and  $0.105$  respectively when compared to only male AS / HFA subjects).

### *6.2 Strange Stories Results*

The second task given to our group of twelve prosopagnosics was the strange stories test. There was not a significant difference ( $p = 0.60$ ) between the mean control age of 36.1 ( $sd = 12.31$ ) and the mean prosopagnosic age of 38.3 ( $sd = 9.24$ ). The prosopagnosia and control subjects had similar results for the ToM (control = 14.3,  $sd = 1.45$ ; proso = 13.8,  $sd = 1.48$ ) and physical stories scores (control = 13.8,  $sd = 1.48$ ; proso = 13.0,  $sd = 2.04$ ), as well as for the reaction times for the ToM (control = 25.3,  $sd = 6.21$ ; proso = 24.8,  $sd = 11.76$ ), physical (control = 35.5,  $sd = 11.61$ ; proso = 28.9,  $sd = 10.95$ ), and unlinked sentences (control = 34.8,  $sd = 11.66$ ; proso = 36.3,  $sd = 24.45$ ).

There was, however, more variability of scores for the unlinked sentences. This was also seen for the case study subjects in the first methods section and was the only score that reached any significance in the t-test analysis ( $p = 0.001$ ). These results can be seen below in table 5.3.

AUTISM – SPECTRUM QUESTIONNAIRE RESULTS

	Gender	Age	AQ score
DG	M	26	25
DH	M	30	14
TU	M	31	15
RR	M	42	19
MT	M	48	26
DF	M	54	18
AM	F	24	14
NG	F	30	18
SA	F	34	10
KW	F	36	11
RS	F	45	37
SM	F	49	19

Table 5.1: A list of the twelve developmental prosopagnosia subjects is presented, including each subject's gender, age, and AQ score.

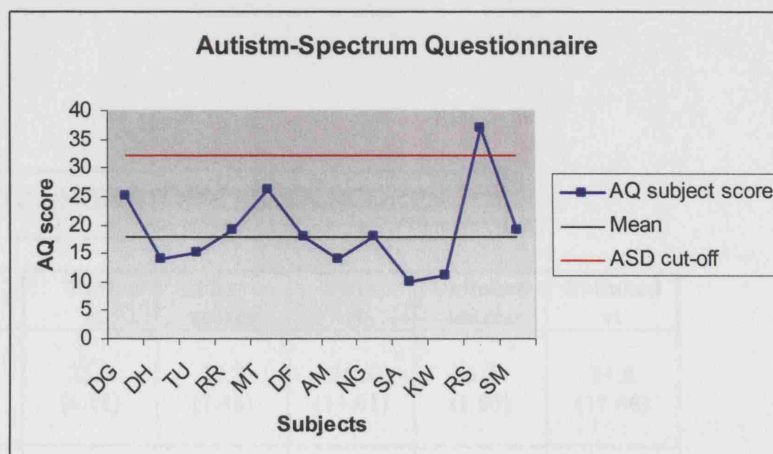


Table 5.2: Each developmental prosopagnosia subject's AQ score in comparison to the group's mean score of 18.0 and the suggested cut-off score of 32.

ANALYSIS OF AUTISM – SPECTRUM QUESTIONNAIRE MEAN SCORES

	Mean (sd)	t-test p value	
<b>AS / HFA</b>	35.8 (6.5)	0.000*	
<b>Prosopagnosia</b>	18.0 (7.5)		0.405
<b>Controls</b>	16.4 (6.3)		

Table 5.2: T-test analysis for the AQ, comparing the mean scores of the prosopagnosia group compared to the AS / HFA group and comparing the prosopagnosia group to the control group. AS / HFA and control mean scores and standard deviations used for calculation were from Baron – Cohen et al. (Baron-Cohen *et al.* 2001)(\*p<0.001)

ANALYSIS OF STRANGE STORY MEAN SCORES
---------------------------------------

	Age	ToM scores	ToM rt	Phys scores	Phys rt	Unlinked scores	Unlinked rt
<b>Controls (n = 24)</b>	36.1 (12.31)	14.3 (1.45)	25.3 (6.21)	13.8 (1.48)	35.5 (11.61)	6.7 (1.05)	34.8 (11.66)
<b>Prosopagnosics (n = 12)</b>	38.3 (9.24)	13.8 (1.48)	24.8 (11.76)	13.0 (2.04)	28.9 (10.95)	10.3 (4.86)	36.3 (24.45)
<b>t-test (p values)</b>	0.60	0.47	0.31	0.22	0.11	0.001*	0.80

Table 5.3: Data showing scores for ToM, physical, and unlinked sentence passages, as well as the reaction times for each. Also included are p values from the t-test analysis comparing the prosopagnosics to the controls. (\*p = 0.001)



## 7.0 DISCUSSION

The aim of this paper is to investigate the social cognition skills of developmental prosopagnosics in order to test the predictions of a theory proposing that a failure to develop face processing leads to the impaired social cognition seen in ASD. To do this, in-depth case studies of two developmental Prosopagnosics were investigated. The second objective was to look at a larger group of twelve prosopagnosics (ten prosopagnosics plus the two case studies from method 1) to see if the results obtained from TH and DU were exceptional, or if they coincided with a larger group.

To establish whether or not the prosopagnosic subjects have character traits that are similar to traits seen in ASD subjects, two types of examinations were used: the ADOS, module 4 and the AQ. Because of time constraints and the need for a qualified examiner to administer the ADOS, this diagnostic tool was only administered to TU and DH. Both subjects performed normally on the ADOS. DH received zero points, while TU only received one point, which was for slightly interrupting the examiner a few times. Average people have traits that are often seen in ASD subjects, but it is the summation of these traits that leads to an ASD diagnosis. It is therefore perfectly normal for TU to receive one point during his ADOS interview and not be considered as having an ASD. The examiner stated that neither subject can be diagnosed as having an ASD based on the ADOS.

