

My Miracle

Diffuse systemic scleroderma survivor Sherlyn Delaney Roberts shares her story

For as long as I can recall, I've always been cold, being repeatedly told that if I'd put some meat on these bones, I wouldn't be so cold. I could deal with that. But I had a harder time dealing with what came next.

How it all started

In June 2003 my hands and feet began to swell. I figured the swelling was a result of the birth control pills I had been taking. But then my elbows and inner thighs became tender. Swallowing was beginning to be a problem—consuming things was a problem. Even liquids would get stuck.

One day in September, I woke up with sore, stiff hands. The day before, I had just played the organ at church services with no difficulties.

Fortunately, my physician, Dr. Susan Dasta, agreed to see me that day. She immediately ordered blood tests. I thought I might have had rheumatoid arthritis, but since there is no family history of the disease, I wasn't sure. Dr. Dasta referred me to a rheumatologist. Within a few days, shooting, stabbing pains began running up and down my forearms, awakening me in the night. It felt like pins and needles and my wrists and elbows began hurting constantly.

My new husband, Kirk, and I moved into our new home on October 11, and I felt frustrated because my hands and arms were preventing me from holding or lifting much of anything. I was also very fatigued.

Two days later I saw Dr. Nancy Becker, a rheumatologist, and was diagnosed with scleroderma and Raynaud's. Dr. Becker told me there was no cure for scleroderma and sent me home with some literature to read. The following day hospital staff collected 18 vials of my blood. I passed out from their digging around in my poor veins and had to return the following day to complete the blood work. I either didn't hear or didn't realize I had the most progressive form of scleroderma, or perhaps at that stage, we

simply didn't know. Things really didn't seem that bad yet; I'd just have to readjust my thinking and realize there would be things I couldn't do the same way I had before.

That positive attitude didn't work for long. My muscle strength—in particular, my upper body strength—was deteriorating at an incredible pace, and by December, things were awful. Because all my extremities were weak, swollen and extremely hard,

even getting out of bed became an incredible chore. My elbows hurt if I touched them on anything and I was having great difficulty swallowing—thus even eating became a challenge. I developed stomach ulcers, spitting up blood regularly. My back was on fire, along with my legs, mostly my inner thighs.

The simple things we take for granted, (holding and writing with a pen/pencil, brushing your teeth, washing your hair, opening a can of soda pop, holding a plate, holding a phone to your ear, buttons, getting dressed, panty hose, socks, tying shoes, reaching for anything, opening doors, turning the steering wheel, holding a cup) all became challenges and eventually

impossible. While trying to hold a cup of coffee in November, I dropped it and burned my leg. The pain that was taking over my body was immense and it happened so incredibly fast. At first there were few visible signs of my disease, which apparently made it difficult for some people to accept. This truly added to the pain.

Education was key

I began educating myself about this dreadful disease via the Internet. I refused to believe there was nothing that could be done. I knew with God's help I could surely find something. I first contacted Methodist Hospital in Houston for treatment, but our insurance denied coverage. I then contacted Carol Blair who works with noted scleroderma physician



Sherlyn at the Scleroderma Foundation's 2006 National Gala



Sherlyn with Sue and Larry Smith, Tarrant County Support Group leaders (Larry is also the VP of the Texas Chapter Board)

Thomas Medsger, M.D., and inquired about a clinical trial involving stem cell transplants. Carol was so kind and understanding. She was also very optimistic about the treatment.

I had read everything I could find on the Internet about treatments and learned that although stem cell transplants were being done on a limited basis in the United States, numerous European countries had been using variations of this treatment successfully for many years. I spoke to Dr. Becker about transplants and she told me I might die. However, she agreed to read the printed material from the Internet and then suggested I might want to make an appointment with Dr. Medsger. I really appreciated her encouragement at that point and told her I had already made an appointment.

By the time I saw Dr. Medsger the week before Christmas, I wasn't faring well at all. I was very ill and in great pain, and had great difficulty walking through the airport. My determination to keep walking gave out when I collapsed in the hospital just prior to my appointment. Dr. Medsger honestly explained my circumstances. He told me my kidneys were in crisis and that I had very high blood pressure. A new rush of pain hit me, and he prescribed some narcotics to make me comfortable.

