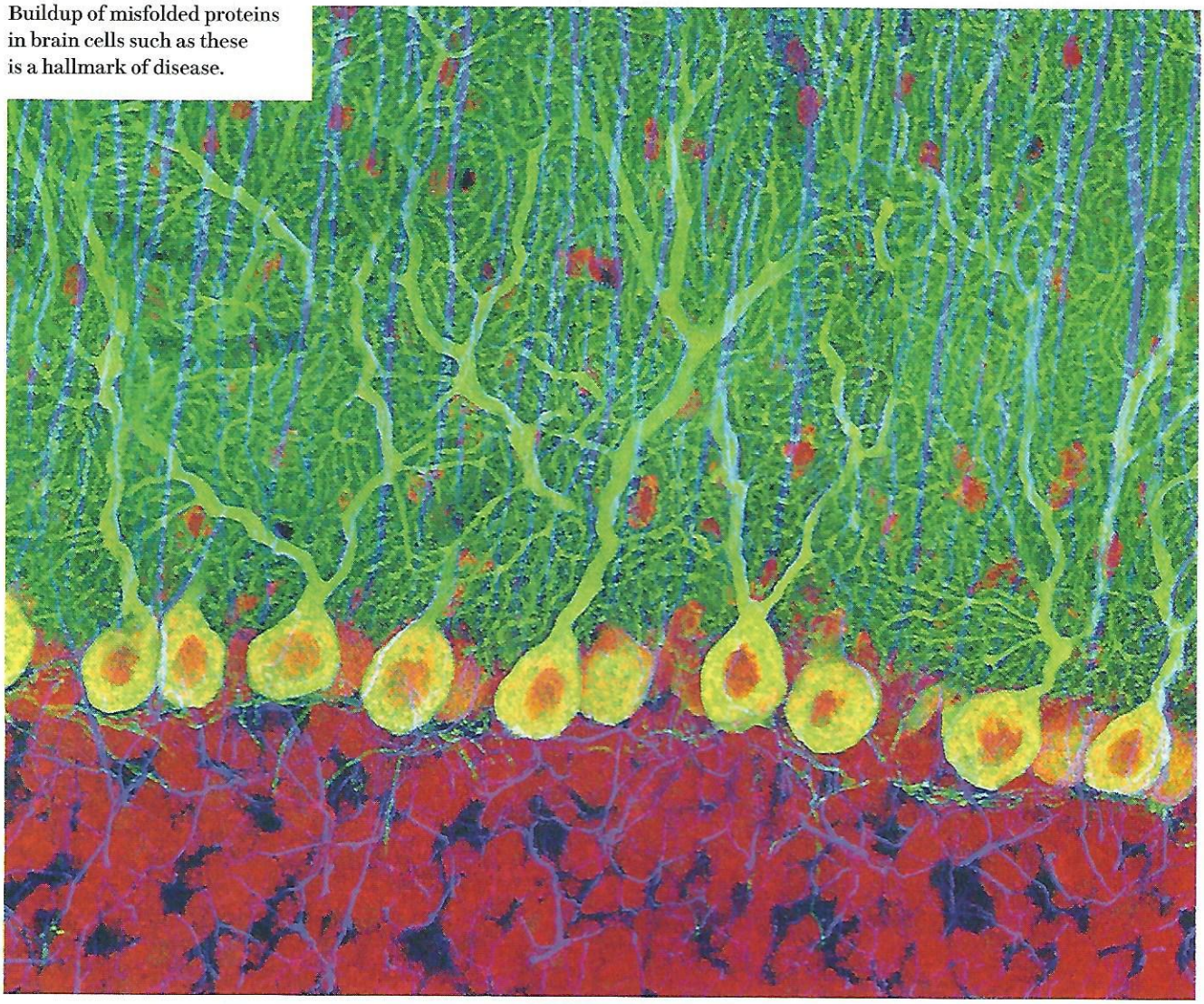

Buildup of misfolded proteins in brain cells such as these is a hallmark of disease.