The AQ was administered to all twelve developmental prosopagnosics involved in this project in an effort to ascertain the presence or absence of any ASD. In method 1, the AQ provided additional support to the conclusions of the more standardized ADOS. TU and DH scored 15 and 14, respectively. These scores are well below the ASD cut-off score of 32 as well as significantly lower than Baron-Cohen et al.'s (Baron-Cohen *et al.*

2001) AS / HFA mean group score of 35.8, providing further support that these two subjects do not have an autism spectrum disorder. When the larger group of prosopagnosics were tested, their mean score (18.0) was compared to Baron-Cohen et al.'s mean scores for an AS / HFA group (35.8) and a control group (16.4). The prosopagnosia mean was significantly lower than the AS / HFA group ( $p < 0.001$ ), but not significantly different from the control group.

When taking into account the individual scores compared to the AS / HFA scores, the only notable subjects to consider are MT, DH, and RS. Although MT and DH's scores are below 32, their scores were not significantly different from the mean AS / HFA scores. This provides support for Baron-Cohen et al.'s (Baron-Cohen *et al.* 2001) observation that "significantly more males than females in the general population show moderate levels of 'autistic' traits". Despite their higher scores, MT and DH most likely would not be diagnosed as having an autism spectrum disorder if tested by a qualified doctor or researcher, but rather they represent the average population who typically show a modest level of autistic traits. RS, on the other hand, has a score (37) that is significantly higher than the control mean and surpasses the suggested cut-off score of 32. Analysis by gender provides further evidence to support a possible diagnosis of an autism spectrum disorder because 0.0% of the control females tested by Baron-Cohen et al. had a score as high as RS, while 61.5% of the AS / HFA females had a score of 37 or higher. It would be interesting to test RS further using a more standardized ASD diagnostic tool, such as the ADOS, in addition to having a discussion with her to see how her facial recognition problems affect the way she interacts with those around her.

ToM tests were used to explore the prosopagnosics' social cognition. An animations test was used to investigate whether TU and DH applied human attributes to

inanimate objects, something that has been shown to be impaired in autistic subjects. This test had to be administered in person, so it was only given to the case studies rather than to the larger group of prosopagnosics. The second ToM test was the strange stories test, which was given to all of the prosopagnosia subjects, and tested the subjects' ability understood concepts such as double bluff, persuasion, and the intentions of eliciting sympathy.

The animations task was scored using three criteria: intentionality, appropriateness, and answer length. There was not a significant difference between the scores of TU and DH and the control group for any of the stories. The intentionality score was important because it was based on the use of mental state terms. TU and DH used more complex mental terms (i.e. terms involving deliberate action with the goal of effecting other's mental state) for the mental state stories, less complex terms (i.e. terms that more simply involved deliberate reciprocal interactions, such as chasing), and finally no mental state terms (i.e. random movement) for the random sequences. This same pattern similarly demonstrated in the control group. These two prosopagnosia subjects also did well on the appropriateness criteria, which is simply how appropriate the answer is to what the animation creator intended. They scored similarly for all three story types, mimicking the mean scores of the controls. Finally, the length reflects the number of clauses used to describe the animations. The prosopagnosia subjects' scores were not significantly different from the controls, however this is not as informative as the other two scoring criteria because autistic subjects tend to answer with a similar number of clauses compared to the controls (Castelli *et al.* 2002).

Regarding the strange stories, a significant difference was seen for the unlinked sentence scores in both the case studies (TU scoring significantly higher and DH scoring

significantly lower than the mean) and with the larger group of prosopagnosics (prosopagnosics scoring significantly higher than controls), however this set of stories may not be the most reliable control to use in comparison to the ToM stories because they rely too heavily on short term memory. The physical stories, on the other hand, provide a more similar control because they are structured like the ToM stories, yet they do not require an inference of mental state beyond the information presented in the story. TU and DH both did well on the mental stories, scoring 15 and 14 points, respectively, out of a total of 16 points. These scores, in addition to the prosopagnosia group as a whole (13.8), were not significantly different from the control group (14.1). On the physical stories, TU did fairly well with a score of 12 out of 16, however DH only scored 8 points, which was significantly lower than the control mean (13.8). Even though he had a significantly lower score for these stories, it is important to make a point that he did well on the mental state stories. This shows he has a good concept of understanding others' intentions, thoughts, and feelings, and is able to understand concepts such as empathy and white lies, something which most autistics do not have a strong comprehension. The prosopagnosia group as a whole did not show any statistically significant differences in mean from the control means for mental or physical stories or reaction times, providing additional support that prosopagnosics do not have an impairment in theory of mind.

In consideration of these results, there are limitations to address in future studies. A stronger argument could be made by using larger sample sizes of prosopagnosics, as well as compare a control and prosopagnosic group to a larger population of ASD subjects. Unfortunately, there was not enough time to do that with this study. With regards to the unlinked sentence passages in the strange stories test, there was a great deal of variability seen in the prosopagnosic subjects. One possible theory to investigate this

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

further is that the poorer results seen in some subjects could be the result of a short term memory problem in facial recognition. Is there a correlation between impairment in facial recognition tests and impairment in the unlinked sentences test? A final negative point to the unlinked sentences is that the control group only had a mean score of 6.7 (sd = 1.0). It does not make sense for a control task to be more difficult than a similar control task (physical stories) or to the test task (mental stories). There are not many highly-used ToM tests used in research, so additional social cognition tests may be useful to see what strengths and weaknesses prosopagnosics may have in comparison to ASD and non-prosopagnosic controls. Finally, it would be useful to do a comparison of scores obtained from tests given in person (strange stories, method 1) and by paper (AQ) compared to tests given online (strange stories and AQ, method 2). This is another variable which could possibly lead to score differences in the test groups.

