Winter 2013 Prosopagnosia Research Center

Face to Face

Prosopagnosia Research & Community

Happy 2013! This winter brings the sixth edition of the *Face to Face* newsletter, and its third year! We are excited as ever to continue to bring you new research and personal stories from the community. Please contact us at any time should you have thoughts or feedback to share!

-Prosopagnosia Research Center (faceblind@faceblind.org)

Deficits Associated with Developmental Prosopagnosia

by Dr. Brad Duchaine

People with developmental prosopagnosia (DP) often have deficits with other abilities that appear to be unrelated to face recognition. For example, many report problems recognizing objects or finding their way in environments they've been in many times. In medicine, when other disorders are present in addition to a primary diagnosis, doctors often refer to the other disorders as *comorbidities*, and this term is now commonly used in psychology and neuroscience. What causes cognitive comorbidities, and what are the comorbidities in developmental prosopagnosia?

Because knowledge about DP is still quite limited, we've looked to research on developmental dyslexia for hints. Like DP, people with developmental dyslexia have no history of brain damage yet they show severe deficits with reading. The leading theories of dyslexia propose that reading problems result primarily from impairments with phonology (the processing of speech sounds), but it has long been recognized that many dyslexics have comorbid conditions affecting a variety of cognitive, sensory, or motor abilities. For example, people with dyslexia are much more likely to be diagnosed with specific language impairment or motor-coordination problems. Autopsy studies of dyslexic brains have shown that dyslexics have cortical anomalies in the left perisylvian cortex where phonological information is represented, but many also have anomalies in other brain regions as well, including brain regions not involved in phonological processing or reading.

To account for the phonological problems in dyslexia and its comorbid conditions, French neuroscientist Franck Ramus and his colleagues have proposed that dyslexia results from neural migration errors. According to their model, these errors occur while the brain is developing prenatally, and the errors lead to cortical anomalies concentrated in the brain regions critical for phonological processing (left perisylvian cortex). For example, small clusters of neurons (approximately 50-100) may end their migration in the wrong layer of cortex, which leads to disorganization in that bit of cortex. Not surprisingly, when neural migration errors are present in an individual, they are not usually limited to one brain area, but rather occur in a number of regions. When multiple brain areas are affected in someone with dyslexia, then that person will have other impairments. The type of impairment will depend on which other brain regions are…

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Prosopagnosia and the Department of Veterans Affairs

by Robert D. Pryor

Like most people, I never thought about face recognition, or my having a particular ability that others didn't have. As a Special Forces soldier in 1969 I received two penetrating head wounds while in the Republic of Vietnam. The injuries cost me about 20% of the brain on the right side. The foreign objects passed more than half way through my head. It was at that moment I lost the ability that I had so taken for granted – my ability to recognize faces.

With four head wounds, 26 other major wounds and about 200 minor wounds, testing for prosopagnosia was certainly not a priority for the VA during my recovery. The greater concerns were regaining the use of my extremities, dealing with hearing loss, and surgeries. After I was medically retired from the Army in 1969 the VA did not test me for prosopagnosia. The condition is not very well understood presently, never mind forty years ago – medical science was woefully deficient when it came to understanding prosopagnosia.

I was having trouble recognizing people, even my own mother, but I never thought to mention the problem to any physician that was treating me. In 1982, I participated in The Vietnam Head Injury Study. It was a collaborative effort of the Department of Defense, the VA and the National Institute of Health. It was during the intake interview that I first mentioned to anyone that I had trouble recognizing people. As part of my…

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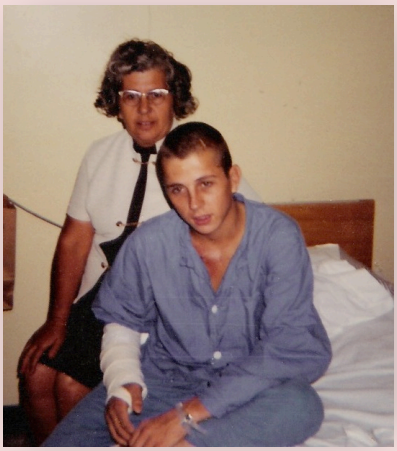
Prosopagnosia and the Department…, continued from pg 1.



