Developmental Prosopagnosia

Kevin T. Blake, Ph.D., P.L.C.
Tucson, Arizona
Cross Country Education
Brentwood, Tennessee
Case Study of a Dyslexic

• Male college student
• 21 years old
• 3.8 GPA in Electrical Engineering
• 145 Full Scale IQ
• Excellent social and conversational skills
• Dresses and acts age appropriate
• Mildly depressed (Dx: Dysthymic Disorder)
• Severe Reading Disorder/Dyslexia
Case Study of a Dyslexic

- “I don’t recognize my own face”!
- *Developmental Prosopagnosia*
Problems
Remembering Faces

– **Prosopagnosia**: Inability to recognize faces, even one’s own face (p. 1168).


– **Joaachim Bodamer, M.D. 1947**: German soldiers with brain injuries who could no longer see faces. Coined term “Prosopon” meaning face + “agnosia” meaning nonrecognition from Greek.

Subtypes of Prosopagnosia

• Acquired Prosopagnosia: Caused by insult to the brain; what Bodamer wrote about in 1947.
• Developmental Prosopagnosia: “...characterized by severely impaired face recognition. Individuals with this disorder, which runs in families, have no history of brain damage and intact early visual systems” (p. 166).

What is a Developmental Disorder?

• A disorder characterized by a significant delay in the rate a normal human trait develops in an individual.

• It takes the individual longer to develop this trait than their age peers.

Subtypes of Prosopagnosia

• Possible Associated Conditions:
  – Problems with recognition of facial expression of emotion
  – Problems with gender of face discrimination
  – Problems with age of face discrimination
  – Problems with **TOPOGRAPHAGNOSIA**: difficulty with personal navigation; getting lost easily
  – Asperger’s Disorder

Subtypes of Prosopagnosia

• Possible Associated Conditions:
  – Central Auditory Processing Disorder (CAPD):
    “The inability to understand spoken language in a meaningful way in the absence of what is commonly considered a hearing loss” (Sineps and Hunter, 1997).


Prosopagnosia

• Remembering Faces:
  – This is an important ability for survival.
    • It lets you know “friends and foes”.
    • It helps you maintain relationships.
    • It helps you remember the social status of others.

Developmental Prosopagnosia

• Affects 2 to 3 percent of the population
• That equates to 6,000,000 Americans!
• Those affected often know something is wrong, but they don’t know exactly what.

Symptoms of Prosopagnosia

• Extreme difficulty recognizing faces. Even with a person who is well known by the sufferer (i.e., a parent, spouse, best friend, etc.).

• Appears aloof/arrogant, does not respond to people they “know” when they see them.

• Often complain they cannot follow movies or TV shows because they cannot remember the identity of characters.

• They tend to recognize people by hair, gait, clothing, voice, context, or other information.

Additional Symptoms of Prosopagnosia Found in Children

• It may take months to recognize their classmates.
• School transition may be a problem.
• Extreme separation anxiety and stranger wariness
• Changes in people’s appearance (i.e., new glasses, new hair style, etc.) may be a problem.
• Feelings of frustration, isolation and embarrassment

Face Perception

• The right Fusiform Gyrus typically does not respond to objects.
• This area reorganizes faces into wholes.
• The fusiform gyrus helps to differentiate between visually similar stimuli.
• Greebles-novices treat them as objects while experts treat them in a holistic manner.

Face Perception

• “Our results show that a man with severe prosopagnosia performed normally throughout the standard greeble training procedure. These findings indicate face recognition and greeble recognition rely on separate mechanisms” (Duchaine, et.al., August, 2004).

Developmental Prosopagnosia

• “The hereditary type of prosopagnosia has an autosomal dominant type of inheritance. This means that men and women are affected in equal numbers. In our experience women are more willing to talk about their face recognition problems, though” (Thomas Grueter, M.D.).

• If one parent has Prosopagnosia their child has a 50% chance of having it.

Whose at Risk for Prosopagnosia?

Those with Learning Disorders, AD/HD, Nonverbal Learning Disorders and Asperger’s Disorder


• There is little data regarding the co-occurrence of developmental prosopagnosia and LD, AD/HD and NVLD.

• Most people he has evaluated with Asperger’s Disorder have face processing problems.

