Researchers Identify New Rare Neuromuscular Disease

An international team of scientists—co-led by researchers in the School of Medicine and Dentistry—has identified a new inherited neuromuscular disorder. The rare condition is the result of a genetic mutation that interferes with the communication between nerves and muscles, resulting in impaired muscle control.

The disease was diagnosed in two families—one in the United States and one in Great Britain—and affects multiple generations. The discovery was published in the *American Journal of Human Genetics*.

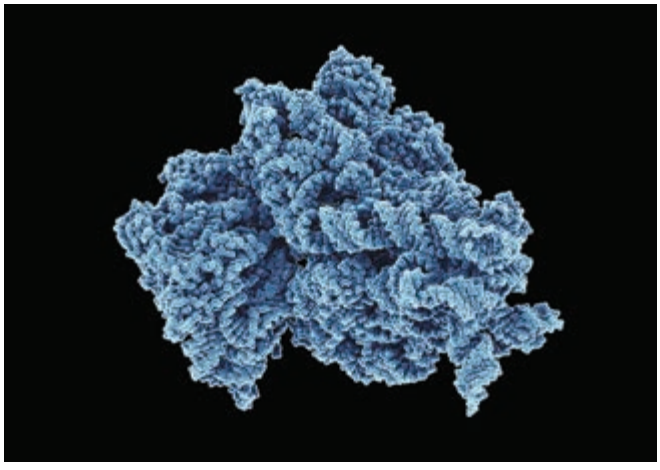
The discovery provides new insight into the mechanisms of diseases caused by a breakdown in neuromuscular signal transmission, says David Herrmann, professor of neurology and a lead author of the study. "It is our hope that these findings will help identify new targets for therapies that can eventually be used to treat these diseases."

The focus of the research is the neuromuscular junction, the point at which the axon fibers that extend from peripheral nerves meet the muscle cells. The chemical signals that pass across the junction are essential for motor function. While the families in the study had at one point been diagnosed with other neuromuscular conditions, the researchers identified them as unique due to their particular motor abnormalities and because the disease was passed from one generation to the next. —Mark Michaud

Going After Harmful Bacteria

One challenge in killing off harmful bacteria is that many of them develop resistance to antibiotics. Now researchers are targeting the formation of the protein-making machinery, or ribosomes, in those cells as a possible way to stop the bacteria. Gloria Culver, professor of biology, has, for the first time, isolated the middle steps in the process that creates the ribosomes.

Published in *Nature Structural and Molecular Biology*, Culver's work—conducted with graduate student Neha Gupta—captures a piece of ribosomal RNA in one of the intermediate states of being pared down to fit with protein molecules. They did so by using genetic tags as markers inside *E. coli* cells. By attaching the tags to nonfunctional regions of the uncut RNA, they were able to isolate the immature RNA strands during the various stages of processing.



STOPPING BACTERIA: New Rochester research uses bacterial ribosomes to advance efforts to kill drug-resistant bacteria.

Targeting ribosomes to kill drug-resistant bacteria is nothing new, but in the past scientists focused on mature ribosomes. While a range of antibiotics was developed to attach the ribosomes, the microbes eventually became resistant to the drugs. While a great deal of work remains to be done, Culver's research creates new possibilities for stopping super-bugs.

The team's discoveries "suggest that there is at least one common step that could be exploited to one day help scientists prevent the ribosomes from developing, which would kill off the bacteria," she says. —Peter Iglinski

Opening a Door to Speed-of-Light Computing?

A new combination of materials can efficiently guide electricity and light along the same tiny wire, a finding that could be a step toward building computer chips capable of transporting digital information at the speed of light.

In a report in the journal *Optica*, optical and material scientists at Rochester and the Swiss Federal Institute of Technology in Zurich describe a basic model circuit consisting of silver nanowire and a single-layer flake of molybdenum disulfide.

Using a laser to excite electromagnetic waves called plasmons at the surface of the wire, the researchers found that the flake at the end of the wire generated strong light emission.

Going in the other direction, as the excited electrons relaxed, they were collected by the wire and converted back into plasmons, which emitted light of the same wavelength.

The "nanoscale light-matter interaction between plasmons and atomically thin material can be exploited for nanophotonic integrated circuits," says Nick Vamivakas, assistant professor at the Institute of Optics and senior author of the paper.

Photonic devices can be much faster than electronic ones, but they're bulkier because devices that focus light can't be miniaturized nearly as well as electronic circuits.

The new results hold promise for guiding the transmission of light and maintaining the intensity of the signal in very small dimensions.

Combining electronics and photonics on the same integrated circuits could drastically improve the performance and efficiency of mobile technology.

—David Barnstone

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Exploring the 'Sublime'

After traveling through Italy in 1699, English writer Joseph Addison noted how the "Alps fill the mind with an agreeable kind of horror."

His observations, along with those of other Englishmen who crossed the mountains on their continental Grand Tours, helped spark an intense interest in the "sublime" as an aesthetic concept—one distinct from "beauty" in its power simultaneously to overwhelm, elevate, and even terrify.

Robert Doran, associate professor of French and comparative literature, explores how and why the distinction became important in his forthcoming book, *The Theory of the Sublime from Longinus to Kant* (Cambridge University Press, 2015).

"The concept of the sublime allows us to conceive of a certain kind of experience that normally would only be accessible using theological concepts," says Doran. "It's a secularized version of religious experience. It's the experience of transcendence, but transcendence in the arts or in the aesthetic appreciation of nature."

Doran argues that the concept of the sublime allowed the middle class to experience a "nobility of mind." "The ordinary individual can experience this kind of nobility of mind, as it were, this heroic feeling that seemed to be only possible for certain classes before," he says.

The concept is still relevant today, he says—one that's possible to experience standing before the Grand Canyon or Niagara Falls, for example—and he contends that the desire for transcendence is even a defining trait of humanity.

—Bob Marcotte

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