Robert and his mother at the hospital in 1969.

…participation in that study I was tested for, and diagnosed with, prosopagnosia.

I had been rated as 100% permanently and totally disabled by the VA after getting out of the Army, so there was no need to apply for increased benefits. But still, out of curiosity, I checked the VA's Schedule for Rating Disabilities to see how it treated prosopagnosia. The U.S. Department of Veterans Affairs did not recognize prosopagnosia as a disability under the law. A somewhat shortened version of how this happened is that the original Schedule for Rating Disabilities was written in about 1944 and the term prosopagnosia was not commonly in use at that time.

It takes extensive work at the highest levels of government, the legislative and the executive branches, to amend the Schedule for Rating Disabilities. It has been amended a few times over the years for conditions affecting large populations, but let's face it; prosopagnosics with confirmed diagnoses and well documented etiology hardly qualifies as a large population. I dropped the idea of having it added to my long list of service-connected disabilities. It would take a lot of work and bring no financial benefit to me. I had been a veterans' benefits counselor, so I was familiar with the regulations and how to investigate disabilities. My experience in this field gave me the knowledge on how to get the VA to rate a condition as service connected, even if it was not listed in the regulations.

Two decades later an extensive coalition was involved with wars in Iraq and Afghanistan. Due to the frequent use of roadside bombs and improvised explosive devices, Traumatic Brain Injuries (TBI) became quite common. National Public Radio did an interview with a recently disabled soldier suffering from TBI. In that interview, the soldier mentioned that he could no longer recognize faces. I felt guilty for not previously trying to get the VA to recognize prosopagnosia as a service connected condition. The law needed to change; and with my experience, I would be able to get them to change it. I felt that I owed it to the new generation of disabled veterans to force the VA to recognize prosopagnosia as a ratable disability.

In September of 2007, 38 years after acquiring prosopagnosia, I formally filed a claim for prosopagnosia as a service connected disability. Because it was not a ratable condition, I filed using two seldom-used VA regulations. I asked for an analogous rating and an extraschedular rating. That gave the VA two options for recognizing the condition. The VA uses an analogous rating when a condition is not listed in the schedule, but when there is a condition listed with a similar set of symptoms. An extraschedular rating is used when a condition is not listed, but it can be shown that it is indeed disabling. In this case it is up to the veteran to also prove exactly how the condition is disabling.

There were still a few other problems to overcome in presenting my claim. I had to prove that my prosopagnosia is the result of the head wounds I received in the Republic of Vietnam. I *then* had to prove that the condition manifested itself to a compensable degree within one year of my separation from the military. Lastly, I had to prove that I had not received any other brain traumas in the 38 years since I got shot.

The real problem is that to guarantee success on a claim, a veteran should file their claim at the time of separation from service, or at least within a year of separation. I was not diagnosed with prosopagnosia until I had been out for 13 years. The longer a veteran waits to file a claim, the lower his or her chances of success. At 38 years post-retirement, my chances were slim at best.

The last problem was that there was no one with whom I could collaborate on this endeavor that understood both VA regulations and prosopagnosia. I asked and received assistance from fellow prosopagnosiacs on the faceblind e-mail group, and asked a few questions of Dr. Ken Nakayama to help me with my quest.

It was a long-shot; however I presented the VA with what I considered to be a very thorough case. I expected the VA to turn me down citing that the condition was not a disability under the law. To my surprise, the VA did not just blow me off by turning my claim down outright. However, they only rated it as 10% disabling. For the first time since the…

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Prosopagnosia and the Department…, continued from pg 2.