Duchaine, B. (June 23, 2010). Personal Communication.
Face Perception in ASD

• The Fusiform Face Area (FFA) responds much more to faces than to other objects.

• Nine different labs have found that those with Autism Spectrum Disorders have a hypoactivation of the FFA when viewing faces.

• Developmental Prosopagnosia and Developmental Agnosia are separate disorders.


Facial Expressions

• Remembering Expressions:
  – The non-disabled are “pre-wired” to find the human face and voice the most important stimuli in the world.
  – Those with Asperger’s Disorder (AD) don’t look at the eyes they look at the mouth. Differentiated those with AD from non-disabled 100% of the time.


“Thus, these data argue for the role of the FFA-amygdala system in social cognition more generally, and retrieval of specific social knowledge about what constitutes a friendly social interaction, or not. Collectively these data suggest the the amygdala-FFA system and its failure to strongly activate during face perception tasks points to a causal mechanism involved in autism...” (p. 137).

How to Assess Developmental Prosopagnosia

• Cambridge Face Memory Test
• Test My Face Recognition - Internet test


www.faceblind.org/people/duchaine06neuropsychologia.pdf#search=%22Cambridge%20Face%20Memory%20Test%22.

Test My Face Recognition (From web site): www.icn.ud.ac.uk/facetests/
Treatment of Prosopagnosia

• “Prosopagnosics cannot be cured, but they can and do learn ways to recognize people” (p. 70).

Treatment of Prosopagnosia: “Are you my Mother?”

• Encourage the person to look at people’s faces when socializing.
• Introduce new people slowly and emphasize their characteristics: “Say hi to Billy with the red hair and freckles”.
• Have children meet teachers long before school starts and have the child meet with them often.
• Have teachers keep their appearance “stable”.
• Play introduction games.
• Post photos of teachers, friends, parents on wall.

Computer Programs to Treat Prosopagnosia

• “Let’s Face It!” – Face Recognition Program and workbook for children with Autism Spectrum Disorders (University of Victoria Brain and Cognition Lab & the Yale Child Study Center)

• Teaches facial recognition and emotion recognition in 20 hours!

• It is **FREE!**

Mnemonic Techniques to Remember Faces


• [www.jerrylucas.com](http://www.jerrylucas.com)
Facial Expression Training & Autism

• “Even when people with autism spectrum disorders can figure out what someone’s eyes or face conveys, they do so in a different way than everyone else, which may be less efficient or take more time” (p. 62).

• The non-disabled use the temporal lobe and fusiform gyrus to decode facial expressions.

Facial Expression Training & Autism

• Looking at pictures of eyes and deciphering the emotion they conveyed activated the non-disabled amygdalas and frontal lobes.

• Those with Asperger’s used the frontal lobes far less and did not activate the amygdala. They used other areas of the brain not designed for such tasks.

• Those with Asperger’s may use voice, touch, etc. to recognize others, not their face.

Prosopagnosia of Facial Expressions
“Face perception can be subdivided into two general types – recognition of person identity via the structures of the face, and recognition of internal affective state on the shape of individual features and changes in their relative distance from one another during the expression” (p. 128).

Recognizing Emotional Facial Expressions

• Emotional Facial Expression Recognition:
  – “Does this mean we come into the world expecting to see human faces and ready to respond with our own prewired facial expressions? Yes!” (Ratey, 2001, p. 300).

Decoding Skill and Facial Expression

• Positive emotions are the easiest to decode.
• Negative emotions are the most difficult.
• Poor interpreters of facial expression have less social acceptance and poorer adjustment.

Decoding Skill and Facial Expression

• Most and Greenbank (2000) stated LD children are less accurate in identifying emotional expressions than non-LD children.

• Brown (2001) indicated AD/HD children are less accurate in facial expression identification than their non-AD/HD peers.

• Attwood (2007) stated those with Asperger’s Disorder have great difficulty decoding faces.


