

How to Cope With and Survive a Terrible, Incurable, Fatal Disease

I am writing to tell my story and help others. Everybody is different, but a terrible, incurable, fatal disease equalizes us all. There is wisdom and understanding that comes through adversity and suffering. It is much easier to read about than experience, but knowledge can be passed on and I can provide some counsel and comfort for those in similar situations. Strength and hope are crucial and I hope my story encourages others to fight the good fight and, hopefully, win.

You never really fully appreciate anything until you think it might be gone. This is particularly true of your own life. There are a lot of really terrible diseases out there, and mine was a very bad one that came out of the blue. My terrible surprise came when I was 58, vigorous and in good health, working out almost every day. I had a wife and two sons who were 11 and 18 years old and a career as an international lawyer, mostly representing sovereign wealth funds. I noticed some shortness of breath and difficulty in exercising at peak levels, so I went to be examined by my general practitioner and passed my annual physical. However, I knew something was different and awry in my body and was not satisfied, so I obtained a referral to a cardiologist. The EKG was abnormal and she conducted an echocardiogram. My film revealed enlarged lower heart ventricles. Amazingly, this led the doctor to immediately suspect that I might have Amyloidosis. She told me that the good news was that I caught it early, it was treatable and great medical advances had been made. She saved my life.

I did not know what Amyloidosis was and, of course, like any modern patient, googled it on the internet. Amyloidosis is an extremely rare (1 in 100,000), incurable, deadly disease in which Lamda light chains in the blood, are overproduced and misfold. This means that the light chains infiltrate the organs and other soft tissue of the body, interfering with normal functioning until something critical fails. It can affect the heart, kidneys, liver, lungs, skin, nerves, and digestive system. Therefore, it has a wide variety of symptoms from a swollen tongue to swollen ankles, to carpal tunnel syndrome. Because it is so rare, it is hardly ever picked up immediately. Most people take months or years to be properly diagnosed and by then it is too late to stop or undo the damage. I was in shock: according to what I read my new anticipated life span was six months untreated, one year treated.

So what did I do? My emotions sent me reeling, from shock to disbelief, to fear, to anger, to denial. However, I am a lawyer and trained to be a logical problem solver, so I focused on how to best move forward with this new reality. I started reading online and became familiar with the literature on the disease, including medical journals and materials from various treatment centers. This was actually calming and helpfully distracting. First, I needed to figure out who was the best doctor for this rare condition, so I marshalled my resources: called my local doctors, friends and colleagues who might have leads and sought information. After speaking to several medical experts, the recommendation that came back was uniform: Rodney Falk, a cardiologist and amyloidosis specialist in Boston. I pushed for an appointment immediately and flew to Boston with my wife and in-laws. I met with the doctor and went through a heart and bone marrow biopsy which confirmed the diagnosis: AL Amyloidosis. An oncologist who was familiar with the disease and who primarily focused on multiple myeloma assisted in developing a treatment plan.

The first step was treatment with chemo drugs developed and tested for multiple myeloma and also used for amyloidosis. They might reduce the light chains and might stop the progress of the disease. If not, and maybe in any event, there was a stem cell transplant. This is a formidable procedure where the stem cells are extracted from the blood, a high dose of mephalin (a mustard gas derivative) is administered to the patient, killing the bone marrow/immune system, and then the stem cells are reinserted. It has a 30-40% success rate and almost equally high death rates for eligible patients with Amyloidosis and cardiac involvement. Although the results from a stem cell transplant tended to last longer, due to the risks there was a schism among doctors as to whether stem cell transplants were always the preferred treatment. Not an easy choice to make and I hoped that the chemo would work and at least buy me time.

I asked what my overall odds of survival were. I was told 50/50 on chemo, 50/50 on the stem cell and 50/50 that they could figure out something else. That was somewhat reassuring, but, unfortunately, none of the treatments was permanent with stem cell lasting the longest.

The chemo treatments did not work. I spent the next three months going from treatments of Bortezomib to Velcade to Revlimid. Weekly infusions were coupled with pill regimens. These are serious drugs with very noticeable side effects. I started to feel fatigue, mood swings from the steroids that were part of the cocktail, insomnia, nausea and itching. Each week I also felt worse from the disease and/or the medicine with increased difficulty breathing, extreme fatigue and lots of anxiety. Unfortunately, the light chains in my blood did not go down materially and, each month, when the blood tests came back, I was more demoralized. My local doctor who administered the treatments did not help. At our first meeting, he told me that he could cure me. Later, as the treatments failed, I was told not to worry as it was about him and his ego, so he would therefore find a solution. He showed me trials that demonstrated that the medicine worked on most people and, because they did not work on me, proved I was unlucky. Very upsetting and not helpful.

During this time period, I also worked on my estate plan. I was very happy to being able to do something productive to help my family under the circumstances.

When it became apparent that I was getting worse and the chemo was not working, I scheduled a stem cell transplant. I chose the University of Pennsylvania because of a combination of factors: it was highly recommended; it was a regional amyloidosis center; I knew a recent transplant patient that had great success there; and it was close enough for my family to visit over the three weeks to a month that I would be hospitalized. I went to visit and the oncologist and cardiologist determined that I was eligible. A date was set and I steeled myself for the unpleasantness of the treatment and very slow recovery from a weakened state. The loss of my hair and perhaps nails, bothered me, but seemed trivial given the overall risks.

