

The Case of Eric, Lou Gehrig's Disease, and Stem Cell Research



by
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Eric loved the fall because it meant the start of soccer season. Soccer was in his blood. He had played since he was five and now, at the age of 31, he was coaching children that very age. In his mind, nothing compared to seeing a five-year-old score his or her first goal—that awesome, untamed joy. Of course, he loved his family, his girlfriend of three years, and his job as a computer programmer, but working with kids was his unabashed passion.

It was on the soccer field that he first realized something was very wrong with his body, more specifically with his leg muscles. The kids thought it was incredibly funny when Eric tripped and fell several times during passing drills. But he knew there was definitely something amiss when his legs were trembling after practice.

It took him several weeks to get up the courage to go see his primary care doctor, Dr. Jonathan Drake, who had also been one of his college roommates. Jon was at first dismissive, joking, “Ah, Eric, you’re just getting old like the rest of us!” But he went ahead and ordered some blood tests. A week later, the test results showed nothing abnormal. But Eric, feeling a little panicked because he had tripped going down his apartment stairs twice that day, persuaded Jon to look further.

Jon referred Eric to a neurologist, Dr. Samantha Reiter. Eric felt fortunate to get an appointment that week and was encouraged by her take-charge approach. She took a complete family history, asked him dozens of questions about his symptoms and general health, and proceeded to schedule him for an MRI (magnetic resonance imaging) and EMG (electromyogram).

The same day he got the results back from Dr. Reiter, just two months after first sprawling on the ground during soccer practice, his arm muscle started to twitch ever so slightly. It seemed like cruel fate that he should feel worse and have to sit in Dr. Reiter’s office, hearing but not really accepting her diagnosis. He did catch that she thought there were several possible causes for his symptoms, including MS (multiple sclerosis) and MD (muscular dystrophy), but the one she was favoring was ALS, amyotrophic lateral sclerosis, also known in the United States as Lou Gehrig’s disease. ALS was difficult to diagnose, but she was going to schedule him for a muscle biopsy in order to assess whether or not muscle damage was present in both the upper and lower body neurons. Deterioration had to be in both regions in order to make a clear diagnosis of ALS.

The next time he saw Dr. Reiter, the results of the muscle biopsy were in and Eric brought his girlfriend LeAnne with him to the doctor’s office. Dr. Reiter, as usual, was very kind, but direct. “Eric, I am 95 percent sure you have sporadic ALS, meaning it is not hereditary. I have no intention of overwhelming you, as I know how difficult an adjustment this will be, but I also want you to be prepared for what lies ahead.” She proceeded to rattle off the frightening facts:



ALS is a fatal neuromuscular disease that affects between seven to 11 people out of every 100,000 (13 new cases are diagnosed every day)... The cause is not known and the progression is generally rapid... Eighty percent of ALS patients die within five years of diagnosis and 50 percent in 18 months....

“There is good and bad news about how the disease progresses,” Dr. Reiter continued. “The bad news is that the disease, by some yet unknown manner, gradually destroys voluntary motor neurons, which stimulate the muscles over which you have control. As the muscles receive fewer and fewer signals from these dying neurons, they weaken and atrophy. Probably, Eric, when you first noticed your symptoms on the soccer field, more than half of your motor neurons were already dead. The good news is the disease does not affect the heart, bowel, bladder or sexual function, and the younger a person is when diagnosed, the longer he or she lives. Also, there is a drug that slightly slows the progression of the disease.”

Dr. Reiter had been talking for about 15 minutes when she paused and looked from Eric to LeAnne. Neither spoke, but she had seen that devastated look in a patient’s eyes before. She proceeded by saying, “I want to assure you that I will always be your advocate. I will monitor you weekly if need be, inform you of the latest treatments, and go head to head with the insurance agency if that is what it takes to make sure you live a comfortable life.”

She had much more to tell him about possible treatments, but she thought it would be better if he went home and talked to his family and LeAnne. She gave him some material from the ALS Association based in Calabasas Hills, California, recommended that he take a look at some ALS web sites, and made an appointment with him for the following Monday.

That weekend Eric felt like he was in a fog. The most difficult part was telling his parents about his diagnosis and prognosis. He and his parents and his older sister Kerry were very close, living in Maryland their entire lives. Eric had attended the University of Maryland for his computer science degree and Kerry received her biology degree from another local school, St. Mary’s College. She was now working at Johns Hopkins School of Medicine as a research technician. They all ate Sunday dinner together twice a month after attending their town’s Catholic church, and even took an annual family vacation to the Eastern Shore in June, after Kerry’s two kids got out of school. As Eric repeated to his parents what Dr. Reiter had told him, in the back of his mind he was wondering if he would even be able to walk on the beach next June. His parents and sister insisted that they meet with Dr. Reiter too when Eric went for his Monday appointment. They wanted to ask the many questions that Eric hadn’t dared to face, at least alone.

