Dear Reader,

It’s hard to believe but the holidays are upon us once again. Feels like it was summer just yesterday. We hope this holiday season finds you and your family well.

We continue to get used to our new home in building AB2, and it’s been fun to see some familiar faces coming in for a visit. We are still working on the interior decorating—just like a home, these things always seem to take longer than you think!

We are very proud of our own Sharon Willock, who has been training for the American Stroke Association Marathon in Honolulu, Hawaii this month. She has raised over $3,000 for stroke research and education. Way to go, Sharon!

Our other big news is that we have a new Center website. Please visit us at: www.ebire.org/CARD. You will find some important links and other information that we hope you will find helpful.

We hope to see you at our annual holiday party which is coming up December 22nd. We’d love to see you if you can make it.

Wonderful wishes to you and your family this holiday season!

Nina F. Dronkers, Ph.D.
Director, Center for Aphasia and Related Disorders
In past issues, we have discussed the various forms of aphasia that can occur following stroke. Here, we discuss a different form of aphasia known as primary progressive aphasia or PPA. Unlike aphasia following stroke, PPA is a language disorder that is very gradual and appears to be caused by a degenerative disorder, similar to Alzheimer’s disease. The most common early symptoms of PPA are word-finding problems during speaking or writing. From there, the disorder progresses so that the person has more and more difficulty expressing themselves and/or understanding what is being said to them.

**Symptoms of PPA**

PPA is a newly recognized disorder, and so there is still a lot of controversy as to exactly what characterizes the disorder, as well as what causes it. According to Mesulam (2003), the diagnostic criteria for PPA are as follows:

1. A gradual onset of the disorder, which gets progressively worse over time. Impairments may include deficits in word finding, naming, and word comprehension.

2. Problems with memory, visual-spatial abilities, and movement are absent during the first two years of the illness.

3. Difficulties with performing simple calculations and pantomiming gestures may be present in the first two years.

4. Language remains the most impaired of all functions, even if other areas such as memory also start to deteriorate over time.

5. Other causes of aphasia, such as stroke, must first be ruled out.

**Forms of PPA**

There are different forms of PPA, just as there are different forms of aphasia following stroke. Non-fluent PPA primarily affects the patient’s speech and their ability to express themselves. For example, they may stumble over difficult, multi-syllabic words, such as “administration” or “Christmas.” At the same time, their ability to understand what is being said to them is relatively intact.

Another form of the disorder is fluent PPA, also referred to as semantic dementia. In this form of the disorder, the patient has fluent speech but may leave out important content words or make mistakes in pronouncing words. Also, with this form of the disorder, the patient may have difficulty understanding what is being said to them, especially if it is too complex. For example, patients with fluent PPA may have difficulty following conversations, especially in large groups with many speakers.

**PPA vs. Alzheimer’s Disease**

Unlike other progressive disorders such as Alzheimer’s disease, patients with PPA do not have difficulty with memory, reasoning, or visual perception. Often they can continue to participate in most of their normal daily activities. Although their memory may get a little worse over time, patients with PPA have the most difficulty with their language. This is a very important feature that neurologists consider when trying to distinguish PPA from Alzheimer’s disease.

**Brain Changes in PPA**

There is a lot of new research aimed at understanding PPA and its causes. Our lab, in conjunction with Drs. Miller and Gorno-Tempini at UCSF, is trying to better understand this disorder (see p. 4). Using MRI, we have found that patients with PPA have atrophy (loss of cells) in differ-
ent parts of the brain, depending on the form of PPA they have. In patients with non-fluent PPA, the left, lower part of the frontal lobe and the insula are most typically atrophied. In patients with fluent PPA, the left, inferior and anterior portions of the temporal lobe are atrophied (Gorno-Tempini et al., 2004). The brain areas affected by PPA are sometimes different than those affected by stroke. As a result, the pattern of language deficits is also somewhat distinct.

What Causes PPA?
The exact cause of PPA is unknown. It has not been studied sufficiently yet to determine whether there is a genetic or even perhaps an environmental component to the disorder. Research in these areas is currently underway. Both genetic and environmental factors have been identified in other progressive disorders such as Alzheimer’s disease, and it is likely that research into PPA will also reveal distinct risk factors.

Treatment for PPA
Patients with PPA may benefit from therapy with a speech-language pathologist. The goal of the therapy is to improve communication, whether it be through speaking, writing, gesturing, or using alternative forms of communication such as “talking” computers. The focus of the therapy is individualized, depending on each patient’s particular needs.

Currently, there are no medications prescribed specifically for PPA, although patients often take medications to manage depression or anxiety that may occur at some point during the course of the disease. Some medications have proven successful at slowing the progression of Alzheimer’s disease, and it is hoped that such medications will also be discovered for use with PPA patients.

How is PPA diagnosed?