THOMAS DEERINCK Science Source

NEUROSCIENCE

When Good Proteins Go Bad

As baby boomers acquire the neurodegenerative diseases that come with age, researchers focus on a potential avenue to new treatments: targeting cell-to-cell transfer of misfolded proteins

The first step to treating or preventing a disease is often finding out what drives it. In the case of neurodegenerative disorders, the discovery two decades ago of what drives them changed the field: all of them—including Alzheimer's, Parkinson's, Huntington's and amyotrophic lateral sclerosis (ALS or Lou Gehrig's disease)—involve the accumulation of misfolded proteins in brain cells.

Typically when a protein misfolds, the cell destroys it, but as a person ages, this quality-control mechanism starts to fail and the rogue proteins build up. In Huntington's, for example, huntingtin protein—used for many cell functions—misfolds and accumulates. Symptoms such as muscular difficulties, irritability, declining memory, poor impulse control and cognitive deterioration accompany the buildup.

Mounting evidence suggests that not only does the accumulation of misfolded proteins mark neurodegenerative disease but that the spread of the proteins from one cell to another causes the disease to progress. Researchers have seen misfolded proteins travel between cells in Alzheimer's and Parkinson's. A series of experiments reported in *Nature Neuroscience* in August suggests the same is true

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in Huntington's. (*Scientific American* is part of Nature Publishing Group.)

In their tests, researchers in Switzerland showed that mutated huntingtin protein in diseased brain tissue could invade healthy brain tissue when the two were placed together. And when the team injected the mutated protein into a live mouse's brain, it spread through the neurons within a month—similar to the way prions spread, says Francesco Paolo Di Giorgio of the Novartis Institutes for BioMedical Research in Basel, who led the research. Prions are misfolded proteins that travel through the body and confer their disease-causing characteristics onto other proteins, as seen in mad cow disease. But it is not known if misfolded proteins involved in Huntington's convert other proteins as true prions do, according to Di Giorgio.

Scientists have yet to establish that the movement of bad proteins is critical for the progression of the disease, notes Albert La Spada, a geneticist at the University of California, San Diego, who was not involved in the study. But if it turns out that traveling is essential, then therapies may be able to target the pathway. "If we can find out how it's occurring," La Spada says, "then we might be able to come up with treatments to prevent it." And those treatments could potentially apply to the other neurodegenerative diseases.

The next step is crucial. Researchers will try to block the spread of misfolded proteins and see if that improves symptoms or slows progression. Finding therapies for these diseases is paramount. Approximately 50,000 new cases of Parkinson's alone are diagnosed every year in the U.S., and experts estimate the prevalence will at least double by 2030 because of an aging population. —*Tara Haelle*

According to the Alzheimer's Association, more than **5.2 million** Americans have Alzheimer's

60% of them are prone to wander

WEARABLE TECH

Safety in a Sock

A teenager wins big for an invention that monitors Alzheimer's patients

Fifteen-year-old Kenneth Shinozuka of New York City won the $50,000 *Scientific American* Science in Action Award in August for his invention of a wearable sensor for Alzheimer's patients. The prize, part of the Google Science Fair, recognizes a teen for an innovation that can make a practical difference by

addressing an environmental, health or resources challenge.

Shinozuka's creation—a small pressure sensor that can be attached to a foot or a sock—notifies caregivers via their smartphones if a patient who should be sleeping gets out of bed. His grandfather, who has Alzheimer's disease, served as inspiration. "I don't think I will ever forget my shock at seeing Grandfather in his pajamas, accompanied by a policeman who found him wandering on a nearby freeway in the middle of the night," Shinozuka says. He designed the sensor to keep his grandfather safe and to provide much needed relief to his aunt, the primary caregiver. Shinozuka recently demonstrated the technology at a local chapter of the Alzheimer's Association and a number of care facilities. He has obtained a U.S. patent for his invention.

—*Rachel Scheer and Annie Sneed*

COURTESY OF KENNETH SHINOZUKA