Dyslexia and Facial Expressions

- Non-LD children did better than Verbal LD, NVLD and children with both Verbal LD and NVLD in interpreting facial expressions.
- Non-LD group did better than Verbal LD only with surprise.
- Non-LD group did better than NVLD and NVLD/Verbal LD groups for 4 facial expressions.
- Older subjects did better identifying fear and disgust than younger subjects.
- No difference between girls and boys in ability (Ref. #1)
Dyslexia and Facial Expressions

• Non-disabled children did better in recognizing facial expressions than those with Verbal LD and those with NVLD. (Ref. #2)

• Those with Verbal LD did better than those with NVLD. (Non-LD>Verbal LD>NVLD) (Ref. #2)

• Teens (12-15 years) with Verbal LD were worse than those with NVLD, and those who where Non-LD in recognizing facial expressions. (Ref. #3)

• There was no difference between the Non-LD and NVLD groups. (Ref. #3)
Dyslexia and Facial Expressions: References


"Importantly, findings of this study show that emotion processing difficulties in children with ADHD extend beyond facial emotion and also effect the recognition of emotions on the basis of contextual information. Our data thus indicate that children with ADHD have an overall emotion-processing deficit” (p. 111).

"Children with ADHD exhibited a general deficit in decoding emotional facial expressions, with specific deficit in identifying anger and sadness. Self-rating of the task difficulty revealed lack of awareness of decoding errors in the ADHD group as compared with control subjects. Within the ADHD group, there was a significant correlation between interpersonal problems and emotional facial expression decoding impairment, which was more marked for anger expressions” (p. 93).

“Boys with ADHD may show poorer task performance because of general cognitive factors, but also show selective problems in matching facial expressions to situations” (p. 398).

AD/HD and Facial Expressions

“Attention deficits in boys with ADHD seemed to account for their difficulty in recognizing facial expressions of emotion. Effective treatment for attention deficits is expected to have a beneficial effect on facial emotion recognition in boys with ADHD”. (p. 323)

AD/HD and Facial Expressions

Research evidence indicates that emotional facial recognition difficulties in people with ADHD may be related to biological impairment of the medial prefrontal cortex and amygdala.

AD/HD and Facial Expressions

• “These results suggest that affect recognition abilities may be impaired in adults with ADHD and affect abilities are more adversely affected by inattentive than hyperactive-impulsive symptoms” (p. 1)

• No difference between non-disabled and those with Combined Type AD/HD in facial expression recognition.

• Those with Inattentive AD/HD made more errors in recognizing expressions of fear.

AD/HD and Facial Expressions

Research indicates there may be altered neurological functioning of identifying affect anger and fear recognition in those with AD/HD that appear to be reduced by methylphenidate (Ritalin). Those with ADHD had left amygdala overactivation when viewing neutral faces.

Prosopagnosia and NVLD (Nonverbal Learning Disorders)

“Hence, it appears that children with NLD have a specific deficit on immediate memory for faces. This facial memory deficit may be linked to a deficit in right hemisphere functioning which has already been implicated in facial processing and may also be linked with other disorders (e.g., autism spectrum disorder) in which similar facial processing deficits have been documented.” (p. 1-2)

“Although not part of current diagnostic criteria, much evidence suggests that persons with ASD have marked deficits in face perception.” (p. 127)

Assessment for Face Perception

*Simon Baron-Cohen’s Tests:*

- Faces Test
- Eyes Test (Adult)
- Eyes Test (Child)
- Cambridge Mindreading (CAM) Face-Voice Battery
- Empathy Quotient (EQ) (Adult)
- Empathy/Systemizing (EQ-SQ) (Child)
- And many others...

Downloadable from:

[www.autismresearchcentre.com/tests/default.asp](http://www.autismresearchcentre.com/tests/default.asp)
Evaluating Nonverbal Behavior

• Diagnostic Analysis of Nonverbal Behavior 2 (DANVA2)
  – Adult faces and voices
  – Child faces and voices
  – African American faces and voices
  – Postures

Available from: Steven Nowicki, Ph.D., Emory University –
www.snowik@emory.edu
FACE READING ASSESSMENT

• **Comprehensive Affect Testing System (CATS)**

“This ensemble of tests enables clinical psychologists, neuropsychologists, neurologists, educators, speech therapists and other related disciplines to assess dysfunctional processing of affect expressed by the human face and voice” (p. 1 of 4).


[www.psychologysoftware.com/CATS.html](http://www.psychologysoftware.com/CATS.html)
Treating Problems Reading Facial Expressions

• FACIAL EXPRESSIONS CAN BE TAUGHT!