This kind, humble man recommended I undergo a stem cell transplant because of my diffuse systemic scleroderma. He was right on target, as usual. I knew in my heart that this was my answer from God.

On to Pittsburgh

The doctors determined I needed a transplant soon,

and they identified January 5, 2004 as the goal date. That didn't happen so another date was set, with January 21 the date for pre-testing. The day before that was supposed to occur, I received a call from a coordinator from Pittsburgh, advising me not to come. She offered no explanation why. I was dumbfounded. Having already pre-paid our airline tickets and hotel, not going was not an option. I quickly pulled myself together and wrote numerous electronic letters to Congress, and even President Bush—well over 50 copies were sent. We decided to leave for Pittsburgh anyway, and the next day I received a call from Dr. Andrew Yeager, an oncologist of the transplant team, who apologized for the mishap. He asked if I could be in Pittsburgh the following day for pre-testing. I said "Praise God," and cried and said "I'm already here in Pittsburgh!" To this day, I have no idea what happened, but it's irrelevant now. Obviously, I did qualify for the procedure, and met Dr. Yeager and his colleague, Dr. Diane Buchbarker, and after more confusion and altered dates, the procedure was scheduled for February.

On February 17, I was admitted to Shadyside Hospital and prepped for surgery so I could have a Hickman catheter placed. My husband asked me if I was nervous. I said, "Are you kidding?" I felt like one blessed lady to have the hope to possibly beat this

horrific disease that had so rapidly taken over my body. I was excited if you can believe that!

I had 24-hour mobilization chemotherapy (high dose chemotherapy) later that day after surgery and I could barely wait to get started.

By the end of mobilization chemotherapy, though quite sick from the chemotherapy, my legs were nearly pain free. A miracle was happening and I knew it.

Two days after the procedure I was dismissed from the hospital and began to receive daily Nupagen injections. How it works is the chemotherapy drug (cytoxan) stimulates the bone marrow to produce large numbers of new stem cells and releases them into the blood. These stem cells replace the blood stream cells that will be damaged by the chemotherapy given at a later time to eliminate the disease-causing cells in your blood. Nupagen shots also increase the growth of white blood cells and stem cells. These injections make your bones ache.

By the following week, my white blood cell count reached the desired number needed for the stem cell collection procedure and I was ready for leukapheresis (a cell separating and collection procedure). These stem cells would be frozen, stored and introduced into

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my body after additional chemotherapy. After my cells were collected, a machine separated different types of cells from the blood, specifically removing the T-cells that started the process that causes systemic sclerosis.

My parents drove to see me from Kansas. Though still very sick from the chemotherapy when they arrived, I was so glad to have their loving, caregiving presence.

The most amazing thing was happening to my body. Every day, through March 5, I was getting better. Yes, I was sick from the chemotherapy, and my long blond hair was falling out (and would eventually all fall out), but I was definitely improving physically and fast. I felt good enough that I decided to walk a mile with my husband. I really walked, with zero pain in my legs.

A turn for the worse

But it would turn out to be a day I would not easily forget. My right arm began to hurt and swell. I was having difficulty concentrating while watching a movie, and my arm was getting worse rapidly. We called Dr. Buchbarker and she suggested we get to the emergency room immediately.

The next round of chemotherapy was supposed to start March 9, but I had developed two life-threatening blood clots as my catheter wasn't working properly. My physicians were debating the removal of my Hickman and putting in another one, but wanted to give the drug Heparin an opportunity to work.

The next morning I had a venogram and consented to TPA, a blood thinner (ultimately the TPA was not administered). Since tests clearly confirmed two blood clots prior to the venogram, it seemed unnecessary to have gone through the procedure when ultimately no TPA was given. I felt relieved and trusted the right decision was

being made. Chemotherapy resumed later that night and Dr. Yeager came in and indicated I was "on" to begin therapy. I always had complete confidence in Dr. Yeager, Dr. Michael Carroll, another oncologist, Dr. Buchbarker, and Dr. Medsger, my transplant team of physicians.