…Schedule for Rating Disabilities was implemented in 1945, the VA, by this time called the Department of Veterans Affairs, had formally recognized prosopagnosia as a service connected disability. The battle victory went to me, but the war was far from over.

They rated my condition at 10% because they stated my symptoms were subjective. In other words, they had to just take my word for it when it came to the severity of the symptoms. My wife Julia has developmental prosopagnosia, and as far as I know, there is no accurate way to determine its severity other than taking the patient's word for it when they respond to a face blind test. But in my case, their calling my symptoms subjective only angered me.

The VA had failed to do any meaningful research into my condition so I filed a formal appeal. I sent off a scathing statement explaining that studies indicate a connection between the fusiform gyrus and facial recognition. I advised them about the prosopagnosia web site from Harvard University and University College London where they could learn about current research. I also acknowledged that the VA had few people on staff that understood this condition as well as Doctors Nakayama and Duchaine. I pointed out that a great portion of my fusiform gyrus had been blown away. I explained that they had a number of CT scans on me verifying this, therefore my prosopagnosia certainly was not subjective.

Meanwhile, unbeknownst to me, the VA was busy writing and implementing regulations that would cover all symptoms of TBI, including prosopagnosia. As part of my appeal, I agreed to have my claim presented to a Decision Review Officer (DRO). It was while my claim was with the DRO that the new regulations for TBI were implemented. The DRO increased my rating for prosopagnosia to 70% disabling. As a point of information, assigned disability ratings represent average earning impairment. A 100% disability rating represents profound earning impairment. But there are ratings that far exceed the basic 100% disability level. For example, a paraplegic receives disability benefits far in excess of the basic 100% level. Someone permanently bed-ridden, or a quadriplegic, receives benefits at about twice the benefits paid to a 100% disabled veteran. Additionally, there are a number of ancillary benefits granted in the case of these more severely disabled veterans.

Robert, his wife Julia, and their daughter.

The only problem I have with the VA and prosopagnosia is that on the VA Advance Directive: Durable Power of Attorney for Health Care and Living Will, VA form 10-0137, they list a number of circumstances under which a reasonably prudent individual might not want life-sustaining medical treatment. One of the six situations described states, "If I have permanent, severe brain damage that makes me unable to recognize my family or friends." That is exactly what I have. It's called prosopagnosia. It's nice to know that our government believes that prosopagnosia is a fate worse than death -- yet ratable as only 70% disabling. My struggle with VA continues on that one.

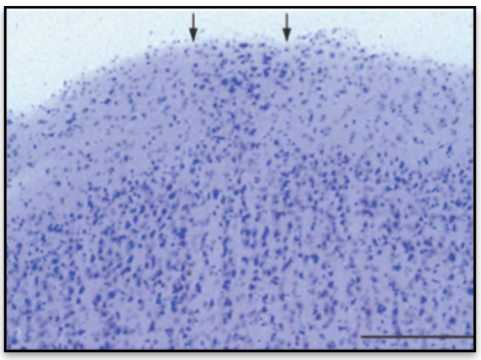
While the VA now recognizes prosopagnosia as a potentially service connected disability under the law, there is still work to be done. The poorly worded statement on the VA form 10-0137 shows how little is understood about prosopagnosia; not only by the general population, but by medical and legal professionals. I, and others with whom I've shared this story, hope that as a result of my efforts real changes will take place. Through those changes, others with prosopagnosia could receive the understanding and benefits to which they should be entitled. ∇

We encourage advocacy, awareness, and any actions like these around the world, and are very happy to provide support and information. If you would like to discuss your ideas, write to us at sarahc@wjh.harvard.edu telling us about them and yourself.

Deficits Associated with…, continued from pg 1.

…disrupted. Support for this neural migration account comes from genetic studies of dyslexia: Four genes have been linked to dyslexia, and all four genes contribute to neural migration.