When Monday rolled around, Eric’s sister, Kerry, had the longest list of questions. His parents wanted to know about prognosis and how the disease is manifested, but Kerry was more interested in “the cure.” She had gone on the Internet and found out much more about the FDA approved drug, Rilutek, and creatine. She also read about the promise of embryonic stem cell therapy for curing diseases such as diabetes, Parkinson’s, and ALS.

Kerry explained to Eric what she had learned: “This type of treatment requires first isolating stem cells, cells not committed to forming one particular cell type but capable of differentiating into many different, functioning cells, from human embryos or young fetuses. There are two different approaches employed, one originally designed by a researcher who works down the hall from me at Johns Hopkins, Dr. John Gearhart, and the other established by Dr. James A. Thomson from the University of Wisconsin School of Medicine. Once the cells are isolated by either method and grown in such a way that they develop into a mature and

normal population of cells, such as the motor neurons malfunctioning in you, these cells could, theoretically, be injected back into you to rescue your damaged neurons and muscles.”

Recent research with mice and rats had shown how promising this curative technique could be and Kerry wanted to find out from Dr. Reiter if any labs were ready to do human trials.

“Kerry, I don’t want to be a guinea pig!” was Eric’s immediate response. “But beyond that, I am very uncomfortable with the idea of having embryonic cells injected into my body.”

Kerry hesitated before she said, “Eric, I am not going to discount what the Catholic Church believes, but other religions have considered this issue very closely as well and some have even argued that embryonic stem cell research must be pursued because it has the potential for saving lives and alleviating pain and suffering. Look, why don’t you let me ask Dr. Reiter about potential cures and the research behind them.”

Eric was relieved. At this point he was like his parents, more concerned with what the immediate future held for his health and lifestyle.

Their meeting with Dr. Reiter was very long, filled with information and some hope. She recommended that Eric join the regional ALS society and attend their meetings and establish contacts. “I know I could spend 24 hours telling you what you should know about ALS and living with the disease, but it is best if you learn on your own time, Eric,” she said. She answered all his questions about the progression of the disease, how to handle insurance issues, what he should be preparing for next, how he should change his job schedule, and what changes he should make in his daily routine.

When Eric felt he’d asked everything he could think of, Kerry asked her questions related to stem cell research. Dr. Reiter was aware of Drs. Gearhart and Thomson as well as others’ latest research using embryonic stem cells to “cure” various neurological diseases. She had even heard that there was talk of a possible human trial with less severe ALS patients. She said she would find out more about it, but in the mean time prescribed Rilutek for Eric to take. As he and his family were leaving, Eric felt more in control than he had in the past five weeks.

The next time Eric met with Dr. Reiter, she told him there was indeed a human trial for testing the effectiveness of injecting human embryonic stem cells into less severe ALS patients. She handed him a phone number of a physician who would set up an initial interview. Although Eric had decided to be proactive and do all he could to slow down the progression of the disease in hopes of a cure, he was uncertain about how he felt about clinical research that used embryonic or fetal tissue. He asked Dr. Reiter for her opinion. She was hesitant to tell him her stance, for fear of swaying him one way or another. Instead, she suggested that he talk to as many people, from as many different fields, as he could.

When Eric left Dr. Reiter’s office he had a list of possible people to contact, some of whom Dr. Reiter personally knew. He immediately began his long search for information, guidance, and hope. He started by talking to a local religious scholar and an ALS specialist. Dr. Reiter had also recommended that if Eric was truly interested in being a part of the stem cell research program, he should contact some of the regional Republican politicians, individuals who were either totally against embryonic stem cell research or very interested in limiting it. She felt Eric needed to understand the political and legal issues that could potentially thwart such a research program if Eric chose to participate. One of the people Eric contacted was Dr. Leon Kass, a philosopher who had publicly presented his position on fetal tissue/embryonic stem cell research and an advisor to the President. Eric also e-mailed and got information from Drs. Gearhart and Thomson, the stem cell research scientists.

Eric was hoping for a simple “yes” or “no” to his ultimate question of whether or not he should participate in the research trial. What he got was a wealth of information, some of which was conflicting, that allowed him to make his own decision.

Your Assignment

Your job is to thoroughly research one of the individuals Eric interviewed/questioned about embryonic stem cell research to find out what this person knows and thinks about this type of research and determine what kind of advice he/she would give Eric. For each person, there are some specific questions you will need to address in addition to his/her opinion of the research. You must understand and articulate why he/she believes what he/she does.