I was again getting Cytoxan infusions now along with Fludarabine, another drug effective in killing lymphocytes. This would continue each night for five days. It was quite a week, as I had episodes of vomiting, diarrhea and an upset stomach—in general, much discomfort. My back pain was horrible too during this week and getting from my bed to the recliner a few inches from my bed was literally impossible without help. I was extremely weak. However, these were all expected, tolerable side effects.

The next day I began another drug, one specific T-lysosome destroyer called rabbit ATG. Everyone reacts to some degree, and mercy did I react. I was one ill lady, but it passed and I received another 8-hour dose the next day. About 4:30 it began: more vomiting, more upset stomach, more diarrhea. It just wouldn't stop, but it was still bearable. I knew it would eventually end—it just would. My folks left my room about 11 p.m. after I reassured them that all was well. A half hour later, things turned for the worse. It was the most difficult night of my life.

My blood pressure was hovering around 40/20 and my temperature was 104.5 and my two amazing, loving caregivers, Bev, my R.N., and Karen, my aide, were at my side constantly. I had diarrhea every few minutes. This all lasted until approximately 4:30 am. During this time, for the first time, I prayed to God to take me home. I was ready. The pain was more than I thought I could bear. I saw no tunnel, no white robe, but my Grandmother Foos appeared as I was praying and clearly told me to



Kirstin Wallin: Another Stem Cell Transplant Survivor

Kirstin Wallin celebrates two birthdays: Her real one, April 27, 1974 and the date she felt her life was saved: March 30, 1998. The latter date was when she became the third person in the United States to undergo a stem cell transplantation for scleroderma. She was treated under the care of Daniel Furst, M.D., a member of the Scleroderma Foundation Medical Advisory Board.

When contacted by the *Scleroderma VOICE* last October, Kirstin reported she is doing "very well." "I have some acid reflux, and some symptoms of Raynaud's, but it is not nearly as bad as it once was," she said.

After her surgery, Kirstin became somewhat of a *cause celebre*, appearing on the NBC show *Dateline* to talk about her surgery.

She reports no joint or muscle pain, though she says she has osteoporosis she attributes to taking a medication to help her with scleroderma-related joint pain.

These days, Kirstin is working a full-time job as an office manager at a small water testing company, and is a Mary Kay cosmetics consultant. She is pursuing her certificate in paralegal studies. She already has a bachelor's degree in general studies.

Kirstin has a boyfriend, and a dog named Moxie, a 15-pound bundle of energy that is a schnauzer/terrier mix.



Thomas Medsger Jr., M.D., oversaw Sherlyn Delaney Roberts' care during her stem cell transplantation procedure.

keep fighting, that it was not my time and more specifically my younger son Mark still really needed me. I listened and after arguing a bit, continued the battle. During this time, many fluids to rid my system of these toxic drugs were being given.

The following morning, Dr. Carroll, my daily hospital oncologist, and his wonderful team came in to ask the magical question. After last night and the reaction you experienced, we will leave it up to you as to whether you'd like the final dose of ATG. The answer was a definite "no!" I finished up the Cytoxan/fludarabine that night and sighed relief early on Saturday morning when it finally ended and it was all over...or so I thought. I can still envision Dr. Buchbarker standing in my room telling me that the worst was surely over for me. I jokingly asked her when that was to start. She looked at the clock and said, right now with a smile on her face.

The following Monday I received all of those precious stem cells earlier frozen and collected for this special transplant day. Cheryl, my attentive P.A., actually administered the cells along with Dr. Carroll, Harry the R.N., Jeanne, and several others,

including my folks and my husband. They were administered through a syringe via my catheter. There were no adverse reactions as expected. I was home free, or so I thought.

The next night I began having difficulty breathing. About 11:30, I shared this with my husband and about two hours later, I was not improving. I buzzed the nurse and she measured my oxygen at 86 percent (normal is 98–100). I was rapidly filling with fluid. Lasix, a fluid remover, was started after a chest X-ray, but this just didn't seem to correct the problem.

My weight was up to 128 pounds and my normal pre-hospitalization weight was 105. One was expected to lose approximately 10 pounds during the hospital process, so in reality, I probably had much more than 28 pounds of excess fluid. The week continued and the fluid problem persisted. On Friday morning, Dr. Medsger visited me following a checkup from my regular attentive transplant team. It was obvious he was concerned, though he tried to be discrete.