## 8.0 CONCLUSIONS

In conclusion, the results on both the ASD and the ToM tests provide support that developmental prosopagnosics have normal social cognition skills. This population may have to make additional effort at times to interact with people in a social situation, however they have developed the important social cognition skills that are lacking in ASD subjects, such as eye contact, reciprocal social communication, empathy, facial expressions, and an interest in relating to other people. These results do not exclude the possibility that ASDs can coincide with prosopagnosia, however they disagree with Schultz's hypothesis by showing that deficits in facial memory and processing do not invariably lead to the social cognition problems that are characteristic of an autism spectrum disorder.

## 9.0 REFERENCES

- Wechsler Abbreviated Scale of Intelligence. 1999. The Psychological Corporation, a Harcourt Assessment Company, USA.
- Autistic Diagnostic Observation Schedule. 2001. Western Psychological Services, Los Angeles, CA.
- Adolphs, R., S. Baron-Cohen, and D. Tranel, 2002 Impaired recognition of social emotions following amygdala damage. *J.Cogn Neurosci.* **14**: 1264-1274.
- American Psychiatric Association Task Force on DSM-IV, 1995 *Diagnostic and statistical manual of mental disorders DSM-IV international version with ICD-10 codes*, 4<sup>th</sup> edition, American Psychiatric Association, Washington, DC.
- Anderson, A. K., and E. A. Phelps, 2001 Lesions of the human amygdala impair enhanced perception of emotionally salient events. *Nature* **411**: 305-309.
- Baron-Cohen, S., and M. K. Belmonte, 2005 Autism: a window onto the development of the social and the analytic brain. *Annu.Rev.Neurosci.* **28**: 109-126.
- Baron-Cohen, S., A. M. Leslie, and U. Frith, 1985 Does the autistic child have a "theory of mind"? *Cognition* **21**: 37-46.
- Baron-Cohen, S., H. A. Ring, E. T. Bullmore, S. Wheelwright, C. Ashwin *et al.* 2000 The amygdala theory of autism. *Neurosci.Biobehav.Rev.* **24**: 355-364.
- Baron-Cohen, S., S. Wheelwright, R. Skinner, J. Martin, and E. Clubley, 2001 The autism-spectrum quotient (AQ): evidence from Asperger syndrome/high-functioning autism, males and females, scientists and mathematicians. *J.Autism Dev.Disord.* **31**: 5-17.
- Barton, J. J., M. V. Cherkasova, R. Hefter, T. A. Cox, M. O'Connor *et al.* 2004 Are patients with social developmental disorders prosopagnosic? Perceptual heterogeneity in the Asperger and socio-emotional processing disorders. *Brain* **127**: 1706-1716.
- Beaumont, R., and P. Newcombe, 2006 Theory of mind and central coherence in adults with high-functioning autism or Asperger syndrome. *Autism* **10**: 365-382.
- Behrmann, M., and G. Avidan, 2005 Congenital prosopagnosia: face-blind from birth. *Trends Cogn Sci.* **9**: 180-187.
- Behrmann, M., G. Avidan, G. L. Leonard, R. Kimchi, B. Luna *et al.* 2006 Configural processing in autism and its relationship to face processing. *Neuropsychologia* **44**: 110-129.

- Bodamer, J., 1947 Die prosopagnosie. *Archiv fur Psychiatrie und Nervenkrankheiten* **179**: 6-53.
- Boutsen, L., and G. W. Humphreys, 2002 Face context interferes with local part processing in a prosopagnosic patient. *Neuropsychologia* **40**: 2305-2313.
- Castelli, F., C. Frith, F. Happe, and U. Frith, 2002 Autism, Asperger syndrome and brain mechanisms for the attribution of mental states to animated shapes. *Brain* **125**: 1839-1849.
- Castelli, F., F. Happe, U. Frith, and C. Frith, 2000 Movement and mind: a functional imaging study of perception and interpretation of complex intentional movement patterns. *Neuroimage*. **12**: 314-325.
- Centers for Disease Control and Prevention. Autism Information Center. <http://www.cdc.gov/ncbddd/autism/index.htm> . 2007. 23-7-2007.
- Crawford, J., and P. Garthwaite, 2007 Comparison of a single case to a control or normative sample in neuropsychology: Development of a Bayesian approach. *Cognitive Nueropsychology* **24**: 343-372.
- de Gelder, B., A. C. Bachoud-Levi, and J. D. Degos, 1998 Inversion superiority in visual agnosia may be common to a variety of orientation polarised objects besides faces. *Vision Res.* **38**: 2855-2861.
- Duchaine, B. C., 2000 Developmental prosopagnosia with normal configural processing. *Neuroreport* **11**: 79-83.
- Duchaine, B. C., Y. Galit, and K. Nakayama, 2007 No global processing deficit in the Navon task in 14 developmental prosopagnosics. *The American Journal of Psychology* **2**: 104-113.
- Duchaine, B. C., and K. Nakayama, 2006 Developmental prosopagnosia: a window to content-specific face processing. *Curr.Opin.Neurobiol.* **16**: 166-173.
- Farah, M. J., K. D. Wilson, H. M. Drain, and J. R. Tanaka, 1995 The inverted face inversion effect in prosopagnosia: evidence for mandatory, face-specific perceptual mechanisms. *Vision Res.* **35**: 2089-2093.
- Fletcher, P. C., F. Happe, U. Frith, S. C. Baker, R. J. Dolan *et al.* 1995 Other minds in the brain: a functional imaging study of "theory of mind" in story comprehension. *Cognition* **57**: 109-128.
- Fombonne, E., 2003 Epidemiological surveys of autism and other pervasive developmental disorders: an update. *J.Autism Dev.Disord.* **33**: 365-382.