Computer Programs to Treat Prosopagnosia


  “Harry Potter” teaches facial expressions.


  “Thomas the Tank-Engine” teaches faces.
Computer Programs to Treat Prosopagnosia

• “Gaining Face”: www.ccoder.com/GainingFace

• Paul Ekman, Ph.D. ("Lie to Me"/SPOT – Surveying Passengers by Observational Techniques) CD ROMS:
  Micro Expression Training Tool (METT)
  Subtle Expression Training Tool (SETT)
Repeate presentations of METT & SETT to those with Autism Spectrum Disorders
Available from: www.paulekman.com
Computer Programs to Treat Prosopagnosia

• “Let’s Face It!” – Face Recognition Program and workbook for children with Autism Spectrum Disorders (University of Victoria Brain and Cognition Lab & the Yale Child Study Center)

• Teaches facial recognition and emotion recognition in 20 hours!

• It is **FREE!**

Treating Problems Making & Reading Facial Expressions

- Cognitive Affective Training-Faces and Feeling Words: [www.CAT-kit.com](http://www.CAT-kit.com)


Teaching Facial Affect Recognition

- Social Skills training by Mental Health Professionals and Speech-Language Pathologists (female teacher with small children)*
- Try an “Emotional Seeing Eye Dog” (Grandin, 1995)


Wearable Prosthetic

“We describe a novel wearable device that perceives and reports on social-emotional information in real-time human interaction. Using a wearable camera, combined with machine perception algorithms, the system records and analyzes the facial expressions and head movements of the person with whom the wearer is interacting. We propose the application of the social-emotional prosthetic to assist the growing number of individuals diagnosed with Autism...
Wearable Prosthetic

...Spectrum Disorder (ASD) in perceiving communication in a natural rather than a structured environment, bootstrapping their ability to learn and develop in social settings…” (p. 1).


Emotional Seeing Eye Dogs

- Dogs separated from wolves about 135,000 years ago.
- Dogs lived with humans for 100,000 years; even before we were “modern humans” (Homo Habilis).
- Dog and humans co-evolved.
- Humans learned to think and act like dogs.
- Dogs allowed humans to hunt big game while they acted as guards and lookouts. Humans did more planning and organization activities.
- 14,000 years ago humans domesticated dogs.
- Homo Sapien Neantathalensis did not have dogs; they are extinct.
- In the past 100,000 years dogs brains shrank by 10 to 30%; mostly in their forebrains while humans’ brains shrank by 10%; mostly in the midbrain sensory and smell areas.
- Dogs have a symbiotic relationship with humans and have a genetic predisposition to understand human emotions.

Dogs may have a rudimentary mirror neuron system!


Emotional Seeing Eye Dogs

• 4Paws For Ability
  253 Dayton Avenue
  Xenia, OH 45385

• Training Center:
  937-374-0385

• Website:
  www.4pawsforability.org
Possible Treatment for Emotional Working Memory Problems

• Stimulant Medication?
  – Lessens Hyperactivity and Impulsivity in AD/HD, Combined Type Individuals
  – Hundreds of Double Blind Studies to Support

**Possible Alternative Medicine Treatment for Working Memory Problems in AD/HD**

- **Working Memory Training:**
  - Torkel Klingberg, M.D., Ph.D.
  - Karolinska Institute- Stockholm, Sweden
  - CogMed software company (RM Program)
  - AD/HD deficient in visual spatial working memory (WM). Gets worse with age.
  - **MAY** help relieve visual spatial WM difficulties and reading comprehension in Combined Type AD/HD.
  - **More Research is needed!**

**References**


Ingersoll, B. (October 26, 2006). Complementary Treatments for AD/HD. Paper Presented at the 18th Annual CHADD International Conference, Chicago, IL.

Problems Making the Appropriate Facial Expression to Match How One Feels and What is Appropriate to The Situation

- Unmedicated AD/HD, Combined Type people have difficulty making facial expressions to match how they feel. They tend to “over-emote” their facial expressions. (Kuehle, et.al., 2002).

