JHU cognitive scientists devise alphabets that allow subjects to read again



Image: Illustration by Greg Stanley

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Diane Slonim recalls the moment last fall she nearly lost faith.

A speech pathologist in New York, Slonim had a unique client, a 12-year-old girl she wanted desperately to help. The girl had two years earlier suffered a stroke and could no longer see letters and numbers except for an "O."



Ultimately, we favored using the double strikethrough font because it was the first we discovered that MTS can see, and she prefers it.

Show the girl an "H," the word "grape," or the number "42," and all she saw was a blur, like a character written in pencil and then

erased.

The girl, who before the stroke read at grade level, could no longer decipher the most basic sentence, and her own writing became unintelligible to her as soon as letters were formed. She was falling behind in school and in danger of failing out.

Confounding many, the girl could see and remember everything else perfectly well. Show her a rabbit, even a crudely drawn one, and she perceived every last whisker.

Slonim suspected that the girl had a severe form of alexia, or acquired dyslexia. She began contacting universities looking for assistance and hit brick walls until a Google search led her to the Department of Cognitive Science at Johns Hopkins' Krieger School of Arts and Sciences.

After an email exchange with Brenda Rapp, chair of the department, Slonim was connected with Michael McCloskey, a professor who focuses on cognitive deficits in children and adults with brain damage or learning disabilities, including issues with visual perception, reading, spelling, and memory.

Within weeks of the initial contact, McCloskey would lead an effort to help find interventions to allow the girl to read again, and start to decipher the complex riddle of what transpired in the girl's brain to cause the deficiency.

McCloskey says he knew from the onset that the girl's problem was extremely unusual. He had seen many cases of brain damage that affected a person's ability to read and use numbers, but the obstacle in many of those cases was that the individual no longer knew what the letters and numbers meant.

"So somebody like that might not be able to tell you it's an 'S' but still see the shape for what it is. They could copy it and describe it to you but not know that it is an 'S," he says. "Or, because the eyes or the visual areas of the brain are damaged, everything can look blurry or distorted, but it will not affect just your ability to see numbers and letters."

McCloskey did, however, have some comparison.

He was working at the time with a 61-year-old man from Baltimore, an engineering geologist who was unable to see numbers and had trouble with a small set of capital letters, including M, N, R, and S.

The man, referred to in McCloskey's research as RFS, was diagnosed with a progressive neurological disease called corticobasal degeneration, which affects the cerebral cortex and basal ganglia. RFS was being seen as an outpatient in Neurology and in Medical Psychology at the Johns Hopkins Hospital.

Like the girl, RFS could see other images, shapes, and symbols correctly.

"The odd part of these cases is it's selective to a specific type of thing that you learn and the form that it takes, and then you can't see them right," McCloskey says. "That is what ties them together. Their issues are not identical, but it's a very similar and unusual type of problem. For both, the processing of these letters and numbers may not be what is affected, but the awareness of what they are seeing [is]. It makes us think about what it takes for us to be aware, for us to see."

McCloskey headed to New York to learn more about how the girl, known as MTS for research purposes, perceives letters and numbers.

MTS' stroke was caused by an arteriovenous malformation. When an AVM occurs, a tangle of blood vessels in the brain, or on its surface, bypasses normal brain tissue and diverts blood directly from the arteries to the veins. The veins are not designed to handle high pressure and can rupture, causing a pooling of blood.

For a time, MTS had some movement issues and slurred speech, but through therapy has since regained her motor abilities, though she continues to have some residual weakness on her left side.

What remained was the problem with seeing letters and digits.

First, McCloskey ruled out that there was a problem with her eyes.

"We would show her a picture of a camel and a shape, like a square, and she would see that fine. She could also write and draw like any other 12-year-old," he says. "It suggests to us that the brain separates numbers and letters from other images."

Knowing the problem was neurological, McCloskey and his team—including graduate students Teresa Schubert and David Rothlein and colleague Brenda Rapp—wanted to see if they could somehow trick MTS' brain into seeing letters and numbers more as shapes.

After some experimentation, they found she could see some numbers and letters if they tinkered with them, pulling apart their components or adding "bits" to standard fonts. For example, they discovered that she could see an "8" if they put a gap between the two circles, and see an "A" if the horizontal line floated between the two inward-leaning lines.

Wanting a more uniform and elegant solution, the team brainstormed with Beth Breakstone and Karen Van Den Heuvel, assistive technology specialists who were working with MTS. Breakstone suggested characters with a single strikethrough, and after some trial and error the group hit pay dirt with a black double strikethrough placed at a certain height on a Comic Sans font.

The strikethroughs instantly allowed her to see all the letters and digits.

"This girl hasn't been able to read a word for two years, and suddenly she can read. Very strange. We don't know exactly what lets her see those and not the standard characters, but it works," McCloskey says. "The really odd part is that a single strikethrough doesn't work, and if the strikethroughs are placed too high or too low, it doesn't work. We don't know what is happening, but somehow we are bypassing whatever triggers her inability to see."