- Gaigg, S. B., and D. M. Bowler, 2007 Differential fear conditioning in Asperger's syndrome: implications for an amygdala theory of autism. *Neuropsychologia* **45**: 2125-2134.
- Hadjikhani, N., R. M. Joseph, J. Snyder, C. F. Chabris, J. Clark *et al.* 2004 Activation of the fusiform gyrus when individuals with autism spectrum disorder view faces. *Neuroimage*. **22**: 1141-1150.
- Hadjikhani, N., R. M. Joseph, J. Snyder, and H. Tager-Flusberg, 2007 Abnormal activation of the social brain during face perception in autism. *Hum. Brain Mapp.* **28**: 441-449.
- Hale, C. M., and H. Tager-Flusberg, 2005 Social communication in children with autism: the relationship between theory of mind and discourse development. *Autism* **9**: 157-178.
- Happe, F. G., 1994 An advanced test of theory of mind: understanding of story characters' thoughts and feelings by able autistic, mentally handicapped, and normal children and adults. *J. Autism Dev. Disord.* **24**: 129-154.
- Harris, A. M., and G. K. Aguirre, 2007 Prosopagnosia. *Curr. Biol.* **17**: R7-R8.
- Heider, F., and M. Simmel, 1944 An experimental study of apparent behavior. *The American Journal of Psychology* **57**: 243-259.
- Hobson, R. P., J. Ouston, and A. Lee, 1988 What's in a face? The case of autism. *British Journal of Psychology* **79**: 441-453.
- Iidaka, T., M. Omori, T. Murata, H. Kosaka, Y. Yonekura *et al.* 2001 Neural interaction of the amygdala with the prefrontal and temporal cortices in the processing of facial expressions as revealed by fMRI. *J. Cogn Neurosci.* **13**: 1035-1047.
- Jolliffe, T., and S. Baron-Cohen, 1999 The Strange Stories Test: a replication with high-functioning adults with autism or Asperger syndrome. *J. Autism Dev. Disord.* **29**: 395-406.
- Lahaie, A., L. Mottron, M. Arguin, C. Berthiaume, B. Jemel *et al.* 2006 Face perception in high-functioning autistic adults: evidence for superior processing of face parts, not for a configural face-processing deficit. *Neuropsychology.* **20**: 30-41.
- Langdell, T., 1978 Recognition of faces: an approach to the study of autism. *J. Child Psychol. Psychiatry* **19**: 255-268.
- Leder, H., and V. Bruce, 2000 When inverted faces are recognized: the role of configural information in face recognition. *Q. J. Exp. Psychol. A* **53**: 513-536.
- Levine, D. N., and R. Calvanio, 1989 Prosopagnosia: a defect in visual configural processing. *Brain Cogn* **10**: 149-170.

- Lord C., Rutter M., DiLavore P.C. & Risi S. Autism Diagnostic Observation Schedule. 2002. Los Angeles, CA, Western Psychological Services.
- Morris, J. S., K. J. Friston, C. Buchel, C. D. Frith, A. W. Young *et al.* 1998 A neuromodulatory role for the human amygdala in processing emotional facial expressions. *Brain* **121** ( Pt 1): 47-57.
- Njiokiktjien, C., A. Verschoor, S. L. de, C. Huyser, h. V. Op, V *et al.* 2001 Disordered recognition of facial identity and emotions in three Asperger type autists. *Eur.Child Adolesc.Psychiatry* **10**: 79-90.
- Nunn, J. A., P. Postma, and R. Pearson, 2001 Developmental prosopagnosia: should it be taken at face value? *Neurocase*. **7**: 15-27.
- Pelphrey, K. A., N. J. Sasson, J. S. Reznick, G. Paul, B. D. Goldman *et al.* 2002 Visual scanning of faces in autism. *J.Autism Dev.Disord.* **32**: 249-261.
- Pessoa, L., M. McKenna, E. Gutierrez, and L. G. Ungerleider, 2002 Neural processing of emotional faces requires attention. *Proc.Natl.Acad.Sci.U.S.A* **99**: 11458-11463.
- Pickles, A., P. Bolton, H. Macdonald, A. Bailey, C. A. Le *et al.* 1995 Latent-class analysis of recurrence risks for complex phenotypes with selection and measurement error: a twin and family history study of autism. *Am.J.Hum.Genet.* **57**: 717-726.
- Quaglino, A., G. B. Borelli, S. S. Della, and A. W. Young, 2003 Quaglino's 1867 case of prosopagnosia. *Cortex* **39**: 533-540.
- Rondan, C., and C. Deruelle, 2007 Global and configural visual processing in adults with autism and Asperger syndrome. *Res.Dev.Disabil.* **28**: 197-206.
- Schultz, R. T., 2005 Developmental deficits in social perception in autism: the role of the amygdala and fusiform face area. *Int.J.Dev.Neurosci.* **23**: 125-141.
- Schultz, R. T., I. Gauthier, A. Klin, R. K. Fulbright, A. W. Anderson *et al.* 2000 Abnormal ventral temporal cortical activity during face discrimination among individuals with autism and Asperger syndrome. *Arch.Gen.Psychiatry* **57**: 331-340.
- Schwarzer, G., S. Huber, M. Gruter, T. Gruter, C. Gross *et al.* 2006 Gaze behaviour in hereditary prosopagnosia. *Psychol.Res.*
- Sorger, B., R. Goebel, C. Schiltz, and B. Rossion, 2007 Understanding the functional neuroanatomy of acquired prosopagnosia. *Neuroimage*. **35**: 836-852.