- Attwood’s (2007) story of the boy with Asperger’s Disorder who saw his mother crying and asked, “What face do I make?” (p. 134)


AD/HD and Making Facial Expressions

- Regarding facial expressions in children and adults with AD/HD Kuhle, Hoch, Rautzenberg and Jansen (2001) concluded, “Altogether, ... the facial expressions, are uncontrolled and jerky and are often wrongly dimensioned in time and space.” (p. 6)

Facial Expressions and AD/HD

- AD/HD children smile abruptly.
- There is little or no transition between emotional states.
- Sometimes their facial expression bleeds over into the next emotional state.
- Expression of emotion often appears exaggerated. The quality of expression can be limited due to this.
- Even body movements are jerky and uncontrolled.

AD/HD and Making Facial Expressions

• AD/HD childrens’ eyes drift away from those they are in conversation with.
• This usually interrupts the flow and their comprehension of the conversation.
• Often parents feel rejected by AD/HD children when they do this.

Possible Treatment of Problems with Facial Expression and AD/HD

- Optimal dosing of a stimulant medication causes a significant reduction in visual attention loss.
- Facial expressions will become smooth and variable.
- Too high a dose can cause a return of the symptoms.
- Can properly ID 80% of the AD/HD children with video procedure.

“Prosopagnosia is a cognitive disorder characterized by a severe deficit in face recognition, which cannot be attributed to lower-level visual problems, higher-level semantic impairments or cognitive alterations such as mental confusion or amnesia. Prosopagnosics can normally recognize that a particular visual stimulus is a face, but they cannot discriminate between different faces, and hence cannot recognize faces of familiar people...”
“...This impairment is severe, and includes not only the faces of close friends and acquaintances, but also family members, siblings, spouses and, in some cases, even their own face. However, prosopagnosics can identify people using alternative cues to recognition, such as hairstyle, clothing, voice or gait. Importantly, this indicates that prosopagnosia is essentially a disorder of visual perception, and general semantic knowledge about familiar people remains intact and accessible from other modalities” (p. 59).

Types of Prosopagnosia

There are two types of prosopagnosia:

- Acquired Prosopagnosia
- Developmental Prosopagnosia

Where The Brain Processes Faces

- It appears the N170 (A.K.A.: M170) waves in the brain are related to the encoding of facial processing.
- The occipital facial area seems to be involved in very early visual face analysis that encoded facial parts.
- The fusiform face area processes facial identity.
- The superior temporal sulcus processes facial expressions and eye gaze direction.

“Ekman and colleagues proposed the existence of six basic emotional expressions that are common to all cultures: anger, disgust, fear, happiness, sadness and surprise...Further work suggests that although these expressions might be universal, we interpret the expressions displayed by individuals in our in-group more accurately than those...This effect may be explained by non-verbal accents, which are subtle differences in the expression of emotions between groups” (p. 45-46).

Children and Facial Recognition

The face processing system continues to develop through adolescence and until about age 30.

Children and Facial Facial Recognition

“...investigations suggest that adult-like face recognition performance is in fact reached by five years age, but lower levels of attention, concentration and memory, and a greater susceptibility to demand characteristics, explain why children perform at a poorer level in face recognition experiments” (p. 121).

Face processing ability differs greatly across the population and those with prosopagnosia may be at the lower end of the continuum.

Depression & Facial Expressions

“In depression, the negative attentional bias and inability to label positive emotions as happy may be explained by increased limbic activity in response to negative expressions but decreased activity for positive emotional expressions” (p. 97).

“Hence, both behavioural and eye movement findings suggest that individuals with social anxiety disorder might be particularly impaired in processing facial expressions of anger” (p. 96).

Types of Facial Processing Disorders

“...the primary evidence that supported the proposed independence of identity and expression processing came from a double dissociation between two neurological disorders. One half of this dissociation comes from individuals with prosopagnosia, who cannot recognize people from their face yet can still recognize different emotional expressions...The other half comes from reports of patients who are impaired at recognizing particular emotional expressions, despite intact identification abilities” (p. 100).

ASD Vs Developmental Prosopagnosia

“Thus, although investigation of individuals who suffer from face-processing impairments alongside SDDs (socio-developmental disorder, sic.) is also of interest in informing our knowledge about other factors that may influence face recognition ability, the bulk of the available evidence supports the viewpoint that DP (developmental prosopagnosia, sic.) should be considered as an independent condition” (p. 127).

