

## A Brief History of Cranial Nerve VII

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The year is 1963 at the University of Cambridge in England. I am a neuron located on the buccal branch of cranial nerve VII, the facial nerve, inside the brain of one of the most brilliant scientists the world has ever known: Stephen Hawking. In about a year's time, he will be diagnosed with 'motor neuron disease', also known to Americans as amyotrophic lateral sclerosis or ALS.

### March 16, 1963

At first, it was slight things. One of my best friends, a neuron from the optic nerve, located a couple of nerves away, was sending me signals so that I could see what was happening to Stephen: stumbling up the stairs, dropping of the chalk he was using on the blackboard, and giving out of his arms with fatigue during rowing. With each day, I could feel myself grow concerned. Something odd was happening to Stephen's motor neurons. Today, I heard some rumors from the vestibulocochlear nerve that the motor neurons in Stephen's extremities hadn't fired an action potential. The muscles in his hand cramped up and knocked over the cup of coffee on his desk. That's so weird! I thought to myself. The message from the bipolar cell to the optic neuron to the interneuron to the motor neuron in his hand had been clear: reach for the handle! Why didn't they fire the impulse? I wondered as I pulled Stephen's cheek muscle down into a frown.

Stephen is a cosmologist, and he has been working on his thesis about black holes. I knew that he had to focus, which is why I hoped that whatever was going on with the motor neurons could be fixed. Their mistakes could be getting in the way of Stephen's success!

To my knowledge, Stephen's neurons were functioning as well as they always had...

Nobody remembers the day they are born, but I do. It was rather sudden. I was born from a cell division, as a stem cell, with a small, round, white body. Time passed, and I began to grow: my cell body was stretching as far as it could. The "growing pains", as I like to call them, lasted for what seemed like forever. Then, one day, they stopped. I was slimmer – I was now a neuron. Out of nowhere, from deep inside my organelles, I could hear a voice telling me to move. I reached up to the tall, towering radial glial cells above me and began to move, listening to the voice inside my head. I climbed through the forest of radial glia cells until I reached my destination: the buccal branch of cranial nerve VII, the facial nerve.

Once I settled in, my axon and dendrites extended to the surrounding neurons and made instant, synaptic connections with them. As soon as the connections were established, action potentials began to pass into my dendrites and through my body. To me, the action potentials were little voices whispering instructions. It was like playing a massive game of Telephone. In the buccal branch, I, along with other neurons, was responsible for contracting the cheek muscle to create facial expressions. The job was easy. And for the next twenty years, I never heard of any problems concerning motor neurons. Until now...

### December 28, 1963

Today was one of the worst days I've ever experienced in my life. Since the last time we talked, the problems with Stephen's motor neurons have gradually gotten worse, and just when I thought it couldn't get any worse, it did.

Stephen had been eating some of his mother's scones. The hypoglossal nerve and the glossopharyngeal nerve, a couple of nerves down from me, were having a field day with all the flavor. Suddenly, a dreadful sensation overcame me. When something goes wrong with one of the cranial nerves around the face, we all sense it because we are all so closely located to each other. I glanced over and saw that some of the neurons in the hypoglossal nerve had...died. The neurons around them were panicking, trying to revive them with action potentials, but nothing worked. The neurons were graying and weakly gripping on the muscles of the tongue. They simply weren't functioning. The neurons around them quickly moved to the tongue to try and compensate for the dead neurons.

It was of no use. Weak control in the tongue muscles were going to affect Stephen's speech – the way he talks will never be the same again. Then, right after the discovery I'd made, he began to talk. I could hear him slurring his words like a happy drunk. His family noticed the slurred speech, and they rushed him to the hospital.

All of it was happening so fast. Seeing those nearby neurons die chilled me to the cytoskeleton. I knew some of them, and it saddens me deeply that I'll never see them again. The diagnosis came: "It's called motor neuron disease. Life expectancy is two to three years, maybe five, if he's lucky."

### June 27, 1986

It's been a long twenty-three years for me since Stephen Hawking's diagnosis with ALS. I still haven't forgotten the day when the diagnosis was given. It was a bittersweet feeling. I had finally understood what had been killed my friends on that day. But I'm no fool – I know the inescapable fate of all motor neurons like me in a body with ALS. The lower motor neurons used to be so alive, with signals rushing through the body. Since Stephen's diagnosis, the motor neuron activity began to quiet down – today, it is completely silent. Absolutely no signals come through. Thus, Stephen had been sentenced to live the rest of his life in a wheelchair. The upper motor neurons are starting to die.

I'm dying, too. I noticed it a couple of years ago while working: the very end of my long axon, right where the terminal buttons began, was exposed. The warm myelin sheath that wrapped around my axon had rotten away. With each year that passed, I lost another layer of myelin. Today, more than half of the myelin on my axon had rotted away, leaving me even more exposed, weak and cold. The disease had robbed me of my myelin and of my will to keep on fighting. I'm tired.

However, I am glad for the life I have lived. The optic nerve had been filling me in on what's happened to Stephen over these years. He got his PhD in cosmology. He's surpassed the life expectancy of ALS by over ten years! I couldn't help being smug. Stephen resisted the effects of the disease and I had resisted with him, every step of the way! After receiving a tracheotomy, Stephen was given a speech device, attached to his wheelchair. It allows him to communicate. The optic nerve told me, with amusement, that the device is attached to his cheek muscle. You can retire early, he'd joked. Maybe he was right. I know Stephen will die someday from this disease, but he's lived a full life and so have I. I smiled to myself and glanced down, just as the last layer of myelin on my axon fell away...

### Neuroscience concepts within the story

- ALS/motor neuron disease (amyotrophic lateral sclerosis)
- Functions of the cranial nerves (the facial nerve, the vestibulocochlear nerve, the optic nerve, the hypoglossal nerve and the glossopharyngeal nerve)
- Neural development
  - Neurogenesis (formation of new neurons, from stem cells)
  - Cell migration (via the radial glial cells)
  - Maturation (dendrite branching and growth)
  - Synaptogenesis (formation of synapses)
- Neural circuit (connections from the sensory neurons to the interneurons to the motor neurons)
- Demyelination (seen in ALS – the loss of myelin sheath around the motor neurons)
- Motor neuron degeneration

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