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

- Stark, L., and S. R. Ellis, 1981 Scanpath revisited: Cognitive models of direct active looking, pp. 193-226 in *Gaze behavior: Cognition and visual perception*, edited by D. F. Fisher, R. A. Monty, and J. W. Senders. Lawrence Erlbaum, Hillsdale, NJ.
- Sweeten, T. L., D. J. Posey, A. Shekhar, and C. J. McDougle, 2002 The amygdala and related structures in the pathophysiology of autism. *Pharmacol. Biochem. Behav.* **71**: 449-455.
- Teunisse, J. P., and B. deGelder, 2003 Face processing in adolescents with autistic disorder: the inversion and composite effects. *Brain Cogn* **52**: 285-294.

### APPENDIX 1 – Autism-Quotient Questionnaire

1. I prefer to do things with others rather than on my own.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
2. I prefer to do things the same way over and over again.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
3. If I try to imagine something, I find it very easy to create a picture in my head.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
4. I frequently get so strongly absorbed in one thing that I lose sight of other things.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
5. I often notice small sounds when others do not.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
6. I usually notice car number plates or similar strings of information.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
7. Other people frequently tell me that what I've said is impolite, even though I think it is polite.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
8. When I'm reading a story, I can easily imagine what the characters might look like.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
9. I am fascinated by dates.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
10. In a social group, I can easily keep track of several different people's conversations.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
11. I find social situations easy.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
12. I tend to notice details that others do not.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
13. I would rather go to a library than a party.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
14. I find making up stories easy.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
15. I find myself drawn more strongly to people than to things.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
16. I tend to have very strong interests, which I get upset about if I can't pursue.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

17.I enjoy social chit-chat.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
18.When I talk, it isn't always easy for others to get a word in edgeways.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
19.I am fascinated by numbers.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
20.When I'm reading a story, I find it difficult to work out the characters' intentions.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
21.I don't particularly enjoy reading fiction.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
22.I find it hard to make new friends.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
23.I notice patterns in things all the time.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
24.I would rather go to the theatre than a museum.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
25.It does not upset me if my daily routine is disturbed.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
26.I frequently find that I don't know how to keep a conversation going.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
27.I find it easy to "read between the lines" when someone is talking to me.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
28.I usually concentrate more on the whole pictures, rather than the small details.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
29.I am not very good at remembering phone numbers.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
30.I don't usually notice small changes in a situation, or a person's appearance.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
31.I know how to tell if someone listening to me is getting bored.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
32.I find it easy to do more than one thing at once.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
33.When I talk on the phone, I'm not sure when it's my turn to speak.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
34.I enjoy doing things spontaneously.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
35.I am often the last to understand the point of a joke.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree

36.I find it easy to work out what someone is thinking or feeling just by looking at their face.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
37.If there is an interruption, I can switch back to what I was doing very quickly.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
38.I am good at social chit-chat.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
39.People often tell me that I keep going on and on about the same thing.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
40.When I was young, I used to enjoy playing games involving pretending with other children.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
41.I like to collect information about categories of things (e.g. types of car, types of bird, types of train, types of plant, etc.)	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
42.I find it difficult to imagine what it would be like to be someone else.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
43.I like to plan any activities I participate in carefully.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
44.I enjoy social occasions.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
45.I find it difficult to work out people's intentions.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
46.New situations make me anxious.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
47.I enjoy meeting new people.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
48.I am a good diplomat.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
49.I am not very good at remembering people's date of birth.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree
50.I find it very easy to play games with children that involve pretending.	Definitely agree	Slightly agree	Slightly disagree	Definitely disagree

**Scoring:** A score of 1 point was give when an answer of “definitely agree” or “slightly agree” were given for questions 1, 2, 4, 5, 6, 7, 9, 12, 13, 16, 18, 19, 20, 21, 22, 23, 26, 33, 35, 39, 41, 42, 43, 45, and 46. And answer of “definitely disagree” or “slightly disagree” were given for questions 3, 8, 10, 11, 14, 15, 17, 24, 25, 27, 28, 29, 30, 31, 32, 34, 36, 37, 38, 40, 44, 47, 48, 49, and 50. All other answer received zero points when circled.

## APPENDIX 2 – Strange Stories Test

**Instructions:** “There are three conditions in this test, consisting of three sorts of materials: social stories having to do with mental states, physical stories, having to do with physical behaviors, and jumbled passages composed of unconnected sentences. On each page, you will find a short story to read. After you’ve read and understood the story, I want you to turn the page. There is a question after each story and I’d like you to tell me the answer to this question. I don’t want you to look back at the story, so make sure you’ve understood it before turning over for the question. For the jumbled passages, the question will relate to one sentence in the passage”.

### Mental State (Social) Stories (0 to 2)

Simon is a big liar. Simon’s brother, Jim, knows that Simon never tells the truth! Now yesterday Simon stole Jim’s ping-pong paddle, and Jim knows Simon has hidden it somewhere, though he can’t find it. He’s very cross. So he finds Simon and he says. “Where is my ping-pong paddle? You must have hidden it either in the cupboard or under your bed, because I’ve looked everywhere else. Where is it, in the cupboard or under your bed?” Simon tells him the paddle is under his bed.