I received by mail the drug (Neupogen) which I was to inject to induce the stem cells to enter the blood stream for harvesting. Following the instructions, I had my son inject me daily. By the second day, I was not feeling very well at all, but trying to fight through it. I went to the gym and could do nothing. That evening after I received the injection, I started passing out. I woke up to have first lost control of my bladder and second my bowels. I passed out again for a third time in

the shower trying to clean up and banged my head which started bleeding. My family called an ambulance and I was rushed to the Emergency Room. Despite my wife's protests, I was left unattended in a treatment room while they waited for blood tests and kept passing out. She finally exploded, threatened legal action and I was then transferred to the ICU.

Later that night in the ICU, I had a cardiac event where I could not breathe and felt like I was drowning. I felt like a fish on land at the edge of a pond. Doctors and nurses filled the room fanning my face and watching my blood pressure drop on the monitors while they waited for the fluids/medicine to kick in. They got out electric paddles and prepared to inject me with a powerful stimulant to restart my heart if it stopped. Fortunately, my blood pressure gradually rose and I recovered. During that incident, my heart released troponins similarly to what occurs with a heart attack.

I spent the next few days in the ICU being watched and tested. The constant prodding, light and noise was very unpleasant and tiring. They could not determine exactly what caused the episode so they decided to insert a pacemaker/defibrillator to protect me from another occurrence. When I left the hospital I was much weaker and demoralized: the doctor at Penn told me that it was now too risky to try the stem cell transplant. Too risky for whom was my question. If I was going to die anyway what did I have to lose? Were the doctor's concerned with their success rates or me?

This was my low point. I focused on finishing my estate planning and recovering from the surgery. However, the doctors had a new suggestion: Darzalex. This was a brand new, monoclonal antibody, that targeted a specific molecule and got the immune system to also attack it. It had been tested and fast tracked on multiple myeloma and no one knew how it would work with Amyloidosis. The weekly infusions took seven hours and about half the patients had immediate allergic reactions that would stop the treatment. I did fine and the drug worked. Within four weeks my number went from 340 to 52! Since then it has gradually gone down to 25 (16 to 27 is normal). If this was cancer I am now in remission. The treatments are now monthly and I do not know how long the effects will last.

So what did I learn from all this? A lot, much of which we intuitively know, but do not focus on in our day to day life. It takes a hard slap to bring us truly into consciousness. Life and time are precious. It is easy to drift through life without paying attention. You need to be your own counselor/advocate. There is power in positive thought and belief. Family and friends are more important than material things. Please permit me to share a few observations and recommendations.

- 1) You can be who you want to be. When you are very sick you fear death, but are also set free. All the trivial day to day things you spend your time worrying about are not all that important. They cannot be ignored, but should not be your primary focus. You have choices about what you care about and devote yourself too. Inevitably, upon consideration and realization, family, friends and helping others become much more important than material things. Do not be afraid to reveal your condition it has become a part of you.

- 2) Do not rely on others to protect yourself. Doctors are human and fallible. No one cares about you or will pay as much attention to you as you will. Do not be afraid to speak up. If you ask enough questions you will surprisingly often ferret out and prevent errors. For example, several times I noted discrepancies between what the doctors suggested for treatment plans and what was recommend by the drug manufacturer or done for other patients. In some cases, there was no special rationale and we ended up following the label. Read, ask questions, prod medical professionals for answers, get multiple opinions and then be your own judge. I read everything I could on my disease and it empowered me and kept the medical practitioners on their toes.
- 3) Keep up hope. If you can believe in something, no matter how improbable, it can change outcomes. I am a logical person, but have come to understand that not everything can be explained. Hope, faith and belief lead to positive results and the absence of those feelings leads to resignation and death. I am jealous of those who have long term religious beliefs that are sustaining, but it is never too late to find faith in God.
- 4) Use your skills to protect yourself. Whatever it is that you are best at, focus on utilizing that talent to fight your disease. If you are well organized get your records straight and make sure that all the doctors have them. If you are creative think outside the box and seek different approaches to treatment that might help. Focus on what you can do to help yourself and it will both distract you in a positive way and make the battle much more winnable.
- 5) Never assume that there is nothing that can be done. I focused on my own treatment plan and read the stories of others. Medical journals often provide critical insights on new discoveries that are coming out all the time. There are often great comeback victories for those who persevere and do not give up. Do not assume anything and stay focused on what you can accomplish. After multiple medical treatment failures and a rapidly worsening physical condition, it was very hard to stay optimistic and stay the course. Darzalex was a life saver for me and it had just been approved for multiple myeloma last year. The first clinical trials on AL Amyloidosis are going on now. Look for options and do not take no for an answer without first trying to specifically understand why.
- 6) Live in the moment. I took up meditation and it taught me how oblivious I had become to myself and my feelings. You need to stop and listen to yourself. Many of your emotions are reactions to things that you are not consciously focusing on. When you listen you realize better how to be positive and control your own feelings and actions.
- 7) Empathize with others. Everyone has difficulties and let your disease open your hearts to others. A dog eat dog world is fine when you are winning, but when you have had bad luck and adversity it should make you better understand the troubles of others. Helping others makes you feel better about yourself.
- 8) There are many good people out there. It is too bad that we have to discover true friendship and loyalty in times of trouble, but when it comes that support is invaluable and truly uplifting. The love and kindness of my family, friends and colleagues was incredibly powerful and reassuring. My family was at my side at so many doctor's appointments and my father in laws, a cardiologist became my liaison and advocate with other doctors. My friends called on contacts and relationships to help me marshal the best medical team and were always there when I needed them. Half my high school class set up weekly prayers sessions to try to help me and another classmate who had a mortal

disease. My law firm and partners stood behind and supported me without question or hesitation. Love and group support show how much good there is in the world.

No matter what my disease will bring, I will always be grateful for what I have learned and for the love and kindness of others which I might not have otherwise experienced. Go forward with a positive attitude and turn adversity into newfound strength and appreciation for the world around you.