

Speech disorder called apraxia can progress to neurodegenerative disease

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It may start with a simple word you can't pronounce. Your tongue and lips stumble, and gibberish comes out.

Misspeaking might draw a chuckle from family and friends. But, then, it keeps happening. Progressively, more and more [speech](#) is lost. Some patients eventually become mute from primary progressive apraxia of speech, a disorder related to degenerative neurologic disease.

Two Mayo Clinic researchers have spent more than a decade uncovering clues to apraxia of speech. Keith Josephs, M.D., a neurologist, and Joseph R. Duffy, Ph.D., a speech pathologist, will present "My Words Come Out Wrong: When Thought and Language Are Disconnected from Speech" on Sunday, Feb. 14, at the American Association for the Advancement of Science annual meeting in Washington, D.C.

Because patients and even many medical professionals don't recognize apraxia of speech, treatment typically is sought in later stages of the disease, says Dr. Josephs. As apraxia progresses, it frequently is misdiagnosed as Alzheimer's disease or [amyotrophic lateral sclerosis](#). One patient received vocal cord injections of Botox by a physician who thought the issue was muscle spasms of the larynx. Apraxia of speech even has been diagnosed as mental illness.

"Because it first presents as 'just' a speech problem, some people are told, 'This is in your head.' We've seen that. It's very sad," Dr. Josephs says.

When it's caused by a stroke, apraxia of speech typically does not worsen and may get better over time. But, apraxia of speech often is ignored as a distinct entity that can evolve into a neurologic disorder, causing difficulty with eye movement, using the limbs, walking and falling that worsens as time passes.

"I don't want the take-home message to be that this condition is benign," warns Dr. Josephs. "It is a devastating disease, in some sense worse than Alzheimer's disease, which typically spares balance and walking until very late in the disease course. It may start with the person simply not being able to pronounce a few words. Six years after that, they are in a diaper, can't speak, can't walk and are drooling."

The benefit to getting an early and correct diagnosis is that people can receive appropriate therapy. "It would be good if people recognized that changes in speech can be the first signs of neurologic disease," Dr. Duffy says. "An important part of treatment is providing information about the condition."

While speech therapy doesn't reverse or halt the progression of apraxia, it can develop compensations for producing better sounds. People with apraxia of speech also can use computers or texting for alternate means of communicating.

Both the value and complexities of speech often are underappreciated. "Speech is what connects us to the world," Dr. Duffy says.

Speech is a complex brain-body achievement, these researchers note. It first requires selection of appropriate words, organizing them into a coherent message. This message activates 100 muscles between the lungs and lips to produce at least 14 distinct sounds per second that can be comprehended by a listener. A problem with speech programming - directing the muscles and structures that move - is apraxia.

People with apraxia of speech or their loved ones may notice:

- Slow speech rate
- Inconsistent mistakes, such as saying a word or sound correctly sometimes and not

others

- Impaired rhythm of speech
- Groping of the mouth to make sounds
- Better automatic speech, such as greetings, compared with purposeful speech

Apraxia of speech differs from aphasia, a language disorder that interferes with a patient's ability to understand or use words. Patients, however, can have apraxia of speech and aphasia.

While the cause of primary progressive apraxia of speech has not been determined, an abnormal accumulation of tau protein—a factor also contributing to Alzheimer's disease—has been found in the brains of those with apraxia of speech who have died.

Mayo Clinic has received National Institutes of Health grants, for which Dr. Josephs is the primary investigator, to focus on apraxia of speech in the context of neurodegenerative cognitive and motor disorders.

Drs. Josephs, Duffy and fellow researchers have published articles about their findings in *Brain*, the *American Journal of Alzheimer's Disease & Other Dementias*, *Neurology* and the *Journal of Neurology*.

Provided by Mayo Clinic

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