*Q: Why will Jim look the cupboard for the paddle?*

2 = reference to Jim knowing Simon lies

1 = reference to facts (that’s where it really is) or Simon hiding it without reference to implication of lying

0 = reference to general non-specific information (because he looked everywhere else)

During the war, the Red army captures a member of the Blue army. They want him to tell them where his army’s tanks are; they know they are either by the sea or in the mountains. They know that the prisoner will not want to tell them, he will want to save his army, and so he will certainly lie to them. The prisoner is very brave and very clever, he will not let them find his tanks. The tanks are really in the mountains. Now when the other side ask him where his tanks are, he says, “They are in the mountains”.

*Q: Why did the prisoner say that?*

2 = reference to fact that other army will not believe and hence look in other place, reference to prisoner’s realization that that’s what they’ll do, or reference to double bluff

1 = reference to outcome (to save his army’s tanks) or to mislead them

0 = reference to motivation that misses the point of double bluff (he was scared)

Brian is always hungry. Today at school it is his favourite meal – sausage and beans. He is a very greedy boy, and he would like to have more sausages than anybody else, even though his mother will have made him a lovely meal when he gets home! But everyone is allowed two sausages and no more. When it is Brian’s turn to be served, he says, “Oh, please can I have four sausages, because I won’t be having any dinner when I get home!”

*Q: Why does Brian say this?*

2 = reference to fact that he’s trying to elicit sympathy, being deceptive

- 1 = reference to his state (greedy), outcome (to get more sausages), or factual
- 0 = reference to a motivation that misses the point of sympathy elicitation / deception, or factually incorrect

Jill wanted to buy a kitten, so she went to see Mrs. Smith, who had lots of kittens she didn't want. Now, Mrs. Smith loved the kittens and she wouldn't do anything to harm them, though, she couldn't keep them herself. When Jill visited she wasn't sure she wanted one of Mrs. Smith's kittens, since they were all males and she had wanted a female. But Mrs. Smith said, "If no one buys the kittens I'll just have to drown them!"

*Q: Why did Mrs. Smith say that?*

- 2 = reference to persuasion, manipulating feelings, trying to induce guilt / pity
- 1 = reference to outcome (to sell them or get rid of them in a way which implies not drowning) or simple motivation (to make Jill feel sad)
- 0 = reference to general knowledge or dilemma without realization that the statement was not true (she's a horrible woman)

One day Aunt Jane came to visit Peter. Now Peter love his aunt very much, but today she is wearing a new hat; a new hat which Peter things is very ugly indeed. Peter things his aunt looks silly in it, and much nicer in her old hat. But with Aunt Jane asks Peter, "Hot do you like my new hat?", Peter says, "Oh, it's very nice".

*Q: Why does he say that?*

- 2 = reference to white lie or wanting to spare her feelings; some implication that this is for aunt's benefit rather than just for his, desire to avoid rudeness or insult
- 1 = reference to trait (he's a nice boy) or relationship (he likes his aunt); purely motivational (so she won't shout at him) with no reference to aunt's thoughts or feelings; incomplete explanation (he's lying, he's pretending)
- 0 = reference to irrelevant or incorrect facts / feelings (he likes the hat, he wants to trick her)

Helen waited all year for Christmas, because she knew at Christmas she could ask her parents for a rabbit. Helen wanted a rabbit more than anything in the world. At last Christmas Day arrived, and Helen ran to unwrap the big box her parents had given her. She felt sure it would contain a little rabbit in a cage. But when she opened it, with all the family standing round, she found her present was just a boring old set of encyclopedias, which Helen did not want at all! Still, when Helen's parents asked her how she liked her Christmas present, she said, "It's lovely, thank you. It's just what I wanted".

*Q: Why did she say this?*

- 2 = reference to white lie or wanting to spare their feelings; some implication that this is for her parent's benefit rather than just for her; desire to avoid rudeness or insult
- 1 = reference to trait (he's a nice girl) or relationship (she likes her parents); purely motivational (so they won't shout at her) with no reference to parent's thoughts or feelings; incomplete explanation (she's lying, she's pretending)
- 0 = reference to irrelevant or incorrect facts / feelings (she likes the present, she wants to trick them)



## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

Late one night Old Mrs. Peabody is walking home. She doesn't like walking home alone in the dark because she is always afraid that someone will attack her and rob her. She really is a very nervous person! Suddenly, out of the shadows comes a man. He wants to ask Mrs. Peabody what time it is, so he walks towards her. When Mrs. Peabody sees the man coming towards her, she starts to tremble and says, "Take my purse, just don't hurt me please!"

*Q: Why did she say that?*

- 2 = reference to her belief that he was going to mug her or her ignorance of his real intention
- 1 = reference to her trait (she's nervous) or state (she's scared) or intention (so he wouldn't hurt her) without suggestion that the fear was unnecessary
- 0 = factually incorrect / irrelevant answers; reference to the man actually intending to attack her

A burglar who has just robbed a shop is making his getaway. As he is running home, a policeman on his beat sees him drop his glove. He doesn't know the man is a burglar, he just wants to tell him he dropped his glove. But when the policeman shouts out to the burglar, "Hey, you! Stop!", the burglar turns round, sees the policeman and give himself up. He puts his hands up and admits that he did the break-in at the local shop.

*Q: Why did the burglar do that?*

- 2 = reference to belief that policeman knew that he's burgled the shop
- 1 = reference to something factually correct in the story
- 0 = factually incorrect / irrelevant answers

### Human Physical State Stories (0 to 2)

Two enemy powers have been at war for a very long time. Each army has won several battles, but now the outcome could go either way. The forces are equally matched. However, the Blue army is stronger than the Yellow army in foot soldiers and artillery. But the Yellow army is stronger than the Blue army in air power. On the day of the final battle, which will decide the outcome of the war, there is heavy fog over the mountains where the fighting is about to occur. Low-lying clouds hang above the soldiers. By the end of the day, the Blue army has won.