Typically, a neurologist will diagnose PPA based on the following:
1) a thorough neurologic exam and medical history
2) an MRI or CT to help determine what areas of the brain have been affected
3) neuropsychological and speech/language evaluations to assess cognitive and linguistic skills and
4) a two-year history of language problems in the absence of memory and other cognitive decline.

PPA Resources
If you wish to find out more about primary progressive aphasia, please refer to the following resources:

“The goal of the therapy is to improve communication, whether it be through speaking, writing, gesturing...”

Primary Progressive Aphasia Newsletter
Cognitive Neurology and Alzheimer’s Disease Center
320 East Superior St.
Searle 11-450
Chicago, IL 60611-3008
Phone: 312-908-9339
http: www.brain.nwu.edu

National Aphasia Association
156 Fifth Avenue, Ste 707
New York, NY 10010
Phone: 1-800-922-4622
http: www.aphasia.org/NAAppa.html

Rare Dementia Registry
This is a registry made up of individuals who have been diagnosed with a rare dementing illness and their primary caregivers. Its purpose is to serve as a telephone support network for patients and their families. The database is confidential and the service is provided by:
The Alzheimer’s Association
Greater Phoenix Chapter
1028 East McDowell Rd.
Phoenix, AZ 85006-2622
Phone: 1-800-392-0550 or 602-528-0550
http: www.alzaz.org
Cross-Linguistic Studies in Aphasia

This study evaluates word and sentence processing in aphasic patients in three different languages: English, Italian, and Mandarin Chinese. Experiments involving aphasic patients are complemented by parallel experiments using functional magnetic resonance imaging (fMRI) in the same three languages. Cross-linguistic comparisons of normal and abnormal language functioning across structurally-different languages allow us to determine which brain structures supporting language are universal and which are language-specific. This study was begun by Elizabeth Bates at the University of California, San Diego, and has produced cutting-edge research regarding the brain areas involved in speaking different languages.

Behavioral Neuroscience and Stroke: Language

This study investigates the brain areas involved in specific components of language. Lesion analysis, functional MRI, and event-related potentials assess the functional roles of particular brain areas in language, the time course with which they are recruited into the language system, the ways in which these areas interact with each other, and how they interact with other areas that support the skills necessary for language. This work is in collaboration with our colleagues at the University of California, Berkeley, and the University of California, Davis.

Primary Progressive Aphasia

The aim of this study is to investigate the brain basis of a progressive form of aphasia that is caused by degeneration of different brain areas over time. Unlike aphasia caused by a stroke, progressive aphasia leads to a unique set of language and cognitive deficits in patients with this disorder. This project also involves using neuroimaging techniques such as MRI to help us understand the changes that occur in this degenerative disorder. This work is done in collaboration with colleagues at the University of California, San Francisco and is headed by Dr. Maria Luisa Gorno-Tempini.

Voxel-based Lesion Symptom Mapping

A new type of neuroimaging analysis we have developed (Voxel-based Lesion Symptom Mapping or VLSM) permits us to analyze stroke lesion data similar to ways used in functional neuroimaging so that the relative contributions of different brain regions to a given cognitive or linguistic function can be analyzed (see Dronkers, Wilkins, Van Valin, Redfern, & Jaeger, 2004; Baldo et al., in press). This technique is allowing us to visualize networks of brain regions responsible for various abilities such as sentence comprehension and problem solving.

Visit our new website for more information: www.ebire.org/CARD
Stroke Support Group
Annual Holiday Party!

**When:** Wed., December 22nd, 12:30-3:00 p.m.

**Where:** Room E8A&B on 1st Floor of AB21 (tallest building at the VA)

**What to bring:** a dish or drink to share, if you are able, and a wrapped ornament to exchange.

Questions: call Juliana (925) 372-4649
Across
1. I saw Mommy _____ Santa Claus.
4. Many people put these on Christmas trees and the outside of their house (but not before electricity).
6. The night before Christmas.
8. Santa's lead reindeer when it is foggy.
9. You should leave _____ and cookies for Santa.
11. The carol that you sing when you are decorating the house.
14. December 25 is ______.
15. Silver and ________.

Down
2. Another name for Kris Kringle.
3. Don't stand under this if you don't want to be kissed.
5. The _____ were hung by the chimney with care.
7. The color of Santa's outfit.
10. Reindeer look forward to this treat that you leave for them.
12. The kind of songs you sing at Christmas time.
13. He is making a _______, checking it twice.


Aphasia News

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Thanks to:
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We would also like to thank the members of the Stroke Support Group and their families, the Speech Pathology staff, and the East Bay Institute for Research and Education.

Newsletter Information

If you would like to receive this newsletter or you have comments/suggestions, call Juliana Baldo at (925) 372-4649 or email her at juliana@ebire.org, or write to:

Center for Aphasia and Related Disorders
VA Northern Calif. Health Care System
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Martinez, CA 94553

We welcome your comments and questions!

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