Ramus's model of developmental dyslexia has clear relevance for DP and other developmental conditions characterized by relatively selective cognitive deficits. Focal cortical anomalies resulting from neural migration errors in temporal and occipital regions that contribute to face processing would be expected to lead to deficits like those seen in DP. Whereas dyslexics have reduced grey matter density in the perisylvian cortex, studies from our lab and others found that DPs have reductions in areas important for face processing such as fusiform gyrus, inferior temporal gyrus, and superior temporal sulcus. More circumscribed cortical anomalies would likely cause specific deficits like those seen in people who only have problems with face recognition. More extended anomalies, however, would also affect abilities like object recognition and navigation mediated by neighboring brain areas. Remote brain areas and the abilities supported by them may be affected in some individuals too.



Cortical anomaly. This image shows an ectopia in a dyslexic subject. Neurons appear as black spots in the micrograph. In the region between the two arrows, neurons are present in unusually high concentrations in the external layer of the cortex due to neural migration errors.

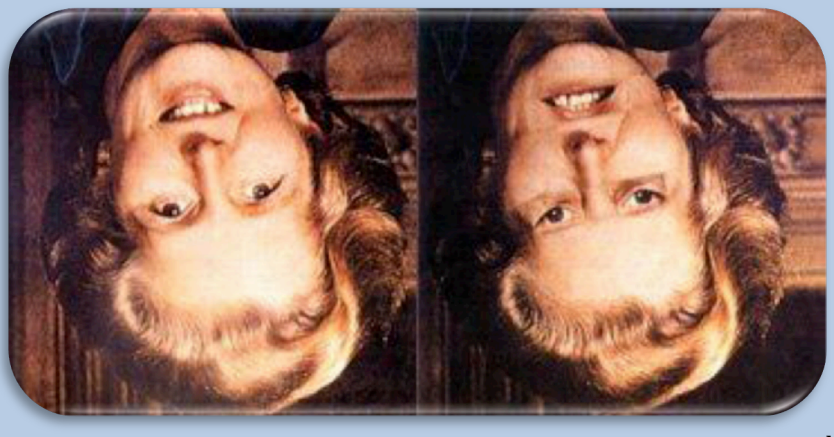
Presently, little is known about comorbidities in DP. Impairments affecting object recognition are certainly more common in DPs than in the control population, and 14% of DPs who responded to our online follow-up questionnaire (n = 3393) reported severe navigational problems. Interestingly, object recognition and navigation deficits are also common in acquired prosopagnosia. Responses to our questionnaire indicate that rates of dyslexia (7.4%) and developmental coordination disorder (5.9%) in DP are comparable to estimates for the general population. This survey data provides tentative support for the possibility that non-face deficits will be elevated for tasks that depend on regions close to the areas involved in face processing, but will diminish or be seen at normal rates for abilities dependent on more remote neural regions. We hope we'll be able to carry out studies in coming years investigating comorbidities in developmental prosopagnosia, because we believe understanding them will help us better understand the nature of DP. ∇

The Classics

In this section, we summarize a classic paper in face recognition research. If you would like access to the original article, or want to know more about follow-up studies, please e-mail lab member Dr. Lucia Garrido at garridolucia@gmail.com.

Thompson (1980). Margaret Thatcher: a new illusion. *Perception, 9*, 483-484.

This article reported a new illusion using a picture of the then UK Prime Minister Margaret Thatcher. If you look at the two pictures below, they look quite normal, right? Do you notice something strange with any of the pictures?



Now, if you rotate this page by 180 degrees (if you are using Adobe Reader, you can go to the menu View, and then Rotate View), you may find that the pictures actually look very different. While, initially, with the inverted pictures, the facial expressions both appeared normal, now the upright faces reveal that one of them has very grotesque features. But if you rotate again to see the two inverted pictures, it may still be very hard to perceive the distortions in the eyes and mouth of the face on the right. This study demonstrated that the ability to perceive faces is significantly disturbed by inverting them, and researchers are still working on understanding the specific cognitive mechanisms that only allow for clearly seeing the distortions when the faces are upright. ∇