You will present your findings in a 12- to 15-minute presentation to the class. You'll end your presentation by saying what you think this person would advise Eric. Following everyone's brief talks, we will discuss the various viewpoints that were addressed, the merits and problems with embryonic stem cell research, and possibly come to some consensus about what Eric should do.

You will also hand in a two-page, single-spaced paper on your understanding of stem cell research and the opinion of your assigned person. You will need *at least five* references that are properly cited. Some of your references can be from the web, but at least two must be from publications that are edited or peer reviewed in some way, such as those found in the *New York Times* or science journals. It is important to learn how to distinguish accurate and credible information from that which is inaccurate or fraught with opinions based on little evidence. Some peer-reviewed scholarly journal articles are accessible freely through the Internet or the library's electronic databases and these are of course acceptable. Standard journal citation and bibliographic format for the hard copy version should be used in the text of your paper.

Before you begin researching the perspective of your assigned role, you must have some general understanding of embryonic stem cell research. Several references (listed below) will provide you with a comprehensive overview of the field and, in some cases, may also assist you in your research on your assigned role. Also listed is a primer published by the National Institutes of Health (2001), which is useful for techniques and basic embryology. The ALS site provides patient information and frequently asked questions about the disease. For each role, there are some crucial articles or documents that must also be read and understood; these are listed after the description of each role.

General Information References and Internet Links

Website of the Amyotrophic Lateral Sclerosis Association.

<http://www.alsa.org/>

Lanza R, Rosenthal N. 2004. The stem cell challenge. *Scientific American*. 290(6):92–100.

http://www.sciam.com/print_version.cfm?articleID=000DFA43-04B1-10AA-84B183414B7F0000

National Institutes of Health Report. 2001. Stem cells: Scientific progress and future research directions.

<http://stemcells.nih.gov/info/scireport/>

Shaw J. 2004. Stem-cell science: When medicine meets moral philosophy. *Harvard Magazine*. July-August: 36–45.

<http://www.harvardmagazine.com/on-line/070483.html>

Silani V, Cova L, Corbo M, Ciammola A, Polli E. 2004. Stem-cell therapy for amyotrophic lateral sclerosis. *The Lancet*. 364:200–202.

List of Eric's Contacts

For your randomly assigned individual, you must address the several points that are listed here, in addition to clearly identifying his/her opinion on stem cell research.

1. ALS Specialist/Physician

- A complete understanding of the disease.
- The current treatment possibilities, including Rilutek, and their effectiveness.
- This person may or may not support embryonic stem cell research—you need to decide what you think seems more likely and then take that stance.
- Crucial resource:
 - Brooks BR. 1994. El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*. 124 Suppl:96–107.
[http://www.alsa.org/files/cms/Resources/Criteria_for_Diagnosis\(1\).pdf?CFID=1608394&CFTOKEN=70500707](http://www.alsa.org/files/cms/Resources/Criteria_for_Diagnosis(1).pdf?CFID=1608394&CFTOKEN=70500707)

2. Dr. John D. Gearhart, Department of Gynecology and Obstetrics, The Johns Hopkins School of Medicine

- His use of embryonic germ (EG) cells derived from the primordial germ cells, which are isolated from five- to 10-week old aborted fetuses.
- The results of his breakthrough research published in 1998 (Shamblott et al., 1998).
- His latest report (Kerr et al., 2003) on partially curing rats paralyzed in a manner similar to ALS, by injecting them with human embryonic stem cells.
- Crucial resources:
 - Kerr DA, Llado J, Shamblott MJ, Maragakis NJ, Irani DN, Crawford TO, Krishnan C, Dike S, Gearhart JD, Rothstein JD. 2003. Human embryonic germ cell derivatives facilitate motor recovery of rats with diffuse motor neuron injury. *Journal of Neuroscience*. 23(12):5131–5140.
 - Shamblott MJ, Axelman J, Wang S, Bugg EM, Littlefield JW, Donovan PJ, Blumenthal PD, Huggins GR, Gearhart JD. 1998. Derivation of pluripotent stem cells from cultured human primordial germ cells. *Proceedings of the National Academy of Sciences*. 95:13726–13731.
 - Transcript of Dr. John Gearhart speaking to the President's Council on Bioethics in 2002. "Medical Promise of Embryonic Stem Cell Research (Present and Projected)."
<http://bioethics.gov/transcripts/apr02/apr25session1.html>

3. Dr. James A. Thomson, Department of Anatomy, University of Wisconsin School of Medicine and the Wisconsin Regional Primate Research Center

- His use of embryonic stem (ES) cells derived from the blastocyst's inner cell mass.
- The theory behind and results of his breakthrough research published in 1998 (Thomson et al., 1998).
- His work on hematopoietic stem cells; although it does not directly relate to curing a neurodegenerative disease, it illustrates the success he has had in stem cell research.
- Crucial resources:
 - Odorico JS, Kaufman DS, Thomson JA. 2001. Multilineage differentiation from human embryonic stem cell lines. *Stem Cells*. 19:193–204.
 - Thomson JA, Itskovitz-Eldor J, Shapiro SS, Waknitz MA, Swiergiel JJ, Marshall VS, Jones JM. 1998. Embryonic stem cell lines derived from human blastocysts. *Science*. 282:1145–1147.

