THE WALL STREET JOURNAL.

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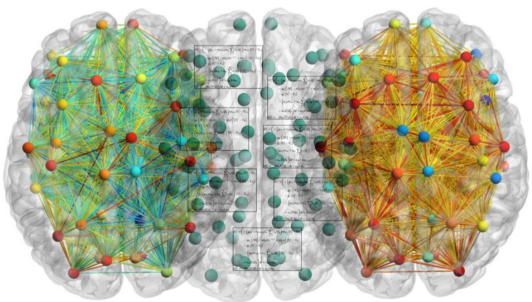
The Brain Wiring Behind a Frustrating Speech Disorder

Neurologists are searching for new ways to crack the riddle of spasmodic dysphonia



On the left, in green, is a simulation of the brain at rest. On the right, in orange, is a simulation of the brain during speech production, a complex task made possible by the activity of many interconnected areas. Both maps are based on brain-imaging data. In the middle are the regions, represented by spheres, whose connectivity was examined. *PHOTO: DR. STEFAN FUERTINGER AND DR. KRISTINA SIMONYAN*

By **DANIELA HERNANDEZ**

Oct. 31, 2016 11:42 a.m. ET

New research on the intricate patterns of brain activity needed to produce speech is giving scientists fresh insights into what goes wrong in various speech disorders.

Kristina Simonyan, a neuroscientist at the Icahn School of Medicine at Mount Sinai, in New York City, uses functional MRI scans to study brain regions involved in the various movements that control speech. Her work aims to decipher how abnormalities in different brain areas give rise to spasmodic dysphonia, a rare neurological disorder that causes spasms of the vocal cords and disrupts people's ability to speak normally.

Another researcher, Edward Chang, a neurosurgeon at the University of California, San Francisco, is mapping brain activity as people pronounce simple syllables. To accomplish this, he relies on electrodes implanted in the brains of epilepsy patients as part of their treatment for that condition. His work could eventually lead to more effective treatment for people with speech problems after a stroke or for those who stutter.

Speaking is among the most complex human behaviors the brain controls. Normal speech depends on the precise coordination of more than 100 muscles, spread across the mouth, lungs and vocal cords. The brain issues a series of rapid-fire commands directing these muscles to move in the exact pattern needed to voice specific syllables and words.

The work is part of a broader trend in neuroscience known as connectomics, the study of how brain cells are functionally connected. The thinking goes that if scientists understand how the brain's networks coordinate tasks like speech, they can home in on

Dr. Kristina Simonyan is a Mount Sinai neurologist studying dysphonia using functional MRI scans. *PHOTO: DANIELA HERNANDEZ/THE WALL STREET JOURNAL*

disease-causing abnormalities and develop diagnostics and treatments.

Brain researchers don't fully grasp yet "how one region influences another" or "what happens when those connections are disrupted," says Jeremy Greenlee, a neurosurgeon at the University of Iowa Carver College of Medicine who studies speech. As a result, doctors "don't have the tools we would like to have to help patients as much as we could."

Dr. Chang says his work using brain electrodes reveals how neurons control a series of so-called articulators—including the larynx, lips, facial muscles and tongue—that work together to coordinate speech.

Some articulators shape the breath. Others push air up through the vocal cords. Still others move the tongue, cheeks and lips. When one of these players falls out of line, speech can sound slurred, shaky or choppy and raspy, as in spasmodic dysphonia.

In the case of tongue twisters, Dr. Chang's research could suggest, the brain may have a hard time differentiating between similar-sounding syllables, and the directions it sends the various voicing muscles get muddled. The findings could someday have implications for people who stutter or have had a stroke, he says.

Another goal: to understand the patterns of speech-related brain activity so precisely that doctors can devise a brain implant that returns patients' ability to speak normally in the future, Dr. Chang says.

Spasmodic dysphonia, which is incurable, is characterized by uncontrollable voice breaks, strained speech and excessive breathiness. It usually strikes in midlife, when patients hit their personal and professional prime.

Some 50,000 people in North America are listed as having the condition, but the number is likely higher, since the disease is often misdiagnosed, according to the National Spasmodic Dysphonia Association, an advocacy group. Treatments, such as Botox injections and in some cases laryngeal surgery, minimize symptoms temporarily.

Spasmodic dysphonia, long believed to be a psychological problem, is now considered to be part of a family of conditions called dystonias—neurological movement disorders characterized by involuntary muscle contractions. Parkinson's disease patients can experience dystonias.

The disorder affects many different interacting brain areas, Mount Sinai's Dr. Simonyan says. Patients with spasmodic dysphonia exhibit different structure, function and connectivity between various brain regions—including the basal ganglia, sensorimotor cortex and cerebellum—than people who don't have the condition, she says.

For a study published in the European Journal of Neurology in June, Dr. Simonyan performed brain scans on roughly 100 patients and healthy controls. She looked for patterns to use in developing a computer algorithm to differentiate between people with normal brains and those with spasmodic dysphonia. The program currently can identify people with the disorder with about 70% accuracy, she says. With further research she


Tommy Johnson, seen with his granddaughter, Emerson, has been battling spasmodic dysphonia for three decades.
PHOTO: TOMMY JOHNSON

hopes to increase the accuracy, as well as study the genetic links of the disorder.

Tommy Johnson, a 62-year-old retired Air Force communications officer, says he developed spasmodic dysphonia three decades ago, when few treatment options existed. He spent nine months in military psychiatric hospitals trying different medications that didn't help.

Unable to do his job in communications, he says he was discharged from duty and moved to Florida, but couldn't land a job. He eventually moved in with his mother in Maine. "I felt so helpless and hopeless. I couldn't speak," he says.

For more than two decades, Mr. Johnson has been getting Botox injections in his vocal cords every few months, which allowed him to find work. Now retired for 13 years, Mr. Johnson helps Dr. Simonyan conduct her research into spasmodic dysphonia. He says he has registered with the National Institutes of Health to donate his brain after he dies. The organ will go to Dr. Simonyan to aid in her research of the disorder.

"She will someday hold my brain in her hands," he says.

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