Our Story

Pain Persisted. . . So Did I

By Delaine Elle

I was born in Westlock, Alberta, Canada more than 50 years ago, and started having nosebleeds at eight months old. No one could tell my parents why. My teenage years were the worst, with many nasal cauterizations and packings.

I was not the only one suffering from frequent nosebleeds. My dad, a farmer, and I spent many summer evenings on the back step trying to get our nosebleeds to stop. I still recall some of the comments people made about my nosebleeds: "Your blood is not clotting properly." "Do not run or play in the sun. Sit in the shade." "Your blood is too thin." And my favorite: "You are just growing too fast!"

Now, I get a nosebleed laughing at how silly these solutions were for those nosebleeds my father, grandmother and I suffered with.

In May 1971, I married a banker, Clarence Elle. We transferred around Alberta, so I had opportunities to consult with many doctors and specialists. No one recognized the telangiectases in my nose, other than to say the blood vessels were close to the surface and bleeding around or beyond the previous cauterizations.

Then, in February 1988, when our son, Stuart, was 14-years-old, and our daughter, Deloyce, was 12, I got up from a kitchen chair and suddenly felt a stabbing, throbbing pain in my left groin and hip. At first, it was intermittent, but it became worse and worse. My stiff hip made it very difficult to walk, sit, climb stairs or bend over to access lower cupboards.

It was years later when I realized the pain was the result of internal bleeding.

Pain persisted, so did I.
My challenging journey of misdiagnosis continued. A bone biopsy on my femur for Paget's Disease went terribly wrong, resulting in hemorrhage and blood transfusions. Again, my leg was stiff and ached from waist to toes. I had to use crutches, and later I learned the specialist involved did not expect me to walk again, unassisted. The biopsy result was benign hemangioma on the bone. What was I to do now? Nothing? Wait? I knew I could not tolerate this constant pain forever. My main concern was for Clarence and our two teenagers. I wanted to attend their graduations.

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Six years later, I was still struggling. I waited months for an MRI, then several weeks for an angiogram. Finally, the physicians identified the existence of an extensive eight-inch AVM in my hip and groin "wrapped around the head of the femur."

Knowing that an AVM could burst and hemorrhage, I inquired about the possibility of having my leg amputated. My cardiovascular thoracic surgeon said, "Definitely no!" He explained that his job was to find me treatment and keep me walking.

He consulted with Mayo Clinic, and I was referred to Doctors terBrugge and Montanera of Toronto Western Hospital AVM Clinic. There, cyanoacrylate, liquid adhesive glue, was injected into some of these AVMs. The staff referred to this as "space age crazy glue." I was told it had been used on the Canada Arm - a robotic appendage used outside the hull of the U.S. Space Shuttle.

This robotic appendage and some HHT patients have something in common: space age crazy glue.

During a consultation, the neuro-interventional radiologists surrounding me asked about my array of symptoms, including birthmarks. "You mean hemangiomas?" I asked. The brain specialist smiled and said, "She speaks our language!" I vowed right then to do two things: Learn to use the doctors' terminology in order to be taken more seriously and to better understand their discussions about my medical condition; and listen carefully and ask questions.

When I asked what the next steps were, they replied that after seven years of tolerating pain, I had a pretty good idea of what I could do and not do, but confirmed what I had already heard - they could not promise pain relief. Without thinking, I emphatically replied, "Well, I am not going home to sit in my rocking chair to wait for the lights to go out!" Even I was surprised, and joined the doctors in hearty laughter.

Pain persisted, so did I.
In November 1996, I told my GP we were moving to Edmonton. He called me "the patient, persistent patient" and offered me my medical file, telling me I would need it. He was right. My medical file came to be a very important lifeline. When I would get frowns and questioning looks from new doctors, I'd produce copies of my medical reports. I have many times been amazed at how conversations suddenly changed for the better.

Some physicians were more receptive than others. One doctor told me he had a very busy practice and did not have time to learn about AVMs and HHT. Several others told me I was "unique" and "an odd duck," so I did not fit into their specialty. I was not about to accept defeat. I handed each of them an HHT booklet. I still pray the information will prove useful someday for someone.

In January 2002, when I was experiencing increased hip pain, I realized I needed to return to Toronto for AVM follow-up and treatment. I spent more than two frustrating years trying to get a referral to meet Alberta Health Care requirements for out-of-province medical care. To travel from western Canada for treatment, 202,000 air miles east, I was required to convince a local specialist that only in Toronto could the expertise I needed be found. Finally, I contacted Doctors Faughnan, Simons and terBrugge directly at the Toronto HHT Center and booked my appointments.

Because of my extensive hip AVM, I was sent to Toronto General Hospital where my cardiac output was measured for a special research project. Instead of my heart pumping the expected 4 to 4.5 liters of blood per minute around my body, my heart was working hard to circulate 10 to 12 liters per minute. Since my late teens, I had complained of left-

side chest pains and consulted with several cardiologists. None of them ever identified that it was blood rushing through the "direct connection" between the artery and vein in my hip that was overloading my heart.

During two subsequent trips, embolizations blocked the blood flow to more AVMs in my hip. My chest and hip pains have been reduced significantly, to minimal pain most of the time. What an enormous relief it is to at last get a break from constant pain.

Small pulmonary AVMs, several hemangiomas on my spine and a few telangiectasias on my esophagus and in my stomach are not presenting any problems at this time. Again, I am reminded of the importance in screening everyone in HHT families for manifestations of the disorder; AVMs and telangiectasias can be found in many organs throughout the body.

Continued Persistence.
Throughout these years of seeking answers, I have been determined to educate more physicians and the public about HHT, and to help solve the mysteries of HHT in any way I can. I urge you all to do the same!

I contacted most of my 122 family members to complete our pedigree (medical family tree), which was sent to Dr. Mary Porteous at the International HHT Mutation Database in Edinburgh, Scotland.

Clarence and I attended the 12th International HHT Conference in Arlington, VA. It was so good to realize we are not alone! We visited and learned from each other - the patients, families, scientists, researchers and all the medical staff who share and dedicate their time to help us. As you know, I had earlier vowed to "speak the doctors' language" and to "listen and ask questions." The conference was an ideal place to do both. We learned so much, and highly recommend the bi-yearly Family and Patient Conference.

Delaine's grandparents with their 12 children at their 50th Anniversary.

In fact, following the Arlington Conference, I was inspired to act on several fronts:

- I approached several local specialists who shared my interest in contacting other families in the Edmonton area to establish an HHT Support Group.

- I mailed the "Learn More About HHT" information booklet to all my former doctors and dentists, as well as to all 39 Ear Nose and Throat specialists in Alberta.

- I contacted the local dental schools and associations, encouraged the idea of a local foundation and contacted newspaper and TV Health programs about featuring stories on HHT.

- Our Square Dance Club has agreed to a special Fun Dance Night with all proceeds going to HHT research.

It has been a long 18 year journey, through much pain and discouragement. Still, I am glad I persisted, and I urge you to do the same on behalf of yourself, your children and all of those with HHT.

Many Thanks to my parents and my family for their patience and care during difficult times, and to those doctors who persist in finding answers and treatment. I have much to be grateful for every day.