Dr. Carroll paid me yet another visit as did Cheryl. I was told after another chest X-ray that a cardiac vascular surgeon named Neil

up a chair very close to me, and clearly and honestly spoke of my serious dilemma. He said that I needed a pericardial window immediately that night, or my heart would most likely collapse from all the fluid. I said, "I think you must have the wrong room and wrong patient" as I was so much better off than the week just prior. Well, I guess I was wrong. Other than having a few breathing problems, I thought I was better.

I persuaded Dr. Christy to wait until Saturday morning and he was not thrilled. The surgery was scheduled. Dr. Christy insisted, however, that I be transferred to the cardiac ICU to be monitored in the event things turned south before morning. I reluctantly agreed. You see, I was having surgery during a time that my immunity was basically absent. The surgery was successful. I wasn't the least bit fearful, but I imagine my family was.

I'm better

Eventually the four chest tubes were removed and Friday I was discharged from Shadyside Hospital. I was still so very sick, but I was one ecstatic girl. I had made it, beating the medical odds

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Christy would be paying me a visit. Hmm, I thought. That really doesn't sound great. It was Friday evening, about 6:30, and my loving, devoted mother was with me. My husband and father were driving my youngest son Mark to the airport. Dr. Christy came in, pulled

more than once, and I knew it! Physically I wasn't ready to leave, but wild horses couldn't have kept me in the hospital one additional minute. It had worked, I had survived with medical odds stacked against me and I was raring and ready to get out of there and begin

my life!

Admittedly the first few days were tough as I was so weak and frail physically. More good news was Nancy Munn, transplanted one week before me with no complications, was doing well and was so supportive of me while I was in the hospital. She would leave her hospital room to offer me encouragement and good cheer. Cathy Fulford, transplanted eight weeks before me, would also visit me. Rosie and Paul Witt became very good friends of ours during this time. Their son Tom was fighting a battle with leukemia. They would all come to visit and encourage me. We also became good friends with Janet Gray. Unfortunately, we lost Cathy in November of 2004 to a heart attack and Janet died shortly after being transplanted due to complications. Every time we all get together in Pittsburgh for appointments, we remember Janet and Cathy. We also remember Tom Witt who died in October 2004.

We flew back home to Kansas City on April 6, nearly seven months after I was first diagnosed. What joy I experienced in my heart. Somehow, with God's help, I sang and directed the church choir on Easter Sunday.

Friends forever

I was still plenty sick while recovering from chemotherapy, the various side effects, and heart surgery, but each day I would grow stronger. We returned to Pittsburgh every two weeks for the first three months. Darrell Lis graciously correlated all of my appointments with Nancy Munn, Joe Dill and Cathy Fulford, whenever possible. We had all become a family. Our reunions were and remain uplifting and encouraging. We have this bond that is just unbelievable. We remain in constant contact.

Two and a half years later, I am eternally grateful that I still have no active disease according to Dr. Medsger. I still have pain in my shoulders, wrists and hands, but the improvement is huge, and although I have some permanent damage, it's pretty overwhelming to have survived this deadly disease and be relatively healthy.



To contact Sherlyn, please contact her at blondyink@aol.com.

Rheumatoid arthritis, Lupus, Scleroderma or Myositis?



Doctors at the National Institutes of Health (NIH) are conducting pioneering research in understanding the genetic and environmental risk factors that may result in autoimmune diseases. The goal of study 03-E-0099 is to assess why one sibling or twin in a family developed an autoimmune disease and why the other brother or sister did not. The study consists of a blood draw, urine collection and completing surveys. There is no charge for evaluations and medical tests at the NIH. Compensation is provided for both participants and referring physician.



You may qualify if:

You have scleroderma or myositis, rheumatoid arthritis/juvenile rheumatoid arthritis, lupus. You were diagnosed within the last 4 years. You have a twin or sibling of the same gender within 4 years of age with out an autoimmune disease.

Both children and adults are eligible.

Duration of Study:

Five years with an annual questionnaire.

Location of the Study:

You may be enrolled in your local doctor's office or at the NIH Clinical Center in Bethesda, MD.



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Nancy Munn, Sherlyn and Joe Dill have become friends because of a common bond: They all had stem cell transplant procedures for scleroderma.