*Q: Why did the Blue army win?*

- 2 = reference to both weather conditions and either relative ground superiority or inability of other army's planes to be useful in fog (names of armies unimportant)
- 1 = reference either or weather or relative superiority on ground versus air (because it was foggy); nothing about why weather makes it especially difficult for tanks or nothing about planes being affected more than tanks; reference to fog to justify incorrect response (the aeroplanes won because the fog meant they could hide from the tanks)
- 0 = reference to irrelevant or incorrect information (they won because they had better planes); justifications for why tanks are better than planes

A burglar is about to break into a jeweler's shop. He skillfully picks the lock on the shop door. Carefully he steps over the electronic detector beam. If he breaks this beam it will set off the alarm. Quietly he opens the door of the store-room and sees the gems glittering. As he reaches out, however, he steps on something soft. He hears a screech and something small and furry runs out past him, towards the shop door. Immediately the alarm sounds.

*Q: Why did the alarm go off?*

- 2 = reference to animal which the burglar disturbed setting off alarm by crossing beam (type of animal unimportant)
- 1 = reference to burglar setting off alarm (he was startled by the animal so crossed the beam); reference to animal setting off alarm without explaining it crossed the beam (he trod on a cat and it set off the alarm)
- 0 = reference to irrelevant or incorrect factors (the animal's screech set off the alarm); alternative reasons for alarm going off (a security camera saw him and set the alarm off)

Old Mrs. Robinson is very frail. One day she slips on her icy door step and falls on her side. She gets up right away, although she feels quite bruised and shaken. The next day her leg feels very stiff and she can scarcely walk. She makes her way to the doctors. As soon as the doctor hears about the fall, and sees her swollen side, he says, "Go immediately to the hospital". At the hospital they take an X-ray.

*Q: Why did they take an X-ray?*

- 2 = reference to possibility that she has fractured / broken her hip / leg; reference to wanting to know or trying to find out ( i.e. 'it was broken' is not enough); must refer to fact that X-rays are for broken things or bones (to see if there's any damage to the bone)
- 1 = reference to general aim (to see what's wrong, because of her fall she might have damaged something) or factually correct (it's bruised and stiff)
- 0 = reference to irrelevant (because she fell) or incorrect factors (that's what doctors do) or to X-rays being cures themselves (to mend her leg)

John is going shopping. He buys a nice new desk lamp for his study. He needs a light bulb for his new lamp. He goes from the furniture department to the electrical department. In the electrical department he finds that there are two brands of light bulbs of the right kind. Everbright light bulbs cost less in single packs than Literite bulbs. However, only Literite bulbs come in multi-packs of six. John buys he multi-pack, even though he only needs one bulb.

*Q: Why does Job buy the Literite bulbs?*

- 2 = reference to saving money by buying the multipack
- 1 = reference to convenience of having more bulbs, or future need for more than one bulb; no mention of saving money
- 0 = reference to irrelevant or incorrect factors (Literite bulbs are brighter)

Mrs. Simpson, the librarian, receives a special book which she has to catalogue and find an appropriate place for. She has to decide which section to file it under. The library is very big, and has different sections on many different subjects. The new book is about

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

plants and their medical uses, and is heavily illustrated. However, Mrs. Simpson does not put it on the shelf with the rest of the books on botany. Neither does she put it with the books on medicine. Instead, she carefully takes it into a separate room. In this room all the books are kept in special cases, and the temperature is kept constant.

*Q: Why did she do this?*

2 = reference to avoiding damage to the book because it is special

1 = reference to the fact that the book is special; no reference to why it might be kept in a special case

0 = reference to irrelevant or incorrect factors (she doesn't know where else to put it)

Henry is preparing for a big dinner party. He is famous for his excellent mayonnaise. He has bought lots of fresh eggs. The recipe says, "Carefully separate the yolks of the six eggs and add oil very gradually". He has already bought easily enough dessert to feed everyone. However, he now looks up the recipe for meringues. Henry will not waste anything.

*Q: Why does Henry make meringues?*

2 = reference to Henry not liking to waste anything and therefore using up the left-over egg whites

1 = reference either to not wasting anything or to having left-over egg whites

0 = reference to irrelevant or incorrect factors (he's having a party)

Paul is very rich, and today he is going to buy an expensive new car. He is considering whether to make a single payment, or whether to spread the cost over the year. If he pays in monthly installments, the dealer will charge five percent interest on the loan. His bank currently gives him eight percent interest on the money in his account. Even though he has easily enough money to pay the full amount, he decides to pay by monthly installments.

*Q: Why does he do that?*

2 = reference to getting more interest from the bank than he's pay on the loan and therefore to saving money

1 = reference to saving money; no explanation why he'd save money

0 = reference to irrelevant or incorrect factors ( he doesn't have enough money)

Sarah is very far-sighted. She has only one pair of glasses, which she keeps losing. Today she has lost her glasses again and needs to find them. She had them yesterday evening when she looked up the television programs. She must have left them somewhere that she has been today. She asks Ted to find her glasses. She tells him that today she went to her regular early morning exercise class, then to the post office, and last to the flower shop. Ted goes straight to the post office.

