Lou Gehrig, Angelina Jolie, and Cancer Genetics

Harold J. Burstein, MD, PhD

In the autumn of 1938, Lou Gehrig was (and remains to this day) the best first baseman in baseball and the most accomplished player on the champion New York Yankees, a team that had won 3 consecutive World Series. For most of his career, Gehrig had batted fourth in the lineup, behind Babe Ruth. By the late 1930s, Gehrig was no longer batting behind Ruth but behind the young Joe DiMaggio. It was a talented group. Gehrig was famous for both his skill and his durability. He set the major league record with 2130 consecutive games, and his nickname, The Iron Horse, was a testimony to his strength and resilience.

But within a year of that autumn, Gehrig had retired from baseball. He had sought medical evaluation for progressive weakness and, after a workup at the Mayo Clinic, was diagnosed with amyotrophic lateral sclerosis (ALS). He went public with his illness. On July 4, 1939, in a ceremony at Yankee Stadium, he announced himself the “luckiest man on the face of the earth.” Less than 2 years later, he died of ALS. So magnificent were his talents as a player and as a person, so prominent was he in the national culture, and so compelling was his battle with ALS, that the disease has been known for generations since as Lou Gehrig’s disease. That eponymous tribute brought global understanding and awareness to what had been a rare, silent, and anonymous illness.

Cancer genetics just had its “Lou Gehrig moment,” and its name is Angelina Jolie. Like Gehrig, she has given a rare medical disorder both a face and a body. On May 14, 2013, Jolie disclosed in a New York Times op-ed contribution that she harbored a hereditary BRCA1 mutation and had undergone preventive surgery. Jolie is a highly visible person in our culture: Academy Award–winning actress, film director, United Nations Goodwill Ambassador for the High Commission on Refugees, familiar paparazzi target, and often named “most beautiful woman in the world.” Still, that kind of public attention and fame paled in comparison to the outpouring of interest that her disclosure generated.

Measuring the extraordinary media attention is difficult. I tried by compiling search results on Google for highly respected, decades-old organizations devoted to breast cancer care and those for Jolie, 1 week after her announcement. Here are the data: NCCN and breast cancer, 250,000 results; Komen and breast cancer, 5,000,000 results; Angelina Jolie and breast cancer, 84,000,000 results and trending up. And Jolie does not even have breast cancer. She has a hereditary predisposition to cancer. Undoubtedly, her disclosure has brought more attention and recognition to hereditary cancer than anyone or anything in history.

Much in Jolie’s essay is valuable, and the whole is worth reading and reflecting on. Start with her title, “My Medical Choice.” It emphasizes that the decisions are hers—not her friends, family, or medical team’s—and that it is a choice. Many people with hereditary cancer syndromes will have choices to make—prophylaxis, prevention, surveillance. A person’s preferences and choices can change over time. Making choices requires information and reflection. She framed all those ideas in her editorial; she sought to help women “make your own informed choices.”

She also got her facts right. Her condition is rare. Her personal risks were high. She received sophisticated medical testing and treatment. She experienced pain and discomfort and got through it.

Jolie’s word for her surgery—preventive—is also a good choice. The medical literature tends to term such surgery prophylactic or risk-reducing. Those words sound...
One is a better word; one that resonates with the concerns of a woman contemplating such surgery. Indeed, the whole tone of her essay was a model of good medical communication: start with the data to frame the situation, outline the clinical experience, give space for various options, capture the emotional angles. Jolie wrote that after her surgery, she did “not feel any less of a woman,” and felt empowered by her choice. Thousands of women will similarly feel empowered and reassured by her remarks.

For what it’s worth, Jolie also received guideline-based care. As the daughter of a woman who had breast and ovarian cancer, Jolie met criteria for genetics evaluation and BRCA1/2 testing, as articulated by the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Genetic/Familial High-Risk Assessment: Breast and Ovarian Cancer. In choosing bilateral preventive mastectomy with breast reconstruction, Jolie followed a set of recommendations outlined in the NCCN Guidelines for Breast Cancer Risk Reduction.

Jolie correctly wrote that BRCA1 and BRCA2 testing can be expensive and that the expense can be an obstacle to testing for many women around the world. However, given her apparent clinical history, the testing, preventive surgery, and subsequent breast reconstruction would probably be covered by most major health care insurers and policies. Most people in the United States do not have access to the jet-set lifestyle, Hollywood spouse, and vast entourage that surround a movie star. But nearly all women with the same clinical story as Angelina Jolie have access to the same kind of counseling, testing, and treatment she received.

It is hard to predict the effect Jolie’s disclosure will have on the choices of people with hereditary cancer and their families. In the near term, I suspect there will be a surge in interest in genetic testing and in surgical prevention for affected individuals, with a shift away from intensive surveillance and chemoprevention for known mutation carriers.

Regardless, Jolie has done a fantastic service to the families of people affected by hereditary breast and ovarian cancer. She has given public voice to their fears and choices. She has shown courage in dealing with the medical information. She has mapped out a route of management that worked for her and might work for others. She has helped to destigmatize mastectomy and reconstruction, and she has assured women that they will still be beautiful afterward. She has started a vast conversation on the biology of cancer genetics and the medical issues related to caring for people with hereditary mutations that cause cancer. She has allowed affected individuals to speak candidly of their own experience.

ALS became “Lou Gehrig’s disease.” With a happier ending, hereditary breast and ovarian cancer is, from now on, “Jolie’s syndrome.”

Reference