Developmental Prosopagnosia

- Developmental prosopagnosia, “...can result from, impairments to different mechanisms in different cases” (p. 132).
- There is great variation of emotional facial expression processing ability across the population.

ASD & Face Processing

“A range of face-processing deficits can present in ASD. Sometimes, individuals have problems recognizing facial identity, gaze direction, gender, expression and lip reading...Most importantly, nearly all individuals with ASD have problems interpreting emotional expression. For some individuals with ASD, the impairment in recognizing emotional expressions seems only to affect certain expressions, most notably fear” (p. 144).

Much neuroimaging evidence suggests that individuals with ASD display abnormalities in both the fusiform gyrus and the amygdala in response to facial stimuli, and some authors have theorized that individuals with ASD might process faces using object-rather than face-specific areas of the brain” (p. 145).

“...not all individuals with ASD are impaired at face processing and developmental prosopagnosics do not all show ASD” (p. 145).

Questions for Face Processing Assessment

- Has the person ever suffered a brain injury?
- Do they have ASD or a related disorder
- Does anyone in their biological family suffer from ASD or a related disorder
- Have they suffered from an uncorrected vision problem for several months?
- Does any biological relative have DP?
- Do they have a neuropsychiatric disorder?
- Are there any memory, cognitive or perceptual difficulties?

Face Processing Assessments

- Benton Facial Matching Test (BFRT)
- Cambridge Face Perception Test (CFPT)
- Glasgow Face Matching Test (GFMT)
- Cambridge Face Memory Test (CFMT)

Powell et al.’s Acquired Prosopagnosia Treatment Program

- **Semantic Association Training**: Client is given additional information about the person.
- **Caricature Training**: Client is trained to recognize a list of caricatures of peoples’ faces and associate them with those they meet.
- **Part Recognition Training**: Client learns to recognize parts of people’s faces.

“At the group level, the three experimental procedures were of similar efficacy, and associated cognitive deficits did not predict which technique would be most beneficial to individual patients; however, there was limited power to detect such associations. Interestingly, a pure prosopagnosic patient who was tested separately showed benefit only from the part recognition technique” (p. 182).

“Thus, although providing promising evidence that face-processing deficits may be treatable in at least acquired cases of prosopagnosia, much work is still needed to determine the optimum intervention strategy with the most long-lasting effects” (p. 191).

Training and Developmental Prosopagnosia

“We designed a task that required discriminating faces by their spatial configuration and, after extensive training, prosopagnosic MZ significantly improved at face identification. Event-related potential results revealed that although the N170 was not selective for faces before training, its selectivity after training was normal. fMRI demonstrated increased functional connectivity between ventral occipital temporal face-selective regions (right occipital face area and right fusiform face area) that accompanied improvement in face recognition. Several other regions showed fMRI activity changes with training; the majority of these regions increased connectivity with face-selective regions. Together, the neural mechanisms associated with face recognition improvements involved strengthening early face-selective mechanisms and increased coordination between face-selective and nonselective regions, particularly in the right hemisphere” (p. 1790)

“Hence, this study provides exciting evidence that face recognition cannot only be improved on a behavioral level in DP (developmental prosopagnosia, sic.), but that intervention can modify the neural mechanisms underpinning the face processing system. This suggests long-lasting improvements might be achievable, at least in developmental prosopagnosia” (p. 193).

“Overall, the authors concluded that these results suggest that children with ASD also exhibit plasticity in their behavioral responses to faces, and a short-term intervention programme can produce measurable improvements in face recognition skills in children with this disorder” (p. 194).

“Attempts to improve both facial identity and facial expression recognition deficits in ASD using computerized intervention programmes have met with much success” (p. 196).

Oxytocin and Prosopagnosia

“Very recently research has indicated an alternative manner in which face-processing skills can temporarily be improved: Using Intranasal inhalation of the hormone oxytocin. Oxytocin is naturally produced and broken down within the human body, and is involved in regulation of basic social and reproductive behaviors, such as cohabitation, gestation and breastfeeding…”
Oxytocin and Prosopagnosia

“...Recently, synthetic forms of oxytocin have been manufactured that can be nasally inhaled, and these sprays have been used in studies that have examined whether oxytocin can improve face-processing abilities in both health and impaired patients” (p. 195).