4. Religious scholar

- The question of does embryonic stem cell research further or hinder the improved health and well being of humans.
- A comparison of major religions and their stance on embryonic stem cell research.
- An overview of Pope John Paul II's opinion of stem cell research.
- Crucial resources:
 - Dobsen P. 2004. Stem cell dilemma. *National Catholic Reporter*. 40(32):13–16.
http://www.natcath.com/NCR_Online/archives2/2004b/061804/061804a.php
 - Peters T, Bennett G. 2001. Theological support of stem cell research. *The Scientist*. 15(17):4.
http://www.the-scientist.com/yr2001/sep/comm1_010903.html
 - Sullivan B. 2004. Religions reveal little consensus on cloning. Online. MSNBC. 2004.
<http://msnbc.msn.com/id/3076930/>

5. Politician who supports President George Bush's stance on stem cell research

- President Bush's August 2001 decision and televised public announcement concerning embryonic stem cell research. What are the present regulations on this science?
- The rationale for Bush's decision.
- What impact has Bush's regulations had on the Gearhart lab getting any government funding for doing research with their EG cells and on the Thomson lab getting funding to create more ES cell lines.
- Make clear what the federal regulations are on embryonic stem cell research that is privately (not federal government) funded.
- Crucial resources:
 - The President's Council on Bioethics. 2004. Monitoring Stem Cell Research. Washington, D.C. January.
<http://bioethics.gov/reports/stemcell/index.html>
 - Transcript of President Bush's August 9, 2001 speech:
<http://www.whitehouse.gov/news/releases/2001/08/20010809-2.html>
 - Wertz DC. 2002. Embryo and stem cell research in the United States: history and politics. *Gene Therapy*. 9:674–678.

6. Dr. Leon Kass, Professor of Social Thought, University of Chicago, and Chair of the President's Council on Bioethics

- His background and education and how he came to be concerned with the "ethical implications of biomedical advances" (report to Committee on Energy and Commerce, Subcommittee on Health, June 20, 2001)
- His argument against the "Brave New World" and his fear that this type of research on embryonic stem cells will lead to designer embryos.
- Address his critics, specifically Dr. Elizabeth Blackburn, who was on the President's Council on Bioethics until her term was not renewed in March 2004.
- Critical resources:
 - Blackburn E, Rowley J. 2004. Reason as our guide. *PloS Biology*. 2(4):420–422.
<http://dx.doi.org/10.1371/journal.pbio.0020116>
 - Kass LR, Wilson JQ. 1998. *The Ethics of Human Cloning*. American Enterprise Institute Press. 122 pages.
 - The President's Council on Bioethics. 2004. Monitoring Stem Cell Research. Washington, D.C. January.
<http://bioethics.gov/reports/stemcell/index.html>
 - The President's Council on Bioethics. 2004. Reproduction and Responsibility: The regulation of new biotechnologies. Washington, D.C. March.
<http://bioethics.gov/reports/reproductionandresponsibility/index.html>

What Everyone Needs to Understand Before We Meet to Discuss the Case Study

1. ALS, the conditions of and the prognosis for the disease.
2. The definition of pluripotent stem cells.
3. The difference between embryonic stem (ES) cells and embryonic germ (EG) cells and the techniques used to produce them.
4. The basic theory and challenges related to how ES or EG cells could be used to cure a disease like ALS.

Grading

1. Presentation—Each person will be graded on his/her contribution to the presentation (20 pts.)
 - Expresses an understanding of the science behind embryonic stem cell research.
 - Understands and clearly states the opinion of person represented.
 - Addresses assigned questions.
 - States what this person would advise Eric; this must reflect the person's viewpoint and beliefs about stem cell research.
 - Clarity and organization of talk.
 - Use of appropriate visual aids.
2. Two-page Paper (10 pts.)
 - Expresses an understanding of the science behind embryonic stem cell research.
 - Understands and clearly states the opinion of person represented.
 - Addresses assigned questions.
 - States what this person would advise Eric; this must reflect the person's viewpoint and beliefs about stem cell research.
 - Writing, clarity, and organization.
3. References (5 pts.)
 - Proper format.
 - Have at least five, two of which are from reviewed, edited sources. Two must *not* be from the ones provided in this handout.

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