

Dying Outside

by [HalFinney](#)

A man goes in to see his doctor, and after some tests, the doctor says, "I'm sorry, but you have a fatal disease."

Man: "That's terrible! How long have I got?"

Doctor: "Ten."

Man: "Ten? What kind of answer is that? Ten months? Ten years? Ten what?"

The doctor looks at his watch. "Nine."

Recently I received some bad medical news (although not as bad as in the joke). Unfortunately I have been diagnosed with a fatal disease, Amyotrophic Lateral Sclerosis or ALS, sometimes called Lou Gehrig's disease. ALS causes nerve damage, progressive muscle weakness and paralysis, and ultimately death. Patients lose the ability to talk, walk, move, eventually even to breathe, which is usually the end of life. This process generally takes about 2 to 5 years.

There are however two bright spots in this picture. The first is that ALS normally does not affect higher brain functions. I will retain my abilities to think and reason as usual. Even as my body is dying outside, I will remain alive inside.

The second relates to survival. Although ALS is generally described as a fatal disease, this is not quite true. It is only mostly fatal. When breathing begins to fail, ALS patients must make a choice. They have the option to either go onto invasive mechanical respiration, which involves a tracheotomy and breathing machine, or they can die in comfort. I was very surprised to learn that over 90% of ALS patients choose to die. And even among those who choose life, for the great majority this is an emergency decision made in the hospital during a medical respiratory crisis. In a few cases the patient will have made his wishes known in advance, but most of the time the procedure is done as part of the medical management of the situation, and then the ALS patient either lives with it or asks to have the machine disconnected so he can die. Probably fewer than 1% of ALS patients arrange to go onto ventilation when they are still in relatively good health, even though this provides the best odds for a successful transition.

With mechanical respiration, survival with ALS can be indefinitely extended. And the great majority of people living on respirators say that their quality of life is good and they are happy with their decision. (There may be a selection effect here.) It seems, then, that calling ALS a fatal disease is an oversimplification. ALS takes away your body, but it does not take away your mind, and if you are determined and fortunate, it does not have to take away your life.

There are a number of practical and financial obstacles to successfully surviving on a ventilator, foremost among them the great load on caregivers. No doubt this contributes to the high rates of choosing death. But it seems that much of the objection is philosophical. People are not happy about being kept alive by machines. And they assume that their quality of life would be poor, without the ability to move and participate in their usual activities. This is despite the fact that most people on respirators describe their quality of life as acceptable to good. As we have seen in other contexts, people are surprisingly poor predictors of how they will react to changed circumstances. This seems to be such a case, contributing to the high death rates for ALS patients.

I hope that when the time comes, I will choose life. ALS kills only motor neurons, which carry signals to the muscles. The senses are intact. And most patients retain at least some vestige of control over a few muscles, which with modern technology can offer a surprisingly effective mode of communication. Stephen Hawking, the world's longest surviving ALS patient at over 40 years since diagnosis, is said to be able to type at ten words per minute by twitching a cheek muscle. I hope to be able to read, browse the net, and even participate in conversations by email and messaging. Voice synthesizers allow local communications, and I am making use of a free service for ALS patients which will create a synthetic model of my own natural voice, for future use. I may even still be able to write code, and my dream is to contribute to open source software projects even from within an immobile body. That will be a life very much worth living.