*Q: Why is the post office the most likely place to look?*

2 = reference to post office being place she would most likely use her glasses (to read/write/look at stamps, etc); may talk about either putting glasses on or taking them off

1 = plausible alternative reason for being in post office (there are lots of people there, you might have posted them by mistake, people take lost things there)

0 = reference to irrelevant or incorrect factors (that was the last place she we you can buy glasses at the post office, she needed the glasses to hear better); general factors, non-specific to post offices)

Unlinked Sentence Passages (0 to 2)

The two countries had been at war. A housewife is about to enter the super-market. Today he is going to buy an expensive new stereo. Mrs. Brown, the post-mistress, receives a special parcel. Mrs. Pearson wouldn't harm a fly. Mary's birthday is in February. Late one evening the old man was watching television.

*Q: When is Mary's birthday?*

2 = February

0 = anything else

Young Simon is very robust. She sees that Fred cannot play. Jeremy is always laughing. Ruth sees her uncle very often, but today he has gone to Brazil. Richard is packing up to go away. Today, at college, it is Jim's worst lecture – statistical mathematics. She has only one dollar left, which she must keep for her bus fare. He buys a bright tie to go with his new shirt.

*Q: How many dollars does she have left?*

2 = One

0 = anything else

Simon takes the special butter from the refrigerator. Each boxer has won several fights. He skillfully picks out the imperfect items. They are either in Boston or in New York. She has to cut the grass and find somewhere to plant the bay tree. The conductor sees that the cellist has broken a string. Tracy took the bus to the station.

*Q: Where did Tracy take the bus?*

2 = the station

1 = anything that seems part way there (eg. The bus station)

0 = anything else

The four brothers stood aside to make room for their sister, Stella. Gill repeated the experiment several times. The name of the airport has changed. Louise uncorked a little bottle of oil. The two children had to abandon their daily walk. She took a suite in a grand hotel. It was already twenty years since the operation.

*Q: Who abandoned their daily walks?*

2 = the two children

1 = anything that seems part way there (eg. The children)

0 = anything else

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

One day Uncle Simon came to visit Alex. The first part of the performance had come to an end. He put away the letter and stuck his hands in his pockets. She was still holding her umbrella. The cats ran back to the boy. Flora came into the middle of the square. The little island had a high rocky shoreline.

*Q: Where did Flora go?*

2 = the middle of the square

1 = anything that seems part way there (eg. The square)

0 = anything else

At the edge of the road a little grass was growing. He reaches out to find the light switch. A sailor who has just left his ship is walking to the town. She has to decide where to keep the pasta. At last daylight came, and Tommy got out of bed to open his presents. Jim knows all about investing money as he works in a large bank. They exchanged a few brief words about the weather.

*Q: Why did Tommy get out of bed?*

2 = to open his presents

1 = anything that seems part way there (eg. Because daylight came / it was morning)

0 = anything else

She is always saying that someone will eventually find the treasure. Everyone is allowed two visits and no more. At the psychiatry department they were interviewing the new nurse. Jim will with the first race of the meeting. She has taken all the children to visit the zoo today. Simon's uncle is wearing a new suit. The same phrase of twenty three notes recurred throughout.

*Q: What will Jim win?*

2 = the first race of the meeting / the first race

1 = anything that seems part way there (eg. The race)

0 = anything else

He needs a new engine for his old car. The prize is an immediate lump sum of \$20,000 tax-free. Japan is stronger than Italy in economic terms. The mother is very brave and long suffering. The new book is about statistics and experimental design, and contains many graphs. The front room contained a little bird in a cage. Although Jim is only twenty one years old, he has an income of \$20,000 per year. There are not many people this evening in the large rectangular dining room.

*Q: Who is brave and long suffering?*

2 = the mother

1 = anything else

### APPENDIX 3 – Animations Test

**Instructions:** “Each animation lasts approximately 40 seconds. The sequences are similar to one another, two triangles are moving about, but different in their content. The triangles act as characters performing different movement, for example, dancing, drifting, or courting each other. There are different types of content: In some animations the behavior of both triangles will appear disconnected from each other. They just move about, with random movement. By contrast, other animations will show the two triangles moving about doing something together, interacting. Their actions are somehow connected to each other, for example, they are imitating each other, or one is feeding the other. Still other animations show the two triangles doing something more complex together, as if they are taking into account their reciprocal feelings and thoughts. By just watching them you will probably imagine they are interacting, for example, courting each other. In this experiment, there is no ‘right’ or ‘wrong’ answer. After each cartoon is over, I will ask you what you think the triangles were doing, whether they were randomly moving about, or whether they were doing something more specific” (Castelli *et al.* 2000).

#### *Intentionality Score (0 to 5)*

0 = the agent acts with no intention and no interaction, randomly, non-deliberate

- bouncing off
- moving around
- floating

1 = the agent acts with a purpose, a goal, with no interaction with another agent, deliberate

- ice-skating
- swimming
- escaping
- walking

2 = the agent acts with a purpose with another agent, the actions of the two agents are parallel in time

- fighting
- following

3 = the agent not only interacts with another agent but acts in response to the other’s action

- chasing
- restraining
- guarding

## Are Social Cognition Deficits Present In Developmental Prosopagnosics?

---

4 = the agent acts in response to a mental state

- arguing
- wanting
- encouraging
- mocking
- mimicking
- teasing
- being happy
- being friendly

5 = the agent acts with the goal of affecting or manipulating the other agent's mental states

- pretending
- deceiving
- coaxing
- surprising
- convincing

### *Appropriateness (0 – 2)*

0 = “don’t know” answers

1 = descriptions that focus solely on a minor aspect of the sequence

2 = partial description of the sequence, description is related to the sequence, but imprecise or incomplete

3 = spot-on description of the story or the actions represented, it may be concise just capturing gist as well as discursive

### *Length (0-4)*

0 = no response

1 = one clause

2 = two clauses

3 = three clauses

4 = four or more clauses