The team also learned that MTS could see some untraditional fonts such as Tranceform, Sparkling, and Demon Metallic.

"She can see all of the characters in these fonts without blurring, although in some she may have difficulty determining what letter some of the shapes are," he says. "Ultimately, we favored using the double strikethrough font because it was the first we discovered that she can see, and she prefers it."

The McCloskey-led team created a double strike-through font for MTS and installed it on a laptop so she could write again. Then, a JHU undergrad, Satyam Ghodasara, created an iPad calculator app to allow MTS to work on math problems. He also crafted a photo app so that she could take pictures of pages of text, like a homework sheet, and it would add strikethroughs to the words. McCloskey says that while the interventions have been effective, they are far from perfect.

"She still can't see what she handwrites, and what happens when she has to read a menu, or is walking around and needs to read a sign?" he says. "The iPad app has some

limitations, such as problems with distinguishing writing on maps and images. The text has to be clean."

MTS also has trouble with any visual stimulus next to a letter or digit. She can see a percent sign by itself, but "4%" could be blurred.

Unassisted by technology, MTS has shown some signs of improvement. Today, she can recognize not only "O" but also "L," "U," and "V."

"Maybe she will gradually see more of the alphabet. I certainly hope so," McCloskey says. "We just have to wait and see, and do as much as we can for her, making our interventions more effective. She just turned 13, and younger brains are very adaptable. Perhaps one day, spontaneously, she will be able to see again normally."

MTS recently underwent some functional magnetic resonance imaging so researchers can study how her brain responds when shown characters she can't see, and those she can.

"We are intrigued to see the results of that," McCloskey says.

McCloskey has also developed effective interventions for the man with whom he is working.

RFS says that when he sees digits, they look like an angry arrangement of black lines with color in the background. (Imagine a 2-year-old scribbling on colored paper.) And each time he looks at the same number, the lines might appear different.

"He might basically see the same blur, but the lines will be in different places," Mc-Closkey says. "And if you show him two digits that are the same right next to each other, he can't tell you whether they are the same or different. He is just completely unable to use numbers."

To allow him to see, McCloskey's team developed a set of characters based on a logical system of horizontal and vertical lines, with a standard "0" that the man is still able to discern. The horizontal lines range in ascending values of 2, so what looks like an "L" would be 2, and an "E" with the top horizontal line removed equals 6.

Ghodasara developed a calculator app with this new digit system and installed it on the man's iPhone so that he's able to continue to work in his number-heavy profession. The

app has a toggle switch to revert to standard fonts so that RFS can show others his results.

The numerical system worked, and he adapted quickly, McCloskey says. "One thing we were concerned about is that once he started to learn these, they would start to look funny to him also. But fortunately that hasn't happened."

The team also replaced the fonts on his computer so that anything with numbers, including the clock and spreadsheets, are displayed in the new number system.

McCloskey says while it's odd to have found two people with similar reading deficiencies within the course of a few months, he suspects there are many more out there. He already has identified a person in North Carolina who may have an impairment with numbers similar to that of RFS.

"What we find could have learning applications," he says. "What could be going wrong that someone is not able to see these symbols that he or she has already learned? Part of my research going forward is to try to find more people like this. They are not going to be exactly the same, but we are looking for this general pattern in not being able to see letters, digits, and symbols."

The research, he says, could tell us more about how the brain works in general.

"It is intriguing where MTS had her stroke, where the damage is done," he says. "It's not an area that you would typically think about reading numbers and letters. You usually think about the gray matter covering the outside of the brain. But her damage is deep in the brain in the basal ganglia. This might help tell us what is going on in this portion of the brain and the role it plays."

McCloskey says that both MTS and RFS may be processing numbers and letters even though they are not aware of it; what may be affected is the awareness of what they're seeing, not their ability to see it directly.

In studying these individuals, his team is using behavioral, electrophysiological, and functional neuroimaging methods to address questions concerning the cognitive and neural representations underlying visual awareness and reading.

"I don't believe the brain is hardwired for reading. We are still too young, evolutionally speaking," he says. "We learn things in specific places in the brain, and that is an interest-

ing mystery to solve."

For MTS, she just knows she can read again.

Slonim says the impact of McCloskey's work on the girl has been profound. This spring, MTS made the honor roll.

"It's been nothing short of amazing, from not being able to read anything but an 'O' to reading pages," Slonim says. "This beautiful young girl really wants to learn in a big way. And now with these new tools, she can read again. I knew I had to try to find somebody who knew something about this condition, who could help find some answers. Dr. McCloskey's work has been incredible. He is not just brilliant; he is caring. He's an enormous human being."

McCloskey says that MTS deserves much credit.

"She's a resourceful little girl," he says. "We wanted to overlay the new strikethrough font on her laptop keyboard, but she refused. She remembers where all they keys are without our help, and she didn't want her laptop to look